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ORAL PRESENTATIONS
Serial Transverse Enteroplasty (STEP) for the Short Gut Syndrome (SGS) Patients with Gut Failure (GF)

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Introduction: With recent evolution of gut rehabilitation, bowel lengthening is increasingly utilized to restore nutritional autonomy in patients with GF. This is the first largest single center experience with STEP particularly among adults with different causes of SGS including Crohn’s and allograft rejection.

Methods: Over 7 years, 65 SGS-GF-patients received a total of 79 STEP procedures. Sixty one(94%) patients were adults and 4(6%) were children with an age ranging from 2 to 79 years. All patients suffered loss of nutritional autonomy with the requirement for total parenteral nutrition (TPN), or IVF-micronutrient replacement. The leading causes were vascular occlusion (n=22), surgical adhesions (n=13), and Crohn’s disease (n=11). Interestingly, the procedure was performed in an isolated intestinal allograft after partial recovery from acute rejection.

The mean pre-STEP length of the residual bowel was $71 \pm 13$ cm with partially or fully preserved colon in 54(83%) and intact ileo-cecal valve in 34(22%). Reestablishment of gut continuity was accomplished preoperatively in 24(37%) and simultaneously performed in 49(63%). The procedure was reproducible in a total of 13(20%) patients. GLP2 was used as an adjunct therapy before and/or after the STEP procedure in 11(17%) patients.

Results: With a total of 79 STEP-procedures and overall total number of 2 to 36 cuts, there was an increment in the bowel length ranging from 2 cm to 50 cm. The increase in the bowel length was influenced by length of the residual bowel, intestine diameter and the cumulative number of the transverse cuts. With a mean follow up of $19 \pm 7$ months, a total of 28(43%) patients achieved full nutritional autonomy with discontinuation of TPN therapy. The remaining 37(57%) experienced partial reduction in the TPN and IVF support. Length of residual bowel, volume/ calorific contents of initial TPN therapy, and presence of the ileocecal valve were significant predictors of successful outcome.

Conclusions: STEP along with autologus gut reconstruction is an effective bowel lengthening procedure for the management of SGS - GF patients. The operation is also technically feasible in Crohn’s disease patients and recipients of intestinal allografts.
Results of Medical and Surgical Rehabilitation of Adult Patients with Type III Intestinal Failure in a comprehensive unit: Is it possible to predict intestinal rehabilitation?

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Hospital Universitario Fundacion Favaloro

Introduction: Short bowel syndrome (SBS) remains the main cause of Intestinal Failure (IF). The management at a comprehensive unit assures an adequate evaluation, maximizing opportunities to achieve intestinal autonomy.

We aim to report the long term results of medical and surgical rehabilitation of patients (pts) with type III IF (III-IF) and to develop a formula to predict PN independency based on anatomical and clinical variables.

Material and Methods: Retrospective analysis of a prospective database of III-IF adult pts, that underwent Autologous Gastrointestinal Reconstruction Surgery (AGIRS) from 3/2006 to 08/2018. Age, gender, primary diagnosis, pre-surgical anatomy, post-surgical intestinal length (PSIL): grouped as a, b and c (table 1); post-surgical anatomy; (PSAT) type: 1, 2 or 3; presence of ICV, PN reduction >2/7 days. Statistical analysis was done on SPSS v2.

Results: From 259 adults pts with IF, 185 (71%) had III-IF. AGIRS was performed in 88 pts; mean age: 49.5 ± 14.6 years; 51 female. Primary diagnosis: post op IF 48, ischemia 20, post bariatric surgery 5, others 15. The anatomy at first visit was: type 1n 78, type 2 in 3, and type 3 in 7 pts. Mean time on PN before AGIRS was 313.14 ± 483.8 days. PSAT was type 1n 3, type 2 in 26 and type 3 in 59 pts. The mean PSIL was 159 ± 103.4 cm; according to the PSIL, 13 pts were type a; 16 type b and 59 type c.

Mean Time on PN after AGIRS was: 817.6 ± 661.3 days. Table 1 shows outcomes based on PSIL and PSAT. Eight pts were started on teduglutide (TED); the mean time on therapy is 764±616.8 days; at the end of follow up, 6/8 discontinued PN support and 2 significantly reduced PN volume. Freedom from PN survival is 83% for the whole group; 91 and 93% for PSIL type b and c respectively, and 33% for a. Figure 1a. represents the Cox Regression for overall PN independency. The logistic regression analysis let us to build a novel formula:

\[ Y(PN \text{ Free}) = \frac{1}{1 + \exp(-5.178 \times \text{Length A} + 3.866 \times \text{Length B} + 1.886 \times \text{ICV Yes} + 2.737 \times \text{GLP-2 Yes} + 0 \times \text{No} = 1)} \]

The ROC curve of the formula results for this cohort was 0.82 (Figure 1b).
Conclusions: AGIRS surgery allows converting anatomy to a favourable type for intestinal rehabilitation, as well as recovering intestinal length. TED treatment allowed later PN discontinuation in patients with unfavourable predictors. The multivariate analysis showed that PSIL, presence of ICV and the TED use, could predict freedom from PN in this cohort of adult pts.

Introduction: A small percentage of children with intestinal failure suffer from congenital enteropathies responsible for chronic diarrhea associated with severe growth failure and dehydration. These disorders usually present early in infancy and result from monogenic mutations in genes affecting intestinal epithelial function. Recently, next-generation sequencing (NGS) technology has revolutionized the approach to molecular diagnosis, although there still remains little information about the associated natural history, and complete clinical phenotype. To better understand the natural history and the impact of NGS, we studied a historical cohort of children with congenital enteropathies.

Methods: A retrospective analysis of children followed in the intestinal failure / congenital enteropathy programs at Necker-Enfants Malades and Boston Children’s Hospital, Harvard Medical School, Boston, USA. Diagnosis was obtained either from pathology, targeted sequencing, and from 2012 (Boston) / 2014 (Paris) by NGS (whole exome, or targeted gene panels). Genetic findings were validated clinically and/or functionally.

Results: 85 patients were included (53% female, age 2 months to 25 years). Three out of 85 patients died. In 82% (70/85) of cases, a diagnosis could be established. 25 (29%) patients were diagnosed with microvillus inclusion disease (MYO5B mutation), 19 (22%) patients were diagnosed with tufting enteropathy (EPCAM mutations). Other diagnoses included DGAT 1 mutations (8,5%), TTC37/SKIV2L, and SLC26A3 [UMO1] mutations. Since 2012, 37 out of 46 patients (80%) obtained a molecular diagnosis, including the discovery of novel genes (DGAT1, UNC45A, WNT2B). 9 out of 42 patients at Necker-Enfants Malades underwent intestinal transplantation. In a number of patients, diagnosis facilitated by NGS led to significant changes in clinical management such as removal of high-risk immunosuppressive medications.

Conclusion: We carried out an analysis of a large cohort of children with congenital enteropathy, across two major pediatric centers. Mutations in MYO5B and EPCAM were responsible for 52% of our cohort, with considerable genetic heterogeneity in the rest of the cohort. Since 2012, most patients underwent NGS, with a molecular diagnosis achieved in 80% of cases. NGS was critical in achieving a diagnosis in atypical or hypomorphic presentations and enabled clinically significant changes in management as well as the discovery of novel disease-causing genes.
220.6 - Effective radiation dose and bone marrow radiation exposure as a result of radiological investigation in a cohort of multivisceral transplant patients
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Introduction: A patient who experienced a rare form of leukaemia following multivisceral transplantation prompted an investigation of cumulative radiation exposure due to radiological investigations in a cohort of multivisceral transplant patients from our centre. In addition, bone marrow dosimetry was calculated in view of the high incidence of bone marrow suppression in this group of patients.

Methods: Dose data for CT examinations was retrospectively collected using PACS. Effective doses for each phase of each scan were estimated using 2017 updated conversion coefficients. The total number of scans and total effective dose were calculated for each patient and the stochastic risks for general cancer risk of 4.3%/Sv and leukaemia risk of 0.63%/Sv stated by the International Commission on Radiological Protection (ICRP) were estimated. Bone marrow dose per Dose-Length-Product (DLP) conversion factors were created for each anatomical region using the ImPACT Calculator, which allowed total bone marrow doses to be estimated for each patient. Bone marrow dose rates were compared against the acute radiation dose rate threshold for reduced haematopoiesis of 0.25-0.5 Gy/year suggested by the ICRP.

Results: Radiation doses were calculated for 80 patients undergoing multivisceral transplant procedures (+/- liver) at Addenbrooke’s Hospital, Cambridge UK between 1998 and 2018. The average age was 49 years (range 18 - 63). The majority of the radiation dose was incurred by CT scans - 370 in total over an average of 4.4 years per patient (SD 2.6yrs). Mean number of scan phases per patient was 33.7 (SD 17.8). Maximum number of scan phases for one patient was 83. Mean effective dose was 29.1 mSv (SD 22.4 mSv). Maximum effective dose was 30.7 mSv (4.3% general cancer risk, 1.1% Leukaemia risk). 5 patients received cumulative doses in excess of 800 mSv and 2 exceeded 1Sv. The highest bone marrow dose was 0.66 Gy, 0.4 Gy/month. 74 patients exceeded the ICRP recommended limit of 0.02 Gy/month and 11 exceeded 0.25 Gy/year and 3 patients exceeded the monthly threshold for >30 days.

Conclusions: Significant radiation exposure is experienced by this cohort of patients and the threshold dose rate for reduced haematopoiesis is often exceeded with significant potential implications for the development of cancer and bone marrow suppression.

220.7 - Human intestinal tissue-resident memory CD8 T cells comprise two transcriptionally distinct populations
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Introduction: Tissue-resident memory (T\textsubscript{TM}) T cells, long-lived effector T cells in tissues that do not recirculate, are critical in inflammatory, infectious, and neoplastic conditions. However, the study of human T\textsubscript{TM} cells has been hampered by an inability to prove long-term tissue residency. We hypothesised that intestinal transplantation could present an opportunity to study long-lived donor-derived T\textsubscript{TM} cell populations.

Methods: Adult intestinal transplantation recipients were identified via longitudinal cohort studies at Oxford University Hospitals NHS Foundation Trust. Thirteen samples of intestinal graft biopsies were obtained from eight patients. Donor- and recipient-derived T cells in intestinal transplant mucosa were identified using flow cytometry and antibodies to discordant HLA Class I proteins. Proposed residency markers CD69 and CD103 were included in the panel. Fluorescence-activated cell sorting was used to sort donor- and recipient-derived intestinal T cells for single-cell RNA sequencing using the 10X Genomics platform.

Results: The proportion of donor-derived T cells was negatively correlated with time post-transplant, and was highly variable between subjects, with some subjects maintaining significant donor-derived populations up to 5 years post-transplant. Donor-derived T cells uniformly expressed CD69 (>99%), and CD63 was highly expressed (84%), with increased expression at late times. Single-cell RNA sequencing was performed on cells sorted from a single healthy subject 1 year post-transplantation. Data from 974 cells were of sufficient quality for analysis. Analysis of gene expression revealed conventional CD4 T cells, CD4 T\textsubscript{REGS}, and 2 distinguishable CD8 T cell populations within the graft-resident donor-derived cells. The CD8 clusters differed in expression of key residency (CD63) and functional markers (GzmB and HLA class II).

Table 1: Demographics of study subject cohort.

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<th>Number of samples</th>
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<td>Age (years)</td>
<td>6</td>
<td>13 (1-4 per study subject)</td>
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Transplant type:
- SBTx
- MM/VTx

Time from transplant:
- 0-3 months
- 3-12 months
- 12 months +

SBTx = Isolated small bowel transplant; MM/VTx = Modified multivisceral transplant.

Results: The proportion of donor-derived T cells was negatively correlated with time post-transplantation, and was highly variable between subjects, with some subjects maintaining significant donor-derived populations up to 5 years post-transplant. Donor-derived T cells uniformly expressed CD69 (>99%), and CD63 was highly expressed (84%), with increased expression at late times. Single-cell RNA sequencing was performed on cells sorted from a single healthy subject 1 year post-transplantation. Data from 974 cells were of sufficient quality for analysis. Analysis of gene expression revealed conventional CD4 T cells, CD4 T\textsubscript{REGS}, and 2 distinguishable CD8 T cell populations within the graft-resident donor-derived cells. The CD8 clusters differed in expression of key residency (CD63) and functional markers (GzmB and HLA class II).
Conclusion: Donor-derived T<sup>RM</sup> cells can persist in the graft for at least 5 years post-transplant, with highly variable dynamics between patients. Single-cell transcriptomic profiling and flow cytometry indicates two transcriptionally and phenotypically distinct CD8 T<sup>RM</sup> populations within the human intestine. Further study of the regulation, differentiation, and biology of these populations is warranted, as well as their role in intestinal transplantation.

Multivisceral Transplant without an ostomy - 5-year experience

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Introduction: Strides continue to be made in the care of patients following Intestinal Transplantation (ITx) yet, rejection continues to be a threat to its success. A temporary ostomy to assess the mucosa and easily perform a biopsy remains the diagnostic gold standard. However, patients with an ostomy may have high output and decreased renal function. A change in intestinal flora, as stool and mucosa are exposed to air, may trigger rejection.

Methods: We retrospectively reviewed 61 primary ITx performed from Jan 2013 to Dec 2018. 27 Isolated Intestine-Colon transplants and 7 Modified MVT were excluded. 67 patients underwent a MVT. 4 died in OR from bleeding and 1 within 72 hours thus excluded as abdominal closure was not performed. Standard immunosuppression was induction with antithymoglobulin/rituximab and maintenance with tacrolimus/mTOR inhibitor.

Results: Of 62 patients, 50 had 1-year follow up. 26 were pediatric (1-15yr) and 24 adult (19-68 yr). In 54% (27/50), no ostomy was performed. In 46%, an ostomy was performed- ileostomy in 12% (6/23), colostomy in 32% (16/23), hybrid colon ostomy in 2% (1/23). There was no difference between groups (Ostomy/No Ostomy) in length of stay (LOS) or admissions within the first year. LOS was 79 days with Ostomy(13-232) and 80 days (14-360) in No Ostomy. There was a trend for earlier discontinuation of parenteral nutrition in No Ostomy(32 days) vs. Ostomy(53 days).

Initial endoscopy occurred earlier in Ostomy at 10 vs. 27 days in No Ostomy[F (1,48) = 15.99, p=0.0002]. Number of scopes performed in the 1st year were significantly less in patients with No Ostomy(mean = 6) vs. Ostomy(mean = 11) [F (1,48) = 15.17, p=0.0003] (Fig 2). Despite this, there was no significant difference in rejection. There was a trend toward increased rejection in patients with Ostomy(35%, 8/23) vs. No Ostomy(35%, 5/27). There was no difference in creatinine pre-transplant (non-renal) nor at 1-year post. However, the majority of patients with an ostomy (colostomy + hybrid colon) had colon in continuity making dehydration less likely. Patient 1-year survival did not differ at 82% (4/23) for those with Ostomy and 78% (6/27) with No Ostomy.

Conclusion: MVT without an ostomy continues to be feasible and safe without an increased risk of rejection nor death. Postop care seems to be easier and preservation of kidney function may impact long term ITx outcomes. Lack of ostomy greatly adds to patient satisfaction and quality of life.
Disease Recurrence After Intestinal and Multivisceral Transplantation

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Introduction: Similar to other organ and cell transplantation, visceral allograft recipients are at risk for disease recurrence. This single center experience addresses the risk of disease recurrence in both the allograft and extra-gastrointestinal systems in patients who underwent transplant due to a potentially recurrent disorder.

Methods: The database of a total of 78 patients with potentially recurring disease including thrombophilia (n = 29), global gut dysmotility (n = 24), Crohn’s disease (n = 38), and gastrointestinal neoplastic disorders (n = 7) were reviewed and analyzed. Of these, 53 were transplanted at Cleveland Clinic and 25 were transplanted elsewhere. All patients were adults with 63% female. The visceral allografts were isolated intestine (n = 52), liver/intestine (n = 6), modified multi-visceral (n = 12), and full multi-visceral (n = 9). All patients were disease free at the time of transplant, with the exception of the thrombophilic patients who were fully anticoagulated. The diagnosis of disease recurrence was established based on clinical, hematologic, radiologic, and histopathologic studies.

Results: With a mean follow-up of 80 ± 75 months from the transplant date, 7 patients developed disease recurrence with an overall incidence of 9% in this cross-sectional study. The risk of disease recurrence was higher with global gut dysmotility (27%) compared to thrombophilia (20%) and Crohn’s (6%). The 4 transplant recipients (3 isolated and 1 modified multivisceral) with gut dysmotility recurrence had no evidence of mechanical obstruction, or allograft rejection. None of the gastrointestinal neoplastic disorder recipients developed de novo disease or recurrence. The hypercoagulable patients developed recurrent vascular thrombosis in the extra allograft vascular system, with the exception of one patient who developed a nonocclusive Carrel patch clot. Recurrent Crohn’s was a histologic diagnosis based on identification of granulomas in surveillance biopsies. None of the recipients with disease recurrence lost the allograft or required retransplantation except two of the gut dysmotility recipients who underwent successful retransplantation.

Conclusions: Disease recurrence is a potential risk after intestinal and multivisceral transplantation with no significant impact on outcome. Longitudinal follow-up is required to determine the long-term impact on allograft function and quality of life.

Early Predictors of Enteral Autonomy in Pediatric Intestinal Failure: Development of a Disease Severity Score

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Introduction: Patients with short bowel syndrome (SBS) are dependent on parenteral nutrition (PN) while their bowel attempts to compensate for loss of function. Our objective was to create a SBS disease severity score that would predict the probability of achieving enteral autonomy (EA) using clinical variables available in the early postoperative period.

Methods: A retrospective cohort study of SBS children managed by our Intestinal Rehabilitation Program (IRP) was completed. Data abstracted included demographic, anatomic and outcome variables including serum conjugated bilirubin, proportion of enteral nutrition (EN) and episodes of sepsis specifically at 6 months post gut loss. A univariate analysis and Cox proportional hazards (CPH) model was performed. A score predicting EA was created based on weighting of Cox model coefficients. For all analyses, an alpha-value of <0.05 was considered significant.

Results: 19 patients were analyzed (63% males). Ninety-five (68%) achieved EA. Those who achieved EA had a longer residual small bowel (75% vs 24%; p < 0.0001) and colon (100% vs 75%; p < 0.0001) and were less likely to have the ileocecal valve removed (26% vs 57%; p = 0.0005). At 6 months, children who achieved EA had higher enteral tolerance (100% vs 30%; p < 0.0001), a lower conjugated bilirubin (0 vs 71.5 umol/L; p < 0.0001) and less septic episodes (10 vs 2.0; p = 0.03). Cox proportional hazards modeling found >50% residual small bowel (HR 2.68 [95% CI 1.60-4.49] p < 0.001), ICV intact (HR 0.61 [95% CI 0.37-1.02] p < 0.06) and >50% enteral tolerance at 6 months (HR 5.70 [95% CI 2.77-11.74] p < 0.001) were positively associated with EA. Conjugated bilirubin >34 umol/L at 6 months was negatively associated with EA (HR 0.42 [95% CI 0.27-0.66], p < 0.001). A severity score was created by weighting CPH parameter estimates [small bowel length >50%, ICV intact, CB <34umol/L and EN >50% for a maximum score of 8]. Disease severity strata were developed (severe 0-2; 25.7% EA), moderate 3-5; 52.9% EA) and mild 6-8; 97.3% EA). Disease severity strata were developed (severe 0-2; 9/35 (25.7%) EA), moderate 3-5; 8/34 (52.9%) EA and mild 6-8; 6/70 (97.2%) EA).

Conclusion: We propose a paediatric intestinal failure disease severity score that predicts probability of EA, stratified into mild, moderate and severe. The score allows prognostication of individual patients, and could assist research by adjusting outcome reporting or stratifying recruitment.
Results of a Multicentric Retrolective Study of Teduglutide Treatment in Benign Short Bowel Syndrome in Germany

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Introduction: Teduglutide (TED) is a medical treatment for intestinal functional rehabilitation of short bowel syndrome (SBS) patients with chronic intestinal failure (cIF). Its application and effectivity is routinely monitored in a structured home care service program by a multicentric approach in Germany. From these prospectively documented data, a retrolective data base was generated in order to study treatment characteristics and outcome parameters in a clinical routine setting of TED-treated cIF-patients.

Methods: For the present interim analysis, prospectively collected clinical data until December 2018 were retrolectively analyzed. Statistical analyses were performed with SPSS using repeated measures ANOVA and Friedman-Test with subsequent Bonferroni-adjusted post-hoc analyses for TED treatment characteristics up to one year.

Results: So far, 29 patients (f:17 / m:12, median age 52 years) were included in this interim analysis. Causes of SBS included vascular (n=12) and inflammatory diseases (n=6), ileus (n=4), injury (n=4) and others (n=3). Median time on parenteral support (PS) before TED was 30 months. At TED start, 72% of patients (21/29) received individually compounded parenteral nutrition (PN); the remaining 28% received standardized PN. PS was administered by patients themselves (n=15), by relatives (n=3), by nurses (n=8) or others (n=3). TED treatment resulted in a significant reduction in PN calories and volume requirements as well as reduced infusion days per week and shortened infusion times (see Table 1).

The reduction of infusion time was positively correlated with the reduction of PN volume (p<0.001; r=0.75) and calories (p=0.003; r=0.59) after one year of TED treatment. After one year, 22 of 29 (76%) patients were considered responders to TED treatment (with a minimum of 20% i.v.-volume reduction).

Conclusion: In this multicentric real-world analysis of severely PS-dependent SBS/ cIF-patients, TED displays effectivity with regards to improvement of intestinal absorptive function as indicated by significantly reduced weekly i.v.-volume and calorie requirements. In addition, the findings demonstrate a reduction of effective infusion days and times, which both importantly affect patients’ wellbeing. These data indicate the effectivity of TED treatment in a national routine treatment setting in concordance with clinical trial data and guideline recommendations.
250.7 - Impact of donor selection on early graft loss in Intestinal Transplantation.

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Introduction: The decision to accept or not an intestinal graft can be difficult and it is based on the donor data and the clinical experience of the procuring surgeon. Our aim was to analyze potential donor-related risk factors impacting survival.

Patient/Methods: Donor data from the last 50 intestinal transplants (IT) performed in 44 pediatric patients were retrospectively reviewed (20 LBT-20 MVT; median age at transplant 3.4 yo; 2 LSBT, 42 MVT, 2 modified MVT, and 4 ISBT; 40% retransplants). Analyzed variables were donor/recipient age and weight ratio, cause of death, chronic or infectious disease, cardiorespiratory arrest (CRA) or hypotension, days in the Intensive Care Unit (ICU), and macroscopic appearance of the graft at the procurement and at reperfusion. Results were measured as early graft loss (<6 months) and/or incidence of immunological events.

Results: Unstable donors and those with prolonged cardiac arrest were directly rejected at the offer. Median donor age and weight was 16 months (1 day-18yo) and 10 kg (range 3 -68Kg), respectively. The main cause of death was traumatic brain injury (n=17), followed by cerebral hypoxia (n=14), cerebral hemorrhage or infarction (n=13), and meningitis (n=6). Ten donors had chronic disease before death, 14 had suffered a CRA (median duration of 20 minutes; range 1-45), and 18 hypotension. The mean length of stay in the ICU was 2.9 days (range 0.5-9). Graft reperfusion was poor in 2 cases.

In total, 19/50 grafts were lost in 17 patients, after an average time of 10 months (range 1-50), 68% early after transplant, and most of them due to immune complications (62%). In 2 patients the graft never functioned until death. Four patients were retransplanted (30% of the ISBT grafts). Patient and graft 1- and 5-year survival was 83%/73% and 70%/69%, respectively.

The data analysis showed a higher incidence of early graft loss in chronically ill donors and in longer CRA times (p<0.05), and when macroscopic appearance after reperfusion was suboptimal. Donor age or weight, donor/recipient age and weight ratio, cause of death, CNS infection, length of stay in ICU, did not significantly impact on graft survival.

Conclusions: Immune complications are the main reason for the early graft loss in IT. However, ischemia reperfusion injuries observed in marginal donors, as chronically ill patients or longer CRA times, can accelerate the appearance of these complications, leading to irreversible consequences.

260.5 - An algorithm for progress from parenteral nutrition to oral nutrition for adult intestinal transplant recipients

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Introduction: Nutritional care is an integral part in the management of patients pre-, peri- and post- intestinal transplant (ITx). However, the published literature is limited on the topic, and there are no clinical trials. The available published literature is based on individual expert opinions and does not include nutrition protocols in ITx. This work aims to combine the available suggestions for nutritional care for adults pre-, peri-, and post-ITx into a step wise, easy-to-follow algorithm of care.

Methods: The current available literature (Pubmed) was reviewed based on following keywords; bowel transplant; intestinal transplant; adult; human; nutrition; and/or their combinations. Since both authors are experienced registered dietitians who have been working with ITx recipients over a decade, expert clinical opinion was also considered.

Results: We created multicenter collaboration of nutrition care for patients undergoing ITx. Based on our literature review and expert opinion, a detailed, easy-to-use nutrition algorithm was developed (Figure 1). This algorithm will act as a prompt for nutritional considerations in the management of adult ITx recipients per-, peri- and post operatively and hopes to generate discussion and information amnesty, harness feedback from the expert community and, in time, develop further iterations as more information is generated and shared.

Conclusion: The current algorithm is a suggested best fit for the aim stated, bearing in mind the lack of published data available. Further development is needed to have an easy-to-use nutritional care algorithm for future clinical practice.
260.6 - Pediatric Home Parenteral Nutrition (Hpn) In France: A National Survey On The Behalf Of The French Pediatric Hpn Network

Olivier Goulet, et al.
The French Pediatric Home-PN network

Chronic intestinal failure (CIF) requires long-term parenteral nutrition (PN). In France, 7 centers are certified as Pediatric Home-PN (HPHN) and supported by the French Social Security. The aim was to review the activity of the 7 HPHN.


Results: 624 patients < 18 years of age (56.9% boys) attended one of the 7 HPHN program during the period. Turn-over: 18.7% entering and 2% leaving. Increasing population from 265 in 2014 to 352 in 2017 (+33%). Mean & median age at inclusion were 33 & 9 months. Primary digestive disease (PDD) involved 95% of the children. Indications for HPN were short bowel syndrome (SBS): 40.8 ± 7.3 % - congenital enteropathies: 219 ±8.5 % - chronic intestinal pseudo-obstruction: 5.7 ± 8.9 % - Total aganglionosis: 8.3 ± 6.8%; IBD: 4.3 ± 4.2%. Broviac type central venous catheter used in 98%. All patients received tailored PN bags made by hospital pharmacy (3%) or by Baxter-Faconnable® (84%). Intravenous lipid emulsions (ILE) were SMOFlipid® (84%), Clinioleic® (8%), MCT/ LCT (6%) or Intralipid® (2%). Mean and median duration of HPN were 62 and 54 months. Causes of resuming HPN were: weaning off PN (79%), transition to adult (35%), death (5%) (90% of death are cancer or immune deficiency) and intestinal Tx (2%). The main complication was CRBSIs caused by Staphylococcus coagulase negative (70 ± 11%) and Staphylococcus aureus (3.0 ± 11.5%) (5 fungal infection). Tauroli dine lock therapy (TLT) made CRBSIs incidence decreasing from 1.02 per 1000 PN days in 2014 to 0.61 in 2017. Fifteen patients (2.6%) had total bilirubin >30 micromol/l, including 5 cirrhosis, listed for Tx.

Conclusions: HPN is a safe and efficient therapy. SBS is the main HPN indication, with the highest rate of PN weaning. CRBSIs and CLD are potentially life-threatening complications, but their rates were low and deaths (4%) were mostly due to the underlying disease. CRBSIs incidence per 1000 days PN decreased dramatically (~40%) with TLT. Need for intestinal Tx is very limited according to the low rate of life-threatening complications. Patients must be referred early to expert centers for optimal management and follow-up.

Data will be updated for the Congress including results of the year 2018

260.7 - Effective transitioning of adolescents into adult intestinal transplant services – survey of NITE members

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Introduction: Transition of paediatric intestinal transplant recipients is important in allowing patients and their families a continuum of care from current paediatric team to an adult team. In some cases this may be within the same hospital/ organisation, and at other times may be to other centres nationally. The long-term outcome and well-being of adolescent patients is clearly much better in terms of compliance and longevity if trust is built up between the patient and the adult centre with the familiarity of their paediatric teams. Over the past 3 years we have transitioned 8 paediatric patients over the age of 18. We have learnt from having a dedicated discussion with transition nurse and psychologist empowers the patient and families towards a smooth transition journey from paediatric to adult centres.

Method: We undertook a survey of members of the NITE network to understand how transition occurs and what typical problems are encountered. A 10 question survey was sent using SurveyMonkey.

Results: From the survey 26/60 (32%) individuals invited responded from 16 centres across Europe. 8/ 26 (42%) were from a paediatric centre, 8/ 26 (42%) were from a combined centre and 3/ 26 (12%) were from adult only. The majority (84%) had a policy for transitioning patients, over 60% thought about commencing transition when patients were aged 14 – 16, but majority (57%) started transitioning at 16 – 18 and the first combined clinic was only 3% >18yrs. The majority of responses did not have access to a dedicated transition nurse (63%) or psychologist (63%). Lastly, majority of endoscopic procedures were carried out using propofol or GA (63%).

Conclusion: From our experience, transition needs careful planning, a dedicated team to facilitate and early conversations to ensure adolescents are ready to transition effectively. This will require centres to develop extended roles for multi-disciplinary team to support transition, eg dedicated nurse and psychologist, but also start transitioning patients to the same endoscopic strategy that adults undertake and consider appropriate immunosuppression when adolescents may be considering starting a family.
300.5 - Milk fat globule epidermal growth factor-8 (MFG-E8) has site specific effects on intestinal lengthening in neonatal short bowel syndrome, studied in piglets

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Introduction: Whether acquired or congenital, short bowel syndrome (SBS) remains the leading cause of intestinal failure in neonates. Structural adaptation is essential for survival including, uniquely for neonates, the potential for intestinal growth in length. Milk fat globule epidermal growth factor-factor-8 (MFG-E8) is present in human milk and known to increase intestinal fat absorption. It has homology with epidermal growth factor (EGF), which is known to enhance structural adaptation, and so the potential role of MFG-E8 treatment in neonatal SBS was explored.

Method: Using neonatal piglets aged 2-5 days, two surgical models for SBS were studied, both with 75% total intestinal resection, but either distal resection with jejunal-colonic (JC) anastomosis, or mid resection with jejuno-ileal (JI) anastomosis. Piglets were maintained on parenteral nutrition and trophic feeds to enhance adaptation, with 3mg/kg of MFG-E8 or saline (control) given daily by gastric feeding tube. On day 7, bowel length was measured and tissue collected for histology and quantitative real-time polymerase chain reaction analysis of mucosal transcripts including trophic peptides, fat transporters and markers of proliferation. Comparisons utilized student t tests/ANOVA.

Results: All piglets gained equivalent weight during the trial (p=0.65). JI piglets demonstrated significant intestinal lengthening (p<0.001), that was more than 2-fold greater in ileum than jejunum (p<0.001). Lengthening was further enhanced by 22% in ileum following MFG-E8 treatment (p=0.03). In contrast, JC piglets did not increase intestinal length and did not benefit from treatment. In JI piglets, insulin like growth factor-1 (IGF-1; p=0.004) and EGF (p<0.001) expression were increased in ileum and, expression for both was enhanced by treatment in ileum as compared to jejunum (see Figure). In addition, EGF-Receptor (p=0.02) and claudin-2 expression (p=0.026) were also increased in ileum. No other differences in transcripts were noted.

Conclusions: MFG-E8 demonstrated site specific trophic effects, only noted in JI model with remnant ileum. This may limit the utility of this treatment for SBS, where loss of ileum is common. However, the mechanisms of these site specific effects and the roles of IGF-1 and EGF in gut growth warrant further exploration in neonates, particularly given that developmentally relevant conditions, like necrotizing enterocolitis, predominantly target the ileum.

300.6 - The REVE study, preliminary results. A Monocentric Single-arm study to characterize the long-term safety, efficacy, and pharmacodynamic of GLP-2 analog (Revestive®) in the management of short bowel syndrome pediatric patients on home-parenteral nutrition (HPN).

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Introduction: GLP2-analog treatment has been demonstrated to be effective in adult and children patients with SBS. This study was designed to evaluate the efficacy and the safety of Teduglutide during one year in children with long-term intestinal failure due to SBS.

Methods: Children with SBS followed in our center with >2 years on HPN, SB length <80cm, stable on long-term PN (no decrease of PN in the past 6 months) were consecutively included in the study. At baseline they underwent a 4 days hospitalization to perform a stool balance analysis with the duplicate meal technique, blood tests, abdominal ultrasound, densitometry, coloscopy if age >12 years old, and to initiate the treatment. Teduglutide was administered sub-cutaneously at the dose of 0.05mg/kg/day. Visits were every 2 weeks for 8 weeks, then every 4 weeks, then every 2 weeks from week 12 until week 48. At week 48, a second hospitalisation will take place to repeat the stool balance analysis. This study was registered on clinical trials NCT03562230.

Results: Six months after the first inclusion, 12 children had been enrolled. Mean age was 12 years old (range 5-18). Two children had SBS type 1, six had type 2, and 4 type 3. The results of their stool balance analysis (baseline) are shown in table 1. At week 12, nine children (75% children who reached the week 12 endpoint) experienced a decrease >20% of PN requirements (mean 30%). For the three children who reached week 24, further decrease of PN intake was achieved (mean 44%, from baseline). All the children experienced a reduction in stool frequency, an improvement of the stool consistency and a reduction of the ostomy flow.

Citrulline plasma levels increased from 34 µmol/l to 26 µmol/l (mean in the nine children who reached week 12). Five children suffered from mild abdominal pain in the first month of treatment. Only one severe adverse event was reported with an increase ostoma output and abdominal pain which led to a hospital admission for 3 days at week 12; no direct link was made with the treatment which was maintained.

Conclusion: All the children included in the study had severe malabsorption as shown by the stool balance analysis. The first results are encouraging on the safety and the efficacy of the treatment. PN reduction still occurred after week 12 and week 24. The results of the second stool analysis at week 48 should confirm the intestinal absorption improvement that is observed clinically.
Isolated intestinal transplantation: Are we shifting the paradigm?

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Introduction: Isolated intestinal transplant (IIT) is the definitive therapy for intestinal failure. Outcomes have improved tremendously in the last decade. Our center has performed almost 500 intestinal inclusive transplants. Intestine-Colon may be the best nomenclature for better defining the current procedure.

Method: We describe all the patients who received IIT from 2013 to 2018 in our center. It represents our most recent Era with the evolution of surgical technique, immunosuppression regimen, graft monitoring and patient management. Immunosuppression consisted of induction with thymoglobulin and rituximab, maintenance with tacrolimus and mTOR inhibitor and use of basiliximab or vedolizumab for the first 3 months.

Results: 31 IIT were performed in 30 patients at our center. 13 were male. 11 of the recipients were pediatric. Recipient age went from 1 year old to 65 years old. Main indication for transplant was short gut. Positive crossmatch was found in 14% of the transplants. Moderate or severe rejection has happened in 32% of the patients. Only 1 patient underwent to enterectomy within 90 days due to rejection. One patient lost the graft due to volvulus. One patient had the graft removed during emergent surgery due to rupture of mycotic aneurism. Two patients developed PTLD. GVHD was not seen in this cohort. Chronic rejection or chronic graft dysfunction was not observed. Kaplan Meyer 5 year patient and graft survival for primary, non-renal transplants, was 92% and 77% respectively. 5 year patient survival including re-transplants and intestine-kidneys transplants is 87%. Patient survival 1 year remains 100% for all the patients.

Discussion: Survival of intestine-colon transplant has greatly improved over time. Historically, rejection was the main reason for graft and patient loss. In this cohort, only half of the graft losses were due to rejection. Mortality or graft loss due to causes unrelated directly to transplant was the majority in our experience. New immunosuppressive strategies have shown excellent results preventing rejection. Chronic rejection is a well-known long term complication for IIT, although no cases were seen up to date. 100% patient survival for the first year and 92% patient survival for 5 years surpasses all the data available not only for intestines but for all other organs. We believe we are entering a new era in intestinal transplant, with extremely high patient survival even when compared with other modalities of treatment for intestinal failure.
Early Intestinal Barrier Dysfunction Early Post Intestinal Transplantation Is Driven by the Absence of Protective Type 3 Innate Lymphoid Cells and the Persistence of Proinflammatory Type 1 Innate Lymphoid Cells

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Introduction: Innate lymphoid cells (ILCs), the most recently described family of lymphoid cells, play fundamental roles in mucosal barrier immunity, tissue homeostasis, and immune regulation through the activation of host-derived cytokine expression; however, their roles in intestinal transplantation (ITx) has yet to be defined.

Methods: Lamina propria cells were isolated and the following phenotypic definition was used for ILCs: lineage negative, viable lymphocytes expressing CD45 were identified as type 1 and 3 ILCs by expression of CD56, NKP44, CD117, and CD127. Four distinct subsets were further defined as NKP44-ILC1, NKP44-ILC3, NKP44+ILC1 and NKP44+ILC3.

Results: We compared ILC phenotypes via flow cytometry in stable ITx recipients with healthy functioning allografts >6 months after ITx to fresh ITx recipients at day 0 after reperfusion. Surprisingly, we found that protective NKP44+ILC3s (p=0.02) were significantly diminished in fresh allografts compared to NKP44+ILC3s in stable recipients 6 months out. In addition, we found comparable numbers of potentially proinflammatory ILCs (NKP44-ILC1, NKP44-ILC3 and NKP44+ILC3) in both fresh and stable ITx recipients, indicating a dysbalance between protective and proinflammatory ILC subsets in fresh but not stable recipients. Intracellular cytokine staining confirmed that NKP44+ILC3 produced protective IL-22, while ILC3s and NKP44-ILC3 produced proinflammatory IFN-γ, TNF-α, and IL-17. Importantly, serial prospective immunomonitoring of fresh ITx recipients revealed that protective NKP44+ILC3s repopulate by 1 month postoperatively, suggesting that protective and proinflammatory ILCs re-equilibrate in ITx patients over time. Critically, the frequencies of repopulating protective NKP44+ILC3s correlated positively with IL-22 dependent antimicrobial peptide (AMP) expression including β-defensins and RegIIIγ, which are important for intestinal barrier protection. In line with this, we further noted that there was a significant increase in the frequency of protective NKP44+ILC3 in healthy ITx tissue compared to inflamed tissue 1 month after ITx.

Conclusion: Our study indicates that reconstitution of protective ILC3, which is absent immediately post transplant, positively correlates with improved epithelial barrier function through increase of IL-22 dependent AMP expression. In contrast, abundance of proinflammatory ILC1 and ILC3 may be contributing to epithelial barrier breakdown and early clinical complications.

Risks Factors For The Development Of Graft-Versus-Host Disease (GVHD) After Intestinal And Multivisceral Transplantation

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Introduction: Graft versus host disease (GVHD) after intestinal and multivisceral transplantation occurs more frequently when compared to other solid organ transplants. This study reports the incidence, location and outcomes for GVHD patients after intestinal transplantation. Analysis was performed in order to establish risks factors in a high volume center

Methods: Medical records were reviewed from the entire intestinal transplant database from 2003 to 2018. The diagnoses of GVHD were based on clinical features, chimerism assays and pathologic specimens. For analysis, a p<0.05 was considered to have statistical significance

Results: During this period, a total of 272 intestinal transplants were performed, with 23 patients (8.5%) being diagnosed with GVHD. The median time to onset was 79 days, with a 20% of cases diagnosed within the first year after transplant. Chimerism data was available for 38 patients and it was positive in 27 with a range of 2 – 98%. The skin was the most frequent affected organ. Immunosuppression was augmented in all cases. After univariate and multivariate analysis the main risks factors for the development of GVHD were retransplantation (p=0.005), multivisceral transplant (p=0.003) and colon-inclusive grafts (p=0.02). Donor thymoglobulin administration was not associated with GVHD risk reduction. Patient survival was severely affected by GVHD, with a one-year mortality of 39%, mainly due to infectious complications.

Conclusion: This study suggests a strong association between Multivisceral transplants as well as retransplants and, interestingly, colon inclusive-grafts with the development of GVHD. This might be in the context of an increased amount of donor-derived lymphoid tissue at the time of transplant. This information regarding immunological derived complications and GVHD - risks factors should be considered at the time of graft selection.
Spleen preservation attenuates GVHD in multivisceral transplant recipients.

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Purpose: The immunological benefits of the spleen are well acknowledged, but splenectomy is often performed in recipients of multivisceral transplantation. Technical reason and the lack of room in the abdominal cavity are the key arguments to justify removal of the native spleen. We aimed to analyze the beneficial role of the spleen in MVTx recipients in a series of pediatric intestinal transplantation.

Patients and methods: A total of 103 IT (55MV, 25SB, 22CLSB, 3MMV) were performed in 84 patients (male-60%, mean age 5.3yrs) between 1999 and 2018. Mean time on the waiting list was 272days waiting list mortality was 25%. Main indications for the transplant on the first instance were short bowel syndrome (69%), motility disorders (22%) and epithelial diseases (9.5%). For the present study we included only pediatric (<18 years) cases that underwent multivisceral transplantation. Patients were divided in three groups, native spleen preservation, native spleen removal and spleen graft inclusion. We analyzed survival, GVHD, hemolytic disorders.

Results: From the whole series, only 55 patients were included, all of them received a multivisceral graft. The spleen was included in the graft in 31.3% of patients, splenectomy was performed in 40%, in several of them after failed attempt of preservation; and native spleen was preserved in 41.8% of patients. Rejection in different forms was more frequent in those patients who underwent splenectomy. None of the patients with the spleen included in the graft presented humoral rejection, 4% presented chronic rejection and acute cellular rejection was two times more frequent compared with patients who underwent splenectomy. None of the patients with the spleen included in the graft presented chronic rejection, PTLD was also (1.8 times) more frequent in this group. The proportion of GVHD was significantly lower in the group of spleen preservation compared to splenectomy and graft inclusion. Survival was characterized by a lower incidence of GVHD and hematologic disorders.

Conclusion: Native spleen preservation demonstrated to attenuate the occurrence and severity of GVHD in pediatric MVTx recipients. This technique should be considered in all patients, although technical difficulties, prolonged ischemia of the graft and other factors can preclude the success of the procedure.
320.2 - Extending the indications of Intestinal transplantation. Cytoreduction and modified multivisceral transplantation for patients with end-stage pseudomyxoma peritonei.

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Background: Pseudomyxoma peritonei (PMP) arising from a low grade appendix tumour can be cured by cytoreductive surgery and hyperthermic intraperitoneal chemotherapy. However, 40% of patients develop residual or recurrent disease. The inevitable disease progression eventually results in nutritional failure from small bowel obstruction and often abdominal wall failure with fistulation. This leads to poor quality of life and is eventually fatal. Median life expectancy is 3 months in patients requiring parenteral nutrition. Radical cytoreduction and modified multi-visceral transplantation could benefit selected patients.

Methods: Between 2013-2018, 13 pmp patients underwent transplantation. Patients had previous surgery for PMP and further cytoreduction was not feasible due to extensive bowel involvement. Patients had complete or intermittent bowel obstruction with intestinal failure ± fistulation. 11 of the 13 patients were on parenteral nutrition and 2 patients had impending gut failure. 4 patients underwent radical debulking and 7 complete cytoreduction. 6 Patients had isolated intestinal transplant and 7 received modified multi-visceral grafts. Median operating time was 13 hours.

Results: Post-op stay on ITU average 4 days (range 1-45). Nine patients surviving at time of review (Follow up 1 month to 42 months following transplantation); 2 died (Day 26 and day 64) due to post operative complications, 1 due to GVHD at 5 months following transplantation and 1 due to recurrent pseudomyxoma at 40 months. Two patients had acute rejection and 1 patient had borderline rejection of intestinal graft treated with methylprednisolone. QOL data using EQ5D and SF36 showed significant improvements following transplantation with marked reduction in pain.

Conclusion: Cytoreductive surgery followed by modified multi-visceral transplantation is technically feasible for end stage PMP. It could potentially extend life giving independence from TPN with an excellent QOL. Our recommendation is to undertake the procedure in patients with low grade disease where complete cytoreduction is likely to be feasible and with impending or established intestinal failure.
To evaluate the outcomes of the USBS with < 10 cm or < 10% of the expected bowel length for gestational age enrolled in the IRP at CNMC. 24 USBS patients, who had at least 2 years follow-up over the past 10 years, were included. Outcomes involved death, transplant, time to normalized conjugated bilirubin (CB) and parenteral nutrition (PN) requirement. Platelets, albumin, CB, weight and height Z score were obtained at entrance and end of the study. At entrance, median age was 3 months, mean bowel length 7 cm; 6 had IC valve, 9 had < ½ of colon. Median PN need was 100%. 21 of 24 had liver disease, 13 had a mean CB of 7.5 mg/dl, liver biopsy in 7/21 showed fibrosis, stage 3-4 in 8). 17/24 (78%) patients with cholestasis normalized their CB with treatment over a median time of 11 weeks. Fourteen patients had 23 lengthening procedures (LP) at CNMC with no complications. 7 had the 1st LP at CNMC; 7 Bianchi; 4 STEP and 1 Bianchi & Step. Median age at 1st LP was 21 months. 8 had a 2nd STEP and 3 had a 3rd STEP (1 had 2 prior STEP in other institution). 2/14 patients with LP were transplanted and 3 weaned off PN (one each after the 1st, 2nd and 3rd LP). The mean PN needs decreased in the non-transplant patients from 75% prior to 1st LP to 50% and to 44% after the 2nd STEP. In 3 patients who had 3rd STEP the PN decreased from 68% to 33%. Of the 24 patients, one is lost to follow up, 7 were considered candidates for a liver-small bowel transplant; 2 declined to be listed (one is now off PN and the 2nd have decreased the PN needs by 35%). Five were listed, 3 were transplanted, one weaned off PN without transplant and one was unlisted, his PN decreased from 100% to 33%. Five were listed, 3 were transplanted, one weaned off PN without transplant and one was unlisted, his PN decreased from 100% to 33%. Of the remaining 13, 8 (69.2%) normalized their CB with treatment over a median time of 11 weeks.

Blood loss and products

<table>
<thead>
<tr>
<th>Blood loss (ml)</th>
<th>Embolised = 9</th>
<th>Median (range)</th>
<th>Non-embolised = 22</th>
<th>Median (range)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>8000 (1395-27400)</td>
<td>15400 (4700-66000)</td>
<td>p = 0.04</td>
<td></td>
<td></td>
</tr>
<tr>
<td>RBCs (units)</td>
<td>6 (0-32)</td>
<td>11 (4-62)</td>
<td>p = 0.02</td>
<td></td>
<td></td>
</tr>
<tr>
<td>FFP (units)</td>
<td>4 (0-26)</td>
<td>11 (4-26)</td>
<td>p = 0.03</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Platelets (units)</td>
<td>0 (0-6)</td>
<td>4 (0-13)</td>
<td>p = 0.04</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cryoprecipitate (units)</td>
<td>0 (0-7)</td>
<td>4 (0-20)</td>
<td>p = 0.36</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Reperfusion lactate (mmol/L)</td>
<td>4.3 (19-110)</td>
<td>7.1 (2.6-16.7)</td>
<td>p = 0.01</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

By performing embolisation in the operating theatre there was minimal delay in the explant procedure and no increase in the cold ischaemic time. There was a reduction in intra-operative blood loss, blood products and metabolic instability. One patient undergoing a full MVT for PMVT required no blood products intra-operatively. There were no complications associated with the embolisation procedure.

Conclusion: We believe that arterial embolisation is a very useful technique to minimize the blood loss associated with severe PHT during intestine containing transplantation. It reduces blood loss, blood products and metabolic instability. The use of arterial plugs substantially reduces the time required for embolisation and the selective occlusion of visceral arterial branches allows for the preservation of the stomach.
Short bowel syndrome (SBS) is the main cause of intestinal failure. The role of the colon as an energy salvage organ has been demonstrated in adult patients but not in children. The aim of this study was to measure the absorption rate in children with SBS using stool balance analysis and to correlate the anatomical SBS type to absorption rate, citrulline plasma level and small bowel length.

Methods: This was a retrospective study over a two years period. All the children older than 1 year of age, with neonatal SBS and PN dependency who underwent a stool balance analysis in our center were included. Children were divided into three groups according to the anatomical type of SBS (ESPEN classification). The level of PN dependency was estimated using the PN/REE index (PN energy intake on resting energy expenditure calculated using Schofield equation). Citrulline plasma levels were measured. Intestinal absorption rate was assessed using the stool balance analysis. Anatomical types of SBS were compared with bowel length, citrulline levels and intestinal absorption rate by analysis of variance. A predicted average percentage of absorption per SBS type, adjusted for relative bowel length was calculated.

Results:

Patients with an intact colon (type 3) had a significantly shorter remnant bowel (p=0.04). Citrulline plasma levels were lower in SBS type 3 patients but not significantly different (p=0.31). Total energy absorption rate did not differ significantly between the three groups. PN dependency index (PN intake/REE) was 30.4% +/- 26%. For the same SB length, absorption rate was 40% in SBS type 1, 70% in SBS type 2, 80% in type 3.

Conclusion: Children with SBS and a remnant colon showed similar absorption rate than children with type 1 SBS although they had significantly lower small bowel length. Citrulline levels did not show a significant correlation with total absorption rate which strengthened the hypothesis that a part of the absorption rate is due to the colon. The citrulline is a strong marker of enterocyte mass but not of colonic mass. This study confirms the role of the colon in energy salvage in children with SBS. Plasma citrulline levels should be interpreted according to the type of SBS - the remnant colon. Efforts should focus on conservative surgery and early restoration of a colon in continuity.
Conclusions: Peripheral T-cell macrochimerism and early recipient T-cell replacement in the graft could serve as potential biomarkers for guiding personalized immunosuppression in intestinal transplant patients.
Identification of biomarkers for risk, diagnosis, and prognosis of GVHD after bowel transplantation and central role of graft resident memory T cells in pathogenesis

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Introduction: Graft-versus-host disease (GVHD) is a relatively common and highly morbid complication after intestinal transplantation. Its pathophysiology remains poorly understood. Resident memory T cells (TRM) are a newly described T cell subset with memory phenotype localizing to peripheral tissue. We hypothesized that the pathophysiology of GVHD might be related to increased donor TRM in the graft that subsequently migrate into host blood and tissue.

Methods: Intestinal transplantation from deceased donors was performed using our standard method. Graft and blood lymphocytes from 10 patients with GVHD and 34 without were longitudinally analyzed using flow cytometry.

Results: Levels of CD4 and CD8 TRM were approximately 20% higher in the grafts of GVHD vs. stable patients prior to implantation and significantly higher at the time of GVHD (p = 0.02 and 0.04). There was also a mean 60.3% higher level of CD8 TRM in the native bowels of GVHD patients compared to controls and 20–30% higher levels of IFN-γ and TNF-α expression in both the grafts and native bowels of GVHD patients. The percentage of CD4 and CD8 TRM in the blood of GVHD vs. stable patients significantly increased during GVHD (p = 0.005), and expression of HLA-DR, CD57, and PD-1 was significantly higher. There were also significant increases in CD8 effector memory cells (p = 0.0056) and decreases in naïve cells (p = 0.0034). Notably, CD8/PD-1 was also significantly elevated prior to transplantation in patients who later had GVHD (p = 0.025), and a pre-transplant CD8/PD-1 level > 45% was an absolute predictor of later GVHD. Moreover, we found significantly higher percentages of HLA-DR, CD57, and PD-1 in patients with GVHD who died vs. those who survived, and values that always correlated with mortality were CD8/CD57 > 35%, CD4/HLA-DR > 15%, CD8/HLA-DR > 45%, and CD4/PD-1 > 35%.

Conclusion: In the largest longitudinal analysis to date, we demonstrate that increased TRM percentage and inflammatory cytokine expression in graft bowel correspond with increased TRM in blood and native bowel as well as increased cytokine expression in native bowel at time of GVHD. Thus GVHD pathogenesis may depend on donor TRM in graft bowel migrating to the blood and native tissue of recipients. Recipients with higher PD-1 expression, indicating T cell exhaustion, might be more vulnerable, providing a possible biomarker for GVHD risk, while increased expression of maturity and activation markers correlate with prognosis.
The Double-Barrel Enteroplasty: A Novel Intestinal Lengthening Procedure for Short Bowel Syndrome

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2 Division of Child and Adolescent Health, Sydney Medical School, University of Sydney.
3 John Hunter Childrens Hospital, Newcastle, NSW

Advances in the management of Short bowel syndrome (SBS) has resulted in children with shorter lengths of small bowel coming off TPN and achieving enteral autonomy. Although existing bowel lengthening procedures such as the Bianchi Longitudinal Intestinal Lengthening and Tailoring (LILT) procedure and Serial Transverse Enteroplasty (STEP) have enjoyed moderated degrees of success, they have not been without complications and better alternatives are still being sought after. We present our experience with a novel bowel-lengthening procedure for SBS termed the double-barrel enteroplasty (DBE) that is simpler to perform, is less disruptive to anatomy and has the potential to achieve similar to superior results to existing bowel lengthening procedures.

Methods: Ten patients have undergone the DBE at the Children’s Hospital at Westmead between January 2011 and November 2018. Baseline characteristics, complications, time to TPN weaning and growth parameters were recorded prospectively.

Results: The mean age at operation was 21 months (range 4-41 months). Mean pre-operative small bowel length was 74.6cm (36-167cm) with a mean length of 35.6cm (17-60cm) undergoing enteroplasty. Mortality and progression to transplantation remains zero. Six patients have achieved complete enteral autonomy within 2.8 months (0.5-5 months). The most recent patients are still weaning TPN. All patients have normalising growth parameters. One patient required an extension of the DBE for on-going proximal dilatation.

Discussion: The DBE is a safe, effective and potentially superior alternative to existing bowel lengthening procedures for SBS. Advantages include its simplicity to perform, less disruption of bowel and mesentery, less anastomoses, decreased possibility of stenosed segments and maintenance of neuromuscular integrity.
Conclusion: ITX-NS appears to be a safe practice model without obvious adverse impact on outcome compared to standard intestinal transplantation with stoma for the purpose of surveillance endoscopy and biopsy. There is theoretical benefit such as better fluid and electrolyte balance, sparing renal function, better abdominal wound care and patient satisfaction. As of now our center’s policy is not to create stoma after ITX except in patients deemed to be at high risk to develop ACR (loop ileostomy) or in patients who do not have distal colon for anastomosis (creating end colostomy). Longer-term follow up with more cases will help confirm safety and benefit of such an approach.

320.10 - "Stoma or no stoma that is the question": a single-center experience of intestinal transplantation without stoma

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Introduction: Our recent data suggests that protocol-driven, routine surveillance endoscopy and biopsy in the absence of clinical features of allograft dysfunction, does not appear to confer any survival advantage to patients and grafts. This observation called into question the necessity of stoma creation in all intestinal transplant (ITX) recipients. At our center, our practice evolved to ITX without stoma (ITX-NS) since 2015 initially in carefully selected patients. We report our preliminary clinical outcomes of ITX-NS.

Methods: Data analysis was limited to adult ITX without liver allograft between 2015 and 2018. There were 34 ITX which were divided into “Control group (with conventional stoma for surveillance)”, n=18 grafts in patients and “Study group (ITX-NS)”, n=16 grafts in 15 patients. We compared clinical outcomes such as graft/ patient survival, episode of acute rejection (ACR), frequency of endoscopic evaluation, initiation of enteral nutrition and duration of parenteral nutrition after ITX, body weight change, usage of anti-diarrheal medicines and renal function between the 2 groups.

Results: Clinical outcome was similar between the 2 groups as table 1.

<table>
<thead>
<tr>
<th>Patient survival (%)</th>
<th>1 year</th>
<th>3 year</th>
</tr>
</thead>
<tbody>
<tr>
<td>Control group (n=18 grafts)</td>
<td>100</td>
<td>96</td>
</tr>
<tr>
<td>Study group (n=16 grafts)</td>
<td>100</td>
<td>96</td>
</tr>
</tbody>
</table>

Conclusion: ITX-NS appears to be a safe practice model without obvious adverse impact on outcome compared to standard intestinal transplantation with stoma for the purpose of surveillance endoscopy and biopsy. There is theoretical benefit such as better fluid and electrolyte balance, sparing renal function, better abdominal wound care and patient satisfaction. As of now our center’s policy is not to create stoma after ITX except in patients deemed to be at high risk to develop ACR (loop ileostomy) or in patients who do not have distal colon for anastomosis (creating end colostomy). Longer-term follow up with more cases will help confirm safety and benefit of such an approach.

350.5 - Acute Rejection Following Intestinal Transplantation in a Cohort of 442 Transplants Performed Over Nearly 25 Years: The University of Miami Experience

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1 Miami Transplant Institute, University of Miami, Miami, Florida, USA 2 Zagazig University School of Medicine, Zagazig, Egypt.

Background: Intestinal transplantation has emerged as a viable therapy for irreversible intestinal failure. Rejection continues to challenge the long-term patient and graft survival. Recently, the incidence of acute rejection has been reduced to 30-40% due to the use of potent immunosuppressive protocols. Herein, we demonstrate the experience of our center in regards to the incidence of acute cellular rejection after intestinal transplantation over almost 25 years.

Methods: Retrospective analysis of 442 intestinal transplants from 1994-2018 with dividing the recipients into 5 groups depending on the induction immunosuppression used: group1: rATG(34/442); group2: anti-CD25(159/442); group3: alemtuzumab(113/442); group4: rabbit antithymocyte globulin(rATG)(34/442); group5: rATG/ rituximab. Types of intestinal transplant included: isolated intestine(LI)(n=38), liver-intestine(LI)(n=38), modified multivisceral(MMV)(n=39), and full multivisceral(MV)(n=24) allografts.

Results: The incidence rate of acute rejection of any grade over the first 60 post-transplant months was 59%(262/442). Actuarial estimates of acute rejection free survival at 13, 36, 24, 60 months were 33%, 22%, 21%, 17% and 11% in group1; 53%, 42%, 37%, 35%, 29% and 25% in group2; 76%, 51%, 44%, 38%, 30% and 23% in group3; 53%, 50%, 50%, 44% and 33% in group4; 85%, 69%, 68%, 63%, 58% and 58% in group5, respectively. Factors associated with a decreased risk of acute rejection were recipients in group5(rATG + rituximab) followed by recipients who received MV or MMV allografts then recipients who received alemtuzumab induction(P<0.0000001). In group5,rATG/ rituximab seemed to be associated with a low risk of developing acute rejection whether during or beyond the first post-transplant month(P<0.0000001). Overall incidence of acute severe rejection was 20.6%(91/442). Within each induction immunosuppressive group, the incidence rate of acute severe rejection among recipients who developed acute rejection was 7.6% in group5, 29.4% in group4, 36.8% in group3, 39.5% in group2 and 27% in group1. Overall incidence rates of graft loss during the first 60 post-transplant months were 33% and 12.2% in recipients who developed acute rejection, respectively. Overall incidence rate of chronic rejection was 16%(7/442).

Conclusion: rATG/ rituximab induction immunosuppression protocol demonstrated a low hazard rate of developing acute rejection even after the 1st month.
The impact of circulating DSA in visceral transplant recipients without histological evidence of rejection: How should we treat or not treat?

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1 Center of Gut Rehabilitation and Transplantation, Cleveland Clinic
2 Allogen Lab, Cleveland Clinic

Backgrounds: There has been accumulating evidence that donor-specific antibody (DSA) has a negative impact on the outcome of visceral allografts. Prospective DSA monitoring allows detection of circulating DSA without histological evidence of acute rejection. However, its association with acute rejection and natural course without treatment are yet to be determined.

Methods: Between 2012 and 2019, 94 visceral transplantation with 25 liver-contained grafts were performed at a single institution. Induction therapy for preformed DSA included rituximab or bortezomib. DSA were monitored at 1 and 2 weeks, followed by 1, 3, and 6 months, and yearly after transplant. De-novo DSA was not treated without histologically proved rejection. The characteristics and impact of DSA in terms of predictability of rejection, and its correlation of severity of acute rejection were analyzed.

Results: One-year cumulative rejection rate in the entire cohort was 67%. With the protocol of anti-B cell therapy, preformed DSA did not increase the risk of rejection, regardless of the status of liver-free allografts, high titer (>4000 MFI) or persistence of DSA. Moreover, there was no difference in severity of rejection with the presence of preformed DSA (n=12) compared to that without DSA (n=36). However, 3-years graft survival was inferior in patients with preformed DSA to those without (52% vs. 64%, p=0.069), due to increased infectious complications.

De-novo DSA was detected in 32 patients (35%) with an HLA-ABDR mismatch as the only predictor (Odds ratio 17, p=0.036). De-novo DSAs were more likely to be class II with high inclusion of anti-DQ DSA (69%). Prospective monitoring detected de-novo DSA in 25 occasions without ongoing rejection. Accordingly, it turned negative without rejection episode in 19 (76%) (Silent DSA), while subsequent rejection occurred in 6 occasions (24%) (DSA before rejection). In 10 occasions, de-novo DSA turned positive during the rejection episodes (DSA during rejection). The rejection episodes that induced DSA circulation was with a more severe grade.

Conclusions: Increased immunosuppressant induction to treat preformed DSA may compensate the rejection risk, but can negatively impact the overall graft survival. With prospective monitoring and no preemptive treatment, high proportion of de-novo DSA was self-limiting, and rarely precipitated rejection episodes particularly with a severe grade.
350.7 - Post-Transplant Proliferative Disorder Following Intestine Transplantation: Contemporary Single Center Experience

Jason Hawksworth, Asha Zimmerman, Alexander Kroemer, Pejman Radkani, Juan Guerra, Khalid Khan, Nada Yazigi, Stuart Kaufman, Sukanya Subramanian, Hannah Sagedy, Thomas Fishbein, Cal Matsumoto

MedStar Georgetown Transplant Institute

Objectives: The purpose of this study was to examine the characteristics and risk factors for post-transplant PTLD in the intestinal transplant population.

Methods: Retrospective review of intestinal transplant recipients between 11/2003 and 11/2017 at a single-center academic institution. Demographic, immunologic characteristics, treatment, graft survival and patient survival were compared between those who developed PTLD and those who did not.

Results: Of the 234 patients who underwent intestinal transplant, 21 (9%) experienced an episode of PTLD. There was no significant difference between the two groups with regards to age, sex, initial disease type, EBV High risk status, donor characteristics, or prior episodes of acute rejection. Those with PTLD were more likely to have received Thymoglobulin induction compared with controls (54% vs 26%, p = 0.005) and a multivisceral transplant including a liver (20% vs 3%, p = 0.024). The most common presenting signs and symptoms were EBV viremia (89%), lymphadenopathy (57%), and fever (38%). Fifty-seven percent of patients had graft involvement. PTLD was fatal in 29% (6/21) of cases and required explant in 14% (3/21). When comparing those who died from PTLD and those who survived, there was no difference in age, Ki-67, CD20 positivity, type of PTLD, rates of CNS or BM involvement, or treatment type. Patient and graft survival at 5 years was lower in those with PTLD compared to those without (50% vs 70%, p = 0.044 and 45% vs 67%, p = 0.019, respectively).

Conclusions: While the incidence of PTLD has improved in the contemporary period, our experience shows that induction immunosuppression with thymoglobulin is associated with PTLD. Additionally, patients with PTLD had worse graft and overall survival than those who did not.

360.5 - Successful Implementation of Remote Consultation for Patients Receiving Home Parenteral Nutrition

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2 Manchester Academic Health Sciences Centre, University of Manchester

Introduction: Our national Intestinal Failure Unit provides care for patients from across the UK and beyond. Type 3 IF patients are routinely reviewed at 3-6 month intervals. Between Mar 2007-2017 there was a 90% increase in type 3 patients attending our outpatient. Coping with the increasing demand whilst maintaining outpatient capacity and standards is a key component of IF care. Telemedicine provides a strategy for achieving this.

Methods: QI methodology was used to implement and evaluate remote video consultations. Implementation began Dec 2015 via patient consultation and small tests of change. Clinical data were obtained from a prospectively maintained database forming part of ESPEN audit standards. A face to face discussion via the internet using the video call service Skype. An anonymous qualitative satisfaction questionnaire was subsequently completed.

Results: During the study period, patients receiving HPN rose by 13.7% to 285. Twenty-one patients used telemedicine service, totaling 55 contacts. Mean potential distance traveled by telemedicine cohort was 136.6 miles (10-441.8), mean cumulative miles saved was 8,600 miles. Twelve patients used the service on multiple occasions. Seventy percent of patients rated their satisfaction with the system at >90%, with the mean satisfaction of 83%. The mean duration between outpatient appointment offered reduced from 103.7 days to 100.4 days in 2017. One patient had a CRBSI following commencement of telemedicine. 9.5% of the telemedicine cohort were admitted with an HPN complication, compared to an admission rate of 23.5% for the whole HPN cohort.

Conclusions: Telemedicine can release some HPN clinic capacity and help reduce the increasing pressure for patient access to HPN services. Whilst maintaining compliance with NICE and ESPEN guidance, patient satisfaction and patient safety
Long term results of a series of pediatric patients with short bowel syndrome treated at an Intestinal Failure, Rehabilitation and Transplant Program

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Introduction: Patients (pts) with short bowel syndrome (SBS) have high morbidity and mortality mainly related to their residual intestinal anatomy/ function and clinical status. Better outcomes are achieved when this condition is managed at a comprehensive multidisciplinary unit.

Aim: To analyze long-term results and chances of achieving Intestinal Sufficiency (IS) of all children with SBS referred to a comprehensive Intestinal Failure, Rehabilitation and Transplant Unit from 3/2006 to 12/2018

Material and methods: This is a retrospective review of a prospectively filled database, including pediatric pts with SBS. Demographic data, diagnosis, nutritional status, parenteral nutrition (PN) support, as % of the basal metabolic rate (no protein Kcal), intestinal anatomy type (1-terminal jejunostomy-; 2-jejuno colonic anastomosis; 3-jejuno-ileo colonic anastomosis; subtypes by length: A< 40 cm and B > 40 cm; treatment implemented [Groups: PN+MR (PN + Medical Rehabilitation); SR+MR (Surgical + Medical Rehabilitation) and ITx (Intestinal Transplantation)], were analyzed on SPSS.

Results: 62 pts with SBS out of 102 pts with chronic intestinal failure admitted in our program, were analyzed. 41 were male, 25 premature, 48 with neonatal SBS. Etiology of SBS included: gastroschisis (15), intestinal atresia (11), Hirschprung (10), NEC (10), post natal volvulus (7) and perinatal volvulus (4).

Pts’ anatomy type, PN at referral, initial and last follow-up visit nutritional status, are shown in Table 1. Median (IQR) time on PN before referral 0.62 (0.25-2.2) years. Figure 1 shows treatment implemented and outcomes. In the long term 30 (48.4%) pts were able to achieve IS; 25 % of the pts of the PN+MR group, Median IQR 12 (1-2.4) years since PN initiation (sPNi); 69% of the SR+MR group Median IQR 5.3 (1.4-8.9) years sPNi; and 76% of the ITx group (Median IQR): 7 (2.5-30.8) sPNi, (PN+MR vs SR+MR and ITx: p= 0.004). From the ITx group, 5 pts failed in the long-term, re-starting PN, 2 died. Other 2 ITx pts died with IS. The overall 1, 5 and 10-year survival sPNi: 97%, 65.4% and 61.2% respectively, independently of the treatment implemented.

Conclusions: Although IS could be achieved under each line of therapy group, SR+MR allowed higher rate of IS with long term maintenance; ITx had higher chances of achieving IS, but there is drop out in the long term. Long-term survival of pts with SBS can be achieved when they are managed under a comprehensive program.
Ex vivo surgery – explant organs en bloc, resect and tumor resection with a reasonable bowel preservation. To date, 17/20 patients have no recurrence with median follow-up of 2 years.

Conclusions: With experience, in selected patients with conventionally unresectable tumors involving superior mesenteric vasculature, ex vivo tumor resection and intestinal autotransplantation can offer effective tumor removal with a reasonable bowel preservation.
370.6 - The Role of T helper 17 cells in Severe Intestinal Transplant Rejection Renders it a Third Form of Inflammatory Bowel Disease

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Introduction: Severe allograft rejection is one of the strongest prognostic factors leading to eventual graft loss and mortality in intestinal transplantation (ITx), specifically if the rejection is refractory to traditional therapeutic maneuvers such as T cell depletion with thymoglobulin(thymo). A characterization of the alloreactive T cell phenotype in severe rejection of both thymo treatment responsive and non-responsive patients has been lacking and is key for unlocking a precision medicine approach.

Methods: A cohort of 57 ITx patients with severe cellular rejection both before, during, and after treatment with thymo, alongside 28 uncomplicated controls, was selected from our IRB-approved Immunomonitoring and Tissue Bank Study. A polychromatic flow cytometry (PFC) panel with and without PMA/Ionomycin re-stimulation and culture was used to analyze peripheral blood and intestinal allograft samples to characterize surface receptor phenotype and cytokine production.

Results: Immunomonitoring of blood via PFC revealed that peripheral CD3 T cells uniformly deplete in both responders and non-responders (p=0.47), confirming that monitoring CD3 T cells in peripheral blood fails to correlate with treatment responsiveness. However, CD3 IHC analysis of rejecting allografts confirmed that the degree of T cell depletion in the allograft correlates with clinical responsiveness. Graft immunomonitoring via PFC showed CD4+ effector memory, terminally differentiated effector memory, and pro-inflammatory CD4+Th17 cells persist both before and during treatment with thymo. PFC cytokine profile evaluation using ex vivo stimulated T cells showed a significant increase in IL-17 production (p<0.05) from the Th17 T cell population in rejection versus controls, which was also corroborated by rtPCR-arrays demonstrating a striking increase in the Th17-related transcriptome signature in rejection vs controls. Moreover, the Th17 cells of these patients co-expressed high levels of TNF-α, reminiscent of immunological features of inflammatory bowel disease (IBD). Indeed, when 3D non-respondor patients were treated with the anti-TNF-α IBD medication infliximab, 9 experienced histologic and clinical recovery from rejection.

Conclusions: Severe ITx rejection is characterized by a Th17-mediated alloimmune response. Given the immunological similarities to IBD, we postulate that severe ITx rejection can be considered a third IBD, which has important clinical treatment implications.

