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Contents

Upper Gastrointestinal SC-UG-0001 to SC-UG-0011 .......................................................... 3
Lower Gastrointestinal SC-LG-0012 to SC-LG-0024 ............................................................ 19
Young Investigator Award SC-YI-0025 to SC-YI-0034 ......................................................... 41
Thoracic SC-TH-0035 to SC-TH-0043 ...................................................................................... 59
Trauma SC-TR-0044 to SC-TR-0055 ....................................................................................... 73
Urology SC-UR-0056 to SC-UR-0067 ..................................................................................... 91
Hepato-biliary SC-HE-0068 to SC-HE-0079 ......................................................................... 109
Podium Poster Presentation SC-PP-0080 to SC-PP-0109 ..................................................... 128
General SC-GE-0110 to SC-GE-0123 ..................................................................................... 171
Basic Science SC-BS-0124 to SC-BS-0135 ......................................................................... 194
Oncology SC-ON-0136 to SC-ON-0147 .................................................................................. 217
IS ROUTINE UPPER GI CONTRAST STUDY NECESSARY PRIOR TO LAPAROSCOPIC GASTROSTOMY TUBE PLACEMENT IN CHILDREN?

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Aims of the Study: Regardless of the indication for gastrostomy tube (GT) placement, many pediatric surgeons request that children undergo a preoperative upper GI contrast study (UGI) to evaluate for the presence of either gastroesophageal reflux (GER) or intestinal malrotation. We hypothesized that routine UGI is unnecessary prior to GT placement.

Method: We performed a retrospective review of the medical records of 500 consecutive children who had a GT placed at a single children’s hospital from 2009-2012.

Results: 403 children underwent UGI prior to planned GT placement; 1 of 403 (0.25%) had a new diagnosis of malrotation on UGI that was treated with a Ladd’s procedure at the time of GT placement. 154 children had evidence of reflux on UGI, 97 of who underwent a fundoplication in conjunction with GT placement. An additional 57 children with no evidence of reflux on UGI also underwent a concurrent fundoplication. Of these 160 children who underwent concurrent fundoplication, only 3 (2%) had a confirmatory pH probe study performed prior to fundoplication. 196 children underwent preoperative UGI and GT placement during the same hospital admission. In these children the median time from UGI to GT placement was 4 days (IQR 2-8 days).

Conclusion: We found that in children undergoing routine preoperative UGI prior to GT placement 1) the risk of malrotation is less than 1%; 2) the decision to perform an anti-reflux procedure weakly correlates with the UGI findings of GER; 3) one in five patients without radiographic GER still underwent concomitant fundoplication with or without confirmatory pH probe study; and 4) the decision to obtain an UGI in hospitalized children prior to gastrostomy was associated with a delay in GT placement. Arguably, malrotation can be evaluated at the time of laparoscopic GT placement. Furthermore, the decision to proceed with concomitant fundoplication is inconsistently directed by the UGI findings. We conclude that the practice of routine UGI prior to laparoscopic gastrostomy placement in children is unnecessary and should be re-evaluated.
GASTROSTOMY PLACEMENT IN CHILDREN DELAYS GASTRIC EMPTYING. A PROSPECTIVE STUDY USING THE 13C-OCTANOIC ACID BREATH TEST.

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Aims of the Study: A laparoscopic gastrostomy placement is frequently performed in pediatric patients who require long-term enteral tube feeding. Unfortunately, postoperative complications such as leakage or feeding intolerance frequently occur. These complications may be due to gastric dysmotility. No large, prospective study on the effects of a gastrostomy on gastric emptying (GE) in children has been published to date. Therefore, the aim of this study is to evaluate the effect of a gastrostomy on GE and to identify parameters associated with failure (feeding intolerance and/or leakage) of a gastrostomy.

Method: Approval of the Medical Research Ethics Committee was acquired. A prospective study was performed including 50 children undergoing laparoscopic gastrostomy placement. Before and 3-4 months after gastrostomy, assessment was performed using 13C-octanoic acid breath tests, 24-hour pH monitoring and upper GI symptom questionnaires. Logistic regression analysis was performed in order to identify parameters associated with failure of gastrostomy.

Results: GE was significantly delayed after gastrostomy, increasing from the 57th percentile (SEM 5.5) to the 79th percentile (SEM 6.0). Severely delayed postoperative GE (P>95) occurred in 40% of patients with preoperatively normal GE. Six patients developed severe leakage at the gastrostomy site. Postoperative GE tests in these patients (n=4) showed severely delayed GE in all cases. Postoperative intolerance of gastrostomy feeding was seen in 8 (17%) patients, of whom six (75%) had severely delayed GE. A positive correlation was found between GE T1/2 and acid exposure time (percentage of time with pH<4) on pH monitoring (R=0.48; p=0.02). No correlation was found between GE T1/2 and reflux symptoms (p=0.84). No parameters could be identified as predictors of gastrostomy failure.

Conclusion: Laparoscopic gastrostomy causes a significant delay in GE. Patients with a normal preoperative GE have an almost 40% chance of developing severely delayed GE after gastrostomy. Unfortunately, failure after gastrostomy cannot be predicted with these current data. However, the results of our current study should be taken into account when a gastrostomy is considered in pediatric patients.
Upper Gastrointestinal

SC-UG-0003

IMPAIRED GROWTH AND A HIGH PREVALENCE OF DYSPHAGIA IN 12-MONTHS-OLD PATIENTS WITH ESOPHAGEAL ATRESIA

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Aims of the Study: Growth below normal, long term feeding problems and dysphagia are reported in children with esophageal atresia (EA). The reported prevalence of dysphagia in different age groups varies, and detailed information about both dysphagia and growth in EA infants are scarce. The purpose of the study was to evaluate growth and to record parents’ experience of feeding problems with special focus on presence of dysphagia in 12-months-old EA infants.

Method: 24 consecutive EA patients born between January 2012 and December 2013 were enrolled in a prospective study. At 12 months of age (corrected for prematurity), we recorded weight and height, and standard deviation scores (SDS) were calculated using Norwegian reference data. In addition, a surgeon performed a systematic parental interview on eating habits, details on swallowing and symptoms of gastroesophageal reflux. Patients were classified as having dysphagia if parents reported repeated slow or difficult passage of bolus or impaction of food in the esophagus.

Results: There were (median, range) 13 (54%) males, gestational age was 36.1 weeks (30.1-41.6), birth weight was 2630 g(1277-3780), EA type C in 21(87.5%), cardiac anomaly (ASD, VSD, tetralogy of Fallot’s) in10 (42%), and VACTERL association in 6(25%). 15 (62%) patients needed 5 (1-16) esophageal dilatations, and 2 (8%) patients had gastrostomy. Dysphagia was reported in 16 patients (67%), 3 patients (12%) coughed during liquid or bolus swallowing, and 5 patients (21%) had no problems with swallowing. Symptoms of gastroesophageal reflux were reported in 10 (42%). Median (range) SDS weight and height were -1.25 (-4.47 – 0.53) and -0.74 (-3.67- 1.00), respectively. All 7 patients with SDS height below -1.5 SDS had problems with dysphagia, and there was a significant negative correlation between reported dysphagia and SDS weight (p< 0.004) and SDS height (p<0.03). No significant correlation was found between SDS height and gestational age or number of esophageal dilatations during first year of life.

Conclusion: Two thirds of 24 one-year-old EA children had parental reported dysphagia. SDS height and weight were impaired at 12 months. There was a significant negative correlation between proxy reported dysphagia and growth.
**Upper Gastrointestinal**

SC-UG-0004

**HIGH PREVALENCE OF BARRETT’S ESOPHAGUS AND HISTOLOGICAL INFLAMMATORY CHANGES IN PATIENTS AFTER ESOPHAGEAL ATRESIA REPAIR**

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**Aims of the Study:** Esophageal atresia (EA) is a rare congenital anomaly, affecting one in 2500 to one in 4000 live births. EA can be classified according to the Gross classification: Type A: EA without trachea-esophageal fistula (TEF), type B: EA with proximal TEF, type C: EA with distal TEF (most common, about 90%) and type D: EA with proximal and distal TEF. Soon after birth EA patients undergo surgical correction. Survival rates now exceed 90% in dedicated centers. Significant gastroesophageal reflux (GER) is a frequent finding after EA repair, with a prevalence of 25-51% in childhood and 39-58% in adulthood. Due to the high prevalence of GER and increased survival in EA patients, concerns arise about an increased risk of developing Barrett’s esophagus (BE) and esophageal carcinoma in these patients. Therefore we have started to assess the prevalence of GER and BE in a cohort of EA patients.

**Method:** Single-center cohort study of all consecutive patients diagnosed with EA in our center. Since 2011, all patients (age≥17 years) with a history of EA repair are invited for gastroscopy with random biopsies at the gastroesophageal junction (GEJ) and 4-quadrant biopsies in case of BE. Clinical, endoscopic and histological data were collected.

**Results:** To date 68 patients (60.3% male) with a median age of 21.9 years (range 16.8-52.8 years) underwent an upper endoscopy. Type of EA according to Gross: type A was found in 8 (11.8%) patients, type C in 59 (86.8%) patients and type D in 1 (1.5%) patient. History of GER, confirmed by pH metry, X-esophagus or upper endoscopy in childhood, was present in 51 (75%) patients, 20 (29.4%) of whom underwent fundoplication surgery. At baseline, 22 (32.4%) patients had GER complaints and 6 (8.8%) patients used PPIs. Endoscopic findings were: BE in 18 (26.5%) patients with a circumferential extent up to 2cm and maximum extent up to 6cm, esophagitis in 4 (5.9%) patients and normal mucosa in 46 (67.6%) patients. Hiatus hernia was observed in 8 (11.8%) patients and inlet patch in 8 (11.8%) patients. Histology revealed BE without dysplasia in 7 (10.3%) patients with a median age of 31 years (range 20.9-52.8 years; 71.4% male) and gastric metaplasia in 1 patient. Inflammation was present in 2/3 of the biopsies: 33 (48.5%) chronic (30 mild, 1 moderate, 2 eosinophilic) and 14 (20.6%) active (11 mild, 1 moderate and 2 eosinophilic). Minimal reactive changes were seen in 8 (11.8%) and in only 5 (7.4%) patients inflammatory or metaplastic changes were absent. One male had developed squamous cell carcinoma of the distal esophagus at the age of 44 years. This was before endoscopic screening and surveillance of EA patients had started in our hospital.
Conclusion: EA patients have a six-fold increase in developing Barrett’s esophagus compared to the general population and at a much younger age. Two-thirds of the adult patients after EA repair have significant inflammation at the gastroesophageal junction. These findings may signify important and relevant clinical implications including use of PPI and lifelong endoscopic follow-up by experts to facilitate early diagnosis of clinically relevant lesions.
EFFECT AND EFFICACY OF LAPAROSCOPIC FUNDOPLICATION IN CHILDREN WITH GERD: THE DUTCH PROSPECTIVE, MULTICENTER STUDY

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Aims of the Study: Laparoscopic antireflux surgery (LARS) in children primarily aims to decrease (acid) reflux events and reduce reflux symptoms in children with therapy-resistant gastroesophageal reflux disease (GERD). The aim was too objectively assess the effect and efficacy of LARS in pediatric patients. Furthermore, we aimed to identify parameters associated with failure of LARS.

Method: We performed a nationwide prospective multicenter study in pediatric patients with therapy-resistant GERD to undergo LARS. From July 2011 until December 2013, 25 patients (12 males, 19 neurological normal development, median age 6 (2-18) years) were included. Before and 3-4 months after LARS clinical assessment was performed using reflux-specific questionnaires, stationary manometry, 24-hour multichannel intraluminal impedance pH monitoring (MII-pH monitoring) and a 13C-labeled Na-octanoate breath test. Linear regression was performed to identify parameters associated with failure of LARS.

Results: Neither conversions to laparotomy nor perioperative complications were observed. After LARS 3/25 (12%) patients had persisting/recurrent reflux symptoms (1/3 had persistent pathological acid exposure on MII-pH monitoring). Postoperative dysphagia was present in 8/25 patients, but resolved spontaneously within 3-4 months after LARS in all patients. 24-hour MII-pH monitoring showed a significant decrease in total acid exposure time (AET) (8.5% (6.0-16.2%) to 0.8% (0.2-2.8%), p<0.001) and in total number of reflux episodes (91 (8-230) to 14 (2-153), p<0.001), both acidic and weakly acidic. LES resting pressure significantly increased after fundoplication from 10 mmHg (7-18) to 24 mmHg (17-26), p<0.0001. Complete LES relaxation (100% (50-100) to 100% (50-100); p=0.311), peristaltic contractions (100% (80-100) to 100% (50-100); p=0.149) and gastric emptying time (77 min (0-113) to 56 min (14-103); p=0.102) did not change after LARS. Both preoperative total AET (B-coefficient=0.971; p<0.0001) and the total number of reflux episodes (B-coefficient = 0.791; p<0.0001) on MII-pH monitoring correlated negatively with postoperative reflux control (p<0.0001).
Conclusion: In children with therapy-resistant GERD, LARS significantly reduces reflux complaints, total AET and the number of (acidic) reflux episodes. LES resting pressure increases significantly after LARS, but esophageal function and gastric emptying are not affected by the procedure. Both preoperative total AET and number of reflux episodes are negatively correlated with postoperative reflux control.
**Upper Gastrointestinal**

SC-UG-0006

**SIMPLE-TRACTION OR TRACTION-AND-GROWTH: IMPACT ON EARLY POST-OPERATIVE OUTCOME IN LONG-GAP ESOPHAGEAL ATRESIA.**

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**Aims of the Study:** Many techniques have been described in order to preserve native esophagus in long-gap esophageal atresia (LGEA). Nonetheless, many surgeons are traditionally skeptical due to the risk of short-term complication and/or of failure surgery requiring redo surgery. Aim of this study is to assess the impact of esophageal simple-traction primary surgery versus preliminary traction-and-growth techniques in LGEA at 3 months follow-up.

**Method:** We enrolled all patients with LGEA, defined as a gap wider than 3 vertebral bodies (vb) and/or absence of distal TEF, treated at our Institution from January 2010 to December 2014. Traction-and-growth techniques were used to induce esophageal growth before the final esophageal anastomosis (FEA), when gap was wider than 3.5 vb. Gap was measured fluoroscopically, under tension, as previously described (Bagolan et al. Dis Esophagus, 2013). Patients were categorized based on the surgical strategy adopted: traction-and-growth group (A) and simple-traction group (B). Main outcomes considered were: first gap, gap at FEA, age at FEA and number of endoscopic dilations in the first 3 months after FEA. T-test and Mann-Whitney test were used. Statistical significance was set at p<0.05. IQR= interquartile range.

**Results:** During the study period, 27 patients with LGEA were managed at our Institution. Traction-and-growth strategy was applied in 12 patients, who underwent 1 to 4 Extra-Thoracic Esophageal Elongation (or Kimura advancements, KA) and in 1 case External Traction (or Foker Technique); four of these patients received at least one KA, but didn’t yet undergo FEA (3 waiting for other KA, 1 died due to major cardiac defects). Primary immediate or delayed anastomosis was achieved in 15 patients using simple-traction strategy; of those, major flap was used in 5 cases.
Table:

<table>
<thead>
<tr>
<th></th>
<th>Group A (12 pts)</th>
<th>Group B (15 pts)</th>
<th>p</th>
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<tbody>
<tr>
<td></td>
<td>(n. of patients)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male/Female</td>
<td>7 M / 5 F</td>
<td>13 M / 2 F</td>
<td></td>
</tr>
<tr>
<td>Gestational age (weeks)</td>
<td>median (IQR)</td>
<td>37.5 (32.5-38)</td>
<td>37 (34-39)</td>
</tr>
<tr>
<td>Birth weight (grams)</td>
<td>median (IQR)</td>
<td>2350 (1643-2915)</td>
<td>2600 (2280-3010)</td>
</tr>
<tr>
<td>Type I-II vs Type III EA</td>
<td>5 vs 7</td>
<td>9 vs 6</td>
<td>0.44</td>
</tr>
<tr>
<td>Referred patients</td>
<td>(%)</td>
<td>10 (83)</td>
<td>4 (26)</td>
</tr>
<tr>
<td>Esophagostomy</td>
<td>(%)</td>
<td>11 (92)</td>
<td>2 (13)</td>
</tr>
<tr>
<td>GAP in vertebral bodies (first measurement)</td>
<td>median (IQR)</td>
<td>4 (3.5 – 6)</td>
<td>3 (2-3.5)</td>
</tr>
<tr>
<td>GAP in vertebral bodies (at FEA)</td>
<td>median (IQR)</td>
<td>1 (1-1.85)</td>
<td>2.5 (2-3.5)</td>
</tr>
<tr>
<td>Age at FEA (days)</td>
<td>median (IQR)</td>
<td>267 (172-422)</td>
<td>65 (27-136)</td>
</tr>
<tr>
<td>Anastomotic dilations in the first 3 months after FEA</td>
<td>median (IQR)</td>
<td>0 (0-1)</td>
<td>2 (1-3)</td>
</tr>
</tbody>
</table>

Conclusion: Our data confirm that preservation of native esophagus is achievable also in LGEA and should be considered the main goal in all patients with EA, including those referred with esophagostomy and extremely LGEA. Moreover, esophageal traction and growth procedure may enhance the possibilities to succeed in direct esophageal anastomosis, although generally realized at older age, reducing early anastomotic stricture and the need for endoscopic dilations. Consequently this approach seems to be a feasible, effective and safe option in all cases of LGEA, not only as rescue strategy. Further studies are needed to confirm our preliminary data.
SURGICAL REHABILITATION TECHNIQUES IN CHILDREN WITH POOR PROGNOSIS SHORT BOWEL SYNDROME

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Aims of the Study: Intestinal failure (IF) requires multidisciplinary management based on nutritional support, surgical and medical rehabilitation and intestinal transplantation. The aim of this study is to review our experience with surgical rehabilitation techniques (SRT: Enteroplasty, Bianchi, STEP) in patients with short bowel syndrome (SBS) and poor prognosis due to complex abdominal pathology.

Method: Single-center retrospective study of all patients with IF evaluated for intestinal transplantation in the Intestinal Rehabilitation Unit who were treated with SRT. Non-parametric tests were used for statistical analysis.

Results: From 1995 to 2014, 205 patients (107 male/98 female) with mean age of 25±7 months were assessed for IF. A total of 433 laparotomies were performed on 130 patients including intestinal resection, enteroplasties, adhesiolysis, and transit reconstruction. SRT were performed in 22 patients: 12 enteroplasties, 8 STEPs and 4 Bianchi procedures; a redo procedure was performed on two patients. All patients were PN-dependent with different stages of liver disease: mild (13), moderate (5) and severe (4). The etiology of SBS was gastroschisis (9), intestinal atresia (5), enterocolitis (5), other (3). Ultrashort bowel (<25cm) was present in 29% of the patients. The adaptation rate for patients who underwent enteroplasty, STEP and Bianchi were 70%, 63% and 25% respectively, although the techniques are not comparable. Overall, intestinal adaptation was achieved in 9(41%) patients and 4(18%) showed significant reduction of PN needs. One child did not respond to SRT and did not meet transplantation criteria. The remaining patients 8 (36%) patients were included on the waiting list for transplant: 4 were transplanted, two are still on the waiting list and two died. Better outcomes were observed in milder cases of liver disease (mild 77%, moderate 40%, severe 25%)(p<0.05). Conversely a trend towards a poorer outcome was observed in cases with ultrashort bowel (p>0.05). One patient required reoperation after a Bianchi procedure due to intestinal ischemia and four needed further re-STEP or adhesiolysis procedure several months later. The median follow-up was 62 (3-135) months. Overall mortality was 19%, and was due to end-stage liver disease and/or central venous catheter related sepsis.

Conclusion: Surgical rehabilitation techniques led to intestinal adaptation in a significant number of patients with poor prognosis short bowel syndrome referred for intestinal transplant. However, SRT requires a multidisciplinary evaluation and should be attempted only in suitable cases. Careful assessment and optimal surgical timing is crucial in order to obtain a favorable outcome.
Upper Gastrointestinal
SC-UG-0008
MOST PEDIATRIC SURGEONS TREAT MUCOSAL INJURIES DURING PYLOROMYOTOMY WITH PRIMARY MUCOSAL REPAIR
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Aims of the Study: Pediatric surgeons deal with mucosal injuries in different ways. The traditional management consists of full thickness closure (FTC) with rotation of the pylorus and repeat pyloromyotomy in a different quadrant. Recently, primary mucosal repair (PMR) has become popular, but has not been accepted as the gold standard. To evaluate current treatment practices, we sent out a survey on mucosal injuries during pyloromyotomy to 650 pediatric surgeons around the world and statistically analyzed the responses.

Method: Survey questions mined data on surgeon experience, operative approach, incidence of mucosal injury, intraoperative management, and postoperative outcome. Binomial multivariate logistic regression was performed on factors associated with complications and outcome. Proportions of the repair methods in relation to demographic data and the occurrence of complications were compared using Fisher Exact test at a significance level of $p \leq 0.05$.

Results: A total of 231 mucosa injuries were included in the study. Of these, 93% were noticed intraoperatively. The incidence of complications was 6.2% and patients were reoperated in 3.8% of cases. The risk of mucosal injuries was no different between laparoscopic or open approach. Overall, two thirds of mucosal injuries were addressed by primary mucosal repair. Surgeons that had one or two mucosal injuries during their career usually performed primary repair (69%), while those with 3 or 4 perforations were more likely to employed full thickness closure and used an omental patch (46%). Despite these findings, we found a positive correlation between experienced surgeons (>20 years) and the use of PMR. Overall, surgeons that reported several mucosal injuries consistently used one type of repair over time. Most surgeons delayed feedings and kept patients in the hospital longer after a mucosal injury, regardless of the repair method. No specific correlation of the repair method and complications was found.

Conclusion: Most mucosal injuries during pyloromyotomy are managed by primary mucosal repair, regardless of whether an open or laparoscopic approach was used. As long as mucosal injuries are noticed and addressed intraoperatively by either method, postoperative complications are infrequent, and patients rarely require reoperation. Surgeons tended to be loyal to one technique throughout their career. Interestingly, almost all surveyed surgeons said they would perform primary mucosal repair in the future.
**Upper Gastrointestinal**

SC-UG-0009

PERCUTANEOUS ENDOSCOPIC GASTROSTOMY WITH T-BAR FIXATION – A RETROSPECTIVE STUDY OF POSTOPERATIVE OUTCOMES

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Aims of the Study: A 1-step endoscopic procedure for establishment of a percutaneous endoscopic gastrostomy (PEG) using a push-through technique and T-bar fixation, allows initial insertion of a balloon-retained device. Few studies have examined this technique in paediatric patients. Therefore, we have investigated postoperative complications related to this particular PEG at our department.

Method: The current study is a retrospective chart review, including patients who underwent PEG placement with T-bar fixation in the period from January 2010 to November 2014. Primary outcomes were twofold. Firstly, we recorded 30-days postoperative complications and secondly, complications after the first 30 postoperative days specifically related to the T-bars, were noted. Complications requiring treatment with systemic medications or a procedure under general anaesthesia were classified as major. Ethical approval was obtained.

Results: 87 patients were included, and the median follow up time was 2.4 years [1 month – 4.9 years]. PEG was inserted at a median age of 1.9 years [9.4 months – 16.4 years], and the median weight was 10.4 kg [5.4 – 33.0]. Operation time was mean 28 minutes [SD 10]. All patients were given prophylactic antibiotics prior to the procedure. During the first 30 postoperative days, complications were reported in 42 (48%) patients of whom 22 (25%) experienced major complications. The major complications included laparotomy (n=4) and endoscopy (n=2) for tube dislodgment, leakage and/or bleeding, peristomal infection requiring systemic antibiotics (n=11), and pneumonia (n=5). Minor complications were local infections not treated with systemic antibiotics (n=12), tube dislodgement without redo operation (n=5), minor bleedings (n=3), and granulation tissue (n=3).

After the first 30 days following PEG insertion, eleven (13%) patients had complications specifically related to the T-bars, of whom seven (8%) had major complications. These major complications were removal of subcutaneously migrated T-bars (n=4) and endoscopy for evaluation of T-bar stuck in the ventricle wall (n=1) under general anaesthesia, and systemic antibiotics for infection around the T-bars (n=2). In addition, three patients with subcutaneous T-bars causing discomfort had these removed without general anaesthesia. Lastly, one patient complained of pain and swelling related to a subcutaneous T-bar, but this healed without specific treatment.
Conclusion: Major complications were reported in 25% of patients during the first 30 postoperative days after PEG insertion. After the first postoperative month, major complications related to the T-bars were noted in 8% of the patients. Thus, PEG with T-bar fixation seems to have a high rate of major complications.
Upper Gastrointestinal
SC-UG-0010

IMPACT OF LAPAROSCOPIC FUNDOPPLICATION ON BELCHING IN PEDIATRIC GERD PATIENTS

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Aims of the Study: Laparoscopic fundoplication (LF) is a well-established treatment option for children with PPI-resistant gastroesophageal reflux disease (GERD). In addition to preventing reflux of gastric contents, LARS may also impair the ability of the stomach to vent intragastric air (i.e. gastric belching). Impaired gastric belching after LARS may lead to gas-related symptoms, such as bloating and/or flatulence, and may induce supragastric belching, an altered mechanism in which air is sucked into the esophagus and immediately expelled without entering the stomach. The aim of the study was to objectively evaluate the impact of LF on gastric belches (GBs) and supragastric belches (SGBs) in children with GERD.

Method: We performed a prospective, multicenter, nationwide cohort study including 25 patients (12 males, median age 6 (range 1-18) years). Twenty-four hour multichannel intraluminal impedance pH monitoring (MII-pH monitoring) was performed before and 3 months after LF. MII-pH-tracings were analyzed manually for reflux episodes and GBs and SGBs according to previous defined criteria (Bredenoord et al, Gut 2004). Belching (reported as a symptom part of the GERD Symptom Questionnaire (GSQ)) was also assessed (scoring 1-7 for severity and frequency).

Results: LF reduced acid exposure time from 8.5\% (6.0-16.2\%) to 0.8\% (0.2-2.8\%), p<0.001. LF decreased the number of reflux episodes from 91 (8-230) to 14 (2-153), p<0.001. The total number of GBs was significantly reduced from 59 (43-77) to 5 (2-12), p<0.001. SGBs occurred both before (16/23 patients) and after (14/23 patients) LF, with no difference in total number of SGBs (from 2 (0-7) to 2 (0-4), p=0.83). The number of patients with moderate to severe belching symptoms decreased from 12 (61\%) before to 4 (17\%), p<0.001 after LF. Postoperative belching symptom scores were associated with GBs (r=0.53, p<0.01), but not with SGBs.

Conclusion: Laparoscopic antireflux surgery significantly reduced the number of GBs in children with GERD, while the already low number of SGBs was not affected. Postoperative higher belching scores in the GSQ correlated with the number of GBs after surgery, but not with SGBs.

References: Bredenoord et al, Gut 2004
**OESOPHAGEAL STENTING IN CATASTROPHIC COMPLICATIONS AFTER OESOPHAGEAL ATRESIA REPAIR**

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¹Pediatric Surgery, Hospital General Universitario Gregorio Marañón, Madrid, ²Pediatric Surgery, Hospitales Universitarios Virgen del Rocío, Seville, Spain

**Aims of the Study:** Difficult cases of Oesophageal Atresia (OA) develop intractable complications: tight esophageal stenosis, anastomotic dehiscence and recurrent tracheo-esophageal fistula (TOF). We present a series of severe cases showing those complications that were treated by esophageal stenting.

**Method:** This is a retrospective study in which relevant clinical and demographic data are analyzed. Patients were very ill to sustain a surgical operation to treat their complications, so temporal stenting were offered. Silicon covered tracheal nonabsorbable stents were used, as there is no small esophageal stents designed for infants and newborns.

**Results:** They were 7 patients, 5 males and 2 females, mean age of 19 months of life (range 15 days to 4 years). Five of them initially had TOF and the other 2 had pure OA with no fistulae. Indications for stenting were recurrent TOF, stenosis and perforation (table). Size of the stent range from 12 to 20 mm in diameter and from 30 to 60 mm in length. Duration of placement from 15 days to 8 weeks. Further dilatation were not needed except in 1 (8 more case 3). One case died after 9 months of the procedure (respiratory failure caused by type A flu virus). Rerrecurrence was observed in two.

**Table:**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Gender</th>
<th>Diagnosis</th>
<th>Stents</th>
<th>Duration</th>
<th>Results</th>
<th>Follow up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20 mo.</td>
<td>Female</td>
<td>OA type A. Foker OE anastomoses. OE perforation + stenosis</td>
<td>12 x 30 mm</td>
<td>4 weeks</td>
<td>Good result but died of flu type A infection</td>
<td>2 years</td>
</tr>
<tr>
<td>2</td>
<td>3 mo.</td>
<td>Male</td>
<td>OA type A. Foker OE anastomoses, dehiscence &amp; OE stenosis</td>
<td>12 x 30 mm</td>
<td>4 weeks</td>
<td>Good</td>
<td>4 years</td>
</tr>
<tr>
<td>3</td>
<td>4 y.</td>
<td>Female</td>
<td>OA type C. Recurrent TOF, 4 operations</td>
<td>14 x 40 mm</td>
<td>4 weeks</td>
<td>Recurrence 2 years later</td>
<td>3 years</td>
</tr>
<tr>
<td>4</td>
<td>15 days</td>
<td>Male</td>
<td>OA type C. Recurrent TOF, dehiscence &amp; OE stenosis</td>
<td>14 x 60 mm</td>
<td>2 weeks</td>
<td>Good</td>
<td>1.5 years</td>
</tr>
<tr>
<td>5</td>
<td>12 mo.</td>
<td>Male</td>
<td>OA type C. Recurrent TOF, dehiscence &amp; OE stenosis</td>
<td>14 x 40 mm</td>
<td>3 weeks</td>
<td>Good</td>
<td>1.5 years</td>
</tr>
<tr>
<td>6</td>
<td>1 mo.</td>
<td>Male</td>
<td>OA type C. Recurrent TOF, dehiscence &amp; OE stenosis</td>
<td>14 x 40 mm</td>
<td>8 weeks</td>
<td>Good</td>
<td>3 mo</td>
</tr>
<tr>
<td>7</td>
<td>4 y.</td>
<td>Male</td>
<td>OA type C. Recurrent TOF, dehiscence &amp; OE stenosis</td>
<td>20 x 40 mm</td>
<td>8 weeks</td>
<td>Recurrence</td>
<td>3 mo</td>
</tr>
</tbody>
</table>
Conclusion: Temporally esophageal stenting after catastrophic OA complications allowed resolution of severe dehiscence and recurrent TOF. Stenosis recurred in one out of 6 patients, and new TOF recurrence in 2. These results using this simple procedure can be lifesaving but they are far from to be perfect. Stenting can play a role in managing these difficult patients.
Aims of the Study: Hirschsprung’s disease (HSCR) is a congenital defect of the enteric nervous system with a lack of ganglion cells in the distal hindgut. HSCR is a multifactorial disease caused by both genetic and environmental factors. The environmental factors, like maternal risk factors, are not well known. The aim of this study was to examine maternal risk factors and perinatal characteristics for HSCR and to study the Swedish perinatal prevalence of the disease.

Method: A Swedish nationwide, population-based, case-control cohort containing all children born in Sweden between 1/1 1982 till 31/12 2013 was used for this study. The cases were identified in the Swedish National Patient Register and data on possible maternal risk factors and patient characteristics were collected from the Swedish National Patient Register and the Swedish Medical Birth Register. Five age- and sex-matched controls were randomly selected among children without HSCR in the cohort. The association between studied risk factors and HSCR were analyzed using conditional logistic regression to calculate odds ratio (ORs) and 95% confidence intervals (CIs). Matching, stratification and multivariable regression were used to adjust for potential confounding.

Results: The study included 600 cases of HSCR (466 males) and 3000 controls (2330 males). The incidence of HSCR was stable during the years showing an incidence of 1,88/10 000 live newborns. The cases had associated malformations in 38 % of the cases; most commonly trisomy 21 and heart defects. The only maternal factor associated with increased risk for HSCR was high maternal BMI (Body Mass Index) 30,0-34,9 OR 1,89 (1,31-2,74). Neither maternal age, parity, maternal smoking nor twin-parity affected the risk for HSCR. Maternal diseases as diabetes, thyroid diseases and multiple sclerosis were not associated with an increased risk for HSCR. The perinatal characteristics showed that children with HSCR were born at a lower gestational age (OR 1,63; 95 % CI, 1,11-2,38) and no significant difference were seen in birth weight correlated for gestational age.
Conclusion: The study shows a Swedish incidence of HSCR of 1,88/10000 live newborns. Children with Hirschsprung’s disease were born at a lower gestational age than controls. Maternal obesity may increase the risk for HSCR.
Lower Gastrointestinal
SC-LG-0014

BOWEL FUNCTION IN HIRSCHSPRUNG’S DISEASE AFTER ENDORECTAL PULL-THROUGH WITH TRANSANAL MUCOSECTOMY: CONTROLLED RESULTS AFTER 4-32 YEARS OF FOLLOW-UP

Malla Neuvonen1, Kristiina Kyrklund1, Antti Koivusalo1, Risto Rintala1, Mikko Pakarinen1

1Department of Pediatric Surgery, Hospital for Children and Adolescents, University of Helsinki, Helsinki, Finland

Aims of the Study: To define bowel function outcomes following endorectal pull-through with transanal mucosectomy for Hirschsprung disease (HD) in a single institution between 1987-2011 in relation to healthy, matched controls.

Method: Patients were approached by an independent investigator and invited to answer a detailed, previously validated questionnaire on bowel function. Each patient was matched for age and gender to three controls randomly selected from the general population. Ethical approval was obtained. Social continence was defined as soiling or fecal accidents <1/week and no requirement for changes of underwear or protective aids.

Results: Of the total 92 responders (response rate 66 %), patients aged <4 years (n = 12), patients with mental retardation (n = 19) and patients with anterograde continence enema (n = 3) or enterostomy (n = 1) were excluded. Of the remaining 57 respondents [median age 15 (range, 4-32 years); 74% male], 89% of patients had classic HD, 7% had long segment disease, and single patients had total colonic disease and aganglionosis extending to ileum.

Compared to controls, patients reported higher rates of impairment for all aspects of fecal control (Table 1; P<0.05 for all). Constipation affected a similar proportion of patients (5%) as controls (4%; P=0.735). By bowel function score (BFS, max 20), 63% of patients had good functional outcome (BFS ≥17), 23% had moderate (BFS 12-16) and 11% had poor functional outcome (BFS <12). 75% of respondents versus 98% of controls were socially continent (P<0.001). Symptoms improved significantly with age; patients aged ≥12 years had less faecal incontinence, improved faecal control, and they felt less urge-symptoms to defecate compared to patients aged <12 years (P<0.05 for each). No patients aged ≥12 years had poor functional outcome (BFS-score <12) [vs 25% of patients aged <12 years (P = 0.009)]. Also a history of recurrent enterocolitis correlated with poor functional outcome; 45% of patients with a history of recurrent enterocolitis had good functional outcome (vs 74% of patients without recurrent enterocolitis (P = 0.043). There was no correlation between the functional outcome by BFS and the level of aganglionosis or operation type (P>0.80 for both).
Table:

<table>
<thead>
<tr>
<th>Variable</th>
<th>HD (n=57)</th>
<th>Controls (n=171)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean ± SD</td>
<td>%*</td>
</tr>
<tr>
<td>Sensation of urge to defecate</td>
<td>2.63 ± 0.616</td>
<td>30</td>
</tr>
<tr>
<td>Urgency period</td>
<td>2.65 ± 0.641</td>
<td>26</td>
</tr>
<tr>
<td>Frequency of defecation</td>
<td>1.46 ± 0.503</td>
<td>54</td>
</tr>
<tr>
<td>Soiling</td>
<td>2.07 ± 0.753</td>
<td>68</td>
</tr>
<tr>
<td>Faecal incontinence</td>
<td>2.53 ± 0.734</td>
<td>35</td>
</tr>
<tr>
<td>Social problems</td>
<td>2.58 ± 0.706</td>
<td>30</td>
</tr>
</tbody>
</table>

* Percentage reporting any degrees of impairment.

** P<0.05 between groups

**Conclusion:** Our results suggest that significant impairment of fecal control continues to prevail in approximately one third of patients with HD without mental disabilities after endorectal pull-through with transanal mucosectomy. Some improvement in symptoms was apparent with increasing age, but recurrent enterocolitis was associated with a reduced functional prognosis.
LONG TERM BOWEL FUNCTION AFTER ENDORECTAL PULL-THROUGH FOR RECTOSIGMOID HIRSCHSPRUNG DISEASE – PATIENT REPORTED OUTCOME

Kristin Bjørnland¹, Mikko Pakarinen², Pernilla Stenstrøm³, Kjetil Stensrud¹, Malla Neuvonen², Anna Granstrøm⁴, Christina Granli³, Ragnhild Emblem¹, Tomas Wester⁴, Risto Rintala²
¹Dep Pediatric Surgery, Oslo University Hospital, Oslo, Norway, ²Dep Pediatric Surgery, Helsinki Central University Hospital, Helsinki, Finland, ³Dep Pediatric Surgery, Skåne University Hospital, Lund, ⁴Dep Pediatric Surgery, Karolinska University Hospital, Stockholm, Sweden

Aims of the Study: Endorectal pull-through (ERPT) performed either total transanally or assisted by laparoscopy or laparotomy, is widely used for the surgical treatment of Hirschsprung disease (HD). Despite being the most popular operation for HD, patient or parent reported outcome is scarce. The aim of this multicenter international study was to report bowel function as reported by patients or parents in a large cohort of HD patients > 4 years operated for rectosigmoid HD using ERPT with transanal mucosectomy.

Method: All patients treated at four pediatric surgical centers for rectosigmoid HD operated with ERPT were eligible. Clinical data were recorded from medical records. Bowel function was assessed by a 7-item Bowel Function Score questionnaire¹. The questionnaires were either filled out by patients/parents themselves or by a nurse interviewing the patients in the clinic or by telephone. Ethical approval was obtained.

Results: Of 258 eligible patients, 55 did not respond, and three were excluded due to language problems. Results were obtained from 200 (77.5%); 157 boys and 43 girls. Median age at follow-up was 9.5 years (3.9-31.9), and 31 had syndromes (19 Downs). 67 were operated by a total transanal procedure, and 19 developed a stricture (= needing dilatations >3 months postoperatively).

73/200 (36.5%) reported no need for bowel management (defined as cleaning the colon via an appendicostomy or by rectal enemas) or stoma and to never experience fecal incontinence (FI= need for change of underwear) or constipation. 7/200 (3.5%) had a stoma, and 33 (16.5%) performed bowel management, 16 using an appendicostomy or 17 regular rectal washouts due to constipation (25) or FI (8). When asked if bowel problems affected social life, 130 answered no, 22 answered “sometimes”, and 20 reported that problems affected social life. All 31 syndromic children reported either FI, constipation, appendicostomy or stoma. Of the children with syndromes, two had stoma, four had appendicostomy and four needed regular rectal enemas. Further details of bowel function in children without stoma or appendicostomy are shown in the table below.
Table:

<table>
<thead>
<tr>
<th></th>
<th>Staining</th>
<th>FI</th>
<th>Gas problems</th>
<th>Constipation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Never</td>
<td>41 (23%)</td>
<td>92 (52%)</td>
<td>43 (24%)</td>
<td>No</td>
</tr>
<tr>
<td>&lt;1/wee k</td>
<td>65 (37%)</td>
<td>50 (28%)</td>
<td>65 (37%)</td>
<td>Diet</td>
</tr>
<tr>
<td>&gt;1/wee k</td>
<td>69 (39%)</td>
<td>32 (18%)</td>
<td>65 (37%)</td>
<td>Laxatives</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Enemas</td>
</tr>
</tbody>
</table>

Caption: Bowel function in the 177/200 HD patients excluding those having a stoma or an appendicostomy. Patients needing regular rectal enemas were given the worst score for the symptom (constipation or FI) needing enemas.

Conclusion: This large, multicenter study shows that bowel problems are common in patients operated for rectosigmoid HD with ERPT. Even though 65% reported that their bowel function did not influence their social life, only 36.5% reported no problems at all. 20% of the patients had a stoma or required appendicostomy or regular rectal washouts.

Lower Gastrointestinal

SC-LG-0016

VIDEO TEACHING PROGRAMME ON MANAGEMENT OF COLOSTOMY: EVALUATION OF ITS IMPACT ON CAREGIVERS

Heena Dabas*, Kamlesh K. Sharma², Sandeep Agarwala¹, Poonam Joshi²
¹Department of Pediatric Surgery, ²College of Nursing, All India Institute Of Medical Sciences, Delhi, India

Aims of the Study: Developing a video based learning resource material and evaluating its effectiveness in terms of knowledge and skill attainment

Method: A video teaching programme (VTP) of nine minutes duration including basic anatomy of the intestines, indications for colostomy, signs of healthy stoma, stoma cleaning and dressing, complication related to colostomy and other important concerns was developed and used to teach the caregivers about colostomy care. Pre-tested and validated knowledge questionnaire, observational checklist and stoma assessment scale (SAS) were used to assess the knowledge and skills of caregivers before and after the administration of VTP immediately (post test 1) and following 2 weeks (post test 2) after the intervention.

Results: There was significant increase in knowledge (from 10.9±2.5 to 16.4±1.67 and 15.9±4.02, p=0.001) and skill scores (from 5.6±2.0 to 9.8±1.6 and 8.6±2.1, p=0.001) immediately and 2 weeks after the video teaching programme. However, decline in skills was observed at 2 weeks when compared with immediate scores, as measured by observation checklist. Though this decrease was statistically significant (p =0.017), it may not be clinically significant. There was no significant increase in skill scores (from 4.3±0.92 to 4.7±0.67 and 4.6±0.56, p=0.40) of caregivers as measured by SAS.
Table 1: Effectiveness of video assisted teaching program on knowledge and skills of caregivers of children having colostomy.

<table>
<thead>
<tr>
<th>Scores</th>
<th>Mean ± SD</th>
<th>Overall p value</th>
<th>Post hoc comparison (mean difference) p value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Pretest vs</td>
<td>Pretest vsPost test I</td>
</tr>
<tr>
<td>Knowledge @</td>
<td></td>
<td>Post test I</td>
<td></td>
</tr>
<tr>
<td>Pretest</td>
<td>10.9 ± 2.5</td>
<td>16.4 ± 1.67</td>
<td>15.9 ± 4.02</td>
</tr>
<tr>
<td>Post test I</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Post test II</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Skill# (based on direct observation)</td>
<td>5.6 ± 2.0</td>
<td>9.8 ± 1.6</td>
<td>8.6 ± 2.1</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Skill ^ (based on SAS)</td>
<td>4.3 ± 0.92</td>
<td>4.7 ± 0.67</td>
<td>4.6 ± 0.56</td>
</tr>
<tr>
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<td></td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

Repeated measures ANOVA with Bonferroni correction, p<0.05
@Maximum score=20, #Maximum score based on direct observation=12, ^ Maximum score based on stoma assessment=5

**Conclusion:** The video teaching program was effective in bringing about an increase in knowledge and skill of caregivers of children having colostomy. So, the video can be utilized for discharge counseling for the caregivers of children with colostomy.

**References:**

**Disclosure of Interest:** H. Dabas Employee of: All India Institute Of Medical Sciences, K. K. Sharma Employee of: Working as lecturer in College of Nursing, AIIMS, S. Agarwala Employee of: working as an assistant professor, P. Joshi Employee of: Working as lecturer in College of Nursing, AIIMS
**Aims of the Study:** Peroperative search for fistula and subsequent fistulotomy is an established treatment for recurrence of perianal abscess (PA) in children but not in first time drainages. The aim was to compare recurrence rates of PA after first time surgical incision with and without search for a fistula and fistulotomy.

**Method:** This is a retrospective chart study of all children 0-16 years old referred because of PA and treated at a tertiary center for pediatric surgery during 2008-2014. Age, gender, size and location of the PA, type of first time treatment, and outcome were recorded and analyzed. The study was ethically approved by the local ethical committee (49/2010).

**Results:** In 104 patients, median age 0.4(0-16) years, 118 first time treatments of PA were performed including 28 conservative treatments, 84 surgical procedures with 49(42%) searches for fistulas followed by fistulotomy in every case where a fistula was found, resulting in 34(29%) fistulotomies. The median follow up time was 3.3 (0.4-7) years. Recurrence after first drainage of PA was more frequent if a fistula was not searched for 16/35(46%) compared to if a peroperative search and subsequent fistulotomy were performed 7/49(14%) (p=0.003). The recurrence rate after any surgical treatment, was higher among infants 0-3 months of age 12/25 (48%) than among children > 3 months 9/49 (18%) (p=0.013). In surgically treated infants of 0-3 months, searches for a fistula were less frequent: 9/25 (36%) compared to in older children 32/49 (65%) (p= 0.026). Among infants 0-3 months with surgical treatment without any search for fistula, PA recurred in 10/16(63%) whereas after incision combined with a search and subsequent fistulotomy, PA recurred in 2/9(22%) (p=0.097). There was no difference in recurrence rates if antibiotics were used in connection to surgical intervention 7/24 (29%) or not 16/60(27%) (p=0.794)

**Conclusion:** In the first time drainage of PA in children, surgery with search for a fistula and subsequent fistulotomy was followed by a lower recurrence rate of PA than after only incisions without search for a fistula. In young infants, where searches for fistulas were scarce, the recurrence rate was higher. Larger prospective studies with standardized surgical procedures are needed.
ACHIEVING A MORE ACCURATE DIAGNOSIS OF APPENDICITIS IN CHILDREN: THE ROLES OF ULTRASOUND AND COMPUTED TOMOGRAPHY

Sophie Sihui Ong 1, Wei Xiang Lim*1, Jason Barco2, York Tien Lee2, Siam Wee Sim2, Shireen Anne Nah2

1Yong Loo Lin School of Medicine, National University of Singapore, 2Paediatric Surgery, KK Women's and Children's Hospital, Singapore, Singapore

Aims of the Study: The use of computed tomography (CT) in the diagnosis or exclusion of appendicitis in children is widespread in some countries, as it demonstrates high sensitivity and specificity. However, detractors cite unnecessary exposure to radiation in children who may be more susceptible to long-term ill effects. We evaluate our use of ultrasound (US) and CT imaging in the diagnosis of appendicitis in children in our institution.

Method: This is an ethically approved review of all children admitted with the chief complaint of abdominal pain to the paediatric surgical unit between January and December 2013. We recorded patient demographics, clinical presentation, imaging investigations done, operative details, histopathology and diagnosis at discharge. Those who had US for genitourinary symptoms were excluded. As part of institutional protocol, all patients are contacted 3 days after discharge to evaluate persistent symptoms. Also, US is our preferred modality when imaging is required for assessment of abdominal pain. We assumed no missed surgical pathologies if there were no physical complaints on follow-up telephone contact. Chi-squared or Mann-Whitney U tests were used as appropriate, with significance level<0.05.

