ABSTRACTS

SCIENTIFIC SESSION I

Saturday 6th October

S-1

IBD Serological Immune Markers ASCA and OmpC are Potential Biomarkers for Hirschsprung-associated Enterocolitis

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3Department of Pediatric Surgery, Astrid Lindgren’s Children’s Hospital, Karolinska University Hospital, Stockholm, Sweden
4Department of Women’s and Children’s Health and Center of Molecular Medicine-CMM, Karolinska Institute, Stockholm, Sweden
5Department of Pediatric Surgery, University of Texas Health Science Center Houston, Houston, TX, USA
6Division of Pediatric Surgery, UCSF Benioff Children’s Hospital Oakland, Oakland, CA, USA
7Division of Pediatric Surgery, C.S. Mott Children’s Hospital, University of Michigan, Ann Arbor, MI, USA
8F. Widjaja Foundation Inflammatory Bowel and Immunobiology Research Institute, Cedars-Sinai Medical Center, Los Angeles, CA, USA
9for the HAEC Collaborative Research Group (HCRG), CA, USA

Objective: The aim of this study was to identify biomarkers to aid in the diagnosis of Hirschsprung-associated enterocolitis (HAEC).

Background: HAEC is the most frequent complication in Hirschsprung (HSCR) patients. Currently HAEC is diagnosed clinically, leaving uncertainty in the diagnosis thereby potentially leading to over- or under-treatment of patients.

Methods: From 2012-2017, 43 children with HSCR enrolled in a multicenter study, underwent retrospective evaluation of their medical records, and questionnaire-directed parent interviews. HAEC status was determined using HAEC score with cutoff ≥4. Plasma was collected and underwent ELISA for the IBD associated antibodies: ASCA, OmpC, CBir, ANCA. Data were analyzed using t-test, univariate, multivariable and binomial regression models.

Results: Eighteen patients had at least one episode of HAEC, 25 had no history of HAEC. The HAEC and NO HAEC groups had similar median ages (3 years), family histories of HSCR. The HAEC group showed markedly elevated ASCA IgA, IgG, and OmpC antibody levels compared with the NO HAEC group while CBir1 and ANCA were similar between the groups. Both univariate and multivariable analysis revealed higher OmpC antibody levels associated with HAEC (OR1.39, CI 1-1.92, p=0.048), while univariate analysis identified a trend toward elevated IgA and IgG ASCA levels with HAEC.

Conclusion: OmpC and ASCA serum antibody levels are associated HAEC, similar to Crohn’s disease suggesting shared gut microbial-host immune responses. Further, these antibodies may serve as potential biomarkers for HAEC diagnosis, although prospective study with larger sample size is needed.

Table 1: Patient Characteristics

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<tr>
<td>Sex</td>
<td>Male</td>
<td>23</td>
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<td>5</td>
<td>28</td>
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<td>Median age (y)</td>
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<td>0.905</td>
</tr>
<tr>
<td>Family history of HSCR</td>
<td>4 (45%)</td>
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<td>1.28 (0.23-6.28)</td>
<td>0.820</td>
</tr>
<tr>
<td>Family history of IBD</td>
<td>8</td>
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<td>1</td>
<td>0.015</td>
</tr>
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<td>Chromosomal Abnormalities</td>
<td>3 (21%)</td>
<td>6 (27%)</td>
<td>2.44 (0.67-12.76)</td>
<td>0.296</td>
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<td>Post-op Complications (&lt;90 days)</td>
<td>2 (11%)</td>
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<td>2.89 (0.76-31.03)</td>
<td>0.096</td>
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<tr>
<td>Length of Anaglognosis</td>
<td>0.000</td>
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<td>Nectrogeniloid</td>
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<td>8</td>
<td>0.13 (0.02-0.81)</td>
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<td>4</td>
<td>21.32 (0.74-61.84)</td>
<td>0.074</td>
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S-2

Hirschsprung-associated Enterocolitis Shares Genetic Pathways with Inflammatory Bowel Disease

Philip K Frykman1,2, Talin Hartunians3, Emebet Mengesha2, Zhenqiu Liu2, Tomas Wester4,5, Agneta Nordenskjöld1,4, Akemi Kawaguchi5, Anna L Granström1,4, Daniel H Teitelbaum6, Dermot P McGovern7

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Purpose: Recent reports have identified Hirschsprung’s disease (HD) patients with chronic Hirschsprung-associated enterocolitis (HAEC) who had clinical and pathological features consistent with IBD, suggesting the possibility of biological similarities between HAEC and IBD. Immune-driven diseases have shared genetic signatures with IBD. The purpose of this study was to identify immune-related genetic associations and pathways in HAEC patients compared with HD patients free from enterocolitis.

Methods: An IRB-approved international multicenter study enrolled 116 consecutive HD patients, median age 6, IQR (3-8) all of whom were scored using the Pastor et al. criteria HAEC score (0-20). To avoid population stratification, only European ancestry subjects were included in the analyses. HAEC score was analyzed as a quantitative trait using linear regression with permutation, adjusting for population sub-structure with 2 principal components. DNA was isolated from blood and run on the immune-focused Illumina Immunochip-v2 array, comprising 253,702 SNPs. Following stringent QC measures, 199,110 SNPs with MAF>3% were available for analyses. Enrichment analysis tool in STRING was used to identify KEGG pathways associated with HAEC.

Results: Eighty-five unrelated HD patients were analyzed and we identified 179 SNPs with (mapped to 47 known genes) nominally significant evidence of association (P<0.001) with HAEC. Genetic pathway analysis identified 13 pathways with false discovery rate <0.05 (Table, Figure); all of which are also associated with IBD. Interestingly no associations were found between HAEC and SNPs located within or near known Hirschsprung-associated genes.

Conclusion: HAEC shares genetic pathways with IBD, strongly suggesting overlapping biological mechanisms.

<table>
<thead>
<tr>
<th>Pathway ID</th>
<th>Pathway Description</th>
<th>Observed Gene Count</th>
<th>False Discovery Rate</th>
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<tr>
<td>4514</td>
<td>MHC Class II Region</td>
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<td>5321</td>
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<tr>
<td>5330</td>
<td>Allergen rejection</td>
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<td>5332</td>
<td>Viral myocardiitis</td>
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<td>5120</td>
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<td>4184</td>
<td>Phagosome</td>
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<tr>
<td>4016</td>
<td>Antigen processing and presentation</td>
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<td>0.00031</td>
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</table>

Table: Kyoto Encyclopedia of Genes and Genomes (KEGG) pathways in HAEC shared with IBD (9 of 13 shown due to space limitations) identified.

S-3 Comparison of Outcomes of Patients with Hirschsprung’s Disease who Underwent Duhamel and Transanal Endorectal Pull-through Procedures

Rob J Meinds1, Cornelius E J Sloots3, L W Ernst van Heurn7, Marieke J Witvliet7, Wim G van Gemert1, Ivo de Blaauw5, Monika Trzpis7, Paul M A Broens1,7

1Department of Surgery, Division of Pediatric Surgery, University of Groningen, University Medical Center Groningen, Groningen, Netherlands, 2Department of Pediatric Surgery, Emma Children’s Hospital, AMC and VU University Medical Center, Amsterdam, Netherlands, 3Department of Pediatric Surgery, Erasmus MC Sophia Children’s Hospital, Rotterdam, Netherlands, 4Department of Pediatric Surgery, Wilhelmina Children’s Hospital, University Medical Center Utrecht, Utrecht, Netherlands, 5Department of Surgery-Division of Pediatric Surgery, Radboudumc- Amalia Children’s Hospital, Nijmegen, Netherlands, 6Department of Pediatric Surgery, University Medical Center Maastricht, University of Maastricht, Maastricht, Netherlands, 7Department of Surgery, Anorectal Physiology Laboratory, University of Groningen, University Medical Center Groningen, Groningen, Netherlands

Aim of the Study: The majority of Dutch Hirschsprung’s disease (HD) patients are operated by either Duhamel or transanal endorectal pull-through (TERPT) procedure. We aimed to compare the long-term functional outcomes of patients who underwent both procedures.

Methods: From a nationwide cross-sectional study we included 52 patients who underwent TERPT (mean age 12±2 years) and 52 patients who underwent Duhamel procedure. We matched them, and also healthy controls, according to age and gender. We assessed functional outcomes using the Constipation Scoring System (CSS) and Continence Grading Scale (CGS). We additionally assessed the outcomes according to the type of surgical approach (laparotomy, laparoscopy or transanal procedure).

Main Results: The median CSS was comparable patients operated with Duhamel and TERPT procedures (5 versus 4), but significantly higher than in controls (P<.001). The CGS was comparable in both patient groups (4 versus 5), but significantly higher than in controls (P=.003, P<.001). Laxative usage was more frequent in patients who underwent Duhamel than TERPT (48% versus 12%, P>.001). The prevalence of soiling (P=.007) and laxative usage (P=.017) was significantly lower among patients who underwent laparoscopic Duhamel procedure compared to open. Patients who underwent transanal TERPT were significantly more often urge incontinent for feces (P<.001) and urine (P=.008) than patients operated laparoscopically.

Conclusions: The differences in functional outcomes following Duhamel and TERPT procedures are limited. Duhamel procedure outcomes can be further improved by opting for laparoscopy instead of an open approach, whereas TERPT outcomes can be improved when performed laparoscopically, instead of completely transanal.
Long-term Functional Outcomes and Quality of Life in Patients with Hirschsprung’s Disease: A Nationwide Survey
Rob J Meinds1, Alida F W van der Steeg2,3, Cornelius E J Sloots4, Marieke J Witvliet5, Ivo de Blaauw6, Wim G van Gemert7, Monika Trzpis8, Paul M A Broens1,8

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Aim of the Study: It is unclear whether functional outcomes improve or deteriorate with age following surgery for Hirschsprung’s disease (HD). Our aim was to determine the long-term functional outcomes and quality of life (QoL) in patients with HD.

Methods: All patients with pathologically proven HD older than eight years were included (N=619). Patients with a permanent stoma or intellectual disability were excluded. Functional outcomes were assessed by Rome IV criteria using the Defecation and Fecal Continence (DeFeC) questionnaire. QoL was assessed by the CHQ-CF87 questionnaire or by the WHOQOL-100 questionnaire. Reference data of healthy controls were available for matching and comparison.

Main Results: A total of 55.9% (n=346) patients responded (median age 18 years). The prevalence of constipation was comparable in pediatric and adult patients (22.0% versus 22.0%) and in patients and controls. Compared to controls, adult patients suffered significantly more often from straining (50.3% versus 36.1%, P=.011) and incomplete evacuation (47.4% versus 27.2%, P<.001). The prevalence of fecal incontinence, most commonly soiling, was lower in adult patients compared to pediatric patients (16.8% versus 37.6%, P<.001), but remained higher than in controls (16.8% versus 6.1%, P=.003). Patients with poor functional outcomes scored significantly lower in several QoL domains.

Conclusions: Our results show that functional outcomes are better in adult patients compared to pediatric patients, but symptoms of constipation and soiling persist in a substantial group of adult HD patients. The persistence of defecation problems are an indication that continuous care may be recommended in a specific group of patients.

Outcome of Hirschsprung’s Disease beyond Adolescence
Johan Mikael Danielson1,2, Elisabet Gustafson1

1Department of Pediatric Surgery, Akademiska Sjukhuset, Uppsala, Sweden, 2Institution of Women and Children’s Health, Uppsala University, Uppsala, Sweden

Aim of the Study: Hirschsprung’s Disease (HD) has often been reported to have better outcome beyond childhood than ARM. The aim of this study was to assess the outcome of HD beyond adolescence.

Methods: All patients operated and/or diagnosed for HD at our department from 1961 to 1995 were identified and their charts were scrutinized to make sure of the diagnosis. 123 patients could be traced and were sent a validated bowel function and SF-36 quality of life questionnaires. 69 patients (mean age 37.8, range 22-58, 13 female) responded and were matched with 138 (2 per patient) age and sex-matched controls who had responded to the same questionnaires in a previous study.

Main Results:

Bowel function: There was no significant difference in Miller Incontinence score. HD-patients had significantly higher incidence of soiling, urgency, permanent stomas, use of laxatives, enemas and loperamide. HD patients had a significantly higher number of bowel moments per week. HD patients scored significantly lower in their satisfaction with their bowel function. Effects on daily life and quality of life: HD patients reported a significantly higher incidence of negative impact by their bowel function on daily life, social interaction, ability to go on vacation. There were no significant differences in SF-36-scores.

Conclusions: Bowel function has a lifelong negative impact on the life of patients with HD. This strongly indicates a need for structured follow-up beyond adolescence.
Resection Margin Histology May Predict Intermediate-term Outcomes in Children with Hirschsprung’s Disease
Laura Veras1, Lauren Brooks1, Kathryn Fowler1, Ming Fu1, Ankush Gosain1

1Division of Pediatric Surgery, Department of Surgery, University of Tennessee Health Science Center, Memphis, TN, USA

Background: Recent studies suggest that some of the post-surgical morbidity in Hirschsprung’s disease (HD) is due to enteric nervous system structural defects in the proximal, ganglionated bowel that remains after surgery. We hypothesized that resection margin histology would predict intermediate-term outcomes in HD patients.

Methods: Following IRB approval, HD patients born between 2009-2016 were reviewed(n=68). N=50 had tissue available for new analyses. Proximal resection margins were analyzed for ganglion size, Hu+ neurons/ganglion, and % nitric oxide synthase (NOS) neurons/ganglion as compared to 7 control (non-HD) patient samples. Chart reviews were performed for 1- and 2-year outcomes. 10 patients were contactable for Stenstrom quality of life survey; 5 had tissue available for analysis.

Results: 44/50 patients had recto-sigmoid disease and were further analyzed. Ganglion size did not differ from HD to controls (4815 um², IQR 3268-7309 vs. 5593, IQR 3776-9670, p=NS). Hu+ neurons/ganglion did not differ from HD to controls (15.6, IQR 13.3-21.3 vs. 16, IQR 11.4-21.2, p=NS). HD patients had higher % NOS neurons (50, IQR 43-61 vs. 30, IQR 23-33, p=0.0006). None of the histology measures correlated with presence/absence of constipation at 1-2 years follow-up. However, increasing % NOS neurons correlated with decreasing patient-reported quality of life (R²=-0.9487).

Conclusions: 1-2 years follow-up may be insufficient to determine if resection margin histology correlates with outcomes. Patient-reported quality of life surveys, although limited in number, suggest that neurotransmitter imbalance may predict poor-outcomes. This study supports the concept that the ganglionated portion of the remaining colon post-surgery may not sustain normal bowel function.

A Novel Diagnosis Support System for Hirschsprung’s Disease Using Deep Learning
Kosuke Chiba1, Hirohisa Oda2, Kensaku Mori1, Yuiiro Tanaka1, Takahisa Tainaka1, Chiyoue Shiroti1, Wataru Sumida1, Kazuki Yokota2, Kazuo Oshima3, Satoshi Makita1, Tomoko Tanaka3, Yukiko Tani1, Hiroshi Kishimoto4, Hiroo Uchida5

1Department of Pediatric Surgery, Nagoya University Graduate School of Medicine, Aichi, Japan, 2Graduate School of Information Science, Nagoya University, Aichi, Japan, 3Graduate School of Informatics, Nagoya University, Aichi, Japan, 4Department of Pathology, Saitama Children’s Medical Center, Saitama, Japan

[BACKGROUND] Hirschsprung’s disease (HD) and its variants are diagnosed by a skilled physician by investigating ganglion cells and examining their maturity level. However, the requisite facilities and pathologists required for HD diagnosis are limited. In this study, we aim to assess a diagnosis support system using Artificial Intelligence (AI) that would permit non-specialists to accurately diagnose HD and its variants.