370.7 - Effectiveness of Infliximab therapy following intestinal transplantation

Robert Venick 1, 2, Arpit Amin 2, Elizabeth Marcus 1, Marjorie Manchandia 1, Jorge Vargas 1, Ronald Busuttil 2, Douglas G Farmer 2
1 Pediatrics, David Geffen School of Medicine, UCLA 2 Surgery, David Geffen School of Medicine, UCLA

Background: We have used Infliximab (INFLX) following intestinal transplantation (ITx) as a rescue agent for early, severe ACR (S-ACR), refractory ACR (R-ACR), and for treatment of chronic mucosal inflammation (CMI). This study aims to review our experience with INFLX in an effort to examine efficacy.

Methods: An IRB-approved, retrospective review of a prospectively maintained single-center database of all ITx recipients receiving INFLX was performed. INFLX therapy was used on a case-by-case basis based on clinical condition/indication starting in 2005. S-ACR was defined as Grade 4 or exfoliative acute rejection. R-ACR was defined as partially treated, ongoing ACR incompletely responsive to standard therapy. CMI was defined as an IBD-like condition including active enteritis with a lymphocyte predominant infiltrate and/or ulcerations not attributable to rejection. INFLX was typically administered at 5mg/kg dose. Doses were administered at 6-8 week intervals for patients with CMI.

Results: 22 patients (12 children; 14 with liver-inclusive allografts) were treated with INFLX for 23 different episodes of allograft dysfunction: S-ACR (n=5), R-ACR (n=8), and CMI (n=9). For S-ACR, the median time for INFLX after ITx was 35 (IQR 30, 44) days. All patients lost their allograft. For R-ACR, the median time for INFLX after ITx was 13 (10, 36) years. In combination with ATG, 3/8 recovered allograft function. For CMI, the median time to for INFLX after ITx was 2.7 (14, 4.7) years. These patient have received a median of 7 (4-13) infusions. 7 of 12 experienced clinical/pathological improvement while 3 had inadequate response. 1 patient stopped INFLX due to PTLD and 1 due to an infusion reaction.

Conclusion: INFLX has been an effective biologic agent for gastroenterology patients with IBD and has been reported to be effective in the treatment of ACR after ITx. Our experience indicates that INFLX is ineffective as a salvage therapy for S-ACR. It may have a role as an adjunct therapy for R-ACR when combined with ATG. INFLX appears to be quite effective in the management of post ITx CMI. Larger, multicenter studies should be considered to further investigate the efficacy of INFLX after ITx.
POSTERS OF DISTINCTION
THURSDAY JULY 4, 2019 SESSION 1
P2A01 - Analysis of the impact of graft reduction techniques on long-term outcome of intestinal transplantation

Girish Gupte 1, Khalid Sharif 1, Maria Coelho 1, Paolo Muiesan 1,2, Jane Hartley 1, Thamara Perera 1,2, Darius Mirza 1

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2 Hepatobiliary Surgery and Transplantation Unit, Queen Elizabeth Hospital

Introduction: The scarcity of paediatric donors has led some units to use larger donors and perform graft reduction to overcome the shortage of small size matched full grafts. The aim of this study was to compare the outcome of patients receiving reduced grafts to those with full size grafts.

Method: The records of 94 Itx performed were reviewed. Patients were divided in two groups, reduced grafts (Group 1) and full size grafts (Group 2) and their details were compared.

Results: 94 Itx were performed in 87 patients, 23 with reduced grafts, 19 Liver and Itx and 4 isolated Itx. Demographics are shown in Table 1.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Group 1</th>
<th>Group 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Recipient Age in months</td>
<td>15 (7-38)</td>
<td>40.5 (8-74)</td>
</tr>
<tr>
<td>Recipient Weight in kg</td>
<td>8.4 (5.58-31)</td>
<td>12.8 (6.53-52)</td>
</tr>
<tr>
<td>Recipient Weight z score</td>
<td>-1.95 (-6.14 to -0.24)</td>
<td>-1.336 (-4.24 to -1.6)</td>
</tr>
<tr>
<td>Surgical procedures</td>
<td>2(1-*)</td>
<td>2(0-8)</td>
</tr>
<tr>
<td>Recipient Residual length</td>
<td>29</td>
<td>60</td>
</tr>
<tr>
<td>Donor age</td>
<td>10.8 (12-480)</td>
<td>72 (2-492)</td>
</tr>
<tr>
<td>Donor weight</td>
<td>30 (20-70)</td>
<td>21(4.5-70)</td>
</tr>
<tr>
<td>D:R weight ratio</td>
<td>2.7 (1.3-6)</td>
<td>134</td>
</tr>
</tbody>
</table>

The length of intestinal resection varied from 45cm to 212cm, usually performed in the mid gut. The cold ischaemic time was a median of 422 minutes in Group 1 and 382 minutes in Group 2. With respect to abdominal closure, both groups had the same incidence of primary closure (56, 62%), however the use of an abdominal prosthesis was commoner in Group 2 (17% vs. 9%). Feeds were started in a median time of 6 days in Group 1 and 5 days in Group 2, while PN was stopped 26 days after SBTx in Group 1 and 18 days in Group 2 (median). Main complications are shown in Table 2.

<table>
<thead>
<tr>
<th>Chronic rejection (%)</th>
<th>Group 1</th>
<th>Group 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>18.18</td>
<td>8.06</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Survival time (days)</th>
<th>Group 1</th>
<th>Group 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>10 (8-6478)</td>
<td>14 (1-6842)</td>
<td></td>
</tr>
</tbody>
</table>

Discussion: Children receiving reduced grafts were younger, more likely to be inpatients at the time of transplant, had longer post-operative ICU and hospital stay, and were transplanted with older donors and with higher D/R weight ratio. This decision of reducing a graft could be influenced by the severity of illness in these patients.

Conclusion: Children with reduced grafts can have a successful long-term outcome comparable with full size grafts.
P2A02 - Developmental and cognitive profile of children with intestinal failure

Riikka Gunnar 1,2, Kaisa Kanerva 3, Silja Salmi 4, Taru Häyrinen 5, Leena Haataja 6, Mikko Pakarinen 1,2, Laura Merras-Salmio 1,2

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7. Dept. of Pediatric Surgery, New Children’s Hospital, Helsinki University Hospital

Introduction: Children with intestinal failure (IF) are at risk of neurodevelopmental problems due to underlying disease, multiple operations, inadequate nutrition and prolonged hospitalisation. In this study we searched for prevalence of neurological impairment among children with IF and its association with risk factors.

Methods: We conducted a prospective cross-sectional study at the nationwide tertiary referral IF centre of the Helsinki University Children’s Hospital during 2017 and 2018. IF patients aged three to sixteen years (n=40) were invited to participate. Those with genetic syndrome or severe neonatal intracerebral haemorrhage (n=12) were excluded. IF was defined as less than 50% of small intestine remaining or requirement of parenteral nutrition for more than three consecutive months. The cognitive and motor skills were evaluated using validated tests; Wechsler Preschool and Primary Scale of Intelligence (WPPSI-III), Wechsler Intelligence Scale for Children (WISC-IV) and Movement Assessment Battery for Children, 2nd edition. To investigate the associations for severe neurocognitive impairment, children who had IQ <66 or serious neurocognitive impairment (n=8, 27%) were compared to those who had IQ ≥ 66 or were estimated to have normal or mildly abnormal cognitive function (n=32, 73%).

Results: Median age of participants (n=30, males=24) was 7.5 (range 3 to 16) years. The median IQ, VCI and PRI scores were 98 (IQR 86 -110), 74 (IQR 60 -92) and 74 (IQR 66 -92) respectively. Ten (35%) patients had an IQ score under 70 (-2 SD), compared to 2.2 % in the normative population. The motor impairment was significant in 10 patients (36%) and milder in 8 (28%) patients. The patient characteristics and comparisons of the patients with severe neurocognitive impairment with others are presented in table 1. The children with severe neurocognitive impairment had less remaining small bowel, greater number of laparotomies and general anaesthesia and longer hospitalisation after birth than others. Children whose PN was started after the age of 12 months had normal median IQ of 96 (86-105), whereas median IQ was 74 (60-85) in children whose PN was started early after birth (P=0.0045).

Conclusion: Clinically significant cognitive and motor impairments are common among paediatric IF patients whose disease manifested during neonatal period. We strongly recommend systematic neurological assessment of all children with IF.

Table 1: Patients characteristic of all patients with intestinal failure and comparison of patients with normal or mildly impaired cognition to those with severe cognitive impairment

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>All study patients (n=40)</th>
<th>Patients with normal or mildly impaired cognition (n=32)</th>
<th>Patients with severe cognitive impairment (n=8)</th>
<th>p-value***</th>
</tr>
</thead>
<tbody>
<tr>
<td>IF etiology:</td>
<td>Gastrodiastasis/other, n</td>
<td>13/5/2</td>
<td>13/5/2</td>
<td>0.1454</td>
</tr>
<tr>
<td>Gestational age at IF, weeks (IQR)</td>
<td>36 (26-38)</td>
<td>36 (26-38)</td>
<td>30 (26-38)</td>
<td>0.0088</td>
</tr>
<tr>
<td>Children born ≤ 28 weeks, n(%)</td>
<td>8 (27%)</td>
<td>6 (27%)</td>
<td>2 (15%)</td>
<td>&gt;0.9999</td>
</tr>
<tr>
<td>Birth weight, grams (IQR)</td>
<td>2230 (1415-3200)</td>
<td>2400 (1600-3799)</td>
<td>1229 (813-3979)</td>
<td>0.3307</td>
</tr>
<tr>
<td>Children with bilirubin ≥ 500 µg/dl, n (%)</td>
<td>13 (43%)</td>
<td>8 (36%)</td>
<td>5 (33%)</td>
<td>0.5420</td>
</tr>
<tr>
<td>Short, lower percentage, % (IQR)</td>
<td>29 (21-72)</td>
<td>40 (29-100)</td>
<td>20 (14-29)</td>
<td>0.2082</td>
</tr>
<tr>
<td>No. of general anaesthesia, n (IQR)</td>
<td>9 (6-14)</td>
<td>8 (5-11)</td>
<td>4 (3-11)</td>
<td>0.8550</td>
</tr>
<tr>
<td>No. of laparotomies, n (IQR)</td>
<td>4 (2-7)</td>
<td>3 (2-5)</td>
<td>10 (**12)</td>
<td>0.2987</td>
</tr>
<tr>
<td>Duration of hospitalisation after birth (months)</td>
<td>48 (1-83)</td>
<td>38 (0-9-3)</td>
<td>6.5 (5.0-11)</td>
<td>0.3471</td>
</tr>
</tbody>
</table>

* Defined as IQ < 66 (n=7) or diagnosed to have serious neurocognitive impairment by neurologist (n=2)
** Defined as IQ ≥ 66 (n=32) or estimated to have normal or mildly impaired cognitive function by psychologist (n=31)
*** p-value is calculated for comparisons between patient groups of normal or mildly abnormal neurodevelopment, and severe abnormal neurodevelopment.

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CIRTA 2019 Book of Abstracts - Posters of Distinction

P2A03 - An assessment of psychiatric needs pre- and post-multivisceral transplant
Elizabeth Fregale, Kelly Bryce, Kelly Collins, Syed-Mohammed Jafri, Yakir Muszkat, Atsushi Yoshida, Michael Rizzari, Nemie Beltran, Marwan Abouljoud, Shunjri Nagai
Henry Ford Health System Transplant Institute Detroit, MI, USA

Introduction: As intestinal transplant becomes a more viable treatment option, there has been increased interest in quality of life and other psychosocial metrics. Systematic review suggests that quality of life improves post-transplant. However, there is limited data regarding psychosocial issues present at initial evaluation and post-transplant. Given that psychiatric issues are associated with increased mortality in solid organ transplant, this study aims to highlight psychosocial patterns that are present at initial evaluation and post-transplant.

Methods: This study was a retrospective chart review of 28 patients who received intestinal transplants between 8/3/10-2/3/17 at an academic hospital in the United States. Psychosocial variables from encounters with health psychology senior staff and trainees were extracted. Descriptive data were evaluated from pre and post-transplant encounters.

Results: Pre-transplant, the most common initial diagnosis was adjustment disorder (68%) with more significant pathology occurring in the remaining patients. 3/28 patients had comorbid substance use issues. 12/28 patients were on psychotropic medications during initial evaluation with 50% of those on multiple medications. Figure 1 describes data from initial psychological assessment. Of those eventually transplanted, 8 required further intervention prior to psychosocial clearance.

Conclusion: Present analysis suggests that many pre-transplant patients have significant psychopathology. Post-transplant a number of psychosocial issues persist and can emerge on an inpatient and outpatient basis. Development of behavioral health protocols for the most common consultations will be a valuable clinical service. These interventions may improve psychosocial outcomes, decrease patient noncompliance and reduce disease burden and cost. Future prospective studies, extending period of follow up, and assessing a wider range of predictors and outcomes are needed.

P2A04 - Successful intestinal transplant recipients have comparable return to work rates as other solid organ transplants
Hannah Sagedy, Sukanya Subramanian, Jason Hawksworth, Alexander Kroemer, Pejman Radkani, Juan Guerra, Annelise Nolan, Ashley Voyles, Thomas Fishbein, Cal Matsumoto
Medstar Georgetown Transplant Institute Washington, D.C.

Background: Intestinal transplantation is an established therapy for patients with intestinal failure suffering from complications of total parenteral nutrition. While the immunogenicity of the bowel has imparted unique hurdles as compared to other solid organs, one and five year survival rates have steadily improved. With increased long-term survival, other metrics of success are of interest. Re-entry to the workforce is an important quality of life indicator that has not previously been reported for intestinal transplant recipients. Re-employment after other solid organ transplantation varies from 22-63%, with kidney recipients demonstrating the greatest rate of re-employment (63%) and lung recipients the lowest (22%).

Methods: All patients who received an intestine inclusive transplant between 2003 and 2018, at our institution, with a post-transplant survival greater than one year were retrospectively identified. Employment, (defined as financially compensated work), was assessed through a questionnaire addressing current work status and return to work within 1-5 years and 5-10 years of transplant. Results are reported as percentages.

Results: 60 patients (20 years of age) transplanted from 2003-2018 were alive at the time of this study. Data from 52 of the 60 patients was available for analysis. 25 (48%) patients were male, 27 (52%) patients were female. Median age at the time of transplant was 41 years (range 20-66) and 40 years (range 13-56), respectively.

20 patients out of 52 reported active employment (38.5%), 22 patients (42.3%) reported employment within 5 years of transplant. Of the 38 patients that were beyond 5 years from transplant, data for 31 was available. 16 of these patients (51.6%) were employed. Among the 32 patients who were unemployed, 3 (9.4%) were retired, 3 (9.4%) were pursuing higher education.

Conclusion: Despite the complexity of intestinal transplant, rates of employment among intestinal transplant recipients are comparable to that of other solid organ transplants. This suggests that intestinal transplant patients are capable of re-integrating into society and leading productive lives. Future aims include elucidating the variables such as support structure, disability prior to transplant, and educational level that may impact re-employment.
P2A05 - Technique of Deceased Donor Vascularized Abdominal Wall Allograft Procurement

Kadivaila Ravindra, Jigesh Shah, Deepak Borle, Detlev Erdman, Debra Sudan
Duke University

Purpose: Vascularized abdominal wall allografts (AWA), which cover solid organs in patients with loss of abdominal domain or extensive abdominal wall damage, remain an essential adjunct to recipients of intestinal and multi-visceral allotransplantation. Ensuring adequate vascular flow to the allograft during solid abdominal organ procurement is critical. Our study aims at developing a standardized technique for AWA procurement.

Methods: Four AWA procurements were performed (2 cadaveric, 2 deceased donors) over a 2-year period. Our study aims to: A) minimize cold ischemia time, B) provide meticulous dissection of the IE vessels while avoiding traction injury and C) provide appropriate vessels to be utilized as extension grafts during re-implantation. In brief, our technique involves incising the skin one inch lateral to the rectus muscle, detaching the flap from the costal margin, identifying the inferior epigastric (IE) vessel pedicles bilaterally and excising the pedicles with the femoral vessels.

Results: Cold ischemia times are prolonged if the AWA is removed prior to the other solid organs. The IE vessels were injured during 2 procurements from traction injury as a result of excess dissection. Iliac vessels are not ideal extension grafts, as these are often sent with liver procurements from traction injury as a result of excess dissection. Iliac vessels are not ideal extension grafts, as these are often sent with liver and pancreas grafts.

Conclusions: The abdominal wall flap should be raised at the start of procurement and placed on the thighs with the inferior rectus attachments intact to avoid traction injury to the IE vessels (Fig.1). The peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2). The IE vessels should be dissected down to the peri-vascular fat around the IE vessels should be preserved to minimize traction injury (Fig.2).
Introduction: Intestinal transplantation (ITx) faces many challenges, among them, the necessity to early detect and treat rejection processes. Improving our understanding on the dynamics of allogenic response will be useful to bring candidate targets to these aims. The objective of our study was to determine the kinetics of mucosal T-cells population in the early phase of small bowel rejection using a Heterotopic IT rat model. We made emphasis in phenotypical, activation and exhaustion markers for the early detection of the rejection phenomena.

Methods: Allogenic heterotopic small bowel IT (n=5) was performed following standard procedure. Brown Norway animals were used as donors and Lewis as recipients. Rats did not receive any immune suppressor treatment. Rejection was monitored by clinical scoring and hematoxylin-eosin staining of intestinal grafts (Gf). T-cells from the peripheral blood (PB) and small bowel mucosa were assessed by flow cytometry. CD3, CD4, CD8, CD25, CD45RC, PD-1 FoxP3 and anti-Lewis MHC I expression was analyzed. Gf samples were taken at day 0, 3 and 7 post-operative (POD).

Results: No clinical signs of rejection or significant histological changes were observed at day 3. At this time, in PB 7.9% ± 0.7 of T-cells were from the donor, meanwhile in the Gf 42.7% ± 6% of T-cells were recipient cells. We observed a significant increase of CD3+CD4+CD25+; CD3+CD4+CD45RC+ and CD3+CD8+CD45RC+ % in comparison with basal levels (PB p<0.05; Gf p<0.001). CD3+CD4+CD25+FoxP3+ cells were also increased in the Gf. At day 7 animals present significant weight loss and histological features compatible with mild to moderate cellular rejection. Differently from day 3 POD, in PB we could not detect donor T-cells. In the Gf 70% ± 7 of T-cells were from the recipient. CD3+CD4+CD45RC+ and CD3+CD8+CD45RC+ % were significantly higher than at day 7 POD (PB p<0.001; Gf p<0.001). We detected increased levels of CD3+CD4+PD-1+ and CD3+CD8+PD-1+ cells in the Gf in comparison to basal levels(p<0.01). No differences in CD25 expression was observed between day 3 and 7.

Conclusion: T-cell replacement in the Gf mucosa is a very fast process and CD25 expression in both PB and the Gf is an early event that precede the histological changes and should be analyzed as a potential biomarker. Considering that T-cells expressing CD45RC have been described as key players in the Gf rejection processes, our results also suggest they could be a relevant target to treat/ prevent ACR.

Conclusion: Our study characterizes ITx rejection as driven by a severely altered Threg/Teff axis with IL-17 producing pro-inflammatory CCR6+Th17 effector and potentially pro-inflammatory Threg cells, which may have strong implications on future clinical therapies.
Introduction: Management of intestinal failure secondary to severe CIPO is based on parenteral nutrition associated to decompression or diversion enterostomies (mainly colectomy). Early prophylactic subtotal enterectomy is not recommended, since small bowel remains partially functional in CIPO. This study evaluates the risk linked to the excluded bowel in such patients.

Methods: Charts of all patients with severe CIPO requiring parenteral nutrition and enterostomies, managed from 1986 to 2017 in our institution, were reviewed. Data about initial management, bowel exclusion, incidence and treatment of enterocolitis and, eventually, transplantation were analyzed.

Results: Forty-two patients were included. Initial management consisted of a complete diverting jejunostomy in 8 patients, ileostomy in 12 and both in 22, leaving an excluded segment of small bowel and colon in 30 and colon in 12 patients.

None of the patient underwent subtotal enterectomy. All patients were treated conservatively, except 3 who underwent 2 ileo-colectomies and 1 diversion, at a median age of 4.6 years (3.6 - 9.6).

Finally, 3 patients underwent a liver and intestinal transplantation and 4 underwent an intestinal transplantation.

Overall mortality was 7% (n=3).

Conclusion: CIPO patients have a risk of enterocolitis but mainly mild episodes that are managed conservatively as compared to long segment Hirschsprung disease patients for whom subtotal enterectomy is frequently required. The main goals of CIPO management remain based on nutritional support, fluid electrolyte restoration, antibiotics for bacterial overgrowth/infections, and control of particularly bothersome symptoms.

11 CIPO patients presented at least one episode of enterocolitis (EC). Median age at first EC was 3.6 years (0.9 - 12). EC episodes occurred in the excluded bowel in 4 patients, in the non-excluded intestine in 2, and both excluded and non-excluded intestine were involved in 5 patients.

None of the patient underwent subtotal enterectomy. All patients were treated conservatively, except 3 who underwent 2 ileo-colectomies and 1 diversion, at a median age of 4.6 years (3.6 - 9.6).

Finally, 3 patients underwent a liver and intestinal transplantation and 4 underwent an intestinal transplantation.

Overall mortality was 7% (n=3).

Conclusion: CIPO patients have a risk of enterocolitis but mainly mild episodes that are managed conservatively as compared to long segment Hirschsprung disease patients for whom subtotal enterectomy is frequently required. The main goals of CIPO management remain based on nutritional support, fluid electrolyte restoration, antibiotics for bacterial overgrowth/infections, and control of particularly bothersome symptoms.
POSTERS OF DISTINCTION
THURSDAY JULY 4, 2019 SESSION 2
P2B17 - Interobserver agreement in histological diagnosis of acute cellular rejection in small bowel allografts: a multicenter study

Thomas Steeelandt 1, Emilio Canovai 2, Inca Hundscheid 1, Rachel M. Brown 3, Olivier Corcos 1, Guido Trentadue 4, J acques Pirenne 1, Kaatje Lenaerts 3, Laurens J. Ceulemans 1, Gert De Hertogh 1, et al.

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3 Department of General Surgery, NUTRIM School of Nutrition and Translational Research in Metabolism, Maastricht University Medical Centre, Maastricht, The Netherlands
4 Birmingham Children's Hospital and University Hospitals Birmingham, Birmingham, United Kingdom
5 Gastroenterology, Beaujon Hospitals, Paris, France
6 Department of Gastroenterology and Hepatology, University Medical Center Groningen, University of Groningen, The Netherlands

Introduction: Outcome after intestinal transplantation (ITx) has improved, however acute cellular rejection (ACR) remains a frequent challenge restricting long-term results. The gold standard for diagnosis of ACR is histological analysis of graft biopsies.

The diagnostic criteria for ACR were defined during the 2003 VIIIth International Small Bowel Transplant Symposium. Although this report provided consensus for international reporting, it has not been validated in a clinical setting. The aim of our study was to evaluate the reproducibility and the interobserver variability of the current histological scoring system in a multicenter study.

Methods: We retrospectively analyzed biopsies on newly stained slides from ITx patients from 3 hospitals. These slides were graded by 2 independent pathologists blinded to clinical information or original diagnosis. One reader was an experienced pathologist from an intestinal transplant center, the second reader was a last year pathology trainee. Biopsies were graded as: no signs of rejection, indeterminate for rejection (IFR) or ACR grade I to III based on current criteria. Only biopsies that were obtained endoscopically with at least 10 identifiable crypts were included in the final analysis.

Diagnoses were subsequently compared between the two readers (weighted Kappa Method) and to the original diagnosis made by several pathologists at other centers (intra-class correlation coefficient).

Results: 40 biopsies were evaluated from 45 patients (27 children, 38 adults) undergoing intestinal transplantation from 2000 to 2014. Median follow-up was 4.3 years (0–12.5). In total, 320 biopsies met inclusion criteria. 38 biopsies in 20 patients were originally described as showing ACR grade 1 of 3. Nine biopsies were IFR.

The overall weighted kappa value for the interobserver agreement was 0.82 (95% CI [0.752;0.882], p<0.001). The intraclass correlation coefficient comparing the pathology panel to the original diagnosis was 0.434 (95% CI [0.365;0.502]).

Conclusion: Despite the difference in clinical experience between the readers, there was a high level of interobserver agreement using the current histological criteria for acute cellular rejection. The lower degree of agreement with the original diagnosis may suggest a learning curve effect, or reflect the focal characteristic of ACR, differentially expressed in the newly cut slides. More importantly, part of the biopsies were evaluated with the old diagnostic criteria.


Jason Hawksworth, Asha Zimmerman, Alexander Kroemer, Pejman Radkani, Juan Guerra, Khalid Khan, Nada Yazigi, Stuart Kaufman, Sukanya Subramanian, Hannah Sagedy, Thomas Fishbein, Cal Matsumoto

MedStar Georgetown Transplant Institute

Objectives: The purpose of this study was to examine the 90-day complications that occurred after intestinal transplantation and to determine whether they effect 5-year patient or graft survivals.

Methods: Retrospective review of intestinal transplant recipients between 1/1/2003 and 12/31/2017 at a single-center academic institution. Complications were classified using the Clavien-Dindo classification system. Five-year graft and patient survival were compared between those with varying degrees of complications.

Results: Of the 214 patients who underwent intestinal transplant, 201 (94%) experienced a Grade II or higher complication, 18 (69%) experienced a Grade III or higher complication, 69 (32%) experienced a Grade IV or higher and 13 (6%) patients died (Grade V) within 90 days of transplant. The average number of complications was 2.6. Medical complications occurred in 166 (75%) patients while 122 (64%) patients had a surgical complication of some kind. Medical complications were categorized as infectious (59%), renal (29%), cardiopulmonary (29%), immunologic (26%), gastrointestinal (2%), hematologic (2%) and neurologic (3%). Surgical complications were categorized as enteric (2%), abscess (15%), chylous (34%), bleeding (34%), wound (13%), thrombosis (7%) and biliary (3%). Patients with Clavien-Dindo Grade IV complications had significantly worse five-year graft and overall survival compared to those with less severe complications (57% vs 72%, p=0.012 and 58% vs 75%, p=0.007, respectively).

Conclusions: Complications are common after intestinal transplantation and are predominantly infectious. Patients suffering from severe complications (Clavien-Dindo IV) within the first 90 days after transplantation have worse graft and overall survival at 5 years.
**P2B13 - Contribution of Video Capsule Endoscopy to the follow-up of intestinal transplantation**

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Introduction: The graft follow-up through conventional endoscopy has technical limitations. The visualization of the entire small bowel by Video Capsule Endoscopy (VCE) may give important information. We collected the experience with VCE after intestinal transplantation, to define its indications and limits.

Methods: Patients with VCE evaluation between January 2014 and December 2018, from one adult and 2 pediatric centers, were included. Clinical and biological data were retrospectively collected.

Results: Thirty-three VCE were performed in 23 patients, including 14 children (22 VCE). Clinical characteristics are summarized in Table. The indications of VCE were control of rejection treatment (24%), protocol control (24%), suspicion of acute rejection (15%) or intestinal inflammation (9%). Adults ingested VCE, whereas in children, endoscopy was required in VCE (68%). The entire small bowel was explored in 23 VCE (70%). Inflammation of the mucosa was seen in 17 VCE (52%), other abnormalities (varices or bleeding) in 3 VCE (9%). VCE was considered in 9 VCE (27%), while VCE was non-contributory in 4 VCE (12%). VCE retention occurred in 3 cases (9%). Interestingly, 5 VCE (15%) demonstrated intestinal inflammation while endoscopy was normal in 3 VCE (9%) or non-contributory in 2 VCE (6%). VCE was useful for diagnosis in 24 cases (73%) and led to a change in management in 6 cases (18%).

Conclusion: VCE is a significant improvement for graft exploration after intestinal transplantation.

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**P2B14 - Predictive Potential of Biomarkers of Intestinal Barrier Function for Therapeutic Management with Teduglutide in Patients with Short Bowel Syndrome**

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Introduction: Teduglutide, an analog of glucagon-like peptide 2, improves intestinal rehabilitation in chronic intestinal failure frequently caused by short bowel syndrome (SBS). However, the mechanisms in the intestinal barrier related to regulation of intestinal permeability (IP) during adaptive response or therapy are not well understood. We analyzed whether measurement of IP or gene expression analysis from mucosal biopsies of selected candidate genes likely associated with IP are useful biomarkers to describe the regulation mechanisms in the intestinal barrier in patients with SBS with or without teduglutide therapy.

Methods: IP was assessed using a sugar drink test containing lactulose and mannitol and urinary recovery rate was used to determine lactulose/ mannitol ratio. Gene expression analyses of mucosal biopsies was performed using qRT-PCR Quantitec Primer Assays for tight junction genes and epithelial markers.

Results: SBS patients showed increased Lac/Man ratio compared to healthy controls (HC), (n=29/34, p=0.0031). Mannitol recovery was decreased in SBS (Mean HC 13.8% vs. SBS 5.4%, p=0.0031), whereas lactulose recovery was similar to HC (mean HC 0.2% vs. SBS 0.36% p=0.2). qRT-PCR analyses showed significant differences in gene expression between SBS patients and HC (n=30/7 ) for YBX3 p=0.048, CRB3 p=0.009, CDK4 p=0.008, CASK p=0.03, and SI p=0.02.

For 12 patients, gene expression data were analyzed before and after up to 12 month of teduglutide treatment. Significant differences in gene expression were received for CASK p=0.01 and SI p=0.02. Further, analysis of citrullin levels in this cohort showed increased citrullin levels in patients with teduglutide therapy (p=0.02).

Conclusions: The analysis of IP gave first insights into changes of intestinal sugar absorption but has not yet been established in SBS patients. Further paired and controlled studies are required to evaluate the specific influence of both altered bowel anatomy and changes in barrier function. Furthermore, altered specific gene expression was shown for both, tight junction formation and genes involved in nutrient transport and should be further analyzed.
Preoperative Arterial embolization for Multivisceral Transplantation for Diffuse Portomesenteric Thrombosis: Performance and Outcome Analysis

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Multivisceral transplant (MVT) for ESLD and stage IV portomesenteric vein thrombosis (PVT), is a highly complex procedure fraught with life-threatening thrombo-hemorrhagic complications. Preoperative visceral arterial embolization is a means to facilitate exenteration and minimize bleeding. Technical success, however, has to be measured clinically through the attainment of poorly perfused/ischemic viscera, and may be further validated by pathological exam of the eviscerated organs.

We present 3 such patients, (2 were redo), who underwent visceral arterial embolization prior to MVT. A descriptive analysis of recipients is presented in Table 1.

A comparative analysis of the embolization procedure, visceral organs perfusion during exenteration, and gross pathological examination of the visceral specimen is outlined in Table 2.

Patient #1 had an angiographically confirmed initial successful deployment of a proximal celiac trunk plug, but in the laparotomy liver perfusion was preserved. Pathological exam of eviscerated organs confirmed the dislodgement and distal migration of the plug into the GDA. In order to preserve liver flow, patients #2 & 3 underwent selective embolization of the splenic and gastroduodenal branches of the celiac trunk. Successful embolization of the SMA was achieved in all 3 patients, and facilitated resection of ischemic intestine, pancreas and spleen with reduced blood loss (4/6/6 pRBCs). However, in all 3 cases a severe coagulopathy with significant hemorrhage occurred during hepatectomy (20/18/12 pRBCs). Patient 1 suffered a full blown DIC, intravascular thrombosis and severe hemodynamic instability that led to intraoperative death. Coagulopathy in patient 2 and 3 subsided post-reperfusion. Circulatory exclusion of the liver in MVT is frequently associated with the development of life-threatening coagulopathy. Heparin prophylaxis is warranted to control consumptive coagulopathy and minimize risk of intravascular thrombosis. Effective heparinization allows a safer management of hyperfibrinolysis with antifibrinolytics when indicated.

In conclusion, MVT in recipients with “hostile abdomen” is associated with severe hemodynamic, hemorrhagic, and hemostatic perturbations. Hepatic sparing embolization of gastrointestinal viscera facilitates exenteration and effectively reduces blood loss. Coagulopathy, however, remains a life-threatening thrombo-hemorrhagic risk, and heparin prophylaxis with diligent hemostatic management are indicated.
Comparison of Short vs. Long Term Anticoagulation Protocols

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Introduction: Catheter related thrombosis (CRT) is a severe and potentially life threatening long-term complication in intestinal failure (IF) but the optimal preventive therapy for CRT is unknown. This study assessed the efficacy and safety of two protocols of secondary anticoagulation (AC) prophylaxis with low molecular weight heparin (LMWH).

Methods: This is a multicenter (N=2), retrospective cohort study of children on home parenteral nutrition (HPN) with a previous episode of CRT. Secondary AC, based on center’s protocol, was initiated once a first CRT was identified. In the short-term protocol (N=10) patients received therapeutic dosing until thrombus resolution or up to 4 months and in the long-term protocol (N=26) prophylactic dosing continued until line removal. All patients underwent routine vascular US at 12m after initiation of AC. The primary outcome was CRT recurrence or progression of CRT at 12 months post therapy.

Results: Patient demographics were similar between groups. Median age at first CRT was 7 months (IQR 2.8–36.5) and median time since PN initiation was 4.5 months (IQR 19–124.4). CRT frequency was similar between PICC line and tunneled central venous catheter (23% vs 28% respectively, p=0.48) and between insertion sides (27% right vs 23% left, p=0.45). Progression/ recurrence of CRT within 12 months of initiation of AC occurred in 7 (70%) patients in the short-term group and 2 (8%) in the long-term group (p=0.001) in a median of 61 days (IQR 35–73). Total CRT recurrence/ progression for the long-term protocol group was 9/26 (35%) patients at a median follow up of 39.5 months (IQR 32–43). Patients with recurrence of CRT had a greater number of catheters/ 300 cutter catheter days (23.5 vs 5.5; p=0.034). The presence of symptoms or complete vessel occlusion at the first CRT episode and frequency of CLABSI between CRT episodes were not associated with an increased risk of recurrence. One patient had a mild allergic reaction, one discontinued treatment following GI bleeding and two at the family and patient’s request. Medication adherence was good (>80% of doses) in all patients in the short-term and 89% of patients in the long-term group.

Conclusions: Long-term secondary anticoagulation prophylaxis with LMWH is more effective than short-term therapeutic AC for the prevention of recurrence or progression of CRT at 12 months. Secondary AC prophylaxis should be considered in children with CRT to maintain long-term venous patency while on PN.

A model to study ischemia-reperfusion injury in human intestinal organoids

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Introduction: Ischemia-reperfusion (IR) injury is inevitable during intestinal transplantation. IR damages the intestinal epithelium, which functions as a physical and immunological barrier and is therefore crucial in maintaining intestinal homeostasis. In order to investigate potential therapeutic targets to protect the epithelium during intestinal IR and promote a regenerative response, we aim to validate a model to study IR in human intestinal organoids. Intestinal organoids have been shown to closely resemble self-renewal kinetics, 3D architecture, and cell-type composition of the intestinal epithelium in vivo.

Methods: A well-established human experimental model to study IR was used for temporal expression profiling of the in vivo intestinal response to IR. The top perturbed pathway was further validated using qPCR. Intestinal epithelial organoids were cultured from crypts isolated from surgical specimens of normal human small intestine. To simulate IR, organoids were subjected to 2 hours of hypoxia (2% O2) followed by 30 and 20 minutes of reoxygenation. Activation of the UPR response was assessed by qPCR for CHOP, GADD34, BiP and XBP1 splicing, and, in addition, signs of endoplasmic reticulum (ER) stress were evaluated using electron microscopy (EM).

Results: The unfolded protein response (UPR) was the top perturbed pathway during reperfusion of the ischemically injured human small intestine in vivo. Subjecting small intestinal organoids to 2 hours of hypoxia followed by 30 minutes of reoxygenation significantly increased expression of UPR-related genes CHOP and GADD34 and splicing of XBP1 mRNA compared to organoids not subjected to hypoxia. In addition, EM showed dilated ER after reoxygenation which is indicative of ER stress.

Conclusion: In line with findings in the in vivo human IR model, revealing the response to unfolded protein as a highly regulated process during reperfusion, hypoxia-reoxygenation in intestinal organoids induces significant activation of the UPR. Intestinal organoids can be used to improve insight in the pathophysiology of epithelial IR injury and regeneration, which could lead to new therapeutic targets.
Feeding difficulties in children with Intestinal Failure

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2 University of Calgary
3 Hôpital Necker-Enfants Malades
4 Departments of Pediatric Gastroenterology, Nutrition and HPN Centre

Introduction: Children with intestinal failure (IF) may be at risk for developing feeding difficulties given the negative stimuli associated with feeding, chronic illness and prolonged hospitalization. There is no literature on the prevalence of feeding difficulties in the IF population, its associated factors or intervention strategies. As a first step, we sought to screen for the frequency of the feeding difficulties and eating behaviours in children and herein provide the results of a preliminary sample.

Methods: Children who were greater than 1 year of age with IF and on home PN for > 3 months were included from two centres: Alberta Children’s Hospital (ACH), Canada and Hôpital Necker-Enfants Malades, France. The validated MCH-Feeding Scale was used to screen for mild, moderate and severe feeding difficulty. The validated parent-rated Child Eating Behaviour Questionnaire (CEBQ) was used to screen for eating styles and food avoidant behaviours.

Results: Preliminary data was gathered for n=34 subjects, median age 6 years with diagnoses of short bowel syndrome (n=22), chronic intestinal pseudo-obstruction (n=4), or other (n=8). Analysis of the MCH-Feeding Scale indicated that 24% of the subjects had no feeding difficulty, while 29%, 18%, and 29% reported mild, moderate and severe difficulty respectively. The subset of n=21 completing the CEBQ rated very high on food fussiness, very high on food enjoyment and very low on satiety responsiveness.

Conclusions: Preliminary data indicate that approximately half of children with IF have moderate to severe eating difficulties. Interestingly, both food fussiness and enjoyment of food are more commonly featured eating behaviours in this sample. It is unclear whether the feeding difficulties of children with IF are related to the impact of the disease etiology and severity, early feeding history, degree of gastrointestinal symptomatology and/or attempts to assert control. Future work will examine how IF specific factors including the use of tube feeds are associated with feeding difficulties.

Kidney Dysfunction after Intestinal Transplantation: Are We Headed in the Right Direction?

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Introduction: Acute kidney injury (AKI) is a common complication after Intestinal transplantation (ITx). There are different etiologies for AKI: high Tacrolimus (TAC) levels, deprivation due to ostomy outputs and acute tubular necrosis (ATN) from sepsis, ischemia, and hemodynamic instability. Modifications in immunosuppressive regimen combined with surgical modifications may decrease the reported incidence of AKI.

Methods: Retrospective analysis of 92 intestinal transplants from 2013-2017: 94.6% (87/92) received the colon. Induction: Thymoglobulin (2mg/kg x5) and Rituximab (150mg/m2 x3); maintenance: 42=TAC + Everolimus (EVL), 24=TAC +Sirolimus (SRL), 26=TAC alone. Target TAC levels: <15ng/ml:0-3 months (mo), 5-15ng/ml:3-6 mo, and 4-6ng/ml:>6 mo. EVL or SRL started after 30 days with target trough levels of 3-5 ng/ml. AKI was defined by recipients who developed an increase in nadir creatinine and required hospitalizations. Estimated glomerular filtration rate (eGFR) was calculated from routine serum creatinine using the CKD-EPI formula for adults and bedside Schwartz formula for children. Median follow-up: 27 [3-56] mo.

Results: AKI occurred in 26.3% (24/92) of ITx, 75% (18/24) in the first 6mo. Median eGFR at 2, 3, and 12 mo was 113.5 [85.0-151.4], 99.8 [65.1-131.0], and 97.3 [55.1-125.9], respectively. Operative time was markedly reduced (median time: 288 minutes) as compared to previous eras. Median TAC levels (ng/ml) at 7days and at 1, 2, 3, and 12 mo: 10 [7.7-14.1], 10 [9.3-14.3], 8.5 [6.8-12.1], 7.5 [5.8-10.5], and 5.7 [4.9-9.0], respectively. Cox regression analysis of the hazard rate (HR) of developing AKI found 2 significant predictors: sepsis (23/92, P<0.0000) and acute rejection (AR) (7/92, P<0.0000). Similarly, the HR of developing an eGFR<60ml/min 173m2: older age at transplant (P<0.0000), AR (7/92, P=0.01), and dehydration on >2 hospitalizations (12/92, P=0.02). Intestinal graft failure (IGF) occurred in 9/92 patients; death with a functioning graft (DWFG) was observed in 12/92. Neither the development of an AKI nor an eGFR<60ml/min 173m2 were associated with the HR of developing graft loss (P>0.3) or DWFG (P>0.5).

Conclusion: Short-term renal dysfunction post ITx still exists. Both surgical modifications and changes in the immunosuppressive regimen have led to diminished AKI and impact on graft failure. Long term follow-up is needed and underway.
Teduglutide Increases Adaptation In A Murine Short Bowel Model By Improving Epithelial Tight Junction Selectivity

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3 University Medical Center Rostock, Institute of Experimental Surgery

Introduction: Teduglutide is used in chronic intestinal failure in order to reduce the need for and to increase time off parenteral nutrition. A trophic effect is induced through GLP-2 stimulation resulting in an improved uptake of water and sodium. It is unclear if the Claudin-10 and -15 mediated sodium selectivity of the paracellular tight junction in the jejunum is influenced. Thus, the effects of Teduglutide were studied in a murine short bowel model.

Methods: By resecting 40% of the ileocecal region (ICR) a severe murine short bowel syndrome was simulated. Subcutaneous Teduglutide (0.1 mg/kg BW) or vehicle administration began 36 h postoperatively. Survival, development of body weight, stool consistency, plasma aldosterone levels, tight junction protein expression, FITC-4kDa-Dextran-Flux and dilution potentials using an Ussing chamber were analyzed.

Results: Compared to vehicle, the development of body weight in Teduglutide treated animals was more favourable (weight nadir vehicle: 83.6 ± 14 % n=15 vs. Teduglutide 87.8 ± 14 % n=12, p<0.05). Lower aldosterone levels in Teduglutide treated animals indicated a better volume state in this group (vehicle 988 ± 172 ng/l, n=6 vs. Teduglutide: 552 ± 915 ng/l, n=9-11, p<0.05). The transmucosal barrier for macromolecules in the jejunum in short bowel conditions increased independently of Teduglutide administration. Occludin and Claudin-15 mRNA expression was increased after ICR, independent of Teduglutide treatment. In the jejunum of vehicle treated animals, cation permselectivity was impaired. At the same time, Claudin-10 mRNA expression in vehicle treated mice was reduced to 55.2 ± 8.5% of baseline, while it was maintained in Teduglutide treated animals at 109 ± 23.7%, n=3-4, p<0.05. In line with this, tight junction localization of Claudin-10 faded towards the villus tips in vehicle treated mice while permselectivity and Claudin-10 expression up to the villus tip remained constant in Teduglutide treated animals.

Conclusion: Teduglutide alleviates intestinal insufficiency in this mouse model of short bowel syndrome. Teduglutide not only induces trophic effects but retains epithelial function by maintaining Claudin-10 expression. This translates to improved paracellular cation permeability, facilitating sodium recirculation and thus sodium coupled nutrient transport, leading to improved nutrition status.
POSTERS OF DISTINCTION
FRIDAY JULY 5, 2019
SESSION 1
P3A01 - The first three years of the New Zealand National Intestinal Failure Service: emerging disparities in ethnicity and social deprivation in children with intestinal failure

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Introduction: The New Zealand National Intestinal Failure Service (NZ NIFS) was established in 2015, with the aim of understanding the aetiology and management of intestinal failure (IF) in NZ. One goal of NZ NIFS is to achieve health equity for patients with intestinal failure in NZ.

Method: The NZ NIFS patient registry collected data for patients with intestinal failure in NZ. Intestinal failure was defined by the requirement for intravenous nutrition (IVN) for ≥ 21 days for paediatric patients up to 3 years of age and ≥ 30 days for preterm neonates (< 34 weeks gestation). Data were reported by the primary clinicians of each IF patient in conjunction with the NZ NIFS from October 2015 to October 2018. Demographic data including ethnicity, social deprivation index, cause of IF and clinical outcome were collected.

Results: Data for 208 patients were collected, 116 (56%) were male, 91 (44%) were preterm. Pasifika and Maori children were marginally over represented accounting for 13% and 28% of the patient cohort, respectively, compared to 9% and 26% of the NZ paediatric population.

More significantly, IF patients with a high NZ socioeconomic deprivation score were over represented, with 36% in the highest deprivation quintile and 9% in the least deprived quintile, compared to 24% and 19% respectively, of the NZ paediatric population.

Most common causes of IF in this group were mechanical obstruction (22%) and short bowel syndrome (18%), although for preterm neonates the prevalence of short bowel syndrome was higher (36%) (Table 1). Ninety seven (47%) children with IF required intravenous nutrition for ≥ 60 days. One hundred and seventy (82%) achieved enteral autonomy, with 14 (7%) discharged on home IVN. No patient required intestinal transplant.

Table 1: Intestinal failure patients in New Zealand by clinical indication

<table>
<thead>
<tr>
<th>Indication for IF</th>
<th>Preterm patients</th>
<th>Paediatric patients</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal wall defects</td>
<td>10</td>
<td>16</td>
<td>26</td>
</tr>
<tr>
<td>Extensive small bowel resection</td>
<td>0</td>
<td>6</td>
<td>6</td>
</tr>
<tr>
<td>Intestinal atresia/malrotation</td>
<td>0</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Mechanical obstruction</td>
<td>19</td>
<td>28</td>
<td>47</td>
</tr>
<tr>
<td>Peritonitis</td>
<td>23</td>
<td>10</td>
<td>33</td>
</tr>
<tr>
<td>Short bowel</td>
<td>31</td>
<td>5</td>
<td>36</td>
</tr>
<tr>
<td>Other</td>
<td>15</td>
<td>16</td>
<td>31</td>
</tr>
<tr>
<td>Total number of IF patients</td>
<td>91</td>
<td>137</td>
<td>228</td>
</tr>
</tbody>
</table>

Figure 1: Clinical outcomes of preterm and paediatric intestinal failure patients in New Zealand

Conclusion: Disease patterns for children with IF are similar to those reported internationally. Future work will focus on further understanding the disparities in ethnicity and social deprivation to achieve equitable outcomes for NZ children with IF.

P3A02 - Racking up the losses: SBS care in Germany - a cost of illness analysis with real life data

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University Hospital of Bonn

Introduction: Short-bowel syndrome (SBS) is considered an orphan disease and no data on the cost of illness (COI) for SBS are available for Germany. This study evaluated the complexity of medical care and economic burden on a center with a specialized intestinal rehabilitation program using real life data.

Methods: 11 consecutive SBS patients of the intestinal rehabilitation program at the University Hospital of Bonn, Germany were included. A total of 979 parameters of treatment-related resources were considered for the year of diagnosis (year0) and a follow-up (FU) of 3 years. This included cost for in- and outpatient care, medication and parenteral nutrition. Subsequently, COI based on these parameters was calculated according to the corresponding service catalogs for Germany.

Results: Median inpatient days were 96 (IQR 59;119) in year0 and decreased to 3 (IQR 0;34) in year3 of FU. In contrast, outpatient visits increased from 1 (IQR 0;11) in year1 of FU to 2 (IQR 0;3) in year3 of FU, with a median of 8 visits (IQR 4;19) per patient in year1. Median total COI for inpatient treatment amounted to 13889€ in year0. This sum consecutively decreased from 33225€ to 22127€ and 4205€ in year2/3 of FU, respectively. While major visceral surgery, open abdomen treatment and reconstructive procedures significantly impacted on COI in year0, catheter-associated complications and related vascular surgery mainly contributed to COI during further follow-up. Contrary to the calculated COI, actual reimbursements for inpatient treatment according to the German Diagnosis Related Groups amounted only to approx. 25%, with a median of 2704€ (IQR 1092;78520). For outpatient care, calculated median total COI was 32645€ per patient, while reimbursement only amounted to approx. 31.5% (540€) in 2017. Median COI for prescribed medication and parenteral nutrition in 2017 was 6752€ (IQR 4990;5313) and 48485€ (IQR 29740;54442), respectively. Only one patient received GLP-2 analogon treatment, which added 5442€ p.a. to the medication cost for this patient.

Conclusions: The complex medical care required for SBS patients resulted in high COI both for in- and outpatient settings. The identified spectrum of surgical/medical interventions reflected a shift in medical treatment over time. Large discrepancies between COI based on "real life" resource expenditure and actual remuneration were found. This imbalance may endanger adequate care for SBS patients in Germany in the future.
P3A03 - Pre-, peri-, and post-operative predictors of survival after intestinal transplantation: results from a single-center analysis.

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Introduction: Intestinal transplantation (ITx) can be a successful procedure for select candidates with intestinal failure (IF). However, there is limited data on variables and characteristics that predict successful outcome. The aim of this study was to analyze a large dataset to determine outcome predictors.

Methods: An IRB approved, prospective database has been maintained since the inception of the program in 1991. Patient inclusion was closed in December 2018. All recipients of intestinal grafts were included. Multiple pre-, peri, and post-transplant variables (n = 35) were collected and included in the analysis. The end point was 1-year patient and graft survival. Standard statistical analysis was undertaken.

Results: 128 patients underwent 151 ITx including 35 isolated intestine (I-ITx), 77 liver-intestine (OLT-SBT), 29 multivisceral (MVT) and 11 modified multivisceral (MMVT).

58% were male, 78% were children, and 77% had surgical IF etiologies. At ITx, the median MELD/PELD at ITx was 34 with 43% hospitalized. The majority of patients received induction immunotherapy with either interleukin-2 receptor antagonist [IL2RA] (58%) or antibody induction with rabbit antithymocyte globulin [rATG]/OKT3 (34%).

Relevant patient clinical characteristics are summarized in the table below:

Significant predictors for improved graft survival were:
Recipient weight (<20 kg), age group (pediatrics), graft type (liver inclusive intestinal graft), panel reactive antibody < 20%, absence of donor specific antibodies, negative crossmatch, warm ischemia time < 1 hr, use of induction immunosuppression, ventilator time < 7 days, length of stay < 80 days, and transplant era (2001-2018).

Significant predictors for improved patient survival were:
Cold ischemia time < 20 hrs, warm ischemia time < 1 hr, absence of recipient splenectomy, use of induction immunosuppression, length of stay < 80 days, and transplant era (2001-2018).

The patient and graft survival at 1 year was 82% and 69%.

Conclusion: The results of ITx have improved over time. Several perioperative factors were identified that impact early outcome. Especially important for graft survival are variables that can be altered including donor: recipient match, preformed donor specific antibodies, ischemia times, and immunosuppression. Further investigations are required to improve ITx outcomes.
P3A04 - Cost-effectiveness of teduglutide in adult patients with intestinal failure: Markov modeling using traditional cost-effectiveness criteria

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Background: Adults with intestinal failure often undergo rehabilitation with the goal of enteral autonomy. However, at 5 years, intestinal failure has a 25% mortality with nearly half of the survivors remaining on parenteral nutrition (PN)[1]. Teduglutide is a GLP-2 analogue that promotes intestinal adaptation and can decrease PN dependence, resulting in enteral autonomy in some and decreased PN in many more. Unfortunately, teduglutide, an orphan drug, is expensive with an estimated cost of over $400,000 per year. The current study evaluated the cost-effectiveness of using teduglutide in adult patients with intestinal failure.

Methods: A Markov model was used to evaluate the costs (in US dollars) and effectiveness (in quality-adjusted life years or QALYs) of treatment with a presumed starting age of 40 years. Parameters were obtained from published data or estimation. The primary effect modeled was the increased likelihood of reduced PN days/week when using teduglutide, leading to greater quality of life and lower daily PN costs. Future costs and utilities were discounted at 3% per year. Sensitivity analyses were performed on all model parameters.

Results: In the base scenario, teduglutide cost $949,910/QALY gained. On one-way sensitivity analysis, only reducing teduglutide cost decreased the cost/QALY gained to below the typical threshold of $100,000/QALY gained. Specifically, teduglutide cost would need to be reduced by over 65% for it to reach the threshold value (Figure 1). Probabilistic sensitivity analysis favored no teduglutide in 80% of iterations at a $100,000/QALY threshold (Figure 2). However, 13% of iterations actually found teduglutide therapy to be cost-saving.

Conclusions: In summary, teduglutide does not meet a traditional cost-effectiveness threshold as treatment for PN reduction in adult patients with intestinal failure compared to standard intestinal rehabilitation. Subpopulations that demonstrate maximum benefit could be cost-saving. Addressing cost alone would require a significant reduction in teduglutide price to improve cost-utility.

References
**P3A05 - Young donors (age ≤ 6 months) for Intestinal transplantation (ITX): are they high risk?**

Khalid Sharif 1, Darius Mirza 1, Paolo Muiesan 1,2, Thamara Perera 1,2, Jane Hartley 1, Girish Gupte 1

1 Liver unit (including small bowel transplantation) Birmingham Women's and Children's Hospital
2 HPB Surgery & Liver Transplantation Queen Elizabeth & Birmingham Childrens Hospital

**Introduction:** ITX is accepted treatment for children with irreversible intestinal failure. Shortage of size matched organs for children especially those with restricted abdominal domain due to various aetiologies resulted in development of various techniques including use of organs from young donors. There is no data on the utilization and outcome of young donors in ITX.

**Subject and Methods:** Retrospective review of medical records between 1993-2018 of all ITX from donors less than 6 months of age. Donor and recipients demographics, intra and early post-transplant complications, short and long term graft and patient outcome were analysed.

**Results:** 4 females median age 20 months & weight 9kg underwent ITX from young female DBD donors median age 3.5 months & weight 6kg with median donor to recipient weight ratio 0.5 (Table 1). All donors with brain death, blood group identical/compatible with median cold ischemia time 5.5 hours. No surgical complications noted intra or early post-transplant. Two died early within 3 months of ITx following severe acute rejection. One established early full enteral feeding and required laparotomy for stoma prolapse 3 months post ITX, but eventually died secondary to complications. The second child took longtime to establish feeding due to fluid and electrolyte imbalance, but eventually died following complications associated with stoma closure. One year patient and graft survival was 50%, whilst 3 year patient and graft survival was 0%.

<table>
<thead>
<tr>
<th>Recipient age months</th>
<th>Recipient age kg</th>
<th>Donor age months</th>
<th>Donor weight kg</th>
<th>ITx graft with liver</th>
<th>Complications</th>
<th>Survival months</th>
</tr>
</thead>
<tbody>
<tr>
<td>216</td>
<td>9.3</td>
<td>6.6</td>
<td>7</td>
<td>No</td>
<td>GVHD,PTLD</td>
<td>18.27</td>
</tr>
<tr>
<td>38</td>
<td>8.6</td>
<td>2.9</td>
<td>5</td>
<td>Yes</td>
<td>GVHD</td>
<td>26.3</td>
</tr>
<tr>
<td>20.9</td>
<td>8.7</td>
<td>19</td>
<td>4.5</td>
<td>Yes</td>
<td>Severe rejection</td>
<td>2.6</td>
</tr>
<tr>
<td>39.6</td>
<td>13</td>
<td>4.2</td>
<td>6</td>
<td>Yes</td>
<td>Severe rejection</td>
<td>14</td>
</tr>
</tbody>
</table>

**Conclusion:** It is technically feasible to consider young donors for intestinal transplantation. In our experience it is associated with a high mortality. Further investigation to understand the complex interactions between the naive immune system of the young donors with the mature immune system of the older recipients may hold the key to understanding the development of the immunological complications.

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**P3A06 - Muscle mass after intestinal transplantation in children is greater than in those on home parenteral nutrition**

Kushila Rupasinghe 1, Jonathan Hind 1, Girish Gupte 2, Pamela Allen 1, Hina Rizvi 2, Arif Wahid 2, Carly Bambridge 1, Dhamyanthi Thangarajah 1

1 King's College Hospital, London, U.K.
2 Birmingham Children's Hospital, Birmingham, U.K.

**Introduction**

Sarcopenia or loss of lean muscle mass is associated with disease states and malnutrition. The effects of both are noted in intestinal failure, intestinal transplant, and adult liver transplantation outcomes. Psoas muscle cross sectional area (PCA) is a validated proxy marker of muscle mass. We look to characterise it in home parenteral nutrition (HPN) and intestinal transplant (IT) patients. Determinants of PCA were explored in IT patients.

**Methods**

This was a multi-centre, retrospective, case-notes based study. Existing axial imaging was analysed, pre- and/or post-transplant. Control images were from a trauma series of healthy children. The PCA was measured at L4 with the psoas muscle examined for ectopic fat content and perinephric fat presence, a validated marker of visceral adipose tissue. Univariate and linear regression analysis was used to identify factors associated with PCA.

**Results**

At total of 81 children were included in the study. 51 with intestinal failure or post-transplant (males=26) and 30 controls (males=22).

![Image of recruitment flowchart](image)

6 scans were analysed with 24 HPN (males=12) and 43 IT (males=23). See figure 2. In the HPN and IT groups after adjusting for sex, age and pubertal status, PCA was found to be significantly lower than controls; HPN having 151cm² and IT patients having 113cm² smaller PCA than controls (p< 0.05). In the total intestinal rehabilitation (HPN and IT) group, after adjusting for sex, age and pubertal status, PCA was 166cm² larger in the IT group than those on HPN (p< 0.05). Length of stay in intensive care was a negative predictor of PCA. There was a trend toward lower PCA in patients that had died. IT had significantly more psoas fat than controls (p< 0.05) and demonstrated presence of perinephric fat on more occasions than controls (p< 0.05).
Conclusion

It is associated with greater muscle mass when compared to HPN. This suggests improved nutritional status after transplantation. Adiposity was found to be aberrant in IT patients. PCA is a novel biomarker of nutritional status and may be developed further in intestinal rehabilitation.

P3A07 - Impact of Donor and Recipient CYP3A5 and ABCB1 Genotype on Tacrolimus Trough/Dose Ratios Following Intestinal Transplant

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2 Department of Pharmacy, Mount Sinai Hospital, New York, NY, USA
3 Department of Genetics and Genomic Sciences, Icahn School of Medicine at Mount Sinai, New York, NY, USA
4 Sema4, a Mount Sinai venture, Stamford, CT, USA
5 Charles Bronfman Institute for Personalized Medicine, Mount Sinai Hospital, New York, NY, USA
6 Department of Population Health Science and Policy, Mount Sinai Hospital, New York, NY, USA

Introduction: Tacrolimus (TAC) is a mainstay of immunosuppression after intestinal transplantation (ITX), but dosing is complicated by a narrow therapeutic index and broad variability. Studies in kidney and liver transplantation have identified the influence of CYP3A5 and ABCB1 genetic variants, both of which are expressed in the intestinal epithelium, on TAC pharmacokinetics; however, no studies have examined the influence of pharmacogenetic (PGX) variation on TAC exposure in ITX.

Methods: This was a single-center, retrospective analysis of adult ITX recipients from 2011-2018. Patients were included if they received a non-liver containing ITX, maintained graft function for 3 months, and had donor and recipient tissue samples stored. DNA was isolated from sample tissue and multiplexed genotyping of CYP3A5*2,*3,*7 and ABCB1 c.3435C>T was performed. Donors or recipients with a *1 allele were CYP3A5 expressers, while patients with a CC genotype were ABCB1 expressers. A combined analysis was also performed that grouped patients by total donor and recipient CYP3A5 and ABCB1 allele score. Weight-normalized TAC trough/dose (T/D) ratios were assessed for each patient during month 1 and at 4, 12, and 52 weeks post-ITX. The primary outcome was difference in mean T/D ratios between the genotypes across time points.

Results: A total of 31 patients were included in the analysis (Table 1). After genotyping, 22.6% of recipients and 32.3% of donors were CYP3A5 expressers; and 35.5% of recipients and 25.8% of donors were ABCB1 expressers. Donor or recipient with a *1 allele were CYP3A5 expressers, while patients with a CC genotype were ABCB1 expressers. A combined analysis was also performed that grouped patients by total donor and recipient CYP3A5 and ABCB1 allele score. Weight-normalized TAC trough/dose (T/D) ratios were assessed for each patient during month 1 and at 4, 12, and 52 weeks post-ITX. The primary outcome was difference in mean T/D ratios between the genotypes across time points.

Conclusion: In the first study evaluating PGX in ITX, our results suggest that both donor and recipient CYP3A5 and ABCB1 expression impact TAC dosing. When both genotypes are combined it may be possible to predict patients at high risk for subtherapeutic TAC dosing.
### Table 1 – Baseline Characteristics

<table>
<thead>
<tr>
<th>Baseline Demographics</th>
<th>Study Cohort (n = 31)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Recipient Age, years</td>
<td>46.0 ± 10.2</td>
</tr>
<tr>
<td>Donor Age, years</td>
<td>9.5 ± 8.1</td>
</tr>
<tr>
<td>Male Recipient Sex</td>
<td>12 (38.7%)</td>
</tr>
<tr>
<td>Recipient Ethnicity</td>
<td></td>
</tr>
<tr>
<td>White/European</td>
<td>18 (58.1%)</td>
</tr>
<tr>
<td>African-American</td>
<td>6 (19.4%)</td>
</tr>
<tr>
<td>Hispanic/Latino</td>
<td>6 (19.4%)</td>
</tr>
<tr>
<td>Asian/Pacific Island</td>
<td>1 (3.2%)</td>
</tr>
<tr>
<td>Recipient Weight, kg</td>
<td>61.7 ± 12.5</td>
</tr>
<tr>
<td>Transplant Type</td>
<td></td>
</tr>
<tr>
<td>Isolated Intestine</td>
<td>23 (74.2%)</td>
</tr>
<tr>
<td>Intestine-Kidney</td>
<td>5 (16.1%)</td>
</tr>
<tr>
<td>Intestine-Pancreas</td>
<td>3 (9.7%)</td>
</tr>
<tr>
<td>Baseline Serum Creatinine, mg/dl</td>
<td>1.2 ± 1.0</td>
</tr>
<tr>
<td>Baseline Total Bilirubin, mg/dL</td>
<td>1.1 ± 1.1</td>
</tr>
<tr>
<td>CYP3A5 Genotype</td>
<td></td>
</tr>
<tr>
<td>De/De</td>
<td>1 (3.2%)</td>
</tr>
<tr>
<td>De/Rf</td>
<td>9 (29.0%)</td>
</tr>
<tr>
<td>Dn/De</td>
<td>6 (19.4%)</td>
</tr>
<tr>
<td>Dn/Rfn</td>
<td>15 (48.4%)</td>
</tr>
<tr>
<td>ABCB1 Genotype</td>
<td></td>
</tr>
<tr>
<td>De/De</td>
<td>4 (12.9%)</td>
</tr>
<tr>
<td>De/Rf</td>
<td>4 (12.9%)</td>
</tr>
<tr>
<td>Dn/De</td>
<td>7 (22.6%)</td>
</tr>
<tr>
<td>Dn/Rfn</td>
<td>16 (51.6%)</td>
</tr>
<tr>
<td>Combined Analysis</td>
<td></td>
</tr>
<tr>
<td>Low Allele Score (0-1)</td>
<td>5 (16.1%)</td>
</tr>
<tr>
<td>Moderate Allele Score (2-3)</td>
<td>14 (45.2%)</td>
</tr>
<tr>
<td>High Allele Score (4-5)</td>
<td>12 (38.7%)</td>
</tr>
</tbody>
</table>
CIRTA 2019 Book of Abstracts - Posters of Distinction

P3A08 - Risks of the excluded bowel in patients with total intestinal Hirschsprung disease

Berénice Tulelli 1, Cécile Tabotet 2, Florence Lacaille 2, Célia Cretolle 1, Olivier Goulet 1, Louise Galmiche 3, Christophe Chardot 1, Carmen Capito 1
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3 Department of Pathology, Hôpital Necker-Enfants Malades, University of Paris Descartes, 29 rue de Sevres 75015 Paris, France

Introduction: Management of intestinal failure secondary to LSHD is based on parenteral nutrition (PN) associated to proximal jejunostomy. Early prophylactic subtotal enterectomy (SE) is not recommended since the excluded intestine allows growth of the abdominal cavity, which may be needed to host a graft in case of intestinal transplantation. This study evaluates the risk of enterocolitis linked to the excluded bowel in such patients.

Methods: The charts of all patients with LSHD requiring PN and enterostomies, managed from 1986 to 2017 in our institution, were reviewed. Subtotal enterectomy was performed, in our center, only after occurrence of complications due to the excluded bowel. Two patients underwent a subtotal enterectomy during the initial management and before referral to our center. They were excluded from this study.

Results: Twenty seven patients were included. Initial management resulted in totally diverting jejunostomy in 21 patients and a loop jejunostomy in one (median length from duodenum to stoma 30 cm [0-90 cm]).

Sixteen patients (59.2%) presented at least one episode of enterocolitis (EC). Median age at first EC was 2.2 years [0.3-7.1]. Fifteen underwent a subtotal enterectomy (4 as an emergency procedure for). Median age at SE was 3.1 years [1.3-7.4].

Three patients underwent a longitudinal seromyotomy with transfer of the jejunostomy downstream at initial management for one and at the time of subtotal enterectomy for two (9 months and 4 years).

Conclusion: Patients with LSHD carry a high risk of early complications linked to the excluded bowel. Since there is a paradigm shift regarding the indications of intestinal transplantation in patients with intestinal failure, these data represent a first step to better evaluate the benefit/ risk balance of preserving the excluded bowel for growth of the abdomen in view of eventual intestinal transplantation. Furthermore, the risks of repeated sepsis on long term liver function should also be assessed in order to discuss more rationally the better timing of enterectomy in this population having already PN and its associated risk of liver disease.

P3A09 - Physical activity, strength and fatigue in children with intestinal failure on parenteral nutrition

Stephanie So 1, 2, 3, Catherine Patterson 1, 2, 3, Zachary Betts 1, Christina Belza 1, 3, Yaron Avitzur 1, 4, Paul W. Wales 1
1 Group for Improvement of Intestinal Function and Treatment (GIFT), The Hospital for Sick Children Toronto, Canada
2 Department of Rehabilitation Services, The Hospital for Sick Children, Toronto, Canada
3 Transplant and Regenerative Medicine Centre, The Hospital for Sick Children, Toronto, Canada
4 Division of Gastroenterology, Hepatology and Nutrition, The Hospital for Sick Children, Toronto, Canada

Introduction: With improved survival of children with intestinal failure (IF), it is important to explore the subsequent impact on long-term physical function. This study evaluates functional outcomes of muscle strength, fatigue and physical activity (PA) and identifies clinical factors that may be predictive of impairment.

Methods: Cross-sectional study in children with IF on long term parenteral nutrition (PN) compared to healthy age and gender matched controls (HC’s). Assessments included: Bruininks-Oseretsky Test of Motor Proficiency-2 (Strength and Agility subtest) (BOT-2), Grip strength, Paediatric Quality of Life (PedsQL) Physical Function subscale and Multi-Dimensional Fatigue Module, and Benefits/Barriers to PA Questionnaire. PA was measured using an accelerometer. Medical data was collected by chart abstraction.

Results: Participants included 21 children with IF (34 male) at a median age of 8.33 (IQR 6.96, 10.04) years and 33 HC’s (20 males), 8.25 (6.67, 10.79) years. In those with IF, 21 (62%) were born prematurely (gestational age 35 (33, 38.5) weeks) and the most common diagnosis was gastroschisis (38%). Children received PN for 34 (12 88) hours/ day, with 18 (58%) dependent since infancy. Since birth, there was a median of 4 (1, 9) septic episodes and 7 (2, 15) in-patient hospitalizations.

Mean BOT-2 standard scores [population norm of 50, SD of 10] were 61.27 +/- 6.8 for HC’s and 45.67 +/- 9.1 for children with IF, with 6/21 (28%) below average (>1SD below mean). 13/21 (62%) children fell >SD below the mean norm for grip strength, compared to only 6/33 (18%) HC’s. Mean steps/ day in children with IF was 9842 +/- 4077 compared to HC’s with 13104 +/- 5416. There was a significant difference between groups in strength and agility (p<.00), grip strength (p=.005), and total steps/ day (p=.047). Medical variables significantly associated with BOT-2 scores are shown in Table 1.

Table 1

<table>
<thead>
<tr>
<th>Medical Factors</th>
<th>R-value*</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gestational Age</td>
<td>0.058</td>
<td>0.037</td>
</tr>
<tr>
<td>Birth Weight</td>
<td>0.068</td>
<td>0.010</td>
</tr>
<tr>
<td>Height 2 Score</td>
<td>0.026</td>
<td>0.105</td>
</tr>
<tr>
<td># of In-patient Hospitalizations/Years of Life</td>
<td>0.026</td>
<td>0.104</td>
</tr>
<tr>
<td>Length of Hospital Stay (Days)/Years of Life</td>
<td>-0.041</td>
<td>0.045</td>
</tr>
<tr>
<td># of Septic Events in first year of life</td>
<td>-0.402</td>
<td>0.035</td>
</tr>
<tr>
<td># of Septic Events/1000 PN Days Later</td>
<td>-0.325</td>
<td>0.005</td>
</tr>
</tbody>
</table>

Results of the PedsQL are shown in Figure 1
Parents report greater fatigue \((r = -0.538, p = 0.012)\) and poorer physical function \((r = -0.650, p = 0.001)\) in children with more hospitalizations. Barriers to PA in children with IF included “I am tired” and “I am worried about my line.”

**Conclusion:** Factors such as prematurity, decreased linear growth, hospitalization and septic events may have a significant impact on physical function outcomes in children with IF on PN. Ongoing development of medical and rehabilitation intervention strategies is vital to optimize outcomes.

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**P3A10 - Accumulated experience with Sirolimus in Pediatric Intestinal Transplantation**

Ane Andres 1, Francisco Hernandez-Oliveros 1, Alba Bueno 1, Alba Sanchez-Galan 1, Javier Serradilla 1, Esther Ramos 2, Alida Alcolea 2, Miriam Nova-Sanchez 2, Manuel Lopez-Santamaria 1


**Introduction:** The aim was to update our experience with Sirolimus (SRL) in intestinal transplanted (IT) pediatric patients.

**Patients and methods:** Between 1999 and 2018, 107 IT in 83 patients were performed at a median age of 38 months at transplant (range 6 m- 30 yo); 18 were retransplanted, 6 of them twice. Initial maintenance treatment protocol was Tacrolimus (TAC). Those patients who were partially or completely converted to SRL were retrospectively reviewed.

**Results:** Since 2008, SRL was used in 31 patients, after a median of 17 months since transplant (range 1 m- 6 years). The main indication was TAC toxicity in 16 (worsening of the renal function +/- hypertension in 13, hypertrophic myocardiopathy in 2, neuropathy in 1 and/or immunological complications in 17 (chronic rejection in 1, hemolytic anemia in 7, GVHD in 5, PTLD in 3, and neutropenia in 1). On the other hand, 10 patients had past medical history of rejection (6 of them moderate/severe), 9 had suffered GVHD, and 8 patients had been retransplanted prior to the conversion. Sirolimus was indicated alone in 20 patients when TAC was contraindicated and combined with TAC in 12 with higher risk of rejection, such as in retransplanted patients.