Results: Of 1359 children admitted with abdominal pain, 814 (59.9%) patients had US for assessment of right-sided and/or lower abdominal pain, of whom 155 had US features of appendicitis. There were 135 US that could not adequately delineate the appendix. Thirty-five (0.03%) patients had CT scans of the abdomen and pelvis: 14 appendicitis, 8 normal scans, 13 other intra-abdominal pathologies. Twenty-two patients had CT following US, 3 of whom had features of appendicitis, including 2 with normal appendices reported on US. A total of 233 patients were diagnosed with acute appendicitis: 166 with radiological confirmation and 67 without. One patient opted for non-operative management, leaving 232 who underwent appendicectomy. Five patients had normal appendices on histology, providing a negative appendidectomy rate of 0.02%. There were no readmissions for appendicitis in those who did not undergo appendicectomy. Those who had US were younger [median 10 years (range 0.2-21.3)] than those who had CT [11.9 years (2.8-16.5)], p=0.03. There was no difference in gender distribution or body mass index between the groups. When the appendix was visualized, US showed higher sensitivity and similar specificity to CT (Table 1 & 2).
Table:

Table 1: Diagnostic value of ultrasound (US) in appendicitis

<table>
<thead>
<tr>
<th>Ultrasound</th>
<th>Appendicitis</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Present</td>
<td>Absent</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Positive</td>
<td>True positive</td>
<td>1</td>
<td>6</td>
<td>9</td>
</tr>
<tr>
<td></td>
<td>False positive</td>
<td>2</td>
<td></td>
<td>171</td>
</tr>
<tr>
<td>Negative</td>
<td>False negative</td>
<td>4</td>
<td></td>
<td>434</td>
</tr>
<tr>
<td></td>
<td>True negative</td>
<td>434</td>
<td></td>
<td>438</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>436</td>
<td></td>
<td>609</td>
</tr>
</tbody>
</table>

Sensitivity 97.7% Specificity 99.5%

Note: Excluded from analysis were 205 scans (70 intussusception, 135 scans where the appendix could not be visualized adequately)

Table:

Table 2: Diagnostic value of computed tomography (CT) in appendicitis

<table>
<thead>
<tr>
<th>Ultrasound</th>
<th>Appendicitis</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Present</td>
<td>Absent</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Positive</td>
<td>True positive</td>
<td>13</td>
<td></td>
<td>13</td>
</tr>
<tr>
<td></td>
<td>False positive</td>
<td>0</td>
<td></td>
<td>13</td>
</tr>
<tr>
<td>Negative</td>
<td>False negative</td>
<td>2</td>
<td></td>
<td>21</td>
</tr>
<tr>
<td></td>
<td>True negative</td>
<td>19</td>
<td></td>
<td>21</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>15</td>
<td></td>
<td>34</td>
</tr>
</tbody>
</table>

Sensitivity 86.7% Specificity 100%

Note: One inconclusive CT report was excluded from analysis

Conclusion: Generous use of ultrasonography as an adjunct to clinical examination can lower negative appendicectomy rates to near zero without underdiagnosis of acute appendicitis. Only a small minority of patients benefit from CT when the diagnosis remains unclear.
Lower Gastrointestinal
SC-LG-0019
YOUNG PATIENTS WITH ANORECTAL MALFORMATION OR HIRSCHSPRUNG’S DISEASE: A COMPARISON OF QUALITY OF LIFE TO HEALTHY CONTROLS
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Aims of the Study: Patients with anorectal malformation(ARM) and Hirschsprung’s disease(HD) face long-term disturbance in bowel function even after definitive surgery. Little is known of their quality of life(QOL) in contrast to a healthy population. This study evaluates the QOL of patients with ARM and HD, and compares them to healthy controls.

Method: This is an ethically approved prospective study with recruitment occurring between September 2013-December 2014. All patients with ARM or HD who had primary surgery done in our institution at least 2 years prior to participation were eligible. Controls were age-matched and gender-matched, and enrolled from our population of patients with minor outpatient complaints and no recent history of gastrointestinal illness. All participants(parent and patient) were approached during scheduled outpatient visits and completed the questionnaires immediately, which consisted of the following PedsQL™ scales(maximum score 100): 4.0 Generic Core Scales(all ages,parent and child versions), 3.0 General Well-Being(GWB) Scale(children aged≥8 years,parent and child versions) and 2.0 Family Impact(FI) Module(all ages,only parent version). All were also scored on bowel function (BFS), maximum score 20. Χ², Kruskal-Wallis and Pearson correlation tests were used as appropriate, with significance level<0.05.

Results: There were 193 participants included: 87 controls, 62 ARM and 44 HD patients (Table). More parents than children answered the questionnaires, with the least proportion of children from the 2-7 year age group. One parent of an ARM patient could not provide responses on BFS, as the child’s severe bowel issues meant complete dependence on medical intervention. When comparing Core, GWB and FI scores, there were no significant differences between the groups although controls had the best scores indicating best QOL and general wellbeing, with least impact of the child’s health on the family. However, BFS were significantly different with the controls having best and ARM having worst scores. For all the questionnaires, when both parent and child provided responses, there were no significant differences in scores between parent and child indicating intradyad consistency. There was a significant positive correlation between BFS and Core (p<0.0001), and between BFS and GWB scores (p<0.005); and significant negative correlation between BFS and FI scores (p<0.0001).
### Table: Characteristics and scores of study participants

<table>
<thead>
<tr>
<th></th>
<th>Controls (n=87)</th>
<th>ARM (n=62)</th>
<th>HD (n=44)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age (years)</strong>#</td>
<td>7.1 (2.1-20.7)</td>
<td>6.9 (2.1-19.8)</td>
<td>9.1 (2.7-0.1)</td>
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<tr>
<td><strong>Gender (Male:Female)</strong></td>
<td>66:25</td>
<td>42:20</td>
<td>33:11</td>
<td>0.65</td>
</tr>
<tr>
<td><strong>Core Scores</strong>@</td>
<td>Parent (n=85)</td>
<td>89.69±7.0 2</td>
<td>84.70±12.95</td>
<td>86.81±14.98</td>
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<td></td>
<td>Child (n=53)</td>
<td>86.34±9.5 4</td>
<td>82.57±11.85</td>
<td>84.42±11.47</td>
</tr>
<tr>
<td><strong>General Well-being Scores</strong>@</td>
<td>Parent (n=35)</td>
<td>83.06±13.29</td>
<td>80.21±16.07</td>
<td>75.97±14.75</td>
</tr>
<tr>
<td></td>
<td>Child (n=38)</td>
<td>80.45±13.28</td>
<td>73.70±21.07</td>
<td>78.20±18.05</td>
</tr>
<tr>
<td><strong>Family Impact Scores</strong>@</td>
<td>(n=87)</td>
<td>14.22±12.89</td>
<td>19.98±17.33</td>
<td>15.18±15.83</td>
</tr>
<tr>
<td><strong>Bowel Function Scores</strong>#</td>
<td>(n=87)</td>
<td>20 (13-20)</td>
<td>16 (6-20)</td>
<td>18 (4-20)</td>
</tr>
</tbody>
</table>

*Scores are mean ± standard deviation.

**Note:** All scores are reported as median (range) except for scores reported as mean ± standard deviation.
*General Well-being Scale only applicable for children age 8 years and above
N.S. Not significant
#Data= median (range)  @Data= mean±standard deviation

**Conclusion:** Good bowel function positively impacts quality of life. Those with ARM and HD can achieve good quality of life comparable to controls, implying sufficient clinical support and internal coping strategies. Further studies on the impact of these diseases in adolescents and young adults would help transition to adult services.

**Disclosure of Interest:** S. A. Nah Conflict with: Singhealth Foundation Grant, C. Ong Conflict with: Singhealth Foundation Grant, L. Y. Ong Conflict with: Singhealth Foundation Grant, A. S. Jacobsen: None Declared, T.-L. Yap: None Declared, Y. Low: None Declared
Lower Gastrointestinal

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COHORT STUDY ON 8,110 APPENDECTOMIES IN GERMANY: LAPAROSCOPIC SURGERY IS ASSOCIATED WITH LESS POSTOPERATIVE COMPLICATIONS

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Aims of the Study: Meta-analyses indicate advantages of laparoscopic appendectomy compared to open appendectomy including lower complication rates. These data were mostly derived from studies performed in centers of excellence. Data on the results of laparoscopic appendectomy in general use are scarce and data from Germany are not available. We aimed to analyze the results of laparoscopic versus open appendectomy using a nationwide unbiased insurance-database.

Method: Data were extracted from the German health insurance TK, covering approximately 10% of the German population. A 3-year period (2010-2012) was analyzed and all patients aged 4 to 17 years with OPS codes for appendectomy were included. In addition, logistic regression analysis for risk of a surgical complication within 180 postoperative days was performed. Complications were defined by ICD and OPS codes.

Results: Appendectomy was performed in 8,110 patients (53% male; 47% female). The intraoperative finding was coded “uncomplicated” in 78.7% of the patients, “complicated” (i.e. perforation, abscess) in 8.5% and in 12.8% information on the type of appendicitis was missing. Appendectomy was conducted laparoscopically in 76.2% of the patients with a conversion rate of 1.2%. Laparoscopic surgery was associated with a shorter length of hospital stay in both, “uncomplicated” (5.2 vs. 5.5 days; p<0.001) and “complicated” appendicitis (7.5 days vs. 9.0 days; p<0.001) compared to open surgery. Patients with “complicated” appendicitis had a lower readmission-rate for a surgical complication after laparoscopic appendectomy (3.1 vs. 8.7%; p<0.05). Logistic regression analysis confirmed a significantly lower risk of readmission for a surgical complication after laparoscopic compared to open operation in patients aged 11 to 17 years when adjusted for the variables “gender”, “type of surgical department”, and “weekday of surgery”. Pediatric surgeons operated 24% and general surgeons 76% of patients. Pediatric surgeons utilized laparoscopy less frequently (61% vs.79%; p<0.01) and their conversion rate was significantly higher (2.9% vs.0.7%, p<0.001).

Conclusion: This first unbiased nationwide German cohort study confirms that laparoscopic appendectomy is associated with less postoperative complications compared to open appendectomy, in particular in patients with complicated appendicitis. Pediatric surgeons use laparoscopy less frequently compared to general surgeons. Laparoscopic appendectomy should be further promoted in pediatric surgery in Germany.
THE RISE OF TRANSANAL IRRIGATION FOR INTRACTABLE FAECAL INCONTINENCE AND CONSTIPATION IN CHILDREN

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Aims of the Study: To assess the impact of transanal irrigation (TAI) systems on the management of children with intractable faecal incontinence (FI) and constipation at our centre. Therapeutic management of FI and constipation in children includes: conservative approaches (education, behavioural therapy, biofeedback), medications (oral and rectal laxatives), neuromodulation techniques (transcutaneous, nerve stimulation, sacral nerve), transanal washouts and surgical interventions (intersphincteric botulinum toxin injection, antegrade continence enema [ACE], distal ACE [DACE], excision of megarectum and stoma).

Method: Retrospective case-note analysis of a prospectively maintained database of patients <17 years old with FI and/or constipation who have been managed with TAI (Peristeen® or Qufora®) and/or ACE between 2004 and 2014. Data collected included: age, gender, diagnosis, TAI and/or ACE functional outcomes (success defined as total continence or occasional soiling) and associated complications. Linear regression analysis was performed and P ≤ 0.05 was considered significant. Approval for this service evaluation was gained through our hospital’s Clinical Effectiveness Unit.

Results: In the 11 year period, 171 patients (of which 7 had incomplete data sets) were managed with ACE and/or TAI. Demographics of patients were 63% male with a median age of 9 (range 3.2 to 16.4) years. Diagnoses were: idiopathic constipation 64% (109/171), anorectal malformation 23% (40/171), Hirschsprung disease 5% (9/171), neuropathic bowel 5% (8/171) and gastrointestinal dysmotility 3% (5/171). TAI was used by 25% (42/171) and ACE was used by 78% (134/171) of patients – 4 TAI patients were subsequently managed with ACE and 1 ACE patient converted to using TAI. In those who tolerated TAI the success rate was 88% (28/32). The success rate of ACE was 79% (103/131). There were no complications with the use of TAI. The complication rate of ACE was 18% (24/131) – this included 9 stomal stenosis, 8 granulation tissue, 4 local infection/leakage, 2 stoma leakage, 2 stoma prolapse, 1 creation of false passage and 1 incisional hernia. On average we performed 11 ACE procedures per year with no significant difference in the absolute number completed over the last decade. However, the proportion of patients managed with TAI has increased significantly over time ($R^2 = 0.717; P = 0.016$). See Figure 1.
Conclusion: TAI offers a safe, effective and potentially complication free management for children with FI and constipation. Management of FI and constipation is, however, limited by patient acceptance. Confidence in the benefits of TAI has led to a proportionally greater use in our centre and we recommend TAI to other centres.
EFFECTIVENESS OF RECTAL SUCTION BIOPSY IN DIAGNOSING HIRSCHSPRUNG'S DISEASE

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Aims of the Study: Rectal suction biopsy (RSB) is performed on clinical and radiological suspicion of Hirschsprung’s disease (HD). Positive histochemical staining for acetylcholinesterase in the lamina propria in the RSB confirms HD. In addition, the presence of ganglion cells in the submucosa is examined. If the diagnosis is still uncertain a second RSB or a full thickness biopsy (FTB) is done. The aim of the study was to evaluate the effectiveness of the RSB in diagnosing HD.

Method: In this retrospective study, all infants younger than 1 year who had RSB for HD suspicion from January 2004 till April 2014 were included. All pathological results were reviewed. Number of all RSB and FTB per patient were recorded. Sensitivity and specificity of the first RSB were calculated.

Results: In total 183 patients were included, of whom 99 were boys, and mean age at the time of biopsy was 11 weeks (range 1-51). Based on the pathological results of the first RSB, HD was diagnosed in 55 patients (30%) and excluded in 128 patients (70%). Due to clinical doubt a second biopsy (FTB or RSB) was performed in 12/55 patients (22%), which confirmed HD in 9 patients. One patient did not undergo a second biopsy, but HD was excluded based on clinical follow up. Thus, HD was eventually excluded in 4 of the 55 patients with a positive first RSB, which means that the sensitivity of the first RSB was 81%.

In 19 of the 128 patients (15%) in whom HD initially was excluded, a second biopsy (FTB or RSB) was done, in which led to the diagnosis of HD in 10 patients. Two patients with negative biopsy results were still operated on because of the high clinical suspicion for HD, and surgery confirmed HD. Thus, HD was eventually diagnosed in 12 out of 128 patients, which means a 97% specificity. In total 63 patients underwent a laparoscopically assisted pullthrough procedure for HD.

A subanalysis was done for the 38 prematurely born infants in the sample, with a mean conceptional age at the time of biopsy of 44 weeks (range 34-82). In 19 of them, RSB had been at a mean conceptional age less than 40 weeks. Sensitivity and specificity of RSB in this sub sample were 83% and 97%, respectively, with one false-positive and one false-negative result. Six prematurely born infants underwent a pullthrough procedure.
Conclusion: The overall sensitivity of the RSB in the studied population was 81%, with 12 false-negative results, in which cases the extra biopsies had been necessary. The specificity was 97%, with 4 false-positive findings. Findings for the sub sample of prematurely born infants were comparable, which implies there is no reason to postpone a RSB in those infants. New methods must be considered to achieve better effectiveness of the RSB for diagnosing HD.
Lower Gastrointestinal

SC-LG-0023

NEUROPSYCHOLOGICAL PERFORMANCE AND NEED FOR BOWEL MANAGEMENT IN SCHOOL-AGED CHILDREN WITH ANORECTAL MALFORMATION AND HIRSCHSPRUNG'S DISEASE

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Aims of the Study: To prospectively evaluate neuropsychological functioning in 8-year-old anorectal malformation (ARM) and Hirschsprung’s disease (HD) patients.

Method: Intelligence (IQ), sustained attention, speed, memory, executive functioning, self-esteem, and quality of life were prospectively assessed in non-syndromal 8-year-old ARM and HD patients with validated neuropsychological tests and questionnaires.

Results: We included 23 ARM and 20 HD patients. Total IQ, memory, executive functioning, working speed, and self-esteem were conform the Dutch norms. Significantly lower was sustained attention: both working speed (median (IQR) Z-score -1.7 (-2.7 to -0.3) and -1.4 (-2.5 to 0.2) for ARM and HD patients) and attention fluctuations (median (IQR) Z-score -3.4 (-5.8 to -1.9) and -3.2 (-7.3 to -0.5) for ARM and HD patients) were below the Dutch norms (both p<0.001).

Educational data showed that 43% of the study cohort needed special education services (18.5% special education and 34.5% remedial teaching). This was significantly higher than the 20% in the normative population (p=0.011). HD patients reported impaired total quality of life, while the parents of both groups reported their child’s total quality of life to be impaired. There were sustained attention problems in 13/14 ARM (93%) and 4/7 HD (57%) patients who required bowel management, vs. 33% and 25% of patients who did not require bowel management (p=0.005 and 0.783, respectively).

Conclusion: Neuropsychological functioning is average in ARM and HD patients at school age with exception of sustained attention. In addition more special education services are needed compared to the normative population. The relationship between sustained attention and bowel management needs to be elucidated. These patients may benefit from a multidisciplinary approach.
**Lower Gastrointestinal**

SC-LG-0024

**CONTROLLED LONG-TERM URINARY AND SEXUAL OUTCOMES IN HIRSCHSPRUNG’S DISEASE TREATED WITH ENDORECTAL PULL-THROUGH AND TRANSANAL MUCOSECTOMY**

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**Aims of the Study:** To study the lower urinary tract symptoms (LUTS) and long-term sexual functional outcomes after transanal endorectal pull-through with transanal mucosectomy for Hirschsprung disease (HD) in relation to healthy controls.

**Method:** All patients with Hirschsprung’s disease > 4 years of age treated at our centre between 1987-2011 were invited to answer a detailed questionnaire on LUTS. For patients ≥16 years, items on sexual function were also included. Of the 92 respondents (response rate 66%), patients ages <4 years (n=12) and with mental retardation due to associated syndromes (n = 19) were excluded. Each patient was matched for age and gender to three controls randomly selected from the general population. Ethical approval was obtained.

**Results:** In total, 61 patients [median age 14 (range, 4-32 years); 72% male] were enrolled, including 16 males and 8 females aged ≥16 years. 89% of patients had classic HD, 7% had long segment disease, one patient had total colonic disease and two patients had aganglionosis extending to ileum. One patient had permanent enterostomy and three patients had significant bowel functional problems requiring antegrade colonic enema (ACE) conduit. The prevalence of LUTS among patients and controls is shown in Table 1. No significant differences compared to controls were apparent in patients operated for HD, apart from straining to void, which was reported by a higher proportion of controls. Patients reported comparable figures for sexual interest [2.57 ± 0.662 vs. 2.44 ± 0.710 (scale 0-3), \(P=0.446\)] and satisfaction [2.14 ± 1.276 vs 2.08 ± 1.351 (scale 0-3), \(P=0.886\)] similar to controls. However, significantly less patients with HD were currently in stable relationships [35% vs. 60% of controls; \(P=0.038\)]. Erectile function in males after endorectal pull-through was comparable to controls [3.00 ± 0.00 vs. 2.85 ± 0.625 (scale 0-3), \(P=0.32\)].
**Table:**

<table>
<thead>
<tr>
<th>Variable</th>
<th>HD (n=61)</th>
<th>Controls (n=183)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Voiding frequency</td>
<td>10 (17)</td>
<td>51 (28)</td>
<td>0.082</td>
</tr>
<tr>
<td>Urge incontinence</td>
<td>9 (15)</td>
<td>38 (21)</td>
<td>0.327</td>
</tr>
<tr>
<td>Stress incontinence</td>
<td>5 (8)</td>
<td>21 (12)</td>
<td>0.472</td>
</tr>
<tr>
<td>Incontinence without urge/stress</td>
<td>0 (0)</td>
<td>11 (6)</td>
<td>0.070</td>
</tr>
<tr>
<td>Straining to begin or continue voiding</td>
<td>7 (12)</td>
<td>48 (27)</td>
<td>0.018 **</td>
</tr>
<tr>
<td>Social/other disadvantages due to urinary problems</td>
<td>3 (5)</td>
<td>12 (7)</td>
<td>0.767</td>
</tr>
</tbody>
</table>

* Percentage reporting any degree impairment  
** P<0.05 between groups

**Conclusion:** Our results support the safety of endorectal pull-through with transanal mucosectomy for HD with regard to preservation of sexual and urinary function. Patients reported erectile function comparable to controls, and no significant differences in the long-term LUTS profile after surgery.
Young Investigator Award SC-YI-0025 to SC-YI-0034

Young Investigator
SC-YI-0025

BMP4 TREATMENT OF ISOLATED FOREGUT IN 3D EXPLANT TISSUE CULTURE INDUCES AN OESOPHAGEAL ATESEIA PHENOTYPE.

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Aims of the Study: Oesophageal Atresia/Tracheo-Oesophageal Fistula (OA/TOF) arises from disturbed foregut division during early embryogenesis. Animal models of abnormal foregut development permit the identification of signals which influence oesophageal and tracheal formation, and have shown that prior correct molecular patterning of tissue into dorsal (oesophageal) and ventral (tracheal) domains is crucial for proper foregut separation. In the Adriamycin Mouse Model (AMM) of OA/TOF, Bone Morphogenetic Protein (BMP) signalling is disturbed in the foregut. In mice which lack Noggin (a BMP antagonist), a similar phenotype to the AMM is observed and Bmp4 knockout mice exhibit tracheal agenesis. We recently established an in vitro system which successfully recapitulates foregut division into oesophagus and trachea. We hypothesised that manipulation of BMP signalling would result in abnormal division of foregut in culture.

Method: Embryonic day 9 CD1 mice underwent micro-dissection to generate isolated foregut explants (License B100/4447 Irish Medicines Board). Explants were embedded in collagen gel and incubated together with Affi-Gel® beads soaked in PBS for control, Noggin at 0.1μg/μl or Bmp4 at 0.1μg/μl and 0.01μg/μl. Explant morphology was investigated by wholemount immunodetection of endoderm marker Hnf3β with 3D Optical Projection Tomography scanning (Fig1B). Molecular patterning was analysed on cryosections for markers: Hnf3β, Sox2 (dorsal/oesophageal) and Nkx2.1 (ventral/respiratory).

Results: Bmp4 treated foreguts exhibited abnormal development while Noggin treated foreguts did not differ from controls. Failure of oesophageal formation, abnormal lung bud morphology and communication between the stomach and respiratory structures occurred in all Bmp4 treated specimens (Fig1D). Proximal foregut atresia or agenesis occurred in most Bmp4 treated specimens (19/23). These malformations occurred together with loss of Sox2 and gain of Nkx2.1 across the entire foregut endoderm, indicating a shift towards respiratory identity (Fig2B). In regions of the foregut abnormally positive for Nkx2.1, Sox2 was ectopically present outside the nuclei (Fig2C). Comparison of Bmp4 treated foreguts to well-established models such as the AMM confirmed that Bmp4 treatment induced an OA phenotype.
Conclusion: These results reveal that BMP signalling is crucial for proper foregut separation through regulation of tissue identity, and confirm that administration of Bmp4 is sufficient to induce tracheo-oesophageal malformations in foreguts in culture. Foregut endoderm cell re-specification from dorsal to ventral fate may be an important mechanism in the pathogenesis of tracheo-oesophageal malformations.
LANDSCAPE OF GENOMIC ALTERATION IN NEUROBLASTOMA

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Aims of the Study: Despite large-scale efforts to identify individual genomic drivers in neuroblastoma pathogenesis, few recurrent genetic alterations have been identified (MYCN, ALK,PTPN11, ATRX). We propose that there may be a global trend underlying patterns of genomic alteration in the cancer genome that predisposes to the accumulation of deleterious change. Through unbiased characterization of the neuroblastoma exome, we set out to identify patterns of risk, hypothesizing that the global frequency and type of single-nucleotide substitution (SNS) in tumor tissue is significantly different from that of matched non-tumor tissue, and that altered patterns of substitution confer increased risk of deleterious change.

Method: Using published data from whole genome sequencing of 40 neuroblastoma and paired normal samples, we extract the exomic data, perform functional annotation with the ANNOVAR software package, and filter to exclude variants with dbSNP identifiers. We then apply prediction algorithms to assess the likelihood that each SNS would result in deleterious change to the protein product. Using the R Statistical package, we perform pairwise analysis of SNS signatures, highlighting global differences between the subsets of SNSs that are unique to the non-tumor sample, unique to the tumor sample, or existent in each.

Results: The SNS signatures of the neuroblastomas and their paired non-tumor counterparts demonstrate a large degree of overlap, however each tumor has a subset of de novo SNSs that does not exist in its paired sample (figure 1a). These subsets show enrichment in certain transversion-type SNSs which cause a shift in the percentage of non-synonymous SNSs from 57% in tumors on the whole, to 68% in the tumor-specific subsets (p < 0.001). Functional analysis demonstrates that this shift carries with it a significant enrichment in the number of SNSs predicted by PolyPhen-2 as “probably damaging” to the structure and function of the protein (38% vs. 28%, p < 0.001)(figure 1b).
Conclusion: Neuroblastomas appear to harbor very few recurrent, localized genomic changes. However, we demonstrate that the global signature of SNSs in the tumors differs from that of matched healthy tissue, and that these substitutions are biased toward non-synonymous change with ensuant risk of deleteriously impacting protein function. Our analysis suggests that there may not exist a single unifying mutation at the heart of the genomic insult in neuroblastoma, but rather an accumulation of transversion-enriched SNS subsets which may aggregate the risk for deleterious change.

Aims of the Study: Anorectal malformations (ARM) and Hirschsprung's disease (HD) are congenital malformations requiring pelvic floor surgery in early childhood, with possible sequelae for psychosexual development. Aim of the study was to assess psychosexual well-being in adult ARM and HD patients related to health-related quality of life.

Method: Eligible for this cross-sectional two-center study were all patients aged ≥18 years who in early childhood had been operated for ARM or HD. Exclusion criteria were mental retardation, comorbidity affecting sexual functioning (e.g. malignancy requiring chemotherapy), and cloacal malformation. Participants completed the International Index of Erectile Functioning (IIEF-15), Female Sexual Functioning Index (FSFI), Female Sexual Distress Scale (FSDS), Hirschsprung and Anorectal Malformation Quality of Life Questionnaire (HAQL), and sexual education questionnaire (SEQ).

Results: Response rates were 32% and 37% for ARM and HD patients, respectively. Forty-five patients did not consent, resulting in 70 participating ARM and 36 HD patients (median age 26 years). Seven/40 men with ARM (18%) reported moderate to severe erectile dysfunction, versus 4/20 men with HD (20%). Sixteen and 11 of 30 women with ARM (53% and 37%) reported sexual dysfunction or sexual distress, respectively, versus 9 and 3 of 16 women with HD (56% and 19%). Quality of life, which was overall good, and type of malformation or operation was not associated with having a psychosexual disorder. Sexual education related to the congenital anomaly was reported to be insufficient by 42 ARM (60%) and 22 HD patients (61%).

Conclusion: Approximately 20% of male ARM and HD patients reported erectile dysfunction, while 53 and 37% female ARM and HD patients reported sexual dysfunction. There was no relation with quality of life or type of malformation. Both ARM and HD patients felt a need for better psychosexual education. Further research is needed to determine optimal form and timing of this education.
LENGTH OF MECHANICAL VENTILATION PREDICTS 2-YEARS NEURODEVELOPMENTAL OUTCOME IN CONGENITAL DIAPHRAGMATIC HERNIA.

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Aims of the Study: Although neurodevelopmental impairment is one of the most significant morbidities among CDH survivors, little is known about predictors of neurodevelopmental outcome (NDO). Length of mechanical ventilation (LMV) was shown to predict NDO at 6 and 12 months of age. Our aim was to evaluate if, in CDH survivors, LMV is correlated to and predicts NDO at 2 years of age.

Method: A prospective longitudinal study was conducted between 2009 and 2012. Mental, motor, and language development at 2 years of age was assessed by Bayley Scales of Infant and Toddler Development — 3rd Edition (BSID-III). Moderate delay: score 85-70; severe delay: score <70. First admission LMV (in hours) was obtained from medical records of all patients. The correlation between LMV and NDO was analyzed using Pearson test. Receiver operating characteristic (ROC) analysis was performed to determine the accuracy and best cut-off value of LMV to predict the risk of neurodevelopmental delay in one or more scales. Statistical significance was set at p<0.05.

Results: Forty subjects were included in the study. Median (interquartile range) Bayley score was 105 (95-110), 100 (97-109), and 100 (91-106) for mental, motor, and language scales, respectively. Seven patients had moderate delay and 2 had severe delay in one or more scales. Median LMV was 204 hours (153-390). There was a significant inverse correlation between LMV and NDO at 2 years of age (r=-0.4297, p=0.0063; r=-0.3089, p=0.0061; r=-0.3710, p=0.0201 for mental, motor, and language scales, respectively). LMV was predictive of both overall moderate (AUC 0.792; p=0.0117) and severe (AUC 0.816; 0.0002) delay. The best cut-off value was 288 hours (12 days) (sensitivity: 83% and 100% for moderate and severe delay, respectively; specificity: 79% and 74% for moderate and severe delay, respectively). When the scales were analyzed separately, LMV was predictive of delay only for the motor development (AUC 0.903, p<0.0001 for moderate delay and 0.816, p<0.0002 for severe delay). The best cut-off was 11 days for moderate delay (sensitivity: 100%; specificity: 69%) and 12 days for severe delay (sensitivity: 100%; specificity: 74%). LMV was not predictive for moderate delay in the language scale. ROC analysis was not possible for severe delay in language scale and both moderate and severe delay in the mental scale as too few patients fell in these categories.

Conclusion: In CDH survivors, LMV confirms to be an important predictor to identify patients at risk of neurodevelopmental delay, in particular for motor development. Strict follow-up and early intervention therapy should be activated in every baby with a history exceeding 12 days of MV.
INTESTINAL EPITHELIAL CELL INJURY IS REVERSED BY ADMINISTRATION OF ACTIVATED AMNIOTIC FLUID STEM CELLS

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Aims of the Study: It has been shown that amniotic fluid stem (AFS) cell administration improves survival, clinical status, gut structure and function in experimental necrotizing enterocolitis (NEC). The mechanism of this effect is unknown. We hypothesize that intestinal epithelial cell injury in vitro could be reversed by administration of AFS cells acting via TLR4 signaling pathway.

Method: AFS cells were treated with 1 or 2µg/ml LPS and PGE2 expression (TLR4 pathway end product) was measured by ELISA. To establish an in vitro model of bowel injury, intestinal epithelial cells (IEC-18) were treated with 50µg/ml lipopolysaccharide (LPS). IEC-18 viability measured by MTT after 3 hours of treatment, was assessed in the following conditions: (a) no LPS + fresh medium; (b) LPS + fresh medium; (c) LPS + supernatant from untreated AFS cells; (d) LPS + supernatant from AFS cells treated with 1µg/ml LPS or (e) 2µg/ml LPS. Results (mean±SEM) were compared using one-way ANOVA with Bonferroni post-test. P<0.05 was regarded as significant.

Results: PGE2 concentration in the supernatant of AFS cells at baseline was 451±134pg/ml and increased to 919±100pg/ml after exposure to 1µg/ml LPS (p<0.05) and to 1233±122pg/ml after exposure to 2µg/ml LPS (p<0.0001). IEC-18 cells treated with LPS had a lower viability (0.38±0.03) than controls (0.50±0.03, p<0.05). IEC-18 viability was not rescued by addition of untreated AFS cells (0.39±0.04) or AFS cells treated with 1µg/ml LPS (0.42±0.04), but it returned to normal using AFS cells treated with 2µg/ml LPS (0.50±0.04, p<0.05; Figure).
Conclusion: This study shows that intestinal cell viability is impaired by exposure to LPS mimicking the epithelial damage occurring in NEC. This damage can be reversed by treatment with supernatant from AFS cells activated by stimulation of the TLR4 pathway. These findings elucidate a paracrine mechanism of AFS cell action and indicate the potential for a pharmacological intervention in NEC.
DEFICIENCY OF PLATELET-DERIVED GROWTH FACTOR RECEPTOR-ALPHA-POSITIVE CELLS (PDGFRALPHA+ CELLS) IN THE COLON OF HIRSCHSPRUNG’S DISEASE

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Aims of the Study: PDGFRα+-cells are a novel interstitial cell type, found within the gastrointestinal tract of various animals, which have come to the attention of neurogastroenterology researchers in recent years. These cells are found alongside interstitial cells of Cajal, neurons and smooth muscle cells (SMCs), and appear to have a role in neurotransmission and smooth muscle contraction. To date, studies have analyzed the expression of PDGFRα+-cells in healthy intestine in both humans and animals, but no data exists regarding the expression of PDGFRα+-cells in human gastrointestinal disease. The aim of our study was to investigate whether the expression of PDGFRα+-cells is altered in Hirschsprung’s disease.

Method: Hirschsprung’s disease tissue specimens (n=10) were collected at the time of pull-through surgery, while colonic control samples were obtained at the time of colostomy closure in patients with imperforate anus (n=10). Immunolabelling of PDGFRα+-cells was visualized using confocal microscopy to assess the distribution of these cells, while Western blot analysis was undertaken to quantify PDGFRα protein expression.

Results: Confocal microscopy revealed PDGFRα+-cells within the mucosa, myenteric plexus and smooth muscle in normal controls, with a marked reduction in PDGFRα+-cells in the Hirschsprung’s disease specimens (Figure). Western blotting revealed high levels of PDGFRα protein expression in normal controls, while there was a striking decrease in PDGFRα protein expression in ganglionic and aganglionic regions of Hirschsprung’s disease colon (Figure).
Conclusion: These findings suggest that the altered distribution of PDGFRα+ cells in the aganglionic bowel may contribute to the motility dysfunction in Hirschsprung's disease.
SYSTEMATIC REVIEW OF COMPLICATIONS IN LAPAROSCOPIC SURGERY FOR HIRSCHSPRUNG'S DISEASE

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Aims of the Study: This study investigated complications associated with Hirschsprung’s disease (HD) with Minimal Access Surgery (MAS) based on 20 year literature review.

Method: Literature search was performed on Google Scholar, Embase, Pubmed and Cochrane on terms “Hirschsprung’s diseases” “laparoscopic” and “complication”. Data was collected for: surgical approach, type of repair - primary or secondary repair, level of aganglionosis, intraoperative- and postoperative- complications.

Results: The search (1994-2014) yielded 61 publications with cumulative 1772 patients for this systematic analysis. Of these, 95.5% (n=1693) underwent a primary repair i.e., a single operation without prior colostomy and 4.5% (n=79) a secondary repair. The level of aganglionosis was restricted to the rectosigmoid colon in 60.0% (n=1064), descending colon 5.2% (n=93) and transverse colon in 3.7% (n=65). Techniques included Laparoscopic Swenson (LS) 7.6% (n=135), Laparoscopic Duhamel (LD) 11.9% (n=211), Laparoscopic Soave (LSO) 18.3% (n=325) and Laparoscopic Georgeson Pull-through (LGP) 59.0% (n=1046). Other procedures were single incision (SILS) (n=49) and Rintala’s procedure (n=6)

Intraoperative complications included: conversions to open 1.8% (n=32), bleeding 0.3% (n=5) and bowel twisting 0.3% (n=4). Morbidity included: enterocolitis 6.9% (n=122), soiling 7.2% (n=128), perianal excoriation 3.7% (n=65), constipation 4.6% (n=82), anastomotic stricture 1.6% (n=29), adhesive bowel obstruction 1.0% (n=18), anastomotic leaks 0.8% (n=14) and perforation requiring reoperation 0.5% (n=9).

Comparing approaches; enterocolitis was highest in LS 11.11% and lowest in LGP 5.54%. Perianal excoriation was highest in LS 7.4%; LD (n=0). Anastomotic stricture rates were highest in LSO 3.69% and lowest in LGP 0.86%. Adhesive bowel obstruction, anastomotic leaks and perforation rates were highest in the LD 1.42%; LS (n=0).
Conclusion: LGP is the most common MAS procedure for HD. Intraoperative complications for MAS in HD are low 2.3%. Morbidity is high (26.4%) in the cumulative series, but may not be specific to MAS. Specific morbidity was correlated with perianal excoriation in LS, anastomotic strictures in LSO and adhesive bowel obstruction, anastomotic leaks and perforations in LD.
SC-YI-0032

IL-17A RELEASED BY PANETH CELLS DRIVES THE INFLAMMATORY RESPONSE IN NECROTIZING ENTEROCOLITIS

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Aims of the Study: Necrotizing enterocolitis (NEC) is characterized by an inflammatory and ischemic destruction of the intestine [1]. It is well known that Paneth cells can initiate intestinal inflammation via releasing the highly proinflammatory IL-17A [2]. As previous studies have suggested a role of Paneth cells in the development of NEC, we tested the hypothesis that IL-17A, released by Paneth cells, could drive the exaggerated inflammatory response in a murine NEC model.

Method: 14d old wild type (wt, n=31) and IL-17A knock out (ko, n=38) Balb/c mice were randomly divided into a breast fed control and a NEC group. NEC mice were injected with dithizone (100mg/kg BW, i.p.) to disrupt the Paneth cells. 6h after the injection, mice were fed twice with LPS milk (10µg/g BW) in a 3h interval. Animals were then sacrificed and small intestines were collected for histologic analysis. RNA was extracted from the distal ileum and MIP-2 expression was quantified by qRT-PCR. Flow cytometry was used to measure intestinal neutrophil accumulation and macrophage/monocyte activation in intestinal leucocyte populations.

Results: Both wt and IL-17A ko treated mice developed necrotic lesions in the small intestine (NEC score: wt=2.8 vs ko=2.4, p>0.05). IL-17A ko mice showed significant less inflammatory response compared to wt mice. MIP-2 expression was markedly reduced in the IL-17A ko mice (Fig. 1). They had significantly less activation of intestinal macrophage/monocyte (% of Ly6-C⁺ in CD45⁺: wt=14.98 vs ko=5.27, p<0.001), and significantly reduced accumulation of neutrophils (% of Ly6-G⁺ in CD45⁺: wt=13.91 vs ko=3.02, p<0.001 and Fig. 2).
The depletion of IL-17A did not prevent the occurrence of small necrotic gut lesions in this paneth cell dependent NEC model. The absence of IL-17A, however, significantly dampened all inflammatory changes. Thus, the ability of Paneth cells to induce the NEC associated inflammation appears to be dependent on IL-17A.

Tissue Engineered Skeletal Muscle Produced in Vitro with Natural Extracellular Matrix and Muscle Precursor Cells for In Vivo Regeneration

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Aims of the Study: This study aims to generate constructs in vitro with decellularised muscular matrices and precursor cells displaying cell survival and functional maturation of skeletal muscle, with the final aim to implant these grafts in vivo for muscle restoration and regeneration.

Method: Effective clinical treatment for volumetric muscle loss resulting from congenital or acquired conditions are extremely limited to date. Tissue engineering aims to mimic neo-organogenesis for the production of living tissue in vitro to be applied in regenerative medicine strategies. To date, the majority of engineered muscle strategies have been using biodegradable synthetic polymers or biologic components to generate scaffolds for cell delivery or three-dimensional culture. However, these systems are far from reproducing the complex and unique highly-organised network of skeletal muscle and they seem to support myogenesis only partially. An expanding recent technology has been developed to cover this experimental gap by decellularizing tissues and organs, creating scaffolding materials that retain the architecture of the native tissue, including vasculature and biofactors present in the extracellular matrix (ECM), making this structures ideal for cell seeding.

For this purpose, muscle stem cells (satellite cells) were isolated from murine hindlimb muscles and seeded into decellularised muscles in combination with other muscle resident cells, and cultured in vitro for 7 and 14 days. The acellular muscles were produced using three different decellularization protocols: detergent-enzymatic treatment (DET), anti-polimerizing agent-enzymatic treatment (Lat B) and detergent alone (SDS). A full characterization of the obtained acellular matrix was performed. In parallel with in vitro cell repopulating experiments, we also developed an in vivo model in mouse for the transplantation of tissue engineered skeletal muscles and the evaluation of their potential for regenerative medicine.
Results: Efficient decellularization was achieved after 3 cycles of DET, 1 cycle of Lat B and 3 cycles of SDS as evidenced by histology and DNA quantification. The remaining matrix showed distinct characteristics in terms of ultra- and nano-architecture, pro-angiogenic properties and ECM composition, with an overall decrease in collagen (50-60%), elastin (100-40%) and glycosaminoglycans (75-60%) content in respect to native tissue. Muscle precursor cells seeded in the 3 scaffolds displayed different degrees of proliferation, migration and differentiation. The in vivo muscle transplantation model in mouse was successful developed and optimised, with 90% of survival after ablation of the extensor digitorum longus (EDL) muscle and replacement with the acellular scaffold. Histology of hindlimb muscles harvested 90 days post-implantation showed strong local muscle regeneration and functional recovery of the limb, as evidenced by animal ability to catch an object.

Conclusion: Our preliminary results strongly suggest that acellular muscles support myogenesis and muscle stem cell maintenance in vitro, and show great potential for their in vivo application for muscle reconstruction in the field of regenerative medicine. This study also highlight the active role of the acellular ECM in affecting cell migration and differentiation.
A NOVEL POPULATION OF PLATELET-DERIVED GROWTH FACTOR ALPHA (PDGFRα) CELLS IN THE HUMAN URETEROPELVIC JUNCTION OBSTRUCTION: ITS RELATIONSHIP TO NEURONS AND ICCS

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Aims of the Study: In the upper urinary tract, peristaltic contractions serve to propel urine from the renal pelvis through the ureter into the bladder. Normal ureteral motility requires coordinated interaction of neuronal cells, smooth muscle cells (SMC) and interstitial cells of Cajal (ICCs). Recently, a new type of interstitial cells characterized by the expression of antibodies against platelet-derived growth factor receptor alpha (PDGFRα) has been discovered in the gastrointestinal tract and the bladder. PDGFRα positive cells mediate neurotransmission in colonic muscles and are adjacent to nerve fibres, ICCs and SMCs. The aim of this study was to determine the morphology and distribution of PDGFRα positive cells and their relationship to nerve cells, ICCs and SMCs in human UPJ.

Method: After obtaining ethical approval, 32 human intrinsic UPJ obstruction specimens and 20 control UPJ specimens were obtained. Cryosections were stained with antibodies against PDGFRα, PGP 9.5, c-kit and phalloidin which labels filamentous actin. Confocal-immunofluorescence-double staining including 3D-reconstruction was performed. PDGFRα and PGP 9.5 protein expression was assessed by western blot and RT-PCR was performed to evaluate gene expression levels.

Results: PDGFRα positive cells had spindle-shaped morphology forming discrete networks in the lamina propria and throughout the inner longitudinal and outer circular muscle layers of the human UPJ. Confocal-immunofluorescence-double staining revealed that PDGFRα positive cells were in close proximity to PGP 9.5 positive nerve fibres. PDGFRα positive cells were adjacent to, but distinct from c-kit positive ICCs and phalloidin positive SMC (Fig.1). PDGFRα gene and protein expression levels as well as immunohistochemical expression in UPJ obstruction were not different from controls (Fig.2) ICCs were markedly reduced in UPJ obstruction compared to controls. Confocal-immunofluorescence showed markedly reduced PGP 9.5 positive nerve fibres in UPJ obstruction compared to controls. Western blot confirmed markedly decreased protein expression levels of PGP 9.5 in UPJ obstruction compared to controls. The relative messenger RNA expression levels of PGP 9.5 was significantly decreased in UPJ obstruction compared to controls (p<0.001) (Fig.3).
Conclusion: We show the expression and distribution of a novel interstitial cell type, PDGFRα, in the human UPJ. PDGFRα positive cells, ICCs and SMCs form a syncytium in the upper urinary tract which may play an important role in motor neurotransmission and peristaltic contraction mechanisms. Decreased PGP 9.5 expression and decreased ICCs observed in UPJ obstruction may have a role in the failure of transmission of peristaltic waves across UPJ obstruction.
Thoracic SC-TH-0035 to SC-TH-0043

Thoracic

SC-TH-0035

GROWTH ASSESSMENTS AND THE RISK OF GROWTH RETARDATION IN CONGENITAL DIAPHRAGMATIC HERNIA: THE MULTICENTER FOLLOW-UP STUDY

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Aims of the Study: Although the long-term morbidities of congenital diaphragmatic hernia (CDH) are an important topic, less information is available regarding the physical growth of patients with CDH. The purpose of this study was to assess the growth of patients with CDH during long-term follow-up, and to identify risk factors for growth retardation (GR).

Method: A multicenter retrospective observational study was conducted in 2013 (ethical approval: No.509). Among the 228 patients with CDH who were born between 2006 and 2010 and who underwent operations at the 9 participating institutions, 182 patients (79.8%) survived to discharge. Of these surviving patients, 174 were included in the present study. Patients with chromosome abnormality (n=5) or very low birth weight (n=3) were excluded. Body weight and height were measured at 1.5, 3, and 6 years old. Other clinical factors were determined by reviewing medical records. GR was defined as a Z-score of either weight or height that was less than -2. Patients who had GR at any of the 3 time points were categorized as the GR group. Clinical variables which had low rates of missing values (<15%) were compared between the GR and non-GR groups using the Wilcoxon rank-sum test and Fisher's exact test. Multiple logistic regression analysis was conducted successively for only the factors that were statistically significant (p<0.01), and had low correlations with other factors (r<0.65). The numerical data was divided into 2 groups by the cut-off value, which was calculated from the receiver operating characteristic curve.
Results: Thirty-five cases (20.1%) were categorized as belonging to the GR group. The rates of GR at 1.5, 3, and 6 years old were 20.0% (26/130), 14.7% (16/109), and 14.3% (5/35), respectively. Among the 26 patients with GR at 1.5 years old, 62.5% (10/16) still had GR at 3 years old. Univariate analyses showed that the risk factors for GR were as follows: birth weight (p<0.001), birth height (p=0.024), small for gestational age (p=0.049), right side of hernia (p=0.038), use of inhaled NO (p=0.002), defect size of C or D (p<0.001), surgical finding of liver-up (p=0.047), patch repair at the primary operation (p<0.001), length of stay (p<0.001), vasodilator administration at discharge (p=0.014), tube feeding at home (p=0.004), and home oxygen treatment (p<0.001). Among these factors, only birth weight under 2,698g (cut-off value) (odds ratio [OR] 6.6, 95% confidence interval [CI] 2.4-21.4, p<0.001), inhaled NO (OR 5.1, 95%CI 1.3-26.5, p=0.022) and home oxygen treatment (OR 7.0, 95%CI 1.8-31.4, p=0.006) were the significant risk factors for GR in a multivariable analysis.

Conclusion: GR was observed in 20% of CDH survivors. The risk factors for GR were small birth weight and the need for inhaled NO and home oxygen treatment.

LUNG TO HEAD RATIO CORRELATES WITH LUNG VOLUME IN CONGENITAL DIAPHRAGMATIC HERNIA SURVIVORS.

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Aims of the Study: Prenatal lung to head ratio (LHR) and observed-to-expected LHR (O/E LHR) are largely used as predictors of postnatal outcome in terms of survival in neonates with congenital diaphragmatic hernia (CDH). Few studies, on small patients’ populations, report on the correlation of LHR with actual lung hypoplasia, only in non-survivors. Aim of our study was evaluate if LHR and O/E LHR correlate with post-natal lung volume, expressed as functional residual capacity (FRC).

Method: Lung function tests were performed by multibreath washout traces with an ultrasonic flowmeter and helium gas dilution technique. For each patient, FRC was recorded at 4 timepoints: T0: after stabilization, before surgery, intubated; T1: 24 hours after surgery, intubated; T2: 48 hours after surgery, intubated; T3: after extubation. LHR and O/E LHR were gathered from prenatal clinical records. Correlation of LHR and O/E LHR with FRC at different timepoints was assessed using Spearman test. Results are median (interquartile range); p<0.05 was considered significant.

Results: Forty patients were included in the study. LHR was 1.8(1.6-2.2)mm and O/E LHR was 51.4(41.5-65.0)%. FRC was 11.5(8.2-13.4)ml.kg⁻¹, 14.4(11.8-17.8)ml.kg⁻¹, 14.7(12.8-18.9)ml.kg⁻¹, and 19.4(17.1-23.3)ml.kg⁻¹ at T0, T1, T2, and T3, respectively. LHR was significantly correlated with FRC only at T3 (r=0.6851, p=0.0024). O/E LHR showed a trend towards a correlation with FRC at T3 (r=0.4608, p=0.0627).

Conclusion: In CDH survivors, LHR is correlated with lung volume only after extubation. Present findings suggest that LHR may represent a predictor not only for survival but also for postnatal lung volume and function.
Aims of the Study: To define patterns in the management of congenital diaphragmatic hernia (CDH).

Method: 180 delegates (77% senior surgeons) from 44 (26 European) countries completed a survey administered at the EUPSA 2014 annual meeting.

Results: 34% surgeons work in centres that treat <5 cases of CDH a year, 38% in centres that treat 5-10 cases a year, and 28% in centres that treat >10 cases a year. 62% surgeons work in ECMO centres and 23% in fetal surgery centres.