[METHODS] We evaluated images of HE-stained preparations of the Auerbach plexus of the normal intestinal tract and the tract without ganglion cells of pathological specimens of 24 children who underwent radical surgery for HD. Using these images and data manually labeled with the ganglion cells region, we applied “U-net,” a deep learning method, to automatically detect ganglion cells from microscopic images, and we evaluated the accuracy of the identification.

[RESULTS] In the learning and testing phase using 190 ganglion cell images of the normal intestinal tract of 24 children, the detection rate of ganglion cells was 85.3% and the number of false positives per photograph was 2.0. The number of false positives observed in the intestinal tract without ganglion cells was 0.39 per photograph.

[CONCLUSIONS] We found that the HD diagnosis support system using AI is fully functional. Because “U-net” can extract the region of cells by estimating the size and shape of the cell, the maturity level (cell or nucleus size) of the intestinal ganglion cell can be estimated. We aim to conduct further analyses of automatic diagnosis of HD and its variants.

Figure: Before and after labeling of intestinal ganglion cells
**S-8**

Transplanted Sox10-Venus Mouse Enteric Neural Crest-derived Cells Colonise the Aganglionic Colon in the Hirschsprung’s Mouse Model

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¹Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, Tokyo, Japan, ²Department of Pediatric Surgery, Juntendo University Nerima Hospital, Tokyo, Japan

**Aim:** Hirschsprung’s disease (HSCR) is caused by the failure of enteric neural crest-derived cells (ENCC) to properly colonise the enteric nervous system (ENS) in the embryonic gut. Cell-based therapies have potential in the treatment of enteric neuropathies by generating enteric neurons in regions that lack an ENS due to developmental defects such as HSCR. The endothelin-B receptor (Ednrb) null mouse model has been widely used as a model for HSCR. Previously, we developed a Sox10 transgenic version of the Ednrb mouse to visualize ENCC with green fluorescent protein, known as the Venus mouse. The aim of the present study was to evaluate cellular migration into the aganglionic gut using Sox10-Venus mice.

**Methods:** Fetal guts from Sox10-Venus mice were dissected on embryonic day 15.5 (E15.5) and cells were dissociated. These cells were cultured for 21 days under non-adherent conditions to generate neurospheres which were transplanted into the aganglionic region from E15.5 Ednrb⁻/⁻ mouse hindgut explants on a semi-permeable membrane. After 4days, the recipient gut was fixed and examined histochemically using confocal microscopy.

**Results:** By 5days, cells had proliferated to form neurosphere-like aggregates that continued to increase in size up to 14 days (Figure 1). Four days after neurosphere transplantation, Sox10-Venus⁺ cells extended from the neurosphere into the aganglionic gut wall (Figure 2 arrow).

**Conclusion:** These findings suggest that ENCC from the gut of Sox10-Venus mice can migrate into explants of recipient aganglionic bowel of Ednrb⁻/⁻ mice. Further study is needed to determine whether they generate functioning neuronal networks in the aganglionic gut.

![Figure 1](image1.png)
![Figure 2](image2.png)

**S-9**

Altered Expression of Acetylated Tubulin in Enteric Neurons in Hirschsprung’s Disease Mouse Model

Nana Nakazawa-Tanaka¹, Katsumi Miyahara², Naho Fujiwara², Masahiko Urao¹, Atsuyuki Yamataka²

¹Department of Pediatric Surgery, Juntendo University Nerima Hospital, Tokyo, Japan, ²Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, Tokyo, Japan

**Aim:** Tubulin acetylation, which regulates microtubule stabilization, plays an important role in neuronal migration and differentiation. However, little is known about the role of acetylated tubulin (Ac-tub) in early development of enteric nervous system (ENS). The aim of this study was to investigate the expression pattern of Ac-tub in enteric neurons in the embryo of HD mouse model.

**Methods:** SOX10-Venus transgenic mice, in which enteric neural crest derived cells (ENCCs) are labeled with a green fluorescent protein, Venus, was crossed with mice lacking the endothelin-B receptor gene, Ednrb⁻/⁻ mice to produce the newly created HD mouse model (KO). Sox10-VENUS+/Ednrb⁻/+ mice was used as control (WT). The fetal guts were dissected on embryonic day 13.5 (E13.5) from KO (n=4) and WT (n=3) and on E16.5 from KO (n=5) and WT (n=3). The expressions of Tuj1, which is a marker for neurons, and Ac-tub were examined by fluorescent immunostaining.

**Results:** Sox10 positive cells contained both Tuj1 positive (+) and negative cells, which proportion was similar in both WT and KO. The expression of Ac-Tub in Tuj1 (+) cells was increased in KO from E13.5 to E16.5, whereas that was not significantly changed in WT during the same period. Furthermore, the expression rate of Ac-tub was significantly higher in
KO compared to WT on 16.5

**Conclusion:** Our results suggest that the altered acetylation of tubulin in enteric neurons may be associated with the failure of ENS development in HD.

### S-10

**Downregulation of Sodium-calcium Exchanger 2 (NCX2) in Hirschsprung’s Disease Colon**

Anne Marie O’Donnell1, Hiroki Nakamura1, Christian Tomuschat1, Naoum Fares Marayati1, Prem Puri1

1National Children’s Research Centre, Our Lady’s Children’s Hospital, Dublin, Ireland

**Aim of the Study:** The sodium-calcium exchanger (NCX) protein is a plasma membrane transporter which plays a key role in the regulation of intracellular Ca²⁺ concentrations in the brain, kidney, and smooth muscle. Nishiyama et al. have previously shown that NCX1/NCX2 double-heterozygote knockout mice displaying decreased acetylcholine release and altered colonic motility. We designed this study to investigate the expression of NCX1 and NCX2 in the normal human colon and in Hirschsprung’s disease (HSCR).

**Methods:** HSCR tissue specimens (n=6) were collected at the time of pull-through surgery, control samples were obtained at the time of colostomy closure in patients with imperforate anus (n=6). qRT-PCR analysis was undertaken to quantify NCX1 and NCX2 gene expression, and immunolabelling of NCX1 and NCX2 proteins was visualized using confocal microscopy.

**Main Results:** qRT-PCR analysis revealed significant downregulation of the NCX2 gene in both aganglionic and ganglionic HSCR specimens compared to controls (p<0.05) (Figure). NCX1 gene expression levels showed no significant difference between groups. Confocal microscopy revealed NCX1 and NCX2 expression within the myenteric plexus and smooth muscle, with a reduction in NCX2 expression in both aganglionic and ganglionic HSCR colon compared to controls.

**Conclusions:** NCX2 gene expression is significantly downregulated in HSCR colon, suggesting a role for this gene in colonic cholinergic neurotransmission. The marked reduction in NCX2 gene expression within ganglionic specimens suggests abnormal cholinergic neurotransmission in this region, which may result in persistent post-operative symptoms in some patients following a properly performed pull through operation.

### S-11

**Altered Expression of KCNG3 and KCNG4 in Hirschsprung’s Disease**

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**Aim of the Study:** Voltage-gated potassium ion channels have long been implicated in gastrointestinal motility. Recent studies by Lee et al. and Ha et al. have highlighted the role of voltage-gated channel subfamily G members 3 (KCNG3) and 4 (KCNG4) genes in the electrical functioning of interstitial cells of Cajal and PDGFRα⁺ cells of the mouse colon. We designed this study to investigate KCNG3 and KCNG4 expression in the normal human colon and in Hirschsprung’s disease (HSCR).

**Methods:** HSCR tissue specimens (n=6) were collected at the time of pull-through surgery, while control samples were obtained at the time of colostomy closure in patients with imperforate anus (n=6). qRT-PCR analysis was undertaken to quantify KCNG3 and KCNG4 gene expression, and immunolabelling of KCNG3 and KCNG4 proteins was visualized using confocal microscopy.
Main Results: qRT-PCR analysis revealed significant downregulation of the KCNG3 and KCNG4 genes in both aganglionic and ganglionic HSCR specimens compared to controls (p<0.05) (Figure). Confocal microscopy revealed KCNG3 and KCNG4 expression within the myenteric plexus and smooth muscle layers, with a reduction in both proteins in aganglionic and ganglionic HSCR colon compared to controls.

Conclusions: KCNG3 and KCNG4 gene expression is significantly downregulated in HSCR colon, suggesting a role for these genes in colonic motility. KCNG3 and KCNG4 downregulation within ganglionic specimens highlights the physiologically abnormal nature of this segment in HSCR patients.

S-12
Innervation of the Entire Internal Anal Sphincter in a Mouse Model of Hirschsprung’s Disease. A First Report
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Background: Impaired function of the internal anal sphincter (IAS) may be implicated in postoperative obstructed defecation (POD) that may complicate Hirschsprung’s disease (HD) patients. While innervation of part of the IAS in HD has been reported, accurate details based on anatomic landmarks that can explain the clinical morbidity seen in POD are lacking, and there appear to be no studies that specifically document the innervation of the “entire” IAS in HD. We used endothelin receptor-B knockout mice to represent HD (HD-mice) and C57B6 mice as controls (C-mice) to investigate the innervation of the entire IAS in order to assess the pathophysiology of POD.

Methods: The end point of the longitudinal muscle layer was used to define the border between the IAS and the circular muscle layer (CML). Specimens of anorectum from HD- and C-mice were immunostained with PGP 9.5 and S100 as general nerve markers, nNOS and VIP as parasympathetic nerve markers, TH as a sympathetic nerve marker, and calretinin as a reliable diagnostic marker for HD. Immunostained cells/fibers were quantified using ImageJ.

Results: On fluorescence microscopy, PGP 9.5, nNOS, and calretinin were significantly lower in the IAS of HD-mice than in C-mice (p<.05, respectively, Figure 1), while there were no significant differences between HD-mice and C-mice for S100, VIP, or TH (Figure 2).

Conclusion: We are the first to confirm that the expression of histochemical markers of innervation is abnormal throughout the “entire” IAS in HD-mice. Application of this finding may be beneficial for preventing POD and requires further research.
Enterocolitis Complicating Hirschsprung’s Disease: The Roll of (C-159t Codon) Polymorphism Biomarkers in Clinical Follow Up
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Aim of the study: Enterocolitis (EC) is the most common and serious complication of Hirschsprung’s disease (HD). The prognosis and outcome of EC depend on early diagnosis and effective management. The present study was established to evaluate the role of (C-159t) gene polymorphism biomarkers in patients developed such complication.

Methods: We conducted a prospective study for all the patients with Hirschsprung’s disease complicating EC admitted and managed in our pediatric surgery unit, from the period 1st of January 2014 to the end of December 2016. All patients developing EC were evaluated with sepsis screen tests, blood culture, subjected to broad-range PCR and PCR-restriction fragment length polymorphism (RFLP). The ethics committees of the health authorities approved the study.

Results: The distribution of (C-159t codon) polymorphism was detected by PCR-RFLP technique. At this locus there are three genotypes; homozygote (TT) at 353bp, heterozygous (CT) at 497bp, 353bp and 144bp, and wild-type (CC) which still undigested. TT genotype was significantly increased by 13.1 times for those patients with EC associated sepsis. Having a CC genotype significantly decreases the risk of EC associated sepsis by 5.9 times. Presence of T allele significantly increased the risk of having sepsis by 5.9 times in the patients with EC.

Conclusion: There is a significantly higher prevalence of detection of (C-159t) gene polymorphism among the Hirschsprung’s patients associated EC. This gene may play a major role in the pathogenesis and EC development. More studies incorporating this gene are warranted.

Is Spinal Ultrasound a Good Screening Method to Look for Tethered Cord in Patients with Anorectal Malformation?
Andrea Bischoff, Alberto Peña, Jill Ketzer, Kristen Campbell, Brent O’Neill, Nicholas Stence, David Mirsky

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Introduction: Tethered cord occurs in approximately 25% of patients with anorectal malformation. The diagnostic value of spinal ultrasound (US), during the first three months of life, as a screening tool for tethered cord is still controversial.

Methods: A search of the electronic medical records of a single institution for patients with anorectal malformation was performed from January 2003 until January 2017, identifying 914 patients. Only patients who were screened for tethered cord by US, MRI or both were included. Data collection comprised: gender, age, type of anorectal malformation, sacral ratio, presence or absence of tethered cord, and surgery for tethered cord release.

Results: 197 patients met the inclusion criteria. 104 (52.8%) were males. 122 patients had MRI only, 41 had US only, and 34 had both US and MRI. Imaging revealed a normal conus position without filar abnormality in 114 patients (57.9%). Forty-three patients (21.8%) had normal conus position with fatty filum, of which 25 of those (58.1%) underwent tethered cord release. Forty patients (20.3%) had a low-lying conus, 29 of those (72.5%) underwent tethered cord release. The sensitivity for detecting tethered cord by US was 0.73 (95% CI:0.39, 0.94) and the specificity was 0.91 (95% CI:0.72, 0.99).

Conclusion: In this series, MRI was used more frequently than US to screen for tethered cord. The sensitivity found for US indicates that it is an effective screening method for low lying conus medullaris (tethered cord).

Table 1: Results for patients who had both US and MRI

<table>
<thead>
<tr>
<th>Ultrasound = Tethered Cord</th>
<th>MRI = Tethered Cord</th>
<th>MRI = Normal</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>8</td>
<td>3</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>3</td>
<td>21</td>
<td>24</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>23</td>
<td>34</td>
<td></td>
</tr>
</tbody>
</table>

Tethered Cord in Patients with Anorectal Malformations with Emphasis on Rectobladder Neck Fistula
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Introduction: The association between anorectal malformations and spinal cord tethering (TC) is well recognized. Untethering of the cord is generally indicated in patients with progressive/new onset of symptoms attributed to TC. Yet, there is controversy regarding the management of asymptomatic patients with TC.

Aim: To find the incidence of TC in ARM patients and to determine the relationship between bowel/urinary control and TC
Methods: The database of a tertiary medical center was retrospectively reviewed for all patients treated for ARM from 1980-2012. All patients with TC and RBNF were identified. Data were collected by chart review.

Results: Among 790 patients, who underwent screening for TC, 285 (36%), were radiologically diagnosed with TC. Eleven of 37 screened patients with RBNF were diagnosed with TC. The median follow-up period was 49 months (range 2-222 months). TC was diagnosed in 3/18 (16.6%) patients with sacral ratio (SR) ≥0.7; 4/12 (33.3%) with SR 0.41-0.69; and 4/7 (57.1%) patients with SR 0-0.4. The association of TC in RBNF patients further reduced prognosis for bowel and urinary control.

Conclusion: The incidence of TC among patients with ARM is 36%. Incidence of TC among patients with RBNF correlates with SR value and is higher the lower the SR. Patients with RBNF and TC have dismal prognosis for bowel control, unrelated to their SR status. Many unresolved questions related to the management of ARM patients with asymptomatic TC, still remain.

S-16
The Conus Medullaris Ratio: A New Way to Identify Tethered Cord on MRI
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Purpose: Approximately 25% of patients with anorectal malformation have tethered cord. The traditional way of determining conus medullaris level on MRI relies on either using lumbar spine images and “counting up” from L5 or acquiring a whole-spine sequence and “counting down” from C1. Either way, the task may be challenging in patients with vertebral numeric variation or transitional vertebrae. Our purpose was to use reliable anatomic landmarks, to differentiate patients with normal from those with low lying conus.