After a median follow-up of 77 months (3m-12years), the renal function improved in all patients along the follow-up (mean creatinine levels decreased from 3.50±0.52 mg/ dl to 1.14±0.34). We did not find more immunological complications after the conversion to SRL compared with those patients on TAC. Six patients died, due to the progression of chronic rejection (3 or GVHD (4), having appeared these complications before the conversion. However, 2 patients developed chronic rejection after the conversion, one of them has recently been retransplanted after 12 years since the first ISBT (10 years with SRL). The second one died 7 years after his third transplant, having taken SRL for the 3 last years. The other 25 are doing well with normal graft function. At the moment of the study, 47 of the global series are alive and 25 of them are on SRL.

**Conclusions:** Although TAC remains to be the maintenance treatment of choice in IT, half of our alive patients needed conversion to SRL along the follow-up. It seems safe and effective, although it does not totally prevent from immunological complications either. Accumulated experience is encouraging to widen its use in younger children, complex scenarios and after less time elapsed since transplant.
POSTERS OF DISTINCTION
FRIDAY JULY 5, 2019
SESSION 2
P38II - Analysis of immune cells draining from the abdominal cavity as a novel tool to study intestinal transplant immunobiology. Prospective multicenter study - INIGMA project.

Maria Virginia Gentilini 1,2, Albanis Parada Moncada 1,2, Melisa Pucci Moliners 1, Mariana Doeyo 1, Hector Solar 1, Carolina Rumbo 2, Mihai Oltean 4, Gustaf Herlenius 4, Laurens Ceulemans 5, Jacques Pirenne 5, Francisco Hernandez Oliveros 6, Javier Serradilla 6, Dominik Meier 1, Martin Rumbo 1, Gabriel Gondolesi 1

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2 Multiorgan Transplant Institute, Nutritional Support Unit, Intestinal Rehabilitation and Transplantation, Favaloro University Hospital. Buenos Aires, Argentina.
3 Immunology and Pathophysiology Studies Institute (IPSI-CONICET) School of Exact Sciences, National University of La Plata, Buenos Aires, Argentina.
4 Department of Surgery, Institute for Clinical Sciences, Sahlgrenska Academy, University of Gothenburg, Gothenburg, Sweden.
5 Department of Abdominal Transplantation Surgery, University Hospitals Leuven, Leuven, Belgium.
6 Pediatric Surgery Service, La Paz University Hospital, Madrid, Spain.

We proved that abdominal drainage fluid collected after intestinal transplant (ITx) contains mainly immune cells trafficking from the implanted intestine, and changes of immune cell composition, especially increase of neutrophils, correlated with the appearance of future clinical events (rejection, infection or other events). During the 34th ISBTS meeting, the development of a prospective multicenter study to analyze the role of performing daily analysis of drainage samples that might serve as a reliable tool for predicting clinical events, was proposed. We aim to present the preliminary results of the INIGMA (International Network for Intestinal Graft Monitoring and Analysis) project.

Material and Methods: This is a prospective study started in 2008 by Favaloro University Hospital, Argentina, and evolved as a multicenter study in 2015, including the University of Gothenburg, Sweden; University of Leuven, Belgium and La Paz University Hospital, Spain. The cell composition of the abdominal draining fluid and blood was analyzed during the first post-op days by differential cell counter and correlation with clinical follow-up was evaluated.

Results: 3 of the 4 centers enrolled 28 patients (pts) with complete biochemical and clinical information (Table 1). The cell composition differs from blood and draining fluid; independently of the immunosuppressive protocol used, blood samples obtained the day of the Tx showed an almost complete lymphopenia, while neutrophils were the main population (B/28 pts). In the drains, lymphocytes were the main population counted in non-complicated pts from post-op day 3 to 4 onwards (7/28 pts). When a new shift to a neutrophil dominant content is observed in the drainage, it anticipates the development of a clinical event in the peritoneal cavity (18/28 pts).

Table 2 summarizes the correlation between neutrophils and clinical events; Fischer test: p=0.002; Sensitivity=90%; Specificity=86%; PPV=94.7%, NPV=75%.

Conclusion: this study supports the concept that multicenter studies are feasible in our field. This analysis validates the findings of the original report, and provides further support to use this innovative approach to monitor the grafts and the transplanted pts. Cell counts from the drainage should be included as part of the daily evaluation of pts receiving an ITx, any shift from lymphocytic to neutrophilic dominance predicts the occurrence of a clinical events, requiring attention, early confirmation and treatment.
P3B12 - Spiral intestinal lengthening and tailoring (SILT) as a rescue procedure for SBS patients with difficult clinical situations.

Riccardo Coletta 1, Antonino Morabito 1,2
1 Paediatric Autologous Bowel Reconstruction and Rehabilitation Unit, Department of Paediatric Surgery, Meyer Children's Hospital, Florence, Italy
2 University of Florence

Introduction: Autologous gastrointestinal reconstructive (AGIR) procedures are essential component of intestinal rehabilitation program for patient with short bowel syndrome (SBS). Unfortunately, several patients develop postoperative bowel re-dilatation with loss of bowel adaptation. Spiral intestinal lengthening and tailoring (SILT) technique promises to regain intestinal physiology by tailoring the bowel even if there is no massive bowel dilatation. The aim of this study was to report our experience using SILT in SBS with difficult clinical situation.

Methods: Retrospective review of the single AGIR surgeon was performed from 2012 to date. Patients' demographics, pre- and post-procedure bowel length, surgical complications, and postoperative parenteral nutrition (PN) requirements were analyzed. Data were compared using independent samples, Mann-Whitney's U-test. Data were expressed in mean (IQR).

Results: 9 children with SBS underwent SILT between 2012 and 2017. Rescue SILT was performed in 4 patients with age at procedure of 58.5 months (45-143.3). 3 patients had previous STEP in history of SBS post gastroschisis while one had mild dilatation after AGIR due to long segment Hirschsprung. Preoperative small bowel length measured 66 cm (49.5-75.7) with a diameter of 7 cm (6-8). SILT allowed a median increase in length of 68.5% (p=0.34) and a significant tailoring of the dilated segment providing a reduction in diameter of 63% (p=0.02). No major complications related to SILT were encountered and none of the children required further surgical intervention following a follow-up of 6.5 months (4.5-39.6). Interestingly, a significant reduction of PN requirement at 4 months (p=0.02) associated with improve of liver function was reported.

Conclusion: In this study SILT was reported to have no peri-operative complication and to have good results in complex SBS patients. This procedure offers a safe alternative for SBS children after the failure of conventional AGIR procedures.

Conclusions
- CMV serostatus for D/R is an important consideration in intestine transplantation but mismatch is not associated with increased graft or patient morbidity.
- Tissue-invasive CMV disease is significantly more frequent in recipients that receive organs from CMV + donors but can be managed with monitoring and current treatment.
Glucocorticoid treatment improves enteral feeding tolerance in pediatric short bowel syndrome patients with chronic intestinal inflammatory changes

Fangfang Wang, Brandi Gerhardt, Sarah Iwansky, David Mercer, Ruben Quiros-Tejeira

University of Nebraska Medical Center, Omaha, Nebraska, United States

Introduction: Studies suggest that chronic local and systemic inflammation are related to short bowel syndrome (SBS). We believe that glucocorticoids may reduce inflammation and improve enteral tolerance in pediatric SBS patients. A group of patients from our intestinal rehabilitation program developed chronic intestinal inflammation under biopsy while struggling weaning off parenteral nutrition (PN). We treated them with oral prednisolone taper, maintenance budesonide and sulfasalazine. We will describe the treatment outcome.

Method: Our study included 15 patients whose lab values, pathology results and clinic notes were reviewed, and excluded patients who took steroids for post-transplant immunosuppression or severe food allergy to maintain the homogenous of our patient cohort.

Results: The most common reason for SBS was gastroschisis. The small bowel length range was 20-108 cm. Thirteen patients had more than ½ colon remaining. The median age of starting steroid therapy was 3.3 years. Twelve patients had decreased PN calorie count or normalized follow-up histology. Six patients were able to wean off PN completely with median treatment duration of 5 months, five of which remained on maintenance budesonide for significant period of time (median: 7.5 months), the other one had steroids stopped 5 months before being off PN due to clinical improvement and concern for medication noncompliance. Six out of fifteen children had significant eosinophils in their initial biopsy, five were able to wean off PN while one had significant symptom improvement (GI bleeding stopped). Two patients were not on steroids continuously, as they resumed steroids months later due to histological recurrence of the chronic inflammatory process. Three patients were still not able to be cut down PN calorie. Two patients’ intestinal biopsies findings were still consistent with chronic inflammatory process.

Conclusion: For SBS children with histologically confirmed chronic intestinal inflammatory changes, steroid therapy may help to promote enteral feeding tolerance and wean off PN. Patients may respond better if their initial histology has chronic intestinal inflammation with significant eosinophilic infiltrate. Patients may need to remain on maintenance steroids for a period of time even if they were able to wean off PN or significantly reduce PN calories as they may experience regression on enteral tolerance or evidence of GI bleeding as weaning off steroids is attempted.
Nine children were included in the study. Mean age at PN resumption was 10 years and 10 months. PN discontinuation lasted six and a half years (mean). All patients presented with stunting which was more important on height than on weight (loss of 1.5 SD and 1.3 SD respectively). All patients were hyperphagic, five of them received tube feeding on top of oral feeding, with mean ingested calories of 2336 kcalories/ day, with a ratio of ingested calories over resting energy expenditure of 205%. Mean number of stools was 4.4 per day, with a mean stool weight of 300g/ day (52g/ kg/ d). One patient presented with recurrent episodes of D-lactic acidosis. Four patients had bicarbonates concentration under 23mmol/ l. Mean total absorption rate was 69%. After PN start, the intestinal symptoms resolved, weight increased by 14 SD and height by 0.8 SD at 12 months follow-up. Biological parameters all improved. Mean citrulline plasma levels remained stable (26mmol/ l at PN re-start - 24mmol/ l at 12 months follow-up).

Conclusion: Overload gut syndrome is a rare complication of SBS in children. It occurs in children with high enteral intake and low absorption rate: the remnant intestine cannot face the calorie load and becomes overwhelmed. Symptoms should be recognized early in order to resume PN in time to avoid consequences on final stature.
P3B17 - Complex relation between timing of acute rejection, severity, and graft loss in isolated intestinal transplantation

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Introduction: Acute rejection (AR) remains a major determinant of morbidity and mortality after ITX. We studied patterns of AR and graft loss to understand potential loci for intervention.

Methods: We reviewed all ITX without concomitant liver graft between 2011 and 2018 at a single-center using a prospectively maintained database. AR, patient and graft survival were outcomes of interest.

Results: We analyzed 72 ITX without liver replacement. Twenty-eight grafts had at least one episode of AR (39%). Majority of AR occurred during first 6 months (57%) post-ITX and remainder occurred sporadically during the entire post-ITX period. Of 35 total episodes of AR among 28 grafts, 13 were mild (37%), 4 moderate, (11%), and 18 (51%) severe AR based on histological grades. Patient survival did not show a difference between ITX with and without AR (p=0.750), though, AR was the most common cause of mortality (n=6, 8%). Censoring death with functioning graft (n=12), AR was the most common cause of graft loss (n=11, 15%). Mild AR tended to develop early (< 12 months) post-ITX. Severe AR was observed evenly during the entire post-ITX period (Figure 1).

There was no graft loss or mortality related to mild or moderate AR. There were 11 graft losses out of 28 episodes of severe AR (graft salvage rate of 39%). Graft enterectomy was performed in 6 patients due to refractory severe AR. Among those, 5 survived and 3 went on to successful re-ITX. Five patients were not candidates for graft enterectomy due to profound sepsis and all died. Early severe AR, occurring < 6 months after ITX, had a poor treatment response (7 severe AR with 6 graft losses). High panel reactive antibody (PRA), positive donor specific anti-HLA antibody (DSA) and cross match appeared related to development of AR. However, there were also 23 episodes of AR in 18 grafts (33%), which occurred despite low PRA, negative DSA and negative cross match; of these 14 were severe AR with 7 graft losses (Figure 2).

Conclusion: Our analysis supports previous reports that severe AR is a major cause for graft loss and mortality in ITX. Presence of PRA, DSA and a positive cross match seem related to risk of developing AR. Of note, patients with no apparent heightened immunological risk factors also develop severe AR with a high frequency of graft loss. Mild to moderate AR has no apparent impact on outcome.
INT-767 - a novel dual Farnesoid-X Receptor (FXR) and Takeda G-protein-coupled Receptor-5 (TGR5) agonist attenuates intestinal ischemia reperfusion injury

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2 Abdominal Transplant Surgery, University Hospitals Leuven, Belgium
3 Translational Research Center for Gastrointestinal Disorders (TARGID), Department Chronic Diseases, Metabolism and Ageing (CHROMETA), KU Leuven, Belgium
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Introduction: Ischemia reperfusion injury (IRI) occurs inevitably during intestinal transplantation and after intestinal infarction. The intestine is especially susceptible to IRI which leads to loss of villi, resulting in systemic translocation contributing to poorer outcomes. The Farnesoid-X receptor (FXR), is a member of the nuclear receptor family. TGR5 is a G-protein-coupled bile activated receptor. Both are abundantly expressed in the gastrointestinal tract. In pre-clinical models, they have shown to reduce inflammation and improve epithelial permeability when administered before ischemia. The aim of our study was to test the effect of a dual FXR/TGR5 agonist as treatment of intestinal IRI, administered intravenously after onset of ischemia.

Material and Methods: In a validated rat model (Sprague-Dawley, male, 300g) of intestinal IRI (laparotomy and clamping of superior mesenteric artery), 3 groups (n=6/group) were investigated: i/ Sham (only laparotomy); ii/ Ischemia 60min + reperfusion 60min (IRI) + intravenous vehicle; iii/ Ischemia 60min + reperfusion 60min + intravenous FXR/TGR5-agonist (IRI+FXR/TGR5). For each group, 10 additional animals were included for a 7-day survival analysis. FXR/TGR5-agonist INT-767 (Intercept Pharma, USA) or vehicle only was administered intravenously in a single dose at 10 mg/kg, 15 minutes after start of ischemia. Analyzed endpoints: i/ Histology: Park/Chiu score and villus length; ii/ intestinal barrier function (transepithelial electrical resistance (TEER) and FD20 permeability measurements in Ussing chambers); iii/ Inflammatory cytokines: IL-6 (ELISA), IL-1β and TNFα (qPCR); and iv/ Anti-inflammatory cytokines: IL-10, IL-13 (qPCR).

Results: IRI led to pronounced damage resulting in high Park/Chiu scores, increased intestinal permeability and systemic inflammation. Dual FXR/TGR5 treatment dramatically improved intestinal histology (Figure) and all other parameters. Survival was substantially improved after treatment (P < 0.05). Results are summarized in the table.

Conclusion: We demonstrated that intravenous treatment with a dual FXR/TGR5 agonist (INT-767) after onset of ischemia significantly decreased intestinal damage caused by IRI. These results show that FXR and TGR5 receptors are promising targets for intestinal graft protection. The ability to administer this substance intravenously greatly enhances the potential applicability for the frequent pathology of intestinal infarction as well as for transplantation.
CONCLUSION: Duodenal mucosa of PN-dependent SBS children showed no structural or molecular signs of adaptive hyperplasia, but was characterized by molecular signature of increased inflammation and disturbed barrier function. These findings have important implications regarding SBS pathophysiology in humans.
POSTERS PRESENTED ON WEDNESDAY JULY 3, 2019
**P101 - Post-transplant vascular complications in isolated intestine and multivisceral transplant patients**

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Indiana University School of Medicine

**Introduction**
Vascular complications such as aortic pseudoaneurysm, AV fistula formation, vascular graft leak, vascular graft thrombosis, and venous outflow obstruction can result in significant morbidity and mortality in abdominal organ transplant patients. In this study, we review our experience with such complications in isolated intestine and multivisceral transplant patients.

**Methods**
All records for isolated intestine, multivisceral, and modified multivisceral transplants over a 15-year period at a single center were reviewed. All cases of aortic pseudoaneurysm, AV fistula formation, vascular graft leak, vascular graft thrombosis, and venous outflow obstruction were included.

**Results**
Of 263 transplants, 16 major post-transplant vascular complications were identified (6%). There were five cases of venous outflow obstruction, three of which required revision of the venous anastomosis. One case resulted in colonic necrosis necessitating colectomy, and another required venotomy and thrombectomy. Four patients developed vascular graft thrombosis, one of which was found to have splenic artery thrombosis for which a distal pancreatectomy was eventually required. The second patient developed distal arterial thrombi of the intestinal graft resulting in small bowel necrosis that required resection of the distal ileum. The third patient developed hepatic artery thrombosis which was successfully treated with intra-arterial tPA infusion and anastomotic revision; this patient also had poor portal flow that was successfully re-established with anastomotic revision. The fourth patient was found to have minimal flow in the aortic jump graft with diffuse necrosis of the transplanted organs and died shortly thereafter from complications of severe acidosis. Four cases of vascular graft leak were identified, all of which involved the aortic graft and resulted in exsanguination and death. There were two instances of aortic pseudoaneurysm, both of which were successfully treated with stent graft placement. One case of AV fistula formation was identified that involved the hepatic artery and portal vein and was successfully treated with coil embolization of the hepatic artery.

**Conclusion**
Vascular complications can result in significant morbidity and mortality in intestine and multivisceral transplant patients. The incidence of vascular complications in this cohort was 6%.

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**P102 - Tacrolimus induced optic neuropathy after multivisceral transplantation**

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2 Abdominal Transplant Surgery, University Hospitals Leuven, Leuven, Belgium  
3 Ophthalmology Department, University Hospitals Leuven, Belgium  
4 Radiology Department, University Hospitals Leuven, Belgium  
5 Gastroenterology and Hepatology, University Hospitals Leuven, Belgium

**Introduction**
Tacrolimus induced optic neuropathy (TION) is a rare condition seen in transplant patients leading to severe vision loss caused by damage to the optic pathway. The underlying pathophysiology is thought to be a combination of ischemic damage due to vasoconstriction of the cerebral microvasculature and direct neurotoxicity.

**Method**
We describe a case of a 51-year old male, combined multivisceral and renal transplant recipient who developed severe, bilateral TION 3 years after transplantation. Furthermore, a literature research was performed for all published cases describing TION after organ transplantation.

**Results**
Optic tract inflammation was clearly detected on MRI (Figure). Treatment with intravenous corticosteroids and immunoglobulins was started. Tacrolimus was reduced but not withdrawn completely to avoid rejection, especially of the intestinal component of the graft. Everolimus was associated to maintain sufficient immunosuppression. After three months, vision had recovered completely. The patient experienced no signs of rejection in any transplanted organ during this period and organ function remained stable. Seven other reports in various organs were found in literature (Table). In most, tacrolimus was discontinued completely and outcomes were poor.

**Conclusion**
Our report demonstrates the importance of swift treatment to reverse optic tract inflammation and highlights the possibility to add everolimus to the immunosuppressive regimen to allow safe reduction of tacrolimus exposure in intestinal transplant patients. By contrast, results from literature show sporadic use of anti-inflammatory medication and poor long-term vision outcomes, often related to delayed diagnosis and treatment (Table).
**P103 - Intravenous glucose nocturnal infusion reduces growth hormone secretion in children on total parenteral nutrition**

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1 Department of pediatric gastroenterology, hepatology and nutrition Hospital Necker-Enfants Malades
2 Department of pediatric endocrinology Hospital Necker-Enfants Malades

**Introduction:** Most children with intestinal failure (IF) benefit from home parenteral nutrition (HPN). It allows a familial life with normal children activities such as going to school, sports, and being at home. In order to do so, PN is provided with cyclic infusion at night. Children with IF on HPN should have normal growth. However, some children grow under their genetic target height and present with decreased height velocity.

**Methods:** We report the case of two children on HPN who presented with decreased height velocity while their weight gain was normal. After ruling out usual etiology (insufficient protein intake, acidosis, sodium deficiency, IFALD...), growth hormone (GH) deficiency was considered. Extensive testings were performed such as IgF1 levels, GH secretion stimulation test and nocturnal GH secretion. PN dependency was assessed using the PN/REE index (parenteral nutrition non-protein energy intake divided by resting energy expenditure calculated using Schofield equation, expressed in percentage).

**Results:** These two children needed extremely high PN caloric intake to achieve a normal weight gain with a PN dependency index of 163% and 150%.

Table 1 shows their clinical characteristics.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yrs)</th>
<th>Height (SD)</th>
<th>Weight (SD)</th>
<th>NCG (mmol/L)</th>
<th>GH Stim (ng/mL)</th>
<th>GH peak (ng/mL)</th>
<th>PN/REE</th>
<th>Full Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11</td>
<td>-0.5</td>
<td>1.8</td>
<td>1.1</td>
<td>0.05</td>
<td>1.1</td>
<td>Full</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>12</td>
<td>-3.5</td>
<td>-3.2</td>
<td>2.0</td>
<td>0.15</td>
<td>2.0</td>
<td>Full</td>
<td></td>
</tr>
</tbody>
</table>

These two children showed a normal GH peak under stimulation, but a low nocturnal GH secretion meaning they were able to produce GH under stimulation, but their nocturnal physiological secretion was inhibited. Their mean glycemia during PN perfusion was high compared to fasting glycemia which should normally be observed at night in children. Hyperglycemia is a well known inhibitor of GH secretion.

**Conclusion:** Children with very high PN dependency are at risk of developing reduced height. In some children, GH secretion seems to be inhibited by nocturnal high glucose infusion. A longer PN infusion time can be proposed and/or a decrease in glucose intake with an increase in lipid intake. Also, growth hormone supplementation might be discussed and should be evaluated in this setting.
Anastomotic Ulcers in Children with Short Bowel Syndrome: A Single Intestinal Rehabilitation Center Experience

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University of Nebraska Medical Center

Anastomotic ulcers can present as a significant problem in children with short bowel syndrome (SBS). They are typically a late complication occurring years after creation of an ileocolonic anastomosis. As the mucosa becomes inflamed and ulcers form complications of GI blood loss (gross or occult), iron deficiency anemia, chronic diarrhea, and feeding intolerance may occur. The purpose for this study is to identify a most common etiology of anastomotic ulcers and best treatment options in patients with SBS.

A retrospective chart review of intestinal rehabilitation patients was completed, and was limited to pediatric patients (<19 yo) with diagnoses of SBS, gastrointestinal bleeding, and endoscopic evidence of ulcers during a 10 year period of time (2006-2016). We collected basic data, including the age, sex, diagnoses, past medical and surgical history, post surgical anatomy, endoscopic findings, small bowel aspiration culture results, laboratory findings, and GI bleeding management and response. Data were analyzed using descriptive statistics and processed to find trends for clinical interpretation.

14 patients met inclusion criteria. The average intestinal length was 40 cm (12/14 ≤ 70 cm), and remaining colon median was 50% (9/14 ≤ 50%). Serial Transverse Enteroplasty (STEP) procedures performed on 9/14. Average age of anastomosis at first step was 2.9 yrs. Of the patients that had a STEP procedure, the median number of procedures was 1 (range 1-5). The median age of surgical anastomosis where ulcer was detected was approximately 3 years. On reviewed laboratory data, 12/14 patients required transfusions. Of the 11 that required a transfusion, the median number of transfusions was 3. The median number of procedures was 6 (range of 1 -19). When treated for SBBO 3/7 showed improvement of ulcers with antibiotics alone. Stopping enteral iron therapy was successful in 2 cases. Surgical intervention was required in 6/14 cases.

Anastomotic ulcers are viewed as multifactorial problem. Factors that seemed to contribute include: past medical and surgical history, ischemic changes, medication exposure (ferrous sulfate), exposure to colonic bacteria and bile acids malabsorption. The one universal factor was the absence of an ileocecal valve. A systematic approach started with antibiotics, cholestyramine and/or steroids should be utilized and consider avoidance of enteral iron therapy. Early consideration of surgical intervention may be necessary.

<table>
<thead>
<tr>
<th>Patient</th>
<th>SB length (cm)</th>
<th>Colon (%)</th>
<th>ICV</th>
<th>STEP</th>
<th>No. of procedures</th>
<th>Resolution of ulcers/symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>32</td>
<td>25</td>
<td>No</td>
<td>Yes</td>
<td>5</td>
<td>Improved</td>
</tr>
<tr>
<td>2</td>
<td>27</td>
<td>75</td>
<td>No</td>
<td>No</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>60</td>
<td>10</td>
<td>No</td>
<td>Yes</td>
<td>1</td>
<td>Improved</td>
</tr>
<tr>
<td>4</td>
<td>70</td>
<td>20</td>
<td>No</td>
<td>Yes</td>
<td>1</td>
<td>Improved</td>
</tr>
<tr>
<td>5</td>
<td>55</td>
<td>50</td>
<td>No</td>
<td>Yes</td>
<td>2</td>
<td>Improved</td>
</tr>
<tr>
<td>6</td>
<td>140</td>
<td>33</td>
<td>Yes</td>
<td>No</td>
<td>1</td>
<td>None</td>
</tr>
<tr>
<td>7</td>
<td>23</td>
<td>50</td>
<td>No</td>
<td>No</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>8</td>
<td>20</td>
<td>67</td>
<td>No</td>
<td>Yes</td>
<td>2</td>
<td>Improved</td>
</tr>
<tr>
<td>9</td>
<td>100</td>
<td>75</td>
<td>No</td>
<td>No</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>10</td>
<td>85</td>
<td>83</td>
<td>No</td>
<td>Yes</td>
<td>1</td>
<td>Improved</td>
</tr>
<tr>
<td>11</td>
<td>65</td>
<td>29</td>
<td>No</td>
<td>Yes</td>
<td>2</td>
<td>Improved</td>
</tr>
<tr>
<td>12</td>
<td>40</td>
<td>67</td>
<td>No</td>
<td>No</td>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>13</td>
<td>54</td>
<td>67</td>
<td>No</td>
<td>Yes</td>
<td>1</td>
<td>Improved</td>
</tr>
<tr>
<td>14</td>
<td>50</td>
<td>33</td>
<td>No</td>
<td>No</td>
<td>0</td>
<td>None</td>
</tr>
</tbody>
</table>

Table 1: Intestinal anatomy: The median intestinal length was 40 cm, and the median percentage of remaining colon was contained to be 50%. All patients included in this study had an ileocecal anastomosis, and as a result there were none with preserved ICV.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Failed management</th>
<th>Duration of time</th>
<th>No. of procedures</th>
<th>Resolution of ulcers/symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Steroids and antibiotics resection of anastomosis, repair of enterotomy</td>
<td>2009-2014</td>
<td>16</td>
<td>Improved</td>
</tr>
<tr>
<td>2</td>
<td>Bed rest and sodium hydrion</td>
<td>2016-2018</td>
<td>1</td>
<td>Improved</td>
</tr>
<tr>
<td>3</td>
<td>Cholestyramine and antibiotics</td>
<td>2010-2015</td>
<td>7</td>
<td>Improved</td>
</tr>
<tr>
<td>4</td>
<td>Paroxetine and resection (2011) of mesoeum</td>
<td>2010-2013</td>
<td>8</td>
<td>Surgical resection</td>
</tr>
<tr>
<td>5</td>
<td>Trials of antibiotics and enteral nutrition for SIBO</td>
<td>2010-2016</td>
<td>3</td>
<td>Surgical resection</td>
</tr>
<tr>
<td>6</td>
<td>Sodium lactate, metronidazole, and mersalin</td>
<td>2006-2015</td>
<td>1</td>
<td>Improved</td>
</tr>
<tr>
<td>7</td>
<td>Cholestyramine</td>
<td>2016</td>
<td>2</td>
<td>SIBO, antibiotics + E. coli (tissue culture)</td>
</tr>
<tr>
<td>8</td>
<td>Antibiotics for SBBO and cholestyramine without improvement</td>
<td>2010-2015</td>
<td>3</td>
<td>Improved</td>
</tr>
<tr>
<td>9</td>
<td>None</td>
<td>2010</td>
<td>4</td>
<td>Surgical resection</td>
</tr>
<tr>
<td>10</td>
<td>Trials of antibiotics, cholestyramine, rectal relaxation of enterotomy, and nutrition</td>
<td>2007-2017</td>
<td>4</td>
<td>Surgical resection</td>
</tr>
<tr>
<td>11</td>
<td>None, primary surgical problems</td>
<td>2016-2017</td>
<td>3</td>
<td>Surgical resection</td>
</tr>
<tr>
<td>12</td>
<td>None</td>
<td>2017</td>
<td>3</td>
<td>None</td>
</tr>
<tr>
<td>13</td>
<td>Enterostomy, sodium lactate, Mesalamine, and Influenza</td>
<td>2008-2017</td>
<td>1</td>
<td>Surgical resection</td>
</tr>
<tr>
<td>14</td>
<td>None</td>
<td>2010</td>
<td>3</td>
<td>None</td>
</tr>
</tbody>
</table>

Table 2: Comparison of management options, period of time, ulcer bleeding was treated, and the ultimate resolution of ulcers. Estimated number of procedures required: diagnosis and treatment bleeding and ulcer tissue culture of SIBO was 1 with average of 1 procedure per patient. Surgical resection was most successful in management of ulcers in 14 cases.
Acute cellular rejection (ACR) is leading cause of graft loss and death in intestinal transplant (ITx) patients. ACR promotes intestinal injury, disruption of the mucosal barrier, organ dysfunction and bacterial translocation. Epithelial regeneration is critical to reverse the situation. The intestinal stem cell (ISC) provides signals supporting normal epithelial maintenance. It has been shown that innate lymphoid cells 3 (ILC3), potent producers of IL-22 after intestinal injury, increase proliferation and expansion of the ISC in an IL-22-dependent fashion. Therefore, we aimed to evaluate the immunological status during the ACR focusing on the axis ILC3/IL-22/IL22R/ISC. To do this, lamina propria cells of biopsies were isolated from ITx [Non-rejection (NR)=17; Mild rejection (MR)=7; Moderate rejection (MoR)=6 and non-transplant patients (NITx)=7. Enrichment of ILC3s and CD4 population was done using MACS technology. ILC3 were determined by flow cytometry. CD4 T cells were isolated and the expression levels of functional markers of Th1 (Tbet, IFN-γ), Th2 (GATA3, IL-13), Th22/17 (RORC, IL-22, IL-17A), Tregs (Foxp3, TGF-β) were evaluated by qPCR. Total levels of IL-22 in biopsies [NR=5 and Moderate rejection (MoR)=6] were measured by qPCR. ILC3, ISC IL-22R+ were detected by immunohistochemical staining [NR=3; MR=2, MoR=2, SR=1]. The results showed that during ACR the expression of Tbet, GATA3, RORC, Foxp3 were significantly decreased (P=0.05). Although the total percentage of ILC3s was not impaired (P=0.47), a reduction of ILC3 NCR+ NCR− ratio was observed. ISC IL-22R number was similar in all groups studied (P=0.28) while IL-22R expression showed a trend to be increased (P=0.06). Levels of IL-22 in ITxMoR were lower than in NRITx patients with normal biopsies (P=0.063) and NITx biopsies. In conclusion, during rejection CD4 population (TH1, TH2, TH22/22, Tregs) and ILC3/IL-22/IL22R axis are affected. In this context ISC number not only is unaffected, but also is able to increase IL-22R expression. Finally, the variation in the ILC3 NCR− NCR+ ratio observed during ACR could explained, in part, the impairment in the epithelial regeneration observed during this process. Thus, our findings reveal that IL-22 can potentially be used as a new therapeutic approach to be used in conjunction with immunosuppressants in order to promote mucosal regeneration. Due to the small number of patients evaluated, this preliminary study is currently being extended with a larger population.
# P107 - The Effects of Minimizing Phlebotomy Blood Volume on Anemia in Pediatric Home Parenteral Nutrition Patients

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2. Loma Linda University Children’s Hospital, Loma Linda, CA, United States of America

**Background & Objectives**

Anemia is a common morbidity in pediatric patients who require home parenteral nutrition (HPN). One factor that increases the risk for anemia in this patient population is frequent phlebotomy as part of routine monitoring. At Loma Linda University Children’s Hospital (LLUCH), the majority of pediatric patients on HPN have monthly labs drawn as part of the standard monitoring protocol. In June 2016, LLUCH began using micro-tubes for routine blood sampling. Traditional phlebotomy tubes require 15-20 mL of blood. Micro-tubes require less than 2 mL for the same tests. The purpose of this study is to understand the effects of phlebotomy blood volume minimization strategies in reducing anemia in our pediatric patients requiring HPN.

**Methods**

This study was IRB-approved and retrospective in design. Since June 2016, the HPN program at LLUCH began using micro-tubes for routine laboratory monitoring. Standard clinical data (age, gender, weight) and laboratory data (CBC, ALT, bilirubin, albumin) were collected prior to June 2016 and after September 2016. The primary outcome of this study was to compare the change in hemoglobin concentrations before and after the use of micro-tubes. The secondary outcome was to estimate the prevalence of anemia for age before and after use of micro-tubes for phlebotomy. Statistical analyses were performed including standard descriptive analysis, t-tests, and bivariate analysis. P-value of <0.05 was considered statistically significant. The sensitivity and specificity of red cell mean corpuscular volume (MCV) and red cell distribution width (RDW) in detecting anemia for ages were also calculated.

**Results**

N = Thirty-eight subjects. Mean age = 6.1 years (SD = 4.2y). Prevalence of anemia for age before micro-tube implementation = 22%

- Prevalence of anemia for age after micro-tube implementation = 34%
- Mean difference in the hemoglobin of before and after implementation = -0.62 g/dL (CI: -0.21 to 1.02; p=0.004)

Secondary bivariate analysis:

- No correlation between anemia for age and the following factors: age, gender, weight, ALT, and albumin level.

**Conclusions**

Strategies such as micro-tubes to reduce the volume of routine blood draws in pediatric patients requiring chronic HPN may reduce the prevalence of anemia and improve hemoglobin levels. Age, weight, and liver enzyme levels were not found to be independent risk factors for the presence of anemia for age.

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# P108 - Risk Factors for Copper Deficiency in Children with Intestinal Failure Receiving Long-Term Parenteral Nutrition

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**Introduction:** Copper is necessary for normal function of hematopoietic, cardiovascular, connective/skeletal and the central nervous systems. A >50% prevalence of copper deficiency has been reported in retrospective case series of children with intestinal failure (IF) during transition from parenteral to enteral nutrition (Yang CJ et al, 2014; Namjoshi SS et al, 2017). We sought to determine the frequency and factors associated with copper deficiency in children with IF who had their copper plasma levels monitored according to a standard protocol.

**Methods:** Prospective cohort study in children with IF followed-up at an intestinal rehabilitation center between July 2015 and November 2017 and who were receiving home parenteral nutrition (PN). Patient’s micronutrient status was routinely monitored at 3-month intervals or once a month when deficiency was detected. Copper was given at a standard dose of 20 mcg/kg as part of a trace-element solution, and was omitted in the PN if the patient developed intestinal failure liver associated disease. The outcome variable was copper plasma level during the follow-up period. The effect of the exposure variables (length of time on PN, prematurity, serum direct bilirubin levels, C-reactive protein, length of remnant small bowel and ostomy) on the outcome was analyzed by generalized estimating equations.

**Results:** Thirteen patients aged 34.2 months (IQR: 25.3; 41.1) were included; median time on PN was 26.4 months (IQR: 5.2 to 32.9). An average of 7 (range 2 to 3) copper measurements/patient were performed; 53.8% of patients had at least 1 copper measurement below normal during the follow-up. Eight patients who had cholestasis had trace elements of PN discontinued for 4 months (IQR: 16 to 22); from June 2017 these patients began to receive copper solution separately in doses to achieve basal requirements or to correct deficiency. Direct bilirubin levels (β coeff. -5.9, 95% CI: -9.0 to -2.7, p=0.04), time on PN without copper (β coeff. -1.7, 95% CI: -3.2; 0.2, p=0.003) and ostomy (β coeff. -20.3, 95% CI: -37.6; -2.9, p=0.02) were associated with decreased copper levels in multivariable analysis. The figure shows adjusted predictions and marginal effects of the length of time without copper on copper serum levels.

**Conclusion:** Direct bilirubin levels, length of time on PN without copper and ostomy are independently associated with the high frequency of...
decreased copper plasma levels in patients with IF receiving long term PN.
P109 - Donor's graft ex vivo T cell depletion with fludarabine reduces GvHD signs and improves survival after intestine transplantation

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Introduction: Intestine passenger T leukocytes are responsible of graft versus host disease (GvHD) in intestine transplantation (ITx). These lymphocytes are known to have inferior tolerogenic qualities compared with leukocytes in the liver, other solid organs and bone marrow. Fludarabine is a routinely used anti neoplasic agent with high cytotoxicity against T cells. We hypothesized that ex vivo fludarabine treatment of the bowel graft could diminish the risk of GvHD and improve post Tx overall survival.

Methods: We performed isolated heterotopic ITx from Lewis (LEW) to Brown Norway (BN) rat strains. The grafts of one of the experimental groups were imbied and sealed in Celsior preserving solution with 1000 µM fludarabine during surgery (1h), before its implantation into recipient animals.

We compared a group of untreated (n = 7) vs a group of fludarabine-treated bowel recipients (n = 5). The mixed hematopoietic chimerism was determined by flow cytometry using strain-specific anti HLA antibodies. Clinical signs of GvHD or as well as post-Tx overall survival were also monitored.

Results: One hour fludarabine treatment of the bowel grafts induced specific apoptosis of its passenger T cells at concentrations from 100 µM while no histological signs of intestinal tissue alterations were observed after 1000 µM fludarabine treatment.

After heterotopic LEW -> BN ITx, untreated intestine recipients showed GvHD signs from the fourth day post-Tx (n = 7). These symptoms include: rash (n = 4), weight loss (n = 3), piloerection (n = 2) and diarrhea (n = 1). The chimerism or percentage of donor’s lymphocytes in the recipient rat, determined in peripheral blood, reached 6.61% (range 1.7-10.8%) at day 3 post-Tx and 2.56% (range 0-9.8%) at day 7 post-Tx (Fig. 1).

Rats transplanted with a fludarabine-conditioned intestine showed statistically significant later and milder clinical signs of GvHD. Additionally, fludarabine treatment reduced total donor cells chimerism at day 7 and the percentage of chimeric T cells at days 3 and 7 post-Tx (Fig. 2).

Both experimental groups died showing clinical signs of graft rejection. Untreated bowel grafts recipients died within 9.2 days ±0.3 days, while fludarabine-treated graft recipients showed prolonged survival (13.5 days ±0.3 days).

Conclusions: Graft immunosupresion with fludarabine during surgical procedure protects bowel recipients of GvHD risk and improves post-Tx overall survival.
**P10 - Mucosal Repair After Small Bowel Transplantation In The Rat: be quick or be dead**

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**Introduction:** Graft cold storage and subsequent reperfusion during small bowel transplantation result in various degrees of mucosal injury ranging from mild edema to extensive mucosal loss. Mucosal barrier impairment favors bacterial translocation and fluid loss and raises nutritional challenges. Moreover, injured enterocytes can produce and release proinflammatory mediators and upregulate various epitopes towards an inflammatory phenotype. We studied the process of mucosal injury and repair during the early period after intestinal transplantation from a histological and molecular standpoint.

**Methods:** Three months old, Sprague Dawley male rats were used as donors and recipients. Donor intestines were perfused and stored in saline for 3 hours, then transplanted heterotopically using microvascular anastomoses. Small bowel graft segments were obtained after preservation, and at 20 minutes, 12 hours and 24 hours after reperfusion. Histology studies (Chiu score, Goblet cell count, morphometry, immunohistochemistry) and ddPCR for tight junctions (tricellulin, claudin-3), apoptosis (Bax, Bcl-2) and inflammation (IL-6, ICAM-1, TLR-4, TLR-9) were performed.

**Results:** Cold storage lead to extensive epithelial detachment (corresponding to Chiu grade 3) and reperfusion lead to extensive villus loss (about 50% of the initial villus length, Chiu grade 5). Goblet cells showed a significant reduction (p<0.01). All these parameters ceased to differ significantly compared to normal intestines after 24 hours of reperfusion. However, the villi appeared shorter and broader than in normal intestines and total mucosal volume was reduced. Bax, Bcl-2, IL-6, ICAM-1 and TLR4 mRNA levels were lower after 24 hours compared with immediately after reperfusion. mRNAs for tight junction proteins tricellulin and claudin-3 remained lower than in normal intestines.

**Discussion:** The current data suggest that early mucosal recovery after intestinal transplantation is mainly due to cell migration and lamina propria remodeling rather than enterocyte proliferation. This rapid phenomenon seems to be accompanied by a local downregulation of the inflammatory response. The very rapid recovery of the rat intestine following moderate/severe reperfusion injury needs to be considered when designing intestinal transplant experiments and choosing sampling and end points.

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**P11 - A Unique Presentation of Pediatric Intestinal Failure: Familial chronic intestinal pseudo-obstruction occurring with diffuse intracranial vasculopathy - a systemic smooth muscle disorder**

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**Introduction:** We present a unique case of familial intestinal failure secondary to chronic intestinal pseudo-obstruction (CIFO). Our patient is a 7-year-old male with longstanding feeding intolerance secondary to distension and vomiting that has required TPN. Additionally, he has a severe intracranial vasculopathy and a history of recurrent strokes. He had a sister who required TPN due to nonobstructive feeding intolerance, who passed away of complications related to stroke. While this association has been described in ACTA2 gene mutation disorders affecting systemic smooth muscle, our patients do not have an identified ACTA2 mutation. Therefore, we suspect a novel genetic mutation causing this phenotype of multi-system smooth muscle dysfunction. This unique case report expands the spectrum of disorders that should be considered in the evaluation of children with intestinal failure and suspected visceral myopathy.

**Case Description:** This is a 7-year-old male with long standing history of vomiting, abdominal distension, and feeding intolerance, required parenteral nutrition for several years. Contrast imaging studies demonstrate diffuse small bowel dilation and slow colonic transit time, without evidence of anatomic obstruction. He also has a history of recurrent urinary tract infection, and markedly distended urinary bladder with bilateral hydronephrosis. These findings are highly suggestive of visceral myopathy. He has an established CNS vasculopathy with a Moyamoya-like presentation, with a history of chronic CNS infarction.

Family history is significant for a sister with similar gastrointestinal symptoms, requiring TPN. This sibling died following complications of a stroke.

Our patient and his sister had undergone genetic testing. ACTA2 mutation was not found. Other investigations are in progress to identify a causative mutation that might offer genotype-phenotype information.

**Discussion:** An alpha-actin smooth muscle mutation can affect all smooth muscle in the body, leading to multi-system myopathy that may cause both intestinal failure and severe CNS vasculopathy. ACTA2 mutation disorders have been described in this presentation, although this spectrum of disorders is rare and not considered in the standard evaluation of patients with myopathic CIFO.

![Figure 1. Upper GI demonstrated dilated small bowel](image)
Figure 2: Abnormal cerebral vasculature

Conclusion:
This case illustrated the importance of multidisciplinary care when treating highly complex patients with intestinal failure, and the role of genetic testing within our field.

P1.12 - Surviving more than 10 years after intestinal transplant. First cohort of patients transplanted at a single centre.

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Hospital Universitario Fundación Favaloro

Introduction: Intestinal transplant (ITx) remains the last therapeutic alternative for patients (pts) with intestinal failure (IF). Although pt and graft survival have improved over time, there are few reports of the long term outcomes of individual pts surviving more than 10 years with functional grafts; there is no long term results report from Latin-America.

Aim: to describe the outcome of those pts underwent ITx at our center and have more than 10 years survival with functional graft.

Materials and methods: This is a retrospective analysis of ITx long term survivors starting in May 2006. Diagnoses, ITx indications, incidence of acute and chronic rejection, complications related to immunosuppression (IS) and social reinsertion over time are reported.

Results: 5/49 (10.2%) ITx done at our center, have more than 10 year survival with a functional graft. Four of them are currently adults, but 3 were less than 18 years of age at the time of ITx; 3 are male. All pts had Short Bowel Syndrome (SBS). The indications for ITx were intestinal failure associated liver disease (40%), lack of central venous accesses (40%), and recurrent line sepsis (20%). The mean duration of hospital stay after ITx was 39.6±18.02 days (4 isolated ITx, 1 combined Liver-ITx). All pts presented at least 1 episode of acute cellular rejection (mean 2.8±1.78); 1 mild, 1 moderate, 2 severe. 7 episodes occurred during the first year post ITx and the rest occurred between the first and the fifth year post ITx. None of the pts developed chronic rejection. Complications related to IS therapy are chronic renal failure requiring kidney transplant (1); diabetes (1); bilateral aseptic necrosis of the femoral head (2) The 5 patients suffered infectious complications episodes (8 viral, 3 bacterial, 1 mycotic, 1 parasitic). One pt had visceral Kaposi's sarcoma. All the long term survivors were able to resume their social and labour activities (3 are students, 1 is a nurse, 1 is a builder); one pt had a child after ITx.

Conclusions: ITx has become feasible option in the Latin-American region with long term survival under the care of a multidisciplinary comprehensive team. Early diagnosis and aggressive management of post ITx complications is required in order to increase the long term survival with a functional graft.
P1.13 - Comprehensive guideline for the management of mesenteric ischemia

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Introduction: Concerning that the leading cause of short bowel syndrome in Asian countries is vascular events, unlike western countries which Chron’s disease lies at the top of the list, and also considering that most of these patients would die without proper management and planning, we had decided to design an algorithmic approach for managing these group of patients including years of experience in our country and also other pioneer studies of the field in other countries, by which we have fortunately come into promising results. Due to limited options and lacking HPN most surgeons in our country prefer resection and anastomosis, which had lead to an unfavorable result during past years. So we as the first small bowl rehabilitation and transplantation center in our country decided to design a comprehensive management guideline for this entity and having a categorized approach which includes, what should the first surgeon confronting this situation do at a local hospital and what would the next treatment plan be in a referral hospital, whether the patient needs more complex procedures as small bowel transplantation or reconstructive surgeries, or not.

Methods: A study group of the Intestinal Rehabilitation Unit (IFU) of Shiraz University of Medical Sciences, Iran was formed in 2018 with the aim of developing guidelines for the management of AMI. The evidence was then reviewed to answer these questions, and recommendations formulated.

Results: Regarding what mentioned in the article, mortality will decrease and a considerable number of patients will survive and by performing AGIR surgeries possible need for bowel transplantation will become less. The resultant recommendations are presented in this paper.

Conclusion: The aim of these guidelines is to provide recommendations for practice that will lead to improved outcomes for patients.

P1.14 - Acquisition of food allergy by multivisceral transplantation

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Introduction: Adoptive transfer of food allergy by transplanted organs has been reported after most solid organ transplants. Here we describe two cases of multivisceral transplantation where the recipients acquired the food allergies of their donor.

Methods: Case note review

Results: Case 1 - A 62 year old male with cirrhosis secondary to NASH was dependent on parenteral nutrition for two years prior to decompensation following small bowel resection for intestinal obstruction. The donor was a young adult female with food allergies to peanut, shellfish and baked beans who died of anaphylaxis after eating a cereal bar. Intestine, liver, pancreas and colon were transplanted. The donor serum demonstrated a total IgE of 259 u/l, and specific IgE positivity to peanut (32.6kua/l), Ara h1, h2 and h3. Following transplant the recipient avoided all nuts, shellfish and baked beans for 6 months. Specific IgE was positive to peanut and ara h1-3 but became negative after one month, and negative to shellfish and tree nuts. Skin prick tests were positive to peanut and grass pollen but negative to other allergens which were subsequently re-introduced. Skin prick positivity to peanut persisted beyond three years post transplant. He avoids peanuts and carries an adrenaline auto-injector.

Case 2 – A 29 year old female who underwent liver transplantation for alpha-1 antitrypsin deficiency at the age of 3 years became parenteral nutrition-dependent following intestinal resections for adhesional obstruction. She underwent liver, intestine, pancreas and colon transplant from a paediatric donor with a history of food allergy who died of anaphylaxis. The donor serum revealed strong specific IgE positivity to cow’s milk (14.9kua/l), whey, casein and hazel nuts and ara H8 peanut allergen only. Total IgE was 301. Post transplant the recipient demonstrated brief IgE positivity to cow’s milk only. Skin prick tests were positive only to cow’s milk and remain positive at 5 months. The patient has reintroduced all other foods with the exception of hazel nuts without incident but continues to avoid milk.

Conclusions: Food allergy transfer by intestine containing grafts may persist longer than with other solid organ transplants, presumably due to persistence of sensitised passenger lymphocytes in the graft. Skin prick tests may be more accurate than specific IgE antibodies and patients should carefully avoid known allergens and take precautions against anaphylaxis.
P115 - Paediatric domino liver transplant following multivisceral transplant (liver-inclusive) with splenic preservation – A case report

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Introduction: A multivisceral transplant followed by domino transplantation of the resected liver to another patient was first reported by Tzakis in 1999. Since then there has not been much development in this technique and its implications for both recipients. We present a paediatric multivisceral transplantation with native spleen preservation followed by the use of the recipient’s liver as a domino graft for another child.

Patients and Methods:

Multivisceral donor: 3 year old boy with hypoxic brain damage, blood group O+, weight 34 kg.

Multivisceral recipient: 7 year old boy, blood group A+, weight 24 kg, with severe intestinal failure (Neuropathic intestinal dysmotility). No enteral feed and 24-h PN, with persistent central line infections. Listed for multivisceral transplant seeking immunological advantage. At transplant, normal native liver (mild portal fibrosis at biopsy) preserved for domino transplant. Splenic artery and left gastric artery were performed preserving native spleen. Native liver perfused with UW solution and stored. Multiorgan graft implanted (liver, stomach, duodenum, pancreas, small bowel and right haemocolon). CIT: 10h.

Domino liver recipient: 3 year old boy, blood group A+, weight 12 kg, with cholestatic liver disease. Domino liver transplant (graft weight: 484 g, CIT: 10h, duct-to-duct anastomosis).

Results: Multivisceral recipient had bowel obstruction (day 3 post-transplant) that required laparotomy. No acute cellular rejection and no serious infections post-operatively. Stoma reversed at 5 m. He remains on enteral feeding and PN support due to rapid bowel transit. The domino liver recipient had acute cellular rejection (day 8 post-transplant). Anastomotic biliary stricture corrected with biliary reconstruction (6 m post-transplant).

Conclusion: A liver-inclusive intestinal transplant seems to give an immunological advantage to the recipient by decreasing rejection episodes but make them vulnerable to lethal infections (Wu & Cruz, 2018), which is maybe worsened by native splenectomy at transplantation. Preserving the native spleen may reduce the incidence of PTLD, GVHD and risk of infections. The above case could be a better option than a modified multivisceral transplantation and using the multivisceral recipient’s liver as a domino graft will reduce the impact on the donor pool. More cases are required to establish the advantages of these surgical techniques.

P116 - Chymie Reinfusion As Treatment Of Temporary Intestinal Failure Type 2 Related To High-Output Double Enterostomies Or Entero-Ateospheric Fistulas.

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Clinique saint yves

Introduction: Temporary double entero stomy (DES) and entero-atmospheric fistulas (EAF) may lead to a type 2 intestinal failure by short bowel syndrome type I. The reinfusion of chyme (CR) from the afferent small bowel to the efferent ileum (EI) restores the intestinal functions until the surgical reestablishment of continuity (SR). Retrospective observational study of 306 patients with CR hospitalized in our centre for 19 years.

Methods: Data from 01/2000 to 12/2018. CR was made with automates and portable pumps with batteries. Height, weight, albumin (Alb), daily intestinal output (IO), nitrogen and lipid absorption (NA, LA), plasma citrulline (Cit) and liver enzymes (ASAT, ALAT, Alkaline phosphatases (AP), γGT were recorded before and during CR. BMI, Nutritional Risk Index (NRI) were calculated. Durations as medians ± SD. Student’s t-test, χ² Fisher’s tests.

Results: 18SH/ 12SF, 64±15 y. DES 266, EAF 40. CR began 5±8d after admission and was pursued 62±31d until SR, at home for 3% of DES patients. The intestinal function improved: IO (2378±775 ml/d - 325±320), NA (51±18 % - 81±12 %), LA (41±22 % - 87±9%), Cit (17.5±6.9 µmol/l - 32.2±17.4). Nutritional status: BMI 24.9±6.5 – 25.1±5.4, Alb 28.2±5.6 – 33.9±5.0 g/l, patients with NRI<83.5 (68% – 27%). The % of patients with plasma liver tests > 2N: ALAT 26% – 13%, ASAT 28% – 10%, AP 42% – 14%, γGT 78% – 38%. Oral feeding was exclusive (n=241), with an enteral complement “en Y” in EI (n=75). The intravenous supplementation (IVS) requirements were nutritional (n=134) or hydration alone (n=44). They were stopped in 19/ 205 cases, 2±8d after the beginning of the RC. (p<0.001 in all results).

Conclusion: In cases of high-output DES or EAF, the CR reestablishes the continuity of the bowel, restores its functions and adapts the downstream small bowel and colon before SR. Some postoperative complications are prevented. Patients are fed with what they eat, their nutritional and liver status improves, IVS are stopped in 92% of the cases, reduced otherwise. CR is feasible at home. CR is much less expensive and avoids complications related to IVS and central venous catheters. This enteral technic was recently recommended by special interest groups of ESPEN and ASPEN.
Ten young people have been transitioned and care handed over to the adult multidisciplinary team. The emphasis is very much on communication between paediatric and adult teams has highlighted differences in protocol processes. This has helped us prepare the young people to cope with the differences. Healthcare professionals need to therefore work collaboratively with young people to ensure they have all the information and resources to engage with the service. This will help to ensure a good long term outcome in young people undergoing transition.

**P17 - Hypomagnesaemia and Long-Term Outcome after Isolated Intestinal Transplantation- Taiwan experience**

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**Introduction:** Hypomagnesaemia is a frequent complication in the early post isolated intestinal transplantation period and particularly associated with the use of calcineurin inhibitor (CNI) which impair renal tubular function causing tubular magnesium loss. Intestinal transplant recipients are at risk of hypomagnesaemia due to the high dosage of immunosuppressant post transplant than other organ transplantation. And we initiated an intestinal transplant program in Taiwan since 2007. We check the magnesium serum level regally after intestinal transplantation and provide magnesium supplement when the level below normal level to prevent hypomagnesaemia.

**Methods:** Twenty-two isolated intestinal transplantations were operated in 21 patients (6 children and 15 adults) with one re-transplantation. Immunosuppression protocol included tacrolimus with combination of low-dose steroids and MMF in some cases. Infectious complications were monitored over CMV and EBV titers. Scheduled biopsy of the graft and biochemical analysis of blood samples were used for the surveillance on the patient condition and graft function. Magnesium level was monitored weekly to monthly and Mg Sulfate 10-20mg was provided according to the level. We analyzed the cases with graft above 1-year survival and calculated the percentage of magnesium supplement among these patients.

**Results:** The causative factors for intestinal failure in these 21 patients include short bowel syndrome (16/21, 80%) and motility disorder (5/21, 24%). Indications for intestinal transplantation were repeated catheter-related sepsis (4/21, 20%), liver function impairment (3/21, 15%), major vein thrombosis (5/21, 25%), and ultra short bowel (12/21, 55%). The survival rates for 3-year, 5-year, and 5-year are 87%, 75%, and 65% for patients. In the 13 grafts that survive longer than 1 year, we found more than 90% patients had hypomagnesium and received supplement with 20 meq Mg Sulfate everyday to maintain their magnesium level around the lower normal limit of 16 mg/dL in early period after isolated intestinal transplantation.

**Conclusion:** Isolated intestinal transplantation is promising and high prevalence of magnesium deficiency was reported in the early post transplant period. Monitor magnesium level and supplementation after intestinal transplantation was essential.

**P18 - Setting up a transition service for intestinal transplant patients**

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**Introduction:** Improving survival of intestinal transplantation resulted in an increase in number of young people surviving into adulthood. The prospect of transition is exciting but “scary” in the words of our young people requiring the development of a service which meets their unique needs. It is vitally important that a robust supportive transition process is established for young people and families.

**Method:** Transition is discussed from 12 years with the family at their annual anniversary admission, introducing the concept and describing the process. Young people meet with the transition nurse, youth worker and psychologist. We use a transition programme called Ready, Steady Go and the HEADSS document to support transition.

Families first have the opportunity to meet the adult team at our biannual family day. The adult team consisting of the Lead Consultant, Specialist Nurse and Dietician contribute to the information session during the day. These can be a talk or information boards/posters. This allows families to gather information to make informed choices about their young person’s future care.

The adult team join us in the outpatient clinic at the children's hospital to meet the young person and their parents/carer. It is the start of the formal transition. The Paediatric Consultant and Specialist Transition Nurse attend the adult clinic. The adult consultant will lead the consultation with the Paediatric Consultant contributing.

**Results:** Ten young people have been transitioned and care handed over to the adult multidisciplinary team. The emphasis is very much on the young person and the time scale will be driven by their needs.

Retrospective review of the medical notes shows that young people are seen once in the paediatric clinic with the adult team, seen two/three times in the adult clinic with the paediatric team in attendance prior to formal handover. Formal handover can take two/three years. Six young people are in the process of being transitioned.

**Conclusion:** Feedback from families has shaped the service. Communication between paediatric and adult teams has highlighted differences in protocol processes. This has helped us prepare the young people to cope with the differences. Healthcare professionals need to therefore work collaboratively with young people to ensure they have all the information and resources to engage with the service. This will help to ensure a good long term outcome in young people undergoing transition.
Introduction: Chronic intestinal failure in children is a rare and heterogeneous disease for which high-quality evidence is lacking to guide clinicians in daily practice. The European Reference Network for rare Inherited and Congenital Anomalies (ERNICA) has installed an intestinal failure working group with the aim to improve the care and treatment for these patients. One of the working group’s strategies is to combine the knowledge on intestinal failure in the European expertise centres united in ERNICA. Areas of research interest and plans for future research topics and development of a research strategy for future projects. The aims of this study are to evaluate the HRQOL in children after ITx with a validated questionnaire, and to compare this population to patients after liver transplantation (LTx), and to patients with IF on long term HPN.

Methods: It is an on-going prospective study including patients between 10 and 18 years old, receiving ITx at Necker-Enfants Malades Hospital, with at least 2 years graft survival. We compared them to patients who underwent LTx or receiving HPN, paired on age, delay from transplantation, diagnosis for children on HPN. We used the Child Health Questionnaire, child report form (CHQ-CF87) and parent report form (CHQ-PF 50), including 3 HRQOL domains: physical functioning, social emotional status, social behavioral status, social physical status, bodily pain, general behavior, mental health, self-esteem, general health perceptions, parental impact, family activities and family cohesion. Mean values for each item will be calculated and converted to percentages. Two-tailed Student t-test will be used to compare the mean scores of subjects.

Results: We enrolled 48 children: 16 after ITx (mean age 14.9 ± 2.7), 16 after LTx (mean age 14 ± 2.9) and 16 on long term HPN (mean age 13.8 ± 3.7). The study just started, with a good participation and feedback from patients and parents. 12 patients and 20 parents already answered. We already observed that in the same subcategory patients and parents scores often differed.

Conclusion: We expect to gain important information about the HRQOL of children after ITx compared to other chronic diseases, to improve the early and long term psychological care of these patients. We hope that better understanding of the impact of these procedures on the child as well as the family’s QOL will assist them in the care.

Discussion: Subgroups were formed to address these different research topics and develop a research strategy for future projects. The ultimate goal is to increase the standard of care for children with chronic intestinal failure.
P121 - Cmv Pan-Drug Resistant Infection In Multivisceral Transplant Recipient: A Case Report Of Successful Treatment

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Introduction: CMV infection is one of the more frequent infection after solid organ transplantation. Several specific antiviral therapy are nowadays available with success in most cases.

Methods: A 38 years old woman underwent total enterectomy due to CMV infection from the same donor. She underwent regular antibiotic, antifungal and antiviral prophylaxis as universally described. Her CMV serological status was D+/R+.

Results: During post operative course a severe abdominal wall infection occurred and vacuum assisted closure needed to avoid abdominal wall graft removal. Despite minimization of the immunosuppressive regimen occurred and CMV viremia increased again. A new drug test resistance showed a viral strain with multiple mutations: M460V on UL97 gene which gives ganciclovir/valganciclovir resistance. Foscarnet treatment was tried, initially successfully, then CMV viremia increased again. A new drug test resistance showed a viral strain with multiple mutations: M460V on UL97 gene (as the previous test), Q578H and E756D on UL54 gene to confer a pan-drug-resistance (PDR) profile towards all drugs anti CMV available (ganciclovir,valganciclovir,foscarnet,cidofovir). Meanwhile the patient developed CMV disease with intestinal and retinal involvement. After multidisciplinary discussion and literature review, a rescue treatment with leflunomide was started. This is a rheumatoid and psoriatic arthritis drug with known anti CMV activity. With the combination of leflunomide and anti CMV immunoglobulins we obtained a complete response of CMV viremia. Leflunomide was discontinued after 1 year of treatment (due to lower limbs side effects) and anti CMV specific immunoglobulins was continued with progressive dosage descalation.

Conclusion: Leflunomide can be considered a rescue therapy in the cases of pan drug resistant CMV infection. The patient is nowadays still alive, with residual blindness but with functioning graft, without parenteral or fluid support and with constantly negative CMV viremia.

P122 - Intestinal failure in a tertiary children’s hospital before and after the establishment of a national intestinal failure service; incidence, aetiology and outcome.

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2 The New Zealand National Intestinal Failure Service.

Introduction: Establishment of a formal intestinal rehabilitation service is associated with improved outcomes for intestinal failure (IF) patients. We compared incidence, aetiology and outcomes of inpatients with IF at the only tertiary children’s hospital in New Zealand, before and after the establishment of the New Zealand National Intestinal Failure Service (NZ NIFS).

Method: A retrospective review of intravenous nutrition (IVN) records identified patients with IF aged 0–32 years, admitted to Starship Child Health between July 2010 and October 2018. IF was defined as inpatients receiving IVN ≥21 days as per the NZ NIFS criteria.

Medical records were reviewed to identify demographics, indication for IVN, biochemical markers for intestinal failure-associated liver disease (IFALD) and clinical outcome. Indication for IVN was defined as per NZ NIFS pathological classifications (adapted from 2016 ESPEN guidelines for chronic IF in adults): extensive small bowel mucosal disease; mechanical obstruction; intestinal dysmotility; intestinal fistula; short bowel syndrome and other diagnosis. IFALD was defined as peak bilirubin >34 mmol/L, for at least 2 weeks, in the absence of another cause.

Results: 882 children received IVN during the 8 year period. 26% (173/682) were classified as having IF. The IF patients came from 17 of NZ’s 20 health care districts. There was no significant increase in patient transfers between the time periods. Three quarters of IF patients came from three of 11 referring teams; oncology 33% (51/171), gastroenterology 28% (48/171) and ICU 39% (32/171).

Common indications for IVN were extensive small bowel mucosal disease (46%), short bowel syndrome (35%) and mechanical obstruction (33%).

IFALD occurred in 9% (15/171), there was no difference between the time periods. There was a slight excess of patients with mechanical obstruction (17%, 4/23) but this was not statistically significant. 73% (125/171) had successful intestinal rehabilitation to enteral autonomy. 6% (10/171) were established on home IVN and existing home IVN patients accounted for 9% (15/171) of IF admissions. Intestinal transplantation was necessary for only one patient. No mortality related to IFALD was identified, however 12% (20/171) died from their underlying diagnosis.

Conclusion: Since the establishment of NZ NIFS in 2015 our inpatient IF demographics have remained broadly the same, the incidence of IFALD remains low and inpatient numbers have not increased.
**P123 - Use of Pureed Green Beans Through a G-tube in Pediatric Patients with Short Bowel Syndrome – A Single Center Experience**

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**Background:** Pectin and other water-soluble fibers have been used in certain patients with short bowel syndrome. Fermented fibers may provide energy contribution via short chain fatty acids produced by colonic bacteria, while soluble fibers that retain a gel may improve the consistency of watery or loose stools and thus decrease stool output. At our institution, we aimed to study the effects of pureed green beans, a good source of pectin, given in a reliable and easy manner, through a G-tube.

**Methods:** Pureed green beans administered via syringe directly into the G-tube was prescribed in a handful of patients with short bowel syndrome. Dose of green beans was adjusted at regularly scheduled clinical visits. Patients and their stools were closely monitored.

**Results:** Of the 71 intestinal rehabilitation patients at our center, 7 are currently receiving pureed green beans through G-tube. All patients experienced a decrease in stool frequency and 6 of them experienced improvement in stool consistency. Over half of the patients also tolerated a decrease in loperamide. Of the 6 patients receiving either TPN or IV hydration, 5 tolerated a decrease in the overall volume. One patient was weaned off of IV hydration fluids entirely. Please refer to Table 1.

**Conclusion:** Overall, our cases highlight the improvement in stool frequency and consistency, TPN volume requirements, and loperamide administration following the administration of pureed green beans into a G-tube. For our patients with G-tubes and difficult to manage loose stools, the introduction of pureed green beans is now being considered more frequently. Potential future research may include: examining if higher doses of green beans increase the risk of malabsorption of certain nutrients (such as magnesium, zinc, and iron), monitoring the incidence of central line infections following stool normalization, and studying the effects of specific fibers or foods on the microbiome in patients with short bowel syndrome.

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**P124 - Vascular stenting for mycotic aneurysm in multivisceral transplant recipients**

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**Introduction:** Severe graft pancreatitis (GP) although uncommon after multivisceral transplantation (MVT) may be associated with necrosis of the gland, infection of necrotic debris and bleeding secondary to erosion of infected necrosis into surrounding vasculature. For MVT the anatomic proximity of graft pancreas to the aortic conduit (AC) poses unique challenge when affected by infection and necrosis as local control of a mycotic aneurysm (MA) by embolization or surgical ligation would lead to ischemia of the transplanted viscera.

**Case 1:** A 40 year old female underwent MVT. Postoperatively, she developed severe GP and intra-abdominal abscesses and had persistent bacterial and fungal sepsis. On day 28, she developed massive intra-abdominal bleed. At emergent laparotomy, aortogram showed irregularities in the AC concerning for MA. An intra-aortic balloon was placed to control hemorrhage with excision of the involved area and primary anastomosis of the conduit. Three days later she had recurrent massive bleeding from multiple sites along the remaining length of the AC which could not be controlled and the patient succumbed.

**Case 2:** A 44 year old male received MVT. His early post operative course was complicated with GP and intra-abdominal abscesses and had persistent bacterial and fungal sepsis. On day 28, he developed massive intra-abdominal hemorrhage which on exploration was noted to be coming from the donor AC. The AC appeared thinned and had blackish discoloration consistent with MA. Vascular surgery colleagues deployed 2 endovascular stents in the celiac axis and superior mesenteric artery to bypass the weakened segment while maintaining blood flow in both the major graft vessels. The space between the stents and wall of the AC was then filled with coils. He had no graft dysfunction although further aneurysmal changes of the proximal AC (Fig1) a few weeks later required extension of the stents from the previously placed stents all the way to his native aorta. He is now more than 1 month after the revision without evidence of infection or extension of the MA, with resolution of the GP and good graft function.
Conclusion: Graft pancreatitis following MVT can lead to fatal hemorrhage from erosion of infected necrosis into the vascular inflow to the graft. Endovascular stenting of the AC vessels (celiac and SMA) offers a potential treatment option in this situation. This is likely a safer and durable solution than attempt at surgical revision of the anastomosis in the presence of infection.
P126 - The impact of advances in intestinal rehabilitation strategies and PN management on the outcome of children with Chronic Intestinal Failure

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Background: Recent advances in the management of children with chronic intestinal failure have presented new therapeutic options for treatment in this complex patient group. To help inform future therapeutic strategies, we assessed the outcome of children with chronic intestinal failure managed by a multidisciplinary intestinal rehabilitation program over a 27-year period.

Methods: A retrospective longitudinal review of children with chronic intestinal failure assessed for home parenteral nutrition (PN) therapy at the Royal Children's Hospital Melbourne from 1991 to 2018.

Results: A total of 64 children with chronic intestinal failure due to short bowel syndrome [SBS; median bowel length 33.5cm] (n=51, 80%), chronic intestinal pseudo-obstruction (n=5), congenital enteropathy (n=4) and other genetic abnormalities (cystic fibrosis n=1, megacystitis microcolon syndrome n=1, immune deficiency n=2) were included. The overall survival was 57 (88%), with no deaths attributed to intestinal failure or its management in home PN patients in the past decade. Weaning from PN was successful in 31/45 (69%) of SBS survivors after a median of 18.5 months. Longer residual small bowel length and the presence of the ileocaecal valve (ICV) and colon were predictors of a good prospect of weaning from PN, particularly if the ICV and colon are preserved. The survival rate and frequency of central line associated blood stream infections have dramatically fallen from ~7.3 episodes/1000 line days in 1991 to 0.5 episodes/1000 line days in 2018.

Conclusion: Advances in the management of children with chronic intestinal failure have had a significant impact resulting in improved outcomes. Children with SBS, even in the presence of small residual bowel length have a good prospect of weaning from PN, particularly if the ICV and colon are preserved. The survival rate and frequency of central line associated blood stream infections have dramatically improved. Targeted individualized therapy and care by a multidisciplinary team expert in intestinal rehabilitation and nutrition can optimize outcomes in children with chronic intestinal failure.

P128 - What do patients with short bowel syndrome eat in real life?

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ESPEN has elaborated guidelines for management of patients with short bowel syndrome (SBS). Those one include dietary recommendations that are difficult to put into practice. Our aim is to evaluate on a cohort of short bowel syndrome the real life food intake in terms of quality and amount.

It is a prospective study including the consecutive adult patients with short bowel syndrome (remaining small bowel length ≤ 200 cm) who had their outpatient visit for regular nutritional monitoring between February 6th, 2018 and May 29th, 2018. As usual, oral intake was evaluated by one of the experimented dietitians involved in our multidisciplinary team. Sixty nine patients (M: 39%; F: 61%), with a mean age of 58.8 years (±15.4) were included. The mean BMI was 22.6 kg/m2 (± 4.5). The mean remaining small bowel length was 99.4 cm (± 54.4). Thirty nine patients (49%) require Home Parenteral Nutrition. Oral intake was evaluated using three days food diary. Total oral intake (percentage of carbohydrates, proteins and lipids), oral rehydration solution (ORS) consumption and amount of fibers were analyzed. The energy expenditure was calculated with Harris and Benedict formulae.