Prenatal workup and delivery: 47% surgeons request prenatal magnetic resonance imaging, 53% routinely offer karyotyping, 22% offer fetal intervention, 74% monitor head-to-lung ratio (LHR), and 55% administer maternal steroids. Delivery of CDH neonates is via caesarean section for 47% surgeons, at 36-38 weeks for 71%, and in a tertiary centre for 94% surgeons.

Postnatal management: 76% surgeons report elective intubation, 65% routinely start antibiotics preoperatively, and 45% administer surfactant. In case of severe refractory hypoxia, 66% surgeons consider extracorporeal membrane oxygenation (ECMO) with a variable course (<14 days for 46%, 14-28 for 8%, >28 for 46%). Parenteral feeding is started preoperatively by 56% surgeons. Only 13% surgeons request contrast studies preoperatively to rule out malrotation. The most used prognostic factors for CDH are liver-up position (81%), defect side (66%), oxygenation index (56%), LHR (48%), and stomach in the chest (36%).

Surgical management: 73% respondents report that the decision to operate is made by surgeons and neonatologists together. If on ECMO, 61% surgeons wait to perform CDH repair after ECMO. Favourite approaches are shown in Figure 1. A hernial sac is excised by 83% surgeons. The favourite suturing technique is interrupted sutures for 90% respondents. In case of large defect, 89% surgeons prefer the use of a patch (Goretex 70%, Permacol 9%, PTFE 9%) and 11% favour a muscle flap repair. Intestinal malrotation is checked intra-operatively by 70% surgeons and corrected by 66%; of these, 69% surgeons perform an appendectomy. Only 35% surgeons leave a chest drain at the end of the surgery.
**Postoperative management:** 56% surgeons electively leave the patient paralysed (<2 days 53%, 2-5 days 34%, >5 days 13%). Enteral feeds are started in less than 2 days by 21% respondents, between 3 and 5 days by 63%, and in more than 5 days by 16%, as continuous feeds by 57% and as bolus by 43%. Anti-reflux therapy is started soon after surgery by 53% surgeons.

**Follow-up:** Patients are variably followed up (<2y 10%, 2-5y 33%, 5-10y 29%, >10y 28%), with chest X-rays requested by 45% surgeons, lung function tests by 56%, hearing test by 52%, and neurodevelopmental assessment by 75%. In case of recurrence, 66% surgeons would have the same operative approach, 29% would do a thoracoscopic repair, 71% would remove the old patch and 39% would use a different patch material.

**Image:**

![Image](image_url)

**Conclusion:** The number of surgeons who work at an ECMO centre is high, compared with the total number of ECMO centres. However, even in these more specialised teams, many aspects of CDH management are lacking consensus. Fetal intervention and minimally invasive surgery are offered by a minority. Controversial management decisions such as early delivery, C-section, antenatal use of steroids, chest drain, and correction of intestinal malrotation are made by a high proportion of surgeons, without evidence from the literature. Guidelines for antenatal and postnatal management of CDH should be developed to support multicentre studies.
Aims of the Study: Prenatally diagnosed congenital pulmonary airway malformations (CPAM) of the lung often present a volumetric reduction during late pregnancy. This reduction has been reported to be complete in a number of patients. Nonetheless, it is not clear how to follow-up those patients, and in particular if thoracic CT scan is always required to uncover, during infancy, those prenatally "regressed" lesions. Aim of the present study is to critically evaluate the outcome of CPAM detected and disappeared during pregnancy, and the role of postnatal CT scan in detecting residual disease.

Method: Prospective collected database of all patients with prenatal diagnosis of CPAM at our Institution between January 2009 and December 2014 was searched. For the purpose of the present study we evaluated only those infants who had a complete regression, during gestation, of the prenatally detected CPAM. Thoracic CT scan was suggested in all patients between 2nd and 3rd months of age. If persistent disease was noted at CT scan, elective surgery was offered, even in asymptomatic patients. Comparison between infants who underwent CT scan and those who did not was performed. T-Test and Mann-Whitney test were used as appropriate. P< 0.05 was considered significant.

Results: During the study period, we diagnosed 78 CPAM, of which 26 (33%) completely regressed during pregnancy at a median age of 34 weeks of gestational age (GA) (interquartile range (IQR): 28-37). In 5 (19%) cases, parents refused to perform thoracic CT scan during infancy (Group A), while 21 (81%) patients did the examination (Group B). Of the 21 CT scans, 100% were positive for persistent residual disease and 10 (48%) patients underwent surgery. The other 11 (52%) patients were followed-up for bronchial anomaly and segmental emphysema. No symptoms developed at mean follow-up of 44-month (range 6-66). 5 patients who did not received CT scan during early infancy, 1 (20%) became symptomatic (pneumonia) at 6 months and required uneventful surgery (intralobar sequestration).
**Table:**

<table>
<thead>
<tr>
<th></th>
<th>Group A: 5 patients</th>
<th>Group B: 21 patients</th>
<th>p</th>
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<tbody>
<tr>
<td>GA at complete regression; median (IQR)</td>
<td>33 (31-37)</td>
<td>34 (28-38)</td>
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<tr>
<td>Prenatal US diffuse hyper-echogenicity; (%)</td>
<td>5 (100)</td>
<td>19 (90)</td>
<td>1</td>
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<tr>
<td>GA at birth; median (IQR)</td>
<td>38 (37-40)</td>
<td>39 (37-40)</td>
<td>1</td>
</tr>
<tr>
<td>Sex; Male/Female</td>
<td>3/2</td>
<td>12/9</td>
<td>1</td>
</tr>
<tr>
<td>Development of symptoms (%)</td>
<td>1 (20)</td>
<td>0 (0)</td>
<td>0.19</td>
</tr>
</tbody>
</table>

**Conclusion:** The high rate of persistent disease even in prenatally disappeared CPAM, indicate the need for close monitoring during early infancy. Thoracic CT scan is a valuable tool to detect residual disease and to correctly inform parents on indications for surgery even in asymptomatic infants.
**Aims of the Study:** The current nationwide survey aimed to clear the clinical features of the congenital cystic lung diseases (CCLD) from prenatal period through adulthood and to establish the suitable surgical strategy in consideration with the late postoperative problems during adulthood.

**Method:** Of 874 CCLD patients identified in the nationwide survey, 428 patients born between 1992 through 2012 and treated at the 10 high-volume centers (194 prenatally and 234 postnatally diagnosed) were involved in the more precise retrospective review with statistical analysis under IRB approval of each center.

**Results:** In the 194 prenatally diagnosed patients, fetal lung abnormality was first recognized during 24th gestational week at median, and 16.7% of the patients presented fetal hydrops in ultrasonography. Among the 241 neonatal patients, the APGAR score (5 minutes) was lower than 5 in 8.3%. On the postnatal day 30, 14.0% of the neonatal patients required respiratory support, and 3.3% had been dead. Overall, 14 patients died in the series; 12 died of pulmonary hypoplasia and 2 died of complication of respiratory therapy during the infantile period. Among 140 patients who were totally asymptomatic immediately after birth, 33.6% of the patients developed the pulmonary infection during their first year of life, and 22.1% did at the age of 1. Of the 428 patients, single lobectomy was performed in 68.2%, whereas segmentectomy was performed in 7.5%, and multiple lobectomy and pneumonectomy was done in 3.0%; 89.0% thru open thoracotomy. Late postoperative complications included thoracic deformity in 30 patients and persistent lung cyst in 4, whereas no malignancy was observed in the whole series. Pathological diagnosis was available in 362 and included CCAM in 189, bronchopulmonary sequestration in 110, and bronchial atresia in 652. Nearly matured respiratory function was assessed beyond the age of 6 in 22 patients, and the prenatally diagnosed patients showed significantly higher %>Vital Capacity (VC) compared to the postnatally diagnosed patients (98.3±11.9 vs. 81.7±9.7, p<0.0222).
Conclusion: The results of the current survey indicated that estimatedly 10-15% of the prenatally diagnosed CCLD patients may carry a high risk for perinatal respiratory distress, early operation, especially with preceding preoperative diagnosis and no episodes of lung infection, seems to be associated with better development of the reserved lung in their later lives, and incidence of carcinogenesis in CCLD may be extremely low.
Aims of the Study: To discuss the prevention and the treatment of the complication of minimally invasive operation (NUSS procedure) for pectus excavatum.

Method: 2954 cases of pectus excavatum age from 3 to 18 years old treated by NUSS procedure from July 2002 to December 2014. Among them, 165 cases were recurrent cases and 596 cases with other diseases. 162 cases (5.48%) developed perioperative complications.

Results: All the cases were accomplished safely. The average operating time was 39.4(19-350) min, the average blood loss was 3.3 (1-400) ml. The intraoperative complications were developed in 68 cases, including pneumothorax 14 cases, heart injury 2 cases, pericardium injury 11 cases, lung injury 23 cases, liver injury by diaphragm injury 2 cases, intercostal muscle avulsion 4 cases, intercostal vascular injury 12 cases. All of the intraoperative complications were found and managed properly during the surgery. The postoperative complication were occurred in 94 cases, including bar displacement 22 cases, pleural effusion 14 cases, long-term chest pain 8 cases with 5 of scoliosis, long-term fever 5 cases, pericardial effusion 1 case, drainage tube breaking 1 case, infection 4 cases, allergy 29 cases, spontaneous pneumothorax 3 cases, thoracic outlet syndrome 3 cases, wound dehiscence 4 cases.

Conclusion: Most of the complications are slight and can be resolved. With the understanding of the complications and the progress of technology, the complications could be greatly reduced. The minimally invasive surgery has become the standard surgery of pectus excavatum.
MDCT EVALUATION OF TRACHEOMALACIA COMPARED TO BRONCHOSCOPY IN CHILDREN WITH VASCULAR RING

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Aims of the Study: The aim of this study is to compare multidetector computed tomography (MDCT) and bronchoscopy in the evaluation of tracheomalacia (TM) in children with vascular ring (VR). TM is a key prognosis factor in management of VR.

Method: We performed a retrospective and monocentric study of all patients operated on for vascular ring from 1997 to 2014. We collected from charts and office notes the clinical data such as: initial symptoms, age and weight at surgery, type of surgery and clinical outcome. When performed and available for analysis, all MDCT were blinded reviewed by two radiologists in consensus, independently of bronchoscopy results. MDCT images and bronchoscopy results were both reviewed according to 3 criteria: 1=percentage of tracheal narrowing (TN), 2=height of tracheal narrowing (TN), 3=severity of tracheomalacia (TM). Criteria 1 included transversal, antero-posterior diameters and cross sectional area measurements. 3D tracheal and vascular reconstructions were also performed in each patient. We evaluate the concordance between radiologic and endoscopic results for each criteria of TM.

Results: From 1997 to 2014, 21 patients with vascular ring were operated on in a single center. All patients presented respiratory symptoms such as stridor (n=8), respiratory distress episodes (n=6), chest infection (n=2), pneumothorax (n=1), “wrong way” swallowing (n=1), associated dysphagia (n=5). Mean weight and age at surgery were respectively 6300 g and 5 months. MDCT images and bronchoscopy results were available and analysed for 10 patients. Concordance between MDCT analysis and bronchoscopy was excellent for percentage of TN and height of TN (respectively, 90 and 88 %), and moderate for the severity of TM (60%). 3D reconstruction combines the 3 criteria and supplementary information about oesophageal compression and vascular malformation useful for surgical planning. Its concordance with bronchoscopy is currently being explored.
Conclusion: MDCT analysis of TM in children with vascular ring is very accurate in the evaluation of the percentage and height of TN consecutive to the vascular compression but bronchoscopy remains mandatory for dynamic analysis of severity of TM. Contribution of 3D reconstruction remains to be explored but surely help the comprehension of the malformation for the surgeon.
PREOPERATIVE SIMULATION WITH 3D MODELS OF PECTUS EXCAVATUM

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Aims of the Study: Recently, 3D printers have made it possible to create models of organs for preoperative simulation. Preoperative simulation increases the safety and reliability of operations. The Nuss procedure is a minimally invasive procedure for pectus excavatum and is currently widely used. However, it is difficult to identify the optimal location for placement of the pectus bar in order to maximize the Nuss procedure effect or predict the effects of this procedure, preoperatively. Furthermore, this procedure is difficult for both family and nursing staff to understand. Therefore we made full scale chest wall models of a number of children’s conditions with pectus exavatum using a 3D printer and simulated the Nuss procedure in order to understand how to maximize the effectiveness. Also, we used the 3D models for preoperative explanation of the surgical procedure to the patient’s family and the nurse in charge.

Method: Three cases of pectus exavatum in children (5-13 y/o) underwent preoperative CT scans. All cases were diagnosed as pectus exavatum before the CT scans. We transformed the CT data into 3D graphical data using Z view (Lexi Co., Ltd., Tokyo, Japan) and customized it using Geomagic Freeform (Geomagic, Cary, NC, USA). This data was then output as an STL (stereolithographical) file and printed with an Objet 260 connex (Stratasys, Eden Prairie, MN, USA) 3D printer. Surgeons then simulated the Nuss procedure in order to determine the optimal location for the pectus bar for maximum improvement of the chest wall. Following the operations, surgeons evaluated the similarity of the chest wall shape of the 3D models and those of the patients. After the preoperative explanations of the surgical procedures to the patient’s families and the nurse, the surgeons asked their opinions regarding their understanding of the surgical procedures.

Results: The 3D models of the chest walls were reported to closely resemble the patient’s chest walls with the shape correction. The preoperatively determined pectus bar locations were found to be satisfactory for the patients. The 3D models were sufficiently sophisticated to permit realistic preoperative simulation. The surgeons were satisfied with the quality of the 3D pectus exavatum models. The patient’s families and the nurse were satisfied with the explanations and understood well the surgical procedure when it was explained with the assistance of the simulated 3D model.

Conclusion: 3D models made using a 3D printer were useful for the Nuss procedure for pectus exavatum. The 3D models can be used both for preoperative simulation and explanation of the surgical procedure. This preliminary study showed the possibility of prediction of the Nuss procedure’s effect. Furthermore, the 3D printer enables customization to individual patient’s needs.
Aims of the Study: Tracheal reconstruction for pediatric tracheal stenosis is a very challenging procedure because the trachea is narrow and delicate. Identifying the clarification and the extent of the lesion by CT images and endoscopy has been an important preoperative examination. Recently, three-dimensional (3D) models created by 3D printers have shown potential for preoperative planning. Surgeons can interact with 3-D models for easier understanding of the morphology. To our knowledge, there are few reports using 3-D models for general pediatric surgery. In this study, we show the use of 3-D models for the preoperative planning of surgery for pediatric tracheal stenosis.

Method: The models were constructed from a CT data. The 2D images of the CT were reconstructed into 3D images using a program named Zview® (Lexi Co., Ltd., Tokyo, Japan). The 3D images were converted to 3D models by an Object 260 Connex® printer (Stratasys, Eden Prairie, MN, USA). We constructed the 3-D models using a rubber-like material offered a variety of elastomer characteristics. Printing layer thickness was 0.016mm. We fabricated the 3D models for four patients with tracheal stenosis. Three of them had subglottic lumen stenosis and one had congenital tracheal stenosis.

Results: The cost of making the 3D models depends on their complexity. A simple tracheal model is a couple of US dollars, and the tracheal models we created for this particular trial cost around 50 dollars each. The entire process required a total of 4-5 hours for a simple structure such as the tracheal stenosis and 24 hours for a complicated structure. Although 3D models can interpret the location and the extent of the tracheal stenosis, minute anatomy or thin structures such as vocal cords cannot be accurately reproduced the original forms. Even though 3D models have their limitations, they also have their advantage, for example: we can share the surgical planning data not only with the surgical parties but with the children’s families.

Conclusion: In contrast to present 2D images, such as CT and MRI, 3D models can provide tactile hands-on feedback to surgeons for surgical planning. Limitations of making 3D models are the relative cost and the length of time required for production. We believe that as this procedure becomes more advanced, the cost-benefit ratio will subsequently decrease. Further improvements in materials and 3-D printing technology can produce more precise models. Creating the 3-D models of pediatric tracheal stenosis is useful and helpful for preoperative surgical planning and patient understanding.
Aims of the Study: To analyze complications and outcomes of end-to-end urethral anastomosis performed for posttraumatic bulbar strictures or posterior urethral injuries in pediatric patients.

Method: The records of 15 boys, age under 18 years, admitted to our tertiary trauma center with urethral injury from 1989 to 2014 were reviewed retrospectively. 7 patients were excluded (2 for iatrogenic trauma, 2 for minor straddle injuries that were not operated on, 2 for incomplete records and 1 lost to followup). 8 analyzed patients were operated for bulbar or posterior urethral injury. Mean followup after the operation was 4.5 years (range 0.5 to 10). To obtain up-to-date followup information all the analyzed patients were contacted by letter and phone in January 2015 and asked about lower urinary tract or erectile dysfunction using The IIEF-5 Questionaire (SHIM).

Results: Mean age at the time of injury was 12.3 years (range 5 to 17). 4 patients with pelvic fracture had complete posterior urethra disruption, 3 after straddle injury developed obliterating stricture of bulbar urethra and 1 patient torn his bulbar urethra apart by a sharp hook. Except immediate exploration of the open perineal wound all others were operated via perineal approach 1 to 6 months after initial suprapubic tube insertion. 5 patients needed cystotomy to identify proximal urethral stump by a probe, 2 had partial pubectomy to gain urethral length. Postoperative complications included stricture in anastomosis in 6 patients (all re-operated, 4 more than once including attempts of endoscopic internal urethrotomy). 1 patient had external massive bleeding around permanent urinary catheter on day 6 after surgery caused by arteriovenous fistula that was stopped by urgent angiography and coil insertion. After discharge 3 patients had transient stress incontinence. All patients had Max Flow above 20ml/s on the last followup uroflowmetry except 2 (12ml/s and 14.6ml/s). None has any lower urinary tract dysfunction symptoms in adulthood, 1 suffers from mild erectile dysfunction and 2 report moderate erectile dysfunction due to penile shortening.

Conclusion: Delayed end-to-end anastomosis for pediatric urethral injury is safe operational option, however high rate of short-term complications and re-operations should be expected. In the long-term the outcomes are much better with penile shortening in 2 patients being the most severe complication in our cohort.
ANALYSIS OF DATA ON BURNS IN CHILDREN TO DEVELOP INJURY PREVENTION PROGRAM

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¹Clinic for Pediatric Surgery and Orthopedics Nis, Clinical Center Nis, Serbia, Nis, Serbia, ²Department of Pediatric Surgery, Clinical Hospital Centre, Rijeka, Croatia, ³Department of Pediatric Surgery, University Clinical Center, Tuzla, Bosnia and Herzegovina

Aims of the Study: This study investigated the epidemiology about causes of burns in different age groups, affected body areas, determination the extent of the burns and classification of burns degree.

Method: A retrospective study was performed in patients who were admitted to our hospital between October 2009 and October 2014 with regards to epidemiological factors leading to burns.

Results: During the 5 year period of investigation period 132 patients were admitted with burns to our Centre. With regards to the age groups:

Group A: 0-1 year (n= 17; 11 males: 6 females) (12.8%) with mean hospital stay of 7.1 days
Group B: 2-5 year (n= 81; 49 males: 32 females) (61.4%) with mean stay of 9.8 days
Group C: 5+ year (n= 34; 24 males: 10 females) (25.8%) with mean hospital stay of 8.4 days

With regards to etiology of burns:
(1) Hot liquids 75.8% (n=100) with n=15 in Group A, n=62 in Group B and n=23 in Group C
(2) Chemical burns 3.8% (n=5) with n=1 in Group A, n=4 in Group B and none in Group C
(3) Hot items 6.8% (n=9) with none in Group A, n=5 in Group B and n=4 in Group C
(4) Electrical burns 3% (n=4) with none in Group A, n=1 in Group B and n=3 in Group C
(5) Open fire 9% (n=12) with n=1 in Group A, n=6 in Group B and n=5 in Group C

Children from the lower social-economic status (Gipsy’s population) comprised of the 18.9% (n=25), with 57.6% (n=76) from urban and 42.4% (n=56) coming from the rural population. There were 56.8% (n=75) children treated for shock, with following body areas affected: genital region 21.2% (n=30), palms 18.9% (n=25) face and neck 30.3% (n=40), upper extremities 44.7% (n=58), lower extremities 38.6% (n=51), abdomen 16.6% (n=22) and thorax 45.5% (n=60).

With regards to area of burns:
Group A: mean 17% (maximum burnt area 30%) with GRll burns in all children
Group B: mean 10% with GRII (n=16) and GRIII (n=5)
Group C: mean 11% with GRII (n=24) and GRIII (n=5), GR IIAB-III (n=8), GR III (n=1) and GRIII-IV (n=1)

In all children primary and secondary treatment were performed. In 61.4% (n=81) Ag Sulfadiazine or fibrinolysis, necrectomy and shavings were performed in 36.4% (n=48) and skin transplantation in 3.8% (n=5) of patients. One boy > 5 years with 95% GRIII-IV burns from electric shock died from multi-organ failure.

**Conclusion:** In our cohort the most common cause of burns is hot liquids which affect age groups 0-5 years. The mean total burn surface area was 20%. Results from this study particularly with regards to the age groups will be helpful in developing injury prevention programs for our cohort targeting parents and kindergartens.
Aims of the Study: In children the majority of spinal fractures are stable compression fractures with a decrease of the anterior height of the affected vertebrae. A certain degree of anterior wedging of the thoracolumbar vertebrae is often seen in children without a history of trauma. Therefore, it remains challenging to differentiate whether a mild degree of anterior wedging is the sequela of trauma or may be a normal developmental variant. The aim of this study was to describe the sagittal index measured on plain radiographs of the lower thoracic and lumbar spine of healthy children and adolescents without a history of trauma in order to determine the amount of physiological wedging during growth.

Method: The sagittal index (SI), as the ratio between the anterior and posterior height of the vertebrae, was measured on 100 randomly selected lateral lower thoracic and lumbar spine radiographs performed in children without a history of trauma, malignancy or steroid use. The subjects were divided into five arbitrary age groups with 20 children each (<3, 4-7, 8-11, 12-14 and 15-17 years). Data are presented in means, range and standard deviation (SD). Measurements were performed by one reader at two time points and intraclass correlation coefficient (ICC 2,1) was calculated for the repeated measurements.

Results: The mean SI of all vertebral bodies (Th11-L5) was 0.932 (range 0.839-1.1, SD 0.03). The mean SI of each of the seven vertebral bodies is listed in the table. There was a significant difference by vertebral body position with higher SI values of the more caudal vertebrae. Moreover, children ages 0-3 years had significantly higher overall SI values compared to all other four age groups. The ICC of the repeated measurements for all vertebrae was 0.901 (95% CI 0.885-0.915, p<0.000). Additionally, the ICC in relation to the individual vertebrae indicated a strong to almost perfect agreement between measurements. ICC for measurements performed in the different age groups gradually increased from the youngest to the oldest patients (0-3a: 0.72; 4-7a: 0.878; 8-11a: 0.814; 12-14a: 0.896; 15-17a: 0.961).
Table:

<table>
<thead>
<tr>
<th>Vertebral Body</th>
<th>mean SI</th>
<th>range; SD</th>
<th>ICC (2,1)</th>
</tr>
</thead>
<tbody>
<tr>
<td>T11</td>
<td>0.915</td>
<td>0.852-0.962; 0.023</td>
<td>0.783</td>
</tr>
<tr>
<td>T12</td>
<td>0.912</td>
<td>0.859-0.963; 0.021</td>
<td>0.815</td>
</tr>
<tr>
<td>L1</td>
<td>0.922</td>
<td>0.839-0.975; 0.020</td>
<td>0.808</td>
</tr>
<tr>
<td>L2</td>
<td>0.926</td>
<td>0.868-0.977; 0.019</td>
<td>0.797</td>
</tr>
<tr>
<td>L3</td>
<td>0.932</td>
<td>0.880-0.963; 0.016</td>
<td>0.727</td>
</tr>
<tr>
<td>L4</td>
<td>0.940 #</td>
<td>0.886-1.000; 0.020</td>
<td>0.870</td>
</tr>
<tr>
<td>L5</td>
<td>0.969 *</td>
<td>0.920-1.102; 0.036</td>
<td>0.876</td>
</tr>
</tbody>
</table>

Caption: Mean sagittal index (SI), range and standard deviation and ICC of vertebrae T11 to L5 (* p < 0.05 vs. all other vertebrae, # p < 0.05 vs. all other vertebrae except L3)

Conclusion: A certain amount of age-dependent vertebral wedging as assessed on lateral radiographs seems to a normal developmental variant in children and adolescents. Higher SI values are found in the caudal vertebrae. From T11 to L5 the SI of all measurements was greater than 0.83 suggesting that lower values raise the possibility of vertebral body injuries.
THE LONG-TERM HEALTH-RELATED QUALITY OF LIFE IN CHILDREN TREATED FOR BURNS AS INFANTS FIVE TO NINE YEARS EARLIER

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Aims of the Study: Assessment of the health-related quality of life (HRQoL) in children and adolescents is gaining growing interest. Aim of this study was to investigate the long-term outcome after infant burn injury 5 to 9 years after the initial accident. All participants had been treated for burn injury before the age of 1 year. We hypothesized that the HRQoL in young burn survivors may be impaired compared to healthy age matched peers.

Method: The HRQoL of 126 infant burn-injured patients with a mean total body surface area (TBSA) of 3.5% was queried with the standardized and validated 17D questionnaire. The HRQoL of the respondents was compared to that of a representative sample of the general age-standardized population comprising a sample of healthy school children from several schools.

Results: A total of 44 (35%) children with a mean age of 7 years responded, and 64% of them were male. The median time from trauma was 6.3 years. Burn injury related features, age at burn injury time, burn size and site, and the treatment given were similar in the respondents group and all 126 children with burn injury approached. The median TBSA of inpatient treated respondents was 5.5%, (range 2.5 to 40%) and of outpatient treated 1.25% (range 0.5 to 7%). The mean HRQoL score of the respondents was better, 0.968, than that of the control population, 0.936 (p<0.05). Comparison of the 17D profiles of the respondents having been treated as inpatients or outpatients showed that those treated on an outpatient basis had better scores on the dimensions of speech, breathing, and friends (p<0.05). The 17D profiles of respondents with scalds or contact burns were similar.

Conclusion: The perceived and expressed long-term HRQoL in children treated for burns as infants was good, and on some dimensions (sleeping, learning, discomfort and symptoms, breathing, depression, and appearance) even better, than that of the control population. Etiology of the burn injury did not affect the perceived HRQoL.


ESOPHAGEAL PERFORATION IN CHILDREN: ANALYSIS OVER A 20-YEAR PERIOD

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Aims of the Study: Esophageal perforations in children are among all the traumatic cases rare and always challenging. We reviewed our experience with esophageal perforations surgically treated to determine the overall morbidity and therapeutic tools.

Method: A retrospective review of esophageal perforations treated from 1995 to 2014 was performed.

Results: We treated 12 children between 5 months and 12 years old with esophageal trauma who all presented esophageal perforation. Median age at time of operation was 3.9 years. Iatrogenic injury was the most frequent cause of esophageal perforation (50%): the perforation by endoscopic dilatation of the stricture of the esophagus (n=3), repeated introducing of NG tube (n=2) and the perforation by rigid esophagoscopy technique (n=1). More esophageal perforations were caused by button battery ingestion (n=3, 25%), accidental ingestion of alkaline detergent (n=1), esophageal transection as a consequence of a blunt trauma (n=1) and a swallowed leaf of Dieffenbachia in an attempted suicide with the developing of aortoesophageal fistula (n=1). Surgical therapy was performed on all patients. The suture of the lesion and gastrostomy was performed on 8 patients, resection of the site of perforation and anastomosis on 2 patients, gastric transposition on 1 patient and a cervical esophagostomy on 1 patient. 6 patients from our study group have no swallowing or feeding difficulty. Esophageal strictures occurred in 5 patients, 3 of them are regularly treated by endoscopic balloon dilatation, 2 patients are fed via gastrostomy. 1 patient lives with a lateral cervical esophagostomy. 2 patients live with tracheostomy. No death occurred in our group.

Conclusion: The specific treatment of the esophageal perforation should be selected individually. Early diagnosis and surgical treatment are life-saving. However, the rate of morbidity in our study group is high.
RESULTS FOLLOWING SURGICAL TREATMENT OF JUDET IV DISPLACED RADIAL NECK FRACTURES IN CHILDREN AND ADOLESCENTS

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Aims of the Study: Pediatric radial neck fractures represent 5-10% of all elbow fractures. Completely displaced fractures (Judet IV) are rarely encountered injuries. However, these injuries compromise the vulnerable blood supply of the radial head leading to a higher ratio of avascular necrosis compared to non-displaced or minimally displaced fractures. An outcome study including solely Judet IV radial neck fractures has not been performed yet.

Method: All patients treated with Judet IV radial neck fractures between 2001 and 2014 were analyzed. The degree of displacement was measured on initial radiographs and only Judet IV fractures were included. In order to assess postoperative outcome range of motion and follow-up radiograph were assessed. Additionally, the Linscheid and Wheeler score was used.

Results: In total 29 patients (15 male, 14 female) were treated with Judet IV displaced radial neck fractures. The mean age of the patients was 9.5 years ranging from 4-17 years. Mechanisms of accident included falls on level surface in 12 children, playground and sports injuries in another 12 children and falls from a height < 1.5 m in 5 children. The majority of the patients (17/29) sustained isolated injuries, 7 patients had additional elbow luxation, 4 patients sustained concomitant fractures of the olecranon and 1 patient a fracture of the ulna shaft. Closed treatment was performed in 80% of the children (n=21 closed reduction and ESIN, n=2 closed reduction and K-wire osteosynthesis). Open reduction was mandatory in 6 patients (n=4 open reduction and ESIN, n=2 open reduction and plate osteosynthesis). In the further course of treatment resection of an avascular radial head was indicated in 2 patients (6 months following open reduction and ESIN, 8 months following open reduction and plate osteosynthesis). Another re-operation was performed in one patient due to impaired elbow movement (extension/flexion 0°-10°-110°, pro-/supination 10°-0°-10°) 6 months post-injury. Outcome for all patients was assessed after a mean of 8 months. The Linscheid-Wheeler score of the patients is presented in the table. Patients with poor outcome had a significantly higher age compared to patients with excellent, good and fair outcome, respectively (p<0.05).
Table:

<table>
<thead>
<tr>
<th>Linscheid-Wheeler-Score</th>
<th>[n]</th>
<th>mean age (range)</th>
<th>primary treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>excellent</td>
<td>10</td>
<td>9 a (4-13 a)</td>
<td>n=9 closed reduction</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>n=1 open reduction</td>
</tr>
<tr>
<td>good</td>
<td>8</td>
<td>8.8 a (5-13 a)</td>
<td>n=7 closed reduction</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>n=1 open reduction</td>
</tr>
<tr>
<td>fair</td>
<td>6</td>
<td>9 a (6-13 a)</td>
<td>n=5 closed reduction</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>n=1 open reduction</td>
</tr>
<tr>
<td>poor</td>
<td>5</td>
<td>13 a (10-17 a)</td>
<td>n=2 closed reduction</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>n=3 open reduction</td>
</tr>
</tbody>
</table>

**Caption:** Linscheid-Wheeler.Score, number, mean age and primary treatment of 29 patients with Judet IV radial neck fractures

**Conclusion:** Treatment of completely displaced radial neck fractures remains challenging. Worse outcome can be expected in older children. It still has to be elucidated to what extent the initial injury and/or a necessary open reduction contribute to the occurrence of avascular necrosis of the radial head leading to poor outcome.
Aims of the Study: Road traffic injuries rank second to interpersonal violence and the road traffic fatality rate in the African Region is higher than for any WHO region. The aim of this study is to explore potential differences in number and patterns of fractures between childhood survivors and non-survivors after a motor vehicle crash in view of developing targeted and more effective prevention strategies.

Method: Total body radiographs were obtained of all children under 13 years presenting to our trauma unit after a motor vehicle crash during a 30 month period between January 2010 and July 2012. In addition, total body radiographs were obtained from the mortuary of all children under the age of 13 who demised after a motor vehicle crash. The number and types of fractures as well as any other skeletal malformations were compared and evaluated. Approval was obtained from our University Ethics Committee.

Results: A total of 1045 children were assessed; 1007 presenting to the trauma unit, and 38 directly to the mortuary. Twenty patients presenting to the trauma unit did not survive. The total amount of non survivors was 58 but 6 were excluded since radiographs were not available. From the 987 survivors 79 were excluded because the radiographs were unavailable. The average age of children was 6.2 years (SD 3.1; Range 0-12). Fractures were far more common in non-survivors than survivors (78.8% vs 46.7%). Fractures of the Skull (p<0.0001), Cervical Spine (p<0.0001), Thoracic Vertebrae (p<0.0001), Trunk including Shoulder (p<0.0001), Lower Arm (p<0.002), Upper Arm (p<0.3) were statistically significantly more frequent in non-survivors than in survivors. Fractures of Facial bones, Elbow, Wrist, Hand, Pelvis, Femur, Knee, Foot and Ankle were different but not statistically significant in survivors and non-survivors.

Conclusion: This is the first study documenting and comparing fracture patterns with outcome (survivors and non-survivors) in young children after motor vehicle crash. Since motor vehicle crashes are the most common cause of unnatural childhood death it is important to study fracture patterns in order to understand the injury mechanism and develop preventative strategies. This study indicates that skull, cervical spine and torso fractures are more intricately associated with severe morbidity and mortality in children than fractures of other body regions. The upper torso, head and neck are the body regions most vulnerable in young children and requiring most protection.
HELIPAD IMPROVES EFFICIENCY WITHOUT INCREASING BURDEN TO PAEDIATRIC TRAUMA

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Aims of the Study: Helipad facilitates helicopter transfer of patients to major trauma centres (MTC). A helipad was recently introduced on the hospital roof of a South London MTC, to replace the older and less efficient helicopter landing facility in an off-site field (hence part of the transfer used to involve road ambulance). Our study aimed to evaluate the case-mix, severity and outcome of paediatric trauma before and after the helipad was introduced.

Method: The prospectively maintained hospital trauma registry and the UK Trauma Audit and Research Network (TARN) database were interrogated. Two months after the introduction of helipad, paediatric traumas treated in 7 consecutive months were reviewed. This was compared with the same period in the previous year (before helipad).

Inclusion criteria were patients aged 18 years or younger who fulfilled TARN criteria for referral. Case-mix comparisons were based on Injury Severity Score (ISS), TARN Probability of survival (Ps), and requirement for surgery. Outcomes measured were Intensive care unit (ICU) admission and length of stay, hospital length of stay, survival, and rehabilitation referral.

Continuous data were presented as median (range). Comparison of medians used Mann-Whitney U-test, and proportions used Fisher’s exact test or two-tailed x² test as appropriate, statistical significance was P<0.05.

Results: Before and after helipad introduction, we treated 180 and 191 paediatric major traumas respectively, both with 10% of patients transferred in by helicopter (18 v 19, P=0.99). 42 patients before helipad (12.1 years) and 36 patients after helipad (13.1 years) (p=0.81) were included for comparison in this study.

The case-mix and helicopter transfer rate were not statistically different between the two periods, when severity of cases were assessed with ISS, Ps, and requirement for surgery. The ICU admission rate, ICU and hospital lengths of stay, survival and rehabilitation referral were also not significantly different between the two periods (Table).
Table:

<table>
<thead>
<tr>
<th>TARN paediatric traumas:</th>
<th>Before helipad (Jun-Dec 2013), N=42</th>
<th>After helipad (Jun-Dec 2014), N=36</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Helicopter transfer (%)</td>
<td>9 (21%)</td>
<td>7 (19%)</td>
<td>0.83</td>
</tr>
<tr>
<td>Injury severity score (ISS)</td>
<td>16 (4-59)</td>
<td>9 (4-54)</td>
<td>0.068</td>
</tr>
<tr>
<td>Probability of survival (Ps)</td>
<td>99.28 (36.6 - 99.8)</td>
<td>99.63 (55.0-99.8)</td>
<td>0.133</td>
</tr>
<tr>
<td>Requirement for surgery</td>
<td>25</td>
<td>21</td>
<td>0.92</td>
</tr>
<tr>
<td>Requirement for ICU admission</td>
<td>22</td>
<td>11</td>
<td>0.086</td>
</tr>
<tr>
<td>ICU length of stay</td>
<td>1 (1-49)</td>
<td>0 (0-25)</td>
<td>0.15</td>
</tr>
<tr>
<td>Hospital length of stay</td>
<td>6 (1-90)</td>
<td>6 (3-67)</td>
<td>0.73</td>
</tr>
<tr>
<td>Survival (%)</td>
<td>41 (98%)</td>
<td>36 (100%)</td>
<td>1</td>
</tr>
<tr>
<td>Referral for rehabilitation (%)</td>
<td>10 (24%)</td>
<td>14 (39%)</td>
<td>0.23</td>
</tr>
</tbody>
</table>

**Caption:** Severity and outcome difference before and after helipad

**Conclusion:** Introduction of the helipad did not significantly change the case-mix, severity, or outcome of paediatric traumas, despite providing a more superior service through logistic and efficient helicopter patient transfer during the acute management of severe trauma. The volume and severity burden to the hospital and ICU were not increased as a result of the helipad.

**References:**

**Disclosure of Interest:** Y.-W. Tan Employee of: YWT was previously employed and ZM is currently employed by the organisation, Z. Mukhtar Employee of: YWT was previously employed and ZM is currently employed by the organisation
NO SPACE FOR BLOOD? THINK AGAIN! THE HAEMODYNAMIC IMPACT OF HEAD INJURY IN CHILDREN

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1Ministry of Health Holdings, 2KK Women's and Children's Hospital, Singapore, Singapore, 3Paediatric Surgery, KK Women's and Children's Hospital, Singapore, Singapore

Aims of the Study: In comparison to adult trauma epidemiology, a much higher incidence of head injury is seen in paediatric trauma. Notably, the resuscitation approach for isolated head injury (IHI) in children differs from that of other solid organ injuries. Influenced by the belief that massive blood loss is unlikely and over-resuscitation may worsen cerebral oedema, a more conservative administration of intravenous fluid and blood product is often practised. By reviewing our experience in paediatric head injury and evaluating the physiological and haematological parameters, this study aims to gain better understanding on the haemodynamic impact of head injury in children and provide information needed to revise our treatment protocol.

Method: With ethical approval, data was extracted from our hospital trauma registry, a prospectively collected database. We reviewed data on all head injury patients with injury severity score (ISS) ≥ 9 admitted to the intensive care unit (ICU) of our institution between January 2011 and December 2014. We categorized patients without other major organ injury or bleeding wound as IHI.

Results: Over the study period, 4186 children were admitted to our centre with head injury. Thirty-six children were included in this study. Median age was 6 years (range 1 month to 14 year). Road traffic accident was the most common injury mechanism (58%). Their median ISS was 25 (range 16 to 50). Out of 29 (81%) children sustaining IHI, 4 (14%) patients had hypotension and 9 (31%) patients received intravenous bolus fluid of more than 20ml/kg during resuscitation. Fourteen (48%) patients were noted to have haemoglobin ≤10g/dL at presentation. Seventeen (59%) patients developed coagulopathy and 10 (34%) had thrombocytopenia. Blood products were transfused to 14 (48%) patients within 12 hours. Ten patients (34%) underwent surgery for intracranial blood clot removal. Their median ICU stay was 4 days (range 1 to 47 days). There were 5 deaths.

Conclusion: While IHI in adults rarely results in major blood loss due to limited intracranial space, we have shown that it is not uncommon for IHI to impact a child’s haemodynamic status. Therefore, clinicians involved in paediatric trauma resuscitation should be mindful of this and adopt a more aggressive but balanced approach for fluid resuscitation and early blood product transfusion.
Aims of the Study: The management of active bleeding with haemodinamic lability in the paediatric trauma patient is difficult and generally leads to damage control surgery. Vascular Interventional Radiology (VIR) techniques are useful for the diagnosis as for the definitive treatment. The aim of our study was to describe our experience and evaluate the effectiveness of VIR in the management of the paediatric trauma patient with active bleeding signs.

Method: Retrospective analysis (2003-2014) of politraumatic patients who showed contrast blush on computed tomography treated by VIR techniques.

Results: In the reported study period 16 patients underwent VIR procedures, medium age was 13 years (5-17). The most frequent lesional mechanism was traffic accident (8 of 17) and 87.5% were blunt traumas. Findings on initial Computed Tomography were 13 contrast bluses and 2 absences of arterial flow. In 2 cases the contrast blush appeared 48 hours after the accident.

Arteriography permitted to localize the bleeding vessels in all the cases, performing selective or supraselective renal (7), pelvic (5), hepatic (3), splenic (1) and intercostal (1) embolization. One patient required an endoprosthesis for renal revascularization.

Two cases could not avoid surgery (2 nephrectomies) because of complete section of the renal artery (1) and disruption of the pieloureteral union (1). One case required haemofiltration in relation to rhabdomyolysis.

Conclusion: In our experience VIR is a valuable diagnostic and therapeutic procedure for the management of paediatric trauma patients, with high effectiveness and a low complication rate.
Aims of the Study: While distal and shaft forearm fractures are frequent and thus frequently analysed, proximal forearm fractures and especially those affecting both bones in the proximal epi-metaphysary region are seen less often in the ER and in literature. Only one larger series has been reported so far (Nenopoulos et al., J Pediatr Orthop 2009, including patients treated from 1984-1999). No data on patients treated by modern means of osteosynthesis such as elastic stable intramedullary nailing (ESIN) are available. Were therefore analysed treatment associated with this injury pattern in a pediatric university hospital emergency room population over a period of 11 years.

Method: From 2004-2014, among > 500 fractures affecting the proximal forearm of children of < 16 years, 100 combined fractures of the radius and ulna were identified by retrospective review of all elbow and forearm x-rays performed in a single institution. Respective fractures were classified as effecting either the radial head and olecranon (type 1), the radial head and proximal metaphysary region of the ulna distal to the coronoid process (type 2), or presenting as monteggia equivalents (proximal ulnar bowing or fracture with radial head subluxation/luxation, type 3).

Results: Patient distribution, gender, age, and operative treatment are displayed below.

<table>
<thead>
<tr>
<th></th>
<th>type 1</th>
<th>type 2</th>
<th>type 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>total number</td>
<td>50</td>
<td>15</td>
<td>35</td>
</tr>
<tr>
<td>male:female</td>
<td>20:30</td>
<td>6:9</td>
<td>21:14</td>
</tr>
<tr>
<td>Age</td>
<td>6.9 ± 2.5</td>
<td>6.0± 2.4</td>
<td>5.2 ± 2.1</td>
</tr>
<tr>
<td>right:left</td>
<td>21:29</td>
<td>5:10</td>
<td>16:19</td>
</tr>
<tr>
<td>OR procedures total</td>
<td>21</td>
<td>3</td>
<td>16</td>
</tr>
<tr>
<td>radius ESIN/open</td>
<td>15/1</td>
<td>1/0</td>
<td>0/0</td>
</tr>
<tr>
<td>ulna ESIN/open</td>
<td>0/13</td>
<td>2/0</td>
<td>13/3</td>
</tr>
<tr>
<td>metal removed (d)</td>
<td>83 ± 35</td>
<td>205 ± 15</td>
<td>94 ± 39</td>
</tr>
</tbody>
</table>

Children of 5-7 years were affected most frequently, with more (53 %) females and more (58 %) left arms involved. 39/100 required an operative procedure including 16 ESIN of the radius, 15 ESIN of the ulna, 1 open operation of the radial head and 15 open ulna tension wire fixations and 1 external fixator to the ulna (in some patients both bones were operated on).
Conclusion: A simple classification of combined proximal forearm fractures is proposed in which each fracture type represents with a typical appearance and standard treatment. As compared to literature, children of the current series were younger (6 vs. 8 years), received more operations (39 % vs. 22 %) while open operations of the radius (1/100 vs. 10/45) were reduced to a minimum by the use of ESIN. We conclude that up to date treatment of combined proximal forearm fractures is increasingly operative and should include ESIN to avoid open access to the radial head.
Aims of the Study: The aim of the study was to present clinical, diagnostic and pathological findings in children with rare diaphragmatic rupture after blunt trauma.

Method: Retrospective analysis of 4 patients with diaphragmatic rupture was particularly focused on diagnostic tools, surgical procedures and outcome. The study followed rules of the Institutional Ethical Committee.

Results: There were 3 girls and one boy, age at presentation ranged from 6 to 11 years. Car accident was the cause of injury in all cases. All patients were sitting in the back seats and were using safety belts. All patients were intubated and ventilated, and were admitted to the intensive care unit. There were two patients with diaphragmatic injury on the right side and two on the left. Diagnosis was established by computed tomography on admission in cases of the left sided injury. Diagnosis was delayed in right sided cases, in which the diaphragmatic rupture was masked by artificial ventilation. The diagnosis was confirmed at the time of weaning off ventilatory support on 3rd day resp. 14th day after injury. Diaphragmatic injuries were associated with pulmonary contusion in 100% of cases, splenic injury (50%), renal contusion (50%), multiple fractures (50%), pericardial rupture (25%), liver contusion (25%). The size of the rupture ranged from 10 to 15 cm. Closure of diaphragmatic rupture was performed via subcostal laparotomy with interrupted nonabsorbable stitches. Associated injuries required thoracic drainage of the affected side in all cases, osteosynthesis of fractures in two, splenectomy in one patient. The postoperative course was uneventful in all cases except one who required thoracoscopic drainage for recurrent fluidothorax. The follow up period ranged from 12 to 72 months and no recurrence of posttraumatic diaphragmatic hernia was observed.

Conclusion: The traumatic rupture of the diaphragm is rare and diagnosis can be delayed. Imaging modality could miss the diagnosis particularly in right sided injury on ventilatory support.
Aims of the Study: Gonadal histology (biopsy or gonadectomy) is required in selected DSD patients with abnormal gonadal development. The existence of a Y chromosome is associated with a higher risk of developing germ cell tumors as gonadoblastoma. The aim of our study was to analyze the spectrum of gonads of these group of patients and assess the incidence of germ cell tumors.

Method: A prospective and observational study of DSD patients who underwent gonadal surgery. Age, sex assigned, scale of external masculinization (EMS), karyotype, molecular analysis, surgical approach and pathology of the gonads were analyzed. Patients were divided into 3 groups: chromosomal dysgenesis (G1); 46XX gonadal dysgenesis; (G2) and 46XY gonadal dysgenesis (G3). More than half of the gonads were intrabdominal and were treated laparoscopically using 3 or 5 mm instruments. All streak gonads were removed, avoiding previous, as it has a 25 to 50% chances to develop a germ cell neoplasm and there is a possibility of an in situ tumor at the time of the procedure. We always waited for the result of biopsy before removing any other gonad than a classical streak. An inguinal approach was indicated in patients with palpable gonads. We still prefer a laparoscopic approach in most of them as it enables not only better visualization of potential Mullerian structures but also allows for treatment of a patent peritoneal sac, when removing the gonads, with better cosmetic results.

Results: In total 94 patients with a mean age of 56.42 months (range, 2-216) were analyzed. Forty eighth patients (19 with a Turner syndrome) with a mean age of 105 months (2-216) were included in G1. The karyotype was 45X0/46XY in 87.5% of them. Male sex was assigned in 19 , with a mean of 7.26 EMS (1-10). Histological analysis of 89 gonads was completed identifying 52 streak gonads, 32 dysgenetic testes and 5 ovotestes. Six GCT were found in 4 patients.

Fifteen patients with a mean age of 27.6 months (2-180) were included in G2. Male gender was assigned to 6 with a mean EMS of 6.82 (range, 4-8.5). Twenty nine gonads were analyzed: 10 ovotestes, 15 dysgenetic testes and 4 ovaries. Bilateral gonadoblastoma was found in a 6 month patient with bilateral ovotestes. Mean age of the 31 patients in G3 was 69.71 months (5-192). Five of them had an SF-1 NR5A mutation, 6 a WT1; 6 a complete and 3 a partial androgen insensitivity syndrome. A new mutation in the SRY (p.MET64VAL) gene was identified in 2 sisters. Male gender was assigned in 10 with a mean EMS of 4.52 (range, 1-10). Fifty nine gonads were analyzed, identifying 41 dysgenetic testes,
10 streak gonads and 8 testes, Eight GCT were found in 5 patients (16%) (7 in streak gonads and 1 in a dysgenetic teste).