Methods: A single institution database search identified 2 groups of patients: 255 with normal and 85 with abnormal spinal MRI, the latter group underwent tethered cord release. The conus ratio was calculated in both groups. The ratio was obtained by dividing the distance between the conus level and the iliac crest by the distance between the foramen magnum and the conus level (figure). IRB approval was obtained (# 16-2330).

Results: The mean ratio was 0.1843 (sd 0.033) for the non-tethered group and 0.1179 (sd 0.093) for the tethered one, with a p-value <0.0001. By ROC curve analysis, the ratio proved to be a good discriminator between normal and abnormal patients, with Area Under the Curve (AUC) equal to 0.749, meaning that at random there is a 75% chance that the tethered cord patient will have a lower ratio than the non-tethered cord patient.

Conclusion: "The Conus Medullaris Ratio" is a good predictor of low lying conus level on MRI and offers an easy alternative to avoid the need for vertebral counting.

S-17
Sacral Agenesis and Fecal Incontinence - How to Increase the Index of Suspicion
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Introduction: Sacral anomalies represent a spectrum of congenital abnormalities ranging from coccyeal aplasia or hypoplasia, to hemi-sacrum, to complete absence of the sacrum with fused iliac bones. The condition is frequently
classified as caudal regression, though this diagnosis does not take into consideration the severity of the defect nor its long-term motor and functional prognosis.

Methods: A retrospective review was performed of patients who presented for bowel management due to sacral agenesis at a children’s hospital between 2012-2017. Data collection included: gender, time of diagnosis, sacral ratio, and associated anomalies. Patients with anorectal malformation and sacrococcygeal teratoma were excluded from this review.

Results: Ten patients were identified (4 female, 6 male). Seven of those had a delayed diagnosis ranging from 22 months to 9 years of age. Most common symptoms included severe diaper rash, as well as failure to toilet train for urine and stool at the appropriate age. All patients older than 3 years of age required daily enemas and intermittent catheterizations. The sacral ratio was zero (6), 0.3 and 0.4 (2), and hemi-sacrum (2). Associated anomalies were present in seven patients.

Conclusion: Sacral anomalies should be suspected in patients who present with early severe diaper rash and in patients who fail to successfully toilet train for urine and stool at the appropriate age. An abdominal radiograph can provide a sacral ratio which, when 0.4 or less, should trigger counseling for parents regarding the potential for fecal incontinence and neurogenic bladder. Patients with fused iliac bones are typically non-ambulatory.

Impact of the Associated Anorectal Malformation on the Outcome of Spinal Dysraphism after Unthethering Surgery

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Aim of the study: To analyze the outcome after unthethering surgery in patients with spinal dysraphism (SD), with or without associated anorectal malformation (ARM).

Methods: Patients operated on for SD, with (Group A) or without (Group B) associated ARM (1999-2015), were included. The post-operative outcome was analyzed in the two groups in terms of improving of clinical symptoms (i.e. neuromotor deficits, abnormal reflex, bladder dysfunction, bowel dysfunction). Groups A and B were also compared for improvement of at least one of the following instrumental analyses: urodynamics, bladder ultrasound, neurophysiology. Fisher’s exact test and X2 test were used as appropriate; p<0.05 was considered statistically significant.

Main results: Ten patients in Group A and 24 in Group B were consecutively treated. One patient was lost at follow up. Six patients (25%) in Group B underwent prophylactic surgery. Table reports main results.

Conclusions: The presence of the associated ARM does not seem to impact the medium-term outcome of patients operated for SD. On the basis of our results, unthethering seems to be effective in selected patients with ARM.
S-19

Functional Anal Sphincter Defects after Repair of Anorectal Malformations Are Correlated to Bowel Outcome
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Aim of the study Our objective was to compare anorectal function with the results of 3D-High Resolution Anorectal Manometry (HRAM).

Methods We used a Manoscan™ anorectal High Resolution Manometry system (Medtronic, MN, USA) applied a 3D probe. Data were compared with the Mann U Whitney test. The study was approved by the National Committee in Health Research Ethics and the Danish Data Protection Agency.

Main Results Twenty one patients were enrolled. Median age was 22 (12-31) years and 14 (67%) were females. Types of anorectal malformations were anocutaneous fistula (n=8), bulbar fistula (n=4), vaginal fistula (n=4), vestibular fistula (n=2), anal stenosis (n=1), no fistula (n=1) and cloaca (n=1). Primary surgical approach was posterior sagittal anorectoplasty (n=11), perineal anorectoplasty (n=5), dilatation only (n=3), abdominoperineal pull-through (n=1) and cutback (n=1). From the visual judgement of the resting anal pressure profile, ten patients (48%) had sphincter defect defined as areas with resting pressure below 25 mmHg. Comparison of subjects with anal sphincter defect and subject without anal sphincter defect are presented in table 1. A higher Wexner score indicating worse bowel outcome was found in participants with a functional sphincter defect.

Conclusions We found sphincter defects detected by HRAM to be common after surgical repair of anorectal malformations and related to Wexner incontinence score but not to the resting anal or squeeze pressure. HRAM may in some cases replace EUS and aid on the choice of optimal treatment.

Table 1 - Comparison of subject with anal sphincter defect and subject without anal sphincter defect detected by High Resolution Anorectal Manometry assembly applied a 3D probe.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Sphincter defect</th>
<th>No sphincter defect</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age(years)</td>
<td>22(12-31)</td>
<td>23(17-31)</td>
<td>NS</td>
</tr>
<tr>
<td>Gender(% female)</td>
<td>66</td>
<td>72</td>
<td>NS</td>
</tr>
<tr>
<td>Resting anal pressure(mmHg)</td>
<td>32</td>
<td>37(23-70)</td>
<td>NS</td>
</tr>
<tr>
<td>Ano sphincter pressure(mmHg)</td>
<td>92</td>
<td>178(56-227)</td>
<td>NS</td>
</tr>
<tr>
<td>Rectal pressure gradient(mmHg)</td>
<td>-16</td>
<td>13(17-41)</td>
<td>NS</td>
</tr>
<tr>
<td>Wexner score</td>
<td>38±14</td>
<td>30±7</td>
<td>0.02</td>
</tr>
</tbody>
</table>

Data are presented as median (interquartile range) if not otherwise indicated. HPZ: High Pressure Zone. NS: not statistical significant.

S-20

Health-related Quality of Life in Infants with Anorectal Malformations
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Objective: To evaluate the health-related quality of life (HRQoL) and analyze the influencing factors in infants ages 1-24 months with anorectal malformations (ARM).

Methods: Generic HRQoL of infants younger than 2 years old with ARM were investigated using PedsQL Infant Scale and PedsQL Family Information Form between Sep 2014 and Aug 2017. Age-matched controls were enrolled from healthy infants. Multiple step-wise regression analysis was used to determine factors that influenced HRQoL in infants with ARM.

Results: One hundred and twenty-eight participants with 68 ARM and 60 controls were included. There were no significant differences in total scale score between ARM group (89.15±4.60) and control group (86.08±13.52). The emotional functioning score of ARM group was significantly higher than the control group (89.15±4.60 vs 81.53±18.10, P <0.05). The type of ARM was associated with the physical health in infants with ARM independently.

Conclusion: The HRQoL is comparable between infants with ARM and healthy infants. Individualized care plan based on the type of ARM should be applied to improve the future HRQoL.
Early Vaginal Replacement in Cloacal Malformation
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PURPOSE: We aimed to study the surgical outcome of Cloacal malformation (CM) with emphasis on need for vaginal replacement.

METHODS: An ambispective study of CM patients was carried out including prospective cases from April 2014 to December 2017 and retrospective cases that came for follow up during that period.

RESULTS: 18 patients with CM were studied with age at presentation from 1 day- 4 years. The associated anomalies included solitary kidneys-2, renal dysplasia-1, Vesico-ureteric reflux (VUR)-9, partial sacral agenesis-2, spina bifida-3, pouch colon-3, clitoromegaly-1, septate vagina-4 and bicornuate uterus in 5. Ten patients had common channel length of >3 cm. All patients underwent PSARPUP with an additional laparotomy in 7. Six patients required vaginal replacement; 2 with ileum; 2 with sigmoid colon; vaginal switch in 1 and hemirectum was used in one. Reduction clitoroplasty was done in 1. 4 developed anal mucosal prolapse that required trimming. Urethro vaginal fistula developed in 1 patient that closed spontaneously after regular vaginal and urethral calibration. 5 patients had incontinence for urine and 2 had fecal soiling. Persistent VUR was found in 8, 5 received deflux; 1 underwent a ureteric reimplantation and 2 remained on chemoprophylaxis. 1 patient of uterus didelphys is having menorrhagia. One patient died due to sepsis at the age of 7 months. Renal functions remained normal in 16. One patient is undergoing dialysis.

CONCLUSION: Early vaginal replacement in CM is feasible. Patients with inadequate introitus may suffer from menorrhagia. A regular follow-up is mandatory.

Menstrual Obstruction in Patients with Anorectal and Urogenital Malformations: Considerations for Management and Surveillance
Veronica Isabel Alaniz1, Duncan Wilcox3, Alberto Peña2, Andrea Bischoff2

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Aim of Study: To review cases of menstrual obstruction in female patients with history of anorectal and urogenital malformations.

Methods: A retrospective review was performed after institutional IRB approval. Relevant ICD-9 codes were used to identify patients with history of anorectal or urogenital malformation and menstrual obstruction from 2008 - 2018.

Main Results: Eight patients were identified which included diagnoses of cloaca (3), cloacal exstrophy (1), posterior cloaca
(1), recto-vestibular fistula (1), perineal fistula (1), and urogenital sinus (1). The five patients with cloacal anomalies developed hematometrocolpos from acquired vaginal stenosis. Of these patients, thee had vaginal replacements and four had uterine didelphys with one hemi-hysterectomy performed at initial reconstruction. The other three patients with recto-vestibular fistula, perineal fistula and UG sinus developed hematometrocolpos from an obstructing vaginal septum. Two of these patients had uterine didelphys with an obstructed hemi-vagina (OHVIRA) and one had a transverse vaginal septum. Pre-operative management included menstrual suppression (6/8), drainage by interventional radiology (2/8), and vaginostomy (1/8). Definitive surgical management included vaginoplasty (3), septum resection (1), abdominal hysterectomy (1), vaginoplasty with buccal graft (1), and vaginoplasty with hemi-hysterectomy (1). One patient has not yet undergone definitive surgical management.

**Conclusions:** Patients with anorectal and urogenital anomalies are at risk for menstrual obstruction from congenital and acquired causes. Menstrual suppression and drainage can be used prior to definitive repair. More importantly, periodic assessment of reproductive structures is recommended to capture patients at risk.

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**S-23**

**Anorectal Malformations and Perineal Hemangiomas: The Arm-NET Consortium Experience**

Inbal Samuk¹, Carlos Gine², Ivo de Blaauw³, Anna Morandi⁴, Pernilla Stenstrom⁵, Stefano Giuliani⁶, Gabriele Lisi⁷, Paola Midrio⁸

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**Aim** Perineal hemangiomas rarely occur in patients with anorectal malformations (ARM), but they can pose a significant challenge and warrant special attention. Surgical incision of posterior sagittal anorectoplasty (PSARP) may involve the hemangioma site resulting in damage to blood supply and muscle complex, leading to complications and adversely affect outcome. The aim of this study was to review the experience of the ARM-Net Consortium in the management of perineal hemangioma associated with ARM and evaluate treatment strategies.

**Materials and Methods** Data on all patients with ARM and a perineal hemangioma located in planes of the PSARP dissection were collected retrospectively by a questionnaire sent to all members of the ARM-Net Consortium.

**Results** Ten patients from eight centers were included. Three patients each had a rectobulbar or rectovestibular fistula, 2 had a rectoperineal fistula, and one had a rectoprostatic fistula; in one patient, the hemangioma was too disfiguring to determine malformation type. Mean follow-up time was 36.6 months. Colostomies were performed before definitive repair in 8 patients. Five patients received systemic beta-blockers before PSARP: 3 were operated uneventfully following partial/complete involution of the hemangioma, and 2 are awaiting surgery. Two patients with rectoperineal fistula were managed expectantly. The remaining 3 patients underwent surgery without preoperative medical treatment, and all had complications: mislocated neoanus in three and complete perineal dehiscence in one.

**Conclusion** Attempting PSARP in the presence of a perineal hemangioma may lead to complications and adversely affect outcome. This study confirms the benefits of beta blocker treatment before surgical reconstruction.

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**Soon after birth**

**Age 4 months**

*Following beta-blockers and prior to PSARP*
Effects of Trapidil on Microvascular Cerebral Thrombosis in an Experimenta Model of Ulcerative Colitis in Mice
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Background: Ulcerative colitis is a significant risk factor for premature atherosclerosis in children and adults including an increased risk of stroke. It has been shown that platelet serve as both effector and target cells in a variety of chronic inflammatory diseases. CD40 and CD40L positive platelets contribute to thrombogenic responses in the vasculature and are elevated in patients after transient ischemic attack and stroke. Trapidil (a 5-methyl-7-diethylamino-s-triazoloprimidin) decreases platelet aggregation/activation by blocking the CD40/CD40L pathway and has been shown to reduce significantly cardiovascular events.

Aim of the study was to investigate whether Trapidil prolongs intracerebral thrombosis time in an experimental mouse model of ulcerative colitis.

Method: Colitis was induced in male wild type mice (25 - 35 g; n = 6) with 3% dextran sodium sulfate (DSS) in drinking water. Control mice received regular water. Disease activity index was recorded daily, platelet and WBC counts were performed on day 6. Thrombus formation in cerebral arteries was observed intravitally through a skull window with a fluorescent microscope using the light dye method. Time of onset and completion of thrombosis was measured. CD40 antibodies were injected prior the experiment and Trapidil (40mg/kg) was injected twice on days 1 and 3 intraperitoneally.

Results: Trapidil significantly reduced platelet and neutrophil counts. CD40 antibodies and Trapidil significantly prolonged thrombosis time similarly to untreated animals.

Conclusion: Trapidil may be used clinically reducing the risk of stroke in patients with inflammatory bowel diseases, too.

Novel Therapy for Congenital Isolated Generalized Hypoganglionosis Using Mesenchymal Stem Cell
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Aim of the study: Congenital isolated generalized hypoganglionosis (CIGH) is a representative disease of the allied disorders of Hirschsprung’s disease, in which the number of ganglion cells are markedly decreased. Currently, the patients are treated with partial bowel resection and high jejunostomy for intestinal decompression and parenteral nutrition. Unfortunately, the survival rate is only 78%, and the patients are hard to withdraw intravenous nutrition for long time, resulting in the low quality of life. Small bowel transplantation is the only curative treatment for the patients; however, it is hard to perform because of varied problems such as severe rejections and infections. Therefore the alternative therapeutics has been desired. Recently, stem cell-based regenerative therapy has been focused on intestinal intractable diseases.

Method: We used stem cells from human exfoliated deciduous teeth (SHED), which is a kind of mesenchymal stem cell. We transplanted SHED to CIGH model mouse intravenously. We performed clinical, histopathological, electrophysiological, molecular biological and nutritional analyses as well as safety evaluation regarding organ morphology, function and immunity following the categorization into three groups as wild type mouse, CIGH model mouse and CIGH model mouse with intravenously SHED transplantation.