Patients with intestinal failure (n=39) required 4.6 ± 1.9 infusions per week, with a volume of 7.030 ± 4.559 kcal per week. Six patients with jejunostomy consumed ORS regularly with a mean intake of 9 041 ± 3 572 ml per week. Oral proteins/glucids/proteins(%) intake is respectively for patients with jejunostomy and patients with anastomosis 20-30% for patients with jejunostomy and 40% of lipids for jejunostomy and 40-50% for patients with anastomosis and 40% of lipids for jejunostomy and 20-30% for patients with anastomosis.

In a large SBS cohort patients with intestinal failure or intestinal insufficiency, even with dedicated dieticians, patients whatever the intestinal anatomy have the same profile of ingesta in terms of quantity and amount of macronutrients. Interestingly, in our population, 40% of patients with jejunostomy have a regular and consistent ORS intake. That’s probably due to a very important engagement of dietician and physician to explain the importance of drinking specific beverage in these patients. So, we can suppose, that a more adapted, dietetic training program could improve the global adhesion of recommended diet for SBS patients.
**P129 - The impact of a nutrition multidisciplinary team**

Michelle Butcher  
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**Introduction:** The evidence-based standard for optimal nutritional support is the Multidisciplinary team (MDT) approach of a Nutrition Support Team (NST). Our hospital team was established in 2015 after a 3 year pilot 2012-2014. The nurse-led team review new parenteral nutrition (PN) referrals and continue to visit assess and advice twice weekly. The aims of the team are to:

- Improve the safety and adequacy of PN delivery to patients
- Provide high quality nutritional outcomes via enteral and PN
- Deliver significant cost savings
- Improve hospital wide nutrition education (nursing, medical and allied health professional)

**Aim:** To assess improvements in clinical care and financial benefits associated with the NST before and after the service was established looking at:

1) PN bag usage  
2) Standard bag usage  
4) Wastage

**Method:** The PN usage pre and post the NST was collected from a pharmacy database. A new NST database collected prospective data on the activity of the team. Data collected included: clinical specialty, recommendations, abnormal blood results, time on PN and refeeding risk. This data was analysed as part of this audit process.

**Results:**

- There has been a 20% reduction in the number of total PN bags/year (fig 1).
- Standard bag usage increased from 6% to 30% between 2012 and 2018 (fig 2).
- Reduction in PN usage and increased % of standard bags has reduced cost of PN provision between 2012 and 2018 by approximately €230,000/year.

**Conclusion:** Regular patient reviews, forward planning and education has improved the safety of PN delivery, decreased the total number of PN bags used and contributed to increased standard bag usage. Choosing other available standard bags to suit our population of patients has also been a contributory factor for this significant increase. Wastage of PN has shown a modest decrease during the study period. Anticipating weaning of PN and patient transfer is an area of focus for the team.
Teduglutide: the new weapon in pediatric short bowel syndrome

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Introduction: Short bowel syndrome (SBS) is generally defined as the loss of an extensive length of the small intestine resulting in an inadequate absorption of enteral nutrients. Teduglutide is a recombinant analog of native human GLP-2 with resistance to in vivo degradation, enhanced intestinal adaptation and increased the absorptive capacity of the remnant intestine in patients with SBS.

Methods: A premature girl (33 weeks) with necrotizing enterocolitis (NEC) underwent extensive intestinal resection (only 30 cm of jejunum anastomosed to the sigmoid colon preserved). Intestinal dilation and the need for PN forced to perform an intestinal lengthening with a modified serial transverse enteroplasty procedure (STEP) twice, at 19 months and 3.5 years (final small bowel length 175 cm).

At the age of 7 years old, the patient still required PN 25 Kcal/kg and high volume of iv fluids (2300 ml; 85 ml/kg) administered for 17 hours/day. She had 7-8 semiliquid bowel movements/day (output 2000 ml/day) and frequent hospital re-admissions owing to catheter-related infections. She only had a single central venous access for PN administration. The written informed consent of the parents was obtained for the administration of teduglutide and the hospital approved its subcutaneous administration in a dose of 0.05 mg/kg/day.

Results: The evolution of PN volume, calorie requirements, days of PN administration and plasma citrulline, before and during teduglutide treatment are shown in Table 1.

Prior to treatment, weight, height and body mass index (BMI) were 27.2 kg (+0.86 SD), 135 cm (+2.04 SD) and 14.9 (-0.39 SD), respectively. After 4 weeks of treatment, less frequent bowel movements, change in stool form (more consistency) and increased urine output were noted, weaning off PN support was initiated. At 22 weeks, the patient only required PN 2 days/week which was stopped at week 30 of treatment. In parallel, a rise in serum citrulline was observed.

After 18 months of treatment we decreased dose to 0.025 mg/Kg/ day and after follow-up of 24 months, the patient remained free of PN at this daily dose. Nowadays, bowel movement frequency is 2-3 per day (type 5 on the Bristol scale). Weight (31.6; -0.24 SD) and BMI remain stable (14.8; -0.85 SD), while her height has increased to 146 cm (+15 SD).

Conclusions: Teduglutide has allowed this child, a candidate for intestinal transplantation, to achieve complete PN-free status with a great improvement in her quality of life.
P131 - Salvage therapy with Infliximab for anastomotic ulcers- a potential treatment for a challenging complication

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4 Division of General & Thoracic Surgery, Hospital for Sick Children, University of Toronto, ON
5 Department of Pediatric Gastroenterology, Starship Child Health, University of Auckland, New Zealand

Background:
Children with short bowel syndrome (SBS) may require bowel lengthening procedures such as the Serial Transverse Enteroplasty (STEP) procedure. STEP may be associated with severe complications such as anastomotic and staple line ulcers with gastrointestinal (GI) bleeding. These complications pose a therapeutic challenge and are often refractory to medical therapy. We present a child with recurrent GI bleeds from staple line ulcers responsive to treatment with infliximab (IFX).

Case:
An 11 year old male with SBS and intestinal failure secondary to gastroschisis with midgut atresia underwent STEP procedures at 4 months of age and again at 4 years. He developed recurrent GI bleeds and was found to have multiple ulcers along staple lines on repeat endoscopies. Initial treatment included mesalamine (65mg/kg/day), oral budesonide (9mg), pantoprazole, omega-3, and cycling antibiotics with no improvement. Due to failure of all treatment attempts by 8 years old, he was started on IFX (6.5mg/kg/dose at week 0, 2, and 6 weeks after 3 doses of IFX). Due to high anti-IFX antibodies, 190AU/mL, IFX was stopped. After 3 weeks of IFX treatment, he started on IFX with methylprednisolone and GI bleeding occurred. The patient was held at dose #4.

Further biologic therapies attempted included adalimumab and golimumab, not effective in preventing recurrent GI bleeding. The failure of medical therapies led to massive small bowel resection with removal of all areas involved with the previous STEP procedures. The most proximal staples were left in situ. Despite resection, new ulcers and GI bleeding occurred at the remaining staple site and in the duodenum. As a result IFX was reintroduced with methylprednisolone for premedication. The patient received IFX (10mg/kg/dose, week 0 and 2), repeat induction at week 4 due to low levels of IFX (<0.035ug/mL) and anti-IFX antibodies <2AU/mL. Almost complete mucosal healing was found on endoscopy after dose #4. At infusion #6, anti-IFX antibodies were 70AU/mL, the child developed a rash and fever during the infusion and IFX was stopped.

Other biologic therapies attempted included adalimumab and golimumab, not effective in preventing recurrent GI bleeding. The failure of medical therapies led to massive small bowel resection with removal of all areas involved with the previous STEP procedures. The most proximal staples were left in situ. Despite resection, new ulcers and GI bleeding occurred at the remaining staple site and in the duodenum. As a result IFX was reintroduced with methylprednisolone for premedication. The patient received IFX (10mg/kg/dose, week 0 and 2), repeat induction at week 4 due to low levels of IFX (<0.035ug/mL). Follow up endoscopy showed excellent mucosal healing after 3 doses of IFX. Due to high anti-IFX antibodies, 70AU/mL, IFX was held at dose #4.

Conclusion:
Anastomotic and staple line ulcers post-STEP procedures were responsive to treatment with IFX in this case. Development of anti-IFX antibody can be a barrier for a successful outcome and should be monitored. The response to IFX suggests a potential role for innate immunity and TNF alpha in the pathogenesis of anastomotic ulcers.

P132 - Concordance of Fibroscan and Biopsy for monitoring liver fibrosis in patient undergoing long term parenteral nutrition.

Yassmin Syagha, Denise Cerqueira Paranaaguá Vezozzo, Mariana Hollanda, Daniel Reis Waisberg, Andre Dong Won Lee, Rafael Soares Nunes Pinheiro, Flavio Silva, Luiz Augusto Carneiro D’Albuquerque, Dan Linetzky Walzberg, FLAVIO GALVAO
Department of Gastroenterology

Background: Home parenteral nutrition (HPN) associated liver disorder remains a major metabolic complication that may requires liver-transplant or multivisceral transplantation in some cases. The decision about the timing and the type of transplantation depends on the degree of liver fibrosis, whose evaluation requires liver histology. Although hepatic fibrosis is the gold standard for detecting liver fibrosis, it is an invasive procedure with some complications. Therefore, noninvasive methods as a Fibroscan were developed to assess liver fibrosis. The goal of this study was to compare liver biopsy and Fibroscan in the evaluation of liver changes in patients with intestinal failure in HPN.

Methods: Inclusion criteria: Patients with short bowel syndrome using HPN for at least six months who underwent liver biopsy. In each patient we evaluated: characteristic of HPN, underlying disease, gut anatomy; clinical assessment; biochemical work-up; liver biopsy: Fibroscan assessment (score of liver stiffness). Liver histologic fibrosis was scored according the Brunt classification (grade: 0 to 4).

Results: Eighteen patients were enrolled (07 women / 11 men, mean age: 37y.). Reasons for liver biopsy were altered hepatic enzymes (n = 18). The median duration of HPN was 31 months (range: 12-133). Indication for HPN was short bowel disease (chronic mesenteric ischemia n = 4; Crohn’s disease n = 4; complications of bariatric surgery n = 4; volvulus n = 3) and chronic intestinal pseudoobstruction (n = 3). Liver histology showed severe fibrosis (Brunt stage > 2) in 5 patients (28%). Abnormal Fibroscan score (F > 3) was observed in 5 patients (28%). The results of liver disease graduation are presented in Table 1. Statistical analysis by Concordance Kappa was evaluated and no correlation was observed between Fibroscan and liver biopsy (p < 0.238).

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Table 1: Comparison between the Fibroscan and the Brunt stage.

Conclusion: No correlation was observed between Fibroscan and liver biopsy in patients on HPN due to short bowel disease.
P133 - Is there a role for ultrasound in the evaluation of graft small bowel?
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Introduction: Intestinal graft recipients are subjected to often significant doses of radiation. MRI studies may not be feasible acutely. Endoscopic examination of the intestine is usually limited to small sections of the graft. Ultrasound (US) is an established technique in the evaluation of disease extent in patients with small bowel Crohn’s. We have utilised US for several years to evaluate the graft small bowel and have retrospectively reviewed our findings.

Methods: This is a retrospective review of small bowel US studies performed post-operatively for our cohort of 97 patients receiving an intestine-containing graft between 2007 and 2019. Imaging interpretation was made on the basis of grey-scale images and Doppler imaging. US images were reviewed with subsequent clinical, imaging, endoscopy and histology findings.

Results: 97 patients received an intestine-containing transplant between 2007 and 2019. There were 45 US studies undertaken in 27 patients. The majority of studies (23 studies in 9 patients) were undertaken in those with biopsy-proven acute cellular rejection (ACR). Imaging findings of mural thickening, loss of mural stratification, reduced peristalsis and mesenteric hypervascularity were observed in 7 patients with ACR. 2 patients with a history of ACR had normal US appearances at follow up, which correlated with endoscopic findings of recovery.

Of the US performed for ACR, concurrent endoscopy (within 1 week) occurred in 11 US studies. 8 endoscopies demonstrated features related to rejection confirmed at histology. 3 demonstrated recovery.

5 patients underwent US immediately post surgery with normal findings consistent with concurrent CT or endoscopy findings.

17 US studies were undertaken for 17 patients for a range of clinical indications: 5 studies were normal, 4 consistent with clinical / imaging findings. 1 patient had drug related ulceration at endoscopy. 11 studies in 6 patients demonstrated minimal mural thickening but no further features to suggest rejection, 4 were followed with endoscopy with no features of rejection. The remainder were followed clinically and radiologically. 1 US demonstrated features suggesting rejection but normal endoscopic findings. An internal hernia was found at surgery.

Conclusion: Small bowel US is a useful technique in establishing normal appearances of the bowel. When interpreted in conjunction with clinical and endoscopic findings in patients with ACR it may have a potential role in surveillance.

P134 - Multivisceral transplantation in the Czech Republic: 4 years single-center experiences
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Michal Kudla
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Introduction: Intestinal/multivisceral transplantation (IT/MVT) is the gold standard treatment for patients with intestinal failure and complications related to total parenteral nutrition, gastrointestinal inoperable indolent tumors, or diffuse portal thrombosis.

Methods: Since 2004 we performed 7 full multivisceral transplantation between December 2013 and May 2018. Indications for transplantation were multivisceral thrombosis (n=5) and desmoid tumours (n=2). Imunosuppression was based on induction with almtuzumab (followed by tacrolimus and steroids) in the first period and infliximab and basiliximab (followed by with tacrolimus and steroids) in the last case.

Results: Four patients are alive. All these patients are without parenteral nutrition. Mortality rate was 43% due to sepsis in 2 cases and in 1 case to cardiac failure in the first day after transplantation. In all patients we diagnosed acute cellular rejection. Graft-vs.-host disease was seen in 1 case.

Conclusion: Intestinal transplantation is a suitable treatment for highly selected patients with intestinal failure who meet specific listing criteria.
P135 - Spectrum of major abdominal surgical procedures within a pediatric intestinal rehabilitation (IR) program

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Background: Surgery plays a key role for the intestinal rehabilitation (IR) of children with short bowel syndrome or chronic intestinal failure. Whereas single procedures – especially for intestinal lengthening – are regularly being evaluated and described, the whole spectrum of surgical procedures within large IR programs in children is often difficult to assess. Our aim was to analyse the complete spectrum of major abdominal surgical procedures in a pediatric IR program.

Methods: We retrospectively analysed patients' data from our pediatric IR program with special focus on all patients undergoing major abdominal surgery. Patients characteristics as well as surgical data were evaluated.

Results: The study period ranged from 2013 until 2018. During that period 384 patients were treated within our IR program. Of these children, 58 (15.1%) underwent major abdominal surgery. There were 35 male and 23 female patients. Diagnoses were anatomical short bowel syndrome (gastrochisis, atresia, volvulus on others) in 40 patients, hypoperistaltic conditions (aganglionosis, hypoganglionosis, MIMHS) in 37 patients and malabsorption (Microvillus Inclusion Disease) in one patient. Ten children originated from our own area, whereas 48 children were referred to us from different national or international regions. Mean number of operations prior to referral to us in those patients coming from other sites was 2.6 (0-7). In the 58 patients, 17 operations were performed (median 1.5, range 1-6). One operation was performed in 29 children, 19 children underwent 2 operations, 6 children underwent 3 operations, 2 children received 4 operations and 2 children underwent 6 operations. The types of surgical procedures were mostly reconstructive (n=100), while lengthening procedures were performed on 7 occasions (6.5%). Mean operating time was 145.2 minutes (range 31-1356). One child died in the early postoperative phase because of the central line sepsis.

Conclusion: The spectrum of surgical procedures in a pediatric IR program displays a wide variety of indications; reconstructive procedures are predominant. The procedures are complex and time consuming and require a comprehensive management including preoperative workup and indication, intraoperative surgical and anaesthesiological care as well as postoperative intensive care treatment.

P136 - Essential fatty acids profile in pediatric patients receiving an intravenous lipid emulsion containing 15% fish oil

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3 Hospital Pharmacy at Necker; University Sorbonne-Paris-Cité; Paris Descartes Medical School.

Composite intravenous lipid emulsion (ILE) containing soybean (30%), medium chain triglycerides (MCT) (30%), olive (25%) and fish (15%) oil (SMOFlipid®) is now widely used in the world for prematures, newborns and children on parenteral nutrition (PN). Long-term use in children on home-PN (HPN) is not documented. The aim was to assess the fatty acids (FA) profile of such HPN children.

Population: 26 children with chronic intestinal failure (CIF) were assessed (SBS = 10, congenital enteropathy = 9, total aganglionosis = 4, CIPOS = 3) aged 6 months-16 years on home-PN for 12 months-16 years, highly dependent on PN (low plasma citrulline levels and high PN intake). All received SMOFlipid® as source of ILE for 12-38 months at the dose of 2.1±0.39 g/kg/day, 6.7±0.7 days/week. They were compared to 26 SBS children aged 7.8±2.9 years, weaned off PN for >2 years.

Methods: Sampling performed after ≥24h fat free PN and 6-8h after PN discontinuation. Red blood cell (RBC) fatty acids (FA) profiles were established by using gas-chromatography. Citrulline plasma levels, ratio non protein energy PN-intake (NPE)/resting energy expenditure (REE) - Schofield equation -, growth parameters in Z-score and total bilirubin were assessed. FA profiles and Holman ratio (triene/tetraene) were compared to those obtained from the control group.

Results:

<table>
<thead>
<tr>
<th>SMOLF (n=26)</th>
<th>Control (n=25)</th>
<th>P values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Citrulline (mean ±SD)</td>
<td>5.9 ± 2.8</td>
<td>27.0 ± 7.8</td>
</tr>
<tr>
<td>NPE/REE (%)</td>
<td>130 ± 29 %</td>
<td>0</td>
</tr>
<tr>
<td>Total bilirubin (umol/l)</td>
<td>11.3 ± 7.0</td>
<td>6.4 ± 3.0</td>
</tr>
<tr>
<td>Body weight (z-score)</td>
<td>-0.1 ± 1.4</td>
<td>-0.2 ± 1.6</td>
</tr>
<tr>
<td>Body size (z-score)</td>
<td>-0.3 ± 1.5</td>
<td>0.1 ± 1.2</td>
</tr>
<tr>
<td>C16:0, Palmitic Ac</td>
<td>26.3 ± 1.7</td>
<td>24.5 ± 2.6</td>
</tr>
<tr>
<td>C18:1, n-9, Oleic Ac</td>
<td>15.8 ± 1.5</td>
<td>15.8 ± 1.1</td>
</tr>
<tr>
<td>C18:2, n-6, Linoleic Ac</td>
<td>7.8 ± 3.4</td>
<td>9.6 ± 1.6</td>
</tr>
<tr>
<td>C18:4, n-3, EPA</td>
<td>8.5 ± 1.4</td>
<td>14.7 ± 1.7</td>
</tr>
<tr>
<td>C20:5, n-3, EPA</td>
<td>3.8 ± 1.4</td>
<td>3.8 ± 0.52</td>
</tr>
<tr>
<td>Holman ratio</td>
<td>0.029 ± 0.014</td>
<td>0.026 ± 0.005</td>
</tr>
</tbody>
</table>

NSD: No significant difference.

Conclusion: Long-term administration of an ILE rich in fish oil (15%) in highly PN dependent children (PN/REE:<20 ± 20 %) is well tolerated. Bilirubin plasma levels remained low and growth was normal. The RBC-FA profile, reflects the n-3/n-6 FA acid composition of this new fish oil rich ILE, without EPA deficiency as assessed by the Holman ratio. This “new” RBC-FA profile, followed on the long term, is not accompanied with any harmful clinical effect and can be considered as safe and probably beneficial for preventing IFALD.
P137 - Incidence and outcomes of exfoliative rejection at a UK transplant centre

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2 Department of Transplant Surgery, Cambridge University Hospitals, Cambridge, UK

Introduction: Exfoliative rejection (ER) of the intestinal graft is associated with significant fluid and electrolyte losses, recurrent sepsis due to microbial translocation and often leads to graft enterectomy, re-transplant or death. The majority of previously reported cases or case series of ER are in paediatrics.

Methods: We present here a case series of ER in adults from a single centre in the UK over the time period 2007-2018. This is a retrospective review of a database containing patient demographics, endoscopy and histology findings and outcomes. ER was diagnosed based on endoscopic findings of widespread areas of denuded mucosa.

Results: 84 patients were transplanted in the specified time period: 34 MVT, 13 Liver/Intestine, 11 MMVT, 25 Intestine only. 7 patients (8.3%) experienced an episode of ER. Precipitating reasons for ER were immunosuppression switch (2), non-compliance (2), low immunosuppression levels because of comorbidity (2) and in one case the patient had concurrent severe adenovirus infection. It is not clear whether the viral infection triggered rejection.

All patients received pulsed steroids. 4/7 patients received second line treatment with Alemtuzumab (1) or Anti-Thymocyte Globulin (3). 3 patients underwent repeat transplant of which one had graft enterectomy 3 weeks prior to the second transplant. 2 further patients had limited graft resection due to strictures and had subsequent full graft function. The only death related to ER occurred in the patient with concurrent adenovirus infection. It is not clear whether the viral infection triggered rejection.

Conclusion: Exfoliative rejection in this case series frequently occurred in the setting of overall lower immunosuppression than would be desirable. In some cases this was unavoidable, but every effort should be made to support patient adherence to treatment and to provide general psychological support. Historically, switching immunosuppression has been a potential trigger for severe/exfoliative rejection in our unit. Protocols are now in place for managing switches and since these have been instigated, no episodes of rejection have occurred in this context. Reducing immunosuppression due to co-morbidity is always a difficult decision and should be made within a full multidisciplinary setting.

P138 - The Dental Health of patients referred for Small Bowel and Multivisceral Transplantation to a UK transplant centre

Cambridge University Hospitals NHS Foundation Trust

Introduction: Dental care during pre-transplant workup is important to reduce potential sources of infection, following immunosuppression to prevent graft rejection. This study documents the prevalence of oral abnormalities and recommended treatment of patients assessed for small bowel and multivisceral transplantation.

Methods: We present a case series of 57 patients who underwent assessment for small bowel and multivisceral transplantation in a single centre in the UK (Cambridge) over the time period 2015-2019. This is a retrospective review of their formal assessment in a maxillofacial clinic and subsequent outcomes.

Results: Of all patients assessed, 18 (32%) required no treatment or crown and polish only; 12 (21%) required at least one filling; 27 (47%) required at least one tooth extraction and 6 (10.5%) required full dental clearance. Of the 28 patients accepted and listed for transplantation, 11 (39%) required no treatment or crown and polish only; 4 (14%) required at least one filling; 13 (46%) required at least one tooth extraction and 4 (14%) required full dental clearance.

Conclusion: This is the first reported study of the dental health of patients referred for small bowel and multivisceral transplantation. Overall these patients have poorer dental health compared with published data for patients referred for liver transplantation1 and over half of those accepted for transplantation required procedural intervention prior to listing. Many patients have intestinal failure at the time of referral. We hypothesise that long-term use of hypertonic fluids may partly account for the poor results described and that patients with intestinal failure should be considered for regular dental review and timely intervention.

P139 - Clinical characteristics of 16 cases of gastroschisis accompanied at a reference hospital in northeastern of Brazil.

Michela Marmo, Jessika Costa, Dayle Vasconcelos, Suelenn Menezes, Camila Dantas, Manuela Camara-Lins, Mara Alves, Georgia de Paula, Paloma Velez

Instituto de Medicina Integral Professor Fernando Figueira - IMIP

Introduction: Gastroschisis is a rare congenital abdominal wall defect. Other associated malformations are not common occurring in 5-35% of cases. However, these patients can present infections, failure to thrive and intestinal failure. This study described a group of patients with gastroschisis and their evolution until hospital discharge born at a reference hospital of Northeastern of Brazil.

Method: this is a study of case series of 16 pediatric patients with a diagnosis of gastroschisis accompanied at a reference hospital during a year (2017-2018). The birth weight, surgical approach (in one or two times with silo use), associated malformations, parenteral nutrition time (considered to be prolonged when greater than 34 days), time to onset of enteral diet (considered early if less than or equal to 7 days), presence of neonatal cholestasis, confection of ostomy, intestinal resection, and death were evaluated. The data evaluated were selected according to the characteristics and risk factors already described at the literature.

Results: sixteen children were evaluated and 6/16 (37.5%) patients had birth weight less than 2,500 g; 11/16 (68.7%) had single-time surgical correction; 3/16 (18.7%) had other associated malformations (2 intestinal atresias, polydactyly). All patients required prolonged parenteral nutrition (NP); 13/16 (81.2%) had onset of diet in more than 7 days of life. The mean time of central venous access were 26.8 days and intestinal atresias, polydactyly). All patients required prolonged parenteral nutrition (NP); 13/16 (81.2%) had onset of diet in more than 7 days of life. The mean time of central venous access were 26.8 days and intestinal resection, and death were evaluated. The patients presented similar characteristics to those described in the literature and observed few hours after the surgery correction. The patients presented similar characteristics to those described in the literature and required multidisciplinary team and specialized care for a long period of time.

Conclusion: The most patients described had presented adequate birth weight and surgical repair in a single time without silo utilization. Associated malformations were not frequent. All patients used NP for a long time and they had had late onset of diet. Two deaths were observed few hours after the surgery correction. The patients presented similar characteristics to those described in the literature and required multidisciplinary team and specialized care for a long period of time.

P140 - Impact of residual bowel length on cost of home parenteral nutrition for short bowel syndrome

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2 Department of Medical Gastroenterology, Rigshospitalet, Copenhagen, Denmark
3 Intestinal Rehab & Transplant Program, Mount Sinai Medical Center, New York, New York

Introduction: Key determinants of costs associated with home parenteral nutrition (HPN) among patients with short bowel syndrome (SBS) are unknown. The aim of this analysis was to examine how charges associated with HPN relate to residual bowel anatomy.

Methods: We conducted a retrospective cohort analysis of medical history and charges for patients with SBS treated at a large Danish referral center. Clinical characteristics for SBS patients from 2006-2016 were obtained from a longitudinal database (LD). Charges assigned to patients for transport, PN, drugs, admissions, and ambulatory care visits were available from hospital databases for the year 2011. The LD contained 210 patients, while the 2011 sample contained a subset of 105 patients. For this analysis, we focused on patients with jejunoileostomies (2011 n=79). Charges are reported in Danish Kroner (DKK). We used linear regression to assess the relationship between anatomy (length of remaining bowel in cm) and 2011 charges.

Results: Patients in both the 2011 cohort and LD averaged 6.5 days of HPN per week. The 2011 sample had an average of 283 HPN days per year (275 in LD), and an average of 5.27 years since HPN initiation (5.9 years in LD). Patients in 2011 received an average 2690.6 mL of HPN fluids per day (2803.8 mL in LD). Sample characteristics were similar across demographic and disease characteristics when comparing 2011 patients to the LD of patients in the 10-year cohort. Among patients with jejunoileostomies and ileostomies, those with the most severe SBS had higher charges compared to those with greater length of residual bowel. Linear regression demonstrated a significant relationship (b=-82657, p<0.001) between anatomy type and total annual charges, with decreasing remnant bowel length resulting in increasing charges.

Conclusion: This study aimed to evaluate costs associated with caring for SBS patients while considering 5 main factors - HPN, hospital admission, ambulatory care visits, transport, and drugs. Our data indicate a linear inverse relation between residual bowel anatomy and costs of care. The potential impact of PN complications and their relation to anatomy merits further study. Improved understanding of these costs will allow more nuanced cost benefit analysis of emerging novel treatments that may allow some patients to be partially weaned off PN.
Double balloon enteroscopy reliably directs assessment for patients with short bowel syndrome.

Andre Lee 1, Adriana Safatle- Ribeiro 1, Flavio Galvao 1, Rafael Pinheiro 1, Mariana Hollandia Rocha 1, Daniel Waisberg 1, Vinicius Rocha Santos 1, RUBENS Macedo 1, Wellington Andraus 1, Lucas Lee 2, Luiz Augusto C. D’Albuquerque 1

1 Department of Gastroenterology
2 Medical Student of Santos- Lusia Medical School

Aim: To evaluated patients with short bowel syndrome(SBS) for determining extension of remnant intestine and aspect of the intestinal mucosa

Methods: We retrospectively reviewed 20 patients who are at least 12 months in Home Parenteral Nutrition (HPN), included type III of intestinal failure characterized chronic condition, metabolic stable Of these patients divided in three groups: Nine had ultrashort bowel syndrome (USBS) characterized type I of SBS, nine had type II of SBS and two presented "no gut syndrome" We considered type I, patients with end-terminal jejunostomy and complex intestinal fistula as the same group. In case of the intestinal dilatation we collected intraluminal fluid for culture to diagnose bacterial overgrowth

Results: The nine patients SBS type I group, the enteroscopy show us in the proximal intestinal mucosa with normal aspect and distal stump of the closed transverse colon, show severe ischemic colitis, except in one patient with mucosal fistula of colon The other nine patients of SBS type II group was observed intestinal stretching and dilatation, which caused difficulty in evaluating the length of the remnant intestine. In this group we diagnosis four cases of bacterial overgrowth and two cases of anastomotic ulcers in jejunum/ transverse colon For the two cases of “no gut syndrome” they have the fourth duodenal stumps closed with gastrostomy, we just observed the duodenal dilatation with some food waste

<table>
<thead>
<tr>
<th>Table 1. Sample Characteristics</th>
<th>2011 (Ind, All Patients, N=111)</th>
<th>2006-2015, Ned.0</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean (SD)</td>
<td>Mean (SD)</td>
</tr>
<tr>
<td><strong>Demographics</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age (yrs)</td>
<td>51.5 (16.3)</td>
<td>51.3 (16.6)</td>
</tr>
<tr>
<td>BMI (kg/m²)</td>
<td>22.8 (3.5)</td>
<td>22.5 (3.9)</td>
</tr>
<tr>
<td>Time since HPN initiation (yrs)</td>
<td>6.2 (4.8)</td>
<td>5.5 (4.3)</td>
</tr>
<tr>
<td><strong>Peritoneal Characteristics</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Remaining Bowel Length (cm)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intestine (cm)</td>
<td>16 (9-9.0)</td>
<td>19.9 (10.6)</td>
</tr>
<tr>
<td>Colon (%)</td>
<td>14 (6-15.9)</td>
<td>14.4 (15.6)</td>
</tr>
<tr>
<td><strong>EPI</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>EPI Bays/Week</td>
<td>6.5 (1.3)</td>
<td>5.5 (1.2)</td>
</tr>
<tr>
<td>Average HPN volume (ml)</td>
<td>269.5 (138.3)</td>
<td>280.1 (181.4)</td>
</tr>
<tr>
<td>Average HPN glycemia (mg)</td>
<td>4943.5 (2795.6)</td>
<td>3092.2 (1962.0)</td>
</tr>
</tbody>
</table>

2011 Charges (in On)

- Total W11 charges: 609445.80 (2567147.90)
- Total submissions charges: 10896.40 (350691.10)
- Total outpatient care charges: 74529.20 (95453.60)
- Total HPN charges: 13121.30 (317828.42)
- Total drug charges: 41359.49 (146611.70)
- Total transport charges: 106754.74 (258591.25)

Table 2. Total charges (Ons) assigned to patients in 2011 (includes EPI, Admissions, Anesthetics, Meals, Transportation, Drugs)
Conclusion: Double balloon enteroscopy is a safe, reliable modality for determining changes in the intestinal mucosal and monitoring the intestine adaptation in SBS. This technique can be used to direct selective surgical reconstruction of intestinal transit in pre-operative time.

P142 - Nutritional, metabolic and hepatic impact associated with HPN use on stable patients undergoing Home Parenteral Nutrition (HPN)

Andre Lee, Maria Hollanda Martins da Rocha, Maria Carolina Dias, Tatiana Rana, Lara Lordello Melo, Gabriela Oliveira Lemos, Yasmin Syagha, Flavio Galvão, Dan Waltzberg

Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo - HCFMUSP

Intestinal Failure is a severe condition defined as the reduction of gut function below the minimum necessary for the absorption of macronutrients and/or water and electrolytes, such that intravenous supplementation is required. Patients with Chronic Intestinal Failure despite being metabolically stable patients, require intravenous supplementation over months or years.

Objectives: The aim of this study was to describe the nutritional, metabolic and hepatic impact associated with HPN use on sixteen adult patients treated at an outpatient clinic of intestinal failure (AMULSIC – Multidisciplinary Ambulatory of Short Bowel Syndrome) located in a public tertiary referral hospital in São Paulo, Brazil. The patients were stable and undergoing Home Parenteral Nutrition (HPN) for at least 12 months.

Methods: Patients were evaluated according to criteria for liver disease progression related to long term parenteral nutrition therapy. The parameters utilized were: elastography (fibroscan), liver biopsy, laboratory tests (AST, ALT, alkaline phosphatase, γ-GT, ferritin, bilirubins, activated partial thromboplastin time, total proteins and fractions), body mass index (BMI), body composition by electrical bioimpedance and also by indirect calorimetry for the accurate determination of individual caloric demands.

Results: Sixteen patients with a mean age of 38.5 years diagnosed with chronic intestinal failure undergoing HPN were followed up for at least 12 months and a maximum of 144 months (mean of 53 months). Fibroscan analysis revealed liver fibrosis grade I in nine patients, grade II in three patients and grades III/IV in four patients. Two patients presented grade III fibrosis in liver biopsy. Eleven patients had excessively elevated levels of ferritin. Eleven patients present a normal range of weight, three patients presented a BMI ≥ 24 (overweight) and two patients presented BMI ≤ 18.5 (underweight). Ten patients out of this group showed muscle mass reduction in electrical bioimpedance. Indirect calorimetry had a 0.8 (SD: ± 0.079) mean respiratory coefficient.

Conclusions: In the population studied, there was no direct correlation between fibroscan results and liver biopsy and there was not a home parenteral nutrition time direct correlation with hepatic impairment. It was also observed an association between parenteral nutrition associated liver disease and high levels of ferritin, previous history of morbid obesity and intestinal pseudo-obstruction.
P143 - Use of transient elastography to determine liver fibrosis in pediatric intestinal failure

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2 Transplant and Regenerative Medicine Centre, The Hospital for Sick Children, Toronto, Canada
3 Division of Pathology, The Hospital for Sick Children, Toronto, Canada
4 Division of Gastroenterology, Hepatology and Nutrition, The Hospital for Sick Children, Toronto, Canada
5 Division of General and Thoracic Surgery, The Hospital for Sick Children, Toronto, Canada

Introduction: Intestinal failure associated liver disease (IFALD) is a significant complication in pediatric intestinal failure. Liver fibrosis with or without cholestasis is a frequent occurrence in IFALD. However, the diagnosis of fibrosis in IFALD is challenging and currently limited to liver biopsy. Our objective was to evaluate the use of transient elastography (TE) in pediatric IF patients to determine its efficacy for monitoring liver fibrosis as an alternative to liver biopsy.

Methods: A retrospective cohort study of IF patients between January 1, 2015 to December 31, 2017. The study cohort included a sample of patients who had a routine liver biopsy during an operative procedure and completed transient elastography at the same time. Liver biopsies were evaluated for stage of fibrosis using the modified Scheuer score. Statistical analysis included univariate analysis and Chi-square methods. High (grade 3-4) versus low (grade 0-2) fibrosis and TE scores were compared to determine sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV). Receiver operating characteristic (ROC) curves were also used to determine the ability of TE to discriminate various stages of fibrosis. Data is presented as medians with interquartile ranges and frequencies.

Results: 30 patients (21 male [70%]) with a median age at biopsy of 320 days (154-1776) were evaluated. Majority of patients had an etiology of abdominal wall defects (24[40%]) and necrotizing enterocolitis (9[30%]). Median length of PN therapy was 190 days (120-789). All TE measurements were completed within a median of 8.5 days (5.0-13.3) of the liver biopsy. Individuals with histologic fibrosis scores of 0-1 (n=8) had a median TE score of 4.9 (3.9-7.2), fibrosis scores of 2 (n=5) had a median TE of 6.7 (4.4-11.4) and those with a fibrosis score of 3 (n=7) had a median TE of 12.1 (6.3-14.6). No patients had grade 4 fibrosis. Dichotomous evaluation of the fibrosis scores (low vs high grade) compared to the chronic cholestatic scale for Fibroscan demonstrated 95.7% sensitivity, 57.1% specificity, 88.0% PPV and 80.0% NPV for low grade fibrosis. The c-statistic for the ROC curve was 0.764.

Conclusion: Fibrosis in patients with IF is often difficult to assess without liver biopsy. TE has only been evaluated in small cohorts but shows potential as a non-invasive method to monitor low vs high grade fibrosis in children with IF. Further studies in a larger cohort are required.

P144 - Exploration of factors impacting on eating in paediatric intestinal transplant recipients: a mixed methods study

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Introduction: The transition to eating after intestinal transplant (IT) can be difficult for some children. Children may have had little experience of eating before IT. Studies suggest that disruptions to the process of learning to eat in early childhood may be implicated in eating difficulties but there is no empirical data in IT recipients regarding factors associated with this. Identifying factors that may influence eating after IT could help manage caregiver expectations and plan targeted interventions. The aim of this study was to describe the nutritional intake and eating behaviours of IT recipient children and to explore factors impacting on eating.

Methods: This mixed methods study consisted of quantitative self-completion questionnaires and a three-day food diary followed by a semi-structured telephone interview. Caregivers of the entire UK paediatric IT population were invited to participate. The questionnaires included the Children’s Eating Behaviour Questionnaire and 26 demographic items. Analysis was by descriptive statistics using SPSS.

Results: The survey response rate was 26% (n=9) with 89% (n=8) of respondents also being interviewed. Of the nine respondents, two were discharged on an exclusive oral diet following IT with three able to wean off tube feeding after between one and five years. Median energy intake was 93% (range, 61-137) of the Estimated Average Requirement and 56% (n=5) were categorised as food avoidant. Transitioning to an oral diet after IT without home tube feeding was statistically significantly associated with complementary foods being introduced at the recommended age, learning to eat, having significant practice and positive eating experiences before IT. The qualitative data supported this as well as providing explanations in relation to three themes: medical, caregiver and child influences.

Conclusion: The study findings present a picture of a complex interplay of factors that can influence eating in IT recipients. This study, although small, provides the first empirical evidence of an association between pre and post-IT eating. The findings suggest that promoting pre-IT eating may be beneficial and indicate that there may be predictors for eating difficulties that could be used to facilitate targeted interventions - further research is required.
**P145 - Mycotic aneurysm after Liver and Small Bowel Transplantation.**

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2 Dept of Radiology, Addenbrooke’s Hospital
3 Dept of Gastroenterology, Addenbrooke’s Hospital

Mycotic aneurysm is a much-feared complication of vascular surgery. Management in the non-transplant situation often utilises placement of an endovascular prosthetic graft and long-term antibiotics. In formally immunosuppressed patients the ongoing likely infection of the prosthesis is of great concern.

We describe the case of a 29-year-old recipient of a liver small bowel transplant who developed a mycotic aneurysm of the donor aortic conduit two months after transplantation.

She had undergone an isolated liver transplant at the age of two for alpha 1 antitrypsin deficiency. This was complicated after transplantation by a volvulus of her small bowel, leaving her with 30 cm of jejunum and dependent on parenteral nutrition till the age of 13. Graft failure, 27 years later, necessitated the need for re-transplantation and at this stage a liver small bowel transplant was performed.

Four weeks post-transplant, she developed a pyrexia of unknown origin and at that stage cross sectional imaging was unremarkable. This was repeated two weeks later and still no source of sepsis could be determined. A further CT two weeks later demonstrated a 2 cm mycotic aneurysm in the mid portion of the donor aortic conduit, (Image 1).

Initial management involved endovascular stenting and antibiotics to gain immediate control, followed by a definitive surgical intervention. A third-party donor thoracic aortic conduit was acquired (blood group compatible) and at laparotomy the infected graft and stent were removed and replaced with the new conduit. Culture of the stent and aortic tissue grew vancomycin resistant enterococci (VRE) and Candida glabrata, necessitating six weeks of antimicrobial treatment (linezolid and liposomal amphotericin).

Five and a half weeks after initiation of linezolid therapy the patient developed profound lactic acidosis and severe pancreatitis requiring ICU admission and haemofiltration for the symptomatic lactic acidosis. Following discontinuation of linezolid, the pancreatitis and lactic acidosis resolved.

She is now four months after the vascular reconstruction and is well with no evidence of recurrence of the aneurysm (Image 2) and no recurrent VRE.

She has developed CMV disease from the third-party tissue. This is an important consideration when considering matching for third party tissues and prophylaxis must be reviewed.
P1.46 - Home Parenteral Nutrition: training of the adolescents
Catherine Poisson, Amelia Rocha, Christelle Alliot, Julie Olivier, Isabelle Eicher, Cécile Talbotec, Cécile Lambe, Olivier Goulet
Home-Parenteral Nutrition Division. Hôpital Necker Enfants Malades; Université Paris-Descartes

The Necker-Enfants Malades Hospital Home Parenteral Nutrition (HPN) program was certified in 1984. It is the largest in France with ~40% of the 380 children currently on HPN. More than 800 infants and children have been enrolled in the Necker program since 1984. In 2018, it supported 156 children (age range: 3 months - 7 years): short bowel syndrome (SBS) (n=78), congenital enteropathy (CE) (n=28), total intestinal aganlionosis (TIA) (n=22); miscellaneous (n=17), > 60% presenting with irreversible intestinal failure (IF): In order to prepare the transition to adulthood and to offer more freedom for leaving the family nest, we train the adolescents (ado) for self cares. In that view, we designed a training program.

Methods: Training involves nurses, MD, pharmacists, psychologist, social worker.... It requires time for the team as well as for the adolescent. It is organized during the school vacation in 3 steps each lasting 5 days. Step 1: Line disconnection & illness knowledge; Step 2: Line connection & body control; Step 3: CVC dressing & information on adult transition. Training also includes an approach focused on their physical and psychological well-being to understand their body and their illness.

Results: In 2018, Z adolescents (age range: 13-17 years) were trained: SBS (n=2), CE (n=2), TIA (n=2) and CIPPOS (n=2). Children received PN 3 to 7 nights a week with bags from the french provider Baxter-Faconnable. At the end of the training program, 4 ado can be considered as fully autonomous on daily basis, 2 are partially (for disconnection only) and 1 failed to finish the training program.

Conclusions: Autonomy is achieved for the majority. However, gaining autonomy does not mean accepting illness or being ready to move into the adult world. It seems that this multidisciplinary approach and the effect of group teaching are beneficial to them. We do continue to adapt the program according to the ado feeling and feedback.

P1.47 - Evaluation of central venous catheters in hospitalized pediatric patients with intestinal failure from a brazilian gastroenterology reference center
Vanessa Scheeffer, luiza nader, caroline dias, Raquel Cechinel, Cintia Steinhaus, Carolina Soares, Melina Melere, Matias Epifanio, Cristina Targa
Hospital da Criança Santo Antonio, Porto Alegre, Brazil

Introduction: Intestinal Failure (IF) is a debilitating condition, defined as the need for parenteral Nutrition (PN) for more than 60 days. One of the most important IF complication is central catheter-associated bloodstream infection (CCABSI). The Taurolidine line prophylaxis seems to be a promising measure for the prevention of these infections.

Materials and Methods: retrospective study, evaluating 89 catheters used by 38 pediatric patients with IF from 2015 to 2017 at a center in the Southern Brazil.

Results and Discussion: Most patients (61%) had short bowel syndrome, and prevalence of CCABSI was higher in this group of patients (P=0.036). Of the 38 children studied, 14 (77.8%) had at least one CCABSI. A total of 89 catheters were used by these children - mean of 4.95/ patient. CCABSI occurred in 27 catheters (30.3%). 66.3% of the total number of catheters were non-tunneled and 32.6% were tunneled. 35.6% of the non-tunneled CVC had CCABSI compared to 20.7% of the tunneled (p=0.289). 34.8% of the catheters were removed due to mechanical problems and 18% were removed due to CCABSI. The frequency of CCABSI was 5.7 per 1000 catheter days. The taurolöine line prophylaxis has been used in our hospital since march/2016 and this paper included patients before this period. We found a statistically significant association between CCABSI and death (p=0.01). Patients with at least one CCABSI needed to change the CVC more often (x= 4 catheters) than children that did not presented infection (x= 2 catheters) (p=0.035). The use of taurolöine improved catheter survival from 20 days to 26 days (p=0.032).

Conclusions: CCABSIs was the main cause of death in this group of patients and frequency of CCABSIs is still high among our population. The use of taurolöine helped to increase the days that the CVC remained in the patient, but more data is necessary to correlate this lock with an improvement in infection rate in our population.
There are limited case reports regarding patients who underwent intestinal transplant after HCT. Clinical evidence suggests to rule out subclinical infection after intestinal transplant in patient with immunodeficiency. To our knowledge, this is the only reported successful case of a patient who received HCT followed by ITx. Despite not having a perfect HLA match between the HCT and intestinal donors, the patient achieved proper immune tolerance with the need for very low level immunosuppression and no significant GVHD.

**Methods:** She received a conditioning regimen of hydroxyurea, alemtuzumab, fludarabine, melphalan and thiopeta, with GVHD prophylaxis of tacrolimus (TAC) and mycophenolate mofetil (MMF). One month after HCT, MMF was discontinued. She received a 6/6 antigen-matched unrelated umbilical cord blood transplant in May of 2013. Nineteen months later, she received a 3/6 HLA matched intestine and colon allograft from a deceased donor. She was maintained on TAC and steroids, and also received basiliximab.

**Results:** One month post-HCT, she had ~50% donor cells in her blood. Her TAC dose was decreased and she received a cryopreserved fraction of her umbilical cord blood graft, with significant improvement. She developed acute GVHD limited to the skin, which was successfully treated with topical creams. Six weeks following her ITx, she was found on surveillance endoscopy to have mild ACR. This resolved with steroids, and also received basiliximab.

**Conclusion:** To our knowledge, this is the only reported successful case of a patient who received HCT followed by ITx. Despite not having a perfect HLA match between the HCT and intestinal donors, the patient achieved proper immune tolerance with the need for very low level immunosuppression and no significant GVHD.
CIRTA 2019 Book of Abstracts - Posters

P150 - Financial hardship faced by our patients and their families Road to Recovery
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Introduction: Many advances have been made in the field of intestinal/multivisceral transplantation over recent years and favourable outcomes are now the expectation, rather than the exception. Despite this, life after small bowel/multivisceral transplantation can be unpredictable. Healthcare professionals often reflect on the physical and psychological ‘costs’ of undertaking this treatment, but rarely consider the financial costs involved.

Methods: Survey of a sub-group of post-transplant patients and their families for estimate of annual personal cost of transplant follow-up care, in a single UK centre performing adult intestinal transplants.

Results: Having surveyed a sub-group of 9 patients we have interesting data which identifies that our patients travel significant distances and this has wide and varied travel costs to enable them to receive the required follow up in our hospital. This data doesn’t reflect parking charges on site. Some patients/relatives stay in accommodation which is funded which varies from £20 per night to £55 per night if on site. Other accommodation would be considerably more and an average in our area would be around £100 per night.

Conclusion: Some patients have not been able to work pre transplant and some cannot regain work post-transplant. This is partly due to physical problems, such as fatigue, difficulty concentrating, tremor. For many, the unpredictability and intensive surveillance requirements are also an anticipated or actual barrier. As a national service extending across the UK, our patients travel notable distances to attend follow-up and the cost of fuel and cost of living make coming here challenging. To mitigate some of these costs, we offer on-site accommodation wherever possible, signpost patients towards charitable funds or organisations that can assist. We as specialist nurses write supportive letters for our patients to receive Personal Independence Payments/Employment Support Allowance(s).

Financial hardship is a real concern for many transplant patients and assistance/support with this should be part of holistic follow-up care, within the limitations of national social support agencies.

P151 - Five year survival of the first pediatric multidisciplinary intestinal rehabilitation program with home parenteral nutrition in the public health system in Brazil
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Introduction: Data on multidisciplinary programs dedicated to the care of intestinal failure (IF) patients on home parenteral nutrition (HPN) in middle-income countries from Latin America are scarce. This study described the results of the first Pediatric Multidisciplinary Intestinal Rehabilitation Program with HPN in the public Health System from a public tertiary hospital in Brazil.

Methods: We conducted a retrospective study that included all children who were referred to our institution with IF from 1-January-2014 to 31-January-2019. Inclusion criterion was the use of parenteral nutrition (PN) for at least 2 months while in stable condition. We assessed the medical records of participants who were discharged while receiving HPN and those who were not discharged and received PN whilst in hospital. All patients who were not discharged on HPN had psychological or social impairments or did not reach a stable clinical condition. The following outcomes were analyzed: total PN period (HPN or PN in hospital period), full PN weaning off and causes of death.

Results: 49 patients were included, median age at onset of long-term PN was 4 months old (17 days - 16 years), 35 (71.4%) were girls and 38 (77.6%) had short bowel syndrome, of whom 9 had remnant intestine <20 cm. IF causes were: intestinal atresia 36 (73.5%); volvulus 9 (18.4%); gastroschisis 7 (14.3%); necrotizing enterocolitis 6 (12.2%); Hirschsprung’s disease 2 (4.1%); pseudo-obstruction syndrome 2 (4.1%); other causes 5 (10.2%). 35 (71.4%) were discharged receiving HPN, of whom 9 weaned off HPN, 23 are currently on HPN (1 patient is on waiting list for multivisceral transplant) and 3 died (1 catheter-related bloodstream infection and 3 death from causes other than intestinal failure). Median period of HPN was 13.9 months (3 days - 4.5 years). From the 14 patients who were not discharged from hospital, 7 weaned off PN, 2 are still receiving PN, and 5 died (3 loss of venous access and 2 CRBSI). Total weaning off PN rate was 32.7%. 5-year survival rate of all patients was 81.4% whereas 5-year survival on HPN was 90.4% (Kaplan-Meier).

Conclusion: Treatment of patients with IF by a multidisciplinary rehabilitation program with HPN is feasible and safe in the public health system in Brazil. The high survival rate supported the use of HPN as the primary treatment for IF.
P152 - Extended Hirschsprung’s Disease: Our Experience And Literature Review

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Introduction: Extended Hirschsprung’s disease (EHD) is a rare condition leading to intestinal failure (IF). Its impact in Latin America (LATAM) is unknown. The aim of this study is to review our experience and published evidence.


Results: N=5. Median age 42.2 m (r:4 -108). Mean RIL: 54.6 cm (r:30 -80). It took 3.2 surgical procedures (r:2 -4) to perform a functional enterostomy. The distal aganglionic bowel was preserved in all cases. 5/5 with IF and mean parenteral nutrition (PN) dependance time of 46 months (r:30-114) . All received enteral nutrition by oral feeds and gastroclisis in 2 cases. 3 patients were listed for intestinal transplantation (ITx), 2 have already received an ITx. Follow up: 2.16 years (r: 0.75-4.25). 80% (4/5) overall survival on home PN, 1 death 9 months after ITx due to PTLD (off PN).

Literature review: 127 articles. No publications from LATAM. 120 excluded: 6 duplications, 114 do not fulfill inclusion criteria. 7 remaining articles, N 47: 0.49 mo.; mean 4.9 years follow up (r:2.67-7.3); 3.57 procedures (r:14-5.75) until reaching an accurate diagnosis; RIL: 44.7 cm (32.2-60); 7 had aganglionic bowel resection; 66% survival; causes of death: 3 after ITx, 1 listed for ITx, 11 due to PN complications. 18/47 patients received an ITx.

Discussion: EHD is an uncommon disorder leading to IF, and its prevalence in our continent is unknown. Our outcomes are similar to those published elsewhere. Home PN provides acceptable medium-term survival, although most patients will need an ITx in the long term.

P153 - Are Femoral Tunnelled Central Venous Catheters safe to use for HPN ?

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Background: Patients with Intestinal Failure (IF) require reliable intravenous access for provision of Parenteral Nutrition. Venous access for Home Parenteral Nutrition (HPN) patients with Type 3 IF can be compromised due to thrombosis of deep veins. Preferred choice of veins for central venous catheter are the supra cardiac veins. Femoral tunnelled CVC are avoided due to concerns of catheter related blood stream infection (CRBSI) and are considered as a last resort when all other thoracic CVC access is exhausted. We assess the outcomes of tunnelled femoral catheters in our cohort of HPN patients.

Methods: We did a retrospective analysis of a prospectively collected data of all HPN patients and venous access from January 2013 to December 2018 managed at a National HPN Unit. The details of venous access, complications of CVC, sex and details of stoma extracted from database.

Results: In a cohort of 9 HPN patients with tunnelled femoral CVC. 6 were females and 3 were males leading to a total of 84 36 days. A total 3 episodes of CRBSI were recorded in 1 patient. All of the 3 episodes (methicillin - sensitive Staphylococcus aureus (MSSA), Escherichia Coli and CNS) were successfully salvaged. The average no of days for femoral CVC was 935 days. The rate of CRBSI was 0.35 per 1000 catheter days. 8 patients had stoma or open abdominal wound. There were no episodes of ipsilateral femoral DVT. The CVC was replaced for 2 patients due to fracture of CVC.

Conclusion: Tunnelled femoral CVC CRBSI rate ( 0.35)is comparable to thoracic veins CVC CRBSI rate(0.31) in our unit. Furthermore femoral access could be considered as a reliable access to provide HPN. It may be considered as an option if more than 2 thoracic deep veins are occluded to prevent complete occlusion of SVC. We feel tunnelled femoral CVC is a safe option to consider for HPN.
P154 - Global gut dysmotility: Patient characteristics and unique clinical features
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Introduction: The pathophysiology and biogenetics of global gut dysmotility has yet to be fully defined. This abstract highlights the results of an observational study that is conducted at a tertiary center with high referral volume. The aim of this study is to define the clinical, neuropsychiatric, and socioeconomic status of this unique patient population.

Methods: A total of 175 patients were referred to our program with global gut dysmotility for possible surgical management including reductive surgery and/or transplantation. The diagnosis of global gut dysmotility was made by exclusion of mechanical bowel obstruction, capsule endoscopy, and sitz marker testing.

Results: Most patients were female adults with a total mean age of 36 ± 14 (range: 5 -70). The medical and surgical histories were significant for oral intolerance, constipation, abdominal pain, orthopedic procedures and cosmetic surgery. There was a common association between the disorder, bariatric surgery, and congenital anomalies. The major neuropsychiatric disorders were anxiety (70%), depression (64%), behavioral disorders (35%), abuse (32%), PTSD (15%), suicidal ideations (13%), and bipolar disorder (7%). The syndrome was associated with autonomic dysfunction (72%), autoimmunity (19%), and mitochondrial disease (3%). Most of the socioeconomic milestones including marriage, education, occupation, and daily activities were severely impaired.

Conclusion: The described herein clinical features may be a valuable tool to assess the spectrum of the disorder, guide therapy, and selection criteria for transplantation.

P155 - Does BMI Correlate with Enteral Autonomy Post Intestinal transplant?
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Introduction: Intestinal transplant recipients often require total parenteral nutrition (TPN) support in the post-operative period until they achieve enteral autonomy. The time to enteral autonomy is a meaningful clinical outcome and indicator of successful graft function. Further, minimizing TPN exposure in the heavily immunosuppressed patient may mitigate infectious risk. TPN dependence after intestinal transplantation is variable and nutritional status at the time of transplant may be a determinant of the duration of TPN dependence. Body Mass Index (BMI) is one surrogate of nutritional status in the adult population. The aim of this study was to assess how BMI at the time of transplant impacts the duration of TPN after transplant.

Methods: Adult patients who received an intestinal transplant at Georgetown University Hospital from 2006 to 2018, and survived beyond one year after transplant were retrospectively identified. BMI at the time of transplant was categorized based on current definitions (<18.5 indicates malnutrition, 18.5 -24.9 healthy weight, 25 -29.9 overweight, and >29.9 obesity), and compared to duration of TPN dependence in days.

Results: 105 adult patients transplanted between 2006 and 2018 were included in the analysis. 51 of the patients were male and 54 were female. Median age was 43 years (range 18 -66). Across all BMI categories the average days of TPN was 19.9 ± 12.6 days. Patients with a higher BMI at the time of transplant had the shortest duration of TPN post-transplant. Patients with a BMI of >30 (n= 11) had a mean of 16.1 ± 7.2 days; patients between 25 -29.9 (n = 21) had an average of 18.9 ± 9.2 days; patients between 18.6-24.9 (n = 63) had an average of 20.6 ± 14.8 days; lastly, patients <18.5 (n = 10) had an average of 23.9 ± 8.7 days. There was a significant difference in duration of TPN in days between the <18.5 BMI and >30 BMI category (p=0.05).

Conclusion: Achievement of enteral autonomy post-intestinal transplant is multifactorial. It is well established that post-transplant surgical and immunological complications are important factors that play a role in determining when TPN can be successfully weaned. Pre-transplant factors, however, may also contribute to enteral autonomy, such as recipient BMI at the time of transplant. Transplant practitioners should assist patients in avoiding a <18.5 BMI pre-transplant in order to potentially decrease the duration of TPN dependence post-transplant.
P156 - Colonic continuity and Incidence of weaning from PN by 5 years of age in children with onset of intestinal failure (IF) related to short bowel syndrome (SBS) by 3 months of age with chronic persistent dependence on parenteral nutrition (PN) at one year of age

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Objective: To review 5-year outcome of children presenting in infancy with chronic intestinal failure (IF) associated with short bowel syndrome (SBS) who were still on parenteral nutrition (PN) treatment at 12 months of age.

Method: Medical records of patients with SBS related IF presenting in neonatal period and up to 3 months of age from 2007-2017 who were still on PN at 12 months of age were reviewed. Data obtained included small intestinal length at time of resection, aetiology, presence/ absence of ileo-caecal (I-C) valve and/or colon and number of patients still on PN at 5 years.

Results: Twenty patients were identified. Eleven were male and 9 female. 14/20 were born prematurely. Diagnoses were necrotising enterocolitis (NEC) in 9, volvulus in 4, long segment Hirschsprung’s disease in 3, gastroscisis in 2, and intestinal atresia in 2. The remaining small intestinal length at time of surgery ranged from 4.5cm - 120cm (median 30cm) in 19 cases (not measured in 1). Four patients had ultra SBS (length <10 cm). Eleven patients weaned from PN by 5 years of age and 4 were still on PN. The median age at time of weaning (corrected gestational age in premature infants) was 2 years 4 months (range 1 year 6 months - 5 years 9 months). Small intestinal length ranged from 7-120 cm (median 30 cm) in the weaned group and from 4-90 cm (median 20 cm) in the persistent IF group. Seven/11 patients who weaned and 2/9 still on PN were female (p=0.09 not significant). 8/11 who weaned and 2/9 still on PN had NEC (p=0.068, not significant). 8/11 who weaned had an intact I-C valve compared to 3/9 on PN (p=0.026, significant). 10/11 or 91% with colon present compared to 5/9 who remained on PN (p=0.026, significant). 11/18 or 61% with colon in continuity weaned.

Conclusion: Over 50% (11/20) infants with SBS and PN dependence for >12 months weaned from PN by or at 5 years of age. There was a statistically significant association with presence of colon with 61% with a colon weaning and even with some cases of ultra-SBS. Larger studies are needed to gain other predictors of weaning in this diverse group of patients.

P157 - Severe Late Onset Acute Cellular Rejection In a Pediatric Patient with isolated small intestinal transplant rescued with aggressive immunosuppressive approach

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Introduction: Small intestinal transplantation has been the standard treatment modality for pediatric patient with intestinal failure who failed other surgical and medical treatment. At its current stage, it carries its own risks including but not limited to acute and chronic cellular rejection, graft malfunctions, development of PTLD, viral and bacterial infections. Acute cellular rejection of the intestinal graft is an important and major complication secondary to its rich lymphatic supply. It is usually results from sub therapeutic immunosuppression or non-adherence to medical management. The current management includes modifying the immunosuppression to achieve therapeutic level. Late severe acute intestinal allograft rejection is associated with increased risks of sepsis, bleeding and in the majority of cases with graft loss that ends up in graft enterectomy.

We present a case of a 20 year old patient who underwent isolated small bowel transplant for complete intestinal Hirschsprung Disease at age 7 years old, but due to medication non-adherence developed severe late-onset acute cellular rejection manifested by large ostomy OP, fever, weight loss. Underwent ileoscopy on presentation that showed complete loss of normal anatomical intestinal landmarks, bleeding, and ulcerated mucosa. Graft biopsies showed ulceration and granulation tissue with severe architectural distortion, and rare residual consistent with severe intestinal graft rejection. She initially received pulse doses of Intravenous corticosteroids and increased dose of tacrolimus without significant improvement. Her immunosuppression plan was escalated to include infliximab and finally had Antithymocyte globulin (ATG). Graft enterectomy option was entertained frequently during the treatment course; however clinical improvement was noted with evidence of histological improvement and salvage of the graft.

The aggressive anti-rejection treatment was complicated with the development of monomorphic, plasmacytoma (PTLD) that was managed with modifying her immunosuppression. Now her graft function is maintained on tacrolimus, oral prednisone, and routine Remicade infusion.

Conclusion: 1-Small bowel graft rejection is associated with increased morbidity and mortality, increased risks of sepsis, bleeding and graft loss. 2-We believe that prompt and aggressive immunosuppressive approach significantly increases the chance of rescuing small bowel transplant rejection.
P158 - Significance of the Extent of Intestinal Resection on the Outcome of a Short-bowel Syndrome in a Porcine Model.

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Background: Insufficient data are available to determine the most suitable extent of intestinal resection required to induce short-bowel syndrome (SBS) in pigs. This study aimed to compare the three main SBS-models published.

Methods: A 75%, 90%, or 30% mid-intestinal resection was performed in groups of n = 5 pigs each. Clinical (body weight, stool consistency) and biochemical (serum electrolytes, citrulline, albumin, prealbumin, and transferrin) parameters were determined daily, functional (D-xylose resorption) and histological (intestinal villus length) parameters were determined after 2 weeks. A t-test and ANOVA were used for statistical analysis.

Results: Only in the 30% group, we observed a persistent weight loss (13.6 ± 3.8%) and diarrhea, as well as a decrease in prealbumin-levels (41%) and transferrin levels (33%). Serum electrolytes remained stable in all groups during the observation period. Citrulline stabilized at different levels: 20% group 13.9 ± 10 μmol/L; 90% group 13.8 ± 10 μmol/L; 75% group 26.3 ± 14 μmol/L; all p < .05). D-xylose resorption was lowest in the 30% group, followed by 90% and 75% group (32.8 ± 4.9 mg/L; 90% group 50.0 ± 13.6 mg/L; 75% group 57.8 ± 8.8 mg/L; p = .393). Intestinal villus length decreased in all groups (20% group 11.0%; 90% group 14.0%; 75% group 19.1%).

Conclusions: 75% intestinal resection is less suitable as an SBS model, as animals tend to recover remarkably. The 90% model is suitable for longer-term studies, as animals might survive longer due to partial compensation. Due to severe nutritional, biochemical, and physiological derangements, the 30% model can only be used for acute experiments and those immediately followed by small bowel transplantation.

P159 - Establishing the first national home parenteral nutrition program for paediatric patients with intestinal failure

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Introduction: Intestinal failure (IF) is a complex condition of severe intestinal malabsorption caused by short bowel syndrome (SBS), congenital diseases of enterocyte development, and severe motility disorders (total or subtotal aganglionosis or chronic intestinal pseudo-obstruction syndrome). Children with IF require long-term total or partial parenteral nutrition (PN) to preserve nutritional status and prevent complications. When a child depends on long-term PN, home parenteral nutrition (HPN) is the best alternative to prolonged hospital stay and is recognized as the best option for improving the quality of life of these children and their families. In our country, during many years all paediatric patients with IF were managed individually within the hospital setting of paediatric gastroenterology referral centres.

Methods: We present the process of developing the first national HPN program for paediatric patients with IF, and the current practice of establishing and managing children on HPN.

Results: Five patients were managed on HPN for prolonged period, four of those are currently on HPN. There is a multidisciplinary team involved in the process. With regards to the aetiology, three patients have SBS, one patient has tufting enteropathy and one total aganglionosis with severe dysmotility and postoperative SBS. For administration of HPN we place a tunnelled single lumen central venous catheter (CVC) surgically. We use commercial premixed PN solutions with individual adjustments. The regime, monitoring and prevention of complications follow the European HPN guidelines.

Conclusion: Although PN is a life-saving therapeutic option for children with IF, it is a high-risk therapy with potential acute and long-term complications. The rate of complications can be high, particularly in inexperienced hands. Management of HPN by centralised units with expertise in the investigation of IF rehabilitation and with a multidisciplinary team to support care at home would minimise complications and improve outcome. This program will continue to expand and to improve the management at all levels.
Totally implantable venous access devices are widely used in nutrition. The authors encountered a 10-year-old boy with implantation of the device at the age of 7 years. In the recent half-year, the device was not used except for regular heparin flushing. However, hydrothorax occurred when fluid therapy was required from the device during this admission. Thoracoscopic approach showed extravascular migration and intrapleural malposition of the catheter. Intrapleural migration of the extravascular portion of the catheter owing to irritation and pressure necrosis of the pleura and gradual shortening of intravascular portion of the catheter when the child grew up may be the pathogenesis of delayed extravascular migration of the catheter.
POSTERS PRESENTED ON THURSDAY JULY 4, 2019
P2.22 - Oral Lactoferrin Increases Intestinal Regulatory T Cells And Decreases Th17 Cells Via Microbiome Alterations

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Background: Lactoferrin (LF) is a mammalian protein produced in all bodily fluids, with its greatest concentration in colostrum and breastmilk. LF is a known iron-binding glycoprotein and iron transporter with both antimicrobial and anti-inflammatory properties, however its exact mechanisms remain unknown. LF likely impacts the intestinal immune system, both through direct effects on immune cell development and activity, and indirect effects mediated through the gut microbiome. Studies in mice demonstrate that the gut microbiome is critical for the development and evolution of gut-associated lymphoid tissues. Segmented filamentous bacteria (SFB), members of the Firmicutes, promote proliferation of gut inflammatory Th17 cells. In contrast, other commensals stimulate Treg differentiation. We hypothesize that recombinant human LF (rhLF) ameliorates inflammation via altering the composition and function of the intestinal microbiome, promoting the development and function of Tregs.

Methods: TNF-αARE mice were administered oral vehicle (PBS), IV anti-TNF antibody (Infliximab) or oral recombinant human lactoferrin (rhLF) for 14 days by gavage. After 14 days of treatment the mice were euthanized and intestinal tissues were isolated for evaluation by Flow cytometry, ELISA, and PCR. Cecal stool samples were collected and bacterial RNA was isolated and sequenced.

Results: Treatment of TNF-αARE mice with orally administered LF decreased ileitis with a significant decrease in Th17 inflammatory cells (12.3% +/- .7% vs 8.2% +/- 1.2%) and a concomitant increase in anti-inflammatory regulatory T cells (18.9% +/- 1.3% vs 28.1% +/- 3.2%). Upon evaluation of the cecal bacteria of treated mice we noted that lactoferrin (rhLF55 or rhLF500) altered both the composition (Figure 1A, B, C) and diversity (Figure 1D) of the cecal microbiota. The alterations in the microbiome correlated well with the changes in immune cell predominance and the increase in the anti-inflammatory genus Faecalibacterium and Treg-skewing SCFA-producing Firmicutes, along with decreased levels of Th17-skewing Segmented Filamentous Bacteria (SFB) (Figure 1C).

Conclusion: The above data demonstrates that rhLF treatment results in a shift of the balance of pro- and anti-inflammatory T cell populations to favor resolution of gut inflammation and identifies a plausible mechanism for this shift.
P2.23 - New alternative to antibiotics: 4% Tetrasodium EDTA is a non-antibiotic antimicrobial solution effective against Canadian microorganisms and associated biofilms found in central venous access devices of total parenteral nutrition patients

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Background information
Infections are problematic for total parenteral nutrition (TPN) patients with chronic central venous access devices (CVADs) prone to microbial colonisation and exposed to multiple antibiotic treatments. This catheter is supposed to provide life-saving treatment but makes them vulnerable to central-line associated bloodstream infections (CLABSI). The current standard of care for CVADs is saline or heparin which do not prevent CLABSI.

Objective
The objective of this clinical evaluation was to confirm the clinical safety and efficacy of a non-antibiotic antimicrobial catheter lock solution containing 4% Tetrasodium EDTA (T-EDTA) to eradicate clinically relevant microbes and associated biofilms colonising the lumen of CVADs of TPN patients.

Materials and Methods
Patients with CVADs were selected across Canada based on a high risk of complications: CLABSI and occlusions. The latter determined from the high use of alteplase. Selected TPN patients used a 4% T-EDTA lock solution as the sole lock solution rather than their standard of care. Main clinical endpoints included reduction of alteplase use and infection, and safety assessment. Before and during data were collected.

Results
Data collected over a 24-month period in 24 TPN patients show both a clinically relevant decrease in CLABSI by 50%, in-hospital days by 75%, catheter replacement by 68% and alteplase use by 100% when the catheter is supposed to provide life-saving treatment but makes them vulnerable to central-line associated bloodstream infections (CLABSI). The current standard of care for CVADs is saline or heparin which do not prevent CLABSI.

Conclusions
Intraluminal microenvironment of a CVAD is ideal for polymicrobial biofilm formation where quorum sensing promotes antibiotic resistance and provides the perfect hiding spot for superbugs. There is a close relationship between clot, bacteria and biofilm propagating CVAD complications: infections and occlusions. Results from this evaluation demonstrate clinically relevant reduction in CVAD complications with the use of a 4% T-EDTA lock solution. Routine use of an effective non-antibiotic antimicrobial solution is crucial in reducing the risk of CLABSI within Canadian hospitals and addresses the need to use alternatives to antibiotic agents.

P2.24 - Gothenburg Intestinal Transplant Endoscopy Score: A Prospective, Single Center Evaluation

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Introduction: The findings during endoscopy in acute cellular rejection (ACR) are well-known, but a grading system for its severity is lacking. Gothenburg Intestinal Transplant Endoscopy Score (GITES) is a novel, five-stage endoscopic score aiming to describe and categorize the endoscopic findings after intestinal transplantation. The aim with this study was to establish the usefulness of GITES in diagnosing acute cellular rejection. This could result in a more objective evaluation of the endoscopic findings and subsequently an earlier diagnosis of rejection.

Methods: We prospectively graded the endoscopic findings with GITES in 13 adult patients (3 isolated intestinal grafts, 10 multivisceral grafts) at one single center using white light high definition endoscopy systems. The scoring was performed at the time of endoscopy and later correlated to the histological findings.

Results: Eighty-five ileoscopies were scored. In 52 (61%) cases the endoscopic findings were normal. Twenty-three (29%) out of the 33 abnormal endoscopies revealed mild alterations represented by mild/moderate edema, erythema or blunted villi (GITES 1 and 2). Acute rejection was found in biopsies from 11 (34%) endoscopy sessions (4 mild & 7 moderate/severe) and in three specimens the biopsies revealed CMV enteritis. GITES above 1 (erythema, edematous villi) had 91% sensitivity and 94% specificity for ACR whereas positive (PPV) and negative predictive values (NPVs) were 78% and 98%, respectively.