**Conclusion:** DSD patients with gonadal dysgenesis have a wide variability. The incidence of gonadoblastoma is not negligible in patients 46XY, and even feasible in 46XX. The incidence of GCT was 8.3; 6.6 and 16% in G1 2 and 3 respectively. Early histological analysis and monitoring of these patients is mandatory. To our knowledge this is the first report of bilateral gonadoblastoma in ovotestes at a very early age.
Urology
SC-UR-0057

RANDOMIZED CONTROLLED TRIAL COMPARING DOUBLE-J STENT AND EXTERNAL URETERIC TRANS-ANASTAMOTIC STENT AMONG CHILDREN UNDERGOING PYELOPLASTY

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Aims of the Study: To compare long term double-J (DJ) stent vs short term external ureteric trans-anastamotic stent and a nephrostomy in children undergoing pyeloplasty for pelviureteric junction obstruction (PUJO).

Method: A randomized controlled trial that included all patients of PUJO undergoing Anderson-Haynes pyeloplasty from November 2011 through June 2014. Fifty patients were randomized to External stent(ES) or DJ stent group(DJ). Pre-operative and Post-operative(at 3 and 6 months) ultrasound was compared with respect to antero-posterior(AP) diameter of pelvis, Society of Fetal Urology(SFU) grade and differential renal function(DRF) on renal dynamic scan. Post-operative complications, duration of hospital stay and cost of the treatment were also compared.

Results: Twenty-five children were included in each arm of the study. The age at pyeloplasty was 7 days-144 months(median 44 months). 35 of 50 patients(70%) had a decrease in hydronephrosis, 19(76%) in ES and 16(64%) in DJ(p=0.538). Relative risk(RR) of worsening of hydronephrosis was 1.5 times greater in DJ[95CI 0.63–3.59; p=0.355]. Among those with pre-operative SFU grade 4, significantly more number of patients in ES showed reduction in hydronephrosis as compared to DJ[RR 2.5, 95CI 0.92-6.73; p = 0.05]. The RR for deterioration in DRF in DJ as compared to ES was 1.25(95CI 0.74 - 2.10, p = 0.395). Post-operative complications occurred in 9(36%) in ES and 8(32%) in DJ[RR 0.90, 95CI 0.40-1.928; p=0.765]. Mean duration of stay in ES was 9.16 days compared to 7.04 in DJ. The difference in the duration of stay was significant (p=0.004). Average cost of treatment for a patient in ES was 335.60 compared to 1356.00 Indian Rupees in DJ.

Conclusion: DJ stent had no significant benefit over External stent in terms of surgical results and complications. Patients in DJ group had shorter duration of hospital stay but had significantly increased treatment cost.
LAPAROSCOPIC, RETROPERITONEOSCOPIC AND ONE-TROCAR ASSISTED PYELOPLASTY IN CHILDREN: REVISION OF LITERATURE AND LESSON LEARNED FROM 1570 CASES.

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¹Department of Paediatric, Gynecological, Microbiological and Biomedical Sciences - University of Messina (ITALY), Unit of Paediatric Surgery, Messina, ²Department of Woman, Child and Urological Diseases, Unit of Paediatric Surgery, Bologna, Italy

Aims of the Study: Anderson-Hynes (AH) pyeloplasty is considered the gold-standard for treatment of ureteropelvic junction obstruction (UPJO). Even if this technique have been classically performed by the open approach, mini-invasive AH pyeloplasty is gaining ground and, nowadays, laparoscopic (LP), retroperitoneoscopic (RP) and one-trocar assisted pyeloplasties (OTAP) are considered safe and feasible also in children. Aim is to compare LP, RP and OTAP, analysing, firstly, the incidence of intra-operative (IO) and post-operative (PO) major complications, the percentage of recurrence; secondly, the incidence of minor complications, the conversion rate to open surgery, the time of procedure and the mean age of patients (pts).

Method: We reviewed International literature from 2000 to 2014 considering LP, RP and OTAP in paediatric age, including papers in English and presentation at International or National Congresses. N. of cases, mean age, operation time, IO and PO complications, incidence of recurrence have been considered. IO complications were classified according to Satava, while the PO ones according to Clavien-Dindo. Grade III IO and ≥ C3b PO complications were considered as major. The incidence for each technique of IO or PO major and minor complication, of recurrence and conversion to open surgery were compared using the Comprehensive Meta-Analysis V.2.0 software. The mean age, the operation time were compared using a student t-test. A value of p < 0.05 was considered as significant.

Results: Thirty-five papers and two presentations were considered eligible and included. On 1570 paediatric pts, 917 pts underwent LP, 273 and 380 RP and OTAP, respectively. Two major IO complication occurred in LP. Sixty-two major PO complications on 917 procedures were described in LP group (6.8%), 27 on 273 RPs (9.9%) and 13 on 380 OTAPs (3.4%). Statistical analysis showed a significant lower incidence of ≥ C3b in OTAP as compared with LP (p=0.021) and RP (p = 0.01). With regard to minor PO complication, 70 occurred on 917 LP (7.6%), 14 on 273 RP (5.1%) and 3 on 380 OTAP (0.8%). Statistical analysis demonstrated a significant lower incidence of < C3b in OTAP as compared with the LP (p=0.001) and RP (p=0.003). Twenty-four recurrences was showed on 917 LP (2.6%), 9 on 273 RP (3.3%) and 380 OTAP (2.3%), without any difference. Conversion rate to open surgery was necessary in 9 LP (1.0%), 10 RP (3.7%) and 17 OTAP (4.5%), significant lower in LP as compared with RP (p=0.001) and OTAP (p=0.004). Operation time and mean age.
was significant lower in OTAP (105min and 28mths) than in LP (175min and 65mths) and RP (153min and 80 mths) (p = 0.001).

**Conclusion:** Our study confirmed that LP, RP and OTAP in children are effective, with a success rate > 95%. The comparison between the three techniques showed a lower incidence of major and minor complications in OTAP and a less operation time. Differently, OTAP has been adversely affected by the higher incidence of conversion to open surgery than LP, but none case was for urgent reason. Moreover, the study pts were not matched for age, with youngest population in OTAP group. It could suggest that the age is considered an advantage for video-assisted technique (less size, easier surgical mobilisation) and a detriment for LP and RP (less size, smaller working space). Our data suggest that OTAP, when applicable, is the most suitable mini-invasive technique, ensuring similar cosmetic results and less incidence of complication.
ACELLULAR COLLAGEN GEL TUBES IN AN IN VIVO RABBIT MODEL FOR URETHRAL REGENERATION, A LONG TERM FOLLOW UP.

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Aims of the Study: Actual surgical procedures using existing autologous tissues for the treatment of congenital malformations or injuries of the urethra are associated, in severe cases, with frequent post-operative complications. Tissue engineered collagen tubes are a promising alternative. As demonstrated in our previous work, there is spontaneous regrowth of urothelium in in vivo rabbit models, using high density collagen gel tubes.

Method: A now modified fabrication procedure results in collagen gel tubes with enhanced mechanical properties; thus increasing their resistance and simplifying the handling of the graft. Leak tests, burst pressures, stress-strain and suture resistance measurements proved a better resistance of those collagen tubes compared to our previous technique. These tubes were used as urethral grafts and surgically placed after a subtotal excision of the urethra. The graft was interposed between the prostatic urethra and the very distal and brittle remaining native urethra. This subtotal urethral replacement (more than 80% of the total length) was done in 20 male New Zealand white rabbits. The constructs were all acellular and no catheter was placed postoperatively. The animals were evaluated at 1, 3, 6, and 9 months by contrast voiding cystourethrography and histological examination.

Results: This multicentric study revealed spontaneous regrowth of urothelial cells and smooth muscle cells (SMC) in all grafts and reduced severe postoperative complications (1 obstructive stenosis).

Conclusion: Those compressed collagen gel tubes are easy to handle and adaptable to current surgical techniques to establish the continuity between tubular anatomical structures. Therefore, they are suitable for clinical applications and may be an alternative to the existing surgical treatment.
LOWER URINARY TRACT SYMPTOMS AMONG CHILDREN WITH ANORECTAL MALFORMATIONS WITH RECTO-PERINEAL FISTULAS

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Aims of the Study: The aim was to describe the frequency of lower urinary tract symptoms (LUTS) among females and males with anorectal malformations with perineal fistulas (ARM-P) and compare it genderwise with healthy controls.

Method: At two tertiary centres of paediatric surgery in two different countries all children 4-12 years old operated on because of ARM-P during 2001-2012 were followed up. The study was retrospective regarding urinary tract malformations (UTM) diagnosed either clinically (hypospadias) or with ultrasonography or voiding urethro cystography. The study was cross-sectional regarding LUTS as defined by the International Children’s Continence Society 2006 [1] with respect to urinary incontinence, enuresis, bladder emptying and lower urinary tract infections. The controls were collected in the catchment area of one of the centres. The statistical analyses were performed by a statistician. The study was approved by the Regional Research Ethics Committee (registration number 2010/49) and the institutional board at the hospital.

Results: After a dropout of 9 patients, 24 females and 33 males with ARM-P, median age 8(4-12) years and equal follow-up time, were studied. Four (17%) females had 7 UTM (bilateral dysplasia and bilateral multicystic kidneys, 1 duplex, 1 vesico ureteral reflux (VUR), ectopic ureter) and 8 (24%) males had 12 UTM (4 hypospadias, 3 VUR, 1 bladder diverticula, unilateral duplex, 2 single kidney, 1 unilateral dysplasia). Operations on the urinary tracts had been performed in 1 female (reimplantation of ectopic urether) and six procedures in 4 (12%) males (1 nephrectomy, 1 kidney transplantation, 4 hypospadias). One female and one male with ARM-P had intermittent catheterizing. Sacrum defects were present in 4 (17%) females and 3 (9%) males. Among patients with ARM-P there were no gender differences in comparisons of urinary incontinence, enuresis, bladder emptying, infections or need of medication. Both females and males with ARM-P with concomitant UTM reported urinary infections more frequently (3/4 females and 4/8 males) compared to those with normal urinary tracts (2/20 females and 0/25 males) (p= 0.018 and 0.002). In comparison between patients and controls, difficulties with bladder emptying were more frequent among the female patients (3/24) than among the female controls (0/55) (p=0.025). The frequency of urinary tract infections was higher among the female patients (5/24) compared with the controls (1/55) (p=0.009) and more females and males with ARM-P (5/24 and 5/33) than controls (1/55 and 2/110) were prescribed urinary prophylaxis (p=0.009 and 0.007). Males with ARM-P reported higher frequency of daytime urinary incontinence (4/33) compared with the male controls (3/110) (p=0.049).
**Conclusion:** There were no gender differences in LUTS in patients with ARM-P. Patients with concomitant UTM had more urinary infections than patients without UTM and both females and males with ARM-P reported more urinary infections than healthy controls.

WHAT HAPPENS WITH THE ILEAL EPITHELIUM FOLLOWING ILEOCYSTOPLASTY IN RATS? – HISTOLOGICAL FINDINGS

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1Department of Paediatrics, 2Department of Pathology, 3Department of Anatomy, University of Pécs, Pecs, Hungary

Aims of the Study: Urothelization of the small intestinal mucosa following ileocystoplasty probably has a role in the genesis of malignant transformation. Little is known about whether the urothelial epithelium migrates to the bowel mucosa or if there are other mechanisms at play. Our main objective was to investigate the histological changes – focusing on mucosal alterations - in rats during a long-term follow-up.

Method: Surgeries were performed on young male Wistar rats (<3 months of age). Animals were divided into four groups. Each group contained 6 rats. Ileocystoplasties were performed and rats were followed for 12 (Group A), 18 (Group B) and 24 (Group C) months. Group D comprised the sham-operated (cystotomy) animals, were followed for 24 months. At the end of the follow-up, all the animals were sacrificed. The augmented bladder was harvested, sliced in mid-sagittal plane, examined macroscopically. Samples were then placed in formaldehyde and fixed for one week. The samples were then sectioned towards the anastomotic line to examine the native bladder, the ileal patch and the anastomotic line between them. Routine HE staining was performed. An independent anatomist and pathologist (both blinded) evaluated the sections.

Results: In Group A (12 months) the urothelium became thicker with more cell layers at the anastomotic line. On the adjacent villi of the ileal patch, epithelium was replaced with varying amounts of urothelial cells. At the anastomotic line some crypts seemed to be completely replaced with nests of urothelial cells. On the bowel mucosa, distally from the anastomosis, urothelium replaced smaller areas of the villi. Furthermore it was found only on the tip of the villi. In one animal a stone was found in the augmented bladder. In this rat histology revealed squamous cell metaplasia in the native bladder.

In Group B (18 months) apart from urothelial hyperplasia, the same histological findings were found as in Group A. In Group C (24 months) thickened, irregularly covered bowel mucosa was discovered in one case, on the ileal patch, representing an invasive transitional cell carcinoma. In another rat polypous overgrowth was detected, microscopically a hyperplastic urothelial epithelium was found at the anastomotic line. The remaining 4 animals showed similar histological appearance as in Group A. The urothelium progressed to replace the majority of columnar epithelium. Chronic inflammation in the mucosa was observed in one animal.

In Group D (control), no abnormalities were detected.
Conclusion: According to our findings, the urothelial replacement of the ileal epithelium probably was caused by the transformation of the columnar cells into urothelial cells instead of migration in rats. Evolution of this mechanism starts at the tips of the villi and moves deeper with time. This change happens first at the anastomotic line and then progresses toward to the bowel epithelium from there. This pattern implies that this mechanism requires continuous contact with the urine and proximity of urothelial cells. Further investigation of the epithelial replacement may help us for better understanding the basics of malignant transformations after urinary bladder augmentation with small intestine.
LONG TERM RESULTS OF THE MITROFANOFF PROCEDURE. A MULTICENTRIC SURVEY

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¹Traslational Medical Sciences, University of Naples "Federico II", Naples, ²Pediatric Surgery, San Bortolo Hospital, Vicenza, Italy, ³Pediatric Surgery, Hôpital Necker Enfants Malades, Paris, ⁴Pediatric Surgery, CHU Centre Hospitalier Universitaire de la Réunion, La Réunion, France

Aims of the Study: We retrospectively reviewed the records of all patients who underwent appendicovesicostomy (Mitrofanoff procedure) between 2000 and 2010 in 3 European centers of pediatric surgery and urology.

Method: Between 2000 and 2010, 35 Mitrofanoff appendicular channels were performed on 8 boys and 27 girls (average age 13 years). The procedure was part of reconstructive procedure in patients with neurogenic bladder (n=22), exstrophy-epispadias (n=3), posterior urethral valve (n=8) and other disease (n=2). Additional procedures included augmentation cystoplasty (n=9) and bladder neck procedure (n=4). Mitrofanoff appendicular channels were performed using open surgery in 34 cases (97%). In all patients the appendix was catheterized with 12 Fr catheter and implanted posterolaterally into the native bladder or the intestinal segment of the augmented bladder with antirefluxing tunnel of 3-4 cm in length. The site of the stoma was the umbilicus in 12 cases (34%) and the right lower abdomen in 23 cases (66%), using a V-flap combined with spatulation of the conduit in 34 cases (97%).

Results: All patients had a Mitrofanoff catheter in situ for 3-4 weeks with administration of oxybutynin (dosage: 0.3 mg/kg) to reduce bladder spasms in 20 cases (57%); after removal of Mitrofanoff catheter all patients performed Clean Intermittent Catheterization (CIC) every 3-4 hours during the day with prescription of antibiotic prophylaxis. With a median follow-up of 7 years (4 to 15 years) complications rate was 54 % (19 cases): stomal stenosis in 9 cases (47%), dehiscence in 3 cases (16 %), false passage in 3 cases (16%), conduit necrosis in 2 cases (11%), conduit fibrosis in 1 case (5%), conduit volvulus around the peduncle in 1 case (5%). Re-operations rate was 34 % (12/35 cases). At follow-up a continent and catheterizable conduit was achieved in 32 patients (92%), with a good aesthetic outcome in all patients.

Conclusion: Mitrofanoff procedure remains a complex procedure to perform but very useful in pediatric patient with neurogenic bladder. With a long term follow-up the complications rate is higher than 50 % with a redo surgery rate of 34 %. However at a long term follow-up the Mitrofanoff channel was continent and catheterizable in 92% of patients of our series. A standardization of the technique together with the use of minimally-invasive surgery probably may improve the results in the next future.
Aims of the Study: Urinary tract infections (UTIs) and vesico-ureteral reflux (VUR) are assumed to predispose children to renal damage. Awareness on the significance of VUR and the possibilities to reduce UTI recurrence and renal damage has warranted guidelines on which patients should undergo imaging after UTI. An authoritative guideline has been issued by the American Academy of Pediatrics (AAP). We assessed the applicability of the AAP guidelines on a subpopulation of patients with UTI, 2–24 month old children with febrile UTI.

Method: The records of 394 children aged 2-24 months with their first UTI were reviewed. Data were recorded on the indications for renal and bladder ultrasonography (RBUS) and voiding cystourethrography (VCUG) according to the AAP guidelines, RBUS results, VCUG results, use of anti-microbial prophylaxis (AMP), anti-reflux procedures and other urological treatment and UTI recurrence.

Results: 344 patients had an indication for RBUS. RBUS results were abnormal in 87, including 53 with urinary tract dilation. An unnecessary RBUS would have been avoided in 43 patients. 7 patients with an abnormal RBUS would not have undergone RBUS. 126 patients had an indication for VCUG. VCUG was performed in 206 patients; VUR was found in 72 patients, including 36 with high-grade VUR. An unnecessary VCUG would have been avoided in 82 patients. High-grade VUR would have been missed in 6 patients. 5 patients would not have undergone surgery.

Conclusion: The AAP guidelines for imaging studies in children aged 2-24 months with febrile UTI should be followed.
RESULTS OF ORCHIDOPEXY IN CHILDREN WITH PRADER-WILLI SYNDROME

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Aims of the Study: Prader-Willi syndrome (PWS) is a rare (1:10,000) genetic condition affecting both males and females. Among other features the syndrome is characterized by cryptorchidism in 86-100% of cases, hypogonadism, delayed puberty and infertility. The aim of the present study is to appraise the results of orchidopexy in this selected population of children.

Method: Following ethical approval (#DA005-2013-06), a retrospective review of children with PWS treated for undescended testes at a single institution over a 20-year period (1994-2014) was performed. Patients were identified from a National PWS registry and reviewed at a special follow-up PWS clinic. Data were collected from electronic and hard copies records and reported as median (range).

Results: Thirty-three children (1-17 years) were identified (Figure). Co-morbidities were present in 22 (66%) and 15 (45%) were on growth-hormone therapy. Twenty-seven (81%) required orchidopexy. Thirteen (48%) underwent a bilateral procedure for a total of 40 orchidopexies. A 2-stage Fowler-Stephens procedure was required in 2 (7%) testes. At surgery hypotrophic testes were documented in 6 (22%) patients. Age at orchidopexy was 1.4 years (0.5-5.5). Age at F-U was 7.2 years (1.7-17). Length of follow-up is 3.5 years (0.4-14). Findings at follow-up are reported in the Table. Twenty-four (60%) testes were palpable in the scrotum. Four (15%) patients over 16 years of age had testosterone levels below normal limits after orchidopexy.

<table>
<thead>
<tr>
<th>FINDINGS AT FOLLOW-UP</th>
<th>NUMBER (%)</th>
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</thead>
<tbody>
<tr>
<td>Testis in scrotum</td>
<td>24 (60)</td>
</tr>
<tr>
<td>Hypotrophic testis</td>
<td>8/24 (33)</td>
</tr>
<tr>
<td>Testis in inguinal canal</td>
<td>10 (25)</td>
</tr>
<tr>
<td>Non-palpable testis</td>
<td>4 (10)</td>
</tr>
<tr>
<td>Atrophic testis</td>
<td>2 (5)</td>
</tr>
<tr>
<td>Redo-orchidopexy</td>
<td>7 (17.5)</td>
</tr>
</tbody>
</table>
Conclusion: This is the first study evaluating the results of orchidopexy in a large population of children with PWS. Our findings suggest that these children have sub-optimal results compared to general population: 40% of testes are not in the scrotum; 33% of testes in the scrotum are hypotrophic; 17.5% require redo-orchidopexy. This information should be taken into consideration for patients' management and pre-operative parents' counselling.
**EVALUATION OF THE DISTAL URETERIC STUMP AFTER NEPHRO-URETERECTOMY IN CHILDREN. A COMPARISON BETWEEN LAPAROSCOPY AND RETROPERITONEOSCOPY.**

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†Traslational Medical Sciences, University of Naples "Federico II", Naples, Italy

**Aims of the Study:** The management of a poorly functioning kidney in children due to a VUR or UPJO consists in a total nephrectomy with ureterectomy. The complete removal of the ureter minimizes the risk of future morbidity associated with the distal ureteric stump (DUS), including febrile UTI, lower quadrant pain and haematuria, the so-called ureteric stump syndrome. To assess the outcome of the distal ureteric stump (DUS) after nephrectomy with ureterectomy we analyzed our recent experience of nephrectomy via retroperitoneoscopy and laparoscopy.

**Method:** The records of 21 patients (median age 2.7 years, range 11 months -10 yrs) who underwent nephrectomy via laparoscopy (11) or retroperitoneoscopy (10) with subtotal ureterectomy were reviewed retrospectively for symptoms caused by DUS and their management. The average of the follow-up was 5 years. Nephrectomy was undertaken for a poorly functioning dysplastic (4), scarred for VUR (10) or hydronephrotic (7) kidney. The patients were evaluated using US to check DUS length and clinically to evaluate symptoms due to a symptomatic DUS.

**Results:** The length of DUS after laparoscopic nephrectomy, measured using US, was statistically significant shorter (range 3-5 mm) compared to residual stump after retroperitoneoscopy (range 2-5 cm) [p<.001]. Laparoscopic patients were all asymptomatic. After retroperitoneoscopy in 2 cases a VUR was present on the residual DUS with recurrent UTIs and after a confirmation using voiding cystography a redo surgery was performed to remove the residual DUS.

**Conclusion:** The risk to leave a long DUS in pediatric patients who undergo nephrectomy and ureterectomy is a truly problem. Long ureteric stumps may cause symptoms. Considering that laparoscopy permits to remove a longer part of the ureter arriving near the bladder dome, it is preferable, in children with non functioning kidney due to VUR, to perform always a laparoscopic rather than a retroperitoneoscopic nephrectomy. In case of symptomatic DUS with recurrent UTIs a redo surgery may be indicated to remove the residual stump as happened in our series in 2 cases after retroperitoneoscopic nephrectomy.


**Aims of the Study:** Endoscopic instillation of inert substance today is the first line treatment for vesicoureteral reflux. Deflux® is a sterile, highly viscous gel of dextranomer microspheres in a carrier gel of non-animal hyaluronic acid. On the other side, Vurdex® consists of positively charged dextranomer-based micro-particles, that stimulates patient own tissue to regenerate collagen at implantation site, and cross-linked hyaluronic acids.

The objective of this study was to compare the clinical efficacy of two similar tissue bulking agents, Deflux® and Vurdex®, used for a treatment of vesicoureteral reflux in our institution.

**Method:** The case records of 104 children, treated endoscopically for primary vesicoureteral reflux from January 2010 to January 2015, were retrospectively reviewed. Most of the patients were treated with Deflux® until 2012, when using of Vurdex® was introduced. Since then, Vurdex® was primarily used in the treatment of vesicoureteral reflux.

Exclusion criteria were patients with secondary reflux due to neurogenic bladder, duplicated refluxing ureters, primary operated patients and patients operated after first or second injection. The children's mean age was 4.8 years (range, 0-13 years). Mean overall follow-up was 2 years (range 1 – 4 years). Mean hospital stay was one day (range, 1–3 days).

**Results:**

Endoscopic treatment of vesicoureteral reflux using Deflux® (group I) was performed in 65 children (106 ureters; grade I: 14; grade II: 30; grade III: 40; grade IV: 19; grade V: 3). Male to female ratio was 15:50. There were 24 patients with unilateral and 41 patients with bilateral VUR. After first injection of Deflux® success was achieved in 74 ureters (69,8%), after second injection in 91 ureters (85,8%) and after third injection in 99 ureters (93,3%).

The same procedure using Vurdex® (group II) was performed in 39 children (58 ureters; grade I: 12; grade II: 17; grade III: 20; grade IV: 7; grade V: 2). Male to female ratio was 9:30. There were 20 patients with unilateral and 19 patients with bilateral reflux. After first injection of Vurdex® success was achieved in 43 ureters (74,1%), after second injection in 52 ureters (89,6%) and after third injection in 55 ureters (94,8%). The costs of the single injection (1 ml) were significantly lower when the Vurdex® was used (£ 481.77) compared to the Deflux® (£ 708.08).
Table:

<table>
<thead>
<tr>
<th></th>
<th>Ureters</th>
<th>Succes after 1st injection</th>
<th>%</th>
<th>Succes after 2nd injection</th>
<th>%</th>
<th>Succes after 3rd injection</th>
<th>%</th>
<th>Unsucces</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Deflux®</strong></td>
<td>106</td>
<td>74/106</td>
<td>6</td>
<td>9</td>
<td>8</td>
<td>91/106</td>
<td>8</td>
<td>99/106</td>
<td>9</td>
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<td></td>
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<tr>
<td><strong>Vurdex®</strong></td>
<td>58</td>
<td>43/58</td>
<td>7</td>
<td>4</td>
<td>1</td>
<td>52/58</td>
<td>8</td>
<td>55/58</td>
<td>9</td>
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<tr>
<td><strong>p</strong></td>
<td>0.557</td>
<td>0.485</td>
<td></td>
<td>0.714</td>
<td></td>
<td></td>
<td></td>
<td>0.71</td>
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</tbody>
</table>

**Conclusion:** Overall success rate for patients treated with Deflux® was 93.3% (99/106) and for patients treated with Vurdex® 94.8% (55/58). No significant difference in success rates between two groups was observed (p=0.714). However, Vurdex® has an advantage because of the significantly lower price, but with same treatment results as Deflux®.
OUTCOMES OF PEDIATRIC LIVING DONOR KIDNEY TRANSPLANTATION: A SINGLE-CENTER EXPERIENCE

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¹Pediatric Urology, ²Pediatric Nephrology, ³Urology, Hospital Virgen del Rocío, Sevilla, Spain

**Aims of the Study:** Renal transplantation is the treatment of choice for children with end-stage renal disease (ESRD) offering advantages of improved survival, growth potential, cognitive development and quality of life. The aim of our study is to compare the outcomes of living donor kidney transplantation (LDKT) versus cadaveric donor kidney transplantation (CDKT) performed in children at a single center.

**Method:** Retrospective chart review of pediatric patients who underwent kidney transplantation from December 2005 to December 2014 at our institution was performed. Demographic data, etiologies of ESRD, pretransplant dialysis, family relationship of the living donors, surgical technique, complications, immunosuppressive drugs, trends in serum creatinine, graft and patient survival rates were assessed. All data were analyzed using the Statistical Package for Social Sciences (SPSS version 19.0 software).

**Results:** 91 renal transplants were performed in children during the last 9 years in our center. Baseline demographic data and etiologies of ESRD were similar between the LDKT and the CDKT groups. 31 cases (38.27%) were LDKT and in 96.7% of the cases the graft was obtained through a laparoscopic approach. For 74 patients (28 LDKT and 46 CDKT) was the first transplant. The family relationship of living donors included the mother in 19 cases (61.2%), the father in 9 (2.7%) and other relatives in 3 (0.9%). Donors median age was 39 years for LDKT and 35 years for CDKT, and the recipients mean age was 9.4 years for LDKT and 10.4 years for CDKT. 34 receptors (37.4%) weighted less than 25 kg, 15 of them were LDKT. Preemptive transplantation was done in 25 patients, being significantly higher in LDKT (13 cases, p<0.009). LDKT group had statistically significant lower cold ischemia times than CDKT one (mean: 5 versus 17 hours, respectively, p<0.001), while warm ischemia did not show significant differences between both groups. Complication rate was 1.4% for LDKT and 7.2% for CDKT. The use of immunosuppressive drugs was equivalent in both groups. Mean serum creatinine and creatinine clearance were lower in LDKT (0.97 mg/dl and 104 cc/min/1.73m², respectively). All children increased weight and height after transplantation (p<0.001). Patient survival rate was 100% for LDKT and 98.3% for CDKT and graft survival rate was 96.7% for LDKT and 80% for CDKT at a year and 5 years.

**Conclusion:** Our program of pediatric kidney transplantation has achieved optimal patient and graft survival rates with low rate of complications. Living donor pediatric kidney transplants have higher patient and better graft survival rates than cadaveric donor kidney transplants. Implementing kidney donation have positively affected the results of our pediatric kidney transplantations.
INCREASED MMP-7 EXPRESSION IN BILIARY EPITHELIUM AND SERUM UNDERPINS NATIVE LIVER FIBROSIS AFTER SUCCESSFUL PORTOENTEROSTOMY IN BILIARY ATRESIA

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1Pediatric Surgery, Children's Hospital, University of Helsinki and Helsinki University Hospital, 2Department of Pathology, 3Oral and Maxillofacial Diseases, University of Helsinki and Helsinki University Hospital, Helsinki, Finland, 4Division of Periodontology, Department of Dental Medicine, Karolinska Institutet, Huddinge, Sweden, 5Department of Surgery, 6Research Program Unit, Translational Cancer Biology, 7Pediatric Nephrology and Transplantation, University of Helsinki and Helsinki University Hospital, Helsinki, Finland

Aims of the Study: The molecular mechanisms underlying progressive liver fibrosis and injury following successful first line surgical treatment of biliary atresia (BA) remain unclear. Our aim was to address hepatic gene and protein expression and serum levels of matrix metalloproteinases (MMP) and their inhibitors after successful portoenterostomy (PE), and relate them to histological signs of liver injury, clinical follow-up data and biochemical markers of hepatic function.

Method: After ethical approval, liver biopsies and serum samples were obtained from 25 children after successful PE at median age of 3.3 years. Serum MMP concentrations were determined by ELISA. Hepatic gene expression of MMPs and TIMPs was analyzed using complementary DNA microarray combined with real-time reverse-transcription PCR. Liver expression of MMP-7, TIMP-1 and cytokeratin-7 was studied using immunohistochemistry. Control liver biopsies were obtained from patients undergoing surgery for cholelithiasis and donor livers.
**Results:** Following PE, median plasma conjugated bilirubin was 4 (interquartile range 2 - 7) µmol/l, histological cholestasis score was 0 (0 - 0) and Metavir fibrosis stage was 3/4 (2 - 4). Despite effective clearance of biochemical and histological cholestasis, BA patients showed markedly increased hepatic gene expression of **MMP-7** (29-fold, \( p < 0.001 \)), **MMP-2** (3.1-fold, \( p < 0.001 \)), **MMP-14** (1.7-fold, \( p = 0.007 \)), and **TIMP-1** (1.8-fold, \( p < 0.001 \)), when compared to controls (Table). Similar to a biliary epithelial marker cytokeratin-7, protein expression of MMP-7 localized in biliary epithelium of bile ducts and ductal proliferations and periportal hepatocytes. Biliary epithelial expression of MMP-7 was increased (\( p < 0.001 \)) in relation to controls. BA patients had 6-fold higher serum levels of MMP-7 (\( p < 0.001 \)), which correlated positively with hepatic **MMP-7** gene (\( r = 0.548, p = 0.007 \)) and protein expression (\( r = 0.490, p = 0.015 \)). Patients showed a positive correlation between MMP-7 staining of biliary epithelial cells and Metavir fibrosis stage (\( r = 0.581, p = 0.002 \)) and portal fibrosis grade (\( r = 0.574, p = 0.003 \)). Serum MMP-7 also showed significant predictive value for portal fibrosis with AUROC of 0.925 (CI 95%: 0.817 – 1.000, \( P = 0.008 \)).

**Table:**

<table>
<thead>
<tr>
<th>MMP-7</th>
<th>Patients</th>
<th>Controls</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum concentration (ng/ml)</td>
<td>14.4 (7.01 – 28.6)</td>
<td>2.23 (1.60 – 2.88)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Liver gene expression *</td>
<td>29.2 (6.11 – 70.3)</td>
<td>1.00 (0.80 – 1.90)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Biliary protein expression (grade 0 – 4)</td>
<td>3 (2 – 3)</td>
<td>1 (0 – 1)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

**Caption:** MMP-7 expression. Data are median (interquartile range). * Transcript levels are expressed after normalization to housekeeping genes and relative to control subjects using the ∆∆Ct method.

**Conclusion:** Our findings support an important role of altered hepatic expression of MMP-7 in the progression of liver fibrosis after successful PE and introduce a potential therapeutic target to pharmacologically extend native liver survival by inhibiting MMP-7 hyperactivity. Serum MMP-7 may be a valuable postoperative prognostic tool in BA.
BILIARY ATRESIA IN THE NETHERLANDS: THE EFFECT OF CENTRALISATION.

Mauri Witt1, Jim Wilde2,3, Henkjan Verkade4, Paul Peeters5, Jan Hulscher 1

1Paediatric Surgery, University Medical Centre Groningen, Groningen, 2Paediatric Surgery, Academic Medical Centre, Amsterdam, Netherlands, 3Paediatric Surgery, Hôpitaux Universitaires de Genève, Genève, Switzerland, 4Paediatric Gastroenterology and Hepatology, 5Hepatobiliary Surgery and Liver Transplantation, University Medical Centre Groningen, Groningen, Netherlands

Aims of the Study: In order to improve the outcome of patients with biliary atresia (BA) in the Netherlands, the treatment of BA patients has been centralised in 2012 to two specialised centres since 2012. After consultation of all paediatric surgical centres a dedicated team of paediatric surgeons and hepatobiliary/transplant surgeons was introduced. Kasai surgery was performed by a team consisting of at least one of the paediatric surgeons and one of the hepatobiliary/transplant surgeons.

The aim of this study was to analyse the early outcome of BA patients in the Netherlands after centralisation.

Method: All patients born between January 1st 1997 and January 1st 2015 who underwent Kasai surgery in the Netherlands were included. All data were obtained from the Netherlands Study group on Biliary Atresia Registry (NeSBAR). Kasai surgery before the age of 60 days and clearance of jaundice (bilirubin < 20 μmol/L) before (Cohort A) and after introduction of the dedicated multidisciplinary team (cohort B) were analysed. Postoperative treatment consisted of ursodeoxycholic acid and antibiotic prophylaxis. Low dose steroids were administered in all patients until December 2013.

Results: Between January 1st 1997 and January 1st 2015 a total of 170 patients underwent Kasai surgery in the Netherlands. Because 15 patients were lost to follow-up, a total of 155 patients were included. After centralisation 27 patients underwent surgery. During the transition period, 8 patients were not operated by the dedicated team. For this reason they were analysed in Cohort A. Cohort A consisted of 136 patients, cohort B contained 19 patients. The percentage of patients who underwent Kasai surgery before the age of 60 days was 52% in cohort A and 67% in cohort B (p=0.25). Clearance of jaundice increased from 26% in cohort A to 56% in cohort B (p=0.01). One patient has only recently been operated and is not included. In cohort B nine patients (47%) underwent liver transplantation. Mortality in cohort B was 11%.
Conclusion: Our first results show a remarkable improvement in surgical outcome of the Kasai procedure after centralisation of the treatment of BA to a dedicated multidisciplinary team. In the long term, this may lead to higher native liver survival and higher overall survival. There was no statistically significant difference in the percentage of patients who underwent Kasai surgery before the age of 60 days. Screening methods might enable us to perform surgery at an earlier age, thereby further improving outcome.

\textbf{Figure 1:} Clearance of jaundice rates in the Netherlands before and after the introduction of a dedicated team, as compared to several other Western countries.

**Hepato-biliary**

**SC-HE-0070**

**HIGH-DOSE STEROIDS IN BILIARY ATRESIA: THE EFFECT OF AGE.**

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**Aims of the Study:** The use of high-dose steroids as adjuvant therapy following Kasai portoenterostomy (KPE) for biliary atresia is controversial with even RCT’s showing conflicting results (1). We hypothesised that response was age-dependent and sought to quantify this effect.

**Method:** Single-centre prospective database. High dose adjuvant steroid (prednisolone starting 5 mg/kg/day: step-down 4 weeks, 103 mg/Kg total) (Jan. 2006 – June 2014). Infants were selected by being ≤70 days at KPE. Outcome was assessed by clearance of jaundice (<20μmol/L) and native liver survival and tested by Chi² and log-rank tests respectively. Infants were divided into sequential cohorts by age at KPE (e.g. ≤ 30, 30 – 39, 40- 49 days etc.). This allowed an individual cohort probability for the outcome statistic which could also be plotted as a cumulative probability (i.e. a cut-off) for various time points. Statistical analysis was carried out by an extended r x c analysis. P ≤0.05 was regarded as significant.

IRB approval was obtained for previous use of steroids in open-label trial.

**Results:** High dose steroid group consisted of 104 infants aged ≤70 days at KPE (range12 – 70). BA was classified as isolated (n = 80), cystic (n = 9), BASM (n = 9) and CMV IgM+ve (n = 6). Overall 70/104 (67%) cleared their jaundice. Age-cohort analysis showed a significant trend over time (P = 0.03) (Fig. 1). The median age in the group was 45 days and this divided the group into two equal cohorts. Furthermore there was a significant native liver survival advantage for younger infants (< 45 days versus >45 days, P = 0.05).

**Conclusion:** There is a marked difference in outcome dependent on age at KPE in infants using a high-dose steroid regimen.

**References:**

USEFULNESS OF A RECANALIZED UMBILICAL VEIN FOR VASCULAR RECONSTRUCTION IN PEDIATRIC HEPATIC SURGERY

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1Department of Surgery, Kanagawa Children's Medical Center, Yokohama, 2Department of Surgery, Ohno Central Hospital, Ichikawa, Japan

Aims of the Study: Pediatric hepatobiliary surgeons involve vascular dissections and reconstructions in the operation for portal hypertension, hepatic neoplasms, and liver transplantation. Vascular grafts are sometimes needed for reconstruction of the superior mesenteric vein (SMV), portal vein (PV), hepatic vein (HV), and inferior vena cava (IVC). Herein, we report our experience using a recanalized umbilical vein (rUV) and long-term outcome.

Method: Case 1-4: Four patients aged two to eleven years with extrahepatic PV obstruction (EHPVO) underwent mesenterico-/porto-left portal vein (meso-Rex) bypass surgery using a short rUV in the round ligament as an anastomotic orifice connected to the left PV. An iliac vein graft was interposed between the rUV and the splenomesenteric confluence or the SMV to create the bypass.

Case 5: A neonate aged 2 days with huge liver tumor (mesenchymal hamartoma) underwent right hepatectomy together with partial resection of the PV. As a UV in the round ligament was still patent, it was directly anastomosed to the proximal PV stump to restore portal perfusion.

Case 6: A patient aged 6 years underwent living donor liver transplantation for recurrent hepatoblastoma in the residual liver after left trisegmentectomy. Because of tumor involvement with the hepatic IVC, it was resected and reconstructed by transposition of the infrahepatic IVC and interposition of the rUV which was obtained from the round ligament of the liver graft.

Periodic blood test, imaging studies (Doppler ultrasonography, contrast computed tomography) and esophagogastric endoscopy (Case 1-4) were performed during the follow-up.

Results: Sufficient flow through the rUV was achieved in all the patients and its patency was maintained without any complications during the follow-up period (0.6 to 6 years). In Case 1-4, while esophageal varices and splenomegaly were alleviated, platelet count, blood ammonia level and prothrombin time were also drastically improved after surgery. In Case 5, the residual liver function was maintained normal without any sign of portal hypertension. In Case 6, the liver graft as well as both kidneys functioned well without HV or IVC obstruction.

Caption: Reconstruction of retrohepatic vena cava in liver transplantation using rUV
**Conclusion:** Our experience suggests the usefulness and long-term patency of the rUV not only as an entry to the intrahepatic PV but also as a free vascular graft to reconstruct the PV or the IVC.

Aims of the Study: Portal hypertension is a rare but potentially life-threatening condition that requires surgical treatment in some situations. Over recent years, surgical options have become more and more developed. However, the process of patient selection, decision making, and choice of adequate procedure has not yet been completely standardised. The aim of this study was to review the respective patient cohort at our institution (September 2002 until January 2015), in order to propose an algorithm for the surgical treatment of children with portal hypertension.

Method: We evaluated 39 children with portal hypertension for surgical treatment. Indications for surgery were: Repeated episodes of gastrointestinal bleeding, severe hypersplenism, and pulmonary affection (hepatopulmonary syndrome or portal-pulmonary hypertension). Diagnostic workup included Ultrasound, Doppler sonography, Angio-MRI or –CT scan, wedge portography, and – if necessary – splenoportography. Selection of the surgical procedure was made based on vascular conditions of the portal-venous system (intra- and extra-hepatic) as well as on the liver function and histology.

Results: Of the 39 evaluated children, 28 underwent surgical treatment. Diagnoses of the children undergoing surgery were: portal vein thrombosis (n=14), portal venous malformation (n=5), tumors (n=5), liver fibrosis (n=4). Surgical procedures consisted of Meso-Rex-Shunts (n=9), Warren Shunts (n=8), Splenectomies (n=8), porto-caval H-Shunts (n=2), and portal vein resection with re-anastomosis (n=2). One child received 2 procedures. Shunt thrombosis requiring interventional treatment occurred in 3 cases (2x Rex Shunt, 1x Warren Shunt). One other major complication (biliary tract stenosis treated by Roux-en-Y portoenterostomy) occurred. The two patients with central H-Shunt have a mild hyperammonemia. One patient died 38 months after Warren Shunt because of a crisis caused by pulmonary hypertension and acute cardiac decompensation. One other patient died 16 months after splenectomy because of his underlying oncological disease. Median follow-up was 57 months (range 0-195).

Conclusion: The Meso-Rex-Shunt represents the first choice in children with portal hypertension, in which surgical treatment is indicated. This procedure represents the only curative approach, especially for extrahepatic portal vein thrombosis. The Warren Shunt is associated with a lower risk for hepatic encephalopathy and should be chosen as second choice if a Rex Shunt is not realisable. Central Shunts should be performed as final option only if the previous two procedures cannot be realised.
Aims of the Study: Portal hypertension (PH) in children is associated with high rate of morbidity and mortality, and its surgical management requiring teamwork and expertise is challenging. The aim of this study is to reveal the role of shunt surgery in the management of PH in children.

Method: Hospital records of children who had undergone shunt surgery for PH between 2006 to 2015 were reviewed, overall outcomes were evaluated, and efficiency of two major surgical techniques (Rex shunt and splenorenal shunts; SRS) were compared.

Results: Forty nine consecutive patients (46 pre-hepatic, 3 hepatic PH) had undergone total of 58 shunt procedures. Primary procedures included Rex (n=17, mean age=124±64 months), SRS (24 distal, 3 side-to-side, and 2 proximal, mean age=117±44 months), mesocaval (n=1), and portocaval (n=2) shunts as appropriate. Mean follow-up was 43.3 months. Most common complication was thrombosis of the graft in 12 and stenosis in one of 17 Rex shunts. Stenosis was managed by transhepatic dilatation. Four of 12 thrombosed patients received salvage SRS whereas 5 patients were managed by revision of the Rex shunt; one of which was re-thrombosed and required further management by SRS. All of the SRS's were functional and did not require any additional intervention. There was one mortality in the SRS group which was performed as an emergency procedure for hemorrhagic shock due to massive variceal bleeding, and the patient was lost in the early postoperative period due to disseminated intravascular coagulopathy.

Conclusion: Shunt surgery has an important role to reduce mortality and morbidity in the treatment of PH. Rex shunt may be the procedure of choice with its physiological nature in limited number of patients; however, patient selection is critical and thrombosis rate seems to be higher than reported. SRS's may successfully be performed in patients who are not appropriate for Rex shunt or as a primary procedure or a salvage surgery for shunt dysfunction.
**Hepato-biliary**

SC-HE-0074

**EPITHELIAL TO MESENCHYMAL TRANSITION IS NOT A FACTOR IN BILIARY ATRESIA RELATED LIVER FIBROSIS**

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**Aims of the Study:** Epithelial to mesenchymal transition (EMT) has been suggested to be important in the development of liver fibrosis. There is still much unclear about the role of EMT in biliary atresia (BA). BA is an inflammatory obliterative cholangiopathy in which liver fibrosis develops quickly and, despite the success of Kasai portoenterostomy, necessitating liver transplantation in ~70% of patients. We analyzed the presence of EMT in a murine model of BA and in liver biopsies of BA patients and correlated these findings with clinical outcome.

**Method:** We used the murine Rhesus Rotavirus (RRV) induced model of BA. RRV and control pups were sacrificed at age 7 or 14 days. Immunohistochemistry (IHC) and rtPCR were used to investigate epithelial (CK 7, CK 19, E-Cadherin) and mesenchymal (αSMA, S100, collagen type 1) markers. Liver biopsies of 11 BA patients obtained during Kasai portoenterostomy (median age 53 days, range 27–85) were analyzed using IHC

**Results:** In the RRV group CK19 staining of bile ducts was absent to weak and significantly lower compared with the control groups at both time points (both p<0.01). Although expression of α-SMA was present in portal tracts of healthy mice, there was greater expansion of αSMA staining in the RRV group (p<0.001). Collagen type I was increased in the RRV group (p=0.02). E-Cadherin staining was moderate/strong in the RRV group versus weak/moderate in the control group (p=0.03). rtPCR analysis (table 1) showed an increase in CK7, CK19, E-Cadherin and S100 in 14 days old RRV mice when compared with 7 days old RRV mice. Compared to controls the RRV mice expressed a lower level of CK7 and a higher level S100 at 7 days. This pattern changed over time: in the 14 days old group the RRV mice had lower level α-SMA and a higher level of E-Cadherin and S100 compared with the healthy group. 3/11 BA patients had cirrhosis. CK7 and CK19 staining were weak in five patients, moderate in two, and strong in three patients. αSMA portal and lobular expression was evaluated as none or weak in the majority of patients. There was no correlation between CK7, CK19 or α-SMA expression and grade of liver fibrosis. Age at Kasai was positively correlated with CK7 and CK19 expression (both r=0.6, p=0.08). None of the markers were correlated to clearance of jaundice after Kasai.
Table:

<table>
<thead>
<tr>
<th>Gene</th>
<th>7 days</th>
<th>14 days</th>
<th>7 days</th>
<th>14 days</th>
</tr>
</thead>
<tbody>
<tr>
<td>CK7</td>
<td>0.52*(0.31-0.90)</td>
<td>3.23^(1.43-6.18)</td>
<td>1.27(0.82-1.77)</td>
<td>1.50(1.22-2.01)</td>
</tr>
<tr>
<td>CK19</td>
<td>0.33(0.23-1.05)</td>
<td>1.17*(1.07-2.70)</td>
<td>0.84(0.68-1.36)</td>
<td>1.33(0.78-1.55)</td>
</tr>
<tr>
<td>E-Cad</td>
<td>1.14(0.87-1.41)</td>
<td>1.62^(1.41-1.99)</td>
<td>0.96(0.78-1.14)</td>
<td>1.01º(0.86-1.09)</td>
</tr>
<tr>
<td>S100</td>
<td>1.50*(1.24-2.21)</td>
<td>5.85^(1.99-8.26)</td>
<td>0.46(0.37-0.51)</td>
<td>0.37º(0.31-0.42)</td>
</tr>
<tr>
<td>αSMA</td>
<td>1.09(0.87-1.34)</td>
<td>0.88(0.58-1.22)</td>
<td>1.17~(0.77-1.63)</td>
<td>1.99º(1.06-2.12)</td>
</tr>
<tr>
<td>Coll. I</td>
<td>1.06(0.55-1.72)</td>
<td>1.40(0.82-3.22)</td>
<td>0.88~(0.74-1.40)</td>
<td>1.34(1.14-1.93)</td>
</tr>
</tbody>
</table>

Median (range)

* significant different comparing RRV 7 days vs control mice 7 days
^ significant different comparing RRV 7 days vs RRV 14 days
~ significant different comparing healthy mice 7 days vs control mice 14 days
º significant different comparing RRV 7 days vs control mice 14 days

Caption: rt-PCR results of murine liver samples

Conclusion: In the murine model for BA there is an increase in both epithelial and mesenchymal markers. During EMT epithelial markers should decrease. In liver biopsies of BA patients we demonstrated an increase in epithelial markers, whilst mesenchymal markers were not increased. There was no association between any of the markers and clinical outcome. Based on present data we conclude that EMT is not involved in BA related liver fibrosis.
**Hepato-biliary**

**SC-HE-0075**

**BILIARY COMPLICATIONS IN MULTIVISCERAL TRANSPLANTATION**

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¹Pediatric Surgery, ²Pediatric Gastroenterology, Universitary Hospital La Paz, Madrid, Spain

**Aims of the Study:** In full multivisceral transplant (MVTx), the portal triad and pancreaticoduodenal complex are not subject to any manipulation or dissection. However, some of these patients develop biliary complications of uncertain ethiology. We describe the clinical characteristics and therapeutic management of those patients who presented biliary complications following MVTx.