Main Results: We initially confirmed the phenotype and morphological findings of CIGH model mouse. The number of myenteric ganglion cells significantly increased after SHED transplantation. Furthermore, we obtained the significant weight gain, and improved the electrophysiological and nutritional parameters.

Conclusion: SHED transplantation may improve quality of life of the patients with intestinal motility disorders.

Decreased Viability/Motility of Spermatozoa after Contact with Colonic Mucosa in Mice May Be Implicated in Infertility of Cloacal Malformation Patients after Colon Vaginoplasty
Seitaro Kosaka1, Masahiro Takeda2, Katsumi Miyahara1, Eri Nakamura2, Norihiro Tada1, Geoffrey Lane1, Atsuyuki Yamataka1

1Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, Tokyo, Japan, 2Research Institute for Diseases of Old Age, Juntendo University Graduate School of Medicine, Tokyo, Japan

BACKGROUND: Colon vaginoplasty is often performed for cloacal malformation. Although the fertility status of these cases is unclear, none of our cases have become pregnant to the best of our knowledge. To better understand possible infertility in colon vaginoplasty cases, we assessed the viability/motility of spermatozoa placed in contact with colonic mucosa in mice.

METHODS: Spermatozoa were isolated from the epididymides of 10-week-old C57BL/6J male mice (n=24), checked for viability and motility, then introduced directly into the vaginas (Vag-group) or per anum into the colons (Colo-group) of 9-week-old female mice (n=4). Deposited spermatozoa were collected at 0, 2, 5, 10, 30, and 60 minutes after introduction, stained with SYBR-14 and Propidium Iodide (PI), and examined under fluorescence stereomicroscopy. Motility was analyzed using the Computer Assisted Spermatozoa Analysis system: IVOSIII.
RESULTS: The mean proportion of viable spermatozoa in epididymal samples was 62±3.46% initially. While viability dropped over time in both Vag and Colo, viability in Colo was significantly lower at each assessment time except at 10 minutes (Figure 1). Final viability in Colo at 60 minutes was only 10.8%. Mean motility, initially 39.4±1.7%, also dropped over time in both Vag and Colo, but the drop was significantly greater and more rapid in Colo, plummeting to only 0.8% at 60 minutes compared with a more gradual decrease in Vag where 10.0% were still viable at 10 minutes, and 6.0% at 60 minutes (Figure 2).

CONCLUSIONS: Decreased viability/motility of spermatozoa when exposed to colonic mucosa may be implicated in infertility in cloacal malformation cases after colon vaginoplasty.

Abnormal Serum Vitamin A Levels and Retinoic Acid Receptor α Expression Patterns in Children With Anorectal Malformation
Yi Wang

Purpose To investigate serum VA levels in ARM newborns and RA receptor (RAR) expression in the rectum of ARM patients and animal models.

Methods Serum VA concentrations were detected in newly diagnosed ARM neonates (n=32) and neonates with non-alimentary tract malformations (n=30). Intestinal specimens were divided into 3 groups: rectum from ARM patients (n=30), colon from a stoma (n=30) and rectum from controls (n=4). RAR mRNA expression was evaluated by RT-qPCR. Rectum specimens from ARM patients were divided into 2 groups by postoperative pathology: the normal and lesion ganglion cell groups. Immunohistochemistry and Western blot were employed to detect RARα protein expression. In addition, the ARM mouse model was induced by ATRA, and the expression levels of RARα and the neuronal marker NeuN in the rectum of mice on embryonic day 16.5-18.5 (E16.5-18.5) were investigated.

Results The serum concentration of VA in ARM neonates was lower than that in control neonates (P<0.0001). RARα mRNA expression was lower in the rectum specimens from ARM patients than in the colon specimens from a stoma and the rectum specimens from controls (P<0.05), and RARα protein expression in the lesion ganglion cell group was significantly lower than that in the normal ganglion cell group (P<0.01). Compared with the control mice, ARM mice at E16.5-18.5 showed decreased fluorescence intensity and relative mRNA expression of RARα and NeuN in the rectum.

Conclusion Serum VA concentration and the RARα expression pattern are abnormal in the rectum in ARM and may contribute to the ENS maldevelopment in ARM.

Sonic Hedgehog Signaling Controls Gut Epithelium Homeostasis Following Intestinal Ischemia-reperfusion in a Rat
Igor Sukhotnik, Yoav Ben-Shahar, Yulia Poliak, Arnold G Coran, Zaki E Assi, Arie Bitterman

Purpose: The purpose of this study was to evaluate the role of Sonic hedgehog (SHH) signaling 24 and 48 hours following intestinal ischemia reperfusion (IR) in a rat.

Materials and methods: Male rats were divided into four: 1) Sham (24h) and 2) Sham (48h) rats underwent laparotomy and were sacrificed after 24 and 48 hours, respectively. 3) IR (24h) and IR (48h) rats underwent occlusion of both SMA and PV for 20 minutes followed by 24 or 48 hours of reperfusion. Intestinal structural changes, enterocyte proliferation and enterocyte apoptosis were determined 24 and 48 hours following IR. Ihh-related genes and protein expression were determined using Real-Time PCR, Western blot and immunohistochemistry.

Results: IR-24 rats demonstrated a significant decrease in GIL mRNA in jejunum and in Shh and PTCH2 mRNA in ileum compared to Sham-24 animals that was accompanied by a significant decrease (2.3-fold decrease) in the number of Shh-positive cells (Immunohistochemistry) in jejunum. After 48 hours, IR rats demonstrated a significant increase in Ihh, PTCH2 mRNA in jejunum as well as in Dhh, Ihh, SMO, GIL, PTCH2 mRNA compared to IR-24 animals that was coincided with increased number of Shh - positive cells in jejunum (2.6-fold increase) and ileum (1.4-fold increase).
Conclusions: 24 hours following intestinal IR, inhibited cell turnover was associated with inhibited SHh signaling pathway. Signs of intestinal recovery appeared 48 hours after IR and were correlated with increase in SHh signaling pathway activity.

S-29

**Experimental Study of DNA Methylation Modified Shh/Bmp4 signaling Pathway in the Regulation of the Enteric Nervous System Dysplasia in the Rectal End with Anorectal Malformations**

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**Objective** To study the influence of gene methylation in Shh/Bmp4 pathway on the ENS in the rectal end of fetal mice with ARM.

**Methods** Pregnant mice were randomly divided into ETU, ETU+5-azaC and normal group. On day 10, ETU and ETU+5-azaC group were gave in ETU via gavage, while ETU+5-azaC group was injected intraperitoneally with 5-azaC. On day 20, cesarian sections were underwent. The content of DNA methyltransferases, methylation status of Shh gene promoter and expression of the key components in Shh/Bmp4 pathway in the rectal end of fetal mice were detected.

**Results** (1) Expression of DNMTs in ETU and ETU+5-azaC group was higher than that of control group. Expression of DNMT1 and DNMT3a in ETU group was higher than that in ETU+5-azaC group. (2) Methylation level of Shh gene promoter in ETU and ETU+5-azaC group was higher than that in control group, and that in ETU group was higher than in ETU+5-azaC group. (3) Shh and Bmp4 expression in ETU and ETU+5-azaC group was lower than that of control group, and that in ETU group was also lower than in ETU+5-azaC group. (4) S-100 labelled submucosa and intermuscular nerve plexus in ETU group were decreased compared with that of control group, and ETU+5-azaC group was different from that in ETU group.

**Conclusions** Low methylation level of Shh gene may promote the expression of the key components in Shh/Bmp4 pathway, and the number of S-100 labeled nerve plexus may be significantly increased, which may promote the ENS development of the rectum.

S-30

**MiR-324-3p Functions as a Suppressor in ARMs by Targeting Wnt5a**

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**Aim to study:** Anorectal malformations (ARMs) represent a variety of congenital disorders that involve abnormal termination of the anorectum. Our previous study indicated that the expression of Wnt5a in ARMs of ETU-exposed rat embryos was decreased. The purpose of this study is to investigate the role of miR-324-3p by target Wnt5a in terminal hindgut in ethylenethiourea (ETU)-exposed rat embryos with anorectal malformations (ARMS).

**Methods:** Mature Wistar rats (body weights from 250g to 300g). ARM rat embryos were induced by 125mg/kg of 1% ETU on embryonic day 10 (E10) and the embryos were collected by cesarean delivery from E14-17. Real time PCR was applied to measure the expression level of miR-324-3p and Wnt5a mRNA in both IEC-6 cells and terminal hindgut tissues, and the expression level of Wnt5a protein was determined by western blot.

**Main results:** The expression of Wnt5a was significantly downregulated and the expression of miR-324-3p was obviously unregulated in terminal hindgut tissues of ARMs. In IEC-6 cells, silencing the expression of miR-324-3p could dramatically increase the expression of Wnt5a by RT-PCR and Western blot.

**Conclusions:** The expression of miR-324-3p increased by target Wnt5a in terminal hindgut in ETU-exposed rat embryos with ARMs and in IEC-6 cells. The miR-324-3p/Wnt5a pathway possibly provides new potential therapeutic clues for ARMs.
Consanguinity and its Relevance for the Incidence of Megacystis Microcolon Intestinal Hypoperistalsis Syndrome (MMIHS)
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Background/Purpose: Megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS) is a rare congenital and generally fatal cause of functional intestinal obstruction in the newborn. The cause of this syndrome is unknown. Familial occurrence and reports of consanguinity in MMIHS implies that genetic factors may have an important role in the pathogenesis of this syndrome. The aim of the study was to determine the consequence of consanguinity for the incidence of MMIHS.

Methods: A literature search was performed using the keywords “megacystis microcolon intestinal hypoperistalsis” for studies published between 1976 and 2018. Retrieved articles, including additional studies from reference lists, were reviewed for consanguinity between parents and recurrence of MMIHS between siblings. Data were extracted for cases where familial MMIHS was present.

Results: A total of 450 patients with the diagnosis of MMIHS have been reported in the literature. There were 76 (17%) cases in which familial MMIHS was confirmed, 25 families with multiple siblings and 23 families with single affected infant. Of the 25 families with multiple siblings, 22 families had two siblings with confirmed MMIHS and 3 families had 3 children each with MMIHS. Consanguinity between parents was confirmed in 30 cases (18 siblings and 12 individual case) (Table 1 and 2). Female to male ratio in the 30 patients was 4.4:1.

Conclusions: The occurrence of MMIHS in the offsprings of consanguineous parents and recurrence in siblings of healthy parents suggests that MMIHS is an autosomal recessive disorder. Pre-maritals and pre-conception counselling of consanguineous populations is recommended to prevent harmful consequences.

S-32

Magnetic Resonance Enterography: A Good Weapon in the Study and Staging of Pediatric Crohn’s Disease
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Aim of the study: The reference standard for diagnosing and monitoring Crohn’s disease (CD) is endoscopy. Magnetic resonance enterography (MRE) may represent an alternative technique to evaluate CD activity in children. Aim of our study is to propose a new simplified MR score for CD and to compare it with clinical and endoscopic indexes.

Methods: We retrospectively included children affected by CD, endoscopically confirmed, studied by MRE. Our Paediatric CD MR Index (PCDMRI) was based on mural/perimural parameters of the most affected intestinal tract and on extramural features. Correlation analysis was performed between the PCDMRI and: Paediatric CD Activity Index (PCDAI), Simple Endoscopic Score for CD (SES-CD), serum C-Reactive Protein (CRP) and fecal Calprotectin (fC). Agreement on disease location and inter-reader reproducibility of PCDMRI were also estimated.

Main results: Forty-two children, aged 13.5±2.6 years, were included, for a total of 80 MRE examinations. PCDMRI positively correlated with PCDAI (r=0.635, P<0.001), SES-CD (r=0.598, P<0.001), fC (r=0.402, P=0.005) and CRP (r=0.500, P<0.001). A positive association was found between PCDAI and all segmental parameters defining PCDMRI (P<0.001). The agreement on disease localization between MRE and endoscopy was substantial (κ=0.754; P<0.001). The inter-reader reproducibility was 91%.

Conclusions: PCDMRI significantly correlates with clinical and endoscopic parameters, proving to be a reliable tool to assess the severity of the disease. MRE advantages include the evaluation of extramural disease extension; the low invasiveness, that makes it an important tool in long-term follow-up; its feasibility in patients with small bowel stenosis, in which the use of capsule endoscopy would be contraindicated.
Pre-operative Management in Pediatric Crohn’s Disease Patients and Its Influence on Post-operative Outcome: 25 Years of Experience in a Tertiary Center

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1Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

Aim of the Study: Preoperative preparation of patients with Crohn’s disease is challenging, particularly in pediatric population. To this date, no specific guidelines exist towards pre-operative preparation and nutritional support in pediatric Crohn’s patients. The aim of this study was to assess whether preoperative preparation influenced the postoperative outcome.

Methods: A retrospective, cohort study including all Crohn’s disease pediatric patients who underwent abdominal surgery at the Schneider Children’s Medical Center of Israel between the years 1993 and 2018 was conducted. Patients clinical and surgical data were recorded and analyzed.

Results: Forty-four patients (aged 10-18) were operated during the study period due to Crohn’s exacerbation and underwent ileo-colic resection. 70% of the patients (n=31) underwent elective or semi-elective surgery due to impending obstruction, unresolved abdominal pain or failed medical therapy while 30% patients (n=13) underwent urgent surgery due to peritonitis or complete obstruction. In 12 patients that underwent elective or semi-elective surgery, preoperative preparation included parenteral and enteral nutrition as well as the use of IV antibiotics and anti-inflammatory drugs. Post-operative complications were noted in 6 (13.6%) patients, all of them underwent urgent surgical intervention (n=6).

Conclusion: Although less investigated than in adults, preoperative preparation and optimization towards better post-operative results is pediatric Crohn’s patients gain more attention in recent years. A multidisciplinary team, including a pediatric surgeon, gastro-enterologist and dietitian should outline a tailor-made strategy for each elective surgery candidate to minimize post-operative complications.

Acquired Hypoganglionosis as a Distinct Entity: Results from a Nationwide Survey

Satoshi Obata, Koichiro Yoshimaru, Kosuke Kirino, Tomoko Izaki, Satoshi Ieiri, Atsuyuki Yamataka, Shigemichi Koshinaga, Jun Iwai, Hitoshi Ikeda, Tomoaki Taguchi

1Department of Pediatric Surgery, Buzzi Children’s Hospital, Milan, Italy, 2Department of Pediatrics, Buzzi Children’s Hospital, Milan, Italy

Aim of the Study: Crohn’s disease (CD) may affect any part of the gastrointestinal tract (GIT). While inflammatory involvement of the proximal GIT varies from 30 to 40% of cases, stenosis of the gastroduodenal site is extremely rare (0.4-5%) especially in children. The aim of the study is to report 4 cases of CD presenting as duodenal and/or pyloric stenosis.

Main Results: All our patients (F:M=2:2, age 7-15 years) experienced acute onset of symptoms: vomiting, weight loss and difficult oral feeding (2 cases). Two patients had delayed puberty. In all cases inflammatory markers were normal. Endoscopy showed gastro-duodenal stenosis, associated with duodenal and pyloric ulceration in two patients. Involvement of other GIT sites was very mild in all cases. Initial management consisted of nutritional implementation and medical therapy. Two patients were refractory to conservative treatment and underwent surgery: gastrointestinal “Roux-en-Y” bypass after temporary video-assisted jejunostomy in one case; duodenal strictureplasty in the other. Post-operative course was uneventful and long-term follow-up (5 years) is excellent. Of the remaining two patients, one was well responsive to medical treatment (steroids, azathioprine and subsequent infliximab with proton pump inhibitors); the other one is now having symptom recurrences and he is scheduled for surgery.