Conclusions: These results suggest that evaluation of the endoscopic findings with GITES results in a satisfactory identification and stratification of rejection. A prospective, multicenter evaluation is needed to confirm these findings.

Table 1 - Comparison of the number of catheter complications before and during the use of 4% T-EDTA as sole lock solution in 24 TPN patients during a 2-year period.

<table>
<thead>
<tr>
<th></th>
<th>Standard</th>
<th>4% T-EDTA</th>
<th>% reduction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of catheter replacement</td>
<td>19</td>
<td>6</td>
<td>68</td>
</tr>
<tr>
<td>Number of catheter-related infections</td>
<td>16</td>
<td>8</td>
<td>50</td>
</tr>
<tr>
<td>Number of occlusions</td>
<td>11</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Number of inpatient days</td>
<td>141</td>
<td>40</td>
<td>75</td>
</tr>
</tbody>
</table>

G6: Normal enteroscopy
G1: Mild erythema and histology, slightly edematous villi
G2: Marked erythema, friability, erosions, blunted villi
G3: Spontaneous bleeding, ulcerations, villi loss
G4: Mucosal loss, visible submucosa

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Background information
Infections are problematic for total parenteral nutrition (TPN) patients with chronic central venous access devices (CVADs) prone to microbial colonisation and exposed to multiple antibiotic treatments. This catheter is supposed to provide life-saving treatment but makes them vulnerable to central-line associated bloodstream infections (CLABSI). The current standard of care for CVADs is saline or heparin which do not prevent CLABSI.

Objective
The objective of this clinical evaluation was to confirm the clinical safety and efficacy of a non-antibiotic antimicrobial catheter lock solution containing 4% Tetrasodium EDTA (T-EDTA) to eradicate clinically relevant microbes and associated biofilms colonising the lumen of CVADs of TPN patients.

Materials and Methods
Patients with CVADs were selected across Canada based on a high risk of complications: CLABSI and occlusions. The latter determined from the high use of alteplase. Selected TPN patients used a 4% T-EDTA lock solution as the sole lock solution rather than their standard of care. Main clinical endpoints included reduction of alteplase use and infection, and safety assessment. Before and during data were collected.

Results
Data collected over a 24-month period in 24 TPN patients show both a clinically relevant decrease in CLABSI by 50%, in-hospital days by 75%, catheter replacement by 68% and alteplase use by 100% when the catheter is supposed to provide life-saving treatment but makes them vulnerable to central-line associated bloodstream infections (CLABSI). The current standard of care for CVADs is saline or heparin which do not prevent CLABSI.

Conclusions
Intraluminal microenvironment of a CVAD is ideal for polymicrobial biofilm formation where quorum sensing promotes antibiotic resistance and provides the perfect hiding spot for superbugs. There is a close relationship between clot, bacteria and biofilm propagating CVAD complications: infections and occlusions. Results from this evaluation demonstrate clinically relevant reduction in CVAD complications with the use of a 4% T-EDTA lock solution. Routine use of an effective non-antibiotic antimicrobial solution is crucial in reducing the risk of CLABSI within Canadian hospitals and addresses the need to use alternatives to antibiotic agents.
P2.25 - Effects of major liver allocation policy changes on waitlist outcomes in multivisceral transplantation in the United States

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Background: Organ allocation in multivisceral transplant (MVT; liver-intestine, liver-pancreas-intestine) is determined based on their ranking in the liver transplant waitlist. MVT candidates do not usually have high laboratory MELDNa (MELD) score and an exception point is given per the OPTN policy. Currently, their exception point is determined as “≥10% increase in mortality risk to their MELD score”. Since 2013, an exception point of 29 has been applied to MVT candidates with approval by regional review board. As major revisions in liver allocation, Share 35 rule and MELDNa score were implemented in 2013 and 2016. The aim of this study was to evaluate effects of these updates in liver allocation policy on waitlist outcomes in MVT.

Methods: We examined adult patients who were registered for liver alone (LTA), liver-kidney (L-K), and MVT between 2011 and 2018 by using the UNOS registry. Registration periods were grouped according to the major revisions of liver allocation; 1) pre-Share 35 period (1/1/2011-6/17/2013), 2) post-Share 35 period (6/18/2013-1/1/2016), 3) MELDNa period (1/1/2016-3/31/2018). 90-day mortality in MVT candidates was evaluated in each period in comparison with those in LTA/L-K candidates who had similar MELD score (score categories of 20-28 and 29-34) to exception points for MVT candidates. Risks were adjusted by using Fine-Gray regression model.

Results: In MVT candidates, while there was no difference between the pre and post-Share 35 periods (HR, 0.96; P=0.29), 90-day mortality significantly increased in the MELDNa period compared with that in post-Share 35 period (HR, 1.08; P=0.02). Mortality within 90 days in LTA/L-K candidates with MELD score of 20-28 continued to decrease over periods (hazard ratio [HR], 0.91 and 0.82; P=0.042 and <0.001 for pre vs. post-Share 35 periods and post-Share 35 vs. MELDNa periods). 90 day-mortality in LTA/L-K candidates with MELD score of 29-34 significantly decreased in the MELDNa period compared with the post-Share 35 period (HR, 0.78; P<0.001), whereas there was no difference between the pre and post-Share 35 periods (HR, 0.99; P=0.9).

Conclusions: While the recent revisions of liver allocation improved waitlist outcomes in LTA/L-K candidates, MVT candidates did not benefit from them and 90 day-mortality significantly increased in the MELDNa period. Exception point for MVT candidates may need to be reconsidered, given the increased number of high score patients.

P2.26 - Optimizing the Management of intestinal pseudo-obstruction with effective bowel decompression using a pan intestinal decompressive tube

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2 Sydney medical school, university of sydney
3 John Hunter Children’s Hospital, Newcastle, NSW

Children with intestinal pseudo-obstruction are notoriously hard to manage. Refractory abdominal distension, small bowel bacterial overgrowth, inability to feed, frequent line infections, septic episodes and intestinal failure related liver damage are unfortunate realities in the management of these children. It’s also hard to determine how contributory adhesive bowel obstruction is to the ongoing abdominal distension. One of the central factors contributing to all this is the inability to effectively decompress the inherently poorly peristaltic long tubular length of the intestine.

We devised a simple solution to effectively decompress the entire length of small bowel by passing a perforated decompressive tube all along its length that went from the DJ flexure to the caecum. Low profile devices at both ends ensured decompression into bags at both ends. The resulting decompressed bowel with reduced luminal calibre prevented stasis and promoted some peristalsis. In addition it stented the bowel in a non-obstructive pattern in the abdomen.

8 children with intestinal pseudo obstruction had pan intestinal decompressive tubes placed. Ages ranged from 5 weeks to 16 years. All children had improvement in their abdominal distension, were able to take oral feeds and started stooling more frequently. 2 children with gastrochisis came off TPN after 4 and 6 months. Serum bilirubin improved initially in all children. In 2 children, after temporary relief, there was progression to liver failure and eventual death. In 3 children, additional procedures to reduce leakage around the tubes and skin excoriation were done. In 7 children, this reduced hospitalization significantly and allowed management to continue from home.

In children with intestinal pseudoobstruction, the placement of a pan intestinal decompressive tube can mitigate several of the problems associated with this condition. In gastrochisis with hypomobility, it has the potential to restore motility and avoid the need for a transplant.
CIRTA 2019 Book of Abstracts - Posters

P2.27 - Bortezomib for refractory autoimmune hemolytic anemia after intestinal transplantation

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2 Dept. Transplant Surgery; UZ Leuven, Belgium

Introduction: Autoimmune Hemolytic anemia (AIHA) after solid organ transplantation is a relatively rare but severe complication. In contrast, after intestinal transplantation the incidence is up to 12.2% and generally occurs between 8-32 months after transplantation. Published therapeutic regimes include high-dose corticosteroids, intravenous immunoglobulin, plasmapheresis, rituximab and a switch of tacrolimus or sirolimus to cyclosporine. Generally, the treatment is started within the first week after transplantation. During the treatment he developed a transient increase in stoma production, within the first week after administration of bortezomib. We ran out of therapeutic options a trial with bortezomib (Velcade®) at a dose of 1.0-1.3 mg/m² was given every 3 days, four times in total (with a second course 10 days later).

Results: We observed a rapid and sustained increase in haemoglobin levels over 12 months. The overall mortality rate of AIHA is about 8%.

Conclusion: Bortezomib is a selective and reversible proteasome 26S inhibitor that directly inhibits antibody production through plasma cell depletion, it is used as a treatment of multiple myeloma and chronic cold agglutinin disease after allogenic hematopoietic stem cell transplantation (HSCT). Bortezomib is a selective and reversible proteasome 26S inhibitor that directly inhibits antibody production through plasma cell depletion, it is used as a treatment of multiple myeloma and chronic cold agglutinin disease after allogenic hematopoietic stem cell transplantation (HSCT).

P2.28 - Eosinophilic gastrointestinal disease following intestinal transplant

Brandon Arnold, Lin Fei, Huaiyu Zang, Patricia Fulkerson, Sam Kocoshis
Cincinnati Children’s Hospital

Introduction: Eosinophilic gastrointestinal disease (EGID) has been reported to be of higher incidence following solid organ transplantation than the general population. We reviewed the EGID incidence following single organ intestinal transplant or multi-visceral transplants at our institution.

Methods: We completed a retrospective chart review on twenty-six patients who were followed at our center after intestinal or multi-visceral transplantation between the years of 2003-2018. Patients transplanted prior to age of 18 and followed at least 1 year from transplant were included. Histological criteria were used for EGID diagnoses. Eosinophil infiltration of the native gastrointestinal (GI) tract only was included and not the allograft. Comparisons were done using Fisher’s exact test and Mann-Whitney two-tailed test.

Results: Of the 26 individuals followed for minimum of a year after intestinal or multi-visceral transplant, 23 (23/26, 88%) individuals were subsequently diagnosed with EGID. Twelve patients (12/26, 46%) were diagnosed with eosinophilic esophagitis (EoE), one of which also had eosinophilic gastritis (EG). One patient had EG and eosinophilic colitis (EC) and one each with EC and EG. The mean age at diagnosis of EGID was 7.56 ± 5.30 years and EGID developed on average 5.20 ± 3.40 years after transplant. There was no statistically significant difference between age at time of transplant in EGID vs non-EGID (2.35 ± 2.91 vs 4.08 ± 5.07). All (15/15) patients who developed EGID were on tacrolimus at the time of diagnosis and 13/15, 87% underwent immunosuppression induction with anti-thymocyte globulin. Five (5/15, 33%) patients also developed food allergies post-transplant, including 2 (2/15, 13%) in the non-EGID group. Three patients (3/15, 20%) were diagnosed with PTLD prior to EGID diagnosis, while no (0/15) PTLD diagnoses occurred in the non-EGID group. Rejection occurred at a greater frequency in the EGID group than the non-EGID groups (13/15, 93% vs 7/11, 64%). Mean time from first rejection episode to developing EGID was 37.4 ± 2.74 years. Eleven (11/15, 73.3%) EGID patients have data on time from first rejection episode to developing EGID was 3.74 ± 2.74 years. Eleven (11/15, 73.3%) EGID patients have data on time from first rejection episode to developing EGID was 3.74 ± 2.74 years. Eleven (11/15, 73.3%) EGID patients have data on time from first rejection episode to developing EGID was 3.74 ± 2.74 years. Eleven (11/15, 73.3%) EGID patients have data on time from first rejection episode to developing EGID was 3.74 ± 2.74 years.

Table: Summary of EGID and Non-EGID Patients

<table>
<thead>
<tr>
<th>Condition</th>
<th>EGID (15)</th>
<th>Non-EGID (11)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>11</td>
<td>15</td>
</tr>
<tr>
<td>Age at transplant in years</td>
<td>4.08 (5.07)</td>
<td>1.35 (2.91)</td>
</tr>
<tr>
<td>Age at transplant (median)</td>
<td>1.36</td>
<td>0.97</td>
</tr>
<tr>
<td>Age at diagnosis of EGID (median)</td>
<td>7.56 (5.10)</td>
<td>5.33</td>
</tr>
<tr>
<td>Time from transplant to EGID (median)</td>
<td>5.20 (2.59)</td>
<td>3.63</td>
</tr>
<tr>
<td>Endoscopic indication</td>
<td>6 (40)</td>
<td>2 (18)</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>2 (13)</td>
<td>1 (7)</td>
</tr>
<tr>
<td>Abdominal Pain</td>
<td>2 (13)</td>
<td>2 (18)</td>
</tr>
<tr>
<td>History of ulceration</td>
<td>1 (7)</td>
<td>2 (13)</td>
</tr>
<tr>
<td>Elevated liver transaminase</td>
<td>1 (7)</td>
<td>2 (13)</td>
</tr>
<tr>
<td>Nausea</td>
<td>2 (13)</td>
<td>1 (7)</td>
</tr>
<tr>
<td>Number of patients with rejection</td>
<td>7 (63.6)</td>
<td>13 (94)</td>
</tr>
<tr>
<td>Grade 1(total episodes of rejection)</td>
<td>7</td>
<td>3</td>
</tr>
<tr>
<td>Grade 2</td>
<td>3 (23)</td>
<td>3 (23)</td>
</tr>
<tr>
<td>Grade 3</td>
<td>3 (23)</td>
<td>3 (23)</td>
</tr>
<tr>
<td>Unspecified grade</td>
<td>5 (33)</td>
<td>5 (33)</td>
</tr>
<tr>
<td>Food allergies</td>
<td>2 (13)</td>
<td>5 (38)</td>
</tr>
<tr>
<td>Time post to food allergies (median)</td>
<td>1.68 (1.39)</td>
<td>1.63 (1.29)</td>
</tr>
<tr>
<td>Peak AEC/CR post-transplant (mean)</td>
<td>7.85 (6.79)</td>
<td>7.35 (6.79)</td>
</tr>
<tr>
<td>Diagnosed with PTLD (%)</td>
<td>0 (0)</td>
<td>2 (18)</td>
</tr>
</tbody>
</table>

Of the 26 individuals followed for minimum of a year after intestinal or multi-visceral transplant, 23 (23/26, 88%) individuals were subsequently diagnosed with EGID. Twelve patients (12/26, 46%) were diagnosed with eosinophilic esophagitis (EoE), one of which also had eosinophilic gastritis (EG). One patient had EG and eosinophilic colitis (EC) and one each with EC and EG. The mean age at diagnosis of EGID was 7.56 ± 5.30 years and EGID developed on average 5.20 ± 3.40 years after transplant.
Conclusion: The prevalence of EGID disorders in intestinal transplants is much higher than the general population and other single solid organ transplants. Close screening for EGID development following intestinal transplant is warranted.

P2.29 - IGL-1 solution for the preservation of human intestine: the first report in the literature
John Söfteland 1 2, Henrik Göransson 1 2, Mats Hellström 1, Gustaf Herlenius 2, Mihai Oltean 1 2

1 Laboratory for Transplantation Biology and Regenerative Medicine, University of Gothenburg, Sweden
2 Transplant Institute, Sahlgrenska University Hospital, Gothenburg, Sweden

Introduction: IGL-1 is a newer preservation solution largely resembling University Wisconsin (UW) solution in terms of ingredients but where the viscous starch has been replaced by polyethylene glycol resulting in lower viscosity. In addition, IGL-1 has low potassium and high sodium concentrations (extracellular-type solution). The solution has been used in several European countries for the preservation of kidneys, pancreases and livers with efficacy and safety comparable to those of UW or Custodiol. However, a systematic appraisal of the intestinal preservation injury using ILG-1s missing.

Methods: Following aortic retrograde perfusion with 4 liters IGL-1 the ileum of fifteen deceased, brain dead multiorgan donors was retrieved and stored at 4°C. Samples were obtained after 8h, 14h and 24 hours of cold storage. Histology (Chiu/Park scale), Goblet cells (GC) count, apoptosis (active caspase 3) and tight junction proteins claudin 3 and Zonula occludens (ZO)-1 were studied.

Results: Eight hours of cold storage resulted in a moderate epithelial detachment form lamina propria (median Chiu grade 2 (range 1-3) and minimal GC depletion. Preservation injury progressed to a median grade of 3 (range 2-5) after 14 hours and remained at similar levels at 24 hours. Active Caspase-3 was very low or absent at all time-points. After 8h ZO-1 expression was well preserved along the entire villus but became discontinuous after 14 hours of cold storage. Claudin 3 staining pattern remained well preserved throughout the entire cold storage.

Conclusions: The development of the preservation injury in the human intestine following perfusion and storage in IGL-1 appears similar with the other solutions. Interestingly, the structural preservation injury did not seem to continue to worsen after 14 hours while the injury at subcellular level advanced further.
A wide range of non-transplant options has been established in the era of successful modern PN, ILT in children with intestinal failure-associated liver disease (IFALD) may be referred for combined liver intestinal transplantation with an aim to becoming parenteral nutrition (PN)-independent and free from liver disease. Isolated liver transplantation (ILT) in these patients is avoided, as most patients who had this procedure developed disease in the graft liver due to the ongoing need for PN. However, with the current paradigm of successful long-term PN managed in an expert centre, ILT may be an option for children at risk of death due to end-stage IFALD where no intestinal graft is available.

Methods: A retrospective review of case notes of three patients with IFALD who underwent ILT in a single centre between 2014 and 2018. Results: Case 1 is an 8 year-old with short bowel syndrome (SBS) secondary to gastroschisis, with 32cm of small bowel, and developed life-threatening IFALD. She was referred for combined liver intestinal transplant, but her parents declined the small bowel so she underwent ILT at age 3. She remains PN-dependent, despite pre-transplant bowel lengthening surgery. She is well and growing along the <0.4th centile with no liver disease at 4 years post-transplant.

Case 2 also developed SBS secondary to gastrochisis, with 32cm of small bowel after resection. She was listed for combined liver intestine transplant at 9 months of age, but developed end-stage liver disease with severe stomal bleeding necessitating a life-saving ILT in 2015. Following transplant she remains PN-dependent with some oral feeds and no signs of liver disease at 2 years post-transplant[1][2].

Case 3 is a 2 year-old with SBS due to antenatal bowel perforation requiring removal of a large portion of small bowel as a neonate. He underwent ILT at 18 months due to development of end-stage liver disease and coagulopathy, with no available intestinal graft. Recovery was complicated with life-threatening line infections, but he has achieved stability, and is growing well with 70% of his calorie and fluid requirement coming from PN and no signs of liver disease at 9 months post-transplant.

Conclusion: In the era of successful modern PN, ILT in children with end-stage IFALD may be life-saving for patients in whom size-matched organs are not available, or intestinal transplantation is declined.
P2.32 - Dynamics of recipient-derived T cell populations infiltrating intestinal transplant mucosa, and their expression of gut-homing chemokines and integrins

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Introduction: Early infiltration of recipient-derived T cells into an intestinal transplant graft is associated with rejection, which can lead to graft loss and death.1 However, the infiltration dynamics of different T cell subsets are poorly understood, as are the mechanisms of entry. With the development of anti-integrin and -chemokine therapeutics, understanding whether pathways involved in gut-homing could be targeted for the prevention or treatment of early rejection is of significance.

Methods: Adult intestinal transplantation recipients were identified via longitudinal cohort studies at Oxford University Hospitals NHS Foundation Trust. Fifteen samples of intestinal graft biopsies were obtained from ten patients (Table 1) at various times after transplantation (23 days to 5 years).

Donor- and recipient-derived T cells in intestinal transplant mucosa were identified using flow cytometry and antibodies to discordant HLA Class I proteins (Fig 1A). Gut-homing molecules CCR9 and β7 integrin, and markers of conventional and innate-like T cell subsets were examined.

Results: The relative frequencies of key T cell subsets altered following transplantation (Fig 1B & C). Early post-transplant, CD8 and non-Vδ2 γδT cells dominated (28% and 49%, respectively). These early non-Vδ2 γδT cells were predominantly donor-derived. Innate-like MAIT and Vδ2+ γδT cells were rare (<1%) early post-transplant but recovered with time (6% and 2%, respectively). The fraction of recipient-derived T cells in the graft increased with time, with highly variable kinetics between subjects.

Early post-transplant, expression of CCR9 and β7 integrin was low on recipient-derived T cells, with 29% of CD8 and 48% of CD4 T cells negative for both markers. In contrast, 52% of CD8 and 33% of CD4 donor-derived T cells co-expressed CCR9/β7 (Fig 1D). CCR9/β7 co-expression increased in recipient-derived T cells with time.

Conclusion: Following intestinal transplantation, the relative abundance of T cell subsets within the graft is dynamic. Innate-like T cells were near-absent in the first year post-transplant, whilst early post-transplant, donor-derived non-Vδ2 γδT cells were over-enriched. Fewer recipient-derived T cells infiltrating the graft expressed putative gut-homing molecules. This questions whether pharmaceutical targeting of these pathways is appropriate in graft rejection, but samples from a rejection episode are needed for validation.

1 Zuber J. et al. Sci Immunol 1, eaah3732 (2016).
P2.33 - Fish oil monotherapy for intestinal failure-associated liver disease on SMOFlipid in the neonatal intensive care unit

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2 Department of Pharmaceutical Services, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea.

Background: Management of IFALD using FO monotherapy with Omegaven has repeatedly been shown to be safe and effective in children and more recently, also in preterm neonates who are on dependent on long-term PN. In the present study we report the outcome of our expanded experience with FO monotherapy for IFALD on fish oil-based combination LE in the pediatric population, particularly in preterm low birth weight infants.

Methods: A diagnosis of IFALD was made when serum direct bilirubin levels were > 2.0 mg/dL in two consecutive measurements that were more than one week apart, without evidence of intrinsic causes of liver dysfunction. Management of IFALD was done by initially decreasing the total calories provided by PN, usually by lowering the amount of lipid to 2.0 g/kg/day. When calorie deceleration did not result in alleviation of cholestasis, LE was switched from SMOF to Omegaven at 1.0 g/kg/day infused over 24 hours.

Results: Fifteen infants met the criteria for IFALD and received FO monotherapy during the study period from March 2017 to June 2018. Median gestational age was 27.5 weeks and median birth weight was 862.5 g. IFALD was successfully reversed in 10 infants (11/15, 73.3%). Four infants died of IFALD and resultant hepatic failure. Median direct bilirubin values were initially elevated and then steadily declined from the third week of treatment onward. Enteral tolerance increased in varying degrees in all but one patient during the treatment period. Mean weight gain was 26.0 g/day during FO monotherapy. Omegaven at a dose of 10 g/kg/day was well tolerated in these infants during the entire period of administration and no adverse events related to Omegaven use were seen. No signs of essential fatty acid deficiency (EFAD) were observed.

Conclusion: Fish oil monotherapy was safe and effective in the treatment of IFALD arising during SMOFlipid use in premature neonates on long-term PN.

P2.34 - Intestinal transplantation: GVHD and different induction immunosuppression protocols over 25 years at a single center.

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1 Miami Transplant Institute, University of Miami, Miami, Florida, USA.
2 Zagazig University School of Medicine, Zagazig, Egypt.

Background: Even though graft versus host disease (GVHD) occurs in 50% of patients receiving allogeneic hematopoietic stem cell transplantation, it is a rare complication after solid organ transplantation. Due to the large amount of lymphoid cells, intestinal and multivisceral transplantation triggers the bidirectional exchange of immune cells resulting in graft versus host and host versus graft interaction. Once it occurs, GVHD is associated with high morbidity and mortality after intestinal transplantation. We describe our center experience with GVHD over almost 25 years.

Methods: We retrospectively reviewed 442 intestinal transplants from 1994-2018 at Miami Transplant Institute and recipients were divided into 5 groups depending on the induction immunosuppression used; group 1 (44/442): high dose steroid (34/44), OKT3 (7/44), or cyclophosphamide (3/44); group 2: anti-CD25 (daclizumab or basiliximab) (159/442); group 3: alemtuzumab (113/442); group 4: rabbit antithymocyte globulin (rATG) (34/442); group 5: rATG and rituximab. Types of intestinal transplant included: isolated intestine (I) (n=124), liver-intestine (LI) (n=38), modified multivisceral (MMV) (n=39), and multivisceral (MV) (n=242) allografts.

Results: GVHD occurred in 8.6% (39/442). Actuarial estimates of GVHD free survival at 3, 6, 12, 24 and 60 months in the 5 induction groups were: 92% through all times in group 1; 94%, 88%, 87%, 86% and 86% in group 2; 99%, 99%, 97%, 96% and 96% in group 3; 88% through all times in group 4; 90%, 87%, 86%, 86% and 86% in group 5, respectively. MV and MMV allografts were associated with an increased incidence of GVHD (P=0.00004) during 60 post-transplant months. Group 3 (Alemtuzumab) was associated with a decreased hazard rate of developing GVHD (P=0.004) but this effect lasted only during the 1st 6 post-transplant months. Graft loss due to GVHD occurred in 31.6% (12/38) with an increased risk in MV and MMV transplant recipients (P=0.05).

Conclusion: MV and MMV transplant recipients experienced increased risk of GVHD related morbidity and mortality while alemtuzumab induction immunosuppression protocol was associated with a decreased risk of GVHD during the 1st 6 post-transplant months.
Introduction: Phenotypic diarrhoea (PD), also known as tricho-hepato-enteric syndrome, is a rare disorder with intestinal and extra-intestinal manifestations. Patients have low birth weight, facial and hair changes and a severe diarrhoeal illness, requiring parenteral nutrition (PN).

In this study, we reviewed outcomes for the largest UK cohort of patients with phenotypic diarrhoea born since 1996.

Methods: We reviewed the medical records of children with phenotypic diarrhoea cared for in our tertiary gastroenterology centre up to 31 December 2018.

Results: 12 patients (7 female, 5 male) were included. 2 patients died, one at age 2 years and one at 10 years, both due to respiratory complications secondary to infection. Median age of the surviving 10 patients was 12.6 years (range 3.7-22.2). Probabilities of survival based on the Kaplan-Meier method was 100% at 1 year, 92% at 5 years, 79% at 10 years and 79% at 15 years.

Median age of PD diagnosis was 11.6 months (range 2.9-114.8). Presenting symptoms included protracted diarrhoea (n=6), failure to thrive and growth (n=6), liver disease (n=3), cholelithiasis (n=2), meconium ileus (n=2), years PN stopped was 9.7 years (range 4.37-13.27); median PN length was 5.7 years (range 3.7-13.4 years). Probability of continuing PN based on the Kaplan-Meier method was 100% at 1 year, 80% at 5 years, 41% at 10 years and 25% at 15 years. Patients continuing PN receive it 3-4 nights/week. One patient restarted PN after 11.4 years to achieve pubertal growth. Patients had improved weight-for-age Z scores with PN but showed a decline (median -0.61, range -3.5 to -0.12) after PN stopped. The majority have short stature, following the lower centiles (0.4th-9th) for growth.

Conclusion: In our cohort, intestinal failure is temporary; half of our patients are off PN, with future plans to wean the remaining patients. Rather than intestinal failure per se, morbidity and mortality is largely caused by immune deficit and resultant infections.
P2.38 - Contemporary outcomes of pediatric intestinal failure: a multicenter study

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Introduction: Important changes in the management of pediatric intestinal failure (IF) have been implemented over the last 15 years.

Objective: To assess the impact of those changes on the outcome of a contemporary cohort of pediatric IF patients from 6 IF programs, 5 of which offer intestinal transplantation.

Methods: Retrospective analysis of a multicenter IF cohort. Entry criteria included patients < 18 years of age receiving PN for more than 42 consecutive days, primary gastroenterological disorder and diagnosis of IF between 2010 and 2015. Primary outcome was death, transplant or enteral autonomy. Kaplan-Meier analysis was used for diagnosis of IF between 2010 and 2015. Primary outcome was death, transplant or enteral autonomy. Kaplan-Meier analysis was used for log rank test and Cox regression analysis.

Results: 443 patients (male 63%) with a median gestational age of 34 weeks (29-37) and birth weight of 2.1kg (1.2-2.8) constituted the study group. Patients were followed for 3.8 years (2.3-5.3). Median time on PN was 384 days (12-947). Patients had 40% (16-100) of expected growth. Main etiologies included necrotizing enterocolitis (29.3%), abdominal wall defects (30.5%) and malrotation (29.3%). Eighty-one patients (38.3%) developed progressive intestinal failure associated liver disease (IFALD) with a bilirubin >75mmol/l. All patients were managed with hepatoprotective nutritional strategies (fish oil based lipids or lipid lowering strategies). The presence of IFALD impacted negatively on transplant free survival (P < 0.001) and achievement of intact full colon in continuity (P < 0.01). Overall, 218 (48.3%) patients achieved enteral autonomy. 137 (30.9%) remained on PN, 53 (12%) were transplanted and 40 (9%) patients died. Achievement of enteral autonomy was associated with short bowel syndrome (P < 0.01), NEC (P < 0.026), intact colon (P < 0.01) and ileocecal valve (P < 0.01).

Conclusions: This contemporary cohort achieved a significantly lower incidence of death and transplantation compared with previous multicenter reports. However, the number of children achieving enteral autonomy has not changed while a larger number of patients remain PN dependent. Although this cohort may represent a more complex group of patients due to potential referral bias, these findings suggest a shift in outcomes and a need for new strategies for the achievement of enteral autonomy.

P2.39 - Visceral Transplantation in megacystis microcolon intestinal hypoperistalsis syndrome

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Introduction: Megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS) is a rare congenital disorder with poor prognosis. It is associated with intestinal pseudo-obstruction and urinary system problems secondary to dilated bladder. MMIHS patients usually have multiple surgical interventions related to pseudo-obstruction episodes and become TPN dependent due to surgical or functional short gut syndrome. We aim to analyze patient and graft characteristics including early and late post-transplant complications in patients with MMIHS with visceral transplantation (VT).

Methods: Data of all patients with MMIHS receiving visceral transplantation between 1990 and 2019 were reviewed retrospectively.

Results: Six MMIHS patients underwent VT at our institution. One had prior isolated small bowel transplantation at another facility and had allograft failure. Transplantation was with multivisceral (MVMT) in 5 and modified multivisceral (MMVT) in one patient with good liver function. Pancreaticoduodenal and splenic complex was preserved in MMVT. Allograft included colon and pyloromyotomy was performed in last 4 patients. Thymoglobulin was administered as induction agent and tacrolimus and prednisolone were given as maintenance immunosuppression. Median age at transplantation was 4.3 years (range=1-12) and a M/F ratio of 5/1. Median follow-up of 3.8 years (range=12-18.4 years). Acute cellular rejection was the most frequent early post-transplant complication (83%) and 2 patients had EBV associated post-transplant lymphoproliferative disorder during the follow up period. One patient needed kidney transplantation because of IgA nephropathy. Two patients currently have end ileostomy and 3 have end colostomy after closure of loop ileostomy. One patient is awaiting ileostomy closure. Five patients have attained nutritional autonomy and were weaned off TPN and intravenous fluids. One of these 5 patients is on jejunostomy tube feeds due to oral aversion. One patient is still on tube feeds and TPN due to esophageal stricture caused by perforation after myotomy for achalasia diagnosed at post-transplant 8 months.

Conclusions: Visceral transplantation for MMIHS patients is a lifesaving procedure with excellent patient and allograft survival. Timing, thorough pre-transplant work-up and appropriate allograft selection are keys for good outcomes.
Objective: Visceral transplantation (VT) is indicated in patients who have failed intestinal rehabilitation and have complications related to parenteral therapy. VT outcomes from an academic center over a 20-year period were studied.

Methods: Between 1/1996 and 12/2015, 212 pediatric patients underwent 235 VT. There were 23 re-transplants. Index VT included isolated small bowel (SB n=90); liver, pancreas, small bowel (L+SB n=86); liver, stomach, pancreas; small bowel (multivisceral MVT n=28); or stomach, pancreas, small bowel (modified multivisceral MMVT n=7) with or without colon. M/F ratio was 129/87. Immunosuppression was antibody induction, tacrolimus and steroids. VT indications were rejection (n=26), technical (n=3), primary nonfunction (n=1), PTLD (n=1), necrotizing enterocolitis (n=24), pseudo-obstruction (n=27), Hirschsprung’s disease (n=27), microvillus inclusion disease (n=12), others (n=11).

Results: 140 patients are alive between 2 and 23 years after transplant (mean 12.8 years). 31 of these underwent alloagraft enterectomy for rejection (n=26), technical (n=3), primary nonfunction (n=3), PTLD (n=3), and are on parenteral nutrition. 11 in this group were re-transplanted. 77 patients died between 4 days and 20.5 years following visceral transplant (mean 44.5 months). Infection and multisystem organ failure were the leading cause of death (n=26, 31%). Maximum 10 and 15-year graft and patient survival were observed in LB recipients confirming liver’s protective role in sustaining small bowel grafts (table and graph; p=0.009).

Conclusions: Visceral transplantation is a viable option for patients with intestinal failure and offers long-term benefit and freedom from parenteral nutrition. Infection continues to be a major factor responsible for graft loss. With careful patient selection and comprehensive management by multidisciplinary team prolonged nutritional autonomy can be restored.
P2.42 - Vascularized composite allografts as immunological monitors
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Introduction: Abdominal wall transplantation (AWTx) offered a potential solution to the often-challenging closure of the abdominal wall at the time of intestinal transplantation (ITx). However, besides facilitating closure, the AWTx has been proven a promising asset for early, patient led rejection monitoring. We have therefore also used sentinel skin grafts for solely graft monitoring purposes when there was no clinical need for AWTx.

Methods: We performed a retrospective analysis of all patients undergoing intestinal and vascularized composite allograft (VCA) transplantation. Clinical presentation of rejection was correlated with histology, stoma output, citrulline levels and endoscopy findings.

Results: From October 2008 to October 2018, 45 patients underwent ITx. Ten underwent a modified multivisceral transplant and 35 an isolated small bowel transplant. Mean age was 42.6 years (range 23-73). M/F: 27/18. Median follow up was 31 days (range 14-365). All patients had Campath induction (30 mg iv) followed initially by Tacrolimus based maintenance (trough level of 8-12 ng/ml). Thirty one patients received a VCA in addition to ITx. Twenty two of these were AWTx. There were 5 intestinal biopsy proven rejections in the IT alone group (36%) and a further 5 patients in the same group were falsely treated for rejection, as this was later labelled as infection. There were 10 patients with rejection in the VCA part of the IT+ VCA group (11/31, 35%). These patients presented with a rash limited to the VCA. Of those 10 patients, there were 5 with concurrent intestinal rejection (5/31, 15%) with a lead-time of 5-7 days between VCA and ITx.

Conclusion: We report on a series of combined VCA and ITx. The skin component has been utilized as a dynamic canvas for remote immune monitoring of visceral grafts. It has so far been useful for patient led monitoring of the ITx graft since it is visible and presents the earliest and only sign of rejection.

P2.43 - Impact of a new Nordic waiting list algorithm for patients awaiting intestinal transplantation in Sweden and Norway
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Introduction: Since 1998, Swedish and Norwegian patients are transplanted in Gothenburg, Sweden. Initially, our institution managed the organ allocation of intestinal grafts for all patients. From 2010, due to a high mortality on the pediatric waiting list (WL) (70%), candidates were listed separately to increase awareness. From 2010 onwards, a joint Nordic shared waiting list (WL) was implemented for children listed for a liver transplantation and both pediatric and adult patients in need of an ITx. Herein, we describe and assess whether the change in WL algorithm implemented in 2010 has had an impact on WL dynamics for Swedish and Norwegian patients.

Materials and methods: Retrospective review of the Nordic Liver Transplant Registry.

Variables studied: Comparison between (ERA 1: 1998 - 2010) and (ERA 2: 2011-2018). WL dynamics, organ donor characteristics, donor to recipient body weight ratio, cold ischemia time, mortality on the WL, proportion of grafts being shared between Swedish and Norwegian patients as well as the number of grafts imported from other Nordic and European countries.

Results: 35 ITx have been performed in 34 patients; pediatric (9), adults (25). Isolated ITx (6) and MV grafts (29). Median time on the WL decreased when comparing ERA 1 and 2 but was not statistically significant (Table 1). Median waiting time for the pediatric recipients was longer than for the adults during both Eras. There were no mortalities on the WL after 2010. The proportion of grafts imported from other countries increased from 28% to 43% and the number of Swedish patients receiving grafts from other countries increased from 15% (n=3) to 33% (n=5). Eight Norwegian patients received 4 Swedish grafts and 4 Norwegian grafts. One isolated intestinal graft was harvested outside the Scandiatransplant procurement area (Switzerland).

Conclusions: Changes in our Nordic waiting list policy appears to have shortened the waiting times, most notably for pediatric ITx candidates. No further mortalities on the WL occurred, but this may be attributed to other factors such as earlier referral for transplantation rather than changes in the shared waiting list. These data suggest an optimized utilization of the donor pool. Broadening the geographic procurement area may explain the longer cold ischemia times. Although many of the studied variables showed clear tendencies as mentioned above, they were not statistically significant. This may be due to the low number of observations.

| Table 1: Waiting List Characteristics (Intestinal Organ Allocation in Sweden) |
|---------------------------------|---------------------------------|
| ERA 1 (n=22) | ERA 2 (n=23) |
| Age (mean) | 42.6 (18.3) | 46.5 (20.7) |
| Body weight (mean) | 77.5 (34) | 78.5 (29.1) |
| Cold ischemia time (mean) | 231 (162-343) | 218 (160-343) |
| Percentage of "important grafts" | 39% (n=8) | 44% (n=10) |
| Pediatric mortality on waiting list | 44% (n=2) | 25% (n=6) |

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P2.44 - Vedolizumab for IBD-like inflammation after intestinal transplantation: does it work?

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Introduction: Vedolizumab (VEDO) is a monoclonal antibody selectively blocking gut lymphocyte migration by blocking the α4β7 integrin. VEDO has proven its efficacy in inflammatory bowel disease (IBD). Based on the selective mode of action we tested this molecule to control chronic IBD-like inflammation.

Methods: Between September 2016 and January 2017, four patients presenting with IBD-like inflammation of the small bowel/colon resistant to conventional treatment were started on VEDO (standard induction therapy at W0-2 6 (6mg/kg) followed by maintenance therapy every 4-8 weeks). Patients were followed one year after VEDO start.

Results: All four patients had received an isolated small bowel transplant for definitive intestinal failure. All four patients had failed conventional treatment for IBD-like ileocolic chronic inflammation. Median age at Vedolizumab onset was 11 years (5-20). Three patients were on triple immunosuppression with steroids, tacrolimus and azathioprine, one on steroids and tacrolimus. They received VEDO induction therapy. No adverse events were reported. Maintenance therapy was started with variable intervals and adjusted to clinical outcome. During maintenance one patient experienced a Norovirus infection and two patients had ITx rejection. The first rejection occurred after the second maintenance infusion, justifying suspension of Vedolizumab and treatment with high dose steroids, thymoglobulin and Infliximab. In the second patient rejection was treated with high dose steroids, thymoglobulin and plasmapheresis, the patient continued Vedolizumab after rejection control. Another patient stopped vedolizumab after the first maintenance infusion because of inefficacy. Two patients completed one year of treatment. Both patients stopped the treatment 15 months after its initiation because of inefficacy. The histological analysis is ongoing to determine the immunological patterns before, during, and after VEDO treatment.

Discussion: The results of treatment for IBD-like inflammation in ITx with VEDO are disappointing. This drug do not prevent form ITx rejection relapse. Immunological pattern to elucidate possible mechanisms are currently under investigation.
P2.45 - Apraglutide Has an Extended Duration and Induces a Greater Intestinotrophic Effect Compared with Teduglutide, Glepaglutide and Elsiglutide

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Introduction: To assess the impact of the long (30 hour) half-life of apraglutide (FE 203799) on intestinotrophic effect and duration of effect compared with teduglutide, glepaglutide and elsiglutide in a rat model.

Methods: Apraglutide was directly compared to teduglutide, glepaglutide and elsiglutide in Sprague-Dawley rats. The compounds, at equivalent dose levels of 30 or 300 nmol/kg (n=6/group), were tested at intervals of 24 hours (once daily for 5 days) or 48 hours (2 doses at times 0 and 48 hours). Rats were euthanized 96 hours after the first dose. The compounds were also tested after a single injection with the rats euthanized 72 or 96 hours post-dosing. Intestinal wet weight was normalized to body weight and was expressed as % increase over a control group run in the same study.

Results: At 24, 48 and 72 hour dosing intervals, apraglutide induced a greater intestinotrophic effect compared to teduglutide, elsiglutide and glepaglutide at identical doses.

At a 96 hour dosing interval, apraglutide at 300 nmol/kg increased intestine weight over the control group. The effect of apraglutide to increase intestine weight was greater than teduglutide at identical dose. Elsiglutide and glepaglutide were not tested at a 96 hour dosing interval.

At the 300 nmol/kg dose, 96 hour dosing interval, teduglutide treated rats had a decrease in intestine weight.

Conclusions: This data indicates that apraglutide has the most robust and longest lasting pharmacodynamic effect of the compounds tested. Apraglutide is currently in Phase II development for patients with short bowel syndrome requiring parenteral support.

P2.46 - RESTORE project (improve underSTanding of small bowel syndRomE in Argentina): First report of a prospective, observational, epidemiological, multicenter study of adult patients with Short Gut Syndrome in Argentina.

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Introduction: There is no centralized registered information regarding patients (pts) suffering from Short Bowel Syndrome (SBS) in Argentina. Estimations based on other countries epidemiology predict an incidence of 8-10 new adult pts/year. In order to compile this information, we started a prospective multicenter observational and epidemiological registry for adults with SBS, independent from the etiology and the treatment proposed. We aim to present the first report of the RESTORE project.

Methods: From 2015 to 2017, design, funding, Institutional Review Board and Ethical approvals were obtained. Since June 2017, 11 centers started activity. Data collection was made using case report forms; a monitor visited and supervised each center initiation and performance. The registered pts are followed at each center at weeks 4, 8, 12, 20 and 24, and yearly thereafter. Dead, intestinal adaptation and transplant have been considered as end-points of the study. The principal investigator developed a prospective database. Statistical analysis was done on SPSS v20.0.

Results: 12 centers were initiated, 11 enrolled 42 pts; 33 pts were actively monitored and analyzed. 12 pts completed 1 year of follow up (FU); 19 pts (57.6%) were female; mean age: 52.15 ± 15.4 years. Diagnoses in Figure 1. Mean intestinal length: 515 ± 42.4 cm. Anatomy type: T1: 22 pts; T2: 8 pts and T3: 3 pts; ileo-cecal valve was present in 11 pts. Colon in 27 pts; Ostomies in 22 pts. Mean ostomy output: 158 ± 78 ml/day; Autologous GI tract Reconstruction Surgery (AGIRS) was done in 11 pts before enrollment. Meantime on parenteral nutrition (PN) before enrollment: 27.4 ± 37.8 months; PN and biochemical variables overtime are shown in Table 1. Espen Clinical Classification in Figure 1.
Treatments proposed at first visit: PN + Medical Rehabilitation: 20 pts (60%), AGIRS: 4 pts (12%); post-surgical teduglutide (TED): 8 pts (24%); transplant: 1 pt (3%); at the end of FU: 1 pt was lost of FU; 4 pts adapted with AG IRS alone, or in combination with TED. One pt was transplanted and another pt, listed due to liver disease. The overall actuarial survival is 87%; sepsis was the main cause of death (3/4 pts).

Conclusions: Although this report still has a limited number of centers, it has registered more SBS adult patients than expected according to theoretical estimations; reinforcing the importance of having registries to understand the behavior of SBS patients overtime and their outcome.

P2.47 - Minimizing intestinal resection strategy for prevention of short bowel syndrome in surgery for chronic radiation enteritis: Shanghai experience

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Introduction: Small intestinal fibrosis induced by chronic radiation enteritis (CRE) is a major complication following radiotherapy and an important underlying disease of short bowel syndrome. This study is to investigate the effect of modified surgical strategy on preventing both major small bowel resection followed by late-term short bowel syndrome and short-term postoperative anastomotic leakage. Methods: We divide all CRE patients with pelvic radiation enteritis-induced intestinal stenosis (REIS) and intestinal obstruction macroscopically into two subtypes:

Type I. Terminal ileum lesion with or without proximal intestine injury (figure 1).

Type II. Isolated intestinal lesion. Records of all 22 type I patients from Apr. 2017 to Dec. 2018 were analyzed. All the patients underwent minimal resection of small intestinal lesion with primary anastomosis between ileum segments and protective proximal loop enterotomy (figure 2).

Postoperatively, stoma effluents were recycled through distal loop where the anastomosis were located. Intraoperative information and postoperative complications were retrospectively analyzed, including patients’ general information, intraoperative bleeding volume, length of resected small bowel, adhesion grade, postoperative complication, postoperative hospital stay, postoperative recovery to total enteral nutrition and recycling time of stoma effluents.
Results: All the operations were performed. Length of resected small bowel was 25-100 cm. No severe complications like anastomotic leakage were observed postoperatively. All the patients recovered to total enteral nutrition in 5-14 days and began ileostomy recycling in 5-14 days. Postoperative stay was 9-23 days. During a follow-up of 1-21 months, one patient died of cancer recurrence and all the other patients kept well nourished.

Conclusion: Protective proximal loop enterotomy is feasible for the surgery of radiation enteritis presenting with terminal small bowel fibrosis. This strategy can both prevent severe postoperative complications and reduce extensive intestinal resection and thus improve both peri-operative recovery and long-term outcome.

P2.48 - Safety and efficacy of mTOR inhibitors following intestine and multivisceral transplantation
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Purpose: Review the indications, outcomes and frequency of the use of mTOR inhibitors (mTORs) after intestine (IT) and multivisceral transplantation (MVT) assessed across two transplant centers in the United States.

Methods: We evaluated patients receiving everolimus or sirolimus following MVT or IT and compiled retrospective data between 2009-2019 at multiple transplant centers.

Results: 22 patients received immunosuppression including an mTOR. Twenty patients were over 18 years. Twelve patients received isolated IT. The most common reason for transplant was short gut syndrome (45%) followed by dysmotility (22%) and neuroendocrine tumor (23%). Mean age at transplant was 46 years (range 22-62) in adults and 2.5 in children. 54% patients were started on mTORs beyond 1 year post transplant. Mean time from transplant to initiation of mTOR was 24 months (range 1-78). 63.6% received sirolimus and 36.4% received everolimus. Other medications at the time of mTORs initiation included tacrolimus, prednisone, mycophenolate mofetil (MMF) and azathioprine. 18%, 13.6% and 59% of patients on mTORs were able to discontinue MMF, prednisone or reduce tacrolimus respectively. Only one patient was weaned to mTOR as single immunosuppression agent. Reason for mTOR use was renal dysfunction in 59% of cases; of these, only one patient had chronic kidney disease pre transplant. Mean glomerular filtration rate (GFR) prior to mTORs initiation was 40 mL/min/1.73sqm. Of those placed on mTORs for renal insufficiency 69.2% had substantial improvement in GFR (defined as increase of 10 points). Nine patients developed some worsening of proteinuria but none were taken off treatment due to this. 27.3% cases developed acute cellular rejection (ACR). 13.6% had cytomegalovirus and two patients died while being on mTOR therapy. mTORs were discontinued in 22 cases due to side effects (54.5%), surgeries (37.5%) or ACR (9%). Sirolimus was discontinued in 8/14 (57%), everolimus was discontinued in 3/8 (37.5%). The mean duration of mTORs use in those stopping therapy was 7 months and 18 months in those remaining on therapy.

Conclusion: Tolerance of therapy remains challenging with almost a third of those started on mTORs unable to tolerate long term treatment due to side effects. If well tolerated, mTORs were generally safe and efficacious following IT and MVT. Further studies with a control group are warranted.
Long-term parenteral nutrition (PN) is essential for the management of paediatric chronic intestinal failure: early experience of a single-centre intestinal rehabilitation program in Singapore

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Background: Long-term parenteral nutrition (PN) is essential for the survival of children with intestinal failure (IF) but it contributes to morbidity & economic burden. We aimed to characterise the clinical & financial impact of long-term PN burden in the 1st 5 yrs of our intestinal rehabilitation program established in 2014 at a single-centre in Singapore.

Methods: A retrospective cohort review of children with IF who had long-term (>90 days) PN & transitioned to home-PN (HPN) from 2014 – 2018 was done. All complications related to long-term PN & the underlying gastrointestinal/surgical disorder were collected. Financial data was retrieved from the hospital’s business office. Costs were categorized into 3 groups: related to PN, underlying disease (UD)–related including investigations, surgery, intensive care, & complications from UD; or unrelated. In Singapore, healthcare cost is borne by the patient, with varying proportion of government subsidy.

Results: 38 children (23 girls) with IF received long-term PN for median duration of 136 (0.27-7.5) yrs. Aetiologies included necrotizing enterocolitis 50%, paediatric intestinal pseudo-obstruction 27%, congenital mucosal disorders 11%, malrotation 5.6% & inflammatory bowel disease 5.5%. The median age at PN-onset was 0.66 (0.03-68) months and at HPN-onset was 2.19 (0.63-7.26) years. 11 patients received hospital-PN while 17 were transitioned to HPN. 64% achieved enteral autonomy after a median duration of 133 (0.27-6.95) years of PN support. Central line-associated bloodstream-infections affected 77% of patients with an incidence rate of 4.6/1000 catheter-days. Other major complications were recurrent venous thrombosis 50% & intestinal failure-associated liver disease 38%. One patient died from sepsis at 6 months of age, no patient has undergone intestine or liver transplantation.

The overall median cost of managing children with IF for the 1st year was US$350,232.30/patient. It dropped to US$59,331.01 in the 2nd yr & US$30,349.91 in the 3rd yr. In the 1st yr, 95% of costs were UD-related while 5% were PN-related. Subsequently, the proportion spent on PN was 43% while costs due to UD dropped to 45% (Fig).

Conclusion: Long-term PN therapy, whilst an effective life-saving option for paediatric IF, incurs substantial financial burden to families and the healthcare system. These findings should form the basis of a cost-benefit analysis of initiating an intestinal/multi-visceral transplantation program for patients with irreversible IF.
P2.51 - A training program for SBS patients treated with GLP2 agonist (Teduglutide) elaborate by a multidisciplinary team

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SBS is the main cause leading to chronic intestinal failure. In France, Teduglutide, a GLP2 agonist is available since 2015. A multidisciplinary team (in particularly 5 specialized nurses, physicians, and a “partner” SBS patient) created an educational program for patients who start Teduglutide. The aim of our study is to describe the program and evaluate his feasibility.

Methods: In a reference centre with a cohort of 280 patients with CIF, 42 SBS patients started Teduglutide between last 2015 and December 2018. The multidisciplinary team have conceived a training program dedicated to teach a) to reconstitute and inject the drug b) clinical and biological parameters to monitor at home and c) to recognize the main signs of complications and/ or efficacy requiring a call or venue to hospital for HPN or drug adjustment.

Results: The program included a) a written guide (“my patient book”) with information about SBS, drug monitoring, measurement sheets of quantifications, b) an injection training session (hand’ on training and video film) d) a smartphone application (Android & iOS) developed together with the patient association “La vie par un fil” called “Mon Suivi – La Vie par un fil” associated to a training video to monitoring (input - output balance, weight, vital parameters, etc.). Forty five patients had started Teduglutide. At the end of one training session, 28/45 patients were considered as totally autonomous in terms of a) injection of Teduglutide, b) knowledge of SBS, action of drug, clinical parameters requiring a monitoring at home and c) principal actions to do in specific situations such as dehydration, fluid overload, abdominal pain. Ten/ 45 patients were trained to use the Smartphone app for the monitoring and used it regularly.

Conclusion: The arrival of new treatments in SBS is really challenging because rapid changes in digestive parameters may require faster adaptation of parenteral support than usual. A complete training program combining human resources, digital and written tools allows the increase of SBS patient autonomy. The next step should be an impact analyse of the impact of the program on mid and long term terms changes such as hospitalisation rate, global adhesion to SBS treatment and quality of life.
Complete Jejunal Exclusion: A novel method to manage Enterocutaneous Fistulas in the background of short bowel syndrome

Sidharth Sharma, Regina Hwang, Peter Liou, Monica Velasco, Shilpa Ravella, Mercedes Martinez, Adam Griesemer, Tomoaki Kato, et al.

Columbia University of Physicians and Surgeons, NY, USA

Introduction: Management of enterocutaneous fistulas (ECF) in patients with short bowel syndrome (SBS) is challenging. High output fistulas are unlikely to close spontaneously; however, the success rate of surgical intervention is limited due to poor nutrition, inflammation, limited bowel length and lack of abdominal wall integrity. Here, we describe 8 cases of utilizing complete jejunal exclusion at the ligament of Treitz with a gastrostomy tube drainage to manage the ECF as a temporary measure for subsequent bowel reconstruction or transplant. To our knowledge, this is the first report of this approach.

Methods: Eight patients who underwent jejunal exclusion at our institution were retrospectively reviewed. In all cases, the jejunum was divided 1-2 inches distal to the ligament of Treitz. Both ends were oversewn. A gastrostomy tube was placed for retrograde decompression of proximal enteric secretions without pyloroplasty. In 4 cases of active ECF's, we accessed the ligament of Treitz via a left subcostal incision to avoid the fistula.

Results: The procedure was done for active ECFs (n=4), impending fistulas (n=3), bridge to transplant following desmoid tumor resection in Gardner’s syndrome (n=2), and after graft enterectomy (n=2). Four cases of active ECFs were managed elsewhere without success. All patients were discharged home on TPN. Of the 4 ECF cases, the fistula output decreased substantially in all patients and closed entirely in 2 cases. Two underwent successful reconstruction and 2 are awaiting reconstruction. One who had secondary biliary cirrhosis due to biliary obstruction and TPN induced liver disease died while waiting for multivisceral transplant (MVT). Of the desmoid tumor cases, one underwent MVT, is alive and well 8 years after transplant, while the other refused to undergo MVT and is living with TPN and g-tube decompression 6 years after the procedure. The patient who underwent graft enterectomy is listed for MVT-this patient developed acute pancreatitis after the jejunal exclusion and required G-J tube decompression.

Conclusions: Jejunal exclusion is an effective and generally well tolerated option in the management of intractable ECF in patients with SBS. Associated complications include pancreatitis and liver failure. This procedure should be considered as a bridge to intestinal rehabilitation or transplantation in patients with ECFs that fail to close with medical or traditional surgical management.

<table>
<thead>
<tr>
<th>Status</th>
<th>Diagnosis</th>
<th>Length of small bowel lost (cm)</th>
<th>ECF</th>
<th>TPN</th>
<th>TPN status</th>
<th>Length of stay (days)</th>
<th>Procedure (s)</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>SMA thrombosis</td>
<td>23</td>
<td>Yes</td>
<td>Yes</td>
<td>Off</td>
<td>32</td>
<td>Uncharged home</td>
<td></td>
</tr>
<tr>
<td>Case 2</td>
<td>Minor/Valvular</td>
<td>50</td>
<td>No</td>
<td>Yes</td>
<td>On</td>
<td>35</td>
<td>Discharged home</td>
<td></td>
</tr>
<tr>
<td>Case 3</td>
<td>Mitral/Valvular</td>
<td>5cm</td>
<td>No</td>
<td>Yes</td>
<td>On</td>
<td>25</td>
<td>Discharged home</td>
<td></td>
</tr>
<tr>
<td>Case 4</td>
<td>Recurrent pancreatitis</td>
<td>25</td>
<td>Yes</td>
<td>Yes</td>
<td>On</td>
<td>41</td>
<td>Discharged home</td>
<td></td>
</tr>
<tr>
<td>Case 5</td>
<td>Severe small bowel obstruction following bowel injury</td>
<td>60</td>
<td>Yes</td>
<td>Yes</td>
<td>TPN-advised with TPN</td>
<td>35</td>
<td>Discharged home</td>
<td></td>
</tr>
<tr>
<td>Case 6</td>
<td>Transplant graft failure</td>
<td>50</td>
<td>No</td>
<td>Yes</td>
<td>Off</td>
<td>45</td>
<td>Uncharged home</td>
<td></td>
</tr>
<tr>
<td>Case 7</td>
<td>Mesodigonal/Thrombotic</td>
<td>50</td>
<td>No</td>
<td>Yes</td>
<td>Off</td>
<td>31</td>
<td>Discharged home</td>
<td></td>
</tr>
<tr>
<td>Case 8</td>
<td>Intestinal Obstruction/Bowel</td>
<td>50</td>
<td>Yes</td>
<td>Yes</td>
<td>On</td>
<td>29</td>
<td>Discharged home</td>
<td></td>
</tr>
</tbody>
</table>
P2.53 - SMOFlipid in pediatric patients with intestinal failure: a single center experience from the United States

Sivan Kinberg, Kimberly Law, Iliana Martin, Eunie Park, Christine Haro, Elizabeth Breza, Mercedes Martinez
Columbia University Irving Medical Center

Introduction: The soy-based lipid component of parenteral nutrition (PN) is a known contributing factor to intestinal failure associated liver disease (IFALD). Although effective in reducing hepatotoxicity, lipid minimization (LM) protocols predispose to essential fatty acid deficiency (EFAD). SMOFlipid (soybean oil, medium-chain triglycerides, olive oil, fish oil) was FDA approved for adult patients with intestinal failure (IF) in the United States in 2016 and contains a more favorable ratio of omega-3 to omega-6 fatty acids compared to Intralipid (IL). We reviewed our experience with SMOFlipid in pediatric patients with IF.

Methods: A retrospective chart review was conducted on pediatric patients with IF and a Center who switched from IL to SMOFlipid between 2016-2017. We reviewed data before and 3-6 months after initiation of SMOFlipid (demographics, IF etiology, PN components, liver biochemistry tests, fatty acid profiles, growth parameters, micronutrient labs, abdominal ultrasound and liver histology).

Results: Sixteen eligible patients were included in this analysis (median age: 4 years, range: 4 months-10 years, 50% male). After switching to SMOFlipid, the mean fat component of PN increased (0.4 g/kg/day vs. 1 g/kg/day, P<0.001), glucose infusion rate (GIR) decreased (1.3 mg/kg/min vs. 1.4 mg/kg/min, P=0.01), serum α-tocopherol levels increased (6.8 mg/L vs. 9.2 mg/L, P=0.01), EFAD improved (mean triene:tetraene 0.07 vs. 0.04, P<0.001), and growth improved (mean BMI/Weight-Length Z-score 0.11 vs. 0.95, P=0.03). There was some improvement in liver biochemical tests, despite increased lipid administration. Two patients had resolution of abnormal sonographic findings, and one patient had histologically-proven resolution of hepatic steatosis and fibrosis. The average cost was $2.70/day for IL and $14.40/day for SMOFlipid (P<0.001). SMOFlipid was not discontinued in any of the patients and no adverse effects were observed.

Conclusion: An increased dose of a balanced lipid emulsion was associated with improved EFAD, improved growth, decreased GIR and increased serum α-tocopherol levels, while not worsening IFALD. Although the daily cost of SMOFlipid is higher than IL, the benefits offered by its better anti-inflammatory profile may prove to outweigh the higher cost. Based on our single-center experience, we recommend the routine use of SMOFlipid in pediatric patients with IF to avoid the potentially harmful effects associated with IL and LM protocols.

P2.54 - Re-analysis of a randomized placebo (PBO) controlled trial of intravenous (IV) choline chloride for IFALD using state-of-the-art analytic and imaging methods and contemporary definition of IFALD


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3 Pharmapace, San Diego, CA, United States
4 University of Illinois, Chicago, IL, United States

Introduction: IFALD, defined as steatosis and cholestasis, is associated with significant morbidity in patients dependent on parenteral nutrition (PN). Choline is an essential nutrient that is a fundamental component of cell membranes, VLDL for triglyceride export from the liver, and normal bile. PN products do not contain sufficient choline. Buchman et al (2001) determined that IFALD may be reversible in choline-deficient patients. The current study reanalyzed, transformed and analyzed data from Buchman et al 2001 using state of the art methods to inform a confirmatory phase 3 trial

Methods: Data were imported from original source documents into an electronic eCTD format. Because MRI-Proton Density Fat Fraction (MRI-PDFF) has become the gold standard method for non-invasive quantification of hepatic steatosis, CT data were transformed to MRI-PDFF using an established linear equation (Kramer et al, 2017). The trial was then re-analyzed using MMRM statistical approach, and in two subgroups of patients meeting contemporary definitions of IFALD. Missing data were reasonably assumed to be missing at random.

Results: This was a study of adults dependent on PN for many years (11.8 years). IV Choline Chloride was safe and well-tolerated. Baseline MRI-PDFF values (mean 13.6%, range [9.8-38.3%]) demonstrate moderate to severe fatty liver in the study population. The benefit of IV Choline Chloride vs PBO achieves significance or trend-significance from Weeks 4-24 despite small sample size. In the sub-group analyses, improvement in ALP is consistent and substantial, with 20-30% improvement over 12-24 weeks of treatment.

Conclusion: Using MRI-PDFF, a contemporary definition of IFALD, modern analytic approaches, and source-verified data, the original findings from Buchman et al 2001 are replicated and extended. These robust results in a rare, but serious condition affecting PN patients can inform future trials attempting to replicate these promising results in a larger cohort to support the development of a safe, choline PN product.
P2.55 - Decreasing Tacrolimus Time-In-Therapeutic Range is Associated with an Increased Incidence and Severity of Rejection Following Intestinal Transplant

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2 Department of Pharmacy, Mount Sinai Hospital, New York, NY, USA
3 Department of Population Health Science and Policy, Mount Sinai Hospital, New York, NY, USA

Introduction: Acute rejection (AR) is a major complication in intestinal transplantation (ITX) that can lead to graft loss. Tacrolimus (TAC) is a mainstay of immunosuppression following ITX to prevent AR. Drug monitoring is routinely employed to ensure adequate exposure to TAC, while limiting adverse effects. Despite established practice, there is limited direct evidence linking therapeutic TAC levels with improved clinical outcomes post-ITX.

Method: This was a single-center review of all adult non-liver containing ITX graft recipients from 2010-2017, who maintained a functioning graft for 1 month. Patients received anti-thymocyte globulin (ATG) induction, with plasmapheresis and IVIG for positive T-cell CDC cross matches (CXM). Maintenance immunosuppression included TAC and prednisone, with mycophenolate or sirolimus added at physician discretion. 12-hour TAC whole blood trough levels were monitored daily in the hospital and every 1-4 weeks following discharge, with target levels of 12-17ng/mL month 1, 10-15ng/mL months 2-3, 8-15ng/mL months 3-6, and 8-12ng/mL months 6-12 post-ITX. Percent time-in-therapeutic range (TTR) for TAC was calculated from the date of transplant until 1 year post-ITX or graft failure using Rosendaal’s method (Rosendaal. Thromb Haemost. 1993;69:236-239.) and patients were divided into TTR quartiles. Multivariable regression was used to identify variables associated with the incidence of AR on biopsy tissue within 1 year of ITX. Additional variables assessed included: graft type, adjunctive immunosuppression, age, sex, ethnicity, indication for ITX, prior transplants, cold ischemia time, DSA > 2,000 MFI, and positive CXM.

Results: A total of 51 patients were included in the analysis, 11 of which had pre-transplant DSA, and 6 patients had a positive CXM at time of ITX (Table 1). Mean TAC-TTR for the cohort was 29.9% ± 11.2, and 19 episodes of AR were observed in the first year post-ITX. Logistic regression demonstrated TAC-TTR <30% was associated with an increasing risk for any rejection episode (Table 2). A similar trend were observed for severe rejection episodes requiring ATG, with TAC-TTR <20% associated with a greater risk for severe rejection (p=0.02; HR: 7.7 [95% CI: 0.9, 65.3]).

Conclusion: Our dataset suggests that decreasing TAC-TTR may be a risk factor for both the occurrence and severity of rejection. If confirmed, our results indicate a need for utmost vigilance in TAC trough monitoring within the first-year post-ITX.

Table 1 – Baseline Characteristics

<table>
<thead>
<tr>
<th>Baseline Characteristics</th>
<th>Final Cohort (n= 51)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Recipient Age, years</td>
<td>46.7 ± 10.1</td>
</tr>
<tr>
<td>Donor Age, years</td>
<td>11.6 ± 10.5</td>
</tr>
<tr>
<td>Male Recipient, Sex</td>
<td>23 (45.1%)</td>
</tr>
<tr>
<td>Recipient Ethnicity</td>
<td></td>
</tr>
<tr>
<td>White/European</td>
<td>34 (66.7%)</td>
</tr>
<tr>
<td>African-American</td>
<td>6 (11.8%)</td>
</tr>
<tr>
<td>Hispanic/Latino</td>
<td>9 (17.6%)</td>
</tr>
<tr>
<td>Asian/Pacific Island</td>
<td>2 (3.9%)</td>
</tr>
<tr>
<td>Graft Type</td>
<td></td>
</tr>
<tr>
<td>Isolated Intestine</td>
<td>40 (86.3%)</td>
</tr>
<tr>
<td>Intestinal-Kidney</td>
<td>7 (13.7%)</td>
</tr>
<tr>
<td>Intestinal-Pancreas</td>
<td>4 (7.8%)</td>
</tr>
<tr>
<td>Etiology of Short-Gut-Syndrome</td>
<td></td>
</tr>
<tr>
<td>Crohn’s Disease</td>
<td>12 (23.5%)</td>
</tr>
<tr>
<td>Vascular Thrombosis</td>
<td>11 (21.6%)</td>
</tr>
<tr>
<td>Trauma</td>
<td>2 (3.9%)</td>
</tr>
<tr>
<td>Dysmotility Syndrome</td>
<td>8 (15.7%)</td>
</tr>
<tr>
<td>Failed Prior Transplant</td>
<td>6 (11.8%)</td>
</tr>
<tr>
<td>Other</td>
<td>12 (23.5%)</td>
</tr>
<tr>
<td>Adjunctive Immunosuppression</td>
<td></td>
</tr>
<tr>
<td>Mycophenolate</td>
<td>27 (52.9%)</td>
</tr>
<tr>
<td>Sirolimus</td>
<td>12 (23.5%)</td>
</tr>
<tr>
<td>Mean Cold-Ischemia Time, minutes</td>
<td>435.2 ± 77.8</td>
</tr>
<tr>
<td>Pre-Transplant DSA</td>
<td>11 (21.6%)</td>
</tr>
<tr>
<td>CDC Cross Match</td>
<td></td>
</tr>
<tr>
<td>B-/T-</td>
<td>45 (88.2%)</td>
</tr>
<tr>
<td>Br/T-</td>
<td>2 (3.9%)</td>
</tr>
<tr>
<td>Br/T+</td>
<td>4 (7.8%)</td>
</tr>
<tr>
<td>Graft Loss Prior to 1 Year</td>
<td>14 (27.4%)</td>
</tr>
<tr>
<td>Overall Incidence of Rejection</td>
<td></td>
</tr>
<tr>
<td>Any Rejection</td>
<td>19 (37.2%)</td>
</tr>
<tr>
<td>Severe Rejection</td>
<td>11 (21.6%)</td>
</tr>
<tr>
<td>Tacrolimus Dosage</td>
<td></td>
</tr>
<tr>
<td>Time-In-Therapeutic Range</td>
<td>28.9% ± 11.2</td>
</tr>
<tr>
<td>Time Supratherapeutic</td>
<td>34.2% ± 15.6</td>
</tr>
</tbody>
</table>

Table 2 – Multivariable Logistic Regression Model for Any Acute Rejection

<table>
<thead>
<tr>
<th>Parameter</th>
<th>p-Value</th>
<th>Hazard Ratio</th>
<th>95% Confidence Interval Limits</th>
</tr>
</thead>
<tbody>
<tr>
<td>TAC-TTR &lt;30%</td>
<td>0.01</td>
<td>2.7</td>
<td>1.4</td>
</tr>
<tr>
<td>Positive CXM</td>
<td>&lt; 0.01</td>
<td>2.6</td>
<td>1.4</td>
</tr>
</tbody>
</table>
Introduction: Thirty-seven intestinal transplant (ITx) programs have performed >1 pediatric ITx since 1990 according to the 2017 United Network for Organ Sharing (UNOS) database. The median number of ITx per center was 8. Our institution is an aggressive center for pediatric intestinal transplantation (ITP), with relatively low volumes for ITx. We hypothesized that outcomes from a medium-size program like ours compared favorably to national data.

Methods: A retrospective review of all ITx done at a main pediatric IRP and ITx program was performed (2004–2018). Demographic, medical, and surgical data were collected. National data was obtained from the UNOS database (n=500). Mann-Whitney and Chi-Square tests were used for analysis. p<0.05 was considered significant. Institutional Review Board approval was obtained (IRB #2019-2474).

Results: Since 2004, 18 ITx were performed at our institution. The proportion of girls at our center, 8/18 (44%), was identical to UNOS, 598/1346 (p=1.0). Gastroschisis was the main cause of short gut syndrome in both groups (7/18, 39% locally, vs. UNOS 34/1500, 23%, p=0.13). Mean age at transplant was similar (3.94±3.43 years, p=0.33). Mean waitlist time was longer for our patients, although not significantly (39.4±11.8 vs. UNOS 220.2±336.9 days, p=0.062). There was a trend in shorter mean distance between donor and recipient hospitals in our program (466.47±335.9 vs. UNOS 547.39±436.09 days, p=0.65) while the mean cold ischemia time was significantly shorter in our group (466.47±335.9 vs. UNOS 788.8±2.58 hours, p=0.049). There was a significant difference between groups regarding intestinal venous drainage through the portal vein for isolated small bowel ITx (3/18, 27%, vs. UNOS 247/440, 57%, p=0.005). The incidence of multivisceral transplants was significantly higher nationally (UNOS 1034/1500, 69%, vs. 220/18, 20%, p=0.001). There was no statistical difference in overall graft survival (18/18, 61%, vs. UNOS 846/1500, p=0.39) although it was higher in our group. There was a trend in improved overall patient survival in our patients (15/18, 83%, vs. UNOS 832/1347, 62%, p=0.062).

Conclusion: Timelier listing for isolated ITx is associated with a lower incidence and need for multivisceral transplantation in aggressive intestinal rehabilitation program. Shorter cold ischemia times associated with shorter travel distance between donor and recipient hospitals may contribute to higher patient and graft survival.

Introduction: Short bowel syndrome (SBS) is the main cause of intestinal failure (IF) and complex gastrochisis (CG) is the most frequent underlying cause in our population. It has been reported that children with SBS related to CG have a worse prognosis. The present work aims to analyze our results in the treatment of SBS secondary to CG and compare them with those obtained in cases of SBS related to other underlying diseases.