**Method:** Patients who developed biliary complications after MVTx were studied retrospectively. Demographic data, biochemical parameters, imaging studies and particular aspects as operating and ischemia time, preservation of the native spleen and immunosuppression protocol were collected from medical records. We also describe the therapeutic strategies used in each case.

**Results:** From 2008 to 2014, 7 (21%) out of 33 MVTx performed showed laboratory or image signs of cholestasis in absence of rejection (3M/4F).

Indications for MVTx were: short bowel syndrome (6), Berdon syndrome (1), retransplantation (1). Median age at transplant was 10(6-93) months and biliary complications appeared at a median of 15(1-36) months after the procedure. Recipient age and sex, operating time, cold ischemia time, preservation of native spleen and immunosuppression protocol were similar among both groups.

The diagnosis was made by ultrasound and confirmed in 6/7 by MRCP. A choledochal stenosis was observed in 6 cases (with or without secondary intrahepatic dilatation) and one case had multifocal cystic dilatation of segmental intrahepatic bile ducts.

Asymptomatic patients with a mild cholestasis pattern (3/7) received only medical treatment. Patients with severe cholestasis or colangitis (4/7) were subject to percutaneous approach. In 3 of them the stenosis was successfully dilated and cholestasis resolved, in the remaining case dilatation was not feasible and a catheter was placed just for external drainage. This patient required Roux-Y hepaticojejunostomy to resolve the problem. The histopathology of the common bile duct excised showed intense parietal fibrosis with complete stenosis of lumen.
Conclusion: Late biliary complications are frequent after en bloc MVTx. Interventional radiology is a useful and safe tool in symptomatic cases. Hepaticoyeyunostomy can be performed when percutaneous dilatation fails. Although immunological, rejection, ischemic and toxic reasons have been proposed, their origin is poorly understood and may be multifactorial, so further investigation is warranted.
TIMING OF SURGERY AND OUTCOME IN ANTENATALLY DETECTED CHOLEDOTHAL MALFORMATIONS: RESULTS FROM THE DUTCH NATIONAL REGISTRY.


1Pediatric Surgery, University Medical Center Groningen, Groningen, Netherlands

Aims of the Study: An increasing number of CM will be diagnosed antenatally. While symptomatic children are operated within a few weeks depending on their physician, the most appropriate timing for surgery in asymptomatic patients remains unclear. Some favour early surgery to prevent progression to symptoms while others prefer to postpone surgery to reduce the surgical complication rate. Aim of this study was to identify the optimal timing of surgery in the asymptomatic children with antenatally diagnosed CM and to identify possible predictive factors for the occurrence of preoperative symptoms.

Method: Using the Netherlands Study group on CHoledochal malformation registry (NeSCHoc) we identified all Dutch patients with a prenatal abnormality leading to postnatal confirmation of diagnosis of CM. Medical records, biochemical and surgical data and short and long term results were studied retrospectively.

Results: Between 1989-2014, 96 patients underwent surgery for CM. We included all patients with antenatal diagnosis based upon prenatal screening (n=17, 18% of total). Four patients (4/17, 24%) became symptomatic within 2 weeks after birth (type Ia, Ic, II and IVa CM according to Todani, each n=1). Thirteen patients (13/17, 76%) were asymptomatic at the age of definitive diagnosis (median age 0.2 months, range 0.03 – 2.1). The asymptomatic patients were diagnosed with type I CM (12/13, 92%), and one type IVb (1/13, 8%). Subtypes of type I were type Ia (n=7), type Ib (n=1) and type Ic (n=4).

Three of the initially thirteen asymptomatic patients became symptomatic (3/13, 23%), at ages 2.0, 2.1 and 21.5 months. All were type Ia CM (p=0.07). There were no differences in liver enzyme tests between patients who progressed to symptoms and those who did not, although the difference in GGT levels at time of postnatal confirmation of diagnosis appeared different: 206 I/L (17-573) in the asymptomatic patients vs. 772 I/L (424-1344) in the patients who progressed to symptoms.

Median age at surgery was 4.1 months (0.3-21.6); median weight was 5.6 kilograms (2.8-10.7). Four open (4/13, 31%) and nine (9/13, 69%) laparoscopic procedures were performed. Four patients (4/13, 31%) developed short term complications (<30 days) of anuria, cholangitis/fever, duodenum perforation and bile leakage/abscesses. Two (2/13, 15%) required surgical or radiological intervention and one (1/13, 8%) required antibiotic treatment. Three patients (3/13, 23%)
developed long term complications of cholangitis. Two (2/13, 15%) were due to biliodigestive stenosis which required surgical intervention. Postoperatively, patients weighing <5.6 kilograms (median in present series) showed significant more short term complications when compared to patients weighing >5.6 kilograms (66% VS 0% p=0.02), long term complications did not show significance(50% versus 0%, p=0.07). There was no significant difference in patients who remained asymptomatic and patients who progressed to symptoms in short and long term complications.

**Conclusion:** Although numbers of this retrospective study are small, it seems that when symptoms do not occur before the age of 3 months, surgery might safely be delayed to the age of at least 6 months/weight >6 kilograms in children with an antenatally detected choledochal malformation. Postponing surgery until > 6 kg decreases the risk for (short-term) postoperative complications. GGT might be used as a marker for biliary stasis and subsequently the progression to symptoms and should be regularly checked.
Aims of the Study: A choledochal malformation (CM) is a rare biliary entity in the Western world. The only option to achieve adequate numbers for research is to create nationwide or even multinational databases. We set out to develop a nationwide database of children who underwent surgery for CM in the Netherlands. With this database, we aimed to identify possible predictive factors associated with long-term complications after surgery.

Method: We included all patients < 18 years of age who underwent a surgical resection for CM into the Netherlands Study group on CHoledochal malformation registry (NeSCHoc). Symptoms, surgical details and short- and long-term complications were noted retrospectively and analyzed. We tested the relation between surgical technique, age/weight at the time of surgery, presence/absence of a common channel and the development of short- and long-term complications. Short- and long-term complications were defined as complications necessitating medical, surgical or radiological intervention, occurring before or after postoperative day 30- surgery respectively.

Results: Between January 1989 and December 2014, 96 paediatric patients underwent surgery for CM, the male:female ratio was 1:2.8. There were 67 Todani type I (70%), two type II (2%), 16 type IVa (17%), one type IVb (1/96, 1%), five type V (5%) and five unknown type (5%). Severe co-morbidity was present in four patients (4%). Seventeen patients (18%) were discovered antenatally. Thirty-eight patients (40%) were diagnosed with CM before the age of 1 year. Ninety-five patients (99%) underwent excision of the affected extrahepatic biliary ducts with reconstruction with Roux-en-Y hepaticojejunostomy, and one patient (1%) underwent a cyst resection (type II). Laparoscopic resection was performed in 11 patients (11%). No hepatic parenchyma resection was performed during primary surgery. Short-term postoperative complications (<30 days) occurred in 22 patients (23%), without significant correlations with weight, age or surgical technique. Long-term postoperative complications occurred in 16 patients (17%) after a median of 1.4 years (range 0.8-23.01; Table 1). Nine patients (9%) required radiological intervention or additional surgery. We did not observe biliary malignancies during the follow-up (median 12.9 years, range 0.1-25.3). Prognostic factors for the overall occurrence of long-term complications were tested univariately (Table 2). The laparoscopic resection (p=0.02) and surgery before 1 year of age (p<0.00) proved to be significant. The relatively low numbers prohibit the meaningful use of multivariate analysis.
Table:

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Long term postoperative complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total of n=17 patients</td>
<td>Observed</td>
</tr>
<tr>
<td>Cholangitis</td>
<td>14 (15%)</td>
</tr>
<tr>
<td>Stricture</td>
<td>4 (4 %)</td>
</tr>
<tr>
<td>Ileus</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>Gallstones</td>
<td>3 (3%)</td>
</tr>
<tr>
<td>Abscesses</td>
<td>2 (2%)</td>
</tr>
<tr>
<td>Secondary parenchyma resection</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>Incomplete resection of cyst</td>
<td>1 (1%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Risk factors for long term complications</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Odds ratio</td>
</tr>
<tr>
<td>Age &lt; 1 year at surgery</td>
<td>9.63</td>
</tr>
<tr>
<td>Weight &lt; 6 kilograms at surgery</td>
<td>6.06</td>
</tr>
<tr>
<td>Laparoscopic technique</td>
<td>4.74</td>
</tr>
<tr>
<td>Absence of a common channel</td>
<td>0.79</td>
</tr>
<tr>
<td>Short term complication</td>
<td>1.15</td>
</tr>
</tbody>
</table>

**Conclusion:** In this 25 years retrospective cohort with patients operated on CM < 18 years of age, no malignancies were found. Long-term complications occur in almost one fifth of patients. Laparoscopic surgery, surgery before 1 year of age and the absence of a common channel are associated with long term complications.
SINGLE CENTRE EXPERIENCE OF MESENCHYMAL HAMARTOMA OF THE LIVER IN CHILDREN

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Aims of the Study: Mesenchymal hamartoma (MH) is a rare, benign tumour of the liver. There is controversy over the need for complete resection. Our aim was to review our experience of this uncommon hepatic lesion.

Method: Retrospective review of patients with MH over 12 years (2001 to 2013). Case notes were reviewed for presentation, radiological findings, management, histological features and outcomes.

Results: 10 (6 female) children were identified with median age at presentation of 13 (2-65) months. There were none with specific antenatal features in this group, although one was complicated by a molar placenta. Co-morbidities included gastroschisis (n=1), port-wine stain (n=1), neuropathic bladder (n=1). Eight presented with symptoms (e.g. abdominal mass) and 2 were diagnosed incidentally on ultrasound for other conditions. There were 2 children with complex features attributable to giant multicystic tumours that had had previous failed surgical attempts at resection elsewhere. Correct preoperative diagnosis by imaging (US/CT/MRI) was only achieved in 4 (40%). Median AFP was 50 (2-120900)KU/L. Jaundice was apparent in two. There was a distinct right lobe predominance (n=9). Median diameter on initial imaging was 120 (22-215) mm. A classical solid/cystic appearance was present in 9 (90%).

All were treated by complete surgical resection including: extended right hepatectomy (n=3); extended left hepatectomy (n=1); bisegmentectomy (n=4) and non-anatomical resection (n=2). Typical histological features were seen in 9, although one contained no cystic elements. One contained atypical hepatocytes showing mitotic activity and quantities of immature parenchyma.

One patient had a bile leak postoperatively. Median follow-up was 2 (0.2-4.8) years, with no evidence of recurrence on liver imaging.

Conclusion: This appears to be the largest reported series from a European centre. Correct preoperative diagnosis can be difficult and extended resection may be necessary for complete excision. Children should be managed in experienced centres.
**Hepato-biliary**

SC-HE-0079

**CONGENITAL PORTO-SYSTEMIC SHUNTS IN CHILDREN: ARE YOUNG BRAINS AS RESISTANT TO PORTAL TOXINS AS WE THOUGHT?**

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**Aims of the Study:** Congenital porto-systemic shunts (CPS) are abnormal connections between the portal and systemic venous system allowing hepatic parenchymal bypass and are biochemically characterised by hyperammoniaemia. One possible consequence is hepatic encephalopathy, typically seen in adults, but believed rare in children due to a degree of "resistance" to portal compounds in the juvenile brain.

We hypothesize that many children with CPS are subject to potentially correctable neuro-cognitive insults. We set out to review our series of children with CPS to determine the incidence of neuro-cognitive symptoms and their potential reversibility after shunt closure.

**Method:** Single-centre retrospective review of children with a diagnosis of CPS treated since 2000 identified from prospectively-maintained database.

**Results:** 32 children were seen with a primary diagnosis of CPS in the period 2000-2014. We excluded 9 with an obvious cause for developmental delay or neurodysfunction (e.g. chromosomal anomaly, microcephaly) and 4 with a true ductus venosus that closed spontaneously. This left 19 (7 F) children who presented at a median of 1 month (antenatal diagnosis – 16 years) of which 11 (58%) had reported neurocognitive symptoms. These included developmental delay (n = 6), and behavioural symptoms (including autistic spectrum) (n = 5). Five children are currently awaiting intervention due to either age <2yrs (chance of spontaneous closure) or awaiting angiographic studies. Nine were closed surgically at median age of 10 (3-18) yrs. Four showed improvement in academic work and/or behaviour post-operatively.

**Conclusion:** Subtle and gross neurocognitive impairment are associated with CPS even in children. This may be reversible with CPS closure and thus should be performed early. Formal neurocognitive assessment is recommended pre-operatively.
Aims of the Study: Epithelial tubular structures are essential units in various organs, including the lungs, intestines, and kidneys. Formation of epithelial tubular structures is regulated by several cell behaviors, including cell adhesion, migration, and proliferation. Understanding of the molecular mechanisms of tubulogenesis is important to clarify the pathogenesis of congenital diseases and for the development of its therapies. We used rat intestinal epithelial cells (IEC6) to analyze epithelial tubulogenesis in vitro and found that a combination of Wnt3a and EGF (Wnt3a/EGF) induced tubular formation in three-dimensional culture conditions with basement membrane components (Fig.1). The aim of this study is to clarify the molecular mechanism of epithelial tubulogenesis induced by Wnt3a/EGF.

Method: DNA microarray analysis, immunofluorescence staining, real-time PCR, and western blot were used to analyze the molecular mechanism of epithelial tubulogenesis induced by Wnt3a/EGF.

Results: Wnt3a/EGF synergistically induced the expression of P2Y$_2$ receptor (P2Y$_2$R), a nucleotide-activated G-protein coupled receptor, and knockdown of P2Y$_2$R suppressed Wnt3a/EGF-induced tubular formation. A P2Y$_2$R mutant that lacks nucleotide responsiveness rescued the phenotypes resulting from P2Y$_2$R knockdown, suggesting that nucleotide-dependent responses are not required for P2Y$_2$R functions in tubular formation. The Arg-Gly-Asp (RGD) sequence of P2Y$_2$R has been shown to associate with integrins, and a P2Y$_2$R mutant that lacks the integrin-binding activity lost the ability to regulate tubular formation. P2Y$_2$R expression inhibited the formation of a complex between fibronectin and integrins, and induced cell morphological changes and proliferation. The appropriate inhibition of the interaction between fibronectin and integrins by the cyclic RGD peptide and fibronectin siRNA enhanced, but a large amount of fibronectin suppressed Wnt3a/EGF-induced tubular formation.
Conclusion: P2Y₂R expression induced by Wnt3a/EGF causes tubular formation of IEC6 cells by preventing the binding of integrins and fibronectin rather than mediating nucleotide responses. We are currently examining whether disruption of the regulatory mechanism of epithelial tubulogenesis triggers congenital diseases of tubular organs using organ culture of mouse embryonic kidney (Fig.2).
Basic Science

SC-PP-0082

PARENTERAL ALUMINUM CAN PRODUCE A Viable MODEL OF PARENTERAL NUTRITION ASSOCIATED LIVER DISEASE IN RATS

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Aims of the Study: For patients with intestinal failure, parenteral nutrition (PN) is a life saving therapy. Unfortunately, hepatic dysfunction will occur in 40-60% of children on long term PN. While the hepatic dysfunction is likely multifactorial, one important chemical component of the disease may be aluminum contamination of the PN. Previous studies have shown a reduction in liver injury by decreasing the aluminum concentration in parenteral nutrition in a pig model. We sought to develop a rat model of parenteral nutrition associated liver disease (PNALD) with parenteral aluminum.

Method: Adult Sprague Dawley rats had intravenous long term catheters placed. The control group underwent daily injections of saline. The study rats had daily injections of either 2 mg/kg or 3 mg/kg aluminum chloride (AlCl3). At the end of 4 weeks, the rats were euthanized and liver and blood samples were taken. The livers were analyzed and graded by a pathologist for histologic evidence of liver degeneration and acute and chronic inflammation. The serum was analyzed for total bilirubin, alkaline phosphatase, ALT, and AST.

Results: There was no difference in serum values of total bilirubin, alkaline phosphatase, ALT, or AST. There was no difference in acute inflammation among the groups (1 [control], 1.2 [2 mg/kg], 1.1 [3 mg/kg]). The rats treated with parenteral aluminum had significantly more Kupffer cells than the control group (0.1 [control], 3 [2 mg/kg], 2.2 [3 mg/kg], p = <0.0001 [control vs 2 mg/kg] and 0.0032 [control vs 3 mg/kg]). There was also more liver degeneration in the parenteral aluminum groups than the control group (1 [control], 2 [2 mg/kg], 2.5 [3 mg/kg], p = 0.0341 [control vs 2 mg/kg] and 0.009 [control vs 3 mg/kg]). However, there was no difference between 2 mg/kg and 3 mg/kg AlCl3 for either variable.

Conclusion: This study suggests that four weeks of parenteral aluminum can induce chronic inflammation and degeneration of the liver in rats. Therefore, we believe that daily injections of parenteral aluminum can produce a viable model of PNALD in rats. However, further studies are warranted, including measurement of serum aluminum levels in infants on parenteral nutrition.
**Hepato-biliary**

SC-PP-0083

**MESOHEPATECTOMY FOR CENTRALLY LOCATED TUMORS IN CHILDREN.**

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¹Pediatric Surgery, Hospital Universitario La Paz, Madrid, Spain

**Aims of the Study:** Central hepatectomy or mesohepatectomy (MH) is a complex surgical technique rarely used in children. It is indicated in central tumors in order to preserve functioning liver mass avoiding an extended right hepatectomy. The purpose of this paper is to analyze our experience with this technique.

**Method:** We reviewed 5 patients who underwent MH in the period 2008-2014. Diagnoses were hepatoblastoma PRETEXT III (2), hepatic embryonal sarcoma (1), focal nodular hyperplasia (1), and vascular tumor with rapid growth in a newborn causing an acute liver failure, compartment syndrome and multiple organ failure (1). In all cases the tumor was centrally located, including the segment IVb, with large displacement of the hepatic pedicle in 2

**Results:** Mesohepatectomy was standard in 3 and under total vascular exclusion in 2. All children are alive with a mean follow up of 38 (6-70) months. None of the children required reoperation because of bleeding. One child developed a biliary fistula in the cutting area that closed spontaneously. The newborn with the vascular tumor, required a mesh of Goretex to relieve compartment syndrome, improving ventilatory support after a few hours. He subsequently underwent partial embolization of the tumor and mesohepatectomy under vascular exclusion.

**Conclusion:** In selected patients, mesohepatectomy is an alternative to trisegmentectomy and should be available in advanced pediatric hepatobiliary units.
Aims of the Study: Posterior sagittal anorectoplasty is considered the gold standard procedure for repair of anorectal malformations (ARM). More recently, laparoscopy has been used for the treatment of such anomalies. In the last four years, we have performed laparoscopic-assisted anorectoplasty (LAARP) for select cases at our institution. The aim of our study is to assess the short term anorectal outcomes of LAARP and evaluate factors affecting our outcomes.

Method: A detailed chart review of all 20 children who underwent LAARP at our institution between July 2010 and September 2014 was performed. All patients had a standard initial diagnostic work-up and associated anomaly screening, with follow-up by the surgical team and colorectal nurse specialists. A prospective database of outcomes was maintained. LAARP was performed using a 3 port technique, with laparoscopic mobilisation of the colon and ligation of fistula if present. The muscle complex was partially divided by a perineal approach, and a laparoscopic port inserted through into the peritoneum to perform the pull through. In 4 patients, we sutured the bowel to the muscle complex in 4 quadrants to prevent prolapse. In all other patients, we sutured the bowel wall only to the posterior aspect of the muscle complex. The outcome measures we assessed were the rate of perioperative complications and conversion to an open procedure. We also examined patient demographics, associated anomalies and type of anorectal malformation with regards to potential effects on outcomes.

Results: Of the 50 patients with ARM referred to our institution in the study period, 20 (40%) were treated by LAARP following neonatal colostomy formation. There were 16 male and 4 female infants, 7 with VACTERL association and 8 with trisomy 21. Subtypes of anorectal malformation were: 7 rectoprostatic, 2 bladder neck, 1 rectobulbar urethral, 1 unspecified fistula and 9 without fistula. Median age at time of LAARP was 7.2 months (range, 3.8-38.8 months). Median time from LAARP to colostomy closure in our cohort was 3.7 months (range, 1.9-5.8 months), with 2 patients awaiting closure. There was no conversion to an open procedure and no patient to date has required revision anoplasty. All 20 patients are under active follow-up for a median of 0.9 years (range, 0.5-4.6 years). Two patients (10%) experienced complications related to the LAARP; 1 patient with a pelvic abscess and 1 with mild mucosal prolapse not requiring treatment. The 4 patients who had 4 quadrant fixation of the bowel developed a definite resistance to dilatation at 2-3cm from the anal verge, causing some difficulty progressing with dilatations. This has not been noted in the patients who had posterior fixation of the bowel only. One further patient has developed anal stenosis due to poor compliance with dilatations. At follow-up, 4 patients have significant perianal excoriation, 3 experience constipation and 1 has high stool frequency.
Conclusion: In our experience, LAARP is a safe and effective approach for the repair of ARM in select cases. In 4 patients, an obvious resistance was noted 2-3cm from the anal verge at initial anal calibration 2 weeks post-operatively. We postulated that this may have been due to the pull through bowel being constricted at the level of the port site defect in the muscle complex, or due to the sutures used to fix the bowel to the muscle complex. Awareness of potential pitfalls and modification of operative technique can improve outcomes for our patients.
ASCA AND OMPC SEROLOGICAL MARKERS ELEVATED IN CHILDREN WITH HIRSCHSPRUNG-ASSOCIATED ENTEROCOLITIS

Philip K. Frykman¹, Zhi Cheng¹, Tomas Wester², ³, Agneta Nordenskjöld², ³, Akemi Kawaguchi⁴, Thomas Hui⁵, Daniel Teitelbaum⁶, Carole Landers⁷ and for the HAEC Collaborative Research Group (HCRG)

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Aims of the Study: Hypothesis: Children with Hirschsprung disease (HD) who develop Hirschsprung-associated enterocolitis (HAEC) have elevated serological immune markers associated with inflammatory bowel disease (IBD) compared with HD patients who have not developed HAEC.

Method: An IRB-approved international multicenter study enrolled n=28 HD patients ages 4 months to 26 years. Each subject’s medical record was systematically reviewed and one parent interviewed for detailed history of enterocolitis symptoms. HAEC status was determined according to Pastor et al. (2009). Blood was collected, plasma isolated and underwent ELISA for Anti-Saccharomyces cerevisiae antibody (ASCA), anti-Escherichia coli outer membrane porin C antibody (OmpC), anti-flagellin antibody (CBir1), Anti-neutrophil cytoplasmic antibodies (ANCA). Results (mean±SEM) were analyzed for significance by t-test, two-tailed, unpaired.

Results: Five patients met criteria for having had at least one episode of HAEC, 23 had no history of HAEC. The HAEC and the HD groups had median ages of 9.1 and 6 years, respectively. HAEC and HD groups were similar in family histories of HD and trisomy 21; HAEC group had a higher proportion of patients with family histories of IBD, long-segment aganglionosis and post-operative complications (Table). The HAEC group showed markedly elevated ASCA IgA, ASCA IgG, and anti-OmpC antibody levels compared with the HD group while anti-CBir1 and ANCA were similar between the groups (Figure).
Serological markers in HD and HAEC patients

<table>
<thead>
<tr>
<th></th>
<th>HD</th>
<th>%</th>
<th>HAEC</th>
<th>%</th>
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<tbody>
<tr>
<td>n</td>
<td>23</td>
<td>5</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Median age (years)</td>
<td>6.1</td>
<td>9</td>
<td>6.1</td>
<td>9</td>
</tr>
<tr>
<td>Family hx of HD</td>
<td>5</td>
<td>22%</td>
<td>1</td>
<td>20%</td>
</tr>
<tr>
<td>Family hx of IBD</td>
<td>1</td>
<td>4%</td>
<td>1</td>
<td>20%</td>
</tr>
<tr>
<td>Short segment (distal to splenic flexure)</td>
<td>22</td>
<td>96%</td>
<td>4</td>
<td>80%</td>
</tr>
<tr>
<td>Trisomy 21</td>
<td>0</td>
<td>0%</td>
<td>0</td>
<td>0%</td>
</tr>
<tr>
<td>Post op complications (&lt;30 days)</td>
<td>4</td>
<td>17%</td>
<td>2</td>
<td>40%</td>
</tr>
</tbody>
</table>

Caption: Patient Characteristics

Conclusion: The HAEC group demonstrated markedly increased ASCA and anti-OmpC serum antibody levels in the absence of ANCA and anti-CBir elevation similar to pediatric patients with Crohn’s disease (CD), but not ulcerative colitis. These findings suggest similarities in gut microbial-host immune response between HAEC and CD, and may have implications for pathogenesis and treatment of this group.

Lower Gastrointestinal

SC-PP-0086

PREVALENCE AND CONTROLLED, LONG-TERM OUTCOMES OF INTRASPINAL ANOMALIES IN PATIENTS WITH INTERMEDIATE AND HIGH ANORECTAL MALFORMATIONS (ARMS) AFTER EXPECTANT, NON-OPERATIVE MANAGEMENT

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Aims of the Study: To define the prevalence of intraspinal anomalies (ISA) in patients with intermediate and high ARMs, and to compare the outcomes for bowel function, lower urinary tract symptoms (LUTS) and lower limb neurology after expectant, conservative management in relation to patients with the same types of ARMs but without spinal cord anomalies.

Method: The spinal cord magnetic resonance imaging (MRI) records were of all patients treated for vestibular fistula (VF) and rectourethral fistula (RUF) at our center from 1983-2006 were cross-sectionally reviewed by a pediatric radiologist. Conus termination below L2 end-plate was considered abnormal. Case records were reviewed for lower limb neurology. Bowel function and LUTS were assessed using a postal questionnaire. Patients with total sacral agenesis (n=2) were excluded from the study. Patients with a diverting colostomy (n=1) or major cognitive disabilities (n=3) were excluded from the functional outcomes analysis. Ethical approval was obtained.

Results: Of a total of 89 patients (median age 15; range 5-28 years), spinal cord MRI was available for 89% (n=80; 40 RUF males). 83% of eligible patients had responded to the questionnaire on bowel function and LUTS (n=64; 31 RUF males). No patients had undergone detethering surgery. Intraspinal anomalies (ISA) were present in 41% in VF and 28% in RUF (p=0.04), comprising a filum terminale lipoma in 29% (n=24), thoracolumbar syrinx in 6% (n=5) and conus termination at L3 in 9% (n=7). Sacral hypoplasia (3-4 segments remaining) was significantly more common in patients with ISA (41vs12%; p=0.01). In the comparison of patients with ISA (n=22) to patients with normal spinal MRI (n=41), no significant difference was found in outcomes by bowel function score (median 16 vs 18/20), soiling (70% vs 63%), constipation (56% vs 39%) or requirement for ACE bowel management (9% vs 12% respectively; p=NS for all). Prevalence of LUTS, including urinary urge (65% vs 54%), urge incontinence (39% vs 24%), stress incontinence (17% vs 22%) or straining to void (32% vs 29%) were also comparable (p=NS for all). UTIs were more common in females with ISA (71% vs 25% with normal MRI; p=0.02), but not in males (33 vs 23%; p=0.65). No patients had abnormal lower limb neurology. All patients with ISA had been followed up regularly by a pediatric neurologist.
Conclusion: Our controlled, cross-sectional study suggests that the long-term functional outcomes for ARM patients with ISA treated with conservative, expectant management are not significantly different from patients with the same type of ARMs but normal spinal cord imaging. The results support a non-operative approach to management of these findings in ARM patients in the absence of abnormal lower limb neurology.
IMMUNOHISTOCHEMICAL CHARACTERIZATION OF SMOOTHELIN, DESMIN AND ALPHA SMOOTH MUSCLE ACTIN IN INTESTINAL MYOPATHIES

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¹Pediatric Surgery, DINOGMI, University of Genoa; Istituto Giannina Gaslini, ²Pathology Unit, Istituto Giannina Gaslini, Genoa, ³Pathology Unit, ⁴Pediatric Surgery, University Hospital of Parma, Parma, ⁵Medical genetics, Istituto Giannina Gaslini, Genoa, Italy, ⁶Pathology Unit, Hospital of the University of Basel, Basel, Switzerland

Aims of the Study: Current guidelines of diagnosis of intestinal visceral myopathies (IVM) are founded on ultrastructural study of the smooth muscle wall. The aim of the study was to compare the histochemical pattern in a population of IVM patients with an established ultrastructural diagnosis.

Method: Authors studied three groups of patients. Group 1: six cases (3 females and 3 males) suffering with severe constipation and chronic intestinal pseudo-obstruction with final ultrastructural diagnosis of IVM. Group 2 (control group): nine pediatric patients (without abnormal ultrastructural intestinal muscle wall defects or innervation abnormalities), plus five other control adults (colonic adenocarcinoma). Group 3: five Hirschsprung cases (ganglionic segment of the colon). One hundred and twenty 4 µm paraffine sections were studied by immunohistochemical staining using 3 different antibodies: A) Anti-smoothelin antibody [R4A] ab8969 abcam(R) at dilution of 1:100 in bond primary antibody diluent; B) Mouse monoclonal antibody desmin [NCL-L-DES-DERII, by Leica biosystems] ) at dilution of 1:100 in bond primary antibody diluent ; C) Mouse monoclonal antibody anti-alpha smooth muscle actin [NCL-SMA by Leica biosystems] at dilution of 1:50 in bond primary antibody diluent.

Results: In all groups anti-desmin and anti-smoothelin immunoreactivity did not present significant qualitative differences. On the other hand Anti-alpha smooth muscle actin showed a general immunoreactivity reduction in the staining of muscle wall of the six IVM cases(Group 1). IVM muscle cells presented a light brown staining and in particular the general aspect of the muscular wall appeared heterogeneous with some elements which preserved positivity and most of the other with variable and low staining.

Conclusion: Alpha smooth muscle actin immunohistochemical marker is a useful complementary diagnostic tool for IVM. Ultrastructural study of IVM muscle cells is characterized by degeneration of myofilaments and variable cytoplasmic electro-density. The same variability of positivity with low immunoreaction is present using alpha smooth muscle actin.
THE SURGICAL TREATMENT OF CLOACAL MALFORMATIONS

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Aims of the Study: The aim of the study was to analyze cases of surgery of cloacal malformations in children.

Method: Among the 22 children aged 1.2±0.1 years old with persistent cloacal malformation 18.2% of the patients had an isolated malformation while the rest of the children displayed a combination of cloaca with a urinary system abnormality (72.8%), a cardiovascular system abnormality (41%) and a locomotor system abnormality (31.8%). 9 patients (40.9%) had a total cloacal canal smaller than 3 cm, 13 children (59.1%) had a total cloacal canal larger than 3 cm.

Results: The choice of the surgical method depended on the cloacal canal length and the “maturity” of vagina walls. 5 children (22.7%) underwent posterior sagittal proctovaginaurethroplasty; 4 children (18.2%) got perineal proctovaginaurethroplasty with laparoscopic support and partial urogenital mobilization; 3 children (13.6%) underwent total urogenital mobilization; 10 children (45.5%) underwent abdominoperineal proctovaginaurethroplasty with the replacement of vagina for colon or small intestine. During the early postoperative period, most children had no complications. The mortality rate amounted to 4.5%.

Conclusion: The treatment efficiency was assessed at the age of 3-7 years old. The children with a shorter cloacal canal displayed better results of the treatment than those with a longer cloacal canal, with fecal matter retention registered in 66.7% and 41.7% of the patients, respectively; urine retention was observed in 77.8% and 75.0% of the children, respectively.

Lower Gastrointestinal

SC-PP-0089

MANOMETRIC FINDINGS IN DIFFERENT TYPES OF ANORECTAL MALFORMATIONS IN THE PSARP ERA: RECTAL SENSATION IS THE MAIN DETERMINANT OF FUNCTIONAL STATUS

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1Pediatric Surgery, Helsinki University Children's Hospital, Helsinki, Finland

Aims of the Study: To study the findings of anorectal manometry (AM) in relation to bowel function in patients with different types of anorectal malformations (ARMs) after treatment with minor perineal procedures for low ARMs, and fistula-saving ASARP/PSARP for intermediate/high anomalies.

Method: All patients ≥7 years of age treated from 1983 onwards at our institution for anterior anus (AA), perineal fistula (PF), vestibular fistula (VF), and rectourethral fistula (RUF) were invited to undergo AM as outpatients. Exclusion criteria were major learning difficulties or total sacral agenesis. Bowel function was assessed using a previously validated questionnaire. AM was performed by a single independent investigator. Participation was voluntary. Ethical approval was obtained. Results are expressed as median (IQR) unless otherwise stated.

Results: A total of 55 patients (26%; 42%male) participated. Anal canal length, resting anal canal pressure and maximal squeeze pressures declined significantly with increasing level of ARM (in AA vs RUF: 2.2 vs 1.7cm; 65 vs 35cmH2O; 120 vs 73cm H2O respectively; \( p<0.05 \) for all). Consistent with preserved internal anal sphincter function, a recto-anal relaxation reflex (RAIR) was observed in 91% of patients, including all patients with low ARMs and 83% of patients with intermediate/high anomalies \( (p=0.06 \) vs low ARMs). The rectal sensation threshold (RST) was significantly lower in patients with low ARMs who had not undergone any form of bowel mobilization (10(10-10)ml) vs (20 (10-40)ml) in high/intermediate anomalies \( (p=0.007) \). The rectal sensation was present to at least some degree in all except one patient (98%). Of all measured parameters, only RST correlated significantly with bowel functional status \( (p=0.0007; \ r_s -0.46) \). Compared to patients with an increased RST of >20ml (25%), patients with normal rectal sensation (≤20ml; 75%) reported significantly less soiling (40% vs 84%; \( p=0.009 \)), fecal accidents (10% vs 62%; \( p=0.0005 \)) and constipation (23 vs 84%; \( p=0.0001 \)). The prevalence of sacral dysplasia (3-4 segments remaining) was not significantly greater among patients with reduced rectal sensation (38%) vs patients with a normal RST (20%; \( p=0.26 \)).

Conclusion: Although the length of the anal canal, resting and maximal squeeze pressures declined with increasing level of ARM, only the RST correlated significantly with functional status. As rectal sensation appears to be preserved in the majority of cases after fistula-saving repair, our results strongly favor continuing this approach for high and intermediate ARMs.
Lower Gastrointestinal
SC-PP-0090
ROLE OF ENTERIC GLIAL CELL STAINING IN THE EVALUATION OF TRANSITION ZONE IN HIRSCHSPRUNG’S DISEASE
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Dublin, Ireland

Aims of the Study: Hirschsprung’s disease (HSCR) is characterised by premature arrest of the migration, proliferation and
survival of the developing enteric nervous system (ENS). Enteric glial cells (EGCs) are nerve supporting cells that form an
extensive network intimately associated with ENS. Both glutamine synthetase (GS) and fatty acid binding protein 7
(FABP-7) can reliably discriminate intrinsic EGCs, which are known to be reduced in aganglionic bowel in HSCR, from
extrinsic EGCs. During development of ENS, glial cell migration and differentiation is known to lag behind that of enteric
neuronal development. We hypothesized that the EGC density and distribution is abnormal proximal to the transition zone
in the ganglionic bowel in HSCR and designed an experiment to test this hypothesis, which would have ramifications for
the accuracy of levelling in HSCR.

Method: Entire colonic specimens were obtained at the time of pull-through surgery for HSCR (n=5). Healthy controls
were obtained at the time of colostomy closure in children with anorectal malformation (n=4). We used double-label
immunofluorescence (IF) to examine the distribution of FABP-7 and GS in serial sections of colon of children with HSCR
in order to determine if the level of the intrinsic EGC histological transition zone corresponded with the neuronal transition
zone.

Results: Expression of GS and FABP-7 was reduced in aganglionic bowel compared to both ganglionic bowel in HSCR
and healthy controls. GS and FABP-7 IF-positive EGCs were intimately associated with calretinin IF-positive ganglion
cells in the submucosal plexus and myenteric plexus in the ganglionic bowel in HSCR. The pattern of GS expression
mirrored that of calretinin across the transition zone and into the ganglionic bowel in HSCR.

Image:

Conclusion: The combination of staining for intrinsic enteric glial cells and ganglion cells may accurately determine the
level of transition zone and ganglionic bowel in HSCR.
**Lower Gastrointestinal**

SC-PP-0091

THE PRESENCE OF MICROBIOTA WITHIN THE BOWEL WALL DURING NECROTIZING ENTEROCOLITIS

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¹Pediatric Surgery, University of Groningen, University Medical Center Groningen, ²Microbiology, University of Groningen, University Medical Center Groningen, ³Pathology, ⁴Neonatology, University of Groningen, University Medical Center Groningen, Groningen, Netherlands

**Aims of the Study:** Necrotizing enterocolitis (NEC) is a devastating gastrointestinal emergency in the premature neonate. Bacteria are believed to play a pivotal role in the development of NEC. Unfortunately, nearly all studies focusing on the bacterial role in NEC used faecal samples, which may not be representative for the NEC affected site of the bowel. Therefore, we aimed to investigate the microbial composition in the affected intestinal NEC specimens compared to age-matched intestinal specimens, derived from autopsy material from children without gastrointestinal disease.

**Method:** Between July 2003 and December 2013 intestinal resection material of 43 surgical NEC patients and intestinal autopsy material of 43 age-matched controls were retrospectively analyzed. In the intestinal NEC specimens the specimen with the most affected tissue and the one with the least affected tissue (based on degree of ischemia and necrosis) were used for analysis. Bacterial populations in the specimens were characterized by fluorescent in situ hybridization (FISH), using bacterial rRNA-targeting oligonucleotide probes. For the NEC specimens we used an universal bacterial probe together with species specific FISH probes for *Clostridium perfringens*, *Enterobacteriaceae*, and *Enterococcus*. In the control specimens we used an universal bacterial probe together with the species specific FISH probe for *Enterobacteriaceae*.

**Results:** Using the universal bacterial FISH probe, moderate to high densities of bacteria were found in 84% respectively 69% of the most affected- respectively least affected NEC specimens. In controls this was 20% (p<0.01) *Clostridium perfringens* was only detected in five of the most affected specimens and twice in the least affected specimen. We found similar results for *Enterococcus* (n=2 for both groups). However, *Enterobacteriaceae* dominated significantly in both NEC specimens (33/43 respectively 32/43) compared to controls (20/43) (p=0.01). In the most affected NEC specimens significantly more colonies were found closely to the mucosa compared to the least affected NEC specimens and the controls (p=0.003 respectively p<0.01). Also, in the least affected NEC specimens we found significantly more colonies closely to the mucosa compared to the controls (p<0.01).
Conclusion: This study demonstrates that higher densities of bacteria are associated with the degree of affected tissue in NEC. Interestingly, Enterobacteriaeeae dominated the NEC specimens, while no other known potential NEC associated bacteria, such as Clostridium perfringens, dominated the specimens. This study offers further evidence that bacteria are involved in the pathogenesis of NEC. Thereby, this study emphasized the importance of examining the microbial composition of affected NEC tissue instead of in faecal material, to avoid biases of detecting bacteria from bowel parts not affected by NEC.
Lower Gastrointestinal

SC-PP-0092

VISUALIZATION OF HUMAN ENTERIC NERVOUS SYSTEM USING CONFOCAL ENDOMICROSCOPY: AS AN ALTERNATIVE TO INTRAOPERATIVE PATHOLOGICAL DIAGNOSIS FOR HIRSCHSPRUNG DISEASE.

Naoki Shimojima¹, ², Masakuni Kobayashi³, Takumi Fujimura¹, Satoshi Ieiri⁴, Tomoaki Taguchi⁴, Junko Takahashi-Fujigasaki⁵, Kazuki Sumiyama³, Tatsuo Kuroda¹

¹Department of Pediatric Surgery, Keio University School of Medicine, ²Department of Surgery, Tokyo Metropolitan Children's Medical Center, ³Department of Endoscopy, The Jikei University School of Medicine, Tokyo, ⁴Department of Pediatric Surgery, Kyushu University Graduated School of Medical Sciences, Fukuoka, ⁵Division of Neuropathology, The Jikei University, Tokyo, Japan

Aims of the Study: Intraoperative pathological diagnosis is mandatory in a surgery for Hirschsprung disease to determine the location of a stoma or the range of resection. It is possible to repeat pathological diagnosis several times in an operation and it can make a waiting time during operation. Recently, confocal laser endomicroscopy (CLE) is used to yield cellular details of mucosa for the differential diagnosis of nonneoplastic versus neoplastic changes. The aim of the present study was to assess a feasibility of visualization of human enteric nervous system (ENS) by CLE.

Method: Subjects were 6 patients who underwent intestinal resection. The primary diseases were anal atresia 2, Meckel's diverticulum 2, jejunal atresia 1, Hirschsprung disease 1. Patient ages ranged from 9 months to 10 years. A part of resected intestines were used for the study. To visualize ENS, a fluorescent dye, cresyl violet, was topically injected in the subserosal layer. Then CLE probe was applied to the serosal surface of the intestines to observe ENS. Pathological findings were also evaluated by hematoxylin-eosin stain. CLE grades and pathological grades were applied as follows.
Grade 0: inadequate stain, 1: plexus visualized, 2: plexus not visualized.

Results: CLE clearly visualized ladder-like structure with negatively identified nuclei inside. This resembles myenteric plexus with ganglion cells and glial cells inside. In samples from a patient with Hirschsprung disease, ladder-like structures were observed in the normal segment. In contrast, no ladder-like structure was seen in the aganglionic segment. CLE grades and pathological grades were all matched in 10 samples from 6 patients.
Conclusion: Visualization of human ENS in resected intestines was confirmed using CLE. This result promises that CLE has the potential of new, speedy, and easy-to-understand system to get the picture of ENS network during a surgery for Hirschsprung disease.

HEPCIDIN AS A DIAGNOSTIC MARKER IN ACUTE APPENDICITIS – A PILOT STUDY

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Aims of the Study: A variety of different parameters have already been tested in the diagnosis of acute appendicitis in pediatric patients. Hepcidin represents an acute-phase protein and is a regulator of iron homeostasis and a component of innate immunity with direct antimicrobial properties. The serum concentrations of this protein have been shown to be increased in several inflammatory conditions. The aim of this study was to evaluate the diagnostic value of serum values of hepcidin in acute appendicitis.

Method: In a cohort of children and adolescents with appendicitis the number of leukocytes, platelets, C-reactive protein (CRP), serum levels of iron, ferritin and transferrin as well as serum concentration of hepcidin were assessed. Values were compared to a group of patients with inpatient treatment due to abdominal right quadrant pain without appendicitis. Additionally, the Pediatric Appendicitis Score (PAS) was assessed at the time of admittance in both groups.

Results: 64 patients were included in the study. While 39 out of these had appendicitis and were treated operatively (Group I), 25 patients were treated conservatively with abdominal right quadrant pain without appendicitis (Group II). Neither the age, the height nor the weight differed among the two groups. Number of leukocytes, platelets, CRP and serum levels of iron, ferritin and transferrin were not significantly different among the two groups. However, Group I revealed significantly higher serum levels of hepcidin compared to Group II (31.3 ± 21.7 ng/ml vs. 20.4 ± 14 ng/ml, p=0.039). 17 out of the 39 operatively treated patients were diagnosed a perforated appendicitis and 22 children were treated with non-perforated appendicitis. Perforated appendicitis was associated with a significantly elevated hepcidin concentration compared to non-perforated appendicitis (38.5 ± 17.6 ng/ml vs. 21.6 ± 23.4 ng/ml, p<0.001). Moreover, PAS, leukocytes, CRP and Ferritin were significantly increased in perforated appendicitis compared with non-perforated appendicitis.

Conclusion: Results of this study show that serum levels of hepcidin might play a future role in the diagnosis of appendicitis in children and adolescents. Further studies are required to unravel the exact role and the prognostic value of hepcidin in the diagnosis of appendicitis.
A TRANSITION RISK ASSESSMENT SCORE (TRAS) TO STRATIFY TEENAGERS WITH ANORECTAL MALFORMATIONS BEFORE THEIR TRANSITION TO ADULT HEALTHCARE. AN EVIDENCE BASED PROPOSAL AND PRELIMINARY DATA.

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Aims of the Study: To create the first evidence based Transition Risk Assessment Score (TRAS) to stratify risk of transition for young teenagers with anorectal malformations (ARM) moving towards adult healthcare services.

Method: An extensive search identifying the common long-term conditions in patients with anorectal malformations older than 10 years was performed. Current published scoring systems were identified and weighted to create a single evidenced based transition risk assessment score for risk stratification. The scoring method divided patients into “Low”, “Medium” and “High” risk categories. A transition pathway derived from evidenced based interventions was developed so that risk categories could easily assist the required frequency and nature of follow-up. The structured assessment and transition pathway involved members of the paediatric and adult multidisciplinary teams (MDT) guiding the transition to adult services. The proposed scoring system was applied to patients visiting our tertiary centre anorectal clinic over a six month period in the second half of 2014.