Conclusions: Despite gastroduodenal localisation of CD is rare, it should be suspected in patients with upper GIT symptoms. An accurate complete GIT diagnostic evaluation is important. Medical treatment should be attempted in first instance but surgery may be required in cases of persistent severe obstructive disease. In our series ¾ cases required surgery.
Conclusion: A-HG is rare but distinct entity characterized by different clinical courses and pathological findings from those of C-HG. The outcome is considered to be favorable after a resection of affected intestine.

S-36
Risk Factors for the Development of Post-operative Enterocolitis in Short Segment Hirschsprung’s Disease
Patrick Ho Yu Chung¹, Kenneth Kak Yuen Wong¹, Michelle On Na Yu¹, Paul Kwong Hang Tam¹
¹Department of Surgery, The University of Hong Kong, Hong Kong, China

Aim of the study The objective of this study is to identify risk factors associated with the development of post-operative enterocolitis (HACE) in short segment Hirschsprung’s disease (HSCR-S)

Methods A retrospective study was carried out to follow-up patients with HSCR-S from 1997 to 2017. HSCR-S was defined as the most proximal extension of aganglionosis limited to the sigmoid colon. An episode of HACE was defined as the presence of i) vomiting or explosive diarrhea; ii) abdominal distension; iii) fever and iv) leukocytosis. Risk factors for the development of HACE were determined using multivariate logistic regression.

Main results The medical records of 96 patients were reviewed. The overall incidence of HACE was 20.8% (n=20) and 65.0% (n=13) of HACE occurred within the first year of operation. After a univariate logistic regression analysis, 3 risk factors for HACE were identified: 1) presence of other major anomalies (OR: 1.43 (1.12-2.32), p = 0.041); 2) pre-operative stoma creation (OR: 2.28 (1.47-3.23), p = 0.035); 3) extension of aganglionosis proximal to the rectum (OR: 1.89 (1.05-3.19), p = 0.049). After multivariate logistic regression analysis, a significant association was demonstrated for pre-operative stoma creation (OR: 1.81 (1.08-3.22), p =0.045) and extension of aganglionosis proximal to the rectum (OR: 1.91 (1.37-2.98), p = 0.038).

Conclusions The requirement of pre-operative stoma creation and a more proximal extension of aganglionosis are risk factors for the development of post-operative HACE in HSCR-S. Patients with these risk factors should be closely followed up especially during the first year after the operation.

S-37
The Importance to Preserve a Small Distal Rectal Reservoir for Idiopathic Constipation. Transanal Partial Proctosigmoidectomy
Lea A Wehrli¹, Luis De La Torre¹
¹Department of Pediatric Surgery, Colorectal and Hirschsprung Center at the Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

We present a new surgical approach for the treatment of idiopathic constipation complicated with a problematic megarectosigmoid.

Methods We had performed the transanal partial proctosigmoidectomy preserving a small distal rectal vault. The resection entails the dilated proximal rectum and sigmoid leaving a rectal reservoir of 5 cm. Thirteen patients with idiopathic constipation complicated by a megarectosigmoid underwent this promising procedure.

Results Thirteen patients underwent transanal partial resection of the rectosigmoid. Postoperatively the laxative dose in our patient cohort was reduced significantly (p = 0.007). The postoperative follow-up time ranged from 2 - 12 months. All patients have fecal control and daily voluntary bowel movements. No intra and postoperative complications occurred.

Conclusion We present a new surgical approach for the treatment of chronic idiopathic constipation. Our preliminary results are promising.

Sunday 7th October

S-38
Successful Surgical Management for a Rectourethral Fistula with a Rectoscrotal Fistula
Reina Hoshi¹, Shuichiro Uehara¹, Kensuke Ohashi¹, Tsugumichi Koshinaga¹
¹Department of Pediatric Surgery, Nihon University School of Medicine, Tokyo, Japan

Aim of the Study: To suggest a new operative strategy for H-type anorectal malformations (ARMs), which often accompany genitourinary anomalies.

Methods We herein report successful operative management in a boy who developed a rectourethral fistula with a rectoscrotal fistula, which is extremely rare among all the H-type ARMs.

Mail Results: A 1-day-old boy was admitted to our department with an anal defect. A rectosigmoid fistula was found, and the meconium was detected in the urine. Therefore, we diagnosed the H-type ARM and created sigmoid colostomy. Voiding cystourethography and fistulography revealed that he had a rectobulbar fistula, and the common wall of the anterior wall of the rectum and the posterior wall of the urethra was nearly 3 cm in length. Before anorectoplasty, we confirmed that his urethra was normal by cystoscopy. Then, posterior-sagittal anorectoplasty (PSARP) was attempted at the age of 8 months. The common wall between the anterior rectum and the posterior urethra were very thin and long, hence, we incised the rectal anterior wall longitudinally in the direction of 1 and 11 o’clock and made the rectum strip-shaped and remodeled it not to injure the urethra. No complications were observed for 15 months after PSARP.

Conclusions: We presented an extremely rare case of a rectourethral fistula with rectoscrotal fistula. To the best our knowledge, this is the third case in English literature. We suggest that our method is useful as one of the operative strategies for rectourethral fistulas with the long common wall.
S-39
Unusual Types of Rectourethral Fistula in Arm
Heidy Tomas1, Anselmo Mondragon2, Jorge Martinez2, Aquiles Uribe2, Maria Cabello2
1Edgardo Rebagliati Hospital, Lima, Peru

The aim of this review is to establish the importance of making a complete study to detect associated urologic malformations.

Methods: Medical charts, radiologic studies and videoendoscopic procedures of 3 male patients with ARM with rectourethral fistula have been reviewed.

Results: The high-pressure distal colostogram and video endoscopy were fundamental to make the diagnosis of uncommon urethral malformations.

Conclusion: In rare cases like posterior cloaca with aphallia and ARM with urethral anomalies, studies like distal colostogram and cystoscopy are an important stool to make the right diagnosis and surgical treatment.

S-40
Constipation in Diagnosis of Multiple Endocrine Neoplasia Type 2B. Three Case Report
Jose Eduardo Frias Mantilla1, Karla Alejandra Santos Jasso1, Pablo Lezama del Valle1, Maria Antonieta Cabrera Hernandez1, Rodolfo Rodriguez Jurado3, Jose Adolfo Peralta Bustamante4
1Pediatric Surgery Department, National Institute of Pediatrics, Mexico City, Mexico, 2Pediatric Surgery Department, Star Medica Hospital Infantil Privado, Mexico City, Mexico, 3Pathology Department, National Institute of Pediatrics, Mexico City, Mexico, 4Angeles Hospital, Leon Guanajuato, Mexico

Intestinal ganglioneuromatosis (GNI) is part of the group of intestinal dysgangloniosis and is a rare disease, corresponding to less than 0.1% within this group. However, its presence has been reported in up to 40-90% of patients with multiple endocrine neoplasia 2B (MEN2B).

The mean age of diagnosis of MEN2B is at 18 years because at lower ages the symptomatology is scarce or nonexistent. Gastrointestinal pathology is the first to manifest in these individuals and may be the only manifestation until adolescence or adulthood.

In about 40% of patients younger than one year of age with diagnosis of MEN2B, gastrointestinal symptoms were the initial manifestation, mainly constipation (72.7%) which is associated with intestinal ganglioneuromatosis. Hence, this may be the only early manifestation in these cases, allowing an accurate diagnosis of MEN2B and its adequate management with an opportune prophylactic thyroidectomy.

In our study, we refer three cases in which the diagnosis of intestinal ganglioneuromatosis was made within the first year of life allowing the suspicion of NEM2B, later confirmed by RET gene tests which lead to an opportune prophylactic thyroidectomy showing histopathological findings of medullary thyroid carcinoma.

Until 2016 just six cases have been reported in the literature where the diagnosis was made during the first year of life, allowing the timely detection of medullary thyroid cancer.

Despite their low frequency, the suspicion of GNI in patients with gastrointestinal disorders and constipation cut allows the detection of MEN2B in early stages.
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<th>S-41</th>
<th>Caudal Duplication Syndrome with Recto-vaginal Fistula</th>
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<tr>
<td>Dmitry A Morozov1,2, Evgeniia S Pimenova1,2, Eduard K Ajrann1,2, Dmitry D Morozov1</td>
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The authors present a case-report of caudal duplication syndrome with recto-vaginal fistula. A full-term female newborn was misdiagnosed after birth. She had coprostasis and coproliitis at the age of 4 months. Recto-vaginal fistula with normal anus was revealed. The patient was referred to a federal medical center. Perineal examination showed two meatuses, two vaginal openings, rectovaginal fistula and normal anus. Contrast enema through the rectum and fistula detected colorectal duplication. Contrast cysto-urethrogram of each meatus showed two bladders without vesico-ureteral reflux. Intravenous pyelography showed two normal kidneys and two ureters. Cystoscopy confirmed complete bladder and urethral duplication, with two normal bladder necks, vaginoscopy revealed two vaginas with a normal uterine cervix. A laparotomy showed intestinal duplication consisted of two rectums, two colons, two appendixes, two ileum with Meckel diverticulum on one. There was non-fixation of colon. Rectovaginal segment was strongly dilate. Appendices and Meckel diverticulum were resected. Modified pull-through procedure was performed: abdominal extrarectal dissection of doubled rectum and submucosal dissection of vaginal fistula. Sigmoideal septum was resected and the duplicated sigmoid colon was anastomosed to the normal rectum on 4-5 cm above the dental line. Sigmostomy was performed for all segment. Ostomy closure was done after 2 months. Girl underwent comprehensive investigation after 4 years. She did not show any complaints, did not have fecal and urinary incontinence. No surgery was performed for the urinary duplication.

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<th>S-42</th>
<th>Laparoscopy-assisted Total Colectomy for an Eight-year-old Girl with Colorectal Cancer Due to Colon Polyposis</th>
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<tr>
<td>Takaharu Oue1, Masahiro Zenitani1, Gakuto Tani1, Natsumi Tanaka1, Kazuo Tamura2, Hiroki Kurahashi3</td>
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**Background:** According to typical adenomatous polyposis series, colorectal cancer usually develops in the third or fourth decade. We herein report a female patient with extremely rare colon polyposis who developed adenocarcinoma at the age of eight years. We performed laparoscopy-assisted total colectomy used for Hirschsprung’s disease.

**Case:** An eight-year-old girl was admitted with a four-year history of occasional bloody stools. Colonoscopy revealed colon polyposis and among 14 adenomatous polyps, pathological assessment confirmed well-differentiated adenocarcinoma in adenoma in four of them. Therefore, laparoscopy-assisted proctocolectomy proceeded. Total colon was mobilized and resected using the single port laparoscopic-assisted methods. Proctectomy and ileo-anal anastomosis was transanal-modified Soave procedure used for Hirschsprung’s disease. The pathological findings revealed scattered well-differentiated adenocarcinomas not only in polyps but also in non-polypoid lesions. Because lymph node metastases were not found, additional adjuvant chemotherapy was not administered. The patient has remained alive and disease-free for one year. Genetic analysis using the real-time PCR for the APC and MUTYH genes, multiple target DNA sequencing including the BMPRA1, SMAD4 and PTEN genes, and multiplex ligation-dependent probe amplification (MLPA) of deletions or duplications within or including APC, MUTYH and GREM1 genes did not identify any responsible genes. Post-operative course was uneventful. The patient has muddy stool around 5 times per day and followed for 1 year without recurrence of disease.

**Conclusion:** Laparoscopy-assisted total colectomy is a feasible procedure for young children who require the total colectomy.

<table>
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<th>S-43</th>
<th>Nerve Sparing Ultra-fine Dissection of Laparoscopic Assisted Anorectoplasty for Recto-bulbar Urethral Fistula Using 4K Image and 3.5 mm Bipolar Scissors</th>
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<td>Satoshi Ieiri1, Masakazu Murakami1, Yoshio Harumatsu1, Keisuke Yano1, Tokuro Baba1, Shun Onishi1, Koji Yamada1, Waka Yamada1, Ryuta Masuya1, Takaumi Kawano1, Seiro Machigashira1, Kazuhiko Nakame1, Motoi Mukai1, Tatsuru Kaji1</td>
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</table>

**Department of Pediatric Surgery, Kagoshima University, Kagoshima, Japan**

**Purpose:** Laparoscopic anorectoplasty for recto-bulbar urethral fistula is technically challenging procedure because of small pelvic space and difficulty of detail anatomical recognition. We presented the nerve sparing fine procedure of laparoscopic assisted anorectoplasty for recto-bulbar urethral fistula using 4K image and 3.5 mm bipolar scissors.

**Case and Operative Procedures:** A 6 months old boy was diagnosed as recto-bulbar urethral fistula by contrast enema. Body weight was 6.5 kg and no associated anomalies was recognized. Transverse colostomy was made on left upper abdomen at neonatal period. A 5-mm 30° laparoscope was inserted through the umbilical incision. After pneumoperitoneum was established, bladder was sustained by stay suture. Peritoneal reflection was opened and rectum was dissected carefully on using 3.5 mm bipolar scissors in small pelvic cavity. Tiny vessels were all coagulated by this bipolar and bleeding was not recognized. A recto-bulbar urethral fistula was ligated by trans-fixing suture and resected. Center of muscle complex including pubo-rectal sling was confirmed by electrical stimulation from both inside and outside. Rectum was pulled through the center of muscle complex and anoplasty was performed.
Conclusion: High resolution 4K image gave us clear anatomical recognition. Based on this image findings and recognition, ultra-fine dissection and the lowest invasive surgery are possible.

S-44
Urinary and Fecal Outcomes in a Patient with Complex Congenital Pouch Colon Anomaly
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INTRODUCTION The anatomy of congenital pouch colon (CPC) anomaly associated with cloacal persistence is complex leading to challenging repairs and questionable outcomes.

CASE REPORT A newborn girl with cloacal persistence presented with sign of intestinal obstruction at another hospital. Laparotomy revealed a type-I CPC and intestinal malrotation with mesenteric hernia. The hernia had been reduced, the mesenteric gap closed and an ileostomy created.

The patient presented to our clinic at the age of three months. The investigations demonstrated the cloacal anomaly with double vagina, urine incontinence and vesicoureteral reflux. The urogenital sinus was 3.5 cm long and the distance between the rectal pouch and perineum was 5.5 cm. At laparotomy we dissected the 7 cm long pouch and the fistula into the septum of the double vagina was ligated and cut. The pouch was converted into a tubular structure.

We performed also a urogenital mobilization, transection of the vaginal septum, urethroplasty and vaginoplasty by a posterior sagittal approach. We completed the anorectoplasty by pull through of the “tubularized” colonic pouch. Two months later, we closed the ileostomy. At five-year follow-up, the patient is fecal continent (two-to-three defecations per day), the appearance of external genitalia is normal. She has a urethrovaginal fistula and urinary incontinence.

CONCLUSION The long-term prognosis of urinary/fecal continence and quality of life is uncertain for cloacal anomalies combined with all subtypes of CPC. Our patient will need further surgical procedures to improve urinary continence.