Methods: A descriptive, retrospective study was carried out, using data collected from institutional electronic records and the specific IF database. Inclusion criteria: patients under 18 years of age, with IF defined as parenteral nutrition (PN) dependency greater than 2 months and diagnosis of SBS. Patients were divided into 2 groups: G1 included cases of SBS associated with CG and G2 patients with SBS related to other diagnosis. Intestinal rehabilitation (sustained suspension of PN), need for intestinal transplantation and mortality were analyzed for both groups. Additionally, the influence of different exposure variables such as anatomical features of the digestive tract and adverse events related to prolonged PN, was analyzed for the group of patients with CG.

Results: We obtained a final number of 86 patients with 24 in G1 and 62 in G2. Both groups were comparable. No statistical difference in intestinal rehabilitation (G1 78.9% vs G2 57.14%) and mortality rate (G1 20.8% vs G2 20.9%) was found, although a trend to better outcome in patients with CG. Additionally, the influence of different exposure variables such as anatomical features of the digestive tract and adverse events related to prolonged PN, was analyzed for the group of patients with CG.

Conclusion: CG is the main individual cause of IF in our population. In the context of specialized multidisciplinary treatment, our results show that the majority of patients with SBS secondary to CG survive and succeed in suspending PN, with similar results as those observed in patients with SBS secondary to other entities. The preservation of the colon whenever possible, as well as the prevention of adverse events related to PN, can improve the outcome in these patients.
P2.58 - Chronic hyperammonemia an important cause of morbidity in isolated intestine transplant patients with venous outflow to the vena cava

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Indiana University School of Medicine

Background: There are limited options for superior mesenteric vein (SMV) outflow in isolated intestine transplant (ITx), the two most common being anastomosis to the recipient superior mesenteric vein or to the vena cava. Previous research comparing these two options demonstrated no difference in early clinical outcomes. This paper reports four cases of symptomatic late hyperammonemia occurring in ITx patients more than 1-year from transplant. Each affected patient experienced marked encephalopathy. In each case, a very low protein diet was effective in lowering ammonia levels and ameliorating symptoms. Each affected ITx patient had direct SMV-vena cava drainage of their graft. This study reports an analysis of all living ITx patient at this single center to determine if hyperammonia is associated with vena cava drainage alone or if it occurs generally in this population.

Methods: ITx recipients at a single center were identified. Random serum ammonia levels were obtained with routine blood draws. Ammonia levels were analyzed in relation to both (1) SMV drainage technique and to (2) time from transplant. Liver function was also assessed using standard lab values.

Results: There were 71 ITx patients between 2003 and 2018, 26 patients with SMV-SMV anastomosis (37%) and 45 with SMV-vena cava anastomosis (63%). Of these patients, 37 are currently living (52%). For SMV-vena cava patients, the available ammonia levels were consistently greater than 50mcg/dL. Among affected patients, the peak levels would reach as high as 150-200mcg/dL. For SMV-SMV patients, the levels were always less than 50mcg/dL. There was one SMV-SMV patient with intermittent encephalopathy, but she had peak ammonia levels of 60mcg/dL. She was ultimately diagnosed with a rare neurocognitive disorder.

Conclusions: This study demonstrates that hyperammonemia is not uncommon among late survivors of ITx, but only in those patients with SMV to vena cava anastomosis. This hyperammonia may result in marked encephalopathy as seen in four patients in this cohort. A very low protein diet is effective in minimizing this process. In ITx patients with SMV-vena cava anastomosis, routine monitoring of serum ammonia levels is indicated.
P2.59 - Simultaneous Serial Transverse Enteroplasty (STEP) in Size Mismatch Small Bowel Transplantations

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4 Division of Liver/Gastrointestinal Transplantation, Miami Transplant Institute, University of Miami Miller School of Medicine, Jackson Memorial Hospital, Miami, FL, USA.
5 Division of General, Visceral, and Transplantation Surgery, University Hospital of Heidelberg, Heidelberg, Germany.

Background: Small bowel transplantation (SBTX) in children receiving larger grafts from adults can be challenging because of size mismatch. The aim of the present study was to assess whether a simultaneous serial transverse enteroplasty (STEP) can address the problem of size mismatch.

Methods: Three different size ratio groups between donors and recipients were compared in a porcine model with a 14-day follow-up. The groups were size matched, size mismatched (13.8 weight ratio), and size mismatched+STEP (each n = 8).

Results: It was technically feasible to simultaneously perform a STEP and SBTX of a mismatched intestinal segment. The postoperative clinical course was uneventful. No signs of bleeding, leakage, stenosis, or ileus were observed and the intestinal segment was well perfused at relaparotomy. Body weight decreased in all groups, but the percentage decrease was lowest in the mismatched+STEP group. Vital enterocyte masses were similar in all the groups (citrulline levels) and the nutritional status was best in the STEP group (transferrin levels, p = 0.04).

Conclusions: We have demonstrated that a simultaneous STEP and SBTX procedure is technically feasible and clinically useful in overcoming the challenges associated with size mismatched SBTX. Our short-term findings justify further investigation in a larger series to elucidate the long-term outcomes of this procedure.
POSTERS PRESENTED ON FRIDAY JULY 5, 2019
P3.21 - Fibrinolytic Shutdown Is Associated With Intraoperative Thrombosis and Hemorrhage During Visceral Transplant
Ramona Nicolau-Raducu, Yehuda Raveh
University of Miami/ Jackson Memorial Hospital

Introduction: End stage liver disease (ESLD) is characterized by a precarious imbalance of hemostasis. Detrimental consequences of hypofibrinolysis, also known as fibrinolytic shutdown, have been recently demonstrated, and its significance in visceral transplant remains unknown.

Method: To fill this gap, following IRB approval, this retrospective study included 49 adult recipients of visceral allografts (14 without the liver, 35 multivisceral (MVT) with the liver) transplanted between 2010-2018 in a single university hospital, and for whom pre-incisional thromboelastography was available. Based on % clot lysis 30 min after maximal amplitude, patients were stratified into 3 fibrinolysis phenotypes: fibrinolytic shutdown, physiologic fibrinolysis, and hyperfibrinolysis.

Results: Fibrinolytic shutdown occurred in 57% of patients, with higher incidence in recipients of multivisceral (69%), compared to visceral (29%) allografts (P=.04). Of the 35 MVT, 26% had normal liver function, 46% had parenteral nutrition-associated liver disease and 28% had severe ESLD with extensive portal vein thrombosis. Fibrinolytic shutdown was statistically associated with ESLD, as incidence of fibrinolytic shutdown in the presence or absence of ESLD was 73% versus 39% (P=0.02). Intraoperative thrombosis (18%) occurred only with MVT, and accounted for 36% of in-hospital mortality. Intraoperative thrombosis occurred solely in recipients with an abnormal fibrinolytic phenotype (fibrinolysis shutdown- 8/9 and hyperfibrinolysis- 1/9). A clinically meaningful reduction in incidence of intraoperative thrombosis was noted in recipients who received iv heparin thromboprophylaxis. Logistic regression identified pretransplant platelet count as a risk factor for fibrinolytic shutdown (OR 0.992, 95%CI 0.984-0.998; χ²=7.8, P=0.005).

Conclusions: This study highlights fibrinolytic shutdown as a dominant and clinically important feature of the hemostatic imbalance in recipients undergoing visceral transplantation.

P3.22 - Current picture of intestinal transplantation for intestinal failure in Japan
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4 Division of Hepato-Biliary-Pancreatic and Transplant Surgery, Department of Surgery, Graduate School of Medicine, Kyoto University, Kyoto, J apan
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6 Division of Gastroenterological Surgery, Department of Surgery, Asahikawa Medical University, Asahikawa, J apan
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Introduction: The prognosis of intestinal failure has improved dramatically in the past few decades with the development of parenteral nutrition. However, PN-dependent patients still have numerous complications. Intestinal transplantation (ITx) is able to significantly improve their prognosis and quality of life. As the ITx has become covered by national health insurance in 2018 in J apan, the number of ITx is expected to increase in the near future. Therefore, we report on the current picture of the ITx for intestinal failure in J apan.

Methods: The ITx have been performed in J apan since 1996. Standardized form were sent to all known ITx programs, asking for information on ITx performed between 1996 and 2017. All programs responded. Patient and graft survival estimates were obtained using Kaplan-Meier method and analyzed with Wilcoxon statistics.

Results: Five institutes provided data on 26 isolated ITx and one simultaneous liver and intestinal transplant in 23 Patients. There were 14 cadaveric and 13 living related donor transplants (Figure 1). Causes of intestinal failure included short bowel syndrome (n=9), motility disorders (n=9), re-transplantation (n=3), and other (n=2). The median age at ITx was 35.2 years (ranged 0.7 to 35 years). The overall 1-, 5- and 10-year patient survival rates were 88%, 70% and 5.8%, respectively. (Figure 2). The overall 1-, 5- and 10-year graft survival rates were 80%, 58% and 39%, respectively. More than 80% of all current survivors discontinued PN with satisfactory performance status.

Figure1
Conclusions
ITx has become an effective therapy for patients with intestinal failure who cannot tolerate PN in Japan. Further improvements are expected with early referral due to suitable donor organ and pre-transplant management.

P3.23 - Copper deficiency in infants with intestinal failure: hematological manifestations
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Introduction: Hematological cytopenias have been reported in copper-deficient children with intestinal failure (IF) and receiving parenteral nutrition (PN). However, no cohort study has investigated a possible association between copper deficiency and low blood cell count in patients who had their copper plasma levels monitored according to a standard protocol. We investigated how much copper plasma levels influence anemia, neutropenia and thrombocytopenia in children receiving long-term home PN.

Methods: Children with IF admitted to a pediatric intestinal rehabilitation program and who were receiving home PN were followed-up prospectively from July 2015 to November 2018. Outcome variables were hemoglobin (Hb, g/dL), neutrophils and platelet counts (mm3); plasma copper levels during the follow-up were considered as the main explanatory variable. Patient’s micronutrient status was routinely monitored at 3-month intervals or once a month when deficiency was detected. All patients were receiving a fixed dose of vitamins, and multi-trace element solution including copper (at a standard dose of 20mcg/kg). Complete blood counts were performed bi-weekly. Generalized estimating equations models were adjusted for vitamin B12, iron and folate plasma levels.

Results: Thirteen patients aged 34.2 months (IQR: 25.3; 41.1) were included; median time on PN was months 26.4 (15.2 to 32.9). An average of 7 (range 2 to 15) copper measurements/patient were performed; 53.8% of patients had at least 1 copper measurement below normal. Eight patients who had cholestasis had trace elements of PN discontinued. All but one patient had anemia; neutropenia was seen in 11 patients (among them 8 had <1000 neutrophils/mm3), and 8 patients had thrombocytopenia.

Copper deficiency (plasma level <72µg/dL) was associated with lower Hb and lower neutrophils and platelet counts. The decrease of 10 µg/dL in plasma copper resulted in decreases in Hb level (β coeff. -0.08 (95% CI: -0.02; -0.14, p=0.009), in neutrophil (β coeff. -201.6, 95% CI: -134.8; -268, p<0.001) and in platelet counts (β coeff. -6278, 95% CI: -2026; -10529, p=0.004). The figure shows predictions and marginal effects of copper serum levels on neutrophils count.

Conclusion: Copper deficiency is associated with lower blood cell counts and higher risk of anemia, neutropenia and thrombocytopenia. This effect was more pronounced for neutropenia. Copper status should be routinely monitored in children with IF receiving long term PN.
P3.24 - First clinical multi-center experience of IGL-1 for intestinal graft preservation
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Introduction: The gold standard for intestinal allograft preservation is University of Wisconsin solution (UW). In animal models, Institute Georges Lopez solution (IGL-1) improved intestinal graft viability and epithelial repair compared to UW. It is an extra-cellular preservation solution with high sodium and low potassium content in contrast to UW and presence of polyethylene glycol, resulting in lower viscosity. The aim of this study is to evaluate IGL-1 for the first time, as a preservation solution for clinical intestinal transplantation (ITx).

Methods: We performed a retrospective analysis (January 2014 to April 2018) of all ITx where the graft was preserved using IGL-1 in 4 European centers.

Results: Thirteen ITx were performed in 13 patients (1 child / 12 adults, 7 females / 6 males) for short bowel syndrome (n=7), motility disorder (n=4) and diffuse portomesenteric thrombosis (n=2). Seven multivisceral and 6 isolated grafts were transplanted. Vascular perfusion with 4-6 liters of IGL-1 was used without luminal preservation. Median cold ischemia time was 485 minutes (range: 192-840 minutes).

In all cases, the bowel appeared macroscopically well vascularized after reperfusion with minimal signs of reperfusion edema. Histology after reperfusion was available in 3/13 cases, with a maximal Park/Chiu score of 2.

One-year graft survival was 76%. Three patients required a transplantectomy (1 for CMV reactivation, 2 for refractory cellular rejection). Two patients died after transplantectomy: 1 from intestinal failure associated liver disease and 1 from bacterial sepsis, resulting in a 1-year patient survival of 83%. Ten patients are alive with a functioning graft and one requires parenteral nutrition following transplantectomy.

Conclusion: This multicenter experience suggests that IGL-1 can safely be used for preservation of intestinal grafts with good short-term results, comparable to the results from the International Intestinal Transplant Registry. Further histological data is being collected from all centers to evaluate preservation capacity of IGL-1.

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P3.25 - Intestinal Cholesterol Absorption Is Preserved After Isolated Intestinal Transplantation Despite Increased Cholesterol Synthesis
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Introduction: We aimed to investigate cholesterol metabolism in relation to nutritional status and serum lipids after isolated intestinal transplantation (ITx).

Methods: Seven patients who had undergone isolated ITx including ascending colon at median age of 17.3 (range, 5.9–52) years due to intestinal failure caused by chronic intestinal pseudo obstruction (n=5) and total intestinal aganglionosis (n=2) were included median 3.5 (IQR 2.9–7.4) years after ITx. Blood samples were analyzed for fat-soluble vitamins (A, D, E, K), prealbumin, and cholesterol (total, LDL, HDL) and triglyceride levels. In six patients and 1 healthy age- and gender-matched controls, cholesterol metabolism was assessed with serum non-cholesterol sterols. The ratios to cholesterol (Σ2 x µmol/mmol of cholesterol) of the serum cholesterol precursors (cholesteno3), desmosterol and lathosterol reflect cholesterol synthesis, while those of plant sterols (campesterol, sitosterol, and avenasterol) and cholestanol reflect cholesterol absorption efficiency. Lathosterol/cholesterol ratio reflects the balance between cholesterol absorption and synthesis and campesterol/cholesterol ratio reflects the plant sterol intake.

Results: Patients were on tacrolimus-based maintenance immunosuppression without current signs of acute or chronic transplant rejection. All patients had weaned off PN median 39 days (IQR 23–25) after ITx and had a median body mass-index of 16.2 (IQR, 15.6–17.3 kg/m2). The levels of fat-soluble vitamins and prealbumin were mostly in normal range while plasma HDL cholesterol was low and triglyceride levels were increased in most patients (Table 1). Cholesterol precursor levels were significantly increased while plant sterols and cholestanol were not significantly different between patients and controls (Table 2). Increased lathosterol/cholesterol ratio suggest that cholesterol synthesis markedly predominated absorption in patients’ whole body cholesterol homeostasis while the campesterol/cholesterol ratio suggests that the plant sterol intake was decreased in patients compared to controls (Table 2).

Conclusion: Following isolated ITx, intestinal cholesterol absorption efficiency is preserved, while cholesterol synthesis increases likely to compensate malabsorption of bile acids. Satisfactory nutritional status, normal fat-soluble vitamin levels and preserved cholesterol absorption suggest that the compensatory increase was sufficient to maintain adequate lipid absorption.
P3.26 - Complications of the aortic conduit after intestinal transplantation with liver containing grafts - A single center experience

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Introduction
Complications originating from the aortic conduit after intestinal transplantation (ITx) are currently not well described in the literature. The aim of this study was therefore to report a single center experience with diagnosis and management of complications affecting the donor aortic conduit after ITx with liver containing grafts.

Methods
From 1998 to 2018, 35 ITx were performed in Gothenburg, Sweden. 29 of 35 grafts were liver containing intestinal allografts supplied by an aortic conduit (figure 1). The recipients were 24 adults (median age 38 years, range 16 – 66 years) and 5 children (median age 8 years, range 3 – 10 years). The aortic conduit was anastomosed to either the aorta (n=20) or the iliac artery (n=9). The most common underlying conditions were intestinal failure (n=18) and non-resectable neuroendocrine pancreatic malignancies with liver metastases (n=6). Immunosuppression consisted primarily of ATG induction, tacrolimus and steroid bolus and tapering. Median donor age was 23 years (range 2-56 years) and median cold ischemic time was 443 min (range 90 - 873 min).

Results
Four patients (4/29, 14%) presented with some type of complication originating from the aortic conduit requiring intervention (figure 2). The complications were: stenosis at the origin of the superior mesenteric artery (SMA) branch (n=1), acute bleeding caused by bacterial and fungal arteritis of the conduit (n=1), stenosis of the conduit caused by kinking of the conduit close to the aortic anastomosis (n=2), inflammatory aneurysm of the donor conduit, affecting the origins of the coeliac trunk and the SMA (n=1). The complications were diagnosed from months up to several years after ITx.

Discussion
In our experience, complications affecting the donor aortic conduit after ITx may present in the early or late post-transplantation period. The insidious clinical presentation and the relatively high prevalence (14%) may warrant specific post-transplantation surveillance of the donor aortic conduit. A timely multidisciplinary therapeutic approach involving interventional radiology was crucial.

Figure 1. Donor aortic conduit anastomosed to the recipient aorta

Figure 2. Complications affecting the donor conduit of 29 ITx with liver containing grafts
P3.27 - Gastric Acid Suppression May Lead to an Increased Risk of Vancomycin-Resistant Enterococcus Colonization

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Introduction: Gastric acid suppression therapy has been reported to change the gastrointestinal microbiome, resulting in increased gastrointestinal infections. This may be of increased importance in intestinal transplantation subjects as immunosuppressive agents are associated with peptic ulcers, leading to increased gastric acid suppression therapy. Also, immunosuppressive therapy leads to a higher risk of vancomycin-resistant enterococcus (VRE) colonization. However, the effect of gastric acid suppression therapy on vancomycin-resistant enterococcus (VRE) colonization has not been investigated.

Methods: Subjects who had surveillance rectal swabs for VRE at a university hospital were investigated. Surveillance was performed on subjects with prior hospitalization within 3 months, history of VRE colonization/infection, or positive VRE findings from an adjacent patient. Gastric acid suppression therapy was defined as use of proton pump inhibitor or histamine-receptor-2 antagonists.

Results: Of 886 subjects who underwent VRE screening, 452 were positive. In univariable analysis, gastric acid suppression therapy, antibiotics use, male gender and prior hospitalization were significant risk factors for VRE colonization. Multivariable analysis showed that gastric acid suppression therapy (OR 2.873, 95% CI 1.473-5.605, P=0.002) and antibiotics use (OR 3.896, 95% CI 2.019-7.520, P<0.001) significantly increased VRE risk. Of antibiotics, carbapenems (OR 3.836, 95% CI 1.603-9.182, P=0.003) glycopeptides (OR 2.784, 95% CI 1.155-6.712, P=0.023), and cephalosporins (OR 2.210, 95% CI 1.213-4.025, P=0.010) were most significantly associated with VRE colonization. The unfavorable effect of gastric acid suppression therapy on VRE colonization was consistent, regardless of antibiotics type.

Conclusion: Gastric acid suppression therapy significantly increased VRE colonization risk. Gastric acid suppression should be administered according to strict guidelines. Physicians should be made aware of the increased VRE risk, and should consider VRE surveillance in those under long-term gastric suppression therapy.

P3.28 - The effect of a novel immunosuppressive drug, PQA-18, in rat small intestinal transplantation

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Introduction: PQA-18 (Prenylated quinolinecarboxylic acid compound 18) has been reported as a novel immunosuppressant that attenuates various cytokine productions, such as IL-2 & TNF-α, and differentiation of macrophages by inhibiting PAK2 inhibitor. In this study, we investigated this drug function mainly on macrophages using the rat small intestinal transplant model.

Methods: Male Dark Agouti (DA) (RT-1a) and Lewis (RT-1l) rats, 7 to 9 weeks old, were used as donor and recipient, respectively. An approximately 15 cm ileal grafts from the donor were heterotopically transplanted to the recipient rats. A Thirty-Vella loop was placed in the right abdominal flank. The recipient rat was treated with PQA-18 (4 mg/kg/day) by intraperitoneal injection (ip) from postoperative day 1 (POD1) to two weeks. The control group, without PQA-18, was treated with just the same amount of Dimethyl sulfoxide (DMSO), the solvent for PQA-18. We firstly compared with graft survival of both groups. The rejection was manifested by progressive stoma coloring from ischemia and necrosis and the development of an abdominal mass, based on our previous study. Next, total cells from intestinal mesenteric lymph nodes (MLN) and graft Payer’s patch (PP) were collected on POD6 and the number of macrophages was investigated using FACS cell analyzer.

Results: The graft survival was significantly prolonged by PQA-18 injection. While the survival time was 7.0 ± 0.77 days in the control group (n=9), the PQA group showed 10.7 ± 1.26 days’ survival (n=10) (P<0.001).

The number of macrophages was also significantly reduced to 9.62 ± 1.22 % in MLN of the PQA group, while the number was 22.05 ± 2.00 % in MLN of the control group (P=0.004) in the control group. In addition, the number of infiltrated macrophages in PP was 6.55 ± 1.22 % in the control group, whereas the PQA-18 group indicated 6.55 ± 1.22 % (P=0.007). The infiltration of macrophage was significantly suppressed in the PQA group.

Conclusion: PQA-18 significantly provided the prolongation of the graft survival in the rat small intestine transplantation model, together with the inhibited number of macrophages in the graft MLN & PP. It was suggested that PQA-18 has the suppressive effect not only on the differentiation but infiltration of macrophages. Further studies of the effect of this drug on macrophages are undergoing.
Intestinal transplantation (IT) remains a challenging procedure. The long term outcome, especially if the graft has been removed, is not well known. We report on clinical and pathological features with and without graft at ≥10 years or more after IT to discuss possible improvements.

Methods: Of 71 children transplanted between 1989 and 2007, 38 survived more than ≥10 years after IT: 26 with a functional graft, 9 on home parental nutrition (PN), 3 lost to follow-up. The median follow-up was 14.6 years, in our pediatric center, then in Beaujon Hospital for University Paris VII, Paris, France.

Results: Indications of IT were: short bowel syndrome (34%), motility disorder (32%), congenital enteropathy (34%), age at IT was 4.1 years and at last follow-up ≥20.1 years. Patient and graft survivals were 53% and 34%. The graft was removed in 12 (34%) for rejection; 5/2 patients were re-transplanted. 3 are well after 9.8-17.4 years follow-up. Five (34%) patients died; 2 transplanted (lymphoma, sepsis), 2 after re-transplantation, one on home PN (sepsis).

The 26 patients with graft were free of PN, 65% had liver-small bowel transplantation. Overall and fat absorptions were 93% and 89%. The height percentile (-0.5 SD) was higher than at IT. The measured renal clearance was 88 ml/min/1.73 m². Five patients (31%) had a lymphoproliferative syndrome, 42% a graft rejection more than one year post-IT, 42% had DSA (donor specific antibodies) without rejection. On biopsies they had a mild to moderate increase of eosinophils. Of the 3 with severe eosinophilia in other areas of the GI tract, 2 were at the time of late onset severe acute rejection and the other is the index case as described above.

Conclusion: IT remains difficult but leads to digestive autonomy and catch-up growth without major complications of immunosuppression. The long-term pathology is reassuring despite the presence of DSA. The prognosis in case of graft removal is acceptable. Re-transplantation carries a significant mortality. Multidisciplinary care with early psychosocial follow-up is essential.

Clinical implications of mucosal eosinophilia in the long term intestinal transplant patient

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Introduction: A child, 6 years following isolated small bowel transplant (SBTx) for intestinal failure secondary to gastroschisis with impaired venous access, developed failling growth associated with increased stool output. The gastrointestinal biopsies (GI) showed severe eosinophilia throughout the GI tract, affecting native as well as graft bowel. It is unclear whether mucosal eosinophilia is causative of gastrointestinal symptoms, a novel pattern of rejection, a feature of infection, or inconsequential.

Aim: Review the histology of patients who are greater than 5 years post SBTx to identify mucosal eosinophilia and correlate this with patient clinical course.

Population: 32 patients (13 boys) underwent annual GI biopsies. Indications for SBTx were short gut =20; motility disorder =12; microvillus inclusion disease =2. Ten had isolated bowel or modified multivisceral, whilst 22 had liver containing grafts.

Results: In total 327 biopsy reports were analysed ranging from 5 to 17 years following transplant. 153/327 (47%) reported to have normal amounts of mucosal eosinophils; 124/327 (37%) moderate; 53/327 (16%) severe eosinophilia.

12 patients had no increase in eosinophils at any time; 12 had a moderate increase; 12 had a severe increase, of which 7 had severe eosinophilia confined to the oesophagus only which was diagnostic of eosinophilic oesophagitis (EO). Of the 3 with severe eosinophilia in other areas of the GI tract, 2 were at the time of late onset severe acute rejection and the other is the index case as described above.

As a proxy for graft function, 4/12 (33%) required enteral supplements in those with normal mucosal eosinophils; 60% with moderate; 60% with severe, of which 3 require PN.

Discussion: Eosinophils are increased in over 50% of biopsies. EO was an isolated finding in 22% of patients indicating a need for long term upper GI endoscopic surveillance. In those confined to EO, there is little impact on graft function. A moderate increase in eosinopils is associated with increased need for enteral supplementation. Eosinophilia is an important finding in late onset acute rejection.

Conclusion: Increased mucosal eosinophilia may be associated with reduced graft function and the inflammatory reaction of late onset acute cellular rejection. Whether this is causative or secondary is unclear.

Longitudinal studies from the time of SBTx will ascertain whether early eosinophilic infiltrates have a bearing on subsequent transplant pathology.
**P3.31** Curcumin shifts the differentiation potential of intestinal stem cells toward the enterocyte lineage

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**Introduction:** Intestinal stem cell (ISC) is the fundamental unit for the regeneration of intestinal epithelium. Curcumin is a natural polyphenol extracted from the plant Curcuma longa. Curcumin has been found to have anti-inflammatory, anti-oxidation, as well as anti-cancer effects. Recent years, curcumin also was reported to regulate the activity of embryonic and mesenchymal stem cells. In this research we aimed to investigate the effects of curcumin on ISCs.

**Methods:** Purified curcumin was purchased from Sigma-Aldrich. Mice ISCs were isolated from B6 mice and subjected to organoid culture under the treatment of different concentrations for 10 days. At the end of treatment, the images of organoids under each treatment group were photographed by microscopic cameras, and the organoid morphology, number, and area were further analyzed by ImageJ software. The ISC markers of proliferation and differentiation were analyzed by real-time PCR, and immuno-fluorescence staining.

**Results:** The average number of the organoids was 33, 47, 43, and 20 per well after the treatment of 0, 1, 5, and 25 μM of curcumin, respectively. The average size of organoid was significantly larger in the curcumin-treated groups (Figure 1). The ISC proliferation markers Lgr5 showed modest decrease after curcumin treatment: 0.66- (1 μM), 0.74- (5 μM), and 0.56- (25 μM) fold compared with the control. Of the four differentiation markers, curcumin treatment resulted in significant increase of villin with 1.87- (1 μM), 2.19- (5 μM), and 2.21- (25 μM) fold increase. Confocal imaging of anti-villin-stained organoids also demonstrated more abundant villin expression in the organoids under the treatment of curcumin (Figure 2).

**Conclusion:** ISCs treated by curcumin showed significant upregulation of villin, which represents the differentiation potential toward “enterocyte”, which makes up the absorptive function of intestinal epithelium. The molecular mechanism of this effect and its clinical application is under investigation in the in vivo mice model.

![Figure 1. Effect of curcumin on the formation of intestinal organoids.](image1)

![Figure 2. Effect of curcumin on the expression of ISC markers.](image2)

**P3.32** Safety and efficacy of standardized versus individualized parenteral nutrition mixtures in a pediatric home parenteral nutrition population

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**Introduction:** Current guidelines on pediatric Parenteral Nutrition (PN) conclude that standardized PN mixtures are not suitable for children with chronic Intestinal Failure (IF) on home PN. However, studies evaluating efficacy or safety of standardized PN are lacking. Possible advantages of standardized PN could be a reduction in costs and a longer shelf life of PN. Therefore, this study was designed to assess the effects on growth and safety of standardized PN compared to individualized PN.

**Methods:** Retrospective cohort study in Dutch children on home PN between J une 2017 and J uly 2018, in which Individualized PN was compared to standardized PN. Growth was assessed by calculating the difference of Weight-for-age (WFA), Height-for-age (HFA) and weight for height (WFH) SD scores between date of inclusion and 6, 24, and 24 months prior to inclusion. Primary outcome was growth over 2 years, secondary outcomes were electrolyte disturbances and biochemical abnormalities such as liver function. Non parametric tests were used to explore differences between groups.

**Results:** 50 patients (50% female, median age 6.5 years) were included, of whom 32% received standardized PN mixtures. Age (12 vs 5 years), gestational age (GA) (39 vs 36.2 weeks) and PN duration (97 vs 39 months) were significantly higher in the group receiving standardized PN (p: <.001; .027; .013 respectively). Type of underlying disease did not differ between groups. For children receiving standardized PN mixtures, median weight gain in 2 years was significantly higher compared to the individualized PN mixtures group of children where a mean SD score decrease was seen (μ: .38 SD score vs -0.55 SD score, p: .003). No significant differences were demonstrated in HFA SD score change, WFH SD score change (Table 1), or electrolyte disturbances (Table 2) between groups. Median total bilirubin was 6.0 μmol/l (5.0 – 13.0) in the standardized PN group and 5.0 μmol/l (4.0 – 9.0) in the individualized PN group (p: 4.73).

| Table 1. Median Change in SD Score over 6, 12, and 24 Months |
|-----------------|-----------------|-----------------|-----------------|
| **Weight**       | **Height**      | **Weight**     |
| 6 months        | 12 months       | 24 months      |
| 1.07 (-0.69 - 1.2) | 1.29 (-1.9 - 0.9) | 2.14 (-0.79 - 2.0) | 0.24         |
| 1.55 (-0.19 - 3.8) | 2.32 (-2.1 - 0.9) | 2.51 (-0.9 - 2.0) | 0.01         |
| 1.90 (-1.93 - 5.7) | 2.90 (-2.9 - 0.9) | 2.78 (-0.9 - 2.0) | 0.89         |

| Table 2. Number of Electrolyte Disturbances |
|-----------------|-----------------|-----------------|
| **Sodium**       | **Potassium**   | **Calcium**     |
| 35 (20-50)       | 35 (20-50)      | 2.00           |
| 50 (50-50)       | 2.14 (1.8 - 2.5) | 35 (20-50)    |
| 50 (50-50)       | 35 (20-50)      | 35 (20-50)    |

All data are presented as median (interquartile range); Mann-Whitney U test was used to determine p-value.
Conclusion: In children receiving standardized PN mixtures, change in WFA SD score was significantly higher compared to children receiving individualized PN. Standardized PN mixtures are at least non-inferior to individualized PN mixtures in terms of electrolyte disturbances and biochemical abnormalities in a home PN cohort. Therefore, standardized PN mixtures can safely be administered to patients with chronic IF if the composition of this mixture meets the nutritional need of the patient.

P3.33 - Clinical outcomes of children weaned from parenteral nutrition after neonatal gastrointestinal surgery
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Introduction: The majority of children with intestinal failure due to short bowel syndrome (SBS) [dependence on parenteral nutrition (PN) for at least 90 days] are nowadays capable to be weaned from PN. Scarcie evidences are available regarding the follow-up setting after intestinal adaptation. The aim of the study was to investigate the occurrence of morbidities after intestinal adaptation in children who underwent neonatal gastrointestinal (GI) surgery.

Methods: In our Home PN Centre we develop a multidisciplinary and long term follow-up to obtain a primary and secondary prevention of the unsuspected and life threatening complications after intestinal resection. Specifically, patients after intestinal adaptation enter in a follow-up program that includes dedicated staff in gastroenterology, nutrition, paediatrics, surgery, nephrology and radiology. Each patient underwent to a serial exams/clinical evaluations including stools assessment for steatorrhea and haematochezia, endoscopy, nutritional intake, urinary analysis, ultrasound and X-ray follow through of GI tract and Breath H2 test.

Results: We included in the analysis 47 children (27 males) with intestinal failure after neonatal GI surgery. The mean gestational age at birth was 32.8±5.1 weeks and the age at the last follow-up visit was: 7.8±8 years (0.6-24 years). Among all children 33 were weaned from PN dependence. Causes leading to SBS were: necrotizing enterocolitis (9/33), multiple bowel perforations (6/33), volvulus (5/33), intestinal aganglionosis (4/33), gastrochisis (2/33), intestinal atresia (7/33). The duration of PN of these children was 11±8 years (3 months-9 years). The length of small bowel residual was 51±39 cm (14-160 cm), 13/33 children had totally preserved the colon, whereas 16/33 had a partial residual colon. Among 33 infants weaned from PN, 42.4% developed morbidities during the follow-up period. Specifically, 3 children were identified as having anastomotic ulcers with clinical anemia, 3 children developed gallstones, 3 children developed kidney stones, 2 children had symptomatic D-lactic acidosis and 1 child developed acute pancreatitis and cholecystitis and 2 child developed venous trombosis. The occurrence of morbidities was found after 4.5±3 years (range 0.1-12.7 years) the weaning from PN.

Conclusions: This study suggest that the occurrence of morbidities after PN weaning is not a rare event. Therefore a multidisciplinary and long term follow-up is mandatory.
P3.34 - Inclusion of the pancreas as a part of the multivisceral allografts: A single center experience.

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Introduction: Intestinal transplantation provides a therapeutic option for patients with irreversible intestinal failure and complications of total parenteral nutrition. For technical reasons, the pancreas has been transplanted en bloc with the multivisceral allografts. Here, we report the advantages and complications related to the pancreas inclusion in the multivisceral (MV) and modified multivisceral (MMV) allografts.

Methods: Retrospective analysis of 64 recipients of MV and MMV allografts transplanted between 2013-2018 was performed at the Miami Transplant Institute. Pancreas was included in all of the 64 recipients. We recorded the pre-transplant history of diabetes, its type and treatment, and the development of new-onset diabetes post-transplant. Pre- and post-transplant acute pancreatitis was documented and recorded. Graft and patient survival due to pancreatitis was also recorded.

Results: The five recipients with pre-transplant history of diabetes (type 2 in four recipients, and type 1 in one recipient) demonstrated clinical cure from diabetes with no need for insulin or oral hypoglycemic drugs at 3, 6 and 12 post-transplant months. The incidence rate of post-transplant acute necrotizing pancreatitis in recipients with no prior history of acute pancreatitis was 3% (2/60) while none of the recipients with pre-transplant history of acute pancreatitis developed it post-transplant (0/4). Acute pancreatitis progressed to fatal necrotizing pancreatitis in 50% (1/2) of the recipients who developed post-transplant acute pancreatitis. No technical problems related to the pancreas were encountered.

Conclusion: Pancreas inclusion in MV and MMV allografts brought the cure for recipients with pre-transplant history of diabetes and those with a past history of acute pancreatitis, as well.

P3.35 - Quantitative dynamics of parenteral support in intestinal failure - importance of sodium for the recovery of body mass index

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Introduction: Intestinal failure (IF) is the inability to maintain protein-, energy- and/or fluid- and electrolyte balance because of reduced absorption with the necessity of parenteral support (PS). The ability to adapt is mainly determined by the functional anatomy (type-I: jejuno-stoma, type-II: jejuno-colic anastomosis, type-III: jejuno-ileo-colonic anastomosis). We determined the dynamical changes of PS over time and as a result of reconstructive surgery or medication with Teduglutide.

Methods: Monocentric analysis of 56 patients with IF. Analysis of the relation of the PS-composition, anatomic type, surgery and Teduglutide to BMI over time (initially, time of max. support, last contact).

Results: 50/56 patients were followed for 869 ± 750 days (range 41-4011) with 8 ± 6 observation time points (mean interval 92 days) per patient. At the initial contact 27 patients had type-I-, 16 type-II-, 2 type-III-anatomy and 5 functional IF. During the observation period 4 type-I-patients were converted to type-II-anatomy and 7 to type-III-anatomy.

At the initial contact type-I-patients received 2926 ml volume, 244 mmol Na and 24 kcal/ kg per day and type-II-patients received 225 ml volume, 17 mmol Na and 19 kcal/ kg per day. At this time they were on parenteral nutrition. For technical reasons, the pancreas has been transplanted en bloc with the multivisceral allografts. Here, we report the advantages and complications related to the pancreas inclusion in the multivisceral (MV) and modified multivisceral (MMV) allografts.

Methods: Retrospective analysis of 64 recipients of MV and MMV allografts transplanted between 2013-2018 was performed at the Miami Transplant Institute. Pancreas was included in all of the 64 recipients. We recorded the pre-transplant history of diabetes, its type and treatment, and the development of new-onset diabetes post-transplant. Pre- and post-transplant acute pancreatitis was documented and recorded. Graft and patient survival due to pancreatitis was also recorded.

Results: The five recipients with pre-transplant history of diabetes (type 2 in four recipients, and type 1 in one recipient) demonstrated clinical cure from diabetes with no need for insulin or oral hypoglycemic drugs at 3, 6 and 12 post-transplant months. The incidence rate of post-transplant acute necrotizing pancreatitis in recipients with no prior history of acute pancreatitis was 3% (2/60) while none of the recipients with pre-transplant history of acute pancreatitis developed it post-transplant (0/4). Acute pancreatitis progressed to fatal necrotizing pancreatitis in 50% (1/2) of the recipients who developed post-transplant acute pancreatitis. No technical problems related to the pancreas were encountered.

Conclusion: Pancreas inclusion in MV and MMV allografts brought the cure for recipients with pre-transplant history of diabetes and those with a past history of acute pancreatitis, as well.
P3.36 - Burden of care for children after establishment of enteral autonomy following intestinal failure: a 12 month follow up study

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2 Starship Child Health, Auckland, New Zealand

Introduction: The importance of a multidisciplinary team for intestinal rehabilitation in children with intestinal failure (IF) is accepted internationally and is a standard of care for patients managed by the New Zealand National Intestinal Failure Service (NZ NIFS). The aim of this study was to understand the ongoing burden of care for children with IF in NZ who achieved enteral autonomy as their primary outcome.

Methods: The NZ NIFS patient registry collected data for patients with IF as defined by the requirement for intravenous nutrition (IVN) for ≥ 21 days for paediatric patients up to 31 years of age and ≥ 30 days for preterm neonates (< 34 weeks gestation). Data were collected from October 2017 to October 2018.

Primary clinicians of patients with IF reported data in conjunction with NZ NIFS and information was obtained from patient records. Data collected included ethnicity, social deprivation index, cause of IF and clinical outcome. For patients that achieved enteral autonomy as their primary outcome and had at least 12 months follow up, further information was collected regarding feeding outcomes and multidisciplinary professionals involved in their care at 12 months after ceasing IVN. The number and indications for readmissions for this patient group over this 12 month period were also collected.

Results: Of 208 children with IF over a 3 year period, 128 (62%) patients achieved enteral autonomy and had 12 months follow up after ceasing IVN. Of this group, most common causes of IF were mechanical obstruction and short bowel syndrome.

At 12 months follow up, 117 (92%) children were having an appropriate oral diet, including 27 (21%) also needing supplementary drinks, and/or enteral feeding in combination with their oral diet. One hundred and seven (84%) children required review by a paediatrician and 56 (44%) and speech and language therapists 42 (33%) were continued for 61 (48%) of children and allied health services including occupational therapy 31 (25%) and physiotherapy 25 (20%) were also required.

Twenty nine (23%) of the children were readmitted to hospital for reasons related to IF during this period.

Conclusion: The burden of care for children with IF does not end with ceasing IVN. Of this group, most common causes of IF were mechanical obstruction and short bowel syndrome.

Table 1: Multidisciplinary professional input for NZ children with IF 12 months after they achieved enteral autonomy

<table>
<thead>
<tr>
<th>Multidisciplinary professional</th>
<th>Number of patients (n = 128)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Paediatric</td>
<td>107</td>
</tr>
<tr>
<td>Neonatologist</td>
<td>47</td>
</tr>
<tr>
<td>Surgeon</td>
<td>56</td>
</tr>
<tr>
<td>Gastroenterologist</td>
<td>19</td>
</tr>
<tr>
<td>Community nurse</td>
<td>91</td>
</tr>
<tr>
<td>Dietitian</td>
<td>56</td>
</tr>
<tr>
<td>Occupational therapist</td>
<td>31</td>
</tr>
<tr>
<td>Pharmacist</td>
<td>11</td>
</tr>
<tr>
<td>Physiotherapist</td>
<td>35</td>
</tr>
<tr>
<td>Psychologist</td>
<td>22</td>
</tr>
<tr>
<td>Speech and language therapist</td>
<td>42</td>
</tr>
<tr>
<td>Social worker</td>
<td>16</td>
</tr>
<tr>
<td>Unknown input</td>
<td>2</td>
</tr>
</tbody>
</table>

P3.37 - Luminal preservation of the intestinal graft: an update on the international multicentre study LUMINALR

Guido Trentadue 1, Anne Marye de Jong 1, J asper van Praagh 2, Emilio Canovai 3, Mathias Clarysse 1, J acques Pirene 1, Henri G. D. Leuvenink 4, J an Willem Haveman 2, Klaas Nico Faber 3, Gerard Dijkstra 1, et al.

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2 Surgery, University Medical Center Groningen, University of Groningen, Groningen, The Netherlands
3 Abdominal Transplant Department, University Hospitals Leuven, Belgium
4 Surgical Research Laboratory, Department of Surgery, University Medical Center Groningen, University of Groningen, Groningen, The Netherlands
5 Laboratory Medicine, University Medical Center Groningen, University of Groningen, Groningen, The Netherlands

Introduction: Graft survival rates in intestinal transplantation (ITx) are the lowest among solid organ transplantation. Unique for ITx is the presence of metabolically-active luminal content consisting of microbes, dietary and waste products. Ischemic damage during cold storage decreases mucosal integrity, which contributes to graft rejection and infections. Cold storage of the bowel is limited to 10 hours, after which it is deemed unsuitable for transplantation. Luminal preservation (LP) with polyethylene glycol (PEG) has shown promising effects in improving graft-viability in experimental animal models. The ITx centres in The Netherlands and Belgium joined forces with the aim to analyse the effect of LP on graft viability.

Methods: A prospective study was set up with 4 study groups:

1. Control: University of Wisconsin solution (UW) for vascular perfusion (VP) only
2. VP + LP: UW + PEG
3. VP + LP: IGL-1 + PEG
4. VP + LP: IGL-1 + PEG

Inclusion criteria follows recommendations from the Organ Procurement and Transplantation Network for ITx, with extended age to 70 years (due to donor shortage) and no anthropomorphic features taken into account. Donors underwent standard dissection for organ procurement. If LP was included, the intestine was perfused with PEG (6.4% w/v) through the nasogastric tube. Samples were taken at 0, 7 and 21 hours after start of preservation. Analyses include histology (Figures 1 a, b, c). Preservation scores reflect these findings (Figure 2).
Conclusion: These preliminary data suggest that LP with PEG reduces preservation injury in intestinal grafts. LP might thus improve graft viability and increase its preservation time-window. Different LP solutions are being tested and further analyses are underway to address barrier function and the cause of the subepithelial oedema.

P3.38 - Two decades of intestinal transplantation (ITx) in Leuven.

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3 Experimental Transplantation Laboratory, Department of Microbiology, Immunology and Transplantation, KU Leuven, Belgium.

Introduction: ITx was initiated at the University Hospitals Leuven in 2000 after preclinical animal studies. We report our long-term experience.

Methods: In 2000-2019 19 ITx were performed in 19 patients. Data were extracted from a prospectively maintained database. Kaplan-Meier was used for survival estimation.

Results: Median follow-up is 6yr 11mth (1yr-16yr). Median age was 40yr 5mth (2yr 9mth-56yr 8mth); male/ female ratio was 9/10; peds/ adult ratio was 4/15. Indications were complicated intestinal failure/disease not treatable by any other means and due to: ischemia (4), volvulus (4), splanchnic thrombosis (4), Crohn (2), chronic intestinal pseudo-obstruction (3), microvillus inclusion disease (1), Churg-Strauss (1). 9 were isolated ITx, 6 combined liver-ITx, and 4 MVTx. 4 received a kidney and 2 a colonic segment. 1 was a live-donor Tx. MVTx was done after visceral artery embolisation, dramatically reducing blood loss. The Leuven immunomodulatory protocol {Donor-specific blood transfusion, low Immunosuppression (IS), reduced periTx inflammation} promoting T-regs was used in 17 recipients. In total 2 severe grade 3 early Acute Rejections (AR) in 2 patients (10.5%) and 4 severe grade 3 late AR in 3 patients (15%) were seen. There was no graft loss to rejection except in the live-donor recipient in whom a transplantectomy was done 7 mths postTx. Renal failure was not seen except in 1 combined kidney Tx recipient. No patients developed PTLD. 4 died (2 aspergillus, 1 NSAID-induced graft ischemia, 1 sepsis). Of the 17 survivors, 13 are nutritionally independent. In 2, a transplantectomy was done (1 graft ischemia > protocol biopsy; 1 multiresistant CMV enteritis). The latter is listed for reTx. So far no proven chronic rejection / late immunological graft loss was seen. 3 3yr patient & graft survival are 90%/ 84% & 84%/ 77%.

Costs of ITx (albeit > other organ Tx) become lower than TPN > 2yrs. Launch of a comprehensive/multidisciplinary intestinal failure center has increased patient referral. 3 are awaiting ITx, 1 combined liver-ITx, and 4 MVTx.

Conclusion: Long-term outcome of ITx under low IS compares favorably with global data and other organ Tx. PreTx embolisation dramatically changed the nature of MVTx. Survival equal or superior to TPN, better quality-of-life, and cost-effectiveness support application of ITx earlier in the course of intestinal failure. With growing waiting list at our center, referral of suitable intestinal donors is critical.
P3.39 - Risk of post-transplant lymphoproliferative disorder (PTLD) with Epstein-Barr virus (EBV) serostatus in donor and recipient in intestinal and multivisceral transplant

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Introduction: Post-transplant lymphoproliferative disorder (PTLD) is one of the most serious complications that can develop after a transplant and rates are reported to be highest in intestinal and multivisceral transplants. This study looks at the risk of developing PTLD with different Epstein-Barr virus (EBV) serostatus in donor and recipient.

Methods: This is a retrospective observation study of case notes. Key data collected includes EBV status in the donor and recipient prior to transplant, time to development of PTLD, histology specimen reports and mortality outcomes.

Results: The intestinal and multivisceral transplant service at Addenbrooke’s Hospital, Cambridge, UK transplanted 80 patients from 2000-2018. There were 12 cases which developed PTLD giving a rate of developing PTLD of 5% in our cohort and mortality of 17.7%. Histology was obtained in 11 cases with one case having an inaccessible lymph node but characteristic features of PTLD. The histology specimens showed 100% stained positive for EBV, whilst 66.7% were lymphomas and 33.3% were pre-lymphomas. The development of PTLD occurred within 3 months in 66% of cases, 3-6 months in 17% of cases, and 6-12 months in 17% of cases.

Information on donor and recipient EBV serostatus was available in 44 consecutive patients, which included 12 of the 12 cases of PTLD. 20 patients developed persistent EBV viraemia, and 12 of these patients developed PTLD, taking approximately 2 weeks to progress.

The risk of persistent EBV viraemia developing with different donor and recipient EBV serostatus and subsequent risk of developing PTLD is shown below in table 1.

<table>
<thead>
<tr>
<th>Donor EBV serostatus</th>
<th>Recipient EBV serostatus</th>
<th>Rate of EBV viraemia</th>
<th>Risk of EBV viraemia developing into PTLD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Positive</td>
<td>Positive</td>
<td>0/0</td>
<td>0</td>
</tr>
<tr>
<td>Negative</td>
<td>Positive</td>
<td>0/0</td>
<td>0</td>
</tr>
<tr>
<td>Positive</td>
<td>Negative</td>
<td>0/0</td>
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</tr>
<tr>
<td>Negative</td>
<td>Positive</td>
<td>0/0</td>
<td>0</td>
</tr>
<tr>
<td>Negative</td>
<td>Negative</td>
<td>0/0</td>
<td>0</td>
</tr>
</tbody>
</table>

Table 1

The risk of PTLD developing in groups with different donor and recipient EBV serostatus is shown below in table 2.

<table>
<thead>
<tr>
<th>Donor EBV serostatus</th>
<th>Recipient EBV serostatus</th>
<th>Rate of PTLD</th>
<th>OR of developing PTLD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Positive</td>
<td>Positive</td>
<td>0/0</td>
<td>0</td>
</tr>
<tr>
<td>Negative</td>
<td>Positive</td>
<td>0/0</td>
<td>0</td>
</tr>
<tr>
<td>Positive</td>
<td>Negative</td>
<td>0/0</td>
<td>0</td>
</tr>
<tr>
<td>Negative</td>
<td>Positive</td>
<td>0/0</td>
<td>0</td>
</tr>
<tr>
<td>Negative</td>
<td>Negative</td>
<td>0/0</td>
<td>0</td>
</tr>
</tbody>
</table>

Table 2

Conclusion: The key finding of this study is that when an EBV seronegative recipient receives and intestinal graft from an EBV seropositive donor, there is 67% risk of developing EBV viraemia and in this group 10% will develop PTLD. Furthermore, the two deaths directly attributable to PTLD occurred in this group, suggesting that primary infection has a more serious disease course. Taken together, this gives a strong argument for matching EBV serostatus, particularly for recipients who are EBV negative. In circumstances where this is not possible, for example a super-urgent listing due to graft failure or a highly sensitised recipient, a persistent EBV viraemia should be treated early and effectively to stop progression to PTLD.

P3.40 - Improving the care of pediatric intestinal failure in the public health care system through a public-private partnership: the Brazilian experience

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Introduction: In Brazil, the majority of the population relies on the public health care system (PHCS). Due to funding restrictions, PHCS has some limitations, as a large part of the budget must be designated to basic health needs, a priority condition in a developing country. In the last years, some initiatives were developed in order to mitigate this condition and improve public access to specialized health care, including the treatment of intestinal failure (IF).

Methods: A model of public-private partnership (PPP) was developed, involving the Federal Government (FG) and Hospital Sirio-Libanes (HSL), a private hospital considered a center of excellence. By this model, HSL is responsible for the development and management of specialized medical programs not available in the PHCS, in exchange for fiscal incentives.

Results: In 2009, a project named PROADI-SUS was initiated between the FG and HSL. This partnership was responsible for the development of a pediatric liver transplant program, which performed more than 200 pediatric liver transplants, promoted research activity and diffusion of medical education to other centers. Due to the scarce of intestinal rehabilitation centers in Brazil, and the absence of a pediatric multivisceral transplant team, by the end of 2018 the same strategy was utilized to develop a program specialized in the treatment of IF in the PHCS. The program was instituted in a pediatric public hospital (Hospital Menino Jesus), and due to the complexity of IF patients, improvements were performed in several areas of the hospital, benefiting not only the IF patients, but the general population. Medical resources not available in the public hospital are obtained in the private hospital; transplants are performed in the private hospital and as soon as the patient is stable, the post-operative is followed by the same team in the public hospital. The intestinal rehabilitation center is now receiving patients from all over Brazil, and has started to teach and support other public hospitals in order to disseminate the treatment of intestinal failure through the country.

Conclusion: The association of a private center of excellence with the public system, through a PPP, promoted a significant improvement in the treatment of IF in the Brazilian PHCS. In the next years, the dissemination of the knowledge acquired with this PPP will permit the creation of new public intestinal rehabilitation centers, benefiting more patients of the public system.
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P3.41 - Pharmacokinetics and Pharmacodynamics of Glucagon-like Peptide (GLP-2) Analogue Apraglutide (FE 203799) in Adult Healthy Volunteers: Results of a Phase I Trial

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Therachon AG, Basel, Switzerland

Introduction: To investigate the safety, tolerability, pharmacokinetics (PK) and pharmacodynamics (PD) of the novel synthetic peptide analog of glucagon-like peptide-2 (GLP-2) apraglutide (FE 203799) in male and female healthy volunteers following subcutaneous (SC) and intravenous (IV) injection.

Methods: A total of 40 subjects were randomized to receive a single ascending dose of either apraglutide or placebo (3:1 ratio) at doses of 2.8, 5.7, 11.4, 28.4, and 56 mg. The first cohort received the lowest dose level of apraglutide by SC injection. Subjects were enrolled into the next dose level cohort sequentially and received the next higher dose level after all subjects in the preceding dose level cohort had been assessed.

Results: Apraglutide was considered safe and well tolerated at all doses with no Serious Adverse Events (SAEs). No immunogenicity was observed. The dose comparisons performed indicated that peak concentrations and exposure were similar for most dose levels when both genders were pooled (Tukey-Kramer p-values >0.05). Dose proportionality analyses demonstrated Cmax was less than dose proportional with a single ascending dose of apraglutide, whereas AUC0 to ∞ showed no significant difference between dose levels. No gender effects were observed. The rate and exposure to apraglutide was proportionate over the dose range when administered via SC injection. Following SC administration, dose-adjusted PK parameters showed dose-proportional kinetics and no accumulation.

The half-life was approximately 30 hours. The single-dose administration of SC apraglutide was generally well tolerated. The AEs reported at 2.8 mg SC, 5.7 mg, and 56 mg dose levels were only mild in severity. Four (4) moderate AEs were reported for 11.4 mg and 28.4 mg dose levels and no severe AEs were reported. Increasing single doses of apraglutide did not affect heart rate or cardiac conduction and in the studied range of plasma concentration, up to approximately 1500 ng/mL, apraglutide did not have a clinically relevant effect on ECG parameters.

Conclusions: This study in healthy volunteers exposed to single ascending doses of apraglutide confirm a favorable safety profile. The long pharmacokinetic half-life of approximately 30 hours for apraglutide supports a dosing interval of one week or longer in clinical trials. Apraglutide is currently in Phase II development for patients with short bowel syndrome requiring parenteral support.

P3.42 - Acute dehydration and hypercalcemia in patients with total intestinal aganglionosis on long term parenteral nutrition

Matilde Rossi 1, Elie Abi Nader 1, Cécile Lambe 1, Marina Charib 2, Cécile Talbotec 1, Dominique Prê 3, Olivier Goulet 1
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2 Hôpital Necker Enfants Malade, Paris-Descartes University, Department of Paediatric Nephrology, Paris, France
3 Hôpital Necker Enfants Malade, Paris-Descartes University, Service des Explorations Fonctionnelles, Paris, France

Objectives and Study: Total intestinal aganglionosis (TIA) is the rarest and the most severe form of Hirschsprung’s disease. Patients are dependent on long term parenteral nutrition (LTPN) and at risk of dehydration episodes, acute renal failure and hypercalcemia. The objective of this study was to identify the risk factors for hypercalcemia in children with TIA on LTPN.

Methods: We retrospectively analysed the medical records of 16 patients with TIA divided into 2 groups. Group A who presented at least one episode of acute dehydration and hypercalcemia and group B (no history of hypercalcemia). Calcium & phosphorus status were analysed in blood and urine, PTH, 25-OHD3 plasma levels, kidney function, PN intake of calcium, phosphorus and bone mineral density (BMD) using X-ray absorptiometry. Values were reported as mean +/- SD or median [IQR].

Results: Both groups were composed of 8 children. No difference were found on steady state between the 2 groups in terms of blood calcium, phosphorus, urea, creatinine and glomerular filtration rate. Urinary calcium divided by creatininuria of group A was lower than in group B (0.64 ±0.60 vs 1.80 ±1.28; p=0.03). Group A patients received higher PN calcium intake (0.49 ±0.05 vs 0.40 ±0.07 mmol/kg/day; p=0.02). On steady state PTH in group A was lower than in group B (20.8 ±15.3 vs 32.5 ±41.5 ng/l; p=0.03) within the normal ranges (normal: 10-50 ng/l). In group A patients received higher PN calcium intake, although they were in line with ESPGHAN guidelines. On steady state the hypercalcemia group had a lower urinary calcium excretion rate than group B. Patients with TIA presents high stool output and are at risk of acute and chronic dehydration. These data underline the importance to assess carefully the calcium metabolism during the follow-up of patients on LTPN for TIA with high stoma output. To prevent hypercalcemia it’s important to correct rapidly acute dehydration episodes and on long term to provide high water-electrolytes supplementation, to limit calcium intake and to monitor plasma and urinary calcium, 25-OHD3 and PTH.
Infection is the leading cause of death in small bowel and multivisceral organ transplantation. It has been reported that the incidence of bloodstream infection (BSI) is higher in children compared to adults, reaching almost 80% in some centers. In the UK there is limited data describing the incidence, aetiology and outcome of BSI in children receiving small bowel and multivisceral organ transplant.

Methods: Retrospective review of patients undergoing small bowel and multivisceral transplants at a tertiary centre from 1993 until 2015.

Results: A total of 92 patients received isolated small bowel (n=30), liver-small bowel (n=49), liver, small bowel and pancreas (n=8), multivisceral (n=3) and modified multivisceral (n=4) transplants. Mean age at transplant was 4.3 years (range 7 months to 16 years). Bacteremia was observed in 58% (53/92) of patients (total of 103 episodes); eight patients had ≥3 episodes of BSI. Gram positive organisms were the most frequently isolated (73/103 episodes), with coagulase negative Staphylococci accounting for 63% of these cultures. Gram negatives and Candida were identified in 12% (12/103) and 5% (5/103) of episodes respectively. All Gram negative BSI occurred >150 days post-transplant, while 60% of infection caused by Staphylococcus aureus and Enterococcus was observed <150 days post-transplant. Overall survival was higher in those without culture confirmed infections later than Gram positive infections. Timing of BSI differed by aetiology, with Gram negative infections observed earlier than Gram positive infections. Overall survival was lower in those with BSI, compared to those without BSI.

Discussion: In this retrospective case series 58% of patients suffered from at least one episode of bacteremia, an incidence lower than that previously reported. Timing of BSI differed by aetiology, with Gram negative infections observed later than Gram positive infections. Overall survival was higher in those with BSI, compared to those without BSI.

Conclusion: Blood stream infections are common following intestinal transplantation and vigilance and prompt treatment may prevent mortality in children following intestinal transplantation.
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P3.45 - Intestinal failure-associated liver disease (IFALD) in the SMOF era: what has changed?

Cecile Lambe, Maud Prevot, Cecile Talbotec, Olivier Goulet, Florence Lacaille

Department of pediatric gastro-enterology, hepatology and nutrition Hôpital Necker-Enfants Malades

Introduction: IFALD is the most severe complication of intestinal failure (IF). Since 2009, we have used a composite lipid emulsion containing fish oil, medium chain triglycerides, olive oil and soybean oil (SMOF lipid) in our cohort of IF children. The aim of this study was to assess the hepatic profile of children with IF receiving this lipid emulsion.

Methods: This study was a retrospective review of the children followed in our center for intestinal failure from 2009 to 2019 who received at least two years of parenteral nutrition (PN) and SMOF lipid and who were still on home PN on January 2019. The data was collected from charts review, using the latest clinical, biological and radiological data available.

Results: One hundred and nine children were included. Of these, 60 (55%) were born after 2009 and received SMOF lipid exclusively. Most children (66%) were followed for short bowel syndrome (whom 17 had extensive Hirschsprung’s disease), 25 (23%) for congenital enteropathy, and 33 (32%) for chronic intestinal pseudo-obstruction. They received PN a mean of 5.4 days/week with SMOF lipid 15 g/kg/day.

Sixty-six percent of patients had abnormal liver tests, 53% elevated ALT, 48% elevated GGT, 22% elevated bilirubin with 9 of them having elevated conjugated bilirubin; 29 patients had low PT and 20 had low platelets rate.

Ultrasound showed splenomegaly in 35 patients (32%). Only 8 patients (7%) had signs of cirrhosis or portal hypertension.

Forty-five children underwent liver biopsy, fibrosis was seen in 33, 5 had F4 or cirrhosis, 3 had steatosis. Thirteen patients had both fibrosis and steatosis.

In the meantime (2009-2019), 43 other children were followed in our HPN center for more than two years but left the program: 19 were weaned from PN, 5 died from non-digestive complications (sepsis, cancer), 15 were transferred to adult centers, 4 underwent liver- small bowel transplantation. Only one child – born in 2011 - showed a rapid progression to cholestasis and severe portal hypertension and received liver-intestine transplantation for life threatening IFALD.

Conclusion: After ten years of using SMOF, children with IF on long term parenteral nutrition rarely evolve to severe and life threatening IFALD. However, most patients still present with abnormal biologic profiles and steatosis/fibrosis on biopsies. Identifying predictive factors for IFALD should help in the prevention of IFALD and the management of these patients.

P3.46 - A Novel Pump Device for Recycling Gastrointestinal Fistula Losses: Design and Feasibility Study

Gregory O’Grady 1,2, 3, Puja Sharma 1, Rob Davidson 3, John Davidson 3, Celia Keane 1, Ian Bissett 1

1 University of Auckland
2 Auckland Bioengineering Institute
3 Surgical Design Studio

Introduction: Enterocutaneous fistulae are a common cause of intestinal failure, and may necessitate parenteral nutrition (PN) and prolonged hospitalisation. Refeeding of fistula losses back into the distal gut is known to be beneficial, however implementation has been limited because devices are not commonly available, and manual recycling is unpleasant and labour-intensive. We present a novel device designed to enable easy and efficient chyme recycling, and report data from a first-in-human feasibility study.

Methods: The novel device comprises a compact centrifugal pump that can be placed inside a standard stoma bag. The pump is connected to an intestinal feeding tube which is inserted into the distal fistula limb. The pump is activated across the stoma bag by magnetic coupling to a custom-designed hand-held driver unit, effecting intermittent bolus refeeding while avoiding contact with the stoma effluent. Five speed settings were included to handle different chyme viscosities. Nutritional, medical, psychological and human-use factors were evaluated in an initial feasibility study.

Results: Following benchtop validation, the device was tested in 30 patients (1 dropout due to unrelated obstruction; median 31d; IQR 22-50d). Indications for inclusion were remediation of high-output fistula / stoma losses (n=7), dependency on PN (n=5), and gut rehabilitation prior to restoration of continuity (n=23). A range of chyme viscosities were successfully recycled with increasing efficiency over the course of the trial, due to iterative device improvements. Patients consumed low residue diets. Once established, chyme recycling was well-tolerated in all patients, using regular boluses of up to 200 ml per episode, performed as many times as needed per day. Patients experienced a variety of benefits including reduced net losses (>65% average volumes), PN cessation (4/5 patients, including all patients employing the final device iteration), liver function improvement, electrolyte normalisation, and improved quality of life. Of 6 patients with continuity restored at the time of reporting, none experienced post-operative ileus.

Conclusion: A novel chyme recycling device was developed and feasibility confirmed in an initial study. The device is easy to use and demonstrates multiple potential benefits including weaning of PN, gut rehabilitation, improved surgical outcomes, and reduced costs of care in these complex patients. A larger efficacy trial is currently planned.
Patients with Global Gut Dysmotility: Techniques and Long-Term Outcome

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Introduction: Global gut dysmotility (GGD) is a severely disabling disorder with the potential to develop gut failure with impaired quality of life. With the lack of effective medical therapy and pacing technology, surgical management with different modalities including gut transplantation (GTx) has recently evolved as part of the multidisciplinary treatment approach for this orphan population.

Methods: Between August 2012 and January 2019, a total of 172 patients with the diagnosis of GGD were referred to Cleveland Clinic, Center of Gut Rehabilitation and Transplantation (CGRT). TPN dependence was documented in 153 (76%). Of those, 82 (73%) underwent rehabilitative surgery (RS) and/or GTx. The diagnosis was confirmed by exclusion of gastrointestinal mechanical obstruction with radiologic evidence of global dysmotility including capsule endoscopy and sitz marker methodology. Most patients were adult female (88%) with extensive psychiatric history in more than 50% of the cases. Previous management included pharmacologic therapy, gastric pacing, partial colon resection, and external venting.

Results: With the intent to treat or as a bridge to GTx, 62 (76%) underwent RS with completion colectomy and ileorectal anastomosis (n=47), pyloroplasty (n=27) and chimney ileostomy (n=40). GTx with liver-free (n=39) and liver-contained (n=1) allograft was performed in a total of 20 (24%). Of these transplant recipients, 7 (35%) failed RS at our institution. With a mean follow up of 32 ±15 months, 30 patients are currently alive with an overall survival rate of (93%). In those with TPN dependency, the survival rate was better after RS (94%) compared to GTx (80%). Restoration of oral tolerance with achievement of full nutritional autonomy was accomplished in 20 (32%) and 30 (63%) of the current survivors, respectively.

Conclusion: Gut rehabilitative surgery and transplantation are effective treatment modalities for GGD patients. RS can be utilized as a long-term therapy or bridge for gut transplantation.

Body composition of pediatric patients with intestinal failure

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Introduction: Infants and children with intestinal failure (IF) are at risk of growth failure and altered body composition with increased fat and decreased lean mass compared to healthy children. Data from our cohort of patients with IF show normal growth on growth charts. However, body composition has not yet been assessed. The goal of the current study was to compare body composition of patients with IF treated by our program to healthy children using the United States National Health statistic database.

Methods: We conducted a retrospective cohort study of patients referred to our program between January 1st 2013 and July 15th 2018. For routine clinical monitoring, all patients with IF have annual Dual-energy x-ray absorptiometry (DXA) to assess bone mass. All patients with a DXA within the timeframe of the study and aged 8–18 years were included. Data related to demographics, residual bowel anatomy, nutritional support and growth anthropometrics were collected. Statistical analysis included means with SD for continuous variables and frequencies with percentages for categorical variables. Height, weight, fat, lean and bone mass were converted to their respective z-scores; regression analysis assessed predictors of body composition.

Results: Thirty-seven patients met inclusion criteria and a total of 68 DXA results were collected. The mean age at the time of the DXA was 10.7±2.2 years. Subjects demonstrated normal growth with weight and height z-scores of -0.67±0.99 and -0.7±1.3, respectively. Lean and fat mass z-scores were -1.61±1.09 and 0.24±0.74. Z-score for total body mass z-scores (p=0.01) and a trend towards increased fat mass with shorter time on parenteral nutrition (PN) (p = 0.09).

Conclusions: The results suggest normal growth and body composition in our patients with IF. This suggests that patients with IF have potential to accomplish normal body composition during growth. Further research is needed in the younger age group as well as separating those on and off PN. It is also important to determine positive contributors to body composition to increase efficiency of care in this population.
P3.49 - GLP-2 therapy for patients with short gut syndrome and intestinal insufficiency: A single center experience with future considerations
Cleveland Clinic

Introduction: The field of gut rehabilitation has recently evolved to restore nutritional autonomy without the need for home parenteral nutrition (PN) and subsequent transplantation.

Methods: A total of 35 short gut syndrome (SGS) patients received glucagon-like peptide-2 (GLP-2) and were followed from April 2013 through February 2019 in the Cleveland Clinic-Center for Gut Rehabilitation and Transplantation. Of these, 34 were adults with a mean age of 53.4±12.6 years (range: 14-72) and a female to male ratio of 2.5:1. Thirty-two of the 35 patients, were PN-dependent and 3 required IV fluid with micronutrient replacement. Leading causes of SGS were vascular occlusion (n=12), inflammatory bowel disease (n=12) and secondary motility disorders (n=5). Autologous gut reconstruction was performed in 32 of the patients (20 at our center and 12 at other institutions) with a mean small bowel length of 88.8±56cm (range 10-220cm). Intestinal lengthening utilizing serial transverse enteroplasty (STEP) was performed in 11 patients. The retained colon was partial in 31 (88.6%) patients and full with intact ileocecal valve in the remaining 4 (11.4%). The duration of GLP-2 therapy ranged from 2 to 63 months with an average dose of 0.32mL/d.

Results: With a mean follow up of 34±20 months, 33 patients (94.3%) are currently alive and 2 died of PN-related liver failure and advanced systemic arteriosclerosis. Full nutritional autonomy was achievable in 12 (54.5%) of the 33 current survivors with an overall success rate of 54.5%. Of these 12 patients, 11 (78%) continued to sustain the restored nutritional autonomy for 1 to 48 months after discontinuation of GLP-2 therapy. Meanwhile, a reduction in PN and IV fluid requirements was observed in 18 patients who failed to restore (n=15) or sustain (n=3) nutritional autonomy. None of the patients developed significant complications that warranted permanent discontinuation of GLP-2 therapy. There were no documentation of recurrent or de novo malignancy except in one adult patient who developed prostate cancer.

Conclusion: GLP-2 is an effective therapy for patients with SGS-associated intestinal insufficiency. This novel treatment should be more frequently utilized as a primary therapy or as an adjunct to autologous reconstruction and bowel lengthening. In addition, such a biologic agent should considered as an innovative approach to enhance the recovery and function of rejected intestinal allografts.

P3.50 - Portal hypertensive surgery for management of diffuse portomesenteric venous thrombosis - Surgical technique and long-term outcome
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Introduction: Patients with diffuse portomesenteric venous thrombosis (PMVT) are often referred to the abdominal organ transplant centers particularly those with experience in gut transplantation. Despite the proven therapeutic efficacy of multivisceral transplantation (MVTx) for these complex patients, the forgotten different modalities of portal hypertensive surgery could be a better alternative particularly for those with compensated liver disease and gut varices in the milieu of a thrombophilic state.

Methods: Between Aug 2012 and Jan 2019, a total of 28 patients referred to our center with diffuse PMVT underwent surgical intervention. Indication was history of variceal bleeding and occult varices in the setting of thrombophilia. All except 1 were adults with a mean age of 42±18 yrs. Precipitating causes of PMVT were thrombophilia (78%) and pancreatitis (18%). All patients had clinical, endoscopic and radiologic evidence of PMVT with stigmata of extra-hepatic portal hypertension. Variceal bleeding was reported in 36% patients. Type of shunt was determined intra-operative suitable visceral vascular anatomy and transplantation dictated by co-existence of advanced liver disease.

Results: Portal hypertensive surgery was performed in 27 patients (96%) while MVTx was performed in one. The surgical modality was atypical non-selective portosystemic shunt in 14 patients (52%) and gastro-esophageal devascularization in the remaining 13 (48%). Partial gastric devascularization was commonly performed in patients with atypical shunt that may not completely decompress the left portal hypertensive compartment. Spleenectomy was unavoidable in 4 patients. The utilization of autologous and synthetic vascular grafts are shown in Table 1

With a mean follow up of 40±26 months, overall shunt patency was radiologically documented in 78% patients. Recurrence of bleed occurred in only 1 patient (7%) despite shunt patency. Throughout the follow up period, surgical complications were minor (grade I & II) in 64% patients and major (grade III & IV) in 22% [Clavien grade]. With one mortality due to acute leukemia, all patients are currently alive with an overall survival rate of 96%.