Results: The TRAS constituents included objective assessment of faecal and urinary continence, quality of life and psychosocial wellbeing and sexual function (Figure 1). Scores ranged from 0 to 19. Scores less than 5 mean “Low” risk, with maximum intervals between follow-up and routine discharge without the automatic referral to adult services. Scores of 5-10 mean “Medium” risk with mandatory yearly follow-up and likely automatic transition to adult services with one or more active problems by aged 18 years. Scores >10 mean “High” risk with severe active problems, not responding to treatment, and need regular 6-month follow-up with automatic referral to a multi-disciplinary adult team at final transition. High and medium risk patients should be discussed at combined adult and paediatric MDT meetings. Irrespective of risk category all patients were seen by a specialist transitional care nurse at time of risk stratification and entered onto the ARM transition register. The author’s present preliminary data on a three patient case series assessed using the TRAS to demonstrate feasibility, outcome and follow-up plan.
Figure 1. Anorectal Malformations Transition Risk Assessment Score (TRAS) clinic pro forma

<table>
<thead>
<tr>
<th>Anorectal Malformations (ARM) Transition Risk Assessment Score (TRAS)</th>
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<tbody>
<tr>
<td><strong>Patient Details</strong></td>
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<tr>
<td>Cloaca</td>
</tr>
<tr>
<td>Operative Procedure:</td>
</tr>
<tr>
<td><strong>Transition Preparation</strong></td>
</tr>
<tr>
<td>Is the patient: female aged 12-13 OR male aged 13-14? (Please Circle):</td>
</tr>
<tr>
<td>Yes / No</td>
</tr>
<tr>
<td><strong>Colorectal</strong></td>
</tr>
<tr>
<td>Voluntary bowel movements:</td>
</tr>
<tr>
<td>(Feeling of urge, capacity to verbalize, hold the bowel movement)</td>
</tr>
<tr>
<td>(0) - Yes</td>
</tr>
<tr>
<td>(1) - No</td>
</tr>
<tr>
<td><strong>Soiling</strong></td>
</tr>
<tr>
<td>(0) - No</td>
</tr>
<tr>
<td>(1) - Occasional*</td>
</tr>
<tr>
<td>(2) - Every day, no social problem</td>
</tr>
<tr>
<td>(3) - Constant, social problem</td>
</tr>
<tr>
<td><strong>Constipation</strong></td>
</tr>
<tr>
<td>(0) - No</td>
</tr>
<tr>
<td>(1) - Manageable by changes in diet</td>
</tr>
<tr>
<td>(2) - Requires laxatives</td>
</tr>
<tr>
<td>(3) - Resistant to laxatives</td>
</tr>
<tr>
<td><strong>Nephro-urological System</strong></td>
</tr>
<tr>
<td>Urinary incontinence/Neurogenic bladder:</td>
</tr>
<tr>
<td>(0) - No</td>
</tr>
<tr>
<td>(2) - Mild dribbling/wetness day and night, no social problem</td>
</tr>
<tr>
<td>(3) - Complete incontinence, social problem</td>
</tr>
<tr>
<td><strong>Quality of Life (QoL) &amp; Psychosocial Wellbeing</strong></td>
</tr>
<tr>
<td>School absence:</td>
</tr>
<tr>
<td>(Never (0))</td>
</tr>
<tr>
<td>(Occasional* (1))</td>
</tr>
<tr>
<td>(Frequent (2))</td>
</tr>
<tr>
<td>Unhappy or anxious:</td>
</tr>
<tr>
<td>(Never (0))</td>
</tr>
<tr>
<td>(Occasional* (1))</td>
</tr>
<tr>
<td>(Frequent (2))</td>
</tr>
<tr>
<td>Food restriction:</td>
</tr>
<tr>
<td>(No (0))</td>
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<tr>
<td>(Moderate (1))</td>
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<tr>
<td>(Severe (2))</td>
</tr>
<tr>
<td>Peer rejection:</td>
</tr>
<tr>
<td>(Never (0))</td>
</tr>
<tr>
<td>(Present (2))</td>
</tr>
<tr>
<td><strong>Sexual function</strong></td>
</tr>
<tr>
<td>(Regular erections, normal external genitalia anatomy, normal menstruation, normal sexual life)</td>
</tr>
<tr>
<td>(0) - Yes</td>
</tr>
<tr>
<td>(3) - No</td>
</tr>
<tr>
<td><strong>Total Transition Risk Assessment Score (TRAS):</strong></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Low</th>
<th>Medium</th>
<th>High</th>
</tr>
</thead>
<tbody>
<tr>
<td>(Less than 5)</td>
<td>(Score 5-10)</td>
<td>(Score &gt;10)</td>
</tr>
</tbody>
</table>
**Caption:** Legend Figure 1: The following adapted scoring systems with were used to create the transition risk assessment score pro forma: Krickenbeck post-operative assessment 1 (Colorectal), Peña 2 (Nephro-urological System), Bai et al. 3 (Quality of Life & Psychosocial Wellbeing). +Occasional defined as “once or twice per week”.

**Conclusion:** Transitional care in congenital surgical anomalies is a rapidly evolving field which is an active process and not a single event. Transition must begin early, be planned and regularly reviewed with each patient. An evidenced based risk assessment score can be an effective clinical tool to help stratify ARM patients early in puberty and identify those who will require intensive multi-disciplinary input to be optimally linked into adult healthcare services.

**References:**
A META-ANALYSIS OF THE DIAGNOSTIC PERFORMANCE OF MAGNETIC RESONANCE IMAGING IN THE DETECTION OF APPENDICITIS IN CHILDREN.

Lea Sibylle Waldron* 1, Matthew Bronstein2, Samir Pandya2, Oliver Muensterer1
1Pediatric Surgery, University Medicine, Johannes Gutenberg University Mainz, Mainz, Germany, 2Division of Pediatric Surgery, New York Medical College, Maria Fareri Children’s Hospital, New York, United States

Aims of the Study: Appendicitis is the leading cause of surgical emergencies children. In inconclusive cases, computed tomography (CT) is sometimes used to aide in the diagnoses. While CT is highly accurate, a major drawback is the exposure of the patient to relatively high levels of ionizing radiation. Magnetic resonance imaging (MRI) has been described as an alternative, but only few studies have evaluated its usefulness. To perform a meta-analysis of the current available evidence on the accuracy of MRI for the diagnosis of acute appendicitis in the pediatric population

Method: The medical literature from January 1995 to January 2015 was searched for studies that evaluated MRI as a diagnostic test for appendicitis in children using Pubmed/MEDLINE. Prospective and retrospective studies were screened and included if they used MRI as a diagnostic test for appendicitis and reported sensitivities and/or specificities, or stated sufficient data to derive these numbers, using surgical pathology as the gold standard. Reported sensitivities and specificities were pooled and summarized with 95% confidence intervals using RevMan version 5.3 and excel.

Results: A total of 8 articles met the inclusion criteria reporting on 766 total patients. All studies reported sensitivities, while only 7 reported specificities. The pooled sensitivity was 96% (80%>99% at 95% confidence interval (CI)) and pooled specificity was 96% (95%>99% at 95% CI). The forest plots are presented below.

<table>
<thead>
<tr>
<th>Study ID</th>
<th>Sensitivity (95% CI)</th>
<th>Specificity (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bayrakutan 2013</td>
<td>0.81 (0.64, 0.92)</td>
<td>1.00 (0.29, 1.00)</td>
</tr>
<tr>
<td>Herliczek 2013</td>
<td>1.00 (0.69, 1.00)</td>
<td>0.96 (0.86, 1.00)</td>
</tr>
<tr>
<td>Johnson 2012</td>
<td>1.00 (0.74, 1.00)</td>
<td>0.97 (0.83, 1.00)</td>
</tr>
<tr>
<td>Koning 2014</td>
<td>0.96 (0.91, 0.98)</td>
<td>0.96 (0.92, 0.98)</td>
</tr>
<tr>
<td>Moore 2011</td>
<td>0.98 (0.87, 1.00)</td>
<td>0.89 (0.76, 0.96)</td>
</tr>
<tr>
<td>Orth 2014</td>
<td>0.93 (0.78, 0.99)</td>
<td>0.98 (0.90, 1.00)</td>
</tr>
<tr>
<td>Thieme 2013</td>
<td>1.00 (0.94, 1.00)</td>
<td>0.89 (0.76, 0.96)</td>
</tr>
<tr>
<td>Hormann 1998</td>
<td>1.00 (0.83, 1.00)</td>
<td></td>
</tr>
<tr>
<td>Overall</td>
<td>0.96 (0.80, 0.99)</td>
<td>0.95 (0.76, 0.99)</td>
</tr>
</tbody>
</table>
**Conclusion:** This meta-analysis found that MRI is an effective and highly accurate imaging modality for diagnosing as well as ruling out acute appendicitis in the pediatric population. Drawbacks of MRI compared to CT include longer scanning times and possibly higher costs.

**References:**
Lower Gastrointestinal

SC-PP-0096

QUALITY OF LIFE AND ANXIETY IN PARENTS OF CHILDREN WITH AN ANORECTAL MALFORMATION OR HIRSCHSPRUNG DISEASE: THE FIRST YEAR AFTER DIAGNOSIS

Marieke Witvliet* 1, Roel Bakx 1, Sander Zwaveling 2, Anton van Dijk 3, Lideke van der Steeg 1, 4

1 Pediatric Surgical Center of Amsterdam, Emma Children’s Hospital AMC and VU University Medical Center, Amsterdam, 2 Wilhelmina Kinderziekenhuis, WKZ, Utrecht, 3 Beatrix Kinderziekenhuis, UMCG, Groningen, 4 Center of Research on Psychology in Somatic Diseases (CoRPS), Department of Medical Psychology, Tilburg University, Tilburg, Netherlands

Aims of the Study: In 2012 we started the KLANKbord-study, a quality of life (QoL) study that follows patients with an anorectal malformation (ARM) or Hirschsprung disease (HD) and their parents from diagnosis till the age of 18 years. We hypothesize that the diagnosis ARM or HD initially has a negative influence on QoL and anxiety levels of parents, but that this influence will diminish over time. The aim of this study is to see whether QoL and anxiety levels of parents change within the first year after the diagnosis.

Method: Parents of all children born, in one of the four participating paediatric surgical centres, with ARM or HD, were eligible for this study. Within 3 months, after the diagnosis ARM or HD, parents received a set of validated QoL questionnaires (e.g. WHOQOL-bref and STAI state) (measurement 1). Measurement two was 12 months after the first questionnaire. Exclusion criteria were the inability to speak the Dutch or English language, and not (yet) completing a follow up questionnaire one year after the first questionnaire. The study was approved by the institutional ethics review board of all hospitals.

Results: During measurement 1 mothers (n=20) scored significantly higher on the social (p-value: 0.01 ; 95% CI 0.3946 – 3.1528) and environmental domain (p-value: 0.01 ; 95% CI 0.4449 – 2.2851) of the WHOQOL-Bref compared to the known reference values. The physical and psychological domain were not significantly different. Fathers (n=19) scored significantly higher on the physical (p-value: 0.01 ; 95% CI 0.2964 – 1.8072), psychological (p-value: 0.001 ; 95% CI 0.7697 – 2.4757) and environmental domain (p-value: 0.003 ; 95% CI 0.5586 – 2.4214) than reference values. The social domain was not significant for fathers. During measurement 2 mothers scored lower on the social and environmental domain, and higher on the physical, psychological and overall domain, of the WHOQOL-bref, compared to measurement 1. During measurement 2 fathers scored higher on the psychological and overall domain. These differences between the measurements were however not significant. Anxiety levels of mothers are lower during measurement 2 compared to measurement 1 (10.60 vs 10.95). Anxiety levels of fathers are higher during measurement 2 compared to measurement 1 (9.76 vs 9.06). These differences are not significant. Anxiety levels of mothers were significantly higher than anxiety levels of fathers during measurement 1 (p-value: 0.002 ; 95% CI 0.808 – 2.956). During measurement 2 this difference in anxiety of mothers versus fathers does not exist (p-value: 0.373 ; 95% CI -1.157 – 2.922).
Conclusion: A significant negative influence on the QoL of parents having a child with ARM or HD, compared with the reference population was not seen in this population. In addition QoL does not change significantly during the first year for both fathers and mothers. Anxiety levels of mothers do decline during this first year. The amount of parents that were included in this study are still small, what might influence our results. That is why we will continue the KLANKbord-study for many more years.
Lower Gastrointestinal
SC-PP-0097

IS PRE-OPERATIVE IRRIGATION WITH SERUM SALINE NECESSARY IN PERFORATED APPENDICITIS, OR IS ONLY ASPIRATION ENOUGH?

Ufuk Tan Aygün1, Cetin Ali Karadağ1, Basak Erginel*1, Melih Akin1, Nihat Sever1, Abdullah Yildiz1, Ali Ihsan Dokucu1
1pediatric surgery, Sisli Etfal Education and Research Hospital, istanbul, Turkey

Aims of the Study: In cases of perforated appendicitis, intra-abdominal irrigation with serum saline is the most commonly used technique to prevent an intra-abdominal abscess (IAA) after an appendectomy. But some authors offer only aspiration. The aim of this study was to compare the efficiency of both methods in preventing IAA in pediatric patients with perforated appendicitis.

Method: One hundred patients who had undergone operations for perforated appendicitis were included to this study. Fifty-two patients underwent aspiration of the abdomen, while 48 underwent intra-abdominal irrigation with serum saline during appendectomy. Of the 52 patients who underwent aspiration during appendectomy, 28 patients were open appendectomies (Group 1: open aspiration, or OA), and 24 patients were laparoscopic appendectomies (Group 2: laparoscopic aspiration, or LA). Of the 48 patients who underwent irrigation during perforated appendicitis operation, 27 patients was operated via laparotomy (Group 3: open irrigation, orOI), and 21 patients was operated via laparoscopy (Group 4: laparoscopic irrigation, or LI). We checked patients’ white blood cell (WBC) counts and C-reactive protein (CRP) levels before the operation and on the fifth post-operative day. On the fifth post-operative day, we applied control abdominal ultrasonography (USG) to check patients for IAA.

Results: Between the irrigation and aspiration groups, pre-operative and post-operative WBC and CRP results were not statistically significant (p > 0.05). The number of patients with IAA was 2 in Group 1 (7.1%), 1 in Group 2 (4.2%), 2 in Group 3 (7.4%), and 2 in Group 4 (9.5%). Between the open surgery (Groups 1 and 3) and laparoscopic surgery (Groups 2 and 4) groups, there was no statistically significant difference in IAA rates (p > 0.05). IAA rates between the aspiration (Groups 1 and 2) and irrigation (Groups 3 and 4) groups were not statistically significant (p > 0.05).

Conclusion: We conclude that the aspiration technique can be used safely in pediatric patients with perforated appendicitis.
References

BIOMARKERS OF KIDNEY INJURY IN NEWBORNS WITH ABDOMINAL COMPARTMENT SYNDROME

Dmitry Morozov1,2, Olga Morozova3, Alexey Tsyplacov4, Ivan Budnik3, Natalia Zacharova4

1Pediatric surgery, Scientific Centre of Children Health, 2Pediatric surgery, 3pathophisiology, I.M. Sechenov First Moscow State Medical University, Moscow, 4Pediatric surgery, Saratov State Medical University, Saratov, Russian Federation

Aims of the Study: To test the levels of urinary biomarkers of kidney injury in newborns with abdominal compartment syndrome (ACS).

Method: The study was approved by the local research ethics committee. A total of 35 newborns with ACS were divided into three groups: Group 1 included newborns with gastroschisis, Group 2 included newborns with false diaphragmatic hernia, and Group 3 included newborns with esophageal atresia with distal tracheoesophageal fistula. Intra-abdominal pressure (IAP) was evaluated by intravesical manometry. The levels of urinary markers of distal tubular damage (π-GST), angiogenesis (VEGF) and inflammation (MCP-1) were measured by ELISA at admission (point 1), on day 4 (point 2) and day 10 (point 3) after surgery. Control group consisted of 20 healthy newborns. Informed parental consent was obtained in all cases. Data are presented as Mean ± SD. Differences were considered statistically significant when the P value was less than 0.05; P – comparison vs. control, P1 – vs. point 1, P2 – vs. point 2.

Results: In control, IAP was 3.1 ± 1.3 mm Hg, π-GST was 1.5 ± 0.2 μg/L, VEGF was 58 ± 4 pg/mL, and MCP-1 was 58 ± 3 pg/mL. In Group 1, at point 1, levels of IAP and π-GST did not differ from those in control, VEGF and MCP-1 increased to 89 ± 4 and 199 ± 50 pg/mL (P < 0.001 for both), respectively. At point 2, all measured parameters reached their maximum values: IAP was 22.0 ± 2.5 mm Hg (P1 < 0.001), π-GST was 10.7 ± 1.9 μg/L (P1 < 0.001), VEGF was 171 ± 7 pg/mL (P1 < 0.001), and MCP-1 was 404 ± 106 pg/mL (P1 = 0.002). At point 3, all of them decreased but were still higher than at point 1: IAP was 12.6 ± 1.6 mm Hg (P1 < 0.001, P2 < 0.001), π-GST was 5.5 ± 1.2 μg/L (P1 < 0.001, P2 < 0.001), VEGF was 139 ± 7 pg/mL (P1 < 0.001, P2 < 0.001), and MCP-1 was 289 ± 31 pg/mL (P1 = 0.002, P2 = 0.006). In Group 2, at point 1, the values of all parameters, except π-GST, were higher than in control: IAP was 6.0 ± 1.7 mm Hg (P = 0.006), VEGF was 82 ± 12 pg/mL (P < 0.001) and MCP-1 was 193 ± 41 pg/mL (P < 0.001). At point 2, all measured parameters reached their maximum values: IAP was 19.0 ± 2.6 mm Hg (P1 < 0.001), π-GST was 9.7 ± 1.6 μg/L (P1 = 0.002), VEGF was 165 ± 20 pg/mL (P1 = 0.002), and MCP-1 was 376 ± 154 pg/mL (P1 = 0.002). At point 3, all the parameters partially decreased: IAP was 11.4 ± 1.2 mm Hg (P1 < 0.001, P2 < 0.001), π-GST was 4.3 ± 1.0 μg/L (P1 = 0.002, P2 = 0.002), VEGF was 135 ± 8 pg/mL (P1 = 0.002, P2 = 0.002), and MCP-1 was 278 ± 24 pg/mL (P1 = 0.002, P2 = 0.04). In Group 3, at point 1, all measured parameters sharply increased: IAP was 13.5 ± 1.9 mm Hg (P < 0.001), π-GST was 6.0 ± 2.4 μg/L (P < 0.001), VEGF was 129 ± 13 pg/mL (P < 0.001), and MCP-1 was 309 ± 30 pg/mL (P < 0.001). At point 2, IAP decreased to 7.3 ± 1.9 mm Hg (P < 0.001, P1 < 0.001), VEGF decreased to 101 ± 9 pg/mL (P < 0.001, P1 = 0.012), MCP-1
decreased to 187 ± 17 pg/mL ($P < 0.001$, $P_1 = 0.012$), and $\pi$-GST remained the same. At point 3, IAP and VEGF remained the same as at point 2, $\pi$-GST decreased to 2.1 ± 1.0 μg/L and did not differ from control ($P = 0.05$, $P_2 = 0.04$), and MCP-1 also decreased but was still higher than in control, reaching 108 ± 44 pg/mL ($P < 0.001$, $P_2 = 0.02$).

**Conclusion:** Changes in the urinary concentrations of kidney injury biomarkers correlate with the severity of ACS. Estimation of the urinary biomarkers is a promising non-invasive approach for early preclinical diagnosis of nephropathy in newborn patients with ACS.
MODE OF DELIVERY IN THE NEONATE SURGICALLY ASSESSED FOR BILIOUS VOMITING

Andrew R Ross¹, Martin F Lister², Amanda McCabe³

¹Dept. of Paediatric Surgery, ²Dept. of Paediatric Medicine, Royal Hospital for Sick Children, Edinburgh, United Kingdom

Aims of the Study: It was postulated that those neonates with bile-stained vomiting in whom a surgical cause has been radiologically excluded, may have experienced a higher incidence of minor birth trauma. We examined the mode of delivery in this cohort.

Method: All neonates with bilious vomiting referred consecutively for surgical opinion over a 5 year period (2008-2013) were included. These data were retrospectively retrieved from neonatal and surgical databases and cross-referenced against radiology records. Outcomes included diagnosis, morbidity and demography. A sub-group analysis of those that had a surgical cause excluded (normal upper gastro-intestinal contrast study and abdominal ultrasound scan) was performed.

Results: Ninety-six neonates were retrieved for surgical opinion with bilious vomiting from 10 regional centres (55 male: 41 female). Median age at time of transfer was 2 days (IQR 1-3d) with a median gestation of 40 completed weeks (IQR 38-41 weeks). Following clinical assessment and diagnostic imaging, 63 had a surgical cause excluded. Twenty-six of these neonates (41.3%) were born by spontaneous vaginal delivery (SVD), 23 (36.5%) were delivered by caesarean section (CS) and 13 (20.6%) had instrumental deliveries (ID). In 2013 the nationally reported percentages for each birth category (Information Services Division Scotland), 58.7% were SVD, 28.5% CS and 12.4% ID. A chi-square test of independence was performed which showed a significant difference between groups (p<0.002).

Conclusion: In the cohort where surgical pathology had been excluded, the frequency of Caesarean section and instrumental delivery was significantly different to that reported in the national mode of delivery data series (ID at almost double reported rates). The increase in ‘stressful’ and traumatic delivery methods described may account for the presence of bile-stained vomitus in the neonatal period. To our knowledge this is the first description of this observation. Further investigation in a wider neonatal population is suggested in order to better understand the presence of these symptoms following the radiological exclusion of surgical pathology.
CONTRIBUTING FACTORS FOR NECROTOIZING ENTEROCOLITIS IN PRETERM INFANTS IN NICU

Zlatan Zvizdic¹, Suada Heljic², Kenan Karavidic¹

¹Department of Pediatric Surgery, ²Pediatric Clinic, NICU, University Clinical Center Sarajevo, Sarajevo, Bosnia and Herzegovina

Aims of the Study: to estimate the influence of potentially contributing factors of this multifactorial disease

Method: The study group included 51 preterm infants <37 gestational weeks with diagnosis of necrotizing enterocolitis, hospitalized in NICU during 5 years period. The control group consisted of 71 patients with approximately the same gestational age and birth weight. Average gestational age in the study group was 30,2 weeks (SD 3, 7), average birth weight 1502, 75 g (SD 781, 5). Average postnatal age in the time of presenting NEC was 18,2 days (SD 12, 8)

Results: The model of logistic regression is done to estimate the influence of risk factors related to the treatment of sick preterm infants on the likelihood of NEC development. Model consisted of 7 independent variables (nosocomial infections, mechanical ventilation, NCPAP, morphine, inotrops, transfusions, and H2 blockers). Whole model was statistically significant, X² (7, n=1222) = 49.522, p<0.0001; two independent variables (nosocomial infections and H2 blockers) made statistically significant contribution to the model. Preterm infants with nosocomial infections prior of development of NEC have 3 time greater chance of developing NEC, and infants receiving H2 blockers have chance to develop NEC 1.5 higher than other.

Conclusion: Underlying pathology contributes to NEC development and identifying risk factors can be crucial for the early diagnosis and outcome of disease.
**Thoracic**

SC-PP-0101

**SPARING-LUNG SURGERY AND CONGENITAL LUNG MALFORMATIONS: 15-YEAR EXPERIENCE**

Francesca Destro¹, Noemi Cantone*¹, Michela Maffi¹, Beatrice Randi¹, Giovanni Ruggeri¹, Mario Lima¹

¹Pediatric Surgery, S. Orsola, Bologna, Italy

**Aims of the Study:** Congenital lung malformations (CLM) require surgical removal to avoid malignancies and respiratory tract infections. Lobectomy has been the procedure of choice for many years. Nowadays patients are identified prenatally and treated prophylactically. In this context, lung-sparing surgery (LS) has gained interest as a lung preservation strategy, especially for asymptomatic patients. We report our experience with thoracoscopic and open sparing-lung resections.

**Method:** We retrospectively reviewed the charts of all patients treated by lung-sparing resection (segmentectomy and atypical resection) for CLM from January 2001 to January 2015. Data were collected regarding preoperative diagnostic workup, type of intervention, and follow up.

**Results:** One-hundred and nineteen patients were treated by LS for CLM. We performed 81 thoracoscopies, with 42 cases requiring open conversions. There were 9 postoperative complications: five asymptomatic pneumothoraces, one tension pneumothorax that required intervention (drainage) and 3 bleedings. During the follow-up (mean time 70.2 months) 4 patients had pneumonia. We experienced one recurrence that required a second-look surgery.

**Conclusion:** LS is safe and effective to treat CLM. Complication rate is acceptable and not related to preoperative symptoms. In our experience, this type of lung surgery does not carry a higher risk of residual disease and recurrence if accurately planned in selected patients, i.e., those with small asymptomatic lesions.
Aims of the Study: The definition and the management of long-gap esophageal atresia (LG-EA) are still debated. The aim of our study is to investigate the effectiveness of our treatment strategy in the management of LG-EA, thus evaluating if a delayed decision on the surgical strategy may influence the options for surgical repair.

Method: Clinical and surgical records of patients operated on for esophageal atresia in our Institution between January 2004 and December 2014 were retrospectively reviewed. LG-EA was considered when, after introducing two endoscopes in the esophageal pouches, a distance ≥ 3 vertebral bodies was found at a radiological investigation. Only patients with LG-EA who underwent at least two subsequent measurements of the gap, before the definitive decision on the surgical repair was done, were included. The variation in the gap between the two measurements was analysed with the two-tailed paired T-test. We analysed the type of esophageal atresia, according to the Ladd-Gross classification, and its influence on the gap variation and on the surgical strategies thanks to the t-test and Fisher exact test respectively. A p-value <0,05 was considered as statistically significant.

Results: 64 patients were operated on for esophageal atresia at our Institution between 2004 and 2014. A diagnosis of LG-EA was done in 17/64, but only 12 patients had at least two subsequent measurements of the gap and were included in the study. 5/12 (42%) had a type A esophageal atresia, 4/12 (33%) a type B, and 3/12 (25%) a type C. The first measurement was performed at 1,5 days of life (range 0-112, DS 35) and the gap was on average 5,25 vertebral bodies (range 3-9, DS 1,62). The second measurement was performed at 70 days of life (range 23-153, DS 45,25) and the gap was on average 3,5 (range 0-6, DS 1,9). 6/12 (50%) underwent delayed esophageal anastomosis while 6/12 (50%) had esophageal replacement. The gap variation between the two measurements was statistically significant (p=0,0228). The type of esophageal atresia seemed not to influence the degree of gap reduction and the selected surgical strategies.

Conclusion: A significant spontaneous reduction of the gap, without adopting any particular kind of lengthening strategy was found in our series. In our opinion the feeding through the gastrostomy and probably the physiological gastroesophageal refluxes may have a role in the development of the distal pouch, thus reducing the gap. We therefore believe that the definitive surgical decision and the opening of a cervicostomy should be taken only after repeating the gap measurement between the second and the third month of life. In fact in our series, thanks to this strategy, we could succeed in saving the native esophagus and performing an esophageal anastomosis in 50% of the LG-EA cases.
SURGICAL STRATEGIES IN CONGENITAL DIAPHRAGMATIC HERNIA; EXPERIENCE WITHIN THE VICI-TRIAL

Kitty Snoek, Pietro Bagolan, Lucas Wessel, Ivo de Blaauw, Francesco Morini, Irwin Reiss, Dick Tibboel, René Wijnen and CDH EURO Consortium

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Aims of the Study: Congenital diaphragmatic hernia (CDH) is a severe congenital anomaly and a surgical repair is performed after cardiorespiratory stabilization of the neonate. Although infants are nowadays treated according to a standardized neonatal treatment protocol implemented within the VICI-trial, the surgical repair is not standardized. We have evaluated the surgical strategies within this RCT to determine which strategy had the best surgical outcome.

Method: In an international multicenter prospective RCT of initial ventilation strategy (NTR 1310) in Europe, antenatally diagnosed CDH infants were included. Indication for MAS depended on hemodynamic stability, extracorporeal membrane oxygenation (ECMO), liver position and scaling of the defect.

Results: Of the 171 included infants, surgical repair was performed in 145 infants (84.8%), 30 of which (20.7%) initially underwent minimal access surgery (MAS). Eleven of these infants were converted to open repair and evaluated in the open repair group, leading to 19 (13.1%) MAS and 126 (86.9%) open repair. Eleven infants (7.9%) had a Boston scale A, 49 infants (35.0%) Boston scale B, 68 infants (48.6%) Boston scale C, 12 infants (8.6%) Boston scale D, and in 5 infants the size of defect was not registered. Patch repair was performed in 92 infants (63.4%) and primary closure in 53 infants (36.6%). Surgical repair was performed in 57 infants (39.3%) in theatre and in 88 infants (60.7%) in the intensive care unit. After exclusion of patients that were operated on ECMO (n=19), there were no significant differences between patients that underwent surgical repair in theatre (n= 54) as compared to in the ICU (n= 72) with regard to mortality (p=0.18) and recurrence within one year (p=0.07) (Table 1). Four infants in the MAS group (21.1%) and 6 infants (4.8%) who underwent an open repair had a recurrence (p=0.009). There was no significant difference in the recurrence between the four Boston scales (p=0.39).
Table:

<table>
<thead>
<tr>
<th></th>
<th>Repair at theatre (n=54)</th>
<th>Repair at ICU (n=72)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left sided CDH</td>
<td>48 (88.9%)</td>
<td>65 (90.3%)</td>
<td>0.80</td>
</tr>
<tr>
<td>Liver up</td>
<td>25 (46.3%)</td>
<td>39 (54.2%)</td>
<td>0.38</td>
</tr>
<tr>
<td>Open repair</td>
<td>36 (66.7%)</td>
<td>71 (98.6%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Mortality</td>
<td>1 (1.9%)</td>
<td>5 (6.9%)</td>
<td>0.18</td>
</tr>
<tr>
<td>Recurrence</td>
<td>7 (13.0%)</td>
<td>3 (4.2%)</td>
<td>0.07</td>
</tr>
</tbody>
</table>

Caption: Table 1

Conclusion: Despite a standardized neonatal treatment protocol for CDH, significant differences within the VICI-trial exist in terms of place of surgical repair, surgical approach and use of patch repair. Place of repair (ICU or theatre) did not influence recurrence and mortality rate but has an impact on type of repair. Despite the increase in use of MAS, only 13.1% of the infants underwent MAS, so it is not standard of care yet. MAS is performed almost exclusively in theatre and seems associated with higher risk of recurrence within one year of life.

**Upper Gastrointestinal**

SC-PP-0104

**LAPAROSCOPIC SLEEVE GASTRECTOMY IN PEDIATRIC AND ADOLESCENT POPULATION: MIDTERM RESULTS.**

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**Aims of the Study:** Obesity is a chronic and progressive disease, associated with related metabolic disorders, causing severe morbidity and mortality in children and adolescents. Aim of this study is to the feasibility of the laparoscopic sleeve gastrectomy (LSG) in pediatric and adolescent population.

**Method:** A retrospective review of all pediatric patients undergoing LSG was performed. Demographic data, access to care, weight, co-morbidities, complication rate were analyzed. Regression analyses were used for estimating the relationships among variables, all values of p<0,05 were considered statistically significant. LSG was performed using a 5 port technique and the stomach was longitudinally resected alongside a 40 Fr bougie with multiple loads of laparoscopic staplers. Postoperative swallow studies with watersoluble contrast were performed on the second postoperative day to detect early leaks and evaluate gastric empty. When this study was satisfactory, patients were placed on clear liquids and discharged two days later with detailed dietary instructions for a semiliquid diet for 1 week and progression to a pureed diet until the first office visit.

**Results:** From June 2013 to July 2014, 15 patients with mean age 15,8 years (range 13,7 – 19,2) underwent to LSG. Nine of these were female (9/6 M/F). Four patients had Prader Willi Syndrome, one had Bardet Biedl Syndrome. Mean pre-op weight was 127,7 ± 26,67 Kg (Range 70 – 198 Kg). Mean pre-op BMI was 46,97 ± 4,48 (range 40,9 – 56,3). Mean Follow-up was 9 months (range 1-12 months). Post-op mean weight was 101,2 ± 21,2, the difference between pre and post-op values was statistically significant (p<005). Post-op mean BMI was 37,6 ± 5,1 the difference between the 2 groups was statistically significant (p<005). Mortality was 0%. Mean EWL was 47.2 %. Mean operative time was 72 minutes (range 65–132 min). There were no complications or deaths. Comorbidities were completely resolved or ameliorated.

**Conclusion:** LSG is feasible and safe in morbidly obese adolescents, achieving efficient weight loss and impressive resolution of comorbidities. Further studies are required to evaluate the long-term results of this procedure as well as its place among other bariatric options especially regarding the complications for long-term follow-up.
Upper Gastrointestinal

SC-PP-0105

LARGE CONGENITAL PARAESOPHAGEAL HERNIA AND THE RISK OF INTRATHORACIC GASTRIC VOLVOLUS

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Aims of the Study: Congenital paraesophageal hernia (CPH) is rare in infants and children. It is unusually large with herniation of most of the stomach which can lead to intrathoracic gastric volvulus. This report describes our experience with twelve infants and children with large CPH, three of them (25%) presented with intrathoracic gastric volvulus.

Method: Twelve infants and children with large CPH, three of them presented with intrathoracic gastric volvulus were reviewed.

Results: Between 1992 and 2012, twelve infants and children with large CPH were treated. There were 7 males and 5 females. Their age ranged from 2 days to 2.5 years (mean 16.3 months). Two presented acutely, immediately after birth with respiratory distress secondary to a large CPH. One of them had intrathoracic gastric volvulus. Four presented with recurrent chest infection and failure to thrive while the remaining three had repeated attacks of vomiting. Another patient was a case of repaired esophageal atresia and tracheoesophageal fistula. He presented at the age of 2.5 years with recurrent attacks of cough and vomiting of one year duration. He was found to have a large left paraesophageal hernia. Two patients were sisters and both of them presented with recurrent chest infection and failure to thrive. At the time of presentation, both of them had large CPH with intrathoracic gastric volvulus.

Conclusion: CPH is rare in the pediatric age group and sometimes unusually large with herniation of most of the stomach which may result in intrathoracic gastric volvulus. Awareness of this and early diagnosis and treatment are important to avoid subsequent morbidity and mortality.

Upper Gastrointestinal

MALROTATION – AGE-RELATED DIFFERENCES IN RE-OPERATION RATE

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Aims of the Study: Intestinal malrotation classically presents in a neonate with bilious vomiting. Population studies suggest that up to two thirds of these patients are diagnosed later in childhood or in adulthood.1 Increased morbidity in the adult population has been reported.2 Local experience suggested that surgery was technically more difficult in older children and led to the hypothesis that it would be associated with increased morbidity.

Method: Retrospective case note analysis. All children presenting with intestinal malrotation to a tertiary referral centre between January 2002 and April 2014 were included. Case notes and operation records were reviewed and those who underwent laparotomy for confirmed malrotation were included. Children were grouped as infants (< 1 year) and older (> 1 year) for comparison. The primary outcome measure was emergency re-operation rate. Statistical analysis was performed using Fisher's exact test.

Results: One hundred thirty five children with malrotation were identified (105 infants, 30 older children; 81 male; age range 0 – 16 years). Twelve patients had emergency re-laparotomy following initial Ladd’s procedure (6 in each group). Risk for reoperation was significantly higher in older children (p = 0.026), requiring revision of Ladd’s procedure in 4, and bypass/resection in 2. One child required careful preservation of collaterals based on the inferior mesenteric vein as a result of thrombotic occlusion of the superior mesenteric vein as a direct consequence of chronic volvulus. There were 3 deaths, all in the infant group. One was directly associated with malrotation with extensive bowel necrosis. The other 2 died of unrelated sepsis several months later.

Conclusion: Malrotation surgery in older children is associated with a significantly higher emergency re-operation rate. Longstanding chronic intermittent obstruction associated with malrotation may require primary duodenal bypass procedure if simple Ladd’s procedure is deemed inadequate.

Aims of the Study: Treating of the long gap esophageal atresia (LGEA) makes a lot of controversies between surgeons around the world and the lack of optimal approach is still observed. Clear definition of LGEA is also missing, however majority of researchers define LGEA as impossibility to achieve true primary esophageal anastomosis. It usually corresponds to gap between both esophageal pouches that exceeds 2,5-3 cm or more than three vertebral heights. Lately, John Foker and colleagues described a new technique of LGEA reconstruction. It uses an axial traction sutures on the esophageal segments to induce growth until a primary repair seems possible. Foker technique was introduced into few pediatric surgery departments around the world, including four major pediatric centers in Europe.

Method: 49 children with LGEA were treated between 2005-2013 in four European pediatric surgery centers: Department of Surgery and Urology for Children and Adolescents, Medical University of Gdansk, Poland, Department of Pediatric Surgery in Leipzig, Germany, Department of Pediatric Surgery Charles University in Prague, Czech and Department od Pediatric Surgery University Hospital Vall d’Hebron in Barcelona, Spain. There were 20 patients with pure esophageal atresia (EA) and 29 with tracheo-esophageal fistula (26 with TEF of distal pouch and 3 with TEF of proximal pouch) and at presentation gap ranged from 2,5 to 14 cm. 10 patients had created spit fistula. Age varied from 1 to 910 days. All patients were operated on according to Foker technique principle and data was analyzed retrospectively.

Results: In our series the true primary esophageal anastomosis was achieved in 92% of patients and only 4 children received esophageal replacement (stomach, jejunum).

Foker technique was associated with high complication rate of which strictures of esophageal anastomosis were most frequent. We observed them in 54% of children with primary esophageal anastomosis but in 84% we were able to restore normal esophageal lumen using only series of dilatations.

Gastro-esophageal reflux was observed in 66% of children after esophageal anastomosis and occurred most frequently in cases where initial gap exceeded 3,5 cm.

Anastomosis leak was observed in 26% of patients and occurred mostly in children with spit fistulas. In the group without spit fistula anastomosis leakage occurred only in 12%.
Conclusion: Unique and undisputed advantage of Foker technique is preservation of the own, native esophagus. According to our results, Foker technique is a safe, repeatable and efficient method of the esophageal lengthening in specialized departments of pediatric surgery. Initially high rate of complications resulted from the lack of experience at the time of introducing the method into departments and could be averted in future. Clear algorithm that was proposed in this paper simplifies schema of LGEA treatment and allows comparing the results in objective ways.
**Urology**

SC-PP-0108

**CURRENT MANAGEMENT OF CRYPTORCHIDISM ASSOCIATED WITH GASTROSCHISIS.**

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**Aims of the Study:** The recent case of a baby boy born with gastroschisis with extrusion of right testis through the abdominal wall defect prompted us to provide a review of available evidence on the management and outcomes of cryptorchidism associated with gastroschisis.

**Method:** Our case was considered in conjunction with an extensive review of the literature conducted by 2 independent researchers searching Medline, EMBASE, Google Scholar, and Cochrane databases, with additional cases chosen from the references of selected articles.

**Results:** A total of 24 studies were retrieved; of these 16 (127 patients, including our own one) fulfilled the entry criteria. The reported rates of cryptorchidism among males with gastroschisis ranged 5 to 40 percent. These rates appeared increased when compared to historical healthy controls1,2, reaching statistical significance in the majority of the studies. Complete data for management and outcome analysis were available in 97 patients, involving 124 cryptorchid testes (27 bilateral cases). Of these, 29 (23%) testes were lying outside the abdominal wall defect. Overall, 11 (9%) testes underwent neonatal orchydopexy, which was predominantly performed for extra-abdominal testes (p=0.0004). Fifty-six (49.5%) of the remaining testes descended spontaneously to the scrotum within a year follow-up. Such spontaneous descent rate was similar among the group of extra-abdominal testes and the remainder of cryptorchid testes, and was statistically significantly lower than that of historical healthy controls2. Overall, successful outcome was achieved in 97 (78%) of the 124 cryptorchid testes, being significantly lower in the extra-abdominal testes group (62% vs. 83%; p=0.02). Notably, such difference did not correlate with the higher percentage rate of neonatal orchydopexy in the extra-abdominal testes group.

**Conclusion:** Cryptorchidism seems frequently associated with gastroschisis, and is likely underreported. Such event is associated with significant testicular morbidity, especially when the ectopic testis lies outside the abdominal wall defect.

**References:**

**Aims of the Study:** There are no evidence-based guidelines for treatments of labial adhesions. The aim of the study was to compare the outcome of treatment with topical oestrogen with that of manual separation.

**Method:** A retrospective chart review was carried out for all females 0-12 years old referred to a tertiary centre for paediatric surgery between 1999 and 2013. Different treatments used for labial adhesions and their effectiveness regarding success rates, recurrences and outcomes for different treatment lengths were registered. Furthermore, a long-term follow-up for late recurrences and family concern was performed as a cross-sectional study based on a telephone questionnaire. A score of 1-5 (5=best) was used for estimation of parents’ degree of inconvenience with the treatment. Manual separation was performed either during sedation with midazolam and locally applied anaesthetics, or under general anaesthesia. The administration of topical oestrogen was demonstrated for the parents by doctors. The study was ethically approved (49/2010).

**Results:** 80 patients were referred and 76 received in total 160 treatments of which 130 were oestrogen courses (81%) and 30 treatments with manual separation (19%). Median age at the first treatment was 19 (2-86) months and follow-up was 3 (0-126) months. The number of initial successes for each oestrogen course was 78/130 (60%) but adhesions recurred in 32 (41%). Manual separation had initial success in 29/30 (97%) and recurrences in 8(28%). The total number of successful treatments without recurrences was higher for manual separation 21/30 (70%) than for oestrogen 46/130 (35%) (p=0.001). Duration of treatment with oestrogen was registered for 85 courses and the median treatment length for each course was 4 (1-12) weeks. The success rate without recurrences did not differ between treatments given for more than four weeks (20/49) or less (11/36) (p=0.369). Side effects for topical oestrogen were reported by 2 patients (3%) and none after manual separation. In the long-term follow-up parents of 42/76 (55%) patients participated and for 35 the last reported treatment was either oestrogen or manual separation. Follow-up was 53 (0-144) months. Persisting or recurrent adhesions after the last treatment was reported by 11/26 (42%) patients treated with oestrogen and 2/9 (22%) treated with manual separation (p=0.431). Parents estimated inconvenience with each treatment as 4 (1-5) and 68% would recommend treatment with oestrogen to others while 73% would recommend manual separation.

**Conclusion:** Manual separation overall had a better outcome with fewer recurrences and no adverse effects, and can be considered either as a first or second line treatment for labial adhesions. A prospective randomised study is needed for more structured treatment recommendations.
General SC-GE-0110 to SC-GE-0123

General

SC-GE-0110

LOW DIASTOLIC BLOOD PRESSURE IN TERM INFANTS WITH CONGENITAL HEART DISEASE AND SEVERE NECROTIZING ENTEROCOLITIS

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Aims of the Study: Necrotizing enterocolitis (NEC) is a devastating disease of the newborn infant that relatively frequently affects term infants with congenital heart disease (CHD). Currently, the underlying mechanism of the increased risk of NEC in term infants with CHD remains poorly understood. One of the hypotheses states that low body perfusion due to the retrograde diastolic flow in the ductus arteriosus, represented by a low diastolic blood pressure, results in ischemia of the intestine. Therefore, we aimed to investigate whether diastolic blood pressure is lower in term infants with CHD who develop NEC (CHD+NEC) compared to term infants with CHD who do not develop NEC (CHD–NEC). Our second aim was to investigate whether a low diastolic blood pressure could be used as a predictive value for severe NEC.

Method: Between December 2003 – December 2013, 13 infants out of 565 infants with CHD and a gestational age >35 weeks, admitted to our NICU, developed NEC and were retrospectively included in this case-control study. These infants were matched 1:1 with patients who did not develop NEC from the same cohort. We matched by type of CHD and date of birth. The mean diastolic blood pressure values, measured either invasively or non-invasively, were collected at first day after admission, two days and one day prior to NEC onset. Severity of disease was assessed by Bell’s stage. We used a Mann Whitney U test to assess differences in diastolic blood pressure between CHD+NEC and CDH–NEC infants, between CHD+NEC (Bell’s stage 1) and CHD+NEC (Bell’s stage 3), between CHD+NEC (Bell’s stage 2) and CHD+NEC (Bell’s stage 3) and between CHD–NEC and CHD+NEC (Bell’s stage 3).

Results: Median age of NEC onset was 5 days (interquartile range (IQR) 2.5-8). We found no significant differences in mean diastolic blood pressure between NEC cases and matched controls at the day of admission and two or one day(s) prior to NEC onset. However, when NEC cases were categorized by Bell’s stage, diastolic blood pressure tended to be lower at day one of admission in infants with Bell’s stage 3 (median 34.4 mmHg, IQR 34.2-36.5) compared to Bell’s stage 1 (median 49.7 mmHg, IQR 41.6-57.8, p=0.08) and compared to Bell’s stage 2 (median 43.9 mmHg, IQR 39.7-47.1, p=0.07). Furthermore, mean diastolic blood pressure in the three Bell’s stage 3 patients (median 34.4 mmHg, IQR 34.2-36.5) compared to their matched controls (median 41.8 mmHg, IQR 40.3-45.9) tended to be lower at day one after admission (p=0.05).
Conclusion: While low diastolic blood pressure might not be associated with the occurrence of NEC in CHD patients, it might be associated with a complicated course of NEC. Diastolic blood pressure however may not be the most accurate measure to predict tissue perfusion.
I-FABP AND CITRULLINE LEVELS IN NEC PATIENTS IN THE FIRST 48 HOURS AFTER ONSET OF DISEASE

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Aims of the Study: Intestinal fatty acid-binding protein (I-FABP) is a promising marker for necrotising enterocolitis (NEC). I-FABP is a small cytosolic protein specifically localized in enterocytes. When enterocytes are being compromised by NEC, I-FABP is released into the bloodstream directly after onset of disease. High I-FABP levels are associated with a complicated course of NEC. It remains unknown whether the subsequent decline of plasma I-FABP levels reflects recovery of the epithelium or reduced turnover of enterocytes with depletion of intracellular levels of I-FABP. Citrulline is a non-protein amino acid, used as an indicator of functional enterocyte mass. The present study focuses on clarifying whether there is an association between I-FABP levels and functional enterocyte mass as measured by citrulline.

Method: We included twenty-two (n=22) neonates with proven NEC (determined via abdominal X-ray or intraoperatively confirmed). Plasma I-FABP levels were measured using a commercially available ELISA, every eight hours for the first 48 hours after onset of symptoms or until surgery, whichever came first. Citrulline levels were measured in the same samples with high-performance liquid chromatography (HPLC). The courses of I-FABP and citrulline levels were analysed over time; post-hoc testing revealed significant differences between the time blocks. Spearman Rho correlation coefficients of I-FABP versus citrulline levels and mean I-FABP:citrulline ratios were determined for each 8-hour time block.

Results: Eleven patients (50%) developed NEC Bell’s stage 2; the remaining patients (50%) NEC Bell’s stage 3. Overall survival rate was 73%. Both citrulline (ANOVA (F(5,59) = 2.963, p=0.019) and I-FABP (Kruskal-Wallis χ²(5) =19.324, P=0.002) levels demonstrated a significant decrease. At baseline (TB1), the mean I-FABP:citrulline ratio measured over the complete cohort was 3.76 (SD 4.67) and remained unchanged during the 48 hours after onset of NEC (Kruskal-Wallis χ²(5) =7.496, p=0.186). Higher mean ratios were found in the NEC Bell’s stage 3 subgroup as compared to the NEC Bell’s stage 2 subgroup. This difference was primarily caused by an increase of I-FABP levels, thereby almost reaching statistical significance in time blocks TB3, TB4 and TB5.

Conclusion: I-FABP levels peak immediately after first symptoms of NEC and rapidly decrease thereafter. Citrulline levels decrease later during the course of disease, resulting in a not significantly changed I-FABP:citrulline ratio within 48 hours after onset of NEC. This result suggests that a NEC-induced loss of functional small bowel enterocytes reflects a reduced enterocyte turnover and depletion of intracellular I-FABP levels. More research is necessary to determine the clinical relevance of monitoring citrulline levels in NEC patients.
ASSOCIATION OF TISSUE OXYGENATION AND INTESTINAL FATTY ACID BINDING PROTEIN IN PLASMA DURING THE DEVELOPMENT OF NECROTIZING ENTEROCOLITIS

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**Aims of the Study:** We aimed to determine whether cerebral and splanchnic fractional tissue oxygen extraction (FTOE) correlate with intestinal fatty acid binding protein in plasma (I-FABPp), a marker for intestinal damage, in infants with NEC. Furthermore, we investigated if cerebral and splanchnic FTOE values, and on the other hand I-FABPp levels could discriminate between uncomplicated (conservative treatment) and complicated NEC (surgery or death).

**Method:** We included 19 preterm infants with NEC (9 uncomplicated, 10 complicated). Using NIRS, regional cerebral and splanchnic tissue oxygen saturations were measured continuously for 48 hours after NEC onset. I-FABPp was measured simultaneously. We used the Spearman correlation tests to calculate correlation coefficients between FTOE and I-FABPp in uncomplicated and complicated NEC.

**Results:** Median (range) gestational age was 28 (25-36) weeks, birth weight 1290 (740-2400) grams. Cerebral and splanchnic FTOE values correlated strongly with I-FABPp levels (rho between .745 and .900; \( P < .001 - .037 \)) in the first 16 hours after NEC onset. Thereafter, splanchnic FTOE increased with concomitant declining I-FABPp in infants with uncomplicated NEC, whilst in infants with complicated NEC both splanchnic FTOE and I-FABPp declined.

**Conclusion:** This study shows strong correlations between cerebral and splanchnic FTOE values and I-FABPp levels in the first 16 hours after disease onset, suggesting that FTOE values can be used to assess information about the intestinal damage. After the first 16 hours after disease onset, cerebral and splanchnic FTOE values may be used to discriminate between progression or recovery of intestinal damage in NEC.
NECROTIZING ENTEROCOLITIS VERSUS ‘CARDIOGENIC NECROTIZING ENTEROCOLITIS’: A COMPARISON OF CLINICAL PARAMETERS AND OUTCOME

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Aims of the Study: NEC predominantly occurs in premature neonates. It is a multifactorial disease and immaturity of both bowel and immune system have been suggested as important pathophysiological factors. NEC also occurs in term neonates. In that case risk factors that lead to a compromised splanchnic blood flow are often present, such as congenital heart disease (CHD). Whether NEC and ‘NEC’ occurring in CHD-patients (CHD-NEC) are similar diseases with comparable pathophysiological mechanisms remains unclear. By comparing the clinical course of NEC patients with CHD-NEC we aimed to gain insight in the underlying pathophysiology of various types of NEC.