S-45
Two Cases of Anorectal Malformation with Anorecto-scrotal Fistula
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Case 1: A boy was born by spontaneous vaginal delivery after threatened preterm labor at 31 weeks gestation, elsewhere; birth weight: 1,380g. At birth, anorectal malformation (ARM) was diagnosed and an orifice was noted to be present low in the scrotum. On X-ray radiography after nasogastric tube insertion, the tube was found to be coiled up and the stomach distended, consistent with esophageal atresia and tracheoesophageal fistula. Emergency sigmoid colostomy and primary anastomosis of the esophagus were performed.

After referral to our hospital at the age of 6 months, examination under general anesthesia and colostography identified an anoscrotal fistula running partially parallel to the urethra (Fig: Case 1). Anterior sagittal anorectoplasty (ASARP) was performed at the age of 15 months. Because of a risk that the fistula may be attached to the ventral wall of the urethra, the fistula was laid open, and the rectum mobilized and anastomosed to the proposed anal site. While laying the fistula open, a segment of the mid-portion of the fistula tract was found to be partially attached to the urethra in the corpus spongiosum, which caused some bleeding when incised, but which was controlled by suturing. The mucosa of the laid open fistula was excised. After surgery, the fistula became completely epithelized.

Case 2: A boy was born by spontaneous vaginal delivery at 39 weeks gestation, elsewhere; birth weight: 2,600g. At birth, ARM was diagnosed which was classified as intermediate-type on ultrasonography, because the distance from the perineum to the pouch of the rectum was 2cm. Emergency transverse colostomy was performed. On day 5 after surgery, meconium was passed from the scrotum.

After referral to our hospital at the age of 1 month, fluoroscopy and examination under general anesthesia identified a rectoscrotal fistula that ran parallel and very close to the urethra, as in Case 1, but which was deep in the perineum, in contrast to Case 1. Posterior sagittal anorectoplasty (PSARP) was performed at the age of 21 months (Fig: Case 2). To prevent injury to the urethra distal to the bulbar urethra, the fistula was divided from the rectum at a level equivalent to the distal end of the prostate and left untouched. After surgery there was no problematic discharge of mucus from the residual fistula. Laser excision of the mucosa of the residual fistula will be performed later, because it is a safer procedure in larger children.
S-46
Transition of Care - The Management of Adult Patients Born with Colorectal Problems
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INTRODUCTION: Traditionally, the care of children and adults has been arbitrarily separated between pediatric and adult surgeons. Despite progress in pediatric surgical techniques, patients still suffer from significant functional sequelae, which persist into adulthood. We aim to describe the needs of adult patients with congenital colorectal malformations.

METHODS: Following IRB approval, a retrospective database review was performed looking for all adult patients who were treated by our group from 1983 until 2017.

RESULTS: 88 cases were identified. 51 patients suffered from an ARM, 18 cloacas, 9 pre-sacral masses, 3 HD, 2 spina bifida and 5 with other diagnoses (3 vaginal anomalies, 1 cloacal extrophy, 1 obstructed seminal vesical). The specific problems that affected the patients were: complications from previous operations (41), rectal prolapse (25), fecal incontinence (11), gynecologic concerns (12), urologic concerns (6), and recurrent fistula (3). We performed 83 surgical interventions, including 13 rectal prolapse repair, 13 continent appendicostomies, 44 PSARP or re-do PSARP, 11 resections of pre-sacral masses, 11 vaginoplasties, 2 examinations under anesthesia, and 2 Mitrofanoff procedures. Five patients were treated medically (bowel management program, obstetric, urologic evaluation).

CONCLUSION: There is an increase need to better prepare adult providers to assume the care of patients born with congenital colorectal disease, as they transition to adulthood. A collaboration between our center with adult colorectal surgeons, urologists and gynecologists has been implemented to guarantee the patients have a smooth transition from pediatric to adult type of care.

S-47
Association of Hirschsprung’s Disease and Anorectal Malformation - It Is Rare and That Is Good
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BACKGROUND: The simultaneous occurrence of Hirschsprung’s disease (HD) with an anorectal malformation (ARM) is rare, with few cases published in the literature. We have seen misdiagnosis and treatment of HD in patients with an ARM, due to severe constipation that ARM patients suffer from. Surgical specimens showed the presence of ganglion cells. The aim of the present study was hence to assess the incidence of simultaneous diagnoses of ARM and HD in a single large cohort of ARM patients.

METHODS: A retrospective review of our database from 1980 to 2018 identified 150 patients with HD and 2316 patients with ARM. Three patients suffered from both, HD and ARM.

RESULTS: The incidence of HD in ARM patients was 3/2316 = 0.13%, and the incidence of ARM in HD patients was 3/150 = 2%. Two patients were female. All patients had chromosomal abnormalities: Trisomy 21, Phox 2B mutation, and multiple chromosomal deletions with the most important being 10p151p14. Full thickness biopsies of the rectum and sigmoid confirmed the diagnosis prior to the pull through. In one patient, the diagnosis was delayed and only suspected after he was admitted in septic shock following colostomy closure. All patients are fecally incontinent and clean with daily enemas.

CONCLUSION: The combination of HD and ARM is rare, but when it happens, patients will be fecally incontinent. Proper counseling should be given to families and, most importantly, clinicians should doubt and confirm biopsies prior to offer a pull through for Hirschsprung in patients with ARM.

S-48
Measuring Common Canal of Persistent Cloaca: Can MRI Substitute Conventional Imaging?
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AIM: To evaluate the role of MRI in preoperative assessments of patients with persistent cloaca and compare MRI versus fluoroscopy contrast study in the accuracy of common canal measurement and classification prediction.

MATERIALS AND METHODS: Thirty-one patients with persistent cloaca were diagnosed and treated at the author’s hospital between March 2011 and December 2017. The length of the common canal was measured using MRI and fluoroscopy contrast study and confirmed by cystoscopy and intraoperative findings. Classification results based on measurements using MRI and fluoroscopy contrast study were compared with cystoscopy and intraoperative findings. The accuracy in predicting the classification by measuring the common canal length were compared between MRI and fluoroscopy contrast study.

RESULT: Among 31 patients, 27 had MRI, 25 underwent fluoroscopy contrast study, and 24 underwent cystoscopy. In 20 patients, MRI-based categorizations were in accordance with cystoscopy or surgery findings, while in 4 patients there was
discordance. In 17 patients, categorizations based on fluoroscopy contrast study were in accordance with cystoscopy or surgery findings, and in 7 patients there was discordance; the difference was not statistically significant (P>0.05).

CONCLUSION: MRI may accurately demonstrate genitourinary anomalies and the length of the common canal in patients with persistent cloaca. Categorization based on MRI measurements of the common canal were precisely accordant with results from cystoscope and findings from the final procedure. The use of this method may help surgeons to develop appropriate reconstruction plans before sending their patients to the operating room.

S-49

Bowel Management in Patients with Spina Bifida

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Purpose: Our center has been successfully implementing a bowel management program (BMP) for fecal incontinence consecutive to anorectal malformation and Hirschsprung. Recently, the number of patients with spina bifida requiring management for fecal incontinence has increased. The purpose of this study was to review the results of bowel management in patients with spina bifida and the challenges unique to this population.

Methods: A retrospective chart review of patients with spina bifida who attended the BMP from February 2016 until April 2018 was performed. Data collection included: prenatal intervention, gender, age, characteristics of contrast enema, success rate and challenges faced.

Results: Twenty-two patients met inclusion criteria of which 13 were female. Three patients had their myelomeningocele repaired prenatally, the remaining were repaired postnatally. Patient’s age ranged from 2 to 24 years. Only nine patients were referred to BMP at proper toilet training age. Two patients came to BMP status post antegrade enema procedure with reported “accidents” on their current regimen. The colon in the contrast enema was non-dilated in all patients but one behaved as hypermotile requiring loperamide. Seventeen patients (77%) were clean of stool and considered successful. The most common challenge, in this population, was leakage of the solution during enema administration due to lack of sphincter tone.

Conclusions: A BMP with enemas is effective for patients with a history of spina bifida. The data support specific considerations for this population including frequent adjustments, close follow up and specific administration techniques.

S-50

Fecoflowmetric Analysis of Postoperative Bowel Functions in Pediatric Patients; Comparison to Krickenbeck Score and Kelly’s Clinical Score

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1Department of Pediatric Surgery, Kurume University, School of Medicine, Fukuoka, Japan

Aim of the Study The aim of the study was to clarify the characteristics of the parameters of fecoflowmetry (FFM) by comparing the conventional bowel function scoring systems in the children underwent anorectal surgery.

Methods This retrospective study enrolled the patients who underwent anorectal surgery for Hirschsprung’s disease (HD) or anorectal malformation (ARM). All patients were assessed postoperative bowel conditions via FFM as well as Krickenbeck score (KS) and Kelly’s clinical score (KCS). All patients were classified according to KS or KCS and the five FFM parameters were compared between each group.

Main Results Sixteen patients (M/F: 11/5, HD: 4 and ARM: 12) were enrolled. According to KS, 9 patients showed soiling (non-soiling: 7, G1: 5, G2: 4), whereas 6 patients showed constipation (non-constipation:10, G2:4, G3:2). Number of patients classified according to KCS were as follows; continence (1):7, (2):9, staining (1):8, (2):8, Sphincter squeeze (0):2 (1):6, (2):8. In KS, TR of non-soiling (79.86±24.04%) and G1 patients (79.53±13.33%) were significantly higher than that of G2 patients (32.11±22.00%), respectively (p=0.0283 and 0.0200). In KCS, Fmax of staining (2) (no staining) patients (64.56±30.08ml/sec) was significantly higher than that of staining (1) (occasionally) patients (36.46±16.44ml/sec) (p=0.0239). Additionally, Fmax of “good” patients in KCS (66.91±25.38ml/sec) was significantly higher than that of “fair”
patients (34.88±20.57ml/sec) (p=0.0177).

Conclusions This study indicated that FFM provides quantitative and detailed informations about bowel functions. In particular, Fmax best reflects bowel functions, and TR could be a good indicator of continence in the pediatric patients with anorectal surgery.

S-51

The Ideal Anorectal Pull-through Route Based on Observations by Three-dimensional (3D) MR Imaging and Intraoperative MR Navigation

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Aim of the Study is to understand and elucidate the ideal anorectal pull-through route for patients with anorectal malformation based upon observations by MR technologies, which have made it possible to visualize the 3D anatomy and apply to the operative field.

Methods: Three-dimensional images of the pelvic structure were obtained from 23 preoperative and 24 postoperative patients with various types of anorectal anomalies as well as 14 controls. Seven who required sacroperineal anorectoplasty were assessed for the pull-through routes by using a 3D MR real-time navigation system EasyGuide Neuro (Philips Medical Systems, The Netherlands).

Main Results: MRI can distinctively demonstrate 3D images of the soft tissues, especially muscle complex. For better postoperative function the anorectum is to be through the muscle in the middle supported from behind and with a sharp anterior angulation to reach the anal orifice even in the very thin high type structure. MR navigation system demonstrated that the inlet of the muscle complex was supposed to be just behind the bulbar urethra and the anal orifice should be in the center of the anal pit confirmed by electrical stimulation. Postoperative 3D images of the pelvic structure from those with poor function demonstrated disfigurement of the muscle complex and/or displacement.

Conclusions: An anorectoplasty should aim to reconstruct the anatomy by placing the anorectum in the middle of the muscle complex with an angulation. When the muscle complex is divided sagittally, it should be carefully approximated not to be disfigured from the ideal 3D structure.

S-52

Fact or Myth? The Long Shared Common Wall between the Fistula and Urethra in Recto-urethral Bulbar Fistula Type of Male ARM

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Aim: It has long been considered surgical dogma that the length of the shared common wall (CW) between the fistula and urethra in males with rectourethral bulbar fistula (RUBF) was considerably longer than the rectourethral prostatic fistula (RUPF). This belief led surgeons who perform laparoscopic-assisted anorectoplasty (LAARP) for RUPF to avoid LAARP for the RUBF on concerns of urethral injury or incomplete removal of the fistula. In this study, we compared CW lengths of RUBF and RUPF using distal colostograms (DCG) and direct intraoperative measurements.

Methods: CWs of 63 DCGs of rectourethral fistula patients were retrospectively measured along with L4 vertebral body height and expressed as a ratio of CW/L4, <0.7 = “short”; 0.71-1.4 = “medium”; >1.41 = “long”. Twenty-four of these patients also had intraoperative CWs measured during LAARP as previously described (Figure) and the results of both techniques compared.

Results: Surprisingly, 48% of RUBF had short CWs, 27% medium, 20% long and 5% indeterminate on DCG, which was similar for mean intraoperative measurements of 7 mm, 8.5 mm, 10.3 mm CWs in short, medium and long patients, respectively (Table). The RUPF had 74% short, 10% medium, 5% long on DCG, while the mean measurements were 5 mm, 7 mm, and 10 mm, respectively. The mean intraoperative RUBF and RUPF CWs were similar (p=NS).

Conclusion: Approximately half of RUBF CWs were “short” using two independent methods, and only about one quarter
were “long”, casting doubt on the long shared common wall notion. These findings suggest LAARP should not be excluded for treatment of select RUBF.

<table>
<thead>
<tr>
<th></th>
<th>DCG common wall as a ratio of iliac vertebral body height (n=63)</th>
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<tbody>
<tr>
<td></td>
<td>RUBF</td>
<td>SP/AP</td>
<td>RUBF/SP/AP</td>
</tr>
<tr>
<td>Short</td>
<td>21</td>
<td>14</td>
<td>7.0 (5-12)</td>
</tr>
<tr>
<td>Medium</td>
<td>12</td>
<td>2</td>
<td>5.0 (4-6)</td>
</tr>
<tr>
<td>Long</td>
<td>9</td>
<td>1</td>
<td>8.5 (8-10)</td>
</tr>
<tr>
<td>Indeterminate</td>
<td>2</td>
<td>2</td>
<td>10.3 (8-15)</td>
</tr>
</tbody>
</table>

**Table:**

<table>
<thead>
<tr>
<th></th>
<th>DCG common wall length versus LAARP common wall length (n=24)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean RUBF length (range)*</td>
</tr>
<tr>
<td>Short</td>
<td>7.0 (5-12)</td>
</tr>
<tr>
<td>Medium</td>
<td>8.5 (8-10)</td>
</tr>
<tr>
<td>Long</td>
<td>10.3 (8-15)</td>
</tr>
<tr>
<td>Indeterminate</td>
<td>NA</td>
</tr>
<tr>
<td>Overall</td>
<td>7.7 (5-15)</td>
</tr>
</tbody>
</table>

LAARP: laparoscopically assisted anorectoplasty; *: mm; NA: not available

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**S-53**

Long-term Follow-up for Anorectal Function after Anorectoplasty in Patients with High/Intermediate Imperforate Anus: A Single Center Experience

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**Aims of the study:** The aim of this study was to evaluate the clinical outcomes and postoperative anorectal function in the patients with high/intermediate imperforate anus (HIA/IIA) treated with the sacroperineal/sacroabdominoperineal pull-through anorectoplasty (SP/SAP), SP with anterior perineal incision (API) and Potts procedure (Potts). In addition, we report our novel procedure, laparoscopically assisted anorectoplasty with API (LAARP-API), which allows direct visualizations of the puborectal sling and muscle complex, and is useful for the creation of a pulling-through route for the rectal pouch.

**Methods:** From 1976 to 2016, 22 patients (19 boys and 3 girls) with HIA and 43 patients (34 boys and 9 girls) with IIA underwent SP/SAP, SP-API, Potts or LAARP-API. Clinical data and anorectal function of those patients were retrospectively evaluated using the Japanese clinical score at the selected ages.