Conclusion: Portal hypertensive surgery is a viable alternative to MVTx for the management of diffuse PMVT and preserved hepatic function. Portal decompressive surgery should always be considered for thrombophilic patients with silent gut varices who are in need of lifelong anticoagulation therapy.
CIRTA 2019 Book of Abstracts - Posters

P3.51 - Sensitivity of differential time to positivity compared to pour plates for diagnosing catheter-related bloodstream infection: an evaluation in patients with chronic intestinal failure

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Background: Clinical findings are unreliable for establishing the diagnosis of central venous catheter-related bloodstream infection (CRBSI) because of their poor sensitivity and specificity. Therefore, in order to establish a diagnosis of CRBSI, when this is clinically suspected and the central venous catheter (CVC) is to be preserved, ESPEN recommend that paired quantitative blood cultures (pour plates) or paired qualitative blood cultures from a peripheral vein and from the catheter are recommended, with continuous monitoring of the differential time to positivity (DTP). However, it is unclear whether quantitative or qualitative cultures provide the optimal method for diagnosing CRBSI in patients with intestinal failure (IF). A retrospective evaluation was undertaken in intestinal failure patients with long term CVCs to evaluate DTP against pour plates for the diagnosis of CRBSI.

Methods: A list of patients with a diagnosis of CRBSI was obtained from the intestinal failure (IF) unit database for a five year period, 2013 to 2017. Microbiology records were reviewed to obtain further information about blood culture and pour plate examinations. Organisms and times of collection, loading and positivity were recorded. Patients with a contemporaneous set of central and peripheral pour plates and blood cultures were included in an analysis of the sensitivity of DTP compared to pour plates.

Results: There were 61 (45.5%) episodes in 56 patients where complete sets of central and peripheral blood cultures and pour plates were received. All 61 episodes had positive central blood cultures, 59 (96.7%) had positive central line pour plates and 17 (27.9%) had positive peripheral pour plates. Using pour plates as the gold standard, DTP sensitivity was 96.0% for 50 episodes where pour plates were consistent with CRBSI. The sensitivity increased to 100% for 17 episodes where there were no delays in either collection or loading of blood cultures.

Conclusions: This is the first evaluation to support the use of DTP as a sensitive test in diagnosing CRBSI in IF patients and provides confidence to IF centres where pour plate cultures are not available. DTP can be used as a primary diagnostic test for CRBSI in patients with IF; however, in order for this to be of maximum value to clinicians, time to positivity needs to be routinely reported with blood culture results.

P3.52 - The Use Of Alemtuzumab As Immunosuppressive Induction In Intestinal Transplantation Among Pediatric Population

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Purpose: Many different pre-conditioning immunosuppression protocols have been reported to improve outcomes of patient and graft survival in intestinal transplantation (IT). The use of Alemtuzumab as an immunosuppressant inducer is not well studied in pediatric IT. Our aim is to analyze its impact on immunological complications among this population, comparing different protocols used in our center.

M&G: A retrospective study of immunosuppression protocols in pediatric (<18yrs) IT was conducted. 23 IT (35 multivisceral, 25 intestinal, 22 hepatointestinal, 3 modified multivisceral) were performed in 84 patients (male predominance -60%-; mean age 5.3yrs) between Oct99 - Oct2018.

Mean time on the waiting list was 272 days and waiting list mortality was 25%. Main indications for the transplant on the first instance were short bowel syndrome (69%), motility disorders (12%) and epithelial diseases (9.5%). Colon inclusion was included routinely from 2012.

Native spleen was preserved in 22/55 (40%) of multivisceral grafts.

Immunosuppression evolved over time in different stages, so we divided patients in 4 groups according to pre-conditioning regime (I-Basiliximab (n=13), II-Thymoglobulin (n=17), III-Basiliximab (n=56) and IV-Alemtuzumab (n=27)). Maintenance was performed in all cases with FK and steroids and also with Azathioprine in group I. Nowadays we use Alemtuzumab for patients >4 yrs and retransplantation of any age and Basiliximab for the rest.

Results: Groups with highest and lowest acute rejection rates were group II (65%) and group IV (1%, p<0.05). The latter presented the highest rates of PTLD (29%) and haematological disorders (35%). Despite this, it is the group with the lowest loss of grafts (45%).

Only 2 cases of humoral rejection were observed in the whole series in group I and group IV, respectively. Regarding chronic rejection, 3 cases in group II (23%) and 3 cases in group III (3%) were observed. There were no cases of GVHD among patients in group I but 3 cases in group II (6%), 3 in group III (6%) and 3 in IV (2%) were registered.

Finally, 35/84 (42%) patients died during follow-up and survival percentages between 53% (group II) and 73% (group III) were recorded.

Conclusions: Alemtuzumab appears to be effective in pediatric IT patients with high immunological risk or retransplanted due to its low rates of acute and chronic rejection; however, further studies are needed to conclude its safety in pediatric and the impact on PTLD, GVHD and hematological disorders compared to other therapies.
Do patients with gastroschisis have worst outcomes after pediatric intestinal transplantation?

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Introduction: Gastroschisis (Gx) is the leading cause of short gut syndrome in our intestinal rehabilitation population and the principal reason for intestinal transplantation (ITx). We investigated the results after ITx in this group of children and compared them to those with other etiologies.

Methods: A retrospective review of all ITx performed at our center was done. Eighteen ITx were done in 18 children between 2004-2018. Outcome data was collected for both surgical and non-surgical factors, and analyzed. Chi-square and independent t-test were used for statistical analysis. p<0.05 was considered significant. The study was approved by our Institutional Review Board (IRB #2019-2474).

Results: Eighteen patients underwent ITx. The most common diagnosis leading to ITx was Gx (7/18, 39%). Other causes included midgut volvulus (3), necrotizing enterocolitis (2), Hirschsprung’s disease (2), motility disorders (2) and other causes (2). Boys predominated in both groups (4/7 Gx vs. 6/11 in all others, p=0.91). Mean age at listing was similar (3.4±5.1 in Gx vs. 3.1±2.7 years in all others, p=0.70). The mean time on the waitlist prior to ITx was also similar in both groups (1.4±1.5 vs. 0.9±1.2 years, p=0.41). Both groups were transplanted at a similar age (5.3±4.7 vs. 4.0±2.2 years, p=0.5). One of 7 patients in the Gx group underwent multivisceral transplant compared to 2/11 in the all others group (p=0.83). Initial feeds were started later after the transplant in the Gx group (48.9 ±72.9 vs. 18.6±13.4 days, p=0.24) and mean postoperative hospital stay was also longer in the Gx group (112.9±165.5 vs. 78.7±53.9 days, p=0.53). However, a significantly greater proportion of Gx patients (6/7, 86%) were discharged on full enteral feeds compared to the patients in the all other group (4/11, 36%), p=0.04. Overall patient and graft survival in the entire cohort at 1 year was 89% and 83% respectively. 1 year patient survival was similar in either group: Gx 6/7 (86%) vs 10/11 (91%) in all others (p=0.73). 1-year graft survival was also equal in both groups: 5/7 (71%) in the Gx group vs 10/11 (91%) in all others (p=0.28).

Conclusions: Outcomes after pediatric ITx appear satisfactory and similar regardless of the underlying diagnosis and represents a viable alternative to long-term parenteral nutrition. Gastroschisis patients appear to have longer lengths of stay after ITx, but also appear to achieve a more complete recovery in terms of independence from parenteral nutrition.
P3.54 - Grafted colon in intestinal transplantation: does it make a difference in the outcome of the patients?

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Introduction: To include the colon as part of an intestinal or multivisceral graft is still a matter of debate. While some groups consider that it might be important for water absorption and residue breakdown without posing risks to patients, others state that its inclusion could result in an increase in infectious complications. Our aim was to analyze if the presence of the grafted colon in our patients has meant differences in their outcome.

Methods: Retrospective review of our historical series of intestinal and multivisceral transplants comparing patients that received a colon as part of the graft to those that didn't. A total of 107 patients received an intestinal or multivisceral graft between 1999-2018. Main indications were short gut syndromes (60 patients, 56.3%), motility disorders (36 patients, 33.6%) and untreatable diarrheas (14 patients, 13.3%). It was used a graft without colon in 61 (group no colon [33 M, 28 F]) and with colon in 46 (group colon [27 M, 19 F]). We compared the appearance of complications (such as rejection or infections) and survival curves adjusted by treatment protocol. A p-value below 0.05 was considered statistically significant.

Results: Average ages and weights at transplant were similar in both groups, with 67.5 months (6 - 361) and 17.2 Kg (4 - 52.7) in patients without a grafted colon and 64 months (6 - 235) and 16 Kg (3 - 59.2) in patients with colon. One hundred and two patients had a small bowel stoma. Timing of stoma take-down was similar in no colon group compared to colon group, with a median of 9.5 (1 - 125) months and 8 (1 - 50) months respectively. Rates of acute and chronic rejection were similar in both groups (39% vs 10% in no colon vs 35%/4% in colon). Rates of GVHD (15% vs 15% in colon vs 15% vs 12%) and PTLD (20% vs 10% in colon vs 13% in colon) were also similar. Median of septic episodes was 2 in both [no colon (0 - 8), colon (0 - 6)]. Among the multiple infectious agents analyzed, only clostridium showed a statistically significant higher rate of infection in colon group (2% vs 25% in colon). Patient and graft survival, adjusted by immunosuppression protocols, did not show differences.

Conclusion: Colon inclusion as part of the intestinal graft does not produce a higher rejection, GVHD or PTLD rates, nor a survival decrease. Even though the infection rate is also similar, there are differences in the infectious agents between the groups.

P3.55 - Allointestine colon conduit urinary diversion in combined intestine and kidney transplant: A case report

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Introduction: In intestinal transplant candidates, the incidence of renal failure is high and is associated with increased mortality. Additionally previous surgeries and loss of abdominal domain may make standard kidney transplant with ureterocystostomy impossible. 

Methods: We present an extreme case of combined small intestine, colon, and kidney transplant in a patient with short gut syndrome due to radiation enteritis, severe loss of abdominal domain, failed previous kidney transplant, and absence of native urinary system.

Results: Our patient is a forty years old female with a past history of bladder Rhabdomyosarcoma as an infant, who underwent bladder resection and post resection radiation. She developed radiation enteritis, renal failure, and eventually received a living related kidney transplant. Her renal allograft subsequently failed due to multiple urinary obstructions despite multiple revision surgeries, as well as chronic immunosuppression toxicity. As a result, she had short gut syndrome and renal failure with no feasible route of urinary drainage. In addition, she had severe loss of abdominal domain due to abnormal abdominal development from the radiation she received as an infant. She had multiple complications of her short gut syndrome and she underwent a simultaneous small intestine, colon, and kidney transplant. Donor was an eight years old female (132 cm, 23.4 kg), who became brain dead after severe exacerbation of asthma attack. Since there were no viable routes for urinary drainage, as there were no viable native intestines, and native and previous transplanted kidney ureter had been obstructed, a 13 cm allograft colon segment was used as a urinary conduit. The patient also underwent a complex abdominal closure with pedicled left anterior lateral thigh perforator flap by the plastic surgery team. One hundred days post transplant the patient is parenteral nutritional independent with normal renal function and her transplant conduit is healthy and functional.
Conclusion: In extreme cases of kidney transplant in intestinal transplant recipient patients with no feasible urinary drainage path, transplant intestine can successfully be used as a conduit. In review of literature, and to the best of our knowledge, this is the first description of utilization of allograft intestine as a urinary conduit.

P3.56 - Lower incidence of catheter related bloodstream infections (CRBSI) in children with short bowel syndrome (SBS) on parenteral nutrition (PN) treatment at home when compared to other aetiologies of IF

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Introduction: Currently the best outcome for chronic intestinal failure (IF) is achieved by discharging children home after parent/ carer(s) have undergone a formal training programme to connect and disconnect PN infusions from the child’s central venous catheter (CVC). Catheter related bloodstream infections (CRBSI) are the most common potentially life-threatening complication. The aim of this study was to prospectively compare the incidence of CRBSI in short bowel syndrome (SBS) with IF related to other aetiologies in children at home with care by parent(s).

Methods: A prospective record was kept of blood culture results from all children managed by our IF rehabilitation service over a 4-year period from January 2015 to January 2019. CRBSI was diagnosed when a child presented with significant fever and/or other symptoms suggestive of septicaemia and blood culture from the central venous catheter (CVC) was positive (in the absence of other focus of infection). The total number of children on long-term PN at home, the number with short bowel syndrome (SBS) and the number of CRBSI in each group were calculated.

Results: A total of 67 children (31 male, 36 female) aged from 6 months to 38 years were treated with PN at home. Eighteen cases had SBS (aetiology: volvulus in 7, necrotising enterocolitis in 6, gastroschisis in 2, long segment Hirschsprungs in 2 and congenital in 1). Ten were male and 8 female and aged from 6 months - 38 years. Other children were diagnosed with motility disorder in 27, primary mucosal disease in 14 and IF secondary to a haematological/immunology disorders in 8 cases. There were a total of 38,606 line days with 16,197 line days in SBS and 22,409 with other conditions. There were a total of 112 CRBSIs giving an overall infection rate of 2.95/10,000 line days. Sixteen infections were in children with SBS, giving an infection rate of 0.98 infections/1000 line days. Non-SBS children had 96 infections in 22,409 line days, an infection rate of 4.2/1000 line days. There was a significant difference in infection rate between those with and without SBS p<0.00001.

Conclusion: Children with SBS had a significantly lower incidence of CRBSI than those with IF due to other aetiologies. Factors related to the underlying disease may contribute to the rate of CRBSI. Further studies are needed comparing disease groups in children.
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P3.57 - Evidence of liver tolerance to severe hyperacute rejection in a pioneer model of multivisceral xenotransplantation.

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Xenotransplantation would be a solution for organs scarcity for transplantation however hyperacute rejection (HR) impair the this therapeutic method. We have described a model of discordant multivisceral xenotransplantation to assess the features of HR in various organs (1-3). Here we compare HR in the liver with the other organs of multivisceral xenograft. Method – After general anesthesia, multivisceral grafts containing esophagus, stomach, small intestine, colon, liver, pancreas, spleen and kidneys were recovered with a vascular pedicle containing the aorta with celiac axis and superior mesenteric artery and vena cava containing the hepatic veins were recovered from rabbits and implanted heterotopically in swine by aorta-aorta and cava-cava anastomosis (n=19). Multivisceral allotransplantation using rabbit-to-rabbit (n=5) composed the negative control group for HR. Three hours after graft reperfusion we sacrificed the animals and collected organs samples for histological study and IgG assessment by immunofluorescence. Features of HR were semi-quantitative graded in (0=normal, I=mild (edema, vascular congestion, and clumping of platelets in the microcirculation), II=moderate (diffuse edema, presence of some fibrin thrombi, intense vascular congestion, epithelial dysplasia, and foci of hemorrhage) and III=severe (presence of fibrin thrombi in all vessels, extensive interstitial edema, diffuse interstitial hemorrhage and confluent necrosis). T Student was used to compare HR in all organs from xenografts. Results – All animals were alive and well at the experimental end. The occurrence of HR was macroscopically noted in xenograft in 15 minutes. The autopsy revealed HR in all organs from multivisceral xenograft; however, we observed less severe HR in liver compared with esophagus, stomach, small intestine, colon, pancreas, spleen and kidneys (figure 3). IgG fixation was strong in in xenografts and absent in allografts. The occurrence of HR was observed in all xenografts. Conclusion - Heterotopic discordant multivisceral xenotransplantation is a relevant tool to study HR. IgG fixation by immunofluorescence was strong in the sites of HR. Therefore, we show for the first time that the liver is more tolerant to HR after multivisceral xenotransplantation than the other abdominal organs.

References:

P3.58 - Modified Spiral Intestinal Lengthening and Tailoring for Short Bowel Syndrome

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Background: The spiral intestinal lengthening and tailoring (SILT) procedure is a new surgical technique for autologous intestinal reconstruction in patients with short bowel syndrome. The aim of this work is to present a first description of a modified SILT technique by which the mucosal layer is left intact to possibly reduce the severe postoperative complications of intestinal leakage and abdominal abscess formation.

Materials and methods: The modified SILT technique was performed on a 30-cm-long intestinal segment in 2 pigs to determine the technical feasibility. Thereafter, the short-term clinical feasibility was monitored clinically in 2 dogs by gastrointestinal X-ray series at postoperative day 4 and by relaparotomy postoperative day 3.

Results: It was technically feasible to lengthen the intestinal segment from 30 cm to 20 cm and tailoring it from 3 cm to 17 cm in diameter, while leaving the intestinal mucosal layer intact. The postoperative course was uneventful for both dogs. The gastrointestinal X-ray series showed an inconspicuous intestinal transit time without any signs of stricture, perforation, or leakage. In the relaparotomy, the initially achieved lengthening and tailoring extents were preserved and the operated intestinal segment was well perfused with no early signs of necrosis, stenosis, or leakage.

Conclusions: Leaving the mucosal layer intact during SILT is technically and clinically feasible in the short term in a large animal model. Further studies are needed to fully assess the impact of this technical modification on the long-term outcome of larger series.
Introduction: Autologous gastrointestinal tract reconstruction surgery (AGIRS) and serial transverse enteroplasty (STEPS) are worldwide accepted strategies for children and adults aiming to achieve intestinal rehabilitation. The aim of this study is to show a series of children with intestinal failure (IF) that underwent surgical rehabilitation at a single center and their outcomes.

Material and methods: This is a retrospective review of a prospectively filled database, from March 2006 to December 2018, including every patient <18 years of age with chronic IF that was referred to this program and underwent abdominal surgery with the aim of intestinal rehabilitation. Demographic data, diagnosis, type of surgery, intestinal anatomy (type 1 - terminal jejunostomy; type 2 - jejunoo colonic anastomosis; type 3 - jejunoo ileo colonic anastomosis; subtypes A < 40 cm and B > 40 cm) and PN independence are analyzed.

Results: From a total number of 122 Pediatric patients (pts) referred with IF, 19 underwent rehabilitation surgery; 17 of them are followed at our center and were included in the analysis. Type of surgery performed was: STEP (6); AGIRS (13). The mean time of follow up after surgery is 65 months (sd ± 49.6). A total of 8 pts gained enteral autonomy after surgery, in a mean time of 12.3 months (sd ± 7), additionally 2 pts reached enteral autonomy on hormonal therapy (teduglutide), initiated 63 and 129 months respectively after surgery (table 1).

Seven pts continued on PN; from those, 2 pts underwent intestinal transplant due to loss of venous access, 1 pt died on the intestinal transplant waiting list due to liver failure and 4 pts remain PN dependent with no indication of transplantation (2 are in the process of starting teduglutide treatment). At the end of the studied period, the Kaplan Meier freedom from PN survival is 83.2% (graphic 1).

Conclusions: Surgical rehabilitation should be considered as the first strategy to achieve intestinal autonomy; the indication for STEP or AGIRS should be done in the context of a multidisciplinary team. The only variable analyzed that showed statistical significance for surgical rehabilitation was residual small bowel length. The introduction of hormonal therapy provides an additional benefit, limiting the need for transplantation.
Native spleen preservation attenuates graft versus host disease in an experimental model of modified multivisceral transplantation.

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**Introduction:** Native spleen used to be removed in patients undergoing modified multivisceral transplantation (MMVT) increasing the risk of sepsis and graft-versus-host disease (GVHD). The mechanistic basis of these effects including mixed chimerism level is poorly understood. Based on a novel experimental procedure of MMVT that triggers GVHD, we aimed to evaluate the effects of native spleen preservation in this model in order to gain insight into the mechanisms that may mediate this phenomenon.

**Material and Methods:** Heterotopic MMVT from Lewis to Brown Norway rats was performed. MMVT graft consisted of stomach, duodenum, pancreas, spleen and the small bowel of the donor. The native spleen was removed (MMVT-S) in half of the MMVT recipients (N=5), while the remaining preserved their own spleen (MMVT+NSP) (N=5). After MMVT mixed hematopoietic chimerism was determined in native spleen and peripheral blood samples by flow cytometry using strain-specific HLA antibodies. Also, histopathological (using Pintar score) and clinical signs of GVHD such as skin rash were evaluated.

**Results:** All animals in the group MMVT-S presented clinical signs of GVHD such as skin rash, weight loss, and diarrhea, among others, between 7-10 days after transplantation. Skin rash was particularly remarkable in the ears and periocular area. Clinical signs of GVHD were less frequent in MMVT+NSP recipients (40%; p<0.05). Also, significant differences between groups were observed in the histopathological study and mixed chimerism in peripheral blood 3 days after MMVT (36.6 and 23.62 % in MMVT-S and MMVT+NSP respectively) (Figure 1). Also, native spleen showed an 18±5 % of chimerism (CD3+ donor cells) 1 week after transplantation. After day 10 post-MMVT, when the signs of transient GVHD disappeared, both groups presented graft rejection.

**Conclusions:** Native spleen preservation in MMVT recipients attenuated the occurrence of GVHD and reduced the level of chimerism compared to recipients that underwent spleen removal during MMVT. Despite more studies are necessary, our preliminary results suggest that native spleen preservation increase the efficacy in removal of anti-recipient reactive clones that would explain the protective effect of native spleen preservation against GVHD.
POSTERS PRESENTED ON SATURDAY JULY 6, 2019
CIRTA 2019 Book of Abstracts - Posters

P4.01 - Reduction of central line associated bloodstream infections and line occlusions in pediatric intestinal failure patients on long-term parenteral nutrition using an alternate locking solution, Kitelock

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Background: Patients with intestinal failure (IF) are dependent on parenteral nutrition (PN) for growth and survival, however are at high risk of central line associated bloodstream infections (CLABSI) and line complications, resulting in significant morbidity and mortality. 70% of ethanol locks have been used for the treatment and prevention of CLABSI with success but reported issues relate to product availability and catheter patency. Kitelock 4%, an EDTA solution, is an effective non-antibiotic antimicrobial, anti-biofilm and anticoagulant agent shown to significantly eradicate clinically relevant microorganisms by 87% within central venous access devices. Adult studies have demonstrated efficacy of EDTA in CLABSI reduction, however there remains a deficit of pediatric data. Our objective was to determine Kitelock efficacy in the prevention of CLABSI and reduction in line occlusions in pediatric IF patients on long-term PN.

Methods: We conducted a retrospective cohort study of patients managed by IF programs at 2 tertiary Canadian pediatric centres between April 1, 2017 to December 31, 2018 who received Kitelock. Data was collected for 12 months prior to and following initiation of Kitelock. CLABSI, line replacements and use of alteplase (per 1000 catheter days) before and after initiation of Kitelock were compared using a Wilcoxon matched-pairs signed-ranks test. Data was reported as medians (interquartile ranges) and frequencies (proportions).

Results: Twenty patients (18 girls; median age 83 months [range, 8-232 months]) began Kitelock for recurrent infections or sluggish/occluded line. The rate of CLABSI prior to starting Kitelock was 2.7(0.4-4) per 1000 catheter days. Patients received Kitelock for a median of 365 (278-365) days with no infections in the 12 months following commencement of Kitelock (p=0.002). Median rates of occlusive episodes for the entire cohort prior to starting Kitelock were 0 (0-5.0) in the 12 months prior to starting Kitelock and 0 (0-2.0) after starting therapy (p=0.018). In patients with occlusions (n=9), the median rate of alteplase use prior to starting Kitelock was 5.5 episodes (278-365) days before and after institution of Kitelock were compared using a Wilcoxon matched-pairs signed-ranks test. Data was reported as medians (interquartile ranges) and frequencies (proportions).

Conclusions: Kitelock was effective in reducing CLABSI and catheter occlusions in pediatric patients with long-term central access requirements.

P4.02 - Micronutrient levels in intestinal failure patients dependent on parenteral nutrition at home over a 5-year period

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Introduction: To analyse longitudinal changes in micronutrient blood levels in all patients already established for >12 months on parenteral nutrition (PN) treatment in our home intestinal failure (IF) rehabilitation service.

Methods: Laboratory reports of blood micronutrient levels and concurrent CRP levels in children with IF established on PN >12 months prior to 2013 were reviewed in 2013, 2015 and 2017. Peditrace® or additrace® supplement was routinely included in PN and doses adjusted according to blood levels with additional enteral supplements if appropriate. Results were categorised as deficient (> 30% below normal range) or normal, based on mean of 2 values (one value only for vitamin D) obtained over 6-months in each time period, 2013, 2015 and 2017. The study was approved as an audit. Two tailed Fisher’s exact test was used to calculate p values.

Results: 30 patients (16 male) diagnosed with motility disorders in 2 (40%), short bowel syndrome (SBS) in 7 (12%), mucosal disorders in 12 (20%). Five children weaned off PN, 2 transitioned to adult care and 1 died (due to underlying disease). IV lipids including vitlipid® were added to PN 2 or 3 nights/week in 27, four nights in 2 and not given to one child. Please see results table for levels obtained in each time period (samples not obtained if patient no longer on PN or blood sample insufficient for analysis). There was a similar incidence of each micronutrient deficiency in the different diagnoses.

<table>
<thead>
<tr>
<th>Micronutrient</th>
<th>2013</th>
<th>2015</th>
<th>2017</th>
<th>P value comparing 2013 with 2017</th>
</tr>
</thead>
<tbody>
<tr>
<td>Copper</td>
<td>16/30 (53.3%)</td>
<td>23/30 (76.6%)</td>
<td>19/27 (70.4%)</td>
<td>0.017 significant</td>
</tr>
<tr>
<td>Zinc</td>
<td>27/30 (90%)</td>
<td>28/30 (93.3%)</td>
<td>21/27 (77.8%)</td>
<td>0.6</td>
</tr>
<tr>
<td>Selenium</td>
<td>24/30 (80%)</td>
<td>27/30 (90%)</td>
<td>19/27 (70.4%)</td>
<td>0.7</td>
</tr>
<tr>
<td>Vitamin A</td>
<td>17/30 (56.7%)</td>
<td>25/30 (83.3%)</td>
<td>26/27 (96.3%)</td>
<td>0.003 significant</td>
</tr>
<tr>
<td>Vitamin E</td>
<td>18/30 (60%)</td>
<td>30/30 (100%)</td>
<td>21/27 (77.8%)</td>
<td>0.41</td>
</tr>
<tr>
<td>Iron</td>
<td>9 (3%) (28.6%)</td>
<td>9/15 (60%)</td>
<td>8/10 (80%)</td>
<td>0.51</td>
</tr>
<tr>
<td>Ferritin</td>
<td>26/30 (86.6%)</td>
<td>26/27 (77.8%)</td>
<td>19/27 (70.4%)</td>
<td>1.0</td>
</tr>
<tr>
<td>Number of patients with no deficiency</td>
<td>5/30 (16.6%)</td>
<td>7/30 (23.3%)</td>
<td>7/27 (25.9%)</td>
<td>0.31</td>
</tr>
</tbody>
</table>

*<30 patients as patients no longer on PN or results not available due to insufficient sample

Table showing prevalence of micronutrient deficiencies in long term home PN patients

Conclusions: The majority of children with chronic IF on home PN had some micronutrient deficiencies with increasing incidence of normal levels with time. Two or 3 nights/week appeared sufficient for maintaining fat soluble vitamin levels. There were significantly less cases of low copper and vitamin A levels with time. Our results emphasise the importance of routine monitoring of micronutrients and the need to prescribe home PN according to individual requirements.

peditrace® commercially available preparation of micronutrients
additrace® commercially available preparation of micronutrients
vitlipid® commercially available preparation of lipids
**P4.03 - The role of parenteral nutrition in children after bone marrow transplantation**

Sunita Amar Rajani, James Evans, Dominika Gayda-Pimlott, Duncan Cartner, Paul Veys, Jutta Köglmayer  
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**Background:**  
The first successful bone marrow transplant (BMT) in a child was carried out in 1968. Since then, it has become an established therapy for a variety of childhood conditions including malignancies not responding to chemotherapy or radiation, congenital immunodeficiencies and haemoglobinopathies. Complications are common and include graft versus host disease (GVHD), graft failure or rejection, veno-occlusive disease and malnutrition. Parenteral nutrition (PN) is used for gut sterilisation in the conditioning phase and offered to children with severe mucositis and GVHD of the gut following transplant and as part of the standard protocol for cord transplant.

**Methods:**  
Patients undergoing BMT in a large nationally commissioned referral centre were identified from the patient database between January 2014 and May 2018. Underlying pathology, date and type of BMT, number, age, sex and outcome of children requiring PN for less or more than 28 days and those referred for home PN were recorded.

**Results:**  
254 (152 males and 102 females) children underwent a BMT during the study period, 64 patients received PN for less than 28 days of which 9 died. PN was required for more than 28 days in 125 of which 41 died. 15 children were discharged on home PN. The indications were chronic GVHD in 9 and colitis or enteropathy in 6. Four children passed away during this period; one from line sepsis and one each due to pulmonary hypertension, lung GVHD and intracranial relapse of leukaemia. 5 children were successfully weaned off intravenous nutrition and 6 remain on home PN.

**Conclusion:**  
The majority of children undergoing BMT requires PN. Long term PN > 28 days is common and a small percentage of patients will need home PN. Although home PN improves the overall outcome the mortality rate amongst those patients is high.

**P4.04 - The Surgical Management Of Acute Bowel Ischemia In Elderly Patients In Order To Avoid Short Bowel Syndrome: A Multicenter Study**

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2 Department of Surgical Sciences - Umberto I University Hospital-Rome (Italy)

**Introduction:**  
This is a multicenter study performed in two Italian tertiary care centers: Emergency General Surgery Unit at St. Orsola University Teaching Hospital-Bologna and Department of Surgical Sciences at Umberto I University Teaching Hospital-Rome. The aim was to compare the results of different approaches among patients with acute bowel ischemia over 65 years of age.

**Methods:**  
Sixty-three patients were divided in two groups: 1) VS-group- 28 patients treated at Department of Surgical Sciences and 2) GE-group- 35 patients treated in Emergency General Surgery Unit. Mean age was 80 years, significantly higher for the GE-group (p<0.001). Gender was predominantly female in both groups, without statistical difference. Pre-operatively, laboratory tests didn’t show any difference in white blood cell count, serum lactate levels or serum creatinine among patients, while increase of c-reactive protein was observed in VS-group with significant difference (p<0.001). The main cause of acute bowel ischemia was embolism in VS-group (p=0.03) and vascular spasm in GE-group (p<0.001). On CT scan, bowel loop dilation was present in 58.7% of patients without statistical difference in both groups.

**Results:**  
The time lapse from diagnosis to operation didn’t show significant differences between two groups (mean 349.4 min). Pre-operative heparin therapy was administered in VS-group more frequently (p<0.001). In VS-group, thrombectomy was the most frequent procedure (19 patients) associated with bowel resection in 9 cases. In GE-group, 22 patients had an explorative laparotomy (p<0.001) and 8 had a bowel resection with anastomosis and 5 a bowel resection plus stoma. A second look was required more significantly in VS-group (p<0.001). Post-operative morbidity affected significantly GE-group (p=0.02). The 3-day survival was significantly higher in the VS-group (p<0.001). At discharge 32 patients (50.8%) were alive, 21 in VS-group (p<0.001). Only one patient among both groups (16%) had a short bowel syndrome.

**Conclusions:**  
In elderly patients with acute bowel ischemia, surgery should be always pursued whenever the interventional radiology is not assessed as a viable option. Both groups of patients showed an excellent outcome in terms of avoiding a short bowel syndrome. A multidisciplinary management by a dedicated team could offer the best results to prevent large intestinal resections.
Three patients were received rice based diet after intestinal transplant. Rice based post-operative diet after intestinal transplant is very important part of transplant, but post-operative diets vary depending on countries because they are based on cultural background. Rice based diet (so called Japanese food) is usually used after post-operative treatment in Japan. We introduced rice based post-operative diet and assess its safety and efficiency.

**Methods:** Patients who received isolated intestinal transplant in our institution were reviewed. All patients received rice based post-operative diet after intestinal transplant. Tacrolimus based immunosuppressions were administrated intravenously then orally. Diet starting date, advancing, parental nutrition off and outcome were assessed.

**Results:** Three patients were received rice based diet after intestinal transplant. Age at transplant was median 17 year olds. (Ranged 14 year olds to 32 year olds) Original diseases were congenital microvillous atrophy, Hirschsprung's allied disease and short gut syndrome. Types of donor were living donor (n=1) and cadaveric donor (n=2). Induction therapies were Daclizumab (n=2) and rabbit anti thymus globulin (n=2). Everolimus was added at one month after intestinal transplant for two patients. Intestinal graft length was median 160cm (150-260cm). Two patients had enteral feeding tube. All patients started rice based post-operative diet as rice gruel (Omoyu) at post-operative day (POD) 26 (ranged POD 7 to POD49) orally. Diet was advanced to rice gruel in three degree (rice: water=1:20), five degree (rice: water=1:13) and regular (rice: water=1:5). Diet reached regular gruel at mean POD 45 (ranged POD7 to POD87). Side dish started later avoiding protein oral intake. Patients are free from parental nutrition at median POD119 (ranged POD18 to POD87). Side dish started later avoiding protein oral intake.

**Conclusion:** Rice based post-operative diet after intestinal transplant was safe after intestinal transplant. Rice based post-operative diet had great advantages its form and less protein. Softness of gruel were easily controlled with rice water ratio. Polished rice has less protein. The contents of side dish were need to be further evaluation.

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**P4.06 - Combined multivisceral and renal transplant in a patient with JAK-2 mutation**

Maria Cristina Segovia, Kadiyala Ravindra, Andrew Barbas, Bradley Collins, Deepalaxmi Borle, Jigesh Shah, Debra Sudan

Duke University Hospital

43 y.o. male with J AK-2 mutation developed cirrhosis due to Budd-Chiari syndrome and had extensive portal and mesenteric vein thrombosis that precluded a liver transplant alone. His liver disease was complicated by refractory ascites, encephalopathy, severe muscle wasting and MELD-Na of 36. In addition, he developed hepatorenal syndrome and hemodialysis was initiated. Despite lack of intestinal failure, the patient required a multivisceral graft for anatomical reasons of lack of ability to restore portal inflow to the liver graft.

Multivisceral transplant (MVT) including liver, pancreas, stomach, intestine and partial colon was performed followed by kidney transplant the following day. Despite preoperative partial arterial embolization of the native superior mesenteric and splenic arteries, he required massive transfusion of 47 units PRBC’s due to incomplete embolization and severe portal hypertension. The patient received Thymoglobulin induction and has been on triple drug immunosuppression (tacrolimus, mycophenolate mofetil, prednisone).

Thrombophilia was managed with IV heparin perioperatively and enoxaparin was started postoperatively. This was associated with gastrointestinal bleed from the gastrojejunal anastomosis during the first postoperative week. The hepatic dose reduction during the period of GI bleed was associated with the development of non-occlusive thrombi in both internal jugular veins. In addition, he was treated with hydroxyurea for thrombocytosis, subsequently plateletphoresis and anagralide when platelet count reached >14 million. By the time of discharge, the patient was on full enteral nutrition and had normal renal function.

This case highlights the indication of MVT for patients with hypercoaguable states and extensive portomesenteric thrombosis. In this patient, his hypercoaguable state was more extensive than many due to involvement of his hepatic veins leading to Budd Chiari syndrome. The cause of his hypercoaguable state was known to be a J AK-2 mutation, although the cause is not always identifiable. Perioperative anticoagulation with standard medications prevented thrombotic complications, however thrombocytosis developed (likely from splenectomy) and was not controlled by hydroxyurea and required the use of plateletphoresis and anagralide. Our plan will be for life-long anticoagulation and we anticipate switching from enoxaparin to apixaban after discontinuation of protocol monthly bowel biopsies and ostomy closure.
**P4.07 - Does our current dietetic input meet the long term needs of our post transplant patient population?**

Laura Johnson, Girish Gupte, Jane Hartley  
*Birmingham Children’s Hospital, Steelhouse Lane*

**Introduction**
As a super-regional centre with patients from all over the UK frequent face to face contact is not feasible, therefore telephone or email dietetic consultations are necessary. Current dietetic review practice is to personally see patients at their annual review and outpatient appointments to provide assessment of nutritional parameters, intake and matching this to need. Telephone consultations are more ad hoc but are amenable to be standardised so to provide a robust dietetic follow up. We therefore report on current practise and recommendations for future standards of care.

**Aim**
To review our current practise of dietetic input and provide recommendations for future standards of care.

**Methods**
Dietetic notes of all patients who have a small bowel containing graft throughout 2018 who are cared for at Birmingham Children’s Hospital, were retrospectively reviewed to identify all patient contact during the study year. We then looked at the intensity of nutritional support and dietetic needs. Of note, some patients will also have local dietetic intervention.

**Results**
32 patients were under review throughout 2018. Table 1 shows the dietetic input required and table 2 the median number of reviews each group received. As expected the patients requiring the most intense dietetic intervention have received the most input. It is vital however to consider individual patient need as some patients on less intense intervention required more frequent review demonstrated in the range of patient contacts. 6 patients also had local dietetic intervention.

**Conclusion**
Dietetic input into small bowel graft patients in imperative. Rationalisation of resources means that those at high risk or high supplemental needs should be prioritised and a standard of care developed.

We propose the following standard:-

- Develop care pathway for each group highlighting frequency of follow up and method to escalate concerns to consultant.
- Written information for families regarding review process to ensure early reporting of concerns.
- Referral to local Dietitian where appropriate.
- Telephone proforma to ensure consistency.

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**P4.08 - Clinical And Histologic Characteristics Of Intestinal Failure Associated Liver Disease In Pediatric Patient Population**

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2 Department of Pathology, HUSLAB, University of Helsinki, Helsinki, Finland

**Introduction**
Our aim was to assess clinical and histologic characteristics of intestinal failure associated liver disease (IFALD) in children.

**Methods**
This was a retrospective single center study. In total, 89 consecutive children with intestinal failure (IF) and liver biopsy obtained between years 2006-2018 were included. Liver biopsies were analyzed for histology with simultaneous measurements of serum liver biochemistry, citrulline, spleen size and cholelithiasis (ultrasound, n=86). Presence of esophageal varices was based on gastroscopy (n=70). Fisher’s exact and Mann Whitney U test were used for comparisons.

**Results**
Median patient age was 3.3 years (IQR 0.6-9.3), age-adjusted small bowel length was 26% (17 -48%) and 49 patients were currently receiving PN at the time of liver biopsy. The causes of IF included short bowel syndrome (SBS, n=67), primary motility disorders (n=20) and enteropathies (n=2). Significant histologic liver fibrosis (Metavir stage ≥2), cholestasis, portal inflammation and steatosis was observed in 36%, 29%, 26% and 31% of patients. Any degree of cholestasis (49% vs 5%), portal inflammation (43% vs 5%) and fibrosis (65% vs 38%) were more frequent during PN (P<0.05 for all), whereas steatosis (30% vs 29%, P=NS) and significant fibrosis were equally common after weaning off PN (Table). Splenomegaly was observed in seven (8%) and esophageal varices in one (14%) patient, mostly during PN delivery. Cholelithiasis was observed in six (7%) patients. Presence of significant liver fibrosis associated with short remaining small intestine, prolonged PN, missing ileocecal region and elevated ALT (Table). One patient died of IFALD and one with associated extrahepatic portal vein occlusion underwent liver transplantation after weaning off PN.

**Conclusion**
Cholestasis and portal inflammation are nearly exclusively associated with PN delivery, whereas significant fibrosis and steatosis frequently persist after weaning off PN. Splenomegaly and esophageal varices were infrequent complications of pediatric IFALD. Although IFALD was uncommon cause of death or liver replacement therapy, long-term significance of persisting fibrosis and steatosis needs to be resolved.
P4.09 - Anastomosis Less Than 10 Cm From The Ileocecal Valve In Intestinal Failure Associated With Short Bowel Syndrome Is Safe. Experience Of 9 Years In The Unit Of Intestinal Failure. National Hospital Guillermo Almenara Irigoyen. Lima Peru.

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1 Emergency Service of Almenara Hospital
2 Coordinator of the Intestinal Failure Unit
3 Professor of the Universidad Peruana Unión- Lima Perú

Introduction: The short bowel syndrome (SBS) associated with intestinal failure (IF) is defined as a state of transient intestinal insufficiency characterized by abundant losses by enterostomy that lead to nutritional, hydroelectrolytic and metabolic alterations. In Peru, it is mainly produced by intestinal ischemia and postoperative complications.

In surgical management it has been tried to recover the largest amount of intestine, however, anastomosis has been avoided very close to the ileocecal valve (ICV) adducing greater risk of dehiscence due to high intraluminal pressure and little vascularity.

A series of patients with anastomosis less than 10 cm from the ICV are analyzed in the present study.

Patient materials and methods: Series of cases of patients older than 18 years with a diagnosis of SBS associated with IF; with ileal jejunal anastomosis less than 10 cm from the ICV. Where a standard protocol was implemented.

Between 2011 and 2018, 120 patients with a diagnosis of SBS associated with IF were treated. We performed 52 surgeries of autologous gastrointestinal restitution (AGIR). In 11 of them, an ileal jejunal anastomosis was performed less than 10 cm from the ICV and 5 of them were made to the same ICV.

We did not find anastomosis dehiscence and 2 complications occurred: a hemoperitoneum reoperated at PO1 for hemostasis and an inadvertent lesion reoperated at PO2 to perform raffia. After the AGIR there were 7 patients with more than 1 meter of residual intestinal length (170 cm average) and 4 patients with less than 1 meter of residual intestinal length (69 cm average); the Nutrition Parenteral was administered 21 and 35 days on average respectively; and the autonomy for the oral diet was given at 3 to 5 weeks.

Conclusion: The AGIR in IF associated with SBS with anastomosis very close to the ICV are safe if they are performed with regulated techniques.

P4.10 - Chronic Intestinal Failure: When Children Become Adults

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Rationale: Major advances in recent years have resulted in improved survival for patients with chronic intestinal failure (CIF). There is very little data on the future of children that became adult with CIF. The aim of our work was to describe this population since the existence of our center.

Methods: In an approved HPN centre for adult with a dedicated activity for CIF since 1984, we collected retrospectively all data related to patients treated with home parenteral nutrition during infancy (at least 4 years before transition to our centre) and transferred in our centre since 1984. We evaluated demographic data, the social evolution and the main complications at the adulthood. The final time of data collected was the 1th December 2017.

Results were expressed as median[±SD].

Results: Among a total of 870 HPN patients since 1984 and 2017, 44 young adults (17F/27M) were transferred from 3 paediatric hospitals. Age of transition was 19±2 years. The principal etiologies of CIF were short bowel syndrome (n=18), CIPO (n=21), mucosal disease (n=5). At the end of follow up defined as the latest news (december 2017) or death, 7/44 patients were deceased (2 after intestinal transplantation, 3 after a sepsis, 2 due to liver failure), 3/44 were weaned off PN (2 due to growth factors, 1 after intestinal transplantation), 33/44 were alive requiring HPN (6±1,7 infusions/week; 2,2±1,3l/day; 29±13kcal/kg/ day). Oral intake was 2000±1085kcal/day but 9/44 presented remaining oral disorders. Seventeen/44 had a regular work (35±6,5hours/week). 23 lived with their parents; 17 lived in partnership and 7 had at least one child.

Conclusions: Despite progress in survival and quality of life in HPN, many children who become adults stay with their parents and do not work. The transition requires probably a better social, educational and psychological preparation if we want to improve the future of these patients.
P4.11 - Intestinal failure management in the middle-income countries without home parenteral nutrition

Hamed Nikoupour 1, Alimohammad Moradi 1, Mohammad Yasin Karami 1, Saman Nikeghbalian 1, Seyed Ali Malek-hosseini 1
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Introduction: Intestinal failure (IF) - a life-threatening medical status - is a rare disease still remains orphan in most countries. However, the management of this condition is still unclear in the middle-income countries (MIC) not benefiting from HPN. The current study aimed at investigating the current management of IF utilized in our center.

Methods: In the current cross-sectional study, data were retrospectively collected from 30 patients (25 males, 5 females) who had IF between January till October 2020 at BOOALI SINA hospital affiliated to Shiraz University of Medical Sciences, Iran.

Results: This cross-sectional survey included 30 patients (mean age: 44.13±10.32 years). The study participants consisted of 5 females (16.7%) and 25 (83.3%) males. Short bowel syndrome and Entero-cutaneous fistula were reported in 60% and 26.7% of patients, respectively. In the present study, the most common Shaffer functional classification type of IF was type 3 (66.67%). Mesenteric Ischemia was the main cause of IF in the present study (46.7%). The most common complication was CI (46.7%). The overall mortality rate for 1 year was 16.7%. STEP procedure was performed on 2 patients (6.7%). Nine patients were uneventful at home without the need for home parenteral nutrition.

Conclusion: For the first time, this study presents a guideline on the development patterns and management exigence of IF in patients from MIC not benefiting from HPN. It is recommended to pursue the development of registries and reproduction of health policies in terms of the promotion of long-term care for patients with IF in MIC not benefiting from HPN in further studies.

P4.12 - Immune response in an experimental model of pig to rabbit abdominal fascia transplantation.

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Introduction: Lack of enough space for safe closure after transplantation leads to the use of fascia and abdominal wall grafts to overcome this problem. Little is known about the immunogenicity of fascial transplants. We have been using an experimental model of fascia transplants in rabbits for the last 4 years. Here we show the results of a subset of 9 pig to rabbit xenotransplants.

Methods: Fascia procurement: a midline abdominal incision in the pig served for a 3 x1 cm fascial graft procurement. The graft was kept at 4ºC in serum (7) or glutaraldehyde (G) (2) and transplanted 24 h later in the rabbits, removing a similar size fascial tissue. Macroscopic examination was also done.

Results: There were not appropriate blood samples from 2 rabbits. One rabbit died of diarrhea on POD 16. All the transplants had an important reaction at 1 month, with seroma or graft detachment. At 3 months, the G grafts maintained an important reaction, but the other were integrated except one that had a big ventral hernia.

Conclusions: All the xenografts showed important immune reactivity at 1 month, with seroma or graft detachment. At 3 months, the G grafts maintained an important reaction, but the other were integrated except one that had a big ventral hernia. Flow cytometry results were positive in all the surviving rabbits at 30 days, except in those with G treated grafts. At 60 days, 4/10 had negative results (2 of them G treated). The antibodies showed cytotoxic activity by CDC-XM test in all serum tested (3 rabbits).

Conclusions: All the xenografts showed important immune reactivity at 1 month, but this decreased after 3 months, and tissue integration was observed at 3 months except in those treated with G. In this model, the immune reaction seem to decrease after the first month, with good integration of the graft in the majority of animals after 3 months. The G solution used had some other components that could explain the tissue reaction.
It is well-known that state insurance in Turkey covers all infections constituted the smallest group (33.3 %) in our CLABSI series. Enterococci, klebsiella and the other gram-negative bacilli and fungal bacterial infectious agents (66.6 %). Staphylococcus aureus (SA), and pseudomonas aeruginosa were the most commonly isolated and all catheters were removed and cultured. Acinetobacter baumannii and pseudomonas aeruginosa were the most commonly isolated bacterial infectious agents (66.6 %). Staphylococcus aureus (SA), methicillin resistant SA (MRSA), coagulase-negative staphylococci, enterococci, klebsiella and the other gram-negative bacilli and fungal infections constituted the smallest group (33.3 %) in our CLABSI series.

Conclusion: CLABSI is a very common problem in the intensive care unit. These infections can cause mortality, and costly. Only through best practices, protocols, checklists, and establishing a culture of patient safety in healthcare institutions can one reduce CLABSI to zero.

**Introduction:** Total parenteral nutrition (TPN) is used when gut fails to provide complete nutrition. Central line associated blood Stream infection (CLABSI) is a major complication of this therapy. It is defined as a laboratory-confirmed blood-stream infection not related to an infection at another site that develops within 48 hours of a central line placement. Of all the healthcare-associated infections, CLABSIs are known as the most costly, and most cases are preventable with proper aseptic techniques, surveillance, and management strategies. The objective of this study was to report the incidence of CLABSI and associated mortality in patients admitted to our intensive care unit (ICU) whom started on TPN.

**Methods:** The study was approved by our institution’s Ethics Committee (no: 23/ 01/ 2018). All patients who received TPN in our adult ICUs from the period between January 1, 2018 till December 31, 2018 were included. Data were retrieved from the hospital electronic data base. Total number of patients receiving TPN and catheter-related complications were recorded. Demographics and CLABSI rates were documented. In the case of tunneled catheters, the definition was changed as follows: signs of inflammation confined to an area (typically < 2 cm) surrounding the catheter exit site and the presence of exudate that proves to be culture positive.

**Results:** A total of 186 patients were included in the study. Mean age was 51 years (range, 18 – 91). The average period of ICU stay was 45 days (range, 1 – 204), including longer hospitalization time due to surgical complications or existing comorbidities. The postoperative stay in intensive care unit (ICU) was also included in this period. In our data, the only health insurance coverage was seen to be supplied by the state (n=186, 100 %). There was no private insurance coverage. No patients used their own financial resources for treatment, as well (including Syrian refugees).

**Conclusion:** It is well-known that state insurance in Turkey covers all polyclinic applications and postdischarge home care (including all refugees), and patients pay only a small amount of fee as employee contribution for the medications at pharmacies. The new information in the present study is the governmental coverage of all Syrian refugees in ICUs, as well. Taking the great amount of bill that an ICU patient should encounter, the importance of state insurance is obvious for the comfort of patient and the ease of financial portion of their treatment.
**P4.15 - IF associated liver disease - a single centre cohort study of adult patients referred for consideration of intestinal transplantation**


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**Introduction:** Patients requiring long term parenteral nutrition for intestinal failure are at risk of developing associated liver disease (IFALD) which can progress to cirrhosis that requires liver transplantation along with the intestine. There is evidence to suggest that pre-emptive intestinal transplantation in this setting can prevent progression of liver disease and may improve fibrosis.

**Methods:** A retrospective review of liver biopsies, liver blood tests (at the time of biopsy), intestinal anatomy and outcomes in patients referred to Addenbrooke's Hospital Cambridge UK between 2008 and 2018 for consideration of transplantation for IFALD. Fibrosis was scored subjectively by pathologists as mild, moderate, severe or cirrhosis.

**Results:** 21 patients (out of a total of 82) underwent transplantation for IFALD. In 13 cases, liver fibrosis was considered too advanced for intestine alone and they required liver containing grafts, the others received intestine without liver. Post transplant survival is shown in fig. 1. In addition, 13 patients have been referred and/or listed and 9 died on the waiting list, 5 of which from advanced liver disease. Liver biopsies which showed at least moderate fibrosis (in the absence of additional pathology) from patients with >20cms intestine to stoma were performed at a mean of 11.6 years (SD 7yrs) after starting PN (n=15), those with <20cms to stoma were performed at a mean of 2.2 years (SD 0.6yrs) (n=10). LFTs in patients (excluding end stage disease and severe cholestasis) were calculated. Mean bilirubin and ALT in patients with cirrhosis or severe fibrosis were 30 umols/l (+/-11) and 52 U/l (+/- 34). Only 12 out of 24 patients with at least moderate fibrosis on biopsy showed bilirubin and/or ALT >1.5x ULN.

**Conclusions:**

Patients with ultrashort intestine are at risk of early progression to significant degrees of IFALD.

Liver blood tests are not predictive of underlying IFALD.

Post-transplant survival in patients with IFALD requiring liver transplantation is inferior to that of those receiving intestine alone

Earlier detection of IFALD and referral for pre-emptive intestinal transplantation is indicated.

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**P4.16 - Clinical presentation and outcomes of Chronic Intestinal Pseudoobstruction in pediatric patients**

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**Introduction:** Chronic intestinal pseudoobstruction (CIPO) presents symptoms of intestinal obstruction without any lesions occluding the intestine. It is a rare and difficult disease entity not diagnosis but also treatment. The standard diagnosis and management has not yet be established enough. In this study, we aim to report clinical outcomes of pediatric intestinal pseudoobstruction and predictive factor for the poor outcomes.

**Methods:** We retrospectively reviewed the 76 patients who were diagnosed as a primary pediatric CIPO between January 1985 and December 2017 in one institute. Age at diagnosis ranged between 0 and 221 months (median 4 months). We categorized the clinical outcomes into four groups and evaluate clinical outcomes.

**Results:** A total of 75 patients underwent the operation including enterostomy, bowel resection and venting gastrostomy. In terms of histopathology, 56 patients were a neuropathy, 11 patients were a myopathy and 3 patients showed both results in specimen. In contrast, we could not abnormal finding of enteric neuron and muscle in specimen of six patients. The overall mortality of primary CIPO was 10.5% (8 of 76). Neuropathic type CIPO patients showed better prognosis than myopathic type patients in significant (p <0.008). Furthermore 41 patients and 21 patients categorized as a good and poor outcome group in respectively. The patients who experienced a home parenteral nutrition (PN) had tendency with poor clinical outcomes due to recurrent hospitalization.

**Conclusion:** In recent, the clinical outcome of CIPO improved because of an early diagnosis, proper surgical treatment and multidisciplinary management. In this study, we showed an acceptable clinical outcome even though many well-known morbidities.

**Disclosure Statement**

The authors have no conflicts of interest.

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Introduction: In the Netherlands, there is only 1 intestinal transplant (ITx) program and 2 centers for home parenteral nutrition. The number of patients as possible candidates for ITx, referred by different centers, is growing. Still, the number of ITx is small. With this overview, we want to present the case mix and complexity in adults over the last decade.

Methods: The nationwide online Dutch Registry of Intestinal Failure and Transplantation (DRIFT) was started (2013) to monitor intestinal failure and transplantation patients, which combines data from patients of the University Medical Center Groningen (UMCG) and three more Dutch centers. In this retrospective overview, we present a small but complex group of patients who were referred to or already known by the UMCG since November 2009 till now.

Results: During this last decade, 37 adult patients were referred to our nutritional support team for possible screening regarding ITx. After a first interview at the UMCG, 14 patients were not screened because enteral feeding or autologous reconstruction was possible. Others lacked condition or feared decreasing quality of life after transplantation. 23 patients were screened, of which ten patients were rejected after screening because of medical or psychological reasons. Eventually, only ten adult patients underwent ITx (of which 1 multivisceral transplantation, 2 ITx with abdominal wall, 2 ITx with kidney transplantation) and three patients are still on the waiting list. At the time of intestinal failure associated liver disease (IFALD) and severe venous access problems.

Conclusion: Over the last decade, a nationwide multidisciplinary approach of intestinal failure has proven to be successful so that only a limited amount of patients needed a transplant. However, the complexity in ITx candidates is increasing with more severe venous access problems and IFALD.

P4.18 - Strategies Developed to Support Families Facing Intestinal Transplantation

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Introduction: A child/young person facing an Intestinal Transplant can be extremely difficult for families. It involves meeting a new team and hospital at a time when medical care of the child is challenging. An MDT approach has been demonstrated to benefit the family and help ensure they are adherent to treatment and care. Working in partnership is essential to ensure children and young people receive the best care. Our aim is to describe the strategies we have developed to meet the needs of our intestinal transplant families, children, and young people.

Methods: Introducing families to our transplant MDT at the time of transplant assessment, ensuring they understand team members' roles and responsibilities.

Giving families regular opportunities to meet with different members of the transplant team to gather information and ask questions. Provide families with information both written and verbal to empower and encourage them to participate in the discussions with healthcare professionals. Daily communication with the family is done in their own language via interpreters.

Establish links with the local support network early and ensure they are kept up to date with treatment and care plans. E.g., Shared care consultant, community nursing teams.

Support the parents to communicate with the nursery/school as appropriate. Discuss return to school plans and educational care plans to ensure the child/young person can resume their education.

Behavioural contracts to establish families understand their roles and responsibilities.

Signpost families to support services e.g., Children’s Liver Disease Foundation, Multi Organ Transplant Support.

Family support team advice parents/carers on welfare benefits, housing issues and financial support in the form of grants available.

Investigate and discuss opportunities for respite and other support services available in the local area.

Summary: Feedback from families is that they feel part of the team looking after their child. They are able to confidently voice their opinions and concerns. Fostering a culture of parent and healthcare professionals working together is required to ensure children and young people’s needs are met.

Future Development: Further development of unit website, links for families to use prior to admission to orientate them to the unit and staff and development of apps.
P4.19 - Incidence and severity of acute cellular rejection in the recipients of small bowel transplantation in Taiwan
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Introduction: Intestinal transplantation is considered as the ultimate life-saving treatment for the patients with irreversible end-stage intestinal failure. The 5-year survival rate is currently around 60% according to international results. Post-transplantation episodes of acute cellular rejection, systemic infection, and, in later stages, chronic rejection are the major prognostic factors for the survival of recipients and grafts. In this research we analyzed the histological findings and the outcomes among the 22 cases of isolated intestinal transplantation in our institute in Taiwan.

Methods: Patients received intestinal transplantation were under periodical graft surveillance by intestinal biopsy through ileostomy. The biopsy histology reports from the 22 cases of intestinal transplantation were retrospectively reviewed and analyzed by IBM SPSS software. The use of data was approved and under the supervision of Institution Review Board (IRB) of Far Eastern Memorial Hospital, New Taipei City, Taiwan.

Results: The 1, 3, and 5-year patient survival rates of intestinal transplant recipients in our institute are 83.7%, 72.2%, and 66.7%, respectively. During year 2007 to 2017, the total amount of biopsies taken from these 22 cases is 904. In these biopsy reports, 734 biopsies (80.9%) were reported as indeterminate for acute cellular rejection (ACR), 39 biopsies (4.3%) as mild ACR, 18 biopsies (2.0%) as moderate ACR, and 103 biopsies (11.4%) as severe ACR (Figure 1). Among the 22 cases, 15 cases (68.2%) had been diagnosed as ACR, in which 6 were reported as mild rejection, whereas the other 9 as moderate to severe rejection. Furthermore, 10 cases within these 15 ACR cases (66.7%) developed ACR in the first 3 months after transplantation.

Conclusion: In our series of small bowel transplantation, the incidence of acute cellular rejection was 68.2%, which is comparable with other American and European centers. Scheduled graft surveillance with histological inspection greatly helped the monitoring of rejection episodes and the subsequent timely treatments for the patients.

P4.20 - Ten years trends in intestinal transplantation in the United Kingdom
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On behalf of the UK national intestinal transplant programmes

Introduction: Intestinal transplantation in the UK has been centrally commissioned at four centres since 2008; two paediatric centres (Birmingham Children’s Hospital and King’s College Hospital) and two adult centres (Cambridge Addenbrooke’s Hospital and Oxford Churchill Hospital). The programme has evolved; this study describes the trends over the last 10 years. Mandatory data are collected centrally by National Health Service Blood and Transplant (NHSBT).

Methods: All patients who were registered for or received an intestinal transplant. These were classified as: bowel only (small bowel +/- colon/ abdominal wall/ kidney, BO), multi-visceral (liver, small bowel, pancreas +/- stomach/ kidney/ colon, MV), or modified multi-visceral (small bowel, pancreas +/- stomach/ kidney/ colon/ abdominal wall, MMV)) between 1April 2008 and 31Mar 2018 were extracted from the UK Transplant Registry held by NHSBT. Trends in age, diagnosis, transplant type and survival post-transplant were analysed.

Results: During the decade, 201 intestinal transplants were performed nationally with an annual increase from 16 in 2008/09 to 26 in 2017/18. Since 2011/12 more transplants have been performed in adults than children, with 65% in adults during the latest year compared with 38% during 2008/09. BO transplants represented 72% of all transplants performed in 2010/11 compared with 27% in the latest year, due to increases in both MV and MMV transplants.

There were 251 registrations onto the transplant list, of which 45% were for short-bowel syndrome, 15% motility disorders, 9% tumour, 9% re-grafts, 6% mesenteric thrombosis, 5% intestinal failure associated liver disease, 4% primary mucosal disorders and 5% other identified causes. Short-bowel syndrome has been reported less in recent years.

Of patients placed on the waiting list, 78% were transplanted. One year post-transplant patient survival rates are 78% (69%-85%) and 85% (73%-92%) for adults and children respectively.

Conclusion: The UK programme transplants 14-26 patients per year, with a trend over time towards adult patients, multi-visceral transplants and away from short-bowel syndrome.
**Introduction:** Experimental intestinal transplantation (ITx) in rats is a widely used tool in the study of ischemia reperfusion injury and rejection. Even so, live donors are used in the majority of these protocols. With the aim of generate a model closer to the clinical reality, our study intends to introduce brain death (BD) protocol to analyze the impact of biological processes associated with ITx.

**Methods:** Wistar rats between 280 - 330g were used, divided into two groups, Brain Death (BD n=5) and Non BD (n=3). Anesthetic induction was with isoflurane at 5%, maintenance was initially with a mask and then endotracheal 2/2.5% with AV (50 RPM - 2.5 TV). Arterial and venous lines are placed for blood pressure and fluid therapy manage. In the BD group, through a skull trepanation, a 4F balloon cannula is subdurally placed, and insufflated at 1ml/h flow rate. When Cushing reflex is observed, BD diagnosis is performed by the apnea test. The rat remains under BD for 2 hours, handling the MAP between 70 and 140 mmhg. NonBD group is ventilated for 2 hours. Afterwards, the graft is flushed with cold saline, harvested and preserved in HTK (Custodiol®) at 4°C. Then, sampled at 0, 4, 8, 12 and 24hs for H&E staining. For histological damage was used Park - Chiu scale. Using ImageJ® morphometric variables were measured. 207 V/C units in 5 individuals of BD group and 169 (3 individuals) for NonBD at 0, 4 and 8hs, Villi and Crypt height and width (VH, VW, CH and CW), with a minimum 10 and maximum 20 V/C units for each sample. In addition, the villus/crypt index (VCI) was calculated.

**Results:** A minor injury in the Non BD group was observed: 0hs NonBD=0.33 (±0.58) vs BD=0.8 (±0.84); 8hs NonBD= 2 (±0) vs BD=2.4 (±0.55); 12hs NonBD= 2.33 (±0.59) vs BD=3 (±12).

Higher scores of focal damage were reached faster in the BD group: BD= 4 (±2) at 8hs vs NonBD= 3.67 (±1.53) at 2hs. Morphometric values (μm) were higher under BD at 0hs for VH, VW and CH, (p<0.001). Decreasing VCI was observed in both groups at different times (BD: p<0.001; NonBD: p=0.005).

**Conclusions:** We could establish a reliable BD model in rats for the study of its impact on the intestine transplantation graft. Our study shows that BD introduced subtle histopathological differences compared to ventilated controls. Future studies will be focused in the consequences of these changes analyzing the impact on graft function after the engraftment.
P4.22 - Reporting on outcome measures in pediatric chronic intestinal failure: a systematic review

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Introduction: Standardized outcome measures provide the ability of comparing outcomes of different clinical trials. Furthermore, standardized outcome measures offer the possibility of reducing potential reporting bias and increase the reporting of patient relevant outcomes. The aim of this study is to systematically assess how definitions and outcome measures are defined in therapeutic trials of children with chronic Intestinal Failure (IF). These results will be used as first step in the development of a Core Outcome Set (COS).

Methods: MEDLINE, EMBASE, and Cochrane Central Register of Controlled Trials (CENTRAL) databases were searched from inception till August 2018. Studies were included if it was an original interventional study or observational study with a control group, concerning children from 0-18 years of age with chronic IF and concomitant parenteral nutrition use, and a definition of IF was provided. No language restriction was used. Outcomes were mapped to one of the five core areas from the OMERACT 2.0 filter. The Delphi List, Newcastle Ottawa Scale for non-randomized studies, or the quality assessment tool for before-after studies (National Heart, Lung, and Blood Institute) were used to assess the quality of included studies.

Results: A total of 1766 articles were found of which 63 studies fulfilled our inclusion criteria (figure 1). 47 studies (75%) did not report any definition of IF. Of the 16 studies (25%) which reported a definition of IF, 5 different definitions were found. A total of 106 different outcomes were reported. The four most reported outcome measures were: mortality (n=23), growth (n=22), liver enzymes (n=18), and parenteral nutrition weaning (n=18). The majority (n=63; 59%) of reported outcomes was mapped to the pathophysiological manifestations core area from the OMERACT 2.0 filter. 17 studies (27%) predefined a primary outcome of which central line related infection was the most frequently used (n=5). Quality of reporting was considered low in most studies (n=47; 75%).

Conclusion: Heterogeneity exists in the definitions of IF, and outcome reporting in research concerning pediatric chronic IF.

P4.23 - Prevalence of Vitamin D Deficiency and Response to Oral Vitamin D Supplement in Children with Short Bowel Syndrome Receiving Home Parenteral Nutrition

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Introduction: Children with short bowel syndrome (SBS) are prone to have vitamin D deficiency or insufficiency due to poor oral intake, malabsorption. In this study, we evaluated vitamin D status of pediatric patients with SBS receiving home parenteral nutrition (PN)

Methods: During two years of study period, we retrospectively reviewed pediatric patients with SBS who had been on home PN for more than six months. Vitamin D level (25-hydroxyvitamin D, 25-OHD) was checked on routine outpatient visit and categorized into normal, insufficiency, or deficiency.

Results: Eighteen patients with SBS included. There were fifteen patients with vitamin D deficiency at least once during study period, and two patients with vitamin D insufficiency. No difference in risk of vitamin D deficiency or insufficiency according to the primary cause of SBS. Followed by supplement of vitamin D, the 25-OHD level of 40% of vitamin D deficiency group had been shown improvement to insufficient status. The rest of patients received high oral doses of vitamin D yet remained in deficient status.

Conclusion: Vitamin D deficiency and insufficiency are common in children with SBS on home PN. Routine surveillance and high dose oral supplement are important for these patients. Large multicenter research is needed to establish the optimal method and dose of vitamin D supplement.
Evaluation of Clinical and Safety Outcomes After Conversion From Brand-Name To Generic Tacrolimus in Adult Intestinal Transplant Recipients

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4 Intestinal Failure Center, Policlinico S. Orsola Malpighi, University of Bologna

Introduction: The introduction of generic immunosuppressants may offer an opportunity for cost savings in intestinal and other solid organ transplantation if equivalent clinical outcomes to the branded counterparts can be achieved.

Methods: At our center 14 clinically stable adult intestinal/multivisceral transplant recipients were switched from Prograf (®) to Adoport (®) starting from 1st January 2018: 8 male and 6 female; 11 of them underwent isolated intestinal transplant, 3 of them multivisceral transplant (1 associated with abdominal wall transplant). After the immunosuppression switch we conducted an acute monitoring with regard to safety and efficacy: acute cellular rejection on intestinal biopses and tacrolimus C0 levels were checked every 3 months. Preconversion tacrolimus level was compared to those revealed at 3, 6, 9 and 12 months after conversion; at the same timepoint an intestinal endoscopy and biopsy was performed.

Results: After 1 year of follow up from the conversion, tacrolimus C0 levels did not change (preconversion mean level 5.1ng/ mL ± 1.8 vs 4.8 ng/ mL ± 17, 5ng/ mL ± 17, 5.1ng/ mL ± 19, 5.2 ng/ mL ± 19 respectively at 3, 6, 9 and 12 months after conversion; at the same timepoint an intestinal endoscopy and biopsy was performed.

Conclusion: Our clinical experience as well as research data showed that the use of generic tacrolimus results in comparable trough concentration. Given the lack of adverse events reported and the cost savings recognized, conversion from brand name tacrolimus to generic one should be encouraged also in intestinal transplant recipients.
P4.26 - Outcomes of three genetic syndromes with intestinal failure (IF), short bowel syndrome (SBS), intestinal failure associated liver disease (IFALD) and parenteral nutrition (PN) dependency, treated in our Intestinal Rehabilitation Program (IRP). Children's National Medical Center Washington DC

Clarivel Torres, Vahe Badalyan, Parvathi Mohan

Children's National Medical Center

3 y old boy with deafness, hypotonia, ichthyosis, osteopenia, IF, chronic enteropathy, initially on PN now on J tube feedings, secondary to MEDNIK syndrome. He had IFALD stage 4 fibrosis, now with normal conjugated bilirubin (CB). Liver biopsy with no iron or copper (Cu) deposition, low Cu quantification, presented with feeding intolerance, persistent diarrhea, dehydration, metabolic acidosis and AKI. Treated with high dose of zinc acetate to avoid accumulation of Cu (brain and liver). He had multiple admissions due to enterocolitis, sepsis and severe anemia related to low Cu. His initial target blood Cu level was ~20’s but it was increased to ~40 mcg/dL to minimized complications. He was started on jejunal feeds of amino-acid (aa) based formula; with control of sepsis was able to wean off of PN, now tolerating oral feeds.

10 y old male with SBS, PN and G/J tube dependent due to multiple congenital atresias, TTC7A gene mutation. History of hypogammaglobinemia, bone marrow transplant and IFALD stage 4 fibrosis, now with normal CB. History of multiple surgeries to correct his intestinal (I) obstruction, left with 75 cm of bowel, a Santulli ostomy and a surgical G and J tube due to severe dysmotility. He had multiple I. strictures dilated endoscopically. He has tolerated progressive advances of J tube feedings decreasing PN needs from 100% to 35%.

3 y old male with adrenal insufficiency and IF initially on PN, now G tube dependent, secondary to congenital osmotic diarrhea related to PCSK1 mutation. His IGF-1, IGF-BP3, Prolactin, TSH, free T4, and MRI of the pituitary were normal. He had history of multiple CLABSI and diabetes Insipidus needing vasopressin during sepsis episodes; enterocolitis and severe metabolic acidosis due to B2 deficiency. His diarrhea persisted on all formulas and he was placed on PN and on a custom formula (amino acid powder, microlipids, electrolytes), but due to his low B2 this formula was changed for aa based formula via G tube, tolerating progressive advances of enteral nutrition, with PN wean. He now takes >50% of caloric needs via PO and the rest as nighttime GT feeds. Although obesity is observed in PCSK1 his weight is at the 50% and height at the 30%.

Patients with complex genetic syndromes and diverse nutritional and electrolyte needs can be successfully managed and transitioned from PN to enteral feeding as was done in our IR program, with a multidisciplinary collaborative approach.

P4.27 - Short Bowel Produced By Intestinal Ischemia Associated With Hypobaric Hypoxia At Height. National Hospital Guillermo Almenara Irigoyen Lima Peru.

EDUARDO HUAMAN 1, 2, 3, 4, SERGIO ZEGARRA 1, 2, 3, 4

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2 Intestinal Failure Unit
3 Professor of the Universidad Peruana Unión- Lima Perú
4 Department of General Surgery National Hospital Guillermo Almenara Irigoyen

Introduction: We describe a series of cases of patients with short bowel, who had intestinal ischemia due to height (hypobaric hypoxia). It has been suggested that polyglobulia and hypoxia can lead to a decrease in splanchnic oxygenation and precipitate intestinal ischemic events. The descent at sea level can reduce the risk of thrombosis and finally, after intestinal rehabilitation therapy, ensure that patients can undergo autologous gastrointestinal reconstruction (AGIR).