Method: We identified all CHD patients (i.e. all term patients with any CHD other than isolated patent ductus arteriosus or patent foramen ovale) who developed NEC Bell’s stage ≥2 in our center between 2004 and 2014 and randomly selected (1:2 ratio) patients with NEC Bell's stage ≥2 without a CHD from the same cohort. Location of NEC was retrieved from operation reports and defined as small bowel, ileocecal or colonic. Various laboratory and clinical variables from onset of the disease until recovery or surgery were retrieved from patient files.

Results: There were 19 CHD NEC cases (GA 37⁺³ [31⁺⁵ – 40⁺⁵]) and 38 control NEC cases (GA 28⁺⁴ [22⁺¹ – 38⁺⁶]). Bell’s stage IIIb occurred in 6 CHD-NEC patients (31.6%) and in 15 control patients (39.5%), P=0.32. NEC patients had a lower Hb (8.0 ± 0.8 vs. 8.5 ± 1.0, P=0.02), higher CRP (126 ± 89 vs. 71 ± 57, P=0.03), lower pH (7.21 [7.01 - 7.47] vs. 7.27 [6.68-7.35], P=0.03) and increased duration of NICU stay compared to CHD-NEC (25 days [10-102] vs. 16 [2-43], P=0.005). Mortality as a consequence of NEC was not significantly different between CHD-NEC and NEC (10.5% vs. 21.1%, P=0.47). There was a clear difference in localisation of disease: CHD-NEC occurred statistically significant more often in the colon compared to the control group of NEC patients (71.4% vs. 16.7%, P=0.02).

Conclusion: While overall severity of disease expressed in Bell’s stage and mortality might be similar, the difference in localisation of NEC in CHD patients compared to control NEC patients was striking. This study suggests that both variants of the disease have a different underlying pathophysiological mechanism that predisposes different enterocolic regions to develop NEC.
THE EFFECTS OF WATER AND LIPID SOLUBLE COMPONENTS OF MECONIUM ON THE INTESTINES OF CHICK EMBRYOS WITH GASTROSCHISIS

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Aims of the Study: Previous studies have shown that contact with amniotic fluid (AF) causes serosal damage on the intestines of the fetus with gastroschisis. Experimentally it has been shown that meconium which is physiologically present in the AF is responsible for the intestinal damage on gastroschisis. At concentrations above the threshold level of 1/400 concentration intraamniotic meconium induce damage. Studies investigating which substances within the meconium directly lead to intestinal damage are still being conducted. Therefore, in this study we separated the water and lipid soluble components in order to investigate their respective roles on intestinal damage.

Method: In this study, 5-day-old fertilized chick embryos (Gallus Domesticus) were used. The chicks were divided into the following five groups: control, sham, meconium, water soluble components of meconium and lipid soluble components of meconium. On the fifth day of fertilization gastroschisis was created. Meconium was fractionated into water and lipid soluble components. Meconium, water soluble components of meconium and lipid soluble components of meconium were added into the amniotic fluid as 1/400 end concentrations. The degree of intestinal damage at histopathological investigation was evaluated from the anti-mesenteric sites the intestinal wall. Kruskal-Wallis analysis of variance and the Mann-Whitney U test were used for statistical analysis. P values <0.05 were considered statistically significant.
Results: Intestinal serosal thickness, inflammation, fibrin and collagen formation were observed in all groups. There was no significant difference between the control and sham groups (p>0.05); however when the control and sham groups were compared with the other groups, there was a statistically significant difference (p<0.001). Comparison between the lipid soluble and water soluble groups showed a statistically significant difference (p<0.001). Comparison between the meconium group and all other groups showed a statistically significant difference (p<0.001) (Table 1).

Table: Table 1: The values of the serosal thickness in the intestines of chick embryos with gastroschisis

<table>
<thead>
<tr>
<th>Groups</th>
<th>Serosal thickness (µm) (mean ± 1SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Control</td>
<td>7.47 ± 0.68 *</td>
</tr>
<tr>
<td>Sham</td>
<td>8.48 ± 0.7 †</td>
</tr>
<tr>
<td>Water soluble meconium</td>
<td>14.15 ± 0.9</td>
</tr>
<tr>
<td>Lipid soluble meconium</td>
<td>23.88 ± 1.69 ‡</td>
</tr>
<tr>
<td>Meconium</td>
<td>36.36 ± 2.80 §</td>
</tr>
</tbody>
</table>

* p<0.05 When compared with water soluble meconium, lipid soluble meconium and meconium groups.
† p>0.05 When compared with control group.
‡ p<0.05 When compared with water soluble meconium group.
§ p<0.05 When compared to the others groups.

Conclusion: This study shows that the lipid soluble components of meconium caused significantly more damage than the water soluble components of meconium. However, damage due to unfractionated meconium was much greater than damage due to lipid and water soluble components alone.
Balancing Family Centered Care with Medical Care and Rights of Children

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Aims of the Study: The focus of care in Pediatric Surgery has changed over the last decade resulting in being mainly family centered. Benefits to this approach are manifold, but they come with occasional conflict situations, particularly when the interest of the child differs from the interest of the family. The objective of this study is to elucidate the experience of Canadian Pediatric Surgeons and Physicians with these ethical challenges.

Method: An online survey was generated. Ten questions on moral values and scenarios were presented to the participants. Pediatric Surgeons were invited to participate through the authors’ tertiary health care center and the Canadian Association of Pediatric Surgeons (CAPS) in December 2014, and data was collected over the time period of 4 weeks.

Results: Fifty-four respondents were recorded. Of the cohort, 40.74% were trained surgeons, 46.3% were Pediatricians/Neonatologists and 12.96% were trainees. The majority had a religious affiliation (64.81%), while 31.48% did not and 3.7% preferred to not disclose. While 68% of staff had a religious affiliation, only 42% of trainees indicated such. Religious beliefs do not contribute to how pediatric surgeons and physicians counsel parents in 81.82%, while 15.91% let their religious values affect counseling in some instances and 2.27% frequently. Pediatric surgeons and physicians accept family-centered decisions over patient interests in 90.74% (3.7% always, 25.93% frequently, 61.11% sometimes). When faced with an isolated lethal potentially correctable abnormality in a pre-term neonate (26 weeks of gestation) and the parent’s wish to not pursue treatment, 83.33% agree with the parent’s choice (3.7% always, 5.56% frequently, 74.07% sometimes). However, 66.67% agree with pursuing a court approval for treatment in such a situation (9.26% strongly agree, 57.41% agree, 20.37% disagree, 12.96% strongly disagree). On the other hand, if faced with poor outcome and parents wish to pursue treatment, 3.7% always, 40.74% agree frequently and 48.15% sometimes with pursuing treatment. However, 73.58% think the medical team should counsel parents to not pursue treatment in such a case. In the case of an Ontario court of justice ruling that the mother on an 11-year old aboriginal girl with leukemia had a constitutional right to refuse a 90-95% effective chemotherapy in favor of traditional medicine, 81.48% of participants did not agree, while 18.52% agreed with the decision. This judgment would not change for the majority of participants (92.31%), if the girl was not aboriginal.
Conclusion: Family and patient interests may conflict with the patient’s interests at times causing ethical dilemmas for the medical care team. Family-centered decisions are accepted widely among pediatric surgeons and physicians, action may be necessary to protect the child’s interests. This is illustrated by three scenarios elucidated in the survey: while pediatric surgeons and physicians respect the parent’s wish to not pursue medical treatment for an isolated lethal potentially correctable abnormality in a pre-term neonate, 80% would advocate for the neonate’s treatment in front of a court of law. In a situation with poor prognosis, 93% of physicians would agree with pursuing treatment, but ¾ would counsel the parents against it. In the case of the 11-year old aboriginal girl, the majority of respondents did not agree with the court ruling of withholding highly effective treatment from the child based on the parent’s wish.
COAGULOPATHY AS A CENTRAL PLAYER IN ADVANCED NECROTIZING ENTEROCOLITIS: COAGULATION AND ANTICOAGULATION GENE EXPRESSION PROFILE

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¹Department of Paediatric and Neonatal Surgery, St.George's Healthcare NHS Hospital and University, ²Division of Biomedical Sciences, St.George's University of London, ³Division of Biomedical Sciences, St George's University of London, ⁴Neonatal Intensive Care Unit, St.George's Healthcare NHS Hospital and University, ⁵Neonatal Intensive Care Unit, St.George's Healthcare NHS Hospital and University, ⁶Cardiovascular and Cell Sciences Institute, St George's University of London, London, United Kingdom

Aims of the Study: Coagulopathy and mesenteric thrombosis are common in necrotizing enterocolitis (NEC). This study aimed to investigate differences in coagulation and anticoagulation gene expression between neonates with advanced NEC and controls.

Method: Consecutive neonates with NEC (Bell’s stage 2-3) were prospectively recruited over 15 months. A control group, matched for birth weight (BW) and corrected gestational age (cGA), was selected based on the absence of inflammation or coagulopathy. Each infant had a 0.5ml blood sample taken, and RNA was extracted from pelleted cells for gene expression analysis using the Human Blood Coagulation 96 StellARay qPCR array. Data were analysed using Global Pattern Recognition software (Bar Harbor BioTechnology), which goes through several iterations among the set of genes being analysed, to extract statistically significant changes in gene expression. Platelet, C-reactive protein (CRP), operative findings, and mortality were noted. Continuous data used Student’s t-test and Mann-Whitney U-test as appropriate; nominal data used Fisher’s exact test; statistical significance was P<0.05.

Results: 11 controls and 12 NEC neonates with comparable demographic characteristics were enrolled. NEC had higher CRP and lower platelet than control (p<0.01). 6/12 NEC had laparotomy and 5/6 because of intestinal perforation. Mortality occurred in 4/12 (33%) NEC and 1/11 (9%) control (P=0.32).

Twelve genes were significantly altered (1.4x-15.5x, p<0.05) in NEC compared to control (Table). These genes play main physiologic roles in the coagulation system: extracellular matrix proteolysis (ELANE, CD63, FN1, FGL2), sensors of proteolysis (F2RL1), coagulation (F12), anticoagulation (PROC), fibrinolysis (PLAT) and transcription regulation (HNF4A).
### Table:

<table>
<thead>
<tr>
<th>Gene Name</th>
<th>Description</th>
<th>Fold Change</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>ELANE</td>
<td>Elastase, neutrophil expressed</td>
<td>15.54</td>
<td>0.0013</td>
</tr>
<tr>
<td>CD63</td>
<td>CD 63 molecule</td>
<td>3.26</td>
<td>0.0006</td>
</tr>
<tr>
<td>F12</td>
<td>Coagulation factor XII</td>
<td>2.62</td>
<td>0.0200</td>
</tr>
<tr>
<td>PROC</td>
<td>Protein C</td>
<td>2.62</td>
<td>0.0004</td>
</tr>
<tr>
<td>FN1</td>
<td>Fibronectin 1</td>
<td>-2.42</td>
<td>0.0196</td>
</tr>
<tr>
<td>PLAT</td>
<td>Plasminogen activator, tissue</td>
<td>-2.02</td>
<td>0.0222</td>
</tr>
<tr>
<td>F2RL1</td>
<td>Coagulation factor II (thrombin) receptor-like 1</td>
<td>-1.97</td>
<td>0.0218</td>
</tr>
<tr>
<td>HNF4A</td>
<td>Hepatocyte nuclear factor 4, alpha</td>
<td>-1.80</td>
<td>0.0405</td>
</tr>
<tr>
<td>F2RL2</td>
<td>Coagulation factor II (thrombin) receptor-like 2</td>
<td>-1.79</td>
<td>0.0388</td>
</tr>
</tbody>
</table>

**Caption:** Table: Coagulation and anticoagulation genes significantly up- or down-regulated in advanced NEC

**Conclusion:** A significant group of coagulation/anticoagulation genes get up- or down-regulated during advanced NEC. These results open a new research field to define the interactions between inflammation and coagulopathy leading to mesenteric thrombosis, gut necrosis and perforation.
USE OF ORAL PROPRANOLOL FOR TREATMENT OF CHILDREN WITH INFANTILE HEMANGIOMA ELIMINATED THE NEED FOR LASER TREATMENTS AND CORTISONE THERAPY BUT STILL DID NOT CHANGE THE NEED FOR SURGICAL RESECTION

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¹Department of Pediatric and Adolescent Surgery, Medical University Graz, Graz, Austria

Aims of the Study: Since its introduction in 2008 the use of propranolol has become the first line of treatment for children with infantile hemangioma. The aim of this study was to evaluate the evolution of different treatment options used for children with infantile hemangiomas at our institution in the last ten years.

Method: From 2005 to 2014 we prospectively assessed all children who were referred to us for evaluation and who were treated for infantile hemangioma at our Institution. Data were analyzed by descriptive statistics.

Results: From 1,397 children referred for evaluation during the study period 362 children (26%) received treatment. Over all four different treatment options were administered: 1) treatment with Neodym-YAG laser (31%), 2) surgical excision (33%), 3) treatment with cortisone (5%), and 4) treatment with propranolol (31%). The changes in distribution of different treatment options over years are depicted in the graph. Increased use of propranolol was associated with a very rapid elimination of use of both, the cortisone therapy, and the previously most commonly used laser treatments (see graph). However, the need for surgical excision of hemangiomas remained unchanged in the last ten years. We identified 3 groups of patients receiving surgical intervention: 1) late presenters with large hemangiomas and excision before the 3rd year of life (52%), 2) optimal localization of hemangioma allowing placement of an “invisible” scar – which parents preferred as compared to propranolol treatment (21%), and 3) late presenters with residual scar tissue and excision after the 3rd year of life (27%).
**Conclusion:** Introduction of propranolol for treatment of children with infantile hemangioma completely eliminated the need for cortisone treatment, and laser treatments, therefore substantially reducing the need for general anesthesia. However, there is still an unchanged need for surgical excision which is partially due to late referrals. We strongly believe that in the near future the surgical excision and with it also general anesthesia will become an exception in the management of children with infantile hemangioma.
LAPAROSCOPIC SURGERY IN INFANTS AND CHILDREN USING RADially EXPANDING ACCESS DEVICES: LESSONS LEARNED FROM 9.270 TROCAR SYSTEMS

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¹Paediatric Surgery, Tallaght National Children Hospital, Dublin, Ireland

Aims of the Study: To evaluate the use of radially expanding access devices in paediatric laparoscopic surgery, focusing on the incidence of major and minor trocar-related complications.

Method: A retrospective chart review of all pediatric patients undergoing elective or emergency laparoscopic procedures between 1998-2014 in our institution was performed. Data was collected on patient demographics, type of procedure, immediate postoperative and long-term complications. Statistical analysis was performed using SPSS software.

Results: 3.088 infants and children with a mean age of 6.1 years (range 2 to 16 years) and weight of 23.4 kg (range 15 to 110 kg) underwent laparoscopic procedures requiring a total of 9.270 radially expanding trocar systems. The most commonly performed procedure was appendicectomy (n=2.670) followed by diagnostic laparoscopy (n=116) and orchidopexy for intraabdominal testis (n=72). Other procedures included cholecystectomy (n=51), varicocelectomy (n=49), Nissen fundoplication (n=20), ovarian cystectomy (n=42), salpingo-oophorectomy (n=28), salpingectomy (n=16), resection of Meckel’s diverticulum (n=12), Ladd’s procedure (n=1), pyeloplasty (n=6) and nephroureterectomy (n=5). In 3020 (97.9%) cases, there were no complications using radially expanding access devices. The most common immediate complication was bleeding from the port-site in 17 (0.55%) cases, with only one case (0.03%) requiring hemostasis. Complications requiring surgical revisions occurred in 7 (0.2%) cases, including bowel (n=2) and great vessel (n=1) injuries. There were 30 (1.0%) minor leakages of carbon oxide, 23 (0.74%) preperitoneal placements and no trocar slippage. Late complications such as wound infections were found in 45 (1.5%) cases with 21 arising from port-site granuloma. There was one (0.03%) case of port site herniation that required surgical repair.

Conclusion: Radially expanding access devices are safe and applicable to a wide range of pediatric surgical conditions. The low rate of complications makes them ideal to be used in infants and children.
A PRELIMINARY STUDY OF THE EFFECT OF KAMPO MEDICINE ON THE HUMAN LYMPHANGIOMA DERIVED LYMPHATIC ENDOTHELIAL CELLS

Mototoshi Kato*, Akihiro Fujino¹, Arhans Ismael¹, Toru Morisada², Nobuhiro Takahashi¹, Motohiro Kano¹, Takumi Fujimura¹, Yohei Yamada¹, Ken Hoshino¹, Tatsuo Kuroda¹

¹Department of Pediatric Surgery, ²Department of Obstetrics and Gynecology, Keio University School of Medicine, Shinjuku-ku, Japan

Aims of the Study: Sclerotherapy and surgical resection are the major treatment options for lymphangiomas. However, a part of the patients are intractable, having difficulty in excision and sclerotherapy because of the involvement of vital organs or cavernous tissue type. We need other good therapeutic option for these cases. Recently, Kampo medicine, traditional Japanese herbal medicine, such as Eppikajutsuto (TJ-28) and Ogikenchuto (TJ-98), have been reported to have an effect of reducing volume of the lymphangioma lesion. To elucidate the mechanism and find the possibility for new therapeutic option, We examined the direct effect of the Kampo medicine on lymphatic endothelium derived from lymphangioma (HL-LEC) in vitro.

Method: HL-LEC is obtained from lymphangioma surgical specimen, and immortalized (Im-HL-LEC). Decoction of TJ-28 and TJ-98, dc28 and dc98, respectively, were obtained by boiling at 80-90°C and filtration. Adding each Im-HL-LEC was cultured with 0.25% 1.0% and 4.0% of dc28 and dc98 and examined for cell proliferation, cell migration, tube formation, and permeability. Our in vitro study supported the application of these Kampo medicine for lymphangioma. To elucidate the mechanism of Kampo for lymphangioma, we will study Kampo medicine farther by examining individual crude drug which is a content of these Kampo medicine and the known ingredient. And we will proceed to the in vivo experiments using the lymphangioma model mouse which is available recently.

Results: In co-culture with both dc28 and dc98, the cell proliferation of Im-HL-LEC was reduced according to the concentration of the decoctions(Fig.1). In tube formation assay, the formation of the tubular structure was suppressed with 4.0% of decoctions. In permeability assay, permeability of the Im-HL-LEC sheet was reduced in 0.25 and 1.0%, however, hyperpermeability was observed in 4.0%(Fig.2). These results suggest that the dc28 and dc98 increase endothelial permeability in proper concentration and these decoctions might be toxic in higher concentration. Clinically, these Kampo medicine have been known as an ‘hydrogogue’ agent (cathartic or diuretic). And our in vitro results of the decrease of permeability in Kampo medicine, might be connected to the decrease of uptake of interstitial fluid in the lymphangioma tissue, resulting in the reduction of the lesion volume.
Conclusion: Our *in vitro* study supported the application of these Kampo medicine for lymphangioma. To elucidate the mechanism of Kampo for lymphangioma, we will study Kampo medicine farther by examining individual crude drug which is a content of these Kampo medicine and the known ingredient. And we will proceed to the *in vivo* experiments using the lymphangioma model mouse which is available recently.

Aims of the Study: Specific alterations in the intestinal microbiota are supposedly associated with the development of necrotizing enterocolitis (NEC) in premature infants. We aimed to identify the microbiome of patients at risk for NEC and investigate several negative and positive associations between certain colonization patterns in the microbiome and the development of NEC.

Method: Prospective trial, aiming to investigate prognostic factors for development of NEC in high risk neonates (NTR4153). 11 NEC cases were age/weight matched with controls from the same cohort with a ratio of 1:2. For the present analysis we used the first feces sample of the study (mostly meconium-like feces) of each patient, as well as the last two samples prior to NEC development. Microbial DNA was extracted and 16s rRNA gene sequences were analyzed on a MiSeq sequencer.

Results: As a large majority in both cases and controls (>75%) (frequently) received antibiotics, a diarrhea like microbiota was often found in which Enterobacteriaceae and/or Enterococcaceae were the most dominant groups. The latter two groups were however not associated with NEC, unlike Staphylococci and Clostridium difficile which were surprisingly negatively associated with the occurrence of NEC (p=0.003 and 0.002 respectively). The abundances of Clostridium perfringens group (p=3*10^{-7}) and Bacteroides dorei (p=5*10^{-5}) on the other hand were found to be strong indicators of NEC.

Conclusion: The presence of certain bacteria in the first feces sample of the study, in particular Clostridium perfringens-like bacteria, are associated with the development of NEC later in life, while other bacterial groups possibly provide colonization resistance against NEC-associated bacteria. The bacteria found in these first feces samples of the study suggest a possible maternal role in the bacterial colonization in high risk neonates.
THE IMPORTANCE OF SOCIAL MEDIA FOR PATIENTS AND FAMILIES AFFECTED BY CONGENITAL ANOMALIES: A CROSS-SECTIONAL ANALYSIS AND USER SURVEY OF FACEBOOK

Robyn Jacobs1, Kirsty Brennan1, Leanne Boyd1, Stefano Giuliani1
1Department of Paediatric and Neonatal Surgery, St. George’s Healthcare NHS Trust, London, United Kingdom

Aims of the Study: Facebook is a popular social media site with over one billion users worldwide. We aimed to define characteristics and needs of people using Facebook with regards to rare congenital anomalies.

Method: Cross-sectional analysis of Facebook (FB) Groups and Pages related to four congenital anomalies (anorectal malformations-ARM; congenital diaphragmatic hernia-CDH; congenital heart disease-CHD; hypospadias/epispadias-HS/ES). Data was collected from 23rd March to 6th June 2014. An anonymous survey was posted on open and closed Groups and Pages to obtain quantitative and qualitative data on their contents and patients’ needs.

Results: 54 Groups and 24 Pages were identified (ARM=10 Groups; CDH=9 Groups, 7 Pages; CHD=32 Groups, 17 Pages; HS/ES=3 Groups). Groups had a total of 16,191 active members. 48,766 individuals 'liked' the related Pages. 1103 people participated in the survey. 868/1103 (79%) of respondents were parents. M:F ratio 1:10.9. 65% were 26-40 years old. 46% used Facebook at least once a day. Common reasons for joining Groups were: seeking support; education; making friends; giving support to others. 932/1103 (84%) would like to have healthcare professionals (HCP) join their current Facebook Group or Page. Specialist nurses, Paediatric surgeons, Paediatricians, Cardiologists and Urologists were the professionals that users would most like to interact with out of a pre-defined option list. 97% of respondents stated they would, or maybe would, join a Facebook Group linked to the hospital where they received care.

Table:

<table>
<thead>
<tr>
<th></th>
<th>ARM N=125</th>
<th>CDH N=185</th>
<th>CHD N=699</th>
<th>HS/ES N=94</th>
<th>Total N=1103</th>
</tr>
</thead>
<tbody>
<tr>
<td>Self-reported open:closed ratio N(%)</td>
<td>1:14.6</td>
<td>1:0.87</td>
<td>1:2.71</td>
<td>1:8.4</td>
<td>1:2.61</td>
</tr>
<tr>
<td>Enough support from healthcare system? N(%)</td>
<td>Yes</td>
<td>56(45)</td>
<td>50(27)</td>
<td>182(26)</td>
<td>49(52)</td>
</tr>
<tr>
<td></td>
<td>No</td>
<td>69(55)</td>
<td>135(73)</td>
<td>517(74)</td>
<td>45(48)</td>
</tr>
<tr>
<td>Would responders want HCP in their Group/Page N(%)</td>
<td>Yes</td>
<td>17(14)</td>
<td>27(15)</td>
<td>119(17)</td>
<td>8(9)</td>
</tr>
<tr>
<td></td>
<td>No</td>
<td>108(86)</td>
<td>158(85)</td>
<td>580(83)</td>
<td>86(91)</td>
</tr>
<tr>
<td>Would responders join a Group linked to their hospital? N(%)</td>
<td>Yes</td>
<td>1(1)</td>
<td>8(4)</td>
<td>16(2)</td>
<td>5(5)</td>
</tr>
<tr>
<td></td>
<td>No</td>
<td>23(18)</td>
<td>34(18)</td>
<td>79(11)</td>
<td>18(19)</td>
</tr>
<tr>
<td></td>
<td>Maybe</td>
<td>101(81)</td>
<td>143(77)</td>
<td>604(86)</td>
<td>71(76)</td>
</tr>
</tbody>
</table>

Caption: How responders use Facebook and possible future use
Table:

<table>
<thead>
<tr>
<th></th>
<th>ARM N=125</th>
<th>CDH N=185</th>
<th>CHD N=699</th>
<th>HS/ES N=94</th>
<th>Total N=1103</th>
</tr>
</thead>
<tbody>
<tr>
<td>Responder age</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>N(%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;25 years</td>
<td>5(4)</td>
<td>22(12)</td>
<td>56(8)</td>
<td>6(6)</td>
<td>89(8)</td>
</tr>
<tr>
<td>26-40 years</td>
<td>89(71)</td>
<td>117 (63)</td>
<td>454(65)</td>
<td>62(66)</td>
<td>722(65)</td>
</tr>
<tr>
<td>&gt;41 years</td>
<td>31(25)</td>
<td>46(25)</td>
<td>189(27)</td>
<td>26(28)</td>
<td>292(26)</td>
</tr>
<tr>
<td>Responder Gender</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>N(%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>8(6)</td>
<td>13(7)</td>
<td>56(8)</td>
<td>15(16)</td>
<td>92(8)</td>
</tr>
<tr>
<td>Female</td>
<td>117(94)</td>
<td>172(93)</td>
<td>643(92)</td>
<td>79(84)</td>
<td>1011(92)</td>
</tr>
<tr>
<td>Responder Role</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>N(%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Affected Individual</td>
<td>4(3)</td>
<td>7(4)</td>
<td>161(23)</td>
<td>15(16)</td>
<td>187(17)</td>
</tr>
<tr>
<td>Parent</td>
<td>118(94)</td>
<td>158(85)</td>
<td>514(74)</td>
<td>78(83)</td>
<td>868(79)</td>
</tr>
<tr>
<td>Other</td>
<td>3(3)</td>
<td>20(11)</td>
<td>24(3)</td>
<td>1(1)</td>
<td>48(4)</td>
</tr>
<tr>
<td>Age of affected</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>N(%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>individual 0-3 months</td>
<td>22(18)</td>
<td>80(43)</td>
<td>91(13)</td>
<td>14(15)</td>
<td>207(19)</td>
</tr>
<tr>
<td>4-12 months</td>
<td>14(11)</td>
<td>23(12)</td>
<td>61(9)</td>
<td>14(15)</td>
<td>112(10)</td>
</tr>
<tr>
<td>1-5 years</td>
<td>49(39)</td>
<td>45(24)</td>
<td>229(33)</td>
<td>40(43)</td>
<td>363(33)</td>
</tr>
<tr>
<td>6-18 years</td>
<td>36(29)</td>
<td>28(15)</td>
<td>148(21)</td>
<td>11(12)</td>
<td>223(20)</td>
</tr>
<tr>
<td>&gt;18 years</td>
<td>3(3)</td>
<td>9(5)</td>
<td>170(24)</td>
<td>15(16)</td>
<td>198(18)</td>
</tr>
</tbody>
</table>

Caption: Demographics of responders

Conclusion: Facebook Groups related to rare congenital anomalies are highly populated and active. There is a clear need for healthcare providers to better understand and utilise this huge contemporary social venue and explore new ways to deliver high quality care and patient-centred support.
General
SC-GE-0122

TONGUE TIE DIVISION: IS IT WORTH IT?
Serena Braccio¹, Manasvi Upadhyaya¹
¹Paediatric Surgery, Evelina London Children Hospital, London, United Kingdom

Aims of the Study: Breastfeeding (BF) is a complex process, influenced by different factors, including social and psychological. A well described impediment to breastfeeding is tongue tie (TT). TT division is routinely recommended to improve BF. However recent studies have shown that there is a fall in BF rate and increased bottle feeding few months after frenotomy. The aim of our study is to demonstrate whether frenotomy is beneficial even though breastfeeding after the procedure is only of short duration.

Method: This is a one year (October 2013 – September 2014) retrospective cohort study of all the patients referred to our TT service with breastfeeding difficulties. The majority had been referred after an assessment by a Breastfeeding Midwife. A telephone survey was performed using a standardised questionnaire.

Data was collected and analysed using McNemar’s test.

Results: 308 babies were referred to TT clinic. 297 attended the clinic and 272 had a clinical diagnosis of tongue tie made by a Consultant Paediatric Surgeon. Complete set of data could be collected in only 158/272 parents. The median age at the time of the procedure was 2 weeks.

The rate of exclusively breastfed babies increased from 58/158 (36.7%) before frenotomy to 85/158 (53.8%) 48 hours post-procedure [p<0.0001, OR = 4.857 (95% CI 2.120 – 12.983)]. However, the rate at the time of follow up (median 4 months) was 44.9% (71/158).

All the breastfeeding-related problems reduced by 48 hours post procedure.

Table 1:
The use of formula reduced from 52/158 (32.9%) before procedure to 45/158 (28.5%) 48 hours post-procedure (p<0.2812). However, at the time of follow up (median 4 months) the use of formula milk increased again to 80/158 (50.6%).

There was no major bleeding, infection or ulceration reported. 5/158 babies (3.2%) underwent a second procedure. 153/158 parents felt adequately supported around the consent procedure (96.8%).

142/158 (89.9%) were very satisfied with the delivery of service. The main reason for dissatisfaction with the service was a perceived delay in the referral.
**Table:**

<table>
<thead>
<tr>
<th>Problems</th>
<th>Pre-procedure</th>
<th>48 hours post-procedure</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bleeding nipples and breast pain</td>
<td>118/149 (79.1%)</td>
<td>36/147 (24.5%)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Frequent / long feeds</td>
<td>116/149 (77.9%)</td>
<td>44/147 (29.9%)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Shallow latch</td>
<td>118/149 (79.1%)</td>
<td>34/147 (23%)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Fussiness at the breast</td>
<td>102/149 (68.5%)</td>
<td>36/147 (24.5%)</td>
<td>&lt;0.0001</td>
</tr>
</tbody>
</table>

**Caption:** Table 1

**Conclusion:** Frenotomy is a well-tolerated surgical procedure accompanied by a very low complication rates. It significantly increases the exclusively breastfeeding rate in short-term period and decreases the breastfeeding-related problems. The decreased breastfeeding rate at the time of follow up may be due to other factors that make mothers give up breastfeeding (factors that were not analysed during this study). We have shown that in the short period TT division is beneficial and very well appreciated by mothers.

**References:**
2. Division of Ankyloglossia and its effectiveness in improving associated breastfeeding difficulties. The next step. *British Journal of Oral and Maxillofacial Surgery, October 2014, vol./is. 52/8(e77), 0266-4356 (October 2014)*
OUR EXPERIENCE IN TRANSCROTAL ORCHIDOPEXY IN CHILDREN AFFECTED BY PALPABLE UNDESCENDED TESTIS

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Aims of the Study: Undescended testis (UDT) is one of the most common congenital anomalies. The majority of UDTs are palpable distal to the inguinal canal. Classically, surgical approach for palpable undescended testis (pUDT) consists is an inguinal orchidopexy. In fact, a double incision allows an adequate mobilization of the spermatic cord and an easy dissection of a patent processus vaginalis, often associated to UDT and also to perform a subdartos pouch where to place the testis. For reduce potential morbility of the inguinal approach, in 1989 Bianchi e Squire proposed a transcrotal orchidopexy, using a high scrotal incision. From January 2012, in our Institution we have started to routinely treat the pUDT using transcrotal orchidopexy.

Method: From January 2012 to June 2014, 217 patients (mean age 35±25 months), affected by PUT were treated at our Institution, for a total of 231 orchidopexies (203 monolateral pUDT, 14 bilateral pUDT). Patients, in which, under anaesthesia, testis could be milked until to neck of the scrotum, have been treated with a transcrotal approach using a high scrotal incision. Differently, patients with pUDT where testis remained higher underwent an inguinal orchidopexy. All patients have been clinically followed at one week and at one, two, three and six months. Besides, scrotal sonography with colour-Doppler was carried out at two, three and six months for evaluation of testicular position, volume, texture and vascularization.

Results: 205 pUDT (88.7 %) were considered eligible for transcrotal approach while the remnant 26 pUDT (11.3%) were treated with inguinal one. 8 (3.9%) pUDT, that were firstly approached transcrotally, have been converted to inguinal approach for difficulty in isolation of patent vaginal processus or to further mobilize the spermatic cord. In pts treated with transcrotal orchidopexy, two moderate scrotal haematomas were found at one week, that were conservatively treated and disappeared at one month after surgical procedure. One inguinal hernia was noted after six months and the pt underwent a successful inguinal herniotomy. No recurrence or testicular atrophy was showed in transcrotal approached testes, while 2 recurrences out of 26 procedures (7.7%) were displayed during follow-up after inguinal orchidopexy. Excepted for atrophic and recurrent cases, sonography and colour-doppler follow-up demonstrated all testes presenting normal volume, regular vascularization and adequate location.
Conclusion: It is reported that the transcrotal orchidopexy offers good results, much less dissection of tissue, greater comfort for the patient, rapid healing, excellent cosmesis, avoiding a groin incision. In our experience, transcrotal approach is possible in almost 90% of pUDT. No major complication, such as recurrence or testicular atrophy, has been complained. Just 3 out of 205 cases (1.5%) reported minor complications and one of which required a day-case successful procedure. In few cases, at beginning of our experience, transcrotal orchidopexy could not be completed with single incision for surgical difficult to isolate patent processus vaginalis and to mobilize enough the spermatic cord. In these cases, an additional groin incision was performed to complete the orchidopexy and scrotal incision was used to create a subdartos pouch. Our data confirm that transcrotal orchidopexy can be considered effective, safe and with a success rate being equivalent or better to classical inguinal approach.
EARLY MATERNAL SEPARATION RESULTS IN COLONIC CRYPT DAMAGE AND ACTIVATION OF LGR5+
INTESTINAL STEM CELLS

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Aims of the Study: Maternal separation in the neonatal period can influence the development of intestinal dysfunction, in diseases such as necrotizing enterocolitis. We have previously demonstrated that early maternal separation leads to colonic epithelial damage and increased trans-cellular permeability. The aim of this study was to investigate whether this phenomenon is mediated by the activation of intestinal stem cells.

Method: Three groups of neonatal C57BL/6 mice were studied (animal license no. 32238): A) no separation (NS, n=10); B) single maternal separation (SS, n=12) for 3 hours on postnatal day 9; C) multiple maternal separations (MS, n=10) for 3 hours daily (postnatal day 5-9). Morphology (H&E), goblet cell density (alcian blue), enterocyte proliferation (Ki67) and intestinal stem cell expression (Lgr5 immunofluorescence and western blotting) were analyzed in the proximal colon. Results were normally distributed and expressed as mean ± SEM. Groups were compared using one-way ANOVA with Bonferroni post-test; p<0.05 was considered significant.

Results: Maternal separation induced reduction of crypt length (NS=138±8μm, SS=124±6, MS=110±5; NS vs. MS p<0.01), decrease in goblets cell number per crypt (NS=13±1, SS=8±0.4, MS=6±1; NS vs. SS and MS p<0.01), and increase in Ki67+ cell number (NS=7.5±0.5, SS=10.1±0.5, MS=10.3±0.7; NS vs. SS and MS p<0.001). In addition, Lgr5+ cell expression in the crypts of MS mice was increased in comparison to NS (Figure A – immunofluorescence; Figure B - western blotting, MS vs. NS 3.7 fold increase, p<0.01).
Conclusion: Neonatal maternal separation results in colonic crypt damage and reduction in goblet cells. This phenomenon induces an increase in enterocyte proliferation and activation of Lgr5+ intestinal stem cells in an attempt to repair the damage. These findings are relevant to further understand the mechanism of intestinal damage and repair in diseases such as necrotizing enterocolitis.
ILEAL AND COLONIC PERMEABILITY IS IMPAIRED IN A NEONATAL MOUSE MODEL OF NECROTIZING ENTEROCOLITIS

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Aims of the Study: It is known that intestinal barrier disruption plays a role in the pathophysiology of necrotizing enterocolitis (NEC). Previous experimental studies have demonstrated impaired intestinal permeability in experimental NEC using sugar absorption tests. The Ussing chamber provides a dynamic electrophysiological measurement of transepithelial ion transport and enables analysis of different intestinal segments. Aim of the present study was to investigate tissue specific intestinal permeability using this technique in a neonatal mouse model of NEC.

Method: Following ethical approval (license 32238), NEC was induced in 31 neonatal C57BL/6 mice using gavage feeding with hyperosmolar formula, hypoxia and oral administration of lipopolysaccharide (4 mg/kg/d). Five age-matched breastfed mice were not induced NEC and served as controls. All mice were sacrificed 96 hours later. The small and large bowel was harvested, stained with haematoxylin/eosin, and evaluated microscopically by 3 independent blinded scorers. Ileum and colon permeability was measured ex vivo by Ussing chamber to investigate trans-cellular and para-cellular transport of molecules. Epithelial permeability to large and small molecules was measured through the mucosal-to-serosal passage of horseradish peroxidase (HRP) and fluorescein isothiocyanate-dextran (FD4). Data are presented as median (interquartile range) and were compared using Mann-Whitney test. P< 0.05 was considered significant.

Results: At histology, NEC mice had severe bowel damage in comparison with controls [2 (1-3) vs. 0 (0-1), p=0.001] (Figure A). Trans-epithelial resistance of ileum and colon did not change indicating that the fresh tissues remained intact during the course of the experiment. In comparison to controls, NEC mice had a higher trans-cellular HRP permeability both in the ileum [35.9 (33.8-51.3) vs. 1.3 (0.6-3.6) ng/ml/cm²/min; p=0.0036] and in the colon [34.7 (33.9-41.3) vs. 10.1(9-11.1) ng/ml/cm²/min; p<0.0001] (Figure B). Conversely, NEC mice and controls had similar para-cellular FD4 permeability in the ileum [148.6 (128.3-165.3) vs. 157.8 (125.6-189.9) ug/ml/cm²/min, p=0.6] and in the colon [146.5 (135.8-148.4) vs. 141.1 (125.5-146) ug/ml/cm²/min, p=0.4] (Figure B).
Conclusion: In a neonatal mouse model of NEC, ileal and colonic trans-cellular permeability is increased but not the para-cellular one. The increase in trans-cellular permeability may be caused by the change of the transmural intestinal morphology. The lack of increasing para-cellular permeability in our experiments could be due to the already high level of permeability in normal feeding conditions (control) and the status of tight junctions in the studied groups. Further studies are needed to investigate the state of ion channels in the intestinal epithelium of experimental NEC.
**BIOREACTOR 3D TISSUE CULTURE FOR THE DEVELOPMENT OF AN ARTIFICIAL OESOPHAGUS FOR CONGENITAL ATRESIA.**
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**Aims of the Study:** Oesophagus tissue engineering has been proposed as a therapeutic alternative to oesophageal substitution for congenital defects, such as oesophageal atresia. The use of a tissue engineered oesophageal conduit could dramatically improve the management of this condition. Tissue engineering aims to mimic neo-organogenesis for the production of living tissue to be applied in regenerative medicine strategies. The majority of engineered strategies have been using biodegradable synthetic polymers or biologic components to generate scaffolds for cell delivery or three-dimensional culture. Decellularised matrices are ideal for oesophageal tissue engineering because they maintain all the extracellular matrix (ECM) information. Mesoangioblasts (MABs) were used as they are able to differentiate into both skeletal and smooth muscle. They have been chosen as best candidate to repopulate the oesophagus muscular layer of the artificial construct, combining cells and acellular ECM in a bioreactor for optimized 3D dynamic culture. The aim is to develop a functional oesophageal construct combining human smooth muscle precursor cells with a decellularized matrix. This tissue engineering approach has the purpose of building a differentiated muscularis externa using dynamic 3D tissue culture with a tailored bioreactor to provide optimal nutrient exchange, oxygenation and pulsatile stimulation to the cells.

**Method:** Rat oesophagi were decellularized with an established protocol: 2 cycles of detergent-enzymatic treatment. Human MABs were isolated from paediatric muscle biopsies, expanded and injected into the muscular layer of the acellular scaffold. Seeded matrices were cultured in dynamic conditions using a chamber for luminal and extra-luminal medium flow and scaffold stimulation.

**Results:** Injections of a suspension of human MABs and subsequent culture in the dynamic setting displayed successful and consistent cell engraftment and migration from the injection sites along the scaffold perimeter/thickness after 9 days of culture. Culturing MAB-seeded matrices in dynamic conditions allowed cell survival and migration with no fibrosis developed, as evidenced by histology, and homogeneous distribution of cells in the oesophageal scaffold wall. An optimized combination of proliferating and differentiating medium provided during the culture allowed cell expansion and
the subsequent differentiation towards smooth muscle. In particular, after 9 days of dynamic culture, MABs were presenting both proliferating and differentiating phenotypes, with 30% of the cells expressing Ki67 (proliferation marker) and a total of 45% positive cells for SM22 (smooth muscle marker). Pulsatile culture with mechanic stimulation seemed to improve cell migratory properties with evident amelioration of cell orientation with pre-existing ECM structures.

**Conclusion:** We identified cell injection into the oesophageal muscle layer followed by static culture as a successful seeding method for re-colonization of decellularised oesophageal ECM. Human MABs appeared as an appropriate cell source for engineering an oesophageal matrix thanks to their migration and smooth muscle differentiation capacity. The use of a bioreactor seemed to be a key factor in supporting and improving cell engraftment, proliferation, migration and differentiation, also leading to more homogeneous cell distribution. Future work will include maximization of cell differentiation to obtain a functional fully engrafted scaffold suitable for *in vivo* transplantations.
The extracellular matrix gene FREM1 is deficient in the developing diaphragm in the nitrofen-induced congenital diaphragmatic hernia

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Aims of the Study: The origin of congenital diaphragmatic hernia (CDH) is assumed to lie in a malformation of the amuscular primordial diaphragm. It is known that fetal diaphragmatic development requires the structural integrity of its underlying mesenchymal tissue. Developmental mutations that inhibit the formation of normal diaphragmatic mesenchyme have been shown to cause CDH. FRAS1-related extracellular matrix 1 (FREM1) plays a critical role in the development of the fetal diaphragm. It has recently been demonstrated that a deficiency of FREM1 can lead to CDH both in humans and mice. Furthermore, FREM1-deficient fetuses exhibit a decreased level of mesenchymal cell proliferation in their developing diaphragms. Although the pathogenesis of diaphragmatic defects has been extensively studied, the molecular basis of the abnormal mesenchymal formation in CDH remains unclear. We designed this study to investigate the hypothesis that FREM1 expression is decreased in developing diaphragms of fetal rats in nitrofen-induced CDH.

Method: After obtaining ethical approval (REC668b), timed-pregnant Sprague-Dawley rats were exposed to either nitrofen or vehicle on gestational day 9 (D9), and fetuses were harvested on selected time-points D13, D15 and D18. Dissected diaphragms (n=72) were divided into two groups: control and nitrofen-exposed samples (n=12 per time-point and experimental group, respectively). Diaphragmatic gene expression levels of FREM1 were analyzed by quantitative real-time polymerase chain reaction. Immunofluorescence staining for FREM1 was combined with the mesenchymal marker GATA4 in order to localize FREM1 protein expression and tissue distribution in developing fetal diaphragms.

Results: Relative mRNA expression of FREM1 was significantly reduced in pleuroperitoneal folds of nitrofen-exposed fetuses on D13 (0.30±0.23 vs. 0.83±0.19; p<0.05), developing diaphragms of nitrofen-exposed fetuses on D15 (0.54±0.22 vs. 1.19±0.28; p<0.05) and fully muscularized diaphragms of nitrofen-exposed fetuses on D18 (0.49±0.53 vs. 0.97±0.53; p<0.05) compared to controls. Confocal laser scanning microscopy revealed markedly diminished diaphragmatic FREM1 immunofluorescence, which was associated with reduced proliferation of diaphragmatic mesenchymal cells in nitrofen-exposed fetuses on D13, D15 and D18 compared to controls (Figure).
**Conclusion:** Decreased expression of FREM1 in the nitrofen-induced CDH model may disturb the formation of the diaphragmatic mesenchyme, causing diaphragmatic hernia.
EVALUATION OF MATERNAL ADMINISTRATION OF CANNABIDIOL PROMOTES ANTI-INFLAMMATORY EFFECT UPON THE INTESTINAL WALL IN GASTROCHISIS RAT MODEL.

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Aims of the Study: In gastroschisis, exposure to bowel loops exposure to amniotic fluid and its components results histological and morphological changes leading to dysmotility and impaired absorption of nutrients. These changes require the use of prolonged parenteral nutrition which is associated with higher risk of infections and postoperative complications leading to increased morbidity and mortality. Recently, due to its anti-inflammatory, antioxidant, and a neuroprotective, Cannabidiol (CBD) has been used as a therapeutic agent in many diseases. Our goal was to test the possible effect of maternal CBD on bowel loops in the experimental model of gastroschisis.

Method: The study was approved by the Committee Animal Research Experimentation No 033/2011. There were two groups of pregnant Sprague-Dawley rats treated intraperitoneally (IP) for 3 days after the establishment of gastroschisis model (day 18.5 of gestation): Control (0.9% NaCl – 0.5mL) and CBD (30mg / kg – 0.5mL). A total of 40 fetuses divided into 4 groups (n = 10) were evaluated: 1) Control (C); 2) Gastroschisis (G), gastroschisis fetus underwent surgery at day 18.5 of gestation; 3) Control + CBD (CCBD), 4) Gastroschisis + CBD (GCBD). On 21.5 of gestation (term) the harvest was performed and the following parameters were evaluated: a) morphological analysis: body weight (BW), intestinal weight (IW) and IW/BW ratio; B) histometric analysis of the intestinal wall (n = 4 per group): serosa (S), the longitudinal muscle (LM), circular muscle (CM), submucosal mucous membrane (SM) and the total wall (TL); C) Immunohistochemical analysis of neuronal nitric oxide synthase (nNOS) and inflammatory (iNOS) (n = 4 per group). ANOVA statistical test were used for the morphological and histological parameters and Kruskal Wallis with post Dunns test were used for the immunohistochemistry, considering p<0.05.
Results: G showed higher IW and IW/BW ratio than the C, CCBD and GCBD groups, respectively [(0.190 ± 0.033) versus (0.107 ± 0.008), (0.126 ± 0.021) and (0.124 ± 0.021) (p<0.05)] and [(0.0530 ± 0.010) versus (0.029 ± 0.002) (0.030 ± 0.004) and (0.038 ± 0.008) (p<0.05)]. There was no difference in BW analysis. G showed increased thickness of the layers in each layer and overall than the C, CCBD and GCBD groups: TL (microns), respectively: [(96.533 ± 19.984) versus (27.252 ± 6.024) (27.523 ± 7.114), and (25.260 ± 5.830) (p<0.005)]. G showed higher expression of iNOS than the C, CCBD and GCBD groups, respectively [(2.67 ± 0.49) versus (2.00 ± 0.30), (1.71 ± 0.50) and (1.42 ± 0.42) (p<0.005)]. There were no changes in nNOS expression.

Conclusion: Maternal use of CBD had a beneficial effect on the intestinal loops with decreased intestinal weight of the intestinal layers and inflammation with less iNOS in experimental model of gastroschisis.
EFFECT OF N-ACETYLSEROTONIN ON INTESTINAL RECOVERY FOLLOWING GUT ISCHEMIA-REPERFUSION INJURY IN A RAT

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Aims of the Study: N-Acetylserotonin (NAS) is a naturally occurring chemical intermediate in biosynthesis of melatonin. Extensive studies in various experimental models have established that treatment with NAC significantly protects heart and kidney injured by ischemia-reperfusion (IR). The purpose of the present study was to examine the effect of NAS on intestinal recovery and enterocyte turnover after intestinal IR injury in rats.