**Main Results:** Of the 22 cases of HIA, 15 were treated by SAP, 2 were SP and 5 were LAARP-API. Of the 43 cases of IIA, 1 was treated by SAP, 31 were SP, 2 were Potts and 9 were SP-API (Table). The mean score of anorectal function of HIA/IIA both increased with age (Figure). In IIA, the score after SP-API was significantly higher than the score after SP through all age groups.

**Conclusion:** Long-term outcomes of our anorectoplasty for HIA/IIA is good with excellent anorectal function score. LAARP-API may be an option for HIA.
**S-54**

The Long-term Bowel Function Outcomes of 73 Consecutive Cases of High and Intermediate Type Anorectal Malformation in a Single Institution

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**Aim of the Study:** The operative results and long-term bowel function outcomes in high- and intermediate-type anorectal malformation (ARM) patients were analyzed and evaluated in our institution.

**Methods:** Patient data were collected from 1984 to 2017. Seventy-three patients with high- and intermediate-type ARM who underwent a definitive operation at our institution were enrolled; cloaca was excluded. The patients’ characteristics, type of ARM, associated anomalies and postoperative bowel function at their most recent evaluation were reviewed based on their medical records and analyzed retrospectively. The bowel function was evaluated according to the evacuation score (ES) of the Japan Society of Anorectal Malformation Study Group. A maximum score of 8 points indicates an excellent bowel function.

**Main Results:** Fifty patients had high-type ARM (recto-prostatic fistula, n=4; recto-urethral fistula, n=46), and all patients were male. The average age at anorectoplasty was 5.2 months. Associated anomalies were urogenital, n=24; cardiovascular, n=8; and abnormal sacrum, n=8. The average ES was 5.2 points. Twenty-three patients had intermediate-type ARM (female, n=8 [no fistula, n=4; recto-vaginal fistula, n=1; recto-vestibular fistula, n=3]; male, n=15 [no fistula, n=6; recto-bulbar fistula, n=9]). The average age at anorectoplasty was 8.2 months. The associated anomalies were urogenital, n=5; cardiovascular, n=5 and chromosomal, n=5 (trisomy 21). The average ES was 5.9 points.

**Conclusions:** Regarding the timing of the definitive operation, intermediate-type was treated later than high-type ARM. The average ES score of intermediate-type ARM was higher than that of high-type, but not significantly. Further efforts should be made to obtain the favorable outcomes by modifying operative techniques using recently introduced laparoscopic surgery.

**S-55**

Results of Peristeen® as a Modality for Enema Delivery in Pediatric Patients

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**Purpose:** The purpose of this study is to review our population of bowel management patients who use or have used a Peristeen® device for daily enema administration to evaluate success and failure rates as well as reasons for discontinuing it.

**Methods:** A retrospective chart review was done from January 2015 until June 2018. Data collection included: age, diagnosis, type of enema, duration of Peristeen® usage, and success or failure reason.

**Results:** Twenty patients were identified with ages ranging from 4 to 16 years old. Duration of Peristeen® use was one month to 3 years. Patients suffered from fecal incontinence due to: anorectal malformation (8), spina bifida (7), profound developmental delay (3), and other (2). Nineteen patients used a combination of pure normal saline or addition of glycerin and/or castile soap, while only one patient used water. 16 patients were successful (table 1) with Peristeen®. Reasons for stopping Peristeen® included: patient fear of the equipment (2), insurance denial (2), technical problems with the balloon (3), Peristeen® not as effective as prior enema regimen (1), and patient went on to have a planned anterograde continent enema procedure (2).

**Conclusions:** Peristeen® is an effective bowel treatment strategy for 80% of our selected patient population. In the United States, insurance plays an important role in deciding who is allowed to try this device. Proper training is needed as well as finding the effective, individualized solution for each patient. Patients who are successful with Peristeen® tend to be older than children who are unsuccessful.

**Table 1.**

<table>
<thead>
<tr>
<th></th>
<th>Continue to Use</th>
<th>No Longer Use</th>
<th>Total</th>
<th>Mean Age (Years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Successful</td>
<td>12</td>
<td>4</td>
<td>16</td>
<td>10.9</td>
</tr>
<tr>
<td>Unsuccessful</td>
<td>0</td>
<td>4</td>
<td>4</td>
<td>5.3</td>
</tr>
<tr>
<td>Total</td>
<td>12</td>
<td>8</td>
<td>20</td>
<td></td>
</tr>
</tbody>
</table>

**S-56**

Clinical Characteristics and Management of Intractable Constipation Combined with Megacolon after Congenital Anorectal Malformation Surgery

Tingchong Zhang1

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**Purpose** This study summarizes the clinical characteristics and treatment efficacy of intractable constipation combined with megacolon after anoaplasty.
Methods Retrospectively analyze the clinical data of 23 cases (10 boys, 13 girls) with intractable constipation combined with megacolon. All 23 patients were treated for constipation after anoplasty. Results Air contrast barium enema showed expansion of the lower colon and rectum, no transition zone, and a delay barium discharge. Soave megacolon radical surgery was used for all patients. During operation, expansion of the intestine, hyperplasia and thickening of the mesentery, and hyperplasia and enlargement of intestinal wall blood vessels were seen immediately on entering the pelvic cavity. Pathological examination showed that 15 cases with resection of the distal segment of intestine had visible ganglion cells, whereas 8 cases did not have; all 23 cases had muscular layer hyperplasia and thickening, myofibrosis, and disorderly arrangement of the muscular layer. Follow-up anus function grading was 5-6 by LiZheng’s scoring system after the second surgery. Conclusion Intractable constipation after anoplasty was often complicated by megacolon. Imaging showed localized intestinal expansion. Surgical treatment to resect pathologically changed intestine combined with necessary repair of anatomical anus defects can achieve excellent postoperative results.

Clean Bum, Happy Mum: The Use of Transanal Irrigation in Children
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Aim: The aim of the study was to assess the treatment efficacy of transanal irrigation (Peristeen) and parental satisfaction in children with anorectal malformation, Spina Bifida, Hirschsprung’s disease, and intractable functional constipation. Methods: A comprehensive literature review was conducted in order to determine the current standards in the bowel management of this patient cohort. A retrospective review of children attending our service was conducted, over 5 years (2012-2017), to determine the numbers using transanal irrigation. A survey was conducted to determine compliance rates, efficacy, and overall parental satisfaction. Efficacy was assessed using the following parameters: soiling, use of pads, impact on daily activities.

Results and Conclusion: The total number of patients using transanal irrigation was 102: anorectal malformation (16), Spina Bifida (54), Hirschsprung’s disease (3), and intractable functional constipation (29). Overall compliance rates were found to be good. Efficacy rates were high amongst responders and this was reflected positively in regards to parental satisfaction. Further results to follow.

Dissynergic Defecation - The Cause of Fecal Incontinence in Children after Surgery
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Fecal incontinence in children has negative impact on quality of life. Usually patient with soiling follows different specialists: pediatricians, neurologists and pediatric surgeons-coloproctologists. Dissynergic defecation (DD, F3b, Rome IV) is functional defecation disorder which is characterized outlet dysfunction (contraction/bad relaxation of the anal sphincters during defecation).

Aim of the Study. To determine the frequency of patients with fecal incontinence and to identify the frequency of dissynergic defecation in this children.

Methods. Patients with fecal incontinence from pediatric coloproctologist reception were investigated. Medical examination, rectal digital investigation, anorectal manometry, defecography were used.

Main results. On 2016-2017 there were 165 children on pediatric coloproctologist admission (single specialist, reception day once a week). 70 children (42.4%) presented with fecal incontinence. Almost all of patients underwent digital rectal examination (except 3 children with negative reaction). There were 55 patients (33.3%) from 3 for 17 years underwent anorectal manometry (ARM), continuous and stationary pull-through technique. ARM used in patients with Hirschsprung's
disease (HD) and anorectal malformations after surgery (22; 40%) and in non-surgical children with defecation disorders (33; 60%). 6 out of 14 patients (42.9%) with anorectal malformations demonstrated DD and 3 out of 8 (37.5%) with HD. Almost all non-surgical patients with fecal incontinence demonstrated DD (30 of 33; 90.1%).

Conclusions. About half of children on the coloproctologist counseling had fecal incontinence. DD (paradoxical anal contraction during defecation) the most common reason of fecal incontinence in non-operated children. Frequency of DD in children with anorectal malformations and HD is 42.9% and 37.5% respectively.

S-59

Institutional Review of the Patients Operated for Vesicoureteral Reflux after Correction of the High Type Anorectal Malformations
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Preface: Repeated urinary tract infections (UTI) by vesicoureteral reflux (VUR) are the most important problem in patients with anorectal malformation (ARM) accompanying urinary tract disorders. To avoid renal demise ending in transplantations is the top priority in these patients’ management. In our institute, the patients after the definite surgery of ARM with urological cases, are managed by the initial preventive antibiotics administrations and following surgical therapy in the refractory cases. But the significance and efficacy of this strategy are not clear.

Aim of the Study: To assess the efficacy of our strategy by overviewing our patients’ background.

Methods: Retrospective study on the medical records of the 9 patients with high type ARM (with urinary fistula) treated from 2012 to 2018.

Results: The 5 patients of 9 had VUR treated by initial medication. The 3 of the 5 needed Cohen’s operations for uncontrollable UTI at the age of 2 4 11 year-old respectively. Clean Intermittent Catheterization had been already started preoperatively in one case. All of kidneys were unilateral with VUR and ectopic ureter or ectopic opening. Spinal lesions were seen in all. And postoperatively, UTI had stopped in 2 cases and seldom seen in one.

Conclusions: Cohen’s operations for VUR in complicated ARM were effective, but not promising complete effect. That may depend on their impaired bladder function by their spinal lesions. Not only to cease UTI, but also reduce bowel bacterial overgrowth by defecation management in corporation with pediatric urologists and surgeons may result in good prognosis.

S-60

How Much Do We Know about Constipation after Surgery for Anorectal Malformation?
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Purpose To evaluate the occurrence of constipation after anorectal malformations(ARM) repair and the results of laxative treatment.

Methods Between August 2012 and July 2017, the clinical data of patients with ARMs was prospectively collected. The patients were divided into two groups, good types and poor types. Good types included rectoperineal, rectovestibular, rectourethral bulbar, and no fistula. Risk factor were defined as Spinal cord anomalies, sacral ratio <0.4, or cognitive impairment. Success was defined as that laxative could be tapered or weaning off.

Results Eighty-four patients were enrolled with mean age of 6.3±7.8(0.6~59.9) years. The onset of constipation was at mean age of 12.8±7.8(0.6~59.9) months. The onset of constipation was at mean age of 12.8±7.8(0.6~59.9) months. Of 27 patients treated with senna-based laxatives, mean age was 2.6±2.2(0.8~9.4) years with mean duration of 14.8±12.2 months. In 23 patients followed >6 months, 14 of 18 (77.8%) patients with good types were classified as success, whereas only 1 of 5 (20%) patients with poor types was (p=0.02). In patients with good types, 9 of 9 (100%) patients with no RF were successful, however, only 5 out of 9 (55.6%) patients with RF were (p=0.02).

Conclusions Constipation occurs shortly after operations. Patients with good types and no RF are susceptible to weaning laxatives.
S-61
Modified Anterior Sagittal Anorectoplasty for Vestibular and Perineal Fistula
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Objective To explore the feasibility of modified anterior sagittal anorectoplasty with preserving partial perineal body.
Methods Retrospectively analysis the perioperative situation and follow-up of 30 girls with modified procedure for perineal fistula or vestibule fistula, and compared with 30 traditional perineal anoplasty with the traditional sagittal approach.
Results There was no significant difference between the 2 groups of operation time and the incidence of fistula and vagina (P>0.05); there was a significant difference in scar and appearance satisfaction between the two group and the difference was statistically significant (P< 0.05). At the first, third and sixth month postoperatively showed that the anal canal resting pressure in the improved operation group was slightly higher than that of the control group, but the difference was not statistically significant (P>0.05), and there was no statistical significance in the incidence of fecal pollution between the 2 groups (P>0.05).
Conclusion The modified Sagittal anorectoplasty with preserving partial perineal body is safe and feasible. On the basis of the traditional sagittal approach, the original perineal body is preserved to the maximum, the appearance is more satisfactory and the clinical feasibility is good.

S-62
Varied Facets of Rectal Duplications
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Background: Rectal duplications are rare anomalies comprising of less than 5% of all alimentary duplications.
Methods: Cases of rectal duplications managed from 1998 till 2018 were studied for their presentation, management and outcome.
Results: Eleven cases of rectal duplication were studied. The age at presentation varied from newborn to 60 years age (median 9 months). Four were females. One had vaginal duplication. One was an old lady who presented with obturator hernia. Five patients had constipation and a retrorectal mass. The diagnosis was incidental during PSARP for ARM in 2. A newborn presented with an incomplete tubular structure about 3 cm long, situated in midline just anterior to the anal verge. A 6 months old male baby presented with and a cystic mass bulging from the anal verge. A twelve-year-old boy presented with perineal hypospadias, anorectal anomaly and a fistulous opening in the perineum with a 4 cm long tract anterior to the rectum ending in a blind tubular structure. The cyst was excised in 10 patients. It was left as such in one patient for fear of incontinence. All patients were continent and retained their continence post surgery.
Conclusion: Rectal duplications are rare anomalies with varied presentations. A high suspicion is vital. A cyst of variable shape and often distended with mucous, lying in the presacral space forms the diagnosis A single stage excision of the cyst, usually by trans-anal, perineal or the posterior sagittal route is curative in most cases.

S-63
Combination Therapy with Traditional Medicine Formulations for Perianal Abscess in Children
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Background: It has been reported that combination therapy with two different traditional medicine formulations called Hainosankyuto and Juzentaihoto in Japan may be effective for perianal abscess (PA), but their actual effectiveness has not been established. The aim of our study was to investigate the effectiveness of combination therapy with Hainosankyuto and Juzentaihoto together as the most effective conservative treatment for PA.
Methods: We identified 69 patients with PA under 2 years old and divided them into 4 groups according to the formulations administered; group 1: Hainosankyuto (n=17); group 2: Juzentaihoto (n=14); group 3: Hainosankyuto and Juzentaihoto (n=19); and group 4: placebo (n=19). Age at presentation, duration of purulent discharge (DP), frequency of surgical intervention (incision and drainage (ID); fistulotomy/fistulectomy), and recurrence rates were analyzed statistically.
Results: We show the outline of result in figure. Mean ages (months) were 8.6±9.2, 6.9±7.3, 5.2±4.7, and 3.8±3.1 in groups 1, 2, 3, and 4, respectively. Age at presentation, duration of purulent discharge (DP), frequency of surgical intervention (incision and drainage (ID); fistulotomy/fistulectomy), and recurrence rates were analyzed statistically. Results: We show the outline of result in figure. Mean ages (months) were 8.6±9.2, 6.9±7.3, 5.2±4.7, and 3.8±3.1 in groups 1, 2, 3, and 4, respectively. Mean DP (weeks) was 2.5±2.2, 7.1±10.8, 2.0±0.0, and 2.7±1.0, respectively. DP was significantly longer in group 2 compared with groups 1 and 3 (p<0.05). Mean ID were 1.0±0.2, 2.3±0.5, 0, and 1.6±0.2, respectively. Group 1 had significantly less ID than group 2 (p<0.01). Recurrence rates were 6%, 36%, 0%, 32%, respectively. Groups 1 and 3 had significantly less recurrences than group 2 (p<0.05) and group 3 had significantly less recurrences than group 4 (p<0.01).
Conclusions: Combination therapy with Hainosankyuto and Juzentaihoto decreased recurrences and surgical intervention to zero in this study, demonstrating how highly effective it is for treating PA in children.
S-64

Is it Safe to Perform Laparoscopic-assisted Endorectal Pullthrough Earlier for Hirschsprung’s Disease? A 16-Year Retrospective Review in a Single Institution

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Aim of the Study
Laparoscopic-assisted endorectal pullthrough (LAPT) for Hirschsprung’s disease (HD) has been well-established. This study aims to investigate the safeness of performing LAPT in the neonatal period.