Patients: 3 adult male patients, with short bowel anatomical who suffered from intestinal ischemia and underwent massive intestinal resection. The patients had in common being males, adults, living in valleys above 3000 meters above sea level, presenting haematological and / or endothelial diseases with levels of polyglobulia up to 60% of hematocrit.

The patients underwent a thrombotic profile, angio-TEM and echocardiogram to determine hematological alterations, atherosclerosis of splanchnic vessels or impaired cardiac function. No abnormality was found in any of them. The hematological and coagulation values, initially altered by the anti-coagulant medication, were reversible.

Discussion: These ischemic events due to physiological changes, polyglobulia and activation of coagulation factors (factor VII), have been described in patients living above 2800 meters above sea level. We believe that patients may have presented ischemic events aggravated by endothelial damage and polyglobulia, in the territory of the mesenteric artery. Intestinal ischemia due to hypobaric hypoxia is a rare entity that usually develops in adult males who live above 3000 meters above sea level, its prognosis is good after massive bowel resection and short bowel syndrome can be resolved with intestinal rehabilitation therapy and AGIR.
Renal dysfunction after intestinal transplantation is a well recognised complication. Calcineurin inhibitors have a direct impact on the kidney with post-operative day 7 tacrolimus associated with kidney function at 3yr. Catheter related sepsis, prolonged usage of intravenous fluid, parental nutrition, diabetes and graft failure have all been associated with chronic kidney disease.

Methods: Review of first-time paediatric intestinal graft recipients at a single centre between August’09-june’18. CKD was defined as Cystatin-C (CysC)>2mg/l measured at baseline, 6mths,3yr,2yr and 5yr post-transplant. Demographic and data associated with CKD post-transplantation were collected (Tac lvs at POD 7,3,6 and 2mths). Immunosuppression protocol consisted of Basiliximab induction then maintenance with tacrolimus and prednisolone +/- sirolimus. Target tacrolimus levels were between 2.5-3µ/L in the first 3wks, 3-6µ/L 3wks -3mths, 5-8µ/L out to 5yr post transplantation. Statistical analysis was applied when appropriate, and analyses performed using SPSS.

Table 1: Cystatin C and Tacrolimus level influencing intestinal transplantation

<table>
<thead>
<tr>
<th>Tacrolimus level (median [IQR])</th>
<th>Cystatin-C (mg/l)</th>
<th>CKD(Cystatin-C &gt;1mg/l)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baseline</td>
<td>1.64 [0.8, 1.16]</td>
<td>1.01 [0.8, 1.2]</td>
</tr>
<tr>
<td>POD 7</td>
<td>3.7 [0.8, 1.18]</td>
<td>1.00 [1.1, 1.18]</td>
</tr>
<tr>
<td>6 months</td>
<td>6.7 [4.4, 8.9]</td>
<td>1.00 [1.1, 1.18]</td>
</tr>
<tr>
<td>12 months</td>
<td>6.8 [4.7, 8.6]</td>
<td>1.00 [1.1, 1.18]</td>
</tr>
</tbody>
</table>

Results: 21 paediatric patients(53% male, 43% short gut syndrome, 38% paediatric intestinal pseudo-obstruction), median age of 5 yrs(4.7,8) underwent either isolated intestinal(43%), liver and intestinal(28%) or multivisceral transplantation(29%). 38% continued with tacrolimus and steroids alone, 57% sirolimus added. 4 patients required re-transplantation at a median time from first transplantation of 33.5mths (3,8,0). 4 patients died at a median time of 9.5mths(4,8,8). CysC level of all patients at baseline was 10.308.12), with 38% of children having evidence of pre-existing CKD (CysC lvl 13 µ/l(11115)). There was significant association with CysC level at 6mths and Tacrolimus lvl at 3mths, r=-0.5 p=0.01. CysC lvl improved between 6mths and 3yr post transplant. No significant association was found between CKD at 6mths or 3yr and type of immunosuppression, prolonged use of intravenous fluids, parental nutrition, diabetes, graft failure, line sepsis or Tac lvl POD 7.

Conclusion: Renal dysfunction is already prevalent at the time of intestinal transplantation therefore continuing close monitoring of renal function and tacrolimus levels is advisable. In our cohort renal dysfunction peaked at 6mths post transplant then improved following titration of tacrolimus level. A larger prospective study is required to identify paediatric variables that effect renal function following intestinal transplantation.
P4.30 - Accelerated transition - a patient centred approach
Samantha Duncan, Victoria Amss-Smith, Lisa Sharkey
Addenbrooke’s Hospital

Introduction: Transition from paediatric to adult services should usually be conducted in a staggered manner, giving the young person time to adapt to the changing environment and healthcare professionals. Occasionally, rapid transition in an acute setting is required and we describe here an unusual case in which this occurred.

Method: A description of a challenging case that required urgent transition from paediatric to adult services in the acute clinical setting of a seriously ill child needing multivisceral transplantation.

Results: CS, a 15 year old male was admitted to a paediatric ward with life-changing diagnoses (nephrotic syndrome, recurrent pancreatitis, cholangiopathy and hypogammaglobulinaemia). Due to his age and requirement for multivisceral transplant he was cared for by multiple specialist teams across paediatric and adult services, coordinated by the paediatric WellChild nurse. Weekly MDT’s were held, with discussions about his medical status, plans for transition, social care and educational needs.

It was agreed that the young person should formally transition to the adult medical and surgical teams at the point of transplant. A document including chronology of patient journey, family perspectives and fears and anxieties, was prepared for staff in ICU/HDU/ward areas. Following his transplant aged 16 years 11 months, support was provided to the patient and family from the WellChild nurse and adult transplant specialist nurses.

Conclusions: Despite occurring under urgent circumstances and entirely in hospital, this transition process was a success. This was due to the engagement and dedication of the staff involved across many aspects of paediatric and adult care. The patient and his family remained at the centre of all discussions and plans, which at times included very difficult decision making.

MDT working encouraged development of new relationships, knowledge of roles and processes within different services.

For the paediatric team it has highlighted the need for a WellChild paediatric transition nurse.

For the adult team it has given much insight into the transition process and the need for transition expertise. Particular lessons learned included how important the emotional wellbeing of the entire family is and how the inpatient environment on the adult ward had to be adapted to accommodate the family.

Overall we “Got it right” for CS and his family.

Reference:
1 DOH document: Transition: Getting it Right for Young People (2006)

P4.31 - Cystatin c as a marker of renal function in children on home parenteral nutrition
Elena Kurteva, SH Hill, J utta Koeglmeier
Great Ormond Street Hospital

Introduction: Well known complications of children with intestinal failure (IF) on home parenteral nutrition (HPN) include sepsis, central access loss, liver disease, pulmonary embolism, metabolic disturbances and bone disease. Renal abnormalities are less understood. Estimation of glomerular filtration rate (eGFR) from serum creatinine, height and a proportionality constant (Schwartz Formula) is the current gold standard in children. Cystatin C (CysC) is freely filtered by the glomerulus, reabsorbed by tubular epithelial cells and measured in serum. The aim of this study was to investigate the incidence of renal disease (RD) comparing eGFR and CysC.

Methods: Children receiving HPN > two years were identified from a tertiary IF clinic and data recorded prospectively from 2017 to 2018. Underlying IF aetiology, duration and proportion of PN were recorded. CysC was measured and eGFR calculated in 2017 and 2018. Normal renal function was defined as CysC < 1 mg/L or eGFR > 90 ml/ min/ 1.73 m².

Results: The underlying IF diagnosis was short bowel syndrome in 15/45, enteropathy in 8/45, motility disorder in 14/45, 6/45 were post bone marrow transplantation and 2/45 had Crohn’s disease. 24/45 patients received HPN for 2 -5 years and 21/45 for 5 -10 years, 4/45 were weaned off PN, 4/45 were on total PN. In 2017 26/45 children had a normal CysC compared to 36/45 with normal eGFR, 19/45 (42%) had an abnormal CysC compared to 9/45 (20 %) with abnormal eGFR. In 2018 30/45 patients had a normal CysC compared to 40/45 with normal eGFR, 15/45 (33 %) had an abnormal CysC compared to 5/45 (11%) with abnormal eGFR.

Conclusion: RD is associated with HPN. A higher proportion of RD is diagnosed by CysC compared to eGFR (p = 0.02/2017; p = 0.006/ 2018).

References:
P4.32 - Digestive autonomy in patients with intestinal failure after small bowel transplantation

Pilar Del Pozo, Jorge Calvo, Iago Justo, Oscar Caso, Javier Martinez Caballero, Alberto Marcacuzco, Alvaro Garcia Sesma, Felix Cambra, Luis Carlos Jimenez, Carmelo Loinaz

Introduction: Home parenteral nutrition (HPN) and all the complications related to it, used to be the only option in patients with intestinal failure. Nowadays small bowel transplant (SBT) is an answer to these patients.

Methods: From December 2004 to January 2019 we have assessed 105 patients as potential candidates, the majority of them not eligible; and have performed 31 transplants in 29 patients (21 IT and 8 MVT).

Results: Short bowel syndrome (SBS) was found in 60% (18/29) of the patients. Potential candidates for SMT were malnourished, with a mean pretransplant BMI of 22.2, average weight of 61.6kg and pre albumin values around 21.7mg/dl. 64% of the patients required total HPN before the SBT and almost 86% had several HPN-related complications due to it (6% thrombosis of major venous access, 37% catheter-related infections, 28% sepsis, refractory balance alterations.

After SBT the patients in our study reached a mean BMI of 23.7, mean weight of 67.8kg and pre albumin values around 26.7mg/dl. All recipients achieved satisfactory oral nutritionat about post-transplant day 12. Almost 80% of the patients required parenteral nutrition support during the first two months post-transplantation.

SBT recipients will need some form of parenteral nutrition support for an average period of 27 months including the time before and after the SBT.

One year after undergoing transplantation, all recipients will achieve a complete digestive autonomy and only 8.3% will need fluid replacement therapy.

Conclusions: Small bowel transplantation is a life-saving procedure for patients with intestinal failure who develop complications related to parenteral nutrition, and it is the most effective treatment in terms of complete digestive autonomy acquisition for these patients.

P4.33 - Operational tolerance can be achieved after intestinal transplant: first report and mechanistic analysis

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Introduction: Transplantation tolerance is a highly sought and rarely achieved state in clinical medicine. Defined as long-term stable and acceptable graft tolerance without immunosuppression and with an otherwise competent immune system, tolerance maximizes graft survival and function and decreases organ demand without the negative side effects of immunosuppression. While there have been prior reports of tolerance after liver and kidney transplantation, we herein report and mechanistically characterize the first case of tolerance after intestinal transplantation.

Methods: Intestinal transplantation was performed using our standard method. Immunosuppression for induction was thymoglobulin and steroids and for maintenance was tacrolimus. Graft and blood lymphocytes from the patient, 8 stable intestinal recipients, and 8 healthy controls (blood only) were analyzed using flow cytometry. Recipient T cell responses to donor and third party antigens were assessed by Pleximmune (proprietary).

Results: We performed intestinal transplantation on a 14-year old male for severe pseudo-obstruction. Subsequently lost to follow up, he presented 7 years post-transplant and 3.5 years after stopping all immunosuppression with no graft problems. Biopsy showed pristine allograft histology. Given his history of GVHD, we hypothesized that central tolerance was mediated by chimerism. However, serial peripheral chimerism studies did not show donor chimerism. Thus, we speculated that he had peripheral tolerance mediated by regulatory T cells (Treg). This was confirmed by flow cytometric analysis showing that, compared to both sets of controls, the patient's blood and graft had higher levels of naïve and lower levels of effector memory CD4/CD8 as well as higher Treg percentages. Demonstrating immunocompetence, recipient T cells robustly produced IL-17, TNF-α, and IFN-γ when stimulated by PMA/IO and highly upregulated antigen-specific CD34 expression in response to CMV and EBV antigen stimulation ex vivo. Critically, donor-specific hyporesponsiveness was confirmed by 2-fold higher CD8+CD154+ T cell response to 3rd party vs. donor lymphocytes in mixed lymphocyte reaction.

Conclusion: This represents the first report of tolerance after intestinal transplantation. Mechanistic analysis demonstrates that peripheral tolerance mediated by increased Tregs and decreased effector memory cells in both graft and blood appears to play a role.
P4.34 - Long term use of Teduglutide (TED) for Type III Intestinal Failure (III-IF) in Adults Patients (pts). A single center experience.

Hector Solar, Mariana Doeyo, Gabriel Gondolesi, Adriana Crivelli
Hospital Universitario Fundacion Favaloro

Introduction
III-IF is a highly invalidating condition which requires long term parenteral nutrition (PN). The most frequent cause is post-surgical short bowel syndrome (SBS). Less than 50% of pts would be able to reach rehabilitation within 2 years. Favorable outcomes rely on, post-surgical intestinal length (PSIL), anatomy type (PSAT) and the presence of ICV. Over the last years the use of TED has changed the course of this disease, challenging the classically accepted predictors for favorable outcome.

Aim:
Report the experience using TED for Type III-IF in adults, single and first center experience in Latin America.

Material and Methods:
Data from TED treated pts from 2014 to 2018 was collected and analyze, including: Age, gender, PSIL: a) < 50 cm., b) 51-99 cm., c) > 100 cm; PSAT: 1) terminal jejunostomy, 2) jejuno-colonic anastomosis and 3) jejuno-ileo-colonic anastomosis; freedom from PN survival and comparison of outcomes to previous reports, and drug adjustments made over time, were analyzed using SPSS v20.

Results:
Twelve adult pts have been treated since 6/2014. 11 received AGIRS. Mean age: 40.5 years, 50% male; 7/12 had PSIL: a), six pts had PSAT: 3); five: 2) and only one: 1). Mean PN volume during admission was 20.37 +/- 5.72 (range: 12-28) Lt/week. All patients received standard treatment for SBS, and most of them could reduce PN volume to 12.5 +/- 4.73 (range:6-21) Lt/week. 7/12 pts are currently PN free, three of them with PSIL: a) and four with PSIL: b). Compared to the 2015 report, the percentage of pts able to achieve intestinal autonomy increased from 74 % to 83%, and from those in the PSIL a) group PN independency grew from 0% to 33%, and in the PSIL b) group went from 67% to 91%. Results are shown in table (1) and figure (1). Two pts are currently off PN, and off TED; and 1 of them has been able to sustain a successful pregnancy; 2 pts have been on every other day doses due to abdominal bloating and discomfort for 14 and 7 months respectively, sustaining weight and urine output.

Conclusions:
TED is the first gut hormone commercially available proven to enhance intestinal rehabilitation in patients with SBS and III-IF. The use of TED in this very selective initial group of patients validates previous reports performed in developed countries. It adds the concept of using it after AGIRS, allowing not only to recover intestinal sufficiency but also to reduce the time to achieve it, even with unfavorably anatomy.
Children with complex intestinal failure benefited from the expert multi-disciplinary review - A retrospective study

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Introduction: Home parenteral nutrition service was established at Birmingham Children’s Hospital since 1987. Since then, the service has grown and the unit provides a second opinion to local, national as well as international complex intestinal failure (IF) patients. There is limited literature looking into the benefits of a multi-disciplinary team (MDT) in intestinal failure. This study aims to provide some initial data.

Methods: We did a retrospective data analysis over a 4-year period from January 2015 to December 2018 of all the children who have been discussed in our monthly multi-disciplinary IF planning meeting. We analysed their background diagnosis/ reasons for referral/ main advice given by the nutritional support and intestinal failure team (NSIF team) and their outcome as of January 2019.

Results: Over this period we had given our input for 61 patients with 70 consults with a male to female ratio of 1.8:1. There were 4 non-UK referrals, 56 (80%) external UK referrals and 10 (14%) internal referrals. The median age at presentation was 5 years, ranging from 1 month to 16 years.

As of January 2019, 19% had either come off parenteral nutrition (PN) or had reduced PN. 4% had small bowel transplant. One patient died post-transplant and the other whilst awaiting transplant. We are not aware of the outcome in 42%. 31% had no change in their clinical outcome.

Conclusion: Gastroschisis is one of the most common referred diagnoses, despite necrotising enterocolitis (NEC) being the most common reason for home parenteral nutrition. This might reflect difficulties faced by the clinicians dealing with complex gastroschisis possibly with dysmotility. The most common reason for referral was feed intolerance or non-progression. 32% either came off PN or had PN reduced after MDT advice/ actioning, signifying the importance of obtaining advice from centres with IF expertise. 42% unknown outcome signifies more regular communication with the referral teams to determine the value of the meetings for the parent site.
P4.36 - Transition from pediatric to adult intestinal transplant services: a multidisciplinary collaboration for development of a successful transition program
Karin Edlund, Annette Lennerling, Anna Brantmark, Marina Jonasson, Ljungvall, Eva Karlsson, Gustaf Herlenius
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Introduction: Thanks to increased knowledge in how to treat children undergoing intestinal or multivisceral transplantation, the number of patients graduating from pediatric to adult care is expected to increase. To this date, 9 intestinal pediatric transplantations have been performed with a 5 year survival, close to 90%. Three patients have transitioned to adults. Intestinal and multivisceral transplanted are fragile patients. According to our experience they don’t mature normally. They risk pain-killer addiction and reduced psychological well-being. In addition, adolescence is a period of increased search for independence and rebellious behavior. Previous studies of young transplanted adults show that the transition process may manifest as non-adherence and lack of engagement with medical services. There are reasons to believe that these findings are highly applicable to young adults with intestinal or multivisceral graft. At Sahlgrenska University Hospital a multidisciplinary, intestinal failure center (TSC) including adult and pediatric care has been founded. This unique organization provides new opportunities to develop an adapted, person-centered transition program, aiming to ensure a successful transition.

Methods: We have used our experience from the transition process of liver transplanted adolescents and modified the program to intestinal or multivisceral transplanted patients. We have established two new roles - transition coordinators; one at the children hospital and one at the adult hospital. These two nurses are responsible for the overall transition process and collaborate closely. Once a month a multidisciplinary conference is held at TSC and the transition coordinators can report progress, discuss patient problems and update the individual transition plan. A set of screening instruments to identify symptoms, evaluate wellbeing and assess adherence has been agreed. Coordinators can report progress, discuss patient problems and update the individual transition plan. A set of screening instruments to identify symptoms, evaluate wellbeing and assess adherence has been agreed.

Results: Preliminary findings from this work is reported. The team conferences enables effective decisions and improved team work. The transition coordination provides security for the patient, the parents and the team. The screening instruments improve assessment, dialog and education in patient contact.

Conclusion: This transition program and new unique organization contribute to a higher level of adherence to both long-term medication and lifestyle changes, and thereby increase chances for improved quality of life and graft survival.

P4.37 - Extra-Intestinal Manifestations of Children with Genetically Confirmed Microvillus Inclusion Disease
Ghada Said, Oluwakemi Ogunmoye, Jutta Koegelmeier
Great Ormond Street Hospital, London, UK

Objectives: Microvillus inclusion disease (MVID): congenital disorder caused by MYO5B or STX3 mutations. Pathognomonic features are severe intractable diarrhoea and malabsorption due to intestinal brush border atrophy, accumulation of lysosomal granules and microvillus inclusions in the apical cytoplasm of enterocytes. All epithelial tissues express MYO5B but gut involvement is classically described.

Method: Two males: A, 2-year-old Pakistani and B, 12-month-old Kuwaiti with MYO5B mutation.

Results: Patient A: Neonatal secretory diarrhea with high fluid and electrolyte requirements, deteriorated on feed introduction. Duodenal histology: blunted villi, hyperplastic crypts, loss of surface epithelium, goblet cell depletion and apoptosis. Periodic acid–Schiff (PAS) and CD10 staining suggested MVID, confirmed by electron microscopy (EM). Genetics: homozygosity of MYO5B gene: c.1087C>T mutation. He was discharged on home parenteral nutrition (PN) at 3 months with ongoing high volume and sodium demands, gradually worsening diarrhea and minor transaminitis in his first year but otherwise stable liver function. Annual PN screening showed low TmP/GFR (0.89mmol/L), elevated cystatin C (1.27mg/L), biochemical evidence of renal tubular leak and radiological signs of rickets but no nephrocalcinosis. Fanconi syndrome was diagnosed. Currently on 24hr PN (250mg/ kg) with large amounts of electrolytes, phosphate, acetate with over 20 watery stools/ day. He was referred for small bowel (SB) transplantation.

Patient B: Neonatal watery diarrhea and severe acidosis with feed introduction. He was commenced on PN, tertiary referral to London at 2 months; arriving in poor nutritional status, marked conjugated hyperbilirubinemia and transaminis. SB mucosal biopsies demonstrated total villous atrophy, focally vacuolated superficial epithelium and few intraepithelial lymphocytes. PAS and CD10 staining suggested MVID confirmed by EM. Genetics: compound heterozygosity for MYO5B gene mutations; c.1576C>T; p. and c.2111del; p. variant. Liver biopsy: lobular cholestasis, hepatocyte giant cell transformation and bridging fibrosis. He currently tolerates small amount of an amino-acid based formula and 170ml/kg/day PN with 10-hours break; with stable diarrhea (6-7/ day). Due to severe liver injury and long term PN, he is being assessed for combined liver and SB transplantation.

Conclusion: MVID has variable phenotypes, with other organ involvement such as liver and kidney apart from the gut.
Introduction: In intestinal transplantation (ITx), bleeding from the intestinal tract is a rare complication and the differential diagnosis include infection, rejection, and PTLD. We report on a case series with bleeding from ectopic varices

Methods:

Case 1: A 15 year old girl (intestinal pseudo-obstruction) presented with recurrent bleeding from the stoma, 8 years after Liver ITx. Graft biopsies excluded rejection and PTLD. An upper GI endoscopy (UGIE) showed no evidence of active bleeding. Repeat UGIE 3 wks later showed a cluster of varices at the upper graft anastomosis. A TIPPS was performed which stopped the bleeding. Two mths after TIPPS, she had recurrent bleeding from the jejunal varices, despite an patent TIPPS. She developed multi-organ failure and she eventually died.

Case 2: A 8 year boy (MVID), presented with malaena & splenomegaly 4 years after isolated ITx. CT Angio revealed a patent portal vein & intra-abdominal varices in small bowel. A further UGIE revealed varices at upper graft anastomosis and were injected with thrombin. A TIPPS was performed, which stopped bleeding. He developed multi-organ failure and subsequently died.

Case 3: A 3 year old girl (gastroschisis) presented with recurrent intermittent bleeding from the stoma 1 year after Liver ITx. A initial UGIE showed no bleeding point and graft biopsies were normal. Video-capsule endoscopy done showed no varices. CT angio could not pinpoint the location of bleeding, but demonstrated occlusion of porta-caval shunt. Due to ongoing blood transfusion requirements, a further UGIE showed varices at the junction of the upper graft anastomosis. The child continues to have intermittent episodes of bleeding but the family has declined a definitive surgical option.

Discussion: The bleeding episodes in our children originated from the ectopic varices that developed at level of the upper graft anastomosis due to altered hemodynamics following portacaval shunt blockage in two liver ITxgrafts. In the child with isolated ITx, it was due to progression of liver disease and possibly because of adhesions from repeated abdominal surgeries, which then predisposes to the development of collaterals at the upper graft anastomosis.

Conclusion: Ectopic varices should be in the differential diagnosis of children presenting with bleeding episodes following ITx especially when the graft biopsies are normal.
P4.40 - Severe hypothyroidism and metabolic encephalopathy in a child receiving long-term home parenteral nutrition without selenium

Keilla Mayumi Uchoa, Heitor Pons Leite, Camila Penteado Genzani, André Ibrahim David, Maria Fernanda Camargo de Carvalho
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Introduction: Selenium is essential in the protection against oxidative stress, for optimum immune response and for thyroid hormone biosynthesis and metabolism.

We describe a case of chronic selenium deficiency with severe hypothyroidism and metabolic encephalopathy in a child with ultra-short bowel syndrome receiving long-term home parenteral nutrition (PN) without selenium supplementation.

Methods: The patient is a 4-year-old girl, born prematurely at 26 weeks of gestation, birth weight 670g. At the 13th day of life she was diagnosed with necrotizing enterocolitis and required exploratory laparotomy. There was necrosis of the small bowel and need of extensive resection, with 3 cm of small bowel and half of colon remaining. After the surgery, she was started on exclusive PN containing a fixed dose of multi-trace element solution but lacking selenium, probably due to drug shortage. After a one-year hospitalization, she was discharged and continued to receive PN at home. At 3 years old she presented with converging strabismus and regression of motor development. Initially she had inability to ambulate, which worsened until she couldn’t walk, sit and talk. At that time, she was diagnosed with decompensated hypothyroidism, requiring T3 and T4 replacement therapy.

Results: At the age of 3 years 8 months, she was referred to our intestinal rehabilitation center for investigation. On examination she had an edematous face, muscle weakness, irritability and depigmented hair. Plasma selenium levels were undetectable. The diagnosis of encephalopathy and myxedema secondary to severe selenium deficiency was made. Intravenous selenium repletion therapy was initiated (4 µg/kg/day), followed by 2 µg/kg/day as maintenance and, as selenium plasma levels normalized, the need of T3 replacement was reduced until it was discontinued and maintenance treatment with levothyroxine isolated was initiated. She was discharged from hospital to home care four months later at the age of 4. After selenium levels restored to normal, neurological signs improved progressively and growing hair was repigmented. She continues to receive physio, speech and occupational therapy and she is progressing well.

Conclusion: Chronic selenium deficiency caused severe hypothyroidism and metabolic encephalopathy in a child receiving exclusive long-term home PN without selenium. Patients on long-term PN need selenium supplementation to avoid serious clinical manifestations of deficiency.

P4.41 - Intestinal Transplantation in children, evolution of its applicability over the last 12 years in a single center.

Carolina Rumbo, Maria Ines Martinez, Dolores García Hervás, Adriana Fernández, Diego Ramisch, Gabriel Gondolesi
Hospital Universitario Fundación Favaloro, Buenos Aires, Argentina.

Introduction: Intestinal transplantation (ITx) has been accepted worldwide as a valid treatment for intestinal failure when other options for medical or surgical rehabilitation have failed and severe complications related to intestinal insufficiency have emerged.

The aim of this study is to show how the prevalence and indications for ITx evaluation and ITx in children have evolved over the last twelve years in a single Intestinal Rehabilitation and Intestinal Transplant Program in Argentina.

Material and methods: This is a retrospective review of a prospectively filled database, from March 2006 to December 2018, including every patient <18 years of age with intestinal failure that was referred to this Program. Demographic data, diagnosis, indication for evaluation and indication given according to evaluation are analyzed. Patients were divided into three periods for analysis and comparison: 1= 2006-2009; 2= 2010-2013 and 3= 2014-2018.

Results: A total number of 32 Pediatric patients with chronic IF were referred to the Unit over the studied period (41 patients in Period 1, 36 patients in period 2 and 25 patients in period 3). From the total number, 66 patients were referred for ITx evaluation and are analyzed here; 23 were female, the primary diagnoses were: intestinal atresia (12); long segment Hirschsprung’s (13); gastroschisis with atresia (9); neonatal volvulus (8); NEC (6); perinatal volvulus (4); CIPO (3); microvillous inclusion disease (2); others (3). At the time of ITx evaluation the mean age was 38 months (SD±44), the mean time on PN was 30 months (SD±37). Table 1 shows indications for ITx evaluation divided by period and Table 2 shows the treatment implemented/patient’s clinical course after the evaluation was completed.

Table 1. Main reason for ITx evaluation

<table>
<thead>
<tr>
<th>Year of ITx evaluation</th>
<th>ITx eval [n]</th>
<th>Pts referred [n]</th>
<th>CVAL*</th>
<th>IFALD**</th>
<th>Hydrotoxicolytic disorders</th>
<th>Recurrent Septis</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>2006-09</td>
<td>28 [41 %]</td>
<td>12 [43 %]</td>
<td>6 [22 %]</td>
<td>2 [22 %]</td>
<td>0 [0 %]</td>
<td>0 [0 %]</td>
<td>0 [0 %]</td>
</tr>
<tr>
<td>2010-12</td>
<td>29 [39 %]</td>
<td>17 [59 %]</td>
<td>7 [25 %]</td>
<td>2 [25 %]</td>
<td>0 [0 %]</td>
<td>0 [0 %]</td>
<td>0 [0 %]</td>
</tr>
<tr>
<td>2014-15</td>
<td>19 [25 %]</td>
<td>8 [53 %]</td>
<td>3 [20 %]</td>
<td>0 [0 %]</td>
<td>0 [0 %]</td>
<td>0 [0 %]</td>
<td>0 [0 %]</td>
</tr>
</tbody>
</table>

*CVAL central venous access loss. **IFALD intestinal failure associated liver disease.

(Χ2 [p < 0.05], χ2 [p < 0.005]
Conclusions: The number of ITx evaluations and the indication of ITx have suffered a significant decrease since 2013. The number of patients excluded from the WL due to clinical deterioration and number of deaths while on WL reflects both, late referrals and scarcity of pediatric donation. Loss of central venous accesses continues to be the main indication for ITx referral and lack of venous accesses has been the main cause for contraindication of ITx in this program.

P4.42 - Teduglutide: Intestinal rehabilitation in children, our initial experience.

Maria Inés Martínez, Carolina Rumbo, Adriana Fernández, Gabriel Eduardo Gondolesi
Hospital Universitario Fundación Favaloro, Buenos Aires, Argentina.

Background and aim: Teduglutide (TED), a synthetic analog of glucagon-like peptide 2, has been recently approved by the European Union to promote intestinal rehabilitation in pediatric patients with chronic intestinal failure secondary to short bowel syndrome. We aim to present the first series of paediatric patients treated in Argentina.

Material and methods: This is a retrospective review of a prospectively filled database, including every patient <38 years of age treated with TED at a single Intestinal Rehabilitation and Transplant Unit. Demographic data, diagnosis, nutritional status, parental support as % of the basal metabolic rate (no protein Kcal), intestinal anatomy, outcome and adverse effects are presented.

Results: From a total of 62 children with short bowel syndrome followed in the Unit, 4 patients started treatment with TED (dose 0.05 mg/ kg/d, administered subcutaneously once a day). Table 1 shows baseline data and Table 2 follow up data.

Table 2. Treatment implemented/patient’s clinical course after ITx evaluation

<table>
<thead>
<tr>
<th>Year of ITx</th>
<th>Transplanted</th>
<th>Complication due to loss of CVA*</th>
<th>Death or withdrawal of ITx ***</th>
<th>Drop out before WL ** due to improvement</th>
<th>Family refusal for WL</th>
<th>No ITx indication</th>
<th>Change of Control</th>
<th>Curiosity on WL **</th>
</tr>
</thead>
<tbody>
<tr>
<td>2018-09</td>
<td>16 #</td>
<td>3</td>
<td>4</td>
<td>2</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>2018-10</td>
<td>10</td>
<td>2</td>
<td>6</td>
<td>1</td>
<td>6</td>
<td>4</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>2019-06</td>
<td>2 #</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>3</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

* CVA: central venous accesses, ** WL: waiting list.

Conclusions: The number of ITx evaluations and the indication of ITx have suffered a significant decrease since 2013. The number of patients excluded from the WL due to clinical deterioration and number of deaths while on WL reflects both, late referrals and scarcity of pediatric donation. Loss of central venous accesses continues to be the main indication for ITx referral and lack of venous accesses has been the main cause for contraindication of ITx in this program.

Table 1. Parenteral support and patients’ data when TED* treatment was started

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Diagnosis</th>
<th>Intestinal Anatomy</th>
<th>Age when TED* started (‘years’)</th>
<th>BMI at Starting Height/ Z-score</th>
<th>PN** support +Protein Kcal + Intravenous fluids + Intravenous Nutrition</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>Atresia IAA</td>
<td>Type 3 A: 30 cm jejunum + 5 cm ileum</td>
<td>0</td>
<td>1.2</td>
<td>0%</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>Intestinal Atresia</td>
<td>Type 2 A: 15 cm jejunum</td>
<td>11.6</td>
<td>-2.2</td>
<td>80%</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>Intestinal Atresia</td>
<td>Type 2 A: 15 cm jejunum</td>
<td>12.7</td>
<td>-2.2</td>
<td>80%</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>Trauma</td>
<td>Type 1: 15 cm jejunum + 5 cm ileum</td>
<td>12.9</td>
<td>0.6</td>
<td>80%</td>
</tr>
</tbody>
</table>

* TED: Teduglutide, ** PN: Parenteral Nutrition.

Table 2. Patients data at last follow up visit and adverse events registered

<table>
<thead>
<tr>
<th>Patient</th>
<th>Week of treatment</th>
<th>PN support (Kcal/ Week)</th>
<th>BMI at Starting Height/ Z-score</th>
<th>Adverse effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>12</td>
<td>0</td>
<td>0.9</td>
<td>Upper respiratory infection, respiratory tract infections</td>
</tr>
<tr>
<td>2</td>
<td>24</td>
<td>0</td>
<td>-1.3</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>34</td>
<td>0</td>
<td>0.7</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>42</td>
<td>0</td>
<td>-0.3</td>
<td>None</td>
</tr>
</tbody>
</table>

Conclusions: TED treatment was well tolerated and has allowed, up to last follow up, achieving intestinal autonomy in half of the treated patients, and a significant PN requirement reduction in the rest. The four treated patients had been chronically PN dependent, and had failed other medical or surgical alternatives to reach intestinal autonomy.
P4.43 - Evaluation of accessibility to intestinal failure therapy and transplant, in a center from an emergent economy country

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Introduction: In Argentina the health care (HC) is provided by a multi-payer system including both, State and Private HC organizations. Chronic IF care is provided by both systems, but intestinal transplant (ITx) is only offered in the private structure. In Argentina, chronic IF was officially included among the visceral disabilities (VD) in 2011. The Federal Government grants a certificate to individuals with VD; this benefit allows them not only the full access to HC coverage including PN and ITx, but also gives them free admission to public transportation and provides them a pension. In addition, VD is not an impairment to get employment in a public or private work. In spite of the existence of a theoretical adequate access to VD certificate, there is still a lack of knowledge and education on how to obtain it, becoming a barrier, delaying or precluding its accessibility.

Aim: To analyze the accessibility to IF therapy and ITx in our center and to measure the impact of the social worker intervention

Material and methods: This is a retrospective review of a prospectively filled database with IF patients that were evaluated for ITx at our Intestinal Failure, Rehabilitation and Transplant Unit, from March 2006 to December 2018.

Patients were divided in three categories according to their HC coverage and it was recorded for each patient when IF was diagnosed and when ITx evaluation was performed:
1) Patient’s paid health insurance (PHI): a) Private insurance (PI); b) Employer and Union sponsored plans (EUSP)
2) Government coverage: (GC) a) Visceral Disability coverage (VDC) b) General government coverage (GGC)
3) Uninsured (UI)

Results: A total of 100 patients were analyzed (Figure 1): when IF was diagnosed, 58 had PHI (58% PI and 8% EUSP) and 42 patients were UI. Due to social worker intervention, when referred for ITx evaluation, all the UI patients could obtain GC (37 patients [88%] got VDC and 5 [12%] got GGC), in order to have access to IF treatment including ITx.

Conclusions: Although many of the IF patients had PHI when IF was diagnosed, a high percentage had no coverage and due to specialized social worker intervention they could obtain GC, allowing them to get appropriate treatment for their medical condition. Rare diseases require a multidisciplinary approach for the diagnosis, treatment and the suitable use of the available social resources to obtain adequate access.

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P4.44 - Transhepatic central venous catheter placement for long-term parenteral nutrition in a patient 10 years after intestinal transplantation: a case report

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Introduction: Patients with end-stage intestinal failure require long-term total parenteral nutrition (TPN) via central venous access; however, recurrent episodes of line sepsis and multiple replacements of central venous catheters may cause venous thrombosis, resulting in loss of all central venous access. Herein, we report our experience in a patient with end-stage central venous access failure 10 years after intestinal transplantation.

Case report: A 12-year-old girl with intestinal failure due to allied disorders of Hirschsprung’s disease underwent deceased intestinal transplantation. The proximal end of the intestinal graft measuring 320 cm in length was Anastomosed in an end-to-end fashion to the recipient’s duodenum and the distal end was brought through the abdominal wall as a single-barrel ileostomy (cold ischemia time, 7 h 39 min; warm ischemia time, 43 min). Venous outflow from the intestinal graft was conveyed into portal venous circulation. At the time of transplant, left internal jugular vein was the only patent central venous access and all other central veins were occluded by frequent episodes of catheter-related infection resulting in venous thrombosis. After the transplantation, she suffered multiple bouts of enteritis both from cytomegalovirus and bacterial infection and acute cellular rejection, which eventually lead to intestinal graft failure. She was forced to go back to long-term TPN. Thereafter, her left internal jugular vein got occluded and a computed tomography scan revealed her left hepatic vein to be the only patent central vein that was accessible percutaneously. We placed a Broviac catheter in the left hepatic vein via percutaneous and transhepatic route, 9 years after transplantation. The catheter was displaced accidentally 8 months later, and we safely exchanged the catheter under the same approach. At the time of this report, her oral intake is limited to fat-free liquid diet. She now weighs 27 kg and is on tacrolimus (target trough level, 2-3 ng/ml), prednisolone 5 mg QD, and mycophenolate mofetil 300 mg QD. She is currently under evaluation for intestinal retransplantation.

Conclusion: Transhepatic central venous catheter placement is a viable option in a patient with end-stage central venous access failure; however, its long-term management to prevent accidental removal has yet to be defined.
CIRTA 2019 Book of Abstracts - Posters

P4.45 - Infective Endocarditis as a complication of central venous catheters used for Home Parenteral Nutrition: experience from a national Intestinal Failure Centre

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Introduction: The management of intestinal failure (IF) requires the safe and sustained delivery of parenteral nutrition (PN) via long-term central venous catheters (CVCs). Infective Endocarditis (IE) is a recognised complication of indwelling CVCs in a number of medical conditions, for example, the incidence of IE was 483 per 100,000 person-years in haemodialysis patients, while it was 6.3 per 100,000 person-years in the general US population. Indeed, recent ESPEN guidelines on the management of acute (type 2) and chronic (type 3) IF highlight IE as a potential complication. However, there are no published data on the incidence and outcomes of IE in patients with IF.

Methods: This is an observational study conducted on all patients with type 2 or 3 IF admitted to a national IF Unit between January 2010 and December 2017. Patients with transthoracic (TTE) and/or transoesophageal (TOE) echocardiography evidence of IE were identified from a prospectively maintained IF database and hospital coded procedures. The diagnosis of IE was based upon Modified Duke’s Criteria, with positive blood cultures, together with a characteristic vegetation appearance on echocardiography, in combination with expert cardiology review. Details of organisms, heart valves involved and antibiotics prescribed were extracted from the patient notes, together with appropriate clinical outcomes.

Results: 332 of 432 patients with indwelling CVCs admitted during the study period underwent a total of 534 echocardiograms. 52 TTE and 2 TOE were performed. 2 patients were diagnosed with IE both involving native aortic valves and both visible on TTE and TOE. One patient (diagnosed with Enterococcus faecalis IE) had type 2 IF at admission from a referring hospital for ongoing treatment for IE with persistent pyrexia, splinter haemorrhages and regurgitant murmur; clinical symptoms resolved with IV Amoxicillin and Gentamicin but valve replacement was required due to severe Aortic Regurgitation. The second patient (Coagulase-negative Staphylococcus with type 3 IF) was admitted with a CRBSI, low grade pyrexia and intermittent rigors and treated with IV vancomycin and currently remains under cardiology follow up pending potential valve replacement.

Conclusions: CVC-associated IE is a very rare complication of CVCs implanted for types 2 and 3 IF. However, as noted in our case series, the potential morbidity associated with this condition means that a high index of suspicion should be maintained.

P4.46 - Pediatric Intestinal Retransplantation: Outcomes In A Referal Center

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Purpose: Intestinal retransplantation is required in more than 10% of long term survivors, especially in those who underwent isolated intestinal transplantation. However, no clear guidelines are available regarding indications, type of graft and immunological management. Our aim is to report our experience after retransplantation in children.

Material and methods: A retrospective study of pediatric intestinal retransplantation performed in our center in the last 15 years (2003-2018) was conducted.

Epidemiological and anthropometric data, number of grafts received by each patient and the type of these with or without liver inclusion, causes of graft loss and recorded DSA data were analyzed.

Results: A total of 38 patients were retransplanted (male predominance, mean age of 5.5yrs), 12 of them twice and 6 three times, totaling 42 grafts implanted. The liver was included in the graft in 22 (4 liver- intestinal-CLSB- and 18 multivisceral-MV-). Overall survival after 5yrs was similar in children undergoing transplantation compared to our general series (63 vs 73%). Regarding the type of graft, survival after 1 and 5yrs for isolated bowel and liver-including grafts was 20% and 0% vs. 68 and 61% respectively (p<0.05).

Preformed DSA were present in 6 cases (34%), 3 of them lost their second grafts. And 2 patients (5%) developed de novo DSA and lost their non-liver included grafts (5B).

Conclusions: Retransplantation in children had long term outcomes similar to the main series. Liver-including grafts showed much better survival. Preformed or the novo DSA had negative impact on graft survival that was attenuated by the presence of the liver. Further studies are warranted to provide better understanding of the role DSA on these patients.

LIVER-INCLUDED GRAFTS

0
1

187
P4.47 - Case Report Of The First Successful Patient Of Isolated Small Intestine Transplantation At Sao Paulo University Hospital

Andre Lee 1, Flavio Galvao 1, RAFAEL PINHEIRO 1, ALICE SONG 1, LILIANA DUCATI 1, RYAN FERREL 2, RUBENS Macedo 1, WELLINGTON ANDRAUS 1, MARIANNA HOLLANDA ROCHA 1, Lucas Lee 3, Luiz Augusto Carneirio D’Albuquerque 1

1 Sao Paulo University School Of Medicine
2 Sao Paulo University School Of Medicine
3 Medical Student Of Santos-Lusiada Medical School

Objective: The case report about the longest survival of small bowel transplantation and their perioperative complications

Material and method: Male, 22 years, which began in childhood due to a pyloric hipertrophic estenosis and subsequently evolves with intestinal occlusion leading to several procedures in the correction of intestinal fistulas and adherences, resulting in six surgical procedures and ultrashort bowel syndrome by several intestinal resections. The patient was submitted at first to parenteral nutrition (PN), in second time to intestinal transit reconstruction and finally isolated intestinal transplant. The evaluation parameters were: various blood test including liver, kidney function and electrolytes level, radiological exams, ultrasound, echocardiography, vascular color doppler, endoscopy, colonoscopy and daily evaluation by multidisciplinary team

Results: The patient presented severe malnutrition and was unable to walk with Body Mass Indice (BMI)=11.18 Kg/ m2. B1 hypovitaminosis (Beriberi) characterized by cardiac disfunction, thrombosis of various central veins caused by recurrent infections and liver test show moderate cholestasis. After six months of nutritional support and three months on post operative period of intestinal reconstruction, the BMI increased to 18 Kg/ m2 and return of normal cardiac function. Then was submitted a Isolated Intestinal Transplantation on the fourth day of december/2017, presenting the following complications: kidney failure, lymphatic fistula, jejunal bleeding and pericardial effusion. But he weaned from NP completely in two months after transplant and reached independent oral feeding.

Conclusion: This case teaches us that despite several severe perioperative complications mainly malnutrition, many central line infections with thrombosis, considered initially palliative treatment, however had excellent evolution and good quality of life until this moment.
P4.48 - Evaluating psychological support services available for intestinal failure patients nationally

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Introduction: New Zealand (NZ) has a population of 4.9 million with a publicly funded health system. The NZ Health Strategy 2030 requires care to be delivered close to home. A multidisciplinary team approach for patients with intestinal failure (IF) is widely accepted and a standard of care (SOC) endorsed by NZ National Intestinal Failure Service (NIFS), this includes access to psychological support services. Clinical network (CN) feedback indicated aspects of the SOC were unable to be delivered due to variability and availability of psychological care. In order to advocate for appropriate services, it was necessary to understand the current psychological and psychiatry services for IF patients.

Method: A web based survey was developed to identify psychological services, referral pathways, appropriateness and desired levels of service provision for IF patients in NZ. The survey contained questions allocated to five categories including occupation details, current psychological care services, access to psychological care and NZ NIFS SOC. The survey was sent to 502 members of the NZ NIFS CN in August 2017. Responses were collated and analysed using Microsoft Excel to identify themes and regional variance.

Results: Forty-one clinicians from 3 of 20 District Health Boards (DHB) completed the survey representing a response rate of 8.2%. Of note, most DHBs contribute multiple staff to the CN and collated responses were sent on behalf of their DHB. Responses confirmed that there is a lack of psychological care services available nationally. This applied equally to both adult and paediatric IF services. It is particularly concerning that respondents identified mental health deficiencies as a reason that 2 of the 10 NZ NIFS SOCs could not be met. Themes highlighted include inconsistencies in availability and adequacy of services between and within DHBs. There is a lack of knowledge of access and referral pathways available for psychological care services.

Conclusion: Findings highlight a disparity in provision of psychological care services in NZ which represents a risk to the NZ Health Strategy 2030 close to home model of healthcare. Every IF patient requires appropriate psychological care as a minimum SOC. Whilst acknowledging that current services have limited capacity the NZ NIFS will continue to advocate for the provision of appropriate and equitable psychological care and engage the CN to identify local psychology and psychiatry services in their DHB.


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2 Pharmacy. Hôpital Necker Enfants Malades; Université Paris-Descartes, Paris; France
3 Social division. Hôpital Necker Enfants Malades; Université Paris-Descartes, Paris; France

Introduction: In France, the Hôpital Necker-Enfants Malades Home Parenteral Nutrition (HPN) program is the oldest (certified in 1984) and the largest with ~40% of the 380 children currently on HPN. We enrolled >800 infants and children in our program since 1984. In 2018, it supported 156 children (ages: 3mths-17yrs): short bowel syndrome (SBS) (n=78), congenital enteropathy (CE) (n=28), total intestinal aganglionosis (TIA) (n=22) or chronic intestinal pseudo-obstruction (CIPOS) (n=47); miscellaneous (n=11). We try to offer the most normal life for the child, parents & siblings: family life, school attendance, physical & cultural activities…, and vacations. In France, holidays are sacred…!! Moreover, some children have their grand-parents living outside France, especially in North Africa, the French islands (Caribbean & Indian Ocean), Turkey, Portugal…. Until recent years, holidays outside of France were like an unreachable dream. Thanks to our experience of vacations within France and with our multidisciplinary approach and mutual trust with families, we crossed progressively the French borders. Nowadays, 23 different countries have been visited by 210 families.

Methods: The HPN vacations organization involves nurses, MD, pharmacists, psychologist, social worker…. It requires time and should be anticipated at least 2 months before departure. Contacts are established with airlines companies for transporting material in the aircraft both in the cabin and in the hold. Local medical and nursing contacts are established, while medical recommendations are delivered regarding emergency situations and the management of heat weather conditions.

Results: In 2018, 20 children went to foreign countries or French Islands. IF causes: SBS (n=9), CE (n=7), TIA (n=4). Children received PN 3 to 7 nights a week, with bags from the French usual provider, completed if needed by a local hospital pharmacy (n=4). Destinations were: Morocco, Algeria, Turkey, Greece, Tunisia, Caribbeans, Portugal, Thailand….Stay in the country ranged from 1 to 8 weeks. Only one child had an harmful event with a dehydration.

Conclusion: According to the very successful and enjoyable experience for the families, we do encourage an increasing number of families to do the same whenever financially possible. We suggest our colleagues and friends in charge of a HPN program to set up a “HPN travel agency”….. for offering families great holidays and happiness.
**P4.50 - Sequential autologous bowel reconstruction for near-total intestinal aganglionosis**

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**Introduction:** Near-total Intestinal Aganglionosis (N-tIA) extending into high jejunum results in a severe short bowel state with large jejunostomy losses, and increased mortality from TPN/sepsis liver injury and loss of venous access, and life-long TPN dependence for those with <20cm jejunum. The alternatives are Autologous Gastrointestinal Reconstruction (AGIR) or Bowel Transplant. We present a female child who underwent Bowel Expansion, Sequential Autologous Bowel Lengthening with SILT (Spiral Intestinal Lengthening) followed by LILT (Longitudinal Intestinal Tailoring and Lengthening), and the addition of a prejejunostomy patch of aganglionic right colon, with an ongoing and improving outcome.

**Methods:** A 14-day old female child with N-tIA and 20cm of ganglionic residual jejunum from the ligament of Trietz, was initially treated by jejunostomy at 50cm from Treitz, retaining 30cm of aganglionic jejunum. At 3months the aganglionic jejunum was excised and the dilated proximal ganglionic 20cm were lengthened by SILT to a total of 50cm (fig1A). The stoma was closed around a 16F Pezzer catheter that was brought out onto the abdominal wall as a Tube Jejunostomy. Timed catheter clamping over 5 months induced Controlled Bowel Expansion, and was followed by LILT (fig1B) to an increased total jejunal length of 80cm. At 29months of age 30cm of opened ascending colon was patched to the side of the distalmost jejunum just proximal to a permanent jejunostomy stoma (fig2).

**Results:** At 5 years of age total TPN caloric requirement has decreased from 100% to 55%, and her Body Mass Index is 15.4 at the 50th centile for her age. Neurological function is normal for her age. Largely after the colonic patch her stoma output reduced from 130ml/kg to 30ml/kg.

**Conclusion:** Patients with N-tIA with a residual short bowel state benefit from AGIR designed to increase full thickness bowel volume and enhancing intestinal adaptation. Following controlled bowel expansion, sequential lengthening by SILT and LILT, and an ascending colon patch (endogenous GLP2?) our patient has had a 50% reduction in TPN requirement and has shown sustained physical and mental growth and an improved quality of life. We recommend management spanning several years by Sequential Autologous Gastrointestinal Reconstruction (expansion, lengthening, increased mucosal contact techniques) to enhance intestinal adaptation and absorption, and with no cut-off point for referral for Intestinal Transplantation.
**P4.51 - Follow-up of nutritional status and growth in children with intestinal failure receiving long-term parenteral nutrition**

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Samaritano Hospital

**Introduction:** Regular and frequent monitoring of the patient’s nutritional status is necessary for optimizing nutritional support in children with intestinal failure (IF). We aimed at examining the changes in anthropometric nutritional status and identifying factors associated with this outcome in children with IF followed at a pediatric intestinal rehabilitation center.

**Methods:** Prospective cohort study in 33 children with IF (8 males and 5 females), mean current age of 32.9 (IQR: 7.7) months. Patients were followed-up at an intestinal rehabilitation center between July 2015 and December 2018; all were receiving long-term parenteral nutrition (PN). The main primary cause of IF was necrotizing enterocolitis (6/13) and 7 patients had ultrashort bowel syndrome. Z scores of weight/age, height, and body mass index for age were routinely monitored at two-week intervals and compared with the WHO reference standards. Data were collected regarding the energy and protein supplied by PN and by oral/tube feeding route. Age, sex, time of follow-up, prematurity, length of the remnant small bowel, ostomy and the diagnosis of intestinal failure associated liver disease were the main explanatory variables for the outcome (Z scores of weight/age and height/age). The effect of the exposure variables on the outcomes was analyzed by generalized estimating equations.

**Results:** The median time of follow up was 16.5 months, with interquartile range (IQR) of 8.9 to 32.4 months. The Z scores of the anthropometric parameters increased significantly during the follow-up. Median weight/age Z score increased from -3.68 (IQR: -4.97 to -2.66) to 0.45 (IQR: -2.0 to 0.44) and mean height/age Z score increased from -3.93 (IQR: -4.66 to -2.63) to -1.2 (IQR: -4.18 to -0.28). Malnutrition based on weight for age Z score decreased from 77% to 23% by the last assessment. Mean energy and protein supply by PN were 88 (23.4) kcal/kg/day and 2.3 (0.7) g/kg/day, respectively, and both decreased significantly along the follow-up period (p<0.001). Mean energy and protein supply by PN were 67.6 (SD 17.5) kcal/kg/day and 1.8 (0.5) g/kg/day respectively. The increase in the anthropometric z scores was not associated with any of the clinical and demographic factors considered in the analysis.

**Conclusion:** There was a significant improvement in the nutritional status of children during the follow-up period. This improvement seems to occur independently of demographic factors and comorbidities.

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**P4.52 - Preventing catheter-associated infections in children with intestinal failure receiving long-term parenteral nutrition in a pediatric gastroenterology unit, 2010-2018: impact of an educational program surveying policies for insertion and care of central venous catheters**

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2 Pediatric gastro-enterology and nutrition, Hospices civils de Lyon, France
3 University Lyon 1 France

**Introduction:** Children hospitalized in gastroenterology units for parenteral nutrition are exposed to central venous catheters (CVC) and the occurrence of infection on CVC. These infections are a major cause of morbidity and mortality for these patients. Prevention methods aiming education of the nursing team and optimization of intravascular medical devices are implemented. The aim of this study is to assess incidence trend and to observe influence of these methods to improve health care quality.

**Methods:** A prospective surveillance study was performed in the pediatric gastroenterology and artificial nutrition unit of Lyon university hospital. Since 2010, any child hospitalized in the ward with a CVC for parenteral nutrition was included. The data were collected by research of bacteriological results in blood culture and discussed with the clinicians. The number of catheter days was counted by the clinicians by number of parenteral nutrition bags administered in the clinical ward. The incidence of blood-stream infections was expressed by 1000 catheter-days.

**Results:** The surveillance conducted since 2010 shows a constant decrease of catheter-related blood-stream infections (CRBSI) since 2012. The annual infection rate was initially superior to 20/1000 catheter days. This high incidence was following the move of the clinical unit to a new pediatric university hospital and the high turn-over of the nursing team. From 2012 to 2016, the incidence decreased constantly between 20 and 10 CRBSI/1000 catheter days. Since 2017, the incidence is inferior to 5 infections for 1000 catheter days.

Several measures of quality care improvement were implemented successively: the revision of protocols, numerous training workshops of care practices, and audits were performed. The newly recruited and interim nurses without training were not allowed to manipulate the central catheters. The catheter material was optimized by the implementation of new perfusion lines adapted to the pediatric context.

**Conclusions:** The surveillance of bacteremia highlights an important decrease of CRBSI in children hospitalized in gastroenterology for parenteral nutrition. The improvement of quality of care seems to be linked to this decrease: reorganization of nursing care, training workshops for perfusion line connection and manipulation, improvement of care protocols and perfusion material optimization.
P4.53 - Unusual indications for multivisceral transplantation

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Introduction: Recent improvements in intestinal rehabilitation programs allowed avoiding intestinal transplantation in many cases. Conversely, improved outcomes prompt the use of intestinal transplantation for complex intestinal pathologies not included in the classical list of indications. Our aim was to analyze our experience in some of these unusual indications for transplantation and to review their outcome when compared to the usual indications.

Methods: Retrospective review of our historical series of intestinal and multivisceral transplants (1999-2018) identifying those cases in which the initial diagnosis was not among the classic indications (short gut syndrome, motility disorders, untreatable diarrheas). Their general outcome (complications, immunosuppression, graft loss, survival) were compared with the rest of the cases, considering statistically significant a P value under 0.05.

Results: We reviewed 107 transplants, finding 9 unusual cases transplanted with an average age of 4 years (1-10). Their initial diagnoses were: 3 benign cystic retroperitoneal teratomas with SMA lesion during surgery, 1 myofibroblastic tumor involving main vessels, 1 multifocal postext IV hepatoblastoma with tumoral portal thrombus reaching beyond the splenic mesenteric junction, 1 vascular accident in an appendectomy, 1 Martinez-Frias syndrome, 1 mitochondrial disease and 1 Alagille syndrome with ileal atresia. Eight patients received a multivisceral graft and 1 an isolated small bowel. Surgical and infectious complications did not shown statistical differences with our main series. PTLD was only seen in 11% (1) and GVHD in 22% (2), being less common than in the main series but not statistically significant. The rate of rejection and retransplantation was also similar. Only the case who had an isolated small bowel graft presented rejection (11%) requiring multivisceral retransplantation. Overall patient survival was 82% with a mean follow-up of 7 years (0-13), with no differences between the groups. No tumor recurrence was observed.

Conclusion: Multivisceral transplant is a safe therapeutic option in unconventional and challenging cases in which the integrity of the intestine is affected. According to our study, these transplants present the same prognosis as the classic indications. Emerging indications, like the 9 cases reported in this work, would appeared in the future. Intestinal transplantation should be at least considered to treat these patients attended in IRUs.

P4.54 - An old technique for a new problem: Bishop Koop stoma to facilitate endoscopic surveillance

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Introduction: Endoscopic surveillance of the transplanted intestine is essential for early diagnosis and successful management of graft rejection. End ileostomies are easy to survey, but patients often have high output leading to renal impairment and electrolyte disturbance. Increasingly colon inclusion has become standard of care but endoscopy via an end colostomy can be difficult and lead to failure to access the terminal ileum. In addition a standard loop ileostomy proximal to a transplanted colon does not take advantage of the fluid balance advantages conferred by colon inclusion. In 2017, we started to use a Bishop Koop anastomosis proximal to an anastomosis between transplanted and native colon.

Methods: This is a retrospective review of a prospectively held database of all intestinal-containing grafts from 2017.

Results: Since 2017, 10/20 transplants have included a Bishop-Koop stoma. In the 10 patients who did not have Bishop Koop stoma this was as a consequence of either concerns regarding the native colon at transplant (2 patients) or recipient pathology that rendered distal continuity inappropriate or technically impossible (pan proctocolectomy, ileal pouch).

Endoscopic surveillance was quicker and more comfortable for patients and ileal visualisation was possible in all cases. Two patients have subsequently undergone stoma reversal and this has proven technically easier than with our previous stoma.

Conclusion: We describe the use of a Bishop Koop stoma for patients undergoing intestinal transplantation. This has resulted in improved endoscopic surveillance, fluid balance and patient satisfaction compared to our previous use of end colostomies. Stoma reversal with the Bishop Koop is a simpler undertaking.
P4.55 - Results from a single institution pediatric intestinal transplant database: Pertinent clinical data absent from national databases

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Division of Transplant Surgery, Intestinal Rehabilitation and Transplant Program, Ann & Robert H. Lurie Children’s Hospital of Chicago
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Introduction: The United Network for Organ Sharing (UNOS) collects data before and after intestinal transplantation (ITx) from all transplant centers across the United States. While much of the information is useful, it fails to capture other data that is important for medical and surgical providers but also for patients and families involved in the care of ITx patients. We present the results of a major intestinal rehabilitation (IRP) and ITx program, focusing on data that is currently not captured by the UNOS database.

Methods: A retrospective review of all ITx (both isolated small bowel and multivisceral) performed at a regional referral pediatric IRP and ITx program between 2004-2019 was performed (n=18). Clinical, surgical, and outcomes data were collected. This study was approved by the Institutional Review Board (IRB #2019-2474). Solely data this is currently not collected in the UNOS ITx database is presented herein.

Results: 18 transplants (3 multivisceral) were performed. The most common indication for ITx was recurrent infections (either central line associated bloodstream infections or small bowel bacterial overgrowth: 12/18, 67%), either alone or combined with loss of vascular access. Intestinal failure associated liver disease was the indication for transplant in 5 patients, including all 3 multivisceral transplants. Loss of vascular access was the indication in one patient. Median time to starting enteral feeds after transplant was 16.5 days [6-210], and 56% (10/18) were discharged on full enteral feeds. Fourteen of 18 patients (78%) were weaned off parenteral nutrition at some point after their transplant. Median time to initially stopping parenteral nutrition after transplant was 74 days [32-244]. Most patients underwent stoma closure (15/18, 67%), and median time to stoma closure was 297 days [122-558]. One-year survival outcomes were 89% (16/18) patients and 83% (15/18) grafts. Ultimately, 5 grafts had to be explanted. 4/5 patients who were explanted suffered from chronic rejection, which was caused by non-compliance in 2 patients.

Conclusion: National databases provide major post-transplant outcome measures. However, much pertinent clinical data that is important for ITx medical and surgical providers but also for patients and families is not collected. A better knowledge and understanding of granular pre- and post-transplant clinical and surgical data can help better counsel and prepare families before transplant.

P4.56 - Muscle and adipose measurement of pre and post-surgical intestinal transplant patients using computed tomography to compare malnutrition diagnosis using nutrition focused physical exam

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Introduction: Sarcopenia has been defined as a low appendicular skeletal muscle mass (kg/m2) two standard deviations below the mean of a young healthy reference group (Baumgartner 1998) coupled with a concomitant loss of muscle function, or severe muscle depletion. Studies have shown that sarcopenic patients undergoing surgical resection diagnosed by computed tomography (CT) prior to surgery were at increased risk for perioperative infections, increased length of stay (LOS) and higher mortality (Moissy et al 2013, Weijs 2014). Sarcopenic obesity measured by CT imaging has been correlated with poorer outcomes in post living donor liver transplantation (Itoh S, 2016). In our institution, highly trained and experienced registered dietitians (RDs) assess muscle loss as part of the nutrition focused physical exam (NFPE) using the new guidelines to diagnose malnutrition set by the Academy of Nutrition and Dietetics and the American Society of Parenteral and Enteral Nutrition (White 2012). This practice, however, has yet to be validated. CT scans are an objective measurement of assessing skeletal muscle mass, and abdominal wall and visceral adipose (Gomez-Perez 2013), and may be useful in determining skeletal muscle loss.

Methods: Our team studied the correlation between malnutrition diagnosis via NFPE and muscle and fat mass on CT scans before and after intestinal transplantation. We also looked at the correlation of CT measurements with predicting post-operative morbidity and mortality.

Results: A total of 42 subjects were included in the study consisting of 31% male and 69% female patients. Sixty percent of subjects did not have any degree of malnutrition during the pre-transplant NFPE, while 40% had moderate to severe protein calorie malnutrition. We found a significant correlation between patients diagnosed with malnutrition and those without malnutrition in subcutaneous fat CT measurements (p=0.037). We also found a significant correlation with LOS (p=0.005) and mortality (p=0.006) with patients who received multivisceral transplants versus all other types of transplant (isolated intestine, modified multivisceral, intestine and pancreas).

Conclusion: This study is the first of its kind to analyze malnutrition in the intestinal transplant population. Based on these results, it may be feasible to analyze CT measurements to assess recent patient nutrition to help identify patients with malnutrition prior to transplantation.
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P4.57 - Intestinal and multivisceral transplantation at hospital das clinicas da faculdade de medicina da universidade de Sao Paulo (HC-FMUSP) - Brazil


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HC-FMUSP is one of the world’s pioneer of isolated intestinal transplantation (IIT) performed by the living legend Masayuki Okumura (2) and currently is the public Hospital reference for Intestinal and Multivisceral Transplantation (IMT) in Brazil. In 2017, HC-FMUSP started IMT program, inspired by animal research (2-6) and financed by the Government. Here we show five cases of IMT performed at HC-FMUSP. CASE 1: A 38 years old male underwent IIT in 1968, 30 days after a massive enterectomy due to mesenteric artery thrombosis. The recipient survived for twelve days (one of the longest survival in that age) and died due to uncontrolled rejection. CASE 2: A 2 years old female underwent IIT in 1969, 34 days after a massive enterectomy due to volvulus. The patient survived for five days and died due to uncontrolled rejection. CASE 3: A 33 years old, male, with a neuroendocrine tumor in the pancreas involving mesenteric vessels and multiple liver metastasis received a multivisceral graft in 2014, survived for five days and died due to uncontrolled bleeding. CASE 4: 51 year old male presenting liver cirrhosis due to alcohol with a complex portal vein thrombosis (grade IV) received a multivisceral graft in May of 2015, survived for 30 days and died due to a liver needle biopsy that caused a progressive internal bleeding. CASE 5: A 21 year old male presenting short bowel syndrome due to mesenteric torsion received isolated intestinal transplantation in 2017. Intestinal reconstruction was performed four months after the transplantation and he is currently very well, gained nine Kg of body weight and accomplished total oral feeding autonomy. This is the longer survival of isolated intestinal transplantation in Brazil (34 months). Currently we have 15 patient referral and five patients listed for IMT, three for isolated and two for multivisceral. The main obstacle for IMT in Brazil is the lack of suitable donors (7). Conclusion: HC-FMUSP is world’s pioneer in IMT and currently have achieved long term survival in a case of intestinal transplantation. Improvements in donor care would improve this actively in Brazil.

References:


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Introduction: Gastroschisis (GS) is the most common abdominal wall defect requiring neonatal operative intervention and it represents one of the most costly congenital defects. It is associated with short and long-term morbidity associated with complications due to preterm birth, sepsis, need for small bowel resections, necrotizing enterocolitis (NEC), associated anomalies such as intestinal atresia (in 1 to 20% of patients with GS) and other birth defects. We report all patients with GS who were surgically treated or underwent intestinal rehabilitation (IR) at our institution in the past 10 years describing features, management and outcome.

Methods: We retrospectively reviewed patients with diagnosis of GS treated at our institution from July 2007 to January 2018.

Results: In this study a total of 91 patients with GS were identified (62% male). Median gestational age was 36 weeks (interquartile range [IQR]: 33-38). Sixty-four patients (71%) had uncomplicated GS. Primary surgical closure in these patients was associated with earlier intestinal autonomy and shorter duration of PN in 33% of patients (p<0.05). In this group, introduction of feeds occurred at a median age of 23 days of life (IQR: 13-35), 90% of them received breast milk with supplemental standard infant formula.

Twenty-seven patients (29%) had complex GS; their complications include: intestinal atresia (n=12), intestinal necrosis at birth requiring bowel resection (n=10), medical NEC (n=3), bacteremia (n=4) and severe intestinal dysmotility requiring promotility medications (n=3). All patients with complex GS initiated treatment by our Intestinal Rehabilitation Program (IRP) at a median age of 2.5 months. Upon initial evaluation, 88% of patients (n=24) were on parenteral nutrition (PN) that provided a mean of 97% of their daily calorie goal. 17 patients (71%) were weaned off PN after a mean of 7 months of IR. 7 patients (29%) remain PN dependent with a median bowel length of 34 cm, their mean daily energy requirement from PN is presently 48%, down from 90% at IRP enrollment.

Conclusion: Multiple prenatal and postnatal events affect outcomes in patients with GS. Patients with complex GS are at higher risk of morbidity. A specialized multidisciplinary IRP is crucial in the management of these patients to optimize their health related outcomes.
P4.59 - Pregnancy during long-term total parenteral nutrition in a patient with intestinal failure

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Introduction: There have been few reports on the use of long-term Total Parenteral Nutrition as the primary nutrition source during pregnancy from conception to birth.

Methods: A 30 year old female who suffered a volvulus resulting in intestinal failure required TPN as her primary source of nutrition. Given the extent of resections and no intestinal rehabilitation possible she required an intestinal transplant. During the evaluation process, she was found to be 5 weeks pregnant. It was decided to put the transplant evaluation on hold. She was transferred to the registered dietitian and physician for TPN management during her pregnancy.

Results: The patient's pre-pregnancy weight was 46kg with a BMI of 18 and was receiving 30 kcals/kg in 3 liters of TPN. Given that she was underweight and pregnant it was our goal for her gain the right amount of weight throughout each trimester. The guidelines for weight gain for underweight pregnant women is 28 to 40 pounds. The patient's calories were gradually increased from 35 kcals/kg to 41 kcals/kg by the end of her pregnancy. She gained 28 pounds during her pregnancy. Her fluid requirements increased to an average of 3500 liters of TPN daily with additional fluids for hydration used as needed. The patient's protein requirements throughout pregnancy averaged 2 g/kg. The patient had been on Intralipid from the time she was started on TPN but given a slight elevation in liver function tests as well as long-term TPN dependence it was decided to change her to SMOF lipid. She required an average of 16 g/kg of SMOF lipid throughout the pregnancy. Her LFT's remained stable during pregnancy with no significant increases.

Conclusion: When properly managed by a multidisciplinary team pregnant women can carry a successful full term pregnancy while dependent on long-term TPN. Labs, weight and outputs should be closely monitored during pregnancy.

P4.60 - Bortezomib for the treatment of chronic Graft Versus Host Disease in Intestinal Transplant recipient

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Introduction: GVHD is a disparate immunological response between host and recipient tissues. When it occurs after Intestinal Transplantation (ITx), it can be deadly. Bortezomib is a proteasome inhibitor that affects dendritic and T-cell processes essential for the development of GVHD but also decreases B cells. The use of Bortezomib at time of bone marrow transplant (BMT) in mice protected against acute GVHD. Bortezomib has been used in ITx for refractory acute rejection with donor specific antibodies. To our knowledge, we present the 1st use of Bortezomib for chronic GVHD in ITx.

Case: 25 yo female with Pseudo-obstruction as a result of Ehlers Danlos received a modified MVT. Induction immunosuppression (ISP) was Antithymoglobulin/ Rituximab/ Methylpred/ Basiliximab and maintenance FK/ Everolimus. Blood chimerism was followed serially. She developed a skin rash and persistent fevers on week (wk) 4 post transplant. Chimerism rose from 33% wk 4, 62% wk 5 and 80% wk 6. Endoscopy on wk 5 was negative for rectal chimerism and GHVD. Given ongoing symptoms, Alemtuzumab 30 mg IV x 2 was given for acute GVHD. Chimerism decreased to 1% wk 8 but again rose to 96% wk 11, fluctuating over the next month, 56 -96%. A 3rd dose of Alemtuzumab was given wk 15. Chimerism decreased to 2% by 5 months (mn) post transplant.

6 mn post transplant, she developed neutropenia. Given possible bone marrow GVHD, a biopsy was done and negative. She again developed rash with alopecia but managed conservatively with prednisone. Course complicated with BK viremia (5.6 million copies) treated empirically with IVIG and decrease in ISP. 9 mn post transplant, she lost 14 pounds due to high ostomy output. Work up ruled out rejection, allergy and infection except BKV pcr + in bowel biopsy. She started Cidofovir, symptoms improved and blood BKV pcr decreased. 12 mn post transplant, liver enzymes increased and biopsy ruled out GVHD. However, she again presented with skin rash despite chimerism <5%.
34 mn post transplant, Bortezomib was given weekly x 4 for chronic GVHD. Thereafter, rash resolved in 4 weeks with no recurrence now 2 years post transplant.

Discussion: GVHD continues to be a threat to ITx recipients with high mortality. Diagnosis is notoriously difficult as biopsies tend to be negative early in the course. A strong index of suspicion is necessary, before life threatening organ involvement develop, as early treatment is key to survival. Bortezomib may be an option for chronic GVHD in ITx.