Method: Male Sprague-Dawley rats were divided into four experimental groups: 1) sham rats underwent laparotomy, 2) Sham-NAS rats underwent laparotomy and were treated with IP NAS (20mg/kg); 3) IR-rats underwent occlusion of both superior mesenteric artery and portal vein for 30 minutes followed by 48 hours of reperfusion, and 4) IR–NAS rats underwent IR and were treated with IP NAS (20mg/kg) immediately before abdominal closure. Intestinal structural changes, Park’s injury score, enterocyte proliferation and enterocyte apoptosis were determined 48 hours following IR. The expression of Bax, Bcl-2, p-ERK and caspase-3 in the intestinal mucosa was determined using real time PCR, Western blot and immunohistochemistry. A non-parametric Kruskal-Wallis ANOVA test was used for statistical analysis with P less than 0.05 considered statistically significant.

Results: Treatment with NAS resulted in a significant increase in mucosal weight in jejunum and ileum, mucosal protein in jejunum, villus height in ileum and crypt depth in jejunum and ileum compared to IR animals. IR-NAS rats had also a significantly lower intestinal injury score as well as lower apoptotic index in jejunum and ileum which was accompanied by a lower Bax levels compared to IR animals.

Conclusion: Treatment with NAS prevents gut mucosal damage and inhibits programmed cell death following intestinal IR in a rat.
Aims of the Study: To investigate the effect of combined prenatal treatment with retinoic acid and tracheal occlusion on the pulmonary vascular morphology in the nitrofen-induced congenital diaphragmatic hernia (CDH) rat model.

Method: Pregnant Sprague-Dawley rats were exposed to nitrofen at 9 days of gestation followed by either no treatment (CDH), prenatal retinoic acid (CDH+RA), tracheal occlusion (CDH+TO), or both prenatal retinoic acid and tracheal occlusion (CDH+RA+TO) (n=16 animals per group). Among fetuses exposed to nitrofen, only the ones found to have CDH on autopsy were included in the study. We measured the median wall thickness (MWT) of resistance pulmonary arterioles and compared data with ANOVA followed by Tukey's post-test. We analyzed expression of VEGF and VEGF receptors (VEGFR1 and VEGFR2) by immunohistochemistry. Two independent blind investigators analyzed the immunohistochemistry and scored staining from 0 to 4 according to intensity. Results were compared with Kruskal-Wallis test for nonparametric data.

Results: The MWT of pulmonary arterioles was increased in animals with CDH (58±7) compared to controls (44±15; p<0.05). We observed an improvement of the MWT in animals treated with retinoic acid (46±9) or tracheal occlusion (42±11) compared to untreated animals with CDH (p<0.05). However, combination of both retinoic acid and tracheal occlusion did not result in further improvement of the MWT compared to either treatment alone (46±8) (Figure 1). We observed significantly decreased immunostaining for VEGF, VEGFR1 and VEGFR2 on animals with CDH (average score 1.0±0.8, 1.25±0.9, 1.2±0.4, respectively) compared to controls (1.8±0.6, 2.4±0.9, 1.7±0.4, respectively). There was an improvement in the immunostaining of VEGF, VEGFR1 and VEGFR2 in animals with CDH treated with prenatal retinoic (2.1±0.8, 2.4±1.1, 2.2±0.5, respectively) or tracheal occlusion (2.9±0.5, 3.2±0.3, 3.0±0.5, respectively), but there was no additional improvement when treatments were combined (2.9±0.3, 3±0.4, 2.7±0.5, respectively) (Figure 2).
Conclusion: Antenatal treatment with either retinoic acid or tracheal occlusion improved the thickness of pulmonary arterioles in the nitrofen-induced CDH rat model. This was accompanied by a recovery of the expression of VEGF, VEGFR1, and VEGFR2. However, combined treatment did not confer any additional benefit compared to either treatment alone.
DETAILED PHENOTYPIC CLASSIFICATION OF THE VENTRAL WALL DEFECT IN MICE LACKING AORTIC CARBOXYPEPTIDASE-LIKE PROTEIN (ACLP): NEITHER GASTROSCHISIS OR EXOMPHALOS

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Aims of the Study: Aortic carboxypeptidase-like protein (ACLP) is secreted into the extracellular matrix and facilitates cell aggregation/adhesion and cell-cell recognition. Mice lacking ACLP exhibit an abdominal wall defect (AWD) which has been described as gastroschisis¹ ². However, a detailed phenotypic description of the AWD is lacking. We aimed to accurately delineate the anatomy of this AWD.

Method: After ethical approval, homozygous ACLP knockout mice were obtained from heterozygote matings. E18.5 fetuses were collected with intact membranes, dissected and imaged. Gut (pylorus to ileocaecal valve) from 10 normal/mutant fetuses were weighed and length measured. Three normal/mutant E13.5 fetuses (time point of physiological hernia) were collected, paraffin embedded in amnio, serially sagittally sectioned, H&E stained and imaged. 69 fetuses were genotyped. Data mean±SEM.

Results: From the 69 fetuses genotyped 22 were ACLP⁻/⁻ (21 exhibited an isolated AWD, 1 normal abdominal wall closure), 21 ACLP⁺/⁻ and 26 ACLP⁺⁺ (all exhibited normal abdominal wall closure). Through the intact fetal membranes, E18.5 mutant ACLP⁻/⁻ fetuses exhibited externalised free floating gut and liver consistent with AWD. However, microdissection revealed that the amniotic membrane (AM) fails to adhere to the umbilical vessels, and attaches directly to the AWD edge. A relatively large central AWD leaves both gut and liver externalised, but within exocoelomic cavity, separated from the amniotic cavity by the AM. The abnormal umbilical cord enters the abdomen to the left of the externalised gut. The externalised gut shows no evidence of inflammation/peel and no difference in weight (mutant 66.6±4.5mg vs. normal 64.4±4.7 p=0.65) or length (mutant 5.8±0.15cm vs. normal 5.9±0.07 p=0.40) compared to normal mice. In-amnio sagittal sectioning (Figure) of E13.5 fetuses shows in the normal fetus (A) a physiological hernia. The gut herniates into the base of the umbilical cord, which is formed by the adherence of the AM to the umbilical vessels. Sectioning of the mutant fetus (B) shows failure of the AM to adhere to the umbilical vessels and directly attaching to the AWD edge; adjacent sections were consistent with this. Normal and mutant phenotypes are schematically represented in C and D.
Conclusion: The AWD exhibited by the ACLP−/− mouse model does not accurately mimic human gastroschisis or exomphalos. The AWD comprises failure of two components 1) the abdominal wall to close and 2) the AM to adhere to the umbilical vessels, likely due to disrupted cell signalling/adhesion secondary to ACLP absence, placing the externalised abdominal viscera in the exocoelomic cavity. Exocoelomic fluid is an ultrafiltrate of mother’s serum, which explains the lack of bowel wall inflammation/peel seen in this model compared to human gastroschisis. Caution should be exercised in using this model for gastroschisis research. However, it still provides a useful model for manipulation of the fluid environment surrounding the gut and studying the impact on gut development.

ALTERED EXPRESSION OF GAP JUNCTION PROTEIN CONNEXIN 26 IS A POSSIBLE CAUSE OF PERSISTENT BOWEL DYSFUNCTION AFTER PULL-THROUGH OPERATION IN HIRSCHSPRUNG’S DISEASE

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Aims of the Study: Many patients continue to have persistent bowel dysfunction such as enterocolitis, constipation and soiling despite having a correctly performed pull-through operation for Hirschsprung’s disease (HSCR). Direct cell-to-cell signalling is facilitated by gap junction proteins called connexins (Cx). Cx26, Cx36 and Cx43 are known to be expressed in the smooth muscle (SM) layers of colon and play an important role in the propagation of organised electrical activity through the colonic smooth muscle. We hypothesized that the expression of these connexins is altered in the bowel of patients with HSCR.

Method: We collected entire pull-through specimens at the time of surgery from children with HSCR (n=10). Healthy controls were collected at colostomy closure in children with anorectal malformations (n=10). Distribution of Cx26, Cx36 and Cx43 was evaluated using immunofluorescence and visualised with confocal microscopy. Protein expression was quantified by western blot analysis.

Results: Cx26 was co-expressed in PGP 9.5-labelled nerve fibres in the SM and myenteric plexus (MP) (Figure 1). Expression of Cx36 was localised to interstitial cells of Cajal (ICCs) in the SM and MP and to some PDGFRα+ cells in the MP. Cx43 was solely co-expressed in ICCs in the MP and SM layers. The expression of both Cx26 and Cx43 was markedly reduced in the aganglionic bowel but the expression of Cx26 was also reduced in the ganglionic bowel in HSCR compared to controls (Figure 2). The expression of Cx36 was similar across all tissue types.

Image:

![Fig. 1](image1.png)

![Fig. 2](image2.png)
Conclusion: Reduced Cx26 expression in the nerve fibres of the ganglionic bowel in HSCR may indicate abnormal intercellular communication between nerve cells and the SM syncytium and may thus explain the basis of persistent bowel symptoms in some patients after a correctly performed pull-through operation.
ROCK INHIBITOR (Y-27632) IMPAIRS SOMITOGENESIS AND ANGIOGENESIS DURING EARLY CHICK EMBRYOGENESIS

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Aims of the Study: In chick embryos, the administration of Cadmium (Cd) interferes with normal embryonic development inducing ventral body wall defects (VBWD) similar to the human phenotype. However, the precise molecular mechanisms causing VBWD are still unknown. Improper somite formation and impaired angiogenesis have been hypothesized to contribute to the failure of VBW closure. Rho-associated kinase (ROCK) signalling is implicated in essential physiological developmental processes during embryogenesis, including somitogenesis and blood vessel formation. ROCK gene expression levels are significantly decreased following Cd administration in chick embryos. ROCK knockout mice have been reported to exhibit a VBWD phenotype and impaired vascular remodelling. We designed this study to investigate the hypothesis that exposure to ROCK inhibitor (Y-27632) in the absence of Cd affects normal morphological development and extra-embryonic blood vessel formation during very early chick embryogenesis, which may contribute to the development of VBWD.

Method: After 60h incubation, chick embryos were explanted into shell-less culture and treated with 50µL of vehicle for controls (n=54) or 50µL of 500µM of ROCK inhibitor (Y-27632) for the experimental group (Y-27, n=80). At 8h post-treatment, embryos were assessed for gross morphological abnormalities and formation of extra-embryonic vasculature with the dissecting microscope. Western blot was performed to confirm Y-27632 interference in ROCK downstream signalling during early embryogenesis using an antibody against phosphorylated myosin light chain (p-MLC). Gene expression levels of proapoptotic signal protein gene of Bcl2-associated X protein (Bax) were evaluated at 1h, 4h and 8h following treatment.

Results: After 8h post-treatment, all controls and 78 (97.5%) Y-27 embryos were alive. In the experimental group, 54 (69.2%) embryos demonstrated morphological abnormalities in the somites and the neural tube (Fig. 1a, square). At examination, 67 (85.9%) of Y-27632 treated embryos revealed improper formation of omphalomesenteric vessels and vitelline vessels in the tail region (Fig. 1b, squares). Western blot confirmed interference in ROCK downstream signalling by decreased MLC phosphorylation in affected embryos compared with controls (Fig. 2a). Gene expression levels of Bax increased during early stages of embryonic development in Y-27 embryos and were significantly higher at 8h post-treatment in relation to controls (p=0.002), whereas mRNA levels of Bax were not different at 1h and 4h following treatment (p>0.05) (Fig. 2b).
Conclusion: Our results provide evidence that ROCK inhibition impairs crucial developmental processes of somitogenesis and angiogenesis during very early chick embryogenesis, which may contribute to failure of anterior body wall formation causing VBWD.
HEART-REMODELLING IN SEVERE CONGENITAL DIAPHRAGMATIC HERNIA

Alessandro Raffaele\(^1\), Piero Romano\(^1\), Noemi Pasqua\(^1\), Mario Fusillo\(^1\), Luigi Avolio\(^1\), Valeria Calcaterra\(^2\), Gloria Pelizzo\(^1\)

\(^1\)Pediatric Surgery, \(^2\)Pediatrics, Fondazione IRCCS Policlinico San Matteo, Pavia, Italy

Aims of the Study: The aim was the evaluation of cardiac adaptation as a response to the malformation during pregnancy. The immunohistochemical distribution of protein, peptide and polypeptide growth factors involved in cardiac structures may help the knowledge of pathological changes during pregnancy in CDH fetuses. Histological study and spatial immunohistochemical distribution of desmin, muscle actin (HHF35), endothelin-1 (ET-1) and TGF-b in the human heart of cases with CDH were investigated.

Method: We analyzed the human cardiac tissue of seven cases with CDH and severe prognosis (abortion or death in perinatal period within the first 72 hours of life). Right and left cardiac sections were studied by traditional histological evaluation and immunofluorescence assay (HHF35, ET-1, and TGF-b antibodies).

Results: Immunohistochemical distribution
Desmin was intensively expressed in the atria and focally in left ventricles in preterm CDH fetuses. On the contrary, the desmin reaction is negative in the atria and positive in ventricular structures in term fetuses with CDH and in control.
In CDH preterm HHF35 is represented in the atria and in the subendocardial and/or subepicardial. A term of gestation in CDH fetuses and in control, HHF35 reaction in ventricular structures with subendocardial and/or subepicardial distribution was only found.
We showed an atrial TGF-b expression only in preterm CDH newborns; a focal or subendocardial ventricular distribution was expressed in all cases.
ET-1 was focally expressed in the atria and negative in ventricles in preterm CDH fetuses. ET-1 expression in ventricular walls was observed sometimes intensively or focally in term CDH cases and only focally in the control. Negative ET-1 expression in atrial section were detected in term CDH newborns and control.

Histological results
Myocardocytes dimensions in left and right ventricules showed no significative difference. In the left ventricular sections, a slower density of the small intramyocardial arteries was noted in comparison to right sections (6.7±1.7 vs 8.3±2.4). Interventricular septum showed higher density of small vessels (10.5±3.1). Vascular wall thickness were not different between two sections (p=0.2). Network of small vessels was unusual: penetrating vessels in trabeculea, subendocardial and papillary muscles were present, particularly in the right ventricle.

Conclusion: Cardiac impairment in CDH affected fetuses is confirmed. Heart immunohistochemical mapping and histological evaluation in severe CDH showed a dissociative maturation of atrial and ventricular sections and supported
the vascular remodelling in response to malformation. Heart remodelling induced several changes of various cardiac elements in order to optimize cardiac output, but it preserved the mycardiomyocyte integrity to maintain heart performance. The immunohistochemical distribution of elements involved in myocardial growth structures would lead to a better knowledge of the fetal cardiac adaptation during pregnancy. Although the real role of the heart in and CDH prognosis is not yet been defined, identification of the new markers of fetal cardiac impairment in CDH fetuses could be crucial in early detection of fetuses with poor outcome.
NECROTISING ENTEROCOLITIS AND MESENTERIC VESSEL THROMBOSIS: HISTOLOGICAL ANALYSIS IN A QUEST FOR PATHOGENESIS

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Aims of the Study: Mesenteric vessel thrombosis (MVT) is commonly encountered during laparotomy for advanced necrotising enterocolitis (NEC). However it is not clear whether thrombosis is a cause or an effect of NEC. This study aimed to establish the incidence of MVT in surgical NEC and to investigate its possible association with inflammatory/coagulation markers and clinical outcomes.

Method: We retrospectively reviewed all neonates operated for NEC in our Department from January 2007 to December 2012. Exclusion criteria were: spontaneous intestinal perforation (SIP), cases where no specimen was sent, or when histology was not available. Primary end points: association of MVT with inflammatory and coagulation markers (i.e. platelet, INR, aPTT, Fibrinogen, CRP). Secondary end points: association between MVT and clinical outcomes (i.e. Bell's staging, perforation, 30-day mortality and transfusion requirement). Continuous data were shown as median (range). Medians were compared using Mann-Whitney U test, and proportions using x² test or Fisher’s exact test as appropriate, statistical significance was P<0.05.

Results: 147 cases of NEC were treated in the 6 years period. 48 NEC cases fulfilled the inclusion criteria for this study. MVT were evident in bowel resection specimens of 12/48 (25%) patients, 6 in arteries and 6 in veins. The group with and without MVT were comparable in term of Bell's staging - stage II (7/12 v 21/36) and stage III (5/12 v 15/36) (P=0.62). The presence of MVT was not associated with significant preoperative changes in inflammatory or coagulation markers. Perforation, survival and tranfusion of platelet and fresh frozen plasma were not different in the two groups (Table).
### Table:

<table>
<thead>
<tr>
<th>End points</th>
<th>No MVT N=36</th>
<th>MVT N=12</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Platelet</td>
<td>40 (8-423)</td>
<td>101 (13-453)</td>
<td>0.35</td>
</tr>
<tr>
<td>INR</td>
<td>1.45 (0.9-2.5)</td>
<td>1.6 (0.9-2.3)</td>
<td>0.37</td>
</tr>
<tr>
<td>aPTTTr</td>
<td>1.50 (0.65-2.42)</td>
<td>1.39 (0.93-2.59)</td>
<td>0.30</td>
</tr>
<tr>
<td>Fibrinogen</td>
<td>1.60 (0.7-5.4)</td>
<td>1.5 (0.8-2.9)</td>
<td>0.99</td>
</tr>
<tr>
<td>CRP</td>
<td>154 (27-372)</td>
<td>134 (20-299)</td>
<td>0.70</td>
</tr>
<tr>
<td>Perforation</td>
<td>15 (42%)</td>
<td>4 (33%)</td>
<td>0.74</td>
</tr>
<tr>
<td>Preop Platelet transfusion, units</td>
<td>2 (0-11)</td>
<td>2 (0-3)</td>
<td>0.76</td>
</tr>
<tr>
<td>Preop FFP transfusion, units</td>
<td>0 (0-5)</td>
<td>0 (0-3)</td>
<td>0.87</td>
</tr>
<tr>
<td>30-day survival</td>
<td>27 (75%)</td>
<td>10 (83%)</td>
<td>0.71</td>
</tr>
</tbody>
</table>

**Caption:** Association between mesenteric vessel thrombosis (MVT) and inflammatory/coagulation markers and clinical outcomes in neonates with advanced NEC.

**Conclusion:** 1/4 of the neonates operated for acute NEC had histological evidences of MVT. There is uncertainty if the mesenteric vessel thrombosis was a cause of ischemia or its late consequence. More research should be targeted to elucidate the relationship between thrombosis, coagulation and inflammatory changes in advanced NEC.
IDENTIFICATION OF PROTEIN PROGNOSTIC MARKERS FOR PATIENTS WITH HEPATOBLASTOMA

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¹Pediatric Surgery Department, Hospital Universitario La Paz, Madrid; ²Liver Childhood Oncology Group, Fundación Institut de la Salut Sciences Germans Trias i Pujol (GIBT, Badalona); ³CIBERehd, Instituto de Salud Carlos III, ⁴Haematology-Oncology and the Paediatric Bone Marrow Transplantation Unit, Hospital Universitario La Paz, Madrid; ⁵CIC bioGUNE. Proteomics Platform, ProteoRed-ISCIII, Derio, Bizkaia, Spain; ⁶Centre Hepatobiliaire Paul Brousse, Villejuif, France; ⁷CIC bioGUNE. Proteomics Platform, ProteoRed-ISCIII, Derio, Bizkaia, Spain

Aims of the Study: Hepatoblastoma (HB) is the main liver tumor in children. However, it is a rare tumor, with an annual incidence of 1.5 cases per million. In contrast to hepatocellular carcinoma, HB is a tumor that responds to chemotherapy and its cure is achieved by combining this treatment with surgery. However, a quarter of patients do not survive, and those who survive may have serious consequences for life. By a previous transcriptomic study, a prognostic factor of 16 genes discriminated the most aggressive molecular subtype of HB as the C2 and predicted prognosis with high accuracy. Subsequently, a study of the proteomic profile of those subclasses was performed in which a panel of biomarkers associated with tumor subtype was identified.

The aim of this study was to identify a prognostic protein marker, extracted from pre-biomarker panel, which may be easier to apply in clinical practice.

Method: 4 protein expression (BM1, BM2, BM3 and BM4) and Ki67 were studied as markers of tumor proliferation by immunohistochemistry

Results: We analyzed 78 HBs (mean age: 2 years, AFP levels: 150–2,000,000 ng/ml, mean follow: 7 years, 9 deaths).

The BM2 and BM3 biomarkers showed high expression in the most aggressive tumors (C2) while a high positivity BM1 in less aggressive tumors was detected. Furthermore, reduced expression was observed for BM4 tumors in Class C2. Finally, 3 of the 4 markers studied and the proliferative index (Ki67) were significantly associated with HB patients survival (p <0.05.)

Conclusion: This immunohistochemical study has allowed us to identify 4 proteins that act as prognostic markers in HB. This could improve the clinical management, helping patients staging at the time of diagnosis.
GENETICS OF FAMILIAL OVARIAN TERATOMA IN CHILDREN

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¹Pediatric surgery, Necker Hospital, ²Genetic department, Necker Hospital and Imagine Institute, Paris, France

Aims of the Study: There is very little data in the literature about familial forms of ovarian teratoma and their genetic determinants. The aim of this study was to review 12 years of experience in management of ovarian teratoma at a single center and to perform genetic studies in familial cases

Method: We retrospectively reviewed all the ovarian tumors operated on in a single center between 2000 and 2012. We focused on ovarian teratoma and looked for familial forms by systematically reviewing clinical notes and re-contacting patients. Clinical characteristics included: alpha fetoprotein (AFP) results, pathologic type of teratoma, bilateralism, clinical presentation, type of surgery, recurrence, clinical outcome, and description of other cases in the family. Family members were asked for their consent to perform a blood test in order to realize an exome study. Capture of exons from genomic DNA was performed with the 51 Mb SureSelect Human All Exon Kit V5 (Agilent technologies) and sequencing was performed on a HiSeq2500 (Illumina) machine. An in-house software (PolyWeb) developed by the Plateforme Bioinformatique of Université Paris Descartes was used for filtering variants under relevant genetic models in each family

Results: 110 ovarian tumors were operated on between 2000 and 2012. 19 were malignant (germ cell tumors, yolk sac tumors, and immature teratoma), 91 were benign tumors from which 74 were teratomas. We identified 10 cases with a familial history of ovarian tumors: 8 benign and 2 with malignant component and elevated AFP. 8 cases were unilateral and two patients presented a bilateral metachronous tumor, (a mature teratoma on one side, a serous cystadenoma on the contralateral ovary). Familial history consisted in: mother with a unilateral or bilateral synchronous mature teratoma (n=3), maternal or paternal aunt with unilateral mature teratoma (n=2), paternal cousins with mature teratoma or serous cystadenoma (n=2), Prader Willi syndrome (n=1), maternal uncle with testicular seminoma (n=1), maternal grand father with testicular seminoma (n=1). Tumorectomy was performed in 6 cases and adnexectomy in 4 cases. All patients are disease free at a mean follow up of 3.7 years (range 1-8). A genetic study (exome sequencing) was performed in the 8 families with mature teratoma but failed to identify any common variant that could be involved in familial teratoma. However, we found an interesting variant in the gene p53-induced protein with a death domain (PIDD1), known to act in the DNA-damage response, within one family. This avenue is currently being explored
Conclusion: This study suggests that some cases of ovarian tumors may be genetically determined. In addition, it shows that in a same patient or family two types of benign tumor of different origin (epithelial, germ cell) may exist. We failed to find a frequent genetic cause of familial forms because there may be a high genetic heterogeneity underlying ovarian teratoma, that the causal mutations fall outside the coding portion of the genome (not investigated here), or that the penetrance of familial ovarian teratoma is frequently incomplete. Enlargement of this cohort is now mandatory to explore the different genetic mechanisms that could be the basis for the generation of ovarian tumors in children.
SURGICAL TREATMENT OF PEDIATRIC AIRWAY TUMORS: EXPERIENCE OF TWO CENTRES AND PROPOSAL OF AN INTERNATIONAL SURVEY

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¹Pediatric Surgery, Istituto Giannina Gaslini, Genoa, Italy, ²Pediatric Surgery, DINOGMI, University of Genoa; Istituto Giannina Gaslini, Genoa, Italy, ³Pediatric Surgery, University of Chile, Santiago, Chile, ⁴Pediatric Surgery, University Hospital of Padoa, Padoa, ⁵Pediatric Oncology, Istituto Giannina Gaslini, Genoa, ⁶Pediatric Oncology, Fondazione IRCCS Istituto Nazionale Tumori, Milan, ⁷Pediatric Oncology, University Hospital of Padoa, Padoa, Italy

Aims of the Study: Airway tumors in children are very rare. The lack of evidence-based data in the literature determines an absence of guidelines for diagnosis and treatment. The aim of this study is to present a two centers series and propose an European survey as first step to share common guidelines for diagnosis and treatment.

Method: In two reference centres, 11 cases were observed in the last ten years. We considered only pediatric primitive airway tumors, excluding papillomatosis, lung, vascular and lymphatic tumors. Ten patients underwent surgical treatment. A form was created for data collection.

Results: Patients presented with pulmonary atelectasis, pneumonias, or respiratory distress. The diagnosis, based on endoscopy and CT, was delayed in many cases. We observed 6 tracheal and 5 bronchial tumors: mucoepidermoid carcinomas (3 cases), carcinoid (3 cases), inflammatory myofibroblastic tumors (2 cases), rhabdomyoma, lipoblastoma, granulosa cell tumor. Surgical treatment was performed in 10 cases (one was lost to follow-up) and included resection anastomosis for tracheal tumors; sleeve resection or lobectomy for bronchial tumors. Two of them, previously treated endoscopically with laser in other centers, required lobectomy, one for achieving oncological radicality and one for persistent atelectasis. All patients were free of disease and in excellent condition. A form was created, divided into the following sections: diagnosis, surgical treatment, post-operative treatment and follow-up. Data will be analyzed by European Cooperative Study Group for Pediatric Rare Tumors.

Conclusion: Pediatric airway tumors although rare should be considered in the differential diagnosis of symptomatic patients. Usually they are benign or low grade malignant tumors. The treatment is based on surgery, endoscopical treatment is controversial. In our hands, open surgery through different approaches offered excellent results in terms of radical resection and resolution of symptoms. A multicentre collection and analysis of clinical data could help in better understanding the best diagnostic and surgical management.
Aims of the Study: To evaluate defecation- and micturition complaints in adults treated for sacrococcygeal teratoma (SCT) during childhood and to identify risk factors for soiling and urinary incontinence at adult age.

Method: After ethical approval, the records of patients aged ≥18 years treated for SCT during infancy in the Netherlands were retrospectively reviewed for patient- and disorder-related characteristics. The frequency and severity of soiling, constipation, urinary incontinence and voiding complaints were evaluated using two questionnaires designed in accordance with the Krickenbeck classification assessing the outcome of anorectal malformation repair. Urinary and defecation complaints during childhood were compared to outcomes at adult age. Logistic regression analysis was performed to identify patient- and disease related risk factors predicting complaints at adult age.

Results: Forty-seven patients (mean age 26.2 years, range 18.3 – 41.1) were analysed. Twenty-three (49%) reported at least one defecation or micturition complaint at adult age. Urinary incontinence was present in 30% and had a larger impact on patients compared to soiling (24%). Ten patients (21%) reported constipation of whom five experienced it as severely bothering. Social restrictions due to defecation- or micturition complaints were reported in three patients (6.4%). Sex, Altman classification, tumor histology and –volume could not be identified as risk factors for soiling or urinary incontinence during adulthood.
Table:

<table>
<thead>
<tr>
<th></th>
<th>Urinary incontinence</th>
<th>Soiling</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Odds ratio (95% CL)</td>
<td>Odds ratio (95% CL)</td>
</tr>
<tr>
<td>Altman</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type III</td>
<td>0.51 (0.12 to 2.26)</td>
<td>1.00 (0.21 to 4.62)</td>
</tr>
<tr>
<td>or IV</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Histology</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Immature</td>
<td>0.27 (0.05 to 1.49)</td>
<td>0.29 (0.06 to 1.47)</td>
</tr>
<tr>
<td>Malignant</td>
<td>0.89 (0.06 to 12.89)</td>
<td>2.62 (0.30 to 23.00)</td>
</tr>
<tr>
<td>Tumor diameter</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5-10cm</td>
<td>3.00 (0.31 to 28.84)</td>
<td>0.28 (0.02 to 4.24)</td>
</tr>
<tr>
<td>&gt;10cm</td>
<td>2.33 (0.36 to 15.10)</td>
<td>0.50 (0.07 to 3.77)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>5.21 (0.60 to 46.07)</td>
<td>1.81 (0.37 to 8.90)</td>
</tr>
</tbody>
</table>

Caption: Risk factors for urinary incontinence & soiling during adulthood

Conclusion: One-third of the patients treated for SCT during childhood report urinary and defecation complaints at adult age. However, in the minority of patients these complaints lead to social restrictions. Risk factors predicting the development of defecation or micturition complaints through adulthood could not be identified. Prolonged surveillance strategies are therefore advised in all SCT patients.

**Oncology**

SC-ON-0140

**WILMS TUMOUR: HOW OFTEN IS PARTIAL NEPHRECTOMY FEASIBLE?**

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**Aims of the Study:** With the continuous improvement of outcomes after Wilms’ tumour treatment has emerged the need to investigate de-escalation strategies. In the process of writing future protocols guidelines, the SIOP-Renal Tumour Study Group has defined criteria, which would allow to perform partial nephrectomy in unilateral non syndromic Wilms tumours. We aimed to investigate in what proportion of unscreened renal tumours these criteria are present.

**Method:** Retrospective analysis of 91 renal tumours, consecutively and incidentally diagnosed in pediatric patients, in two university pediatric surgical oncology departments from 2005 to 2014. Blind reviews of preoperative abdominal CT-scans were correlated to specimen pathology reports, to determine the relative incidence of the following criteria: tumour volume <300mL, potential for sparing >50% healthy kidney, polar or peripheral tumour, absence of invasion of calyces and vessels, surrounding organs, vessels and hilum.

**Results:** Criteria ruling out feasibility of partial nephrectomy were suspected as follow on preoperative imaging: tumour volume >300mL: 35% of patients; invasion of surrounding organs, hilum, and calyces: 12%, 48%, 23% respectively; vascular thrombus: 12%; no potential for >50% healthy kidney preservation: 90%; non polar or peripheral location: 88%.

An a posteriori comparison with pathology reports revealed a tendency of CT-scan to over-estimate the risk of invasion of adjacent structures (false-positive). Specimens pathology analysis eventually confirmed invasion of surrounding organs, hilum, and calyces in 9%, 40%, and 12% respectively.

Altogether, one or more of the criteria contra-indicating nephron sparing surgery were present in all but 6% of children. The most frequent exclusion criteria were the volume of preservable parenchyma, the tumour volume, and the position of the tumour in relation to the poles or the hilum.

**Conclusion:** Among the population of sporadic Wilms tumour not diagnosed by ultrasound screening, the proportion of tumours potentially amenable to nephron-sparing surgery seems marginal. When considering the undisputed risk of increased local relapse, this finding suggests that these cases should be centrally reviewed and discussed, and the procedures be performed in a limited number of institutions.
ELIGIBILITY CRITERIA FOR MINIMALLY INVASIVE SURGERY IN PEDIATRIC ONCOLOGY ACCORDING TO IMAGE-DEFINED RISK FACTORS

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1Pediatric Surgery, 2Radiology, Buzzi Children's Hospital, Milan, Italy

Aims of the Study: The application of minimally invasive surgery (MIS) in the management of pediatric solid tumors is still debated in its indications and thus reserved to carefully selected patients. The availability of high-resolution imaging techniques and the extension of Image-Defined Risk Factors (IDRFs) to pediatric solid tumors allow to select patients that could be submitted to MIS procedures. Aim of our study is to evaluate the role of MIS in the diagnosis and treatment of pediatric solid tumors, presenting our single-centre experience and analyzing indications, rate and type of MIS procedures performed.

Method: In this retrospective study, records of pediatric patients affected by solid tumors, diagnosed and treated at our centre over the last 6 years, were reviewed. Tumor location, anatomical features, vascular supply and relationship of the tumor with vital structures were defined through angio-CT and angio-MRI, allowing to determine proper IDRFs for each neoplasm. Cases eligible for a MIS procedure and the type of procedure to be scheduled – biopsy or surgical excision – were decided through a multidisciplinary discussion involving surgeon, radiologist, oncologist and pathologist.

Results: Between 2009 and January 2015, a total of 228 patients with solid tumors, aged 3 months – 14 years, were surgically managed. 59 patients (25,4 %) were considered eligible for a MIS approach through a multidisciplinary assessment. 33 of them underwent a MIS diagnostic biopsy (55,9%) through a laparoscopic (21, 63.7%), retroperitoneoscopic (3, 9%), thoracoscopic (6, 18.3%) or endoscopic (3, 9%, endoscopic US-guided fine-needle biopsy) approach. Once established the absence of IDRFs, primary surgical excision was planned in 26 patients (44,1%), through a laparoscopic (22, 84.6%), retroperitoneoscopic (1, 3.8%) or thoracoscopic (3, 11.6%) approach. All the procedures were successfully completed with MIS technique. No postoperative complications occurred. No secondary localizations at trocar sites were observed. MIS biopsies proved to be diagnostic in all cases. Resection was macroscopically complete in all cases. The median postoperative hospital stay was 48 hours for patients undergone a diagnostic biopsy and 5 days for patients submitted to primary surgery.
Conclusion: MIS can be effectively applied for both diagnosis and treatment of paediatric solid tumors, although in selected patients. The process of this accurate selection has to be guided by both IDRFS and multidisciplinary considerations. In our series, only the 25% of patients were considered eligible for MIS procedures; the majority of them had a diagnostic purpose and only the 44% had a therapeutic indication. The limits still applied to MIS surgery allow it to be more safely performed, with low complication and recurrence rate in experienced hands; nevertheless further studies, possibly on larger or multicentre series, could help in expanding the spectrum of indications of MIS in oncologic surgery.
CAN IMAGE DEFINED RISK FACTORS PREDICT SURGICAL COMPLICATIONS IN LOCALIZED NEUROBLASTOMA?
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¹Paediatric Surgery, Osaka City General Hospital, Osaka, ²Radiology, Osaka Medical Center and Research Institute for Maternal and Child Health, Izumi, ³Paediatric Surgery, Osaka University Graduate School of Medicine, Suita, ⁴Paediatric Surgery, Hyogo College of Medicine, Nishinomiya, ⁵Paediatric Surgery, ⁶Hematology/Oncology, Osaka Medical Center and Research Institute for Maternal and Child Health, Izumi, Japan

Aims of the Study: Image-defined risk factors (IDRFs) have been propounded for predicting the surgical risks of localized neuroblastoma (NB) since 2009. In 2011, a new guideline (NG) for assessing IDRFs was published. According to the NG, “even if the tumor is in contact with renal vessels only,” this situation should be considered “IDRF present,” which was previously diagnosed as “IDRF not present.” In this study, we evaluated IDRFs in localized NB patients in order to clarify the predictive capability of IDRFs for surgical complications, as well as the usefulness of the NG.

Method: One hundred and seven localized NB patients diagnosed between 1990 and 2012 were included in this study. All images at diagnoses were evaluated by a single radiologist (MN). Additionally, we analyzed clinical factors, such as stage, primary tumor location, timings of the operation, extent of resection, age, tumor biology and IDRFs (before and after applying the NG), in association with surgical complications.

Results: Of the 107 patients, 33 and 74 patients were diagnosed to have IDRFs (Group OP), and to not have IDRFs (Group ON) before the NG, respectively. According to the NG, there were 76 and 31 patients who were classified as IDRF present (Group NP) and IDRF not present (Group NN), respectively. Thus, 43 (40%) patients in ON group were reassigned to the NP group by the NG. Surgical complications were observed in 17 of 82 patients who underwent surgical resection. Twenty-five patients had not undergone surgical resection because their tumors regressed spontaneously or by chemotherapy. Among 11 patients who underwent primary operations in the OP group, 2 patients (18%) had surgical complications. Conversely, among the 46 patients who underwent primary operations in the NP group, surgical complications were observed in 10 patients (22%). Thirteen patients in the OP group and 16 patients in the NP group underwent delayed primary operations or second-look operations. Of these patients, surgical complications were observed in 6 patients (55%) in the OP group and 7 patients (44%) in the NP group. According to a univariate analysis, non INSS 1, IDRFs before the NG, IDRFs after the NG and delayed or second-look operations were significantly associated with surgical complications. In a multivariate analysis, non INSS 1 and IDRFs after the NG were significantly associated with surgical complications.
Conclusion: The NG increased the ratio of patients with IDRF present from 31% to 71%. Additionally, the NG improved the sensitivity of the IDRF for predicting surgical complications (47%→100%) but reduced the specificity (75%→32%) and the accuracy (71%→46%). Although IDRFs after the NG was significantly associated with surgical complications in the multivariate analysis, there were a considerable number of patients who would have been treated by chemotherapy after applying the NG, but could have been safely cured by surgery alone. In addition, preoperative chemotherapy did not decrease the complications in patients in the OP group or in the NP group. Therefore, the usefulness of the NG should be carefully validated in larger prospective studies.
Aims of the Study: Prospective study of presentation, management and outcome (survival, recurrence and progression) of children with clear cell sarcoma of kidney (CCSK).

Method: All cases of CCSK treated at the center from August 1999 through November 2013 were included. Staging was as per NWTS-5 recommendations and treatment included resection (upfront or after pre-op chemotherapy), chemotherapy (Regime I of NWTSG: vincristine, doxorubicin, cyclophosphamide and etoposide) and radiation therapy. Kaplan Meier analysis was done for 2-year overall survival (OS), Recurrence free survival (RFS) and Event free survival (EFS). Death, recurrence and progression of disease were taken as events.

Results: A total of 18 children of CCSK with age ranging from 3.5-144 months (mean age 41.7 months) were included. Seventeen of these 18 (94%) presented with a large renal mass with hematuria in 2(11%), fever in 2(11%), paraplegia with neurogenic bowel/bladder in 1 (5.5%) and mass over frontal region of scalp in 1(5.5%). Three patients were stage 1(16.6%), 1 stage 2(5.5%), 11 stage 3(61.1%) and 3 were stage 4 tumors(16.6%). Stage 4 children had multiple metastases (long bones 3, vertebral 2, orbit, lungs, liver, pancreas and spinal column 1 each) at presentation. In addition 2 patients also had IVC and intra-atrial thrombus that required resection under cardio-pulmonary bypass. There were 3 deaths (16.6%) and 6(33.3%) relapses. The site of relapses included local 5; bone/bone marrow 4; lungs 3; liver 3 and lymph nodes 1. Two of 6 relapses were after 6 years of follow-up. Though the 2-year OS was 82% (95CI 55-91), the 2-year RFS was only 50.7% (95CI 41-91) and the EFS was only 35.2% (95CI 6.6-67.1).

Conclusion: CCSK usually presented with massive tumors in advanced stages (78% stage 3 and 4) had a poor 2 year EFS of 35.2% (95CI 6.6-67.1).
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**PULMONARY PLEUROBLASTOMA IN CHILDREN: IS A CONSERVATIVE SURGICAL STRATEGY POSSIBLE?**

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**Aims of the Study:** Pulmonary pleuroblastoma (PPB) is a rare tumor that comprises three types of different prognosis: subtype I that shares high similarities with cystic lung malformation, and subtype II and III that are treated as aggressive malignant lesions. Constitutional mutation of DICER 1 gene has recently be identified as a predisposition status for PPB. Conclusions of the French and European pediatric rare tumors registries (FRACTURE and EXPERT groups) suggested that histological subtypes, quality of tumor resection, invasiveness and type of chemotherapy could be prognosis factors. Initial management and therapeutical surgical treatment are however not completely defined due to the rarity of this tumor. The aim of this study was to analyze a homogenous series of patients treated for PPB in order to propose some surgical principles of treatments.

**Method:** From 2000 to 2014, nine patients were treated for PPB. There were 6 boys and 3 girls with a median age at diagnosis of 2.3 years (range 6 months – 14 years) with only one patient over 4 years. Circumstances of diagnosis, tumor extension/effraction, histological type, imaging and treatment were analyzed.

**Results:** PPB classification was 2 subtype I, 6 subtype II and 1 subtype III. Circumstances of diagnosis were suspicion of congenital lung malformation in 3 cases, pneumothorax in 2 cases and respiratory distress with fever and thoracic mass in 4 cases. In 4 cases chest tube was placed at diagnosis in emergency. Percutaneous biopsy was performed for diagnosis in 6 cases but was contributive in only 3 cases. First surgery was performed between 2 and 92 days after diagnosis. It was considered as complete in 4 cases (2 isolated lobectomies and 2 lobectomies associated to local pleurectomy) and surgery was followed by adjuvant chemotherapy (IVADo); and incomplete in 5 cases (1 debulking, 2 wedges resections and 2 lobectomies) who received post-operative chemotherapy. Four of these 5 patients underwent a second look procedure. No radiotherapy was proposed then because of the absence of any viable residue. The fifth, aged 13 years at diagnosis, died after 11 months for uncontrolled tumoral progression despite salvage radiotherapy. Two patients presented recurrence at 12 and 22 months, diagnosed on follow-up imaging, one initially considered as complete resection and the other one as incomplete. None had chest tube insertion at diagnosis. They were both treated by complementary surgery, second line chemotherapy and radiotherapy. All patients but one were alive without disease at a median follow-up of 16 months (6-80.4 months).
Conclusion: Although small, this series allows pointing out some elements. First, rupture of the PPB at diagnosis is a frequent event, either spontaneous or linked to a chest tube placement because of respiratory distress or misdiagnosis. It seems however not to impair the prognosis in this series. Secondly, the performance of a second look procedure when resection is incomplete or when the lesion is initially ruptured is of high interest as it could help to decide for radiotherapy. Finally extent of primary surgery should be discussed regarding the high chemosensitivity of PPB type II-III. Complete pneumonectomy is discouraged at diagnosis and should only be discussed in case of persistent unresectable tumor and/or residual tumor incompletely resectable despite a second look surgery. In this case, multidisciplinar discussion should balance irradiation risks with the ones of a total pleuro-pneumonectomy.
Aims of the Study: In spite of recent progress in multimodal therapies against pediatric solid tumors, the prognosis of INSS 4 neuroblastoma (NB) remains poor. As we consider metastatic NB to be a systemic disease, it was thought to be important to control the metastatic lesions initially rather than the local lesions. We therefore introduced delayed local treatment with surgery and radiation following high dose chemotherapy (HDC) with hematopoietic stem cell transplantation (HSCT) in patients with INSS 4 NB, initially to control metastatic lesions rather than local lesion. It still remains controversial as to whether surgical treatment for advanced NB should be aggressive or limited. The aim of this study was to compare the efficacy, complications and outcomes of the following two delayed local treatments: tumor resection and systemic lymph node dissection vs. tumor resection and lymph node sampling with local irradiation.

Method: We retrospectively analyzed seventeen patients with INSS 4 NB who were treated with delayed local treatment between 1992 and 2012. Eleven patients underwent tumor resection and systemic lymph node dissection (designated as the LNR group), and six patients underwent tumor resection and lymph node sampling with 20Gy of local irradiation after surgery (designated as the RT group). There were no significant differences between the LNR and RT groups in terms of the biological prognostic factors. An unpaired t-test was utilized to perform the statistical analysis.

Results: The length of the operation in the LNR group (653min) was significantly longer than that in the RT group (197min) (p<0.01) and the blood loss in the LNR group (1090ml) was significantly higher than that in the RT group (90ml) (p<0.01). Viable tumor cells were observed in the excised tumors of both groups except for one patient in each group. Postoperative complications were observed in eight patients in the LNR group (73%), but only one patient in the RT group had postoperative complications (17%) (p<0.01). The complications observed in the LNR group included infections in five patients, bowel obstruction in four patients, ischemic liver dysfunction due to hepatic artery in one patient, lymphorrhea in one patient and hydronephrosis in one patient; however, in the RT group, only one patient had an infection. Recurrence was observed in five patients from the LNR group and one patient from the RT group (45.4% vs 16.6%; not significant). Distant metastasis was observed in 4 patients in the LNR group and one patient in the RT group; furthermore, local recurrence was observed in only one patient in the LNR group.
**Conclusion:** Our results indicate that the removal of the primary tumor and lymph node sampling with local irradiation is a safe and effective delayed local treatment for patients with INSS 4 NB in terms of surgical invasiveness, the incidence of postoperative complications and recurrence. The control of distant metastasis may be the most important factor to achieve better outcomes following the treatment of INSS 4 NB.
Aims of the Study: Inflammatory myofibroblastic tumours (IMT) are rare but have a predilection for children and adolescents. Mutations and altered expression of the anaplastic lymphoma kinase (ALK) receptor tyrosine kinase have been described in up to 50% in these tumours, identifying a potential target for novel medical management. Surgery is the mainstay of treatment and recurrences are rare after complete resection. Those not amenable to resection may be treated with a variety of agents including steroids and conventional chemotherapy. We present the outcomes of our single-centre cohort of patients.

Method: All patients in the last 10 years with a diagnosis of IMT were included in the study. The diagnosis was confirmed on histopathology records and ALK expression was tested in all cases. All patients were followed up for any evidence of recurrence.

Results: Four patients had a diagnosis of IMT with a median age at presentation of 9 years (6 months - 10 years). The anatomical locations of these tumours were intra-abdominal (n=1 omental, n=1 distal ileum), lung (n=1) and thigh (n=1). Three patients underwent primary surgical excision. The fourth patient presented at 10 years of age with a central lung lesion that would have required a complete pneumonectomy. In view of this, a biopsy was taken to confirm the diagnosis. ALK expression was negative in all of our cases except this patient who was commenced on a novel mTOR/ALK inhibitor. Cross sectional imaging following treatment demonstrated a dramatic reduction in size. Near-total resolution was seen on further follow-up imaging. No recurrences have occurred in the remaining three patients.

Conclusion: Complete surgical excision remains the mainstay for treatment of this condition. Novel drug therapies are emerging with a potential to manage unresectable lesions.

BLADDER PRESERVING SURGERY AND HIGH DOSE RATE BRACHYTHERAPY FOR PATIENTS SUFFERING FROM BLADDER-PROSTATE RHABDOMYSARCOMA: TECHNICAL ISSUES, RESULTS AND PITFALLS

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Aims of the Study: Although treatment results of patients suffering from bladder-prostate-rhabdomyosarcoma (BPRMS) have been improved in the past decades regarding survival, the bladder preservation rates are still too low. In the past, bladder preserving surgery followed by low dose rate (LDR)-brachytherapy has been advocated as a successful novel treatment approach in France. Nevertheless, LDR-brachytherapy is not available in many centers in Europe. The aim of this study was to establish a new treatment modality combining high dose rate (HDR)-brachytherapy and conservative surgery.

Method: Eight patients (median age: 2 years±0.8) suffering from BPRMS underwent non-mutilating bladder preserving tumor resection (R₀/R₁) with intraoperative placement of 4 to 6 brachytherapy tubes around the urethra. This was followed by CT-planned HDR-brachytherapy (3 Gy / fraction, 2 fractions / d with an interval of 6 hours) for 6 days. The study was conducted according to the IRB guidelines.

Results: In patients with BPRMS, bladder preservation was possible in all patients. Conservative surgery and HDR brachytherapy was well tolerated without acute toxicity. In 2 patients a revision of the brachytherapy tubes was necessary due to an observed inappropriate dose distribution during therapy planning (close position of the tubes to the rectum, rigidity of the tubes). The median follow-up was 11.5 months [4-66]. Local recurrence occurred in one patient at the dorsal wall of the urethra six months after treatment, which was treated with endoscopic tumor resection leading to secondary complete remission. All other patients are in first complete remission. 7/8 patients show a normal voiding pattern and 1/8 patients developed a neurogenic bladder requiring intermittent catheterisation.

Conclusion: Combined bladder preserving surgery and HDR brachytherapy is a novel promising treatment option for patients with BPRMS. Surgery is technically challenging especially in regard to the tumor resection, placement of the tubes and the possible need for revison of the them due to an inappropriate dose distribution during treatment planning.