Methods
A 16-year retrospective review of all HD patients undergoing single-staged LAPT from May 2002 to May 2018 in a single tertiary institution was conducted. Safeness of surgery within 30 days of life (Group A) was compared to that beyond (Group B). The outcome was assessed in terms of operation duration, intraoperative and postoperative complications and postoperative length of stay. Results were analysed statistically using Chi-square test and t-test.

Main Results
Eighty-one patients received LAPT with 25 in Group A (median age 21 days) and 56 in Group B (median age 98.5 days). There was no difference in gender and disease involvement. Preoperatively, no patients in Group A had enterocolitis but 16 in Group B (p = 0.0018). The median postoperative stay was similar (9 days versus 8 days; p = 0.14). There was no difference in operative time and intraoperative complications. There was 1 mortality secondary to sepsis in Group A (p = 0.31). Eight patients in Group B developed postoperative complications including pneumonia, intestinal obstruction, intra-abdominal abscess, anastomotic leakage and transition pullthrough (p = 0.09). There was no difference in development of postoperative enterocolitis (19.2% versus 19.6%; p = 0.49).

Conclusions
It is safe to perform LAPT for Hirschsprung’s disease in the neonatal period and may minimize preoperative enterocolitis.

S-65

Treatment of Hirschsprung’s Disease through Laparoscopic-assisted Soave Pull-through Procedure

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Objective: To observe the effect of Laparoscopic-assisted Soave pull-through procedure.

Method: The observation was conducted on 360 cases from January to December in 2006. 250 cases were in the control group, treated with laparoscopic-assisted treatment, while 110 cases in control group, treated with traditional Soave pull-through procedure. Compared variables were operation time, amount of blood loss, anal dissection time, recovery time for gastrointestinal function and length of stay. One Month after the procedure, the patients’ anal function was evaluated according to the Anal Function Standard Evaluation set by China Medical University (PRC), and patients’ quality of life were evaluated according to The Short Form (36) Health Survey (SF-36).
**Result:** In the observation group, the amount of blood loss, anal dissection time, gastrointestinal function recovery time and length of stay were all significantly lower than the control group (p-Value < 0.05). Anal function evaluations of observation group were significantly higher than control group (p-Value < 0.05). Patients in observation group had less postoperative, with higher physical function, emotional function, social function, role function and quality of life (p-Value < 0.05).

**Conclusion:** Laparoscopic-assisted Soave pull-through procedure is better than traditional Soave procedure, with the benefits such as less blood loss and less postoperative complications.

**S-66**
**The Operative Results for Consecutive 200 Cases of Hirschsprung’s Disease with the Focus on Complications in a Single Institution Experiences**
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**Aim of the Study:** Hirschsprung’s disease is considered to be curable when performing definitive surgery. The operative procedure is changing with the times. We analysed operative results and redo surgery in our institution.

**Methods:** Patient data were collected over half century. From 1963 to 2014, 200 patients with Hirschsprung’s disease underwent definitive diagnosis at Kyushu University Hospital. The operative data with these patients were collected and analysed retrospectively.

**Main Results:** Extent of aganglionosis is as follows: short segment (ultra-short and recto-sigmoid):175 (87.5%), long segment: 12 (6.0%) total colon aganglionosis with or without small intestine involvement: 13 (6.5%). As a result, 184 (92.0%) of 200 those patients underwent definitive operation. Totally 195 radical operation including primary and redo were performed for these 184 patients. Breakdown of the primary procedures as follows: Z-shaped anastomosis (Modified Duhamel, Figure1):132, laparoscopy assisted Transanal endorectal pull through (TAEPT):22, Duhamel: 14, Lynn: 8, Soave: 2, Swenson: 1, Rebein: 1, Martin: 1. From 1963 to 1997, main procedure is Z-shaped anastomosis. Since 1998, main procedure is Lap-assisted-TAEPT. Totally 11 Redo surgery were performed. Procedures for redo was as follows: Z-shaped anastomosis after Lynn: 3, Z-shaped anastomosis after Duhamel: 2, Z-shaped anastomosis after Swenson: 1, Swenson after Duhamel: 1, Lynn after Z-shaped anastomosis: 1, and Z-shaped anastomosis after TAEPT: 3.

**Conclusions:** The authors analysed 200 patients over half century in single institution. A primary operation without laparotomy has thus become the procedure of choice for a definitive operation. Open Z-shaped anastomosis was mainly performed as redo procedure.

**S-67**
**Which Is Better for Classical Hirschsprung’s Disease, Rectoplasty with Posterior Triangular Colonic Flap or Transanal Endorectal Pull-through with Rectoanal Myotomy?**
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We employ rectoplasty with a posterior triangular colonic flap (RPTCF) for classical Hirschsprung’s disease. Recently we employed a modified Soave procedure: transanal endorectal pull-through with rectoanal myotomy (TEPTRAM). In both, the internal sphincter muscle is completely divided vertically at the 6 o'clock position. Unlike RPTCF, TEPTRAM does not require abdominal manipulation, is minimally invasive, and highly esthetic. We aimed to verify the usefulness of TEPTRAM. Sixty-four patients with classical Hirschsprung’s disease who underwent surgery between 1970 and 2017 and had a postoperative follow-up period of 6 months or more were divided into 47 cases of RPTCF (group R) and 17 cases of TEPTRAM (group T). We compared the defecation function of the groups. The mean age at operation was 754 and 118 days in groups R and T, respectively (P<0.01). The mean postoperative follow-up period was 7.8 years and 5.7 years in groups R and T, respectively (P=0.08). Tree patients in group R (6%) and four in group T (24%) developed postoperative enterocolitis (P=0.16). No patient above 4 years showed fecal incontinence (R:0/41, T:0/10, ns). Three patients in group R (10%) and one in group T (20%) needed an enema (P=0.36). As there was no incontinence in either procedure, there was no adverse effect of rectoanal myotomy. Although there is no significant difference, the incidences of enterocolitis and constipation were slightly higher in group T. It may be because of the remaining muscle cuff, so it is necessary to provide the best care with attention to constipation soon after surgery.

**Figure 1. RPTCF procedure**
A) The posterior wall of the rectum is split in the midline. The colon is split along its taenia libera. And the anastomosed is performed between them. B) The incision of the posterior wall of the rectum is extended through the dentate line. The triangular colonic flap is pulled down through the rectum and the colorectal anastomosis is performed. C) Completion of colorectal anastomosis.

**Figure 2. Rectoanal myotomy in TEPTRAM**
The internal sphincter muscle is completely divided vertically at the 6 o'clock position.
**S-68**

**How Best to Expose the Entire Surgical Anal Canal in the Operative Field during Transanal Pull-through for Hirschsprung’s Disease. A Crucial Step That Determines Success**

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**Aim:** During transanal pull-through (TAPL) for Hirschspring’s disease (HD), exposing the entire surgical anal canal (SAC) including the squamo-columnar junction, or anorectal line (ARL) is a crucial step for minimizing problematic postoperative bowel function. We present a hint for exposing the entire SAC.

**Method:** Histologically, the ARL represents the junction of proximal unilayer columnar colorectal mucosa with distal stratified squamous epithelium and is the proximal limit of the SAC. It is an obvious landmark; proximal mucosa is vivid pink and distal mucosa is more whitish. We use the Lone Star (LS) self-retaining retractor system to expose the ARL. Before we attach the LS-hooks to the anal sinuses on the dentate line full circle, we place 3/0 sutures at 0, 3, 6, and 9 o’clock around the anus to expose the anal sinuses (Figures 1A and 2). If a patient’s buttocks cannot be positioned as described or the patient is too high on the table, the LS ring doesn’t sit well, resulting only in dilatation and lengthening of the SAC without prolapse (Figure 1B). By hanging the patient’s buttocks 5cm over the end of the table, the LS ring sits snugly and the ARL and entire SAC prolapses to the anal verge (Figures 1C and 2).

**Results:** Good positioning, as described, greatly facilitated dissection in 61/68 TAPL cases, while poor exposure hindered treatment in 7/68.

**Conclusion:** Thorough exposure of the entire SAC, which is crucial for adequate TAPL, is greatly facilitated by patient positioning.

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**S-69**

**Straight Ileo-rectal Anastomosis in Treatment of Total Forms of Hirschsprung’s Disease**

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**Background:** The purpose of the study was to discuss the management of a patient with a total colonic aganglionosis and to present individual cases of straight ileo-rectal anastomosis (SIRA) for this pathology and to assess outcomes of its shaping.

**Material/Methods:** In this study we evaluated postoperative complications and long-term functional outcome of 8 patients with total colonic aganglionosis treated by SIRA in our departments during last 10 years. Among of 8 patients, who included into the study, were 5 males and 3 females. Patients were in age from 6 to 9 month old at the moment of SIRS formation. All patients had proctocolectomy with ileostomy as the first stage of surgical treatment. SIRA have been performed in all patients with preservation of preoperative ileostomy with «protective» purpose.

**Results:** Not one patient had preoperative enterocolitis before SIRS. Diarrhea was preserved in all patients during 4 - 6 month after surgical treatment. 2 patients suffered from postoperative enterocolitis which required temporary treatment with metronidazol with positive clinical effect. Complete anal continence was restored in 7 patients near 1 year after operation. One patient required in continuous medical therapy and bowel management more than 1 year. One patient had complication of ileostoma (necrosis), but others didn’t have any surgical postoperative complications.

**Conclusions:** SIRA with a «protective» ileostomy is a simple and successful procedure with good long-term functional outcome for patients with total colonic aganglionosis. Results of surgical treatment patients with SIRA are good in majority children and complications not systemic.
Quality of Life (QOL) in School-aged Children Who Underwent Bowel Management Program for Fecal Incontinence

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Aim of Study: Fecal incontinence is one of the most annoying and emotionally stressful complications in children. The aim of this study is to evaluate the quality of life (QOL) in school-aged children who underwent Bowel Management Program because of fecal incontinence.

Methods: Children with fecal incontinence who were referred to our colorectal follow up center and underwent BMP since May 2017 were included. The quality of life was evaluated by the children form of assessment PedsQL4.0. The quality of life was evaluated in the aspects of physical function, emotional aspect, social aspect, and school function, pre and post use of BMP. The assessment’s validity and cronbach’s alpha are 0.84 and 0.82 respectively.

Main Results: A total of 11 children with fecal incontinence underwent BMP. Five were male and the mean age was 8.5±1.77 years. The quality of life was in moderate level in both pre and post BMP and there was no significant difference between them (P=0.06). The physical function, emotional and social aspects were improved by the use of BMP (P<0.05). No significant difference was seen in pre and post educational function (P=0.06). School performance was lower than normal in patients. Nine mothers were satisfied by the use of BMP.

Conclusions: Fecal incontinence disturbs all aspects of quality of life in children and BMP can improve the total psychological aspects.

Cytomegalovirus and Human Herpesvirus 6 in the Development of Acute Appendicitis in Children

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Aim: Acute appendicitis has been associated with viral infections. These observations were mostly done in studies focusing on immunocompromised patients without using controls. We studied the association of cytomegalovirus (CMV) and human herpesvirus 6 (HHV-6) with acute appendicitis in immunocompetent children.

Methods: In this prospective study we included 79 children (5-18 years old) suspected of appendicitis. Acute appendicitis was diagnosed in 25 patients. The remaining 54 patients served as controls. Real-time PCR for CMV and HHV-6 on whole blood was performed in 79 children, while serology was performed in 74 children. Also real-time PCR on appendix tissue was performed in 29 children who underwent an appendectomy.

Results: CMV IgG antibodies were present in 20% of patients with appendicitis compared to 29% of control patients (P=0.28). CMV DNA was not detected in whole blood or appendix tissue of any patient. All patients with appendicitis and 98% of patients without appendicitis had HHV-6 IgG antibodies. In 21 out of 25 patients with appendicitis we detected HHV-6 in their appendices compared to two out of four patients without appendicitis (P=0.29). In addition, HHV-6 DNA was detected in all appendices of patients with perforated appendicitis compared to 79% of patients with non-perforated appendicitis (P=0.54).

Conclusions: In our study population of immunocompetent children, no association between CMV and acute appendicitis was found. Nevertheless, due to low CMV seroprevalence, a possible relation cannot be ruled out. No association between HHV-6 and acute appendicitis was found, since HHV-6 DNA was detected in patients with and without appendicitis.

<table>
<thead>
<tr>
<th>CMV</th>
<th>Appendicitis N=25</th>
<th>No appendicitis N=54</th>
<th>P value</th>
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<tr>
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<td>0/25</td>
<td>0/54</td>
<td>0.99</td>
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<tr>
<td>IgM+</td>
<td>0/25</td>
<td>1/51</td>
<td>0.68</td>
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<tr>
<td>IgG+</td>
<td>5/25</td>
<td>15/51</td>
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<table>
<thead>
<tr>
<th>HHV-6</th>
<th>Appendicitis N=25</th>
<th>No appendicitis N=54</th>
<th>P value</th>
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<td>Blood DNA+</td>
<td>0/25</td>
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<tr>
<td>IgG+</td>
<td>24/24*</td>
<td>52/53</td>
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CMV and HHV-6 detection in whole blood and appendix tissue of patients with and without appendicitis.
* Missing data
S-72
Risk Stratification for Surgery in NEC
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Aim: To compare the biochemical and haematological response of neonates with NEC (Bell stage 2): those who respond to medical management and those who require surgery. To form a risk stratification score to identify neonates who will need surgery.

Methods: Three year retrospective study including all neonates with NEC. Platelet count and C-reactive protein (CRP) values were collected from the day of diagnosis. Statistical analysis was performed using the Mann-Whitney U test.

Results: Total of 73 patients. Medical NEC group (42 patients): median gestational age 28+5 weeks, median birthweight 802g. Surgical NEC group (31 patients): median gestational age 27+3 weeks, median birthweight 1010g. The median CRP and platelet count was significantly reduced in the group requiring surgery throughout the period of treatment. This was most significant on day 3 with p<0.0001: Medical group - median 193 (IQR 117-274) and Surgical group - median 44 (IQR 30-62).

Conclusion: This stratification score provides a bedside formula that can identify neonates with NEC for surgery early. A prospective study is in progress to validate this tool.

S-73
Postoperative Intussusception in Children: A Report of 51 Cases
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Objective To summarize clinical features, risk factors, and anatomic patterns of pediatric postoperative intussusception (POI) in past 37 years.

Methods Clinical data of 51 POI cases were retrospectively reviewed. Variables analyzed included patient demographics, time of occurrence, type of intussusception, and surgical procedure.

Results Among 51 POI patients, 33 of them were younger than 2 years of age, 46 cases occurred in first week, 49 cases occurred in 2 weeks after initial surgery, and 17 cases underwent retroperitoneal operation. As expected, ileoileal and jejunojejunal intussusceptions were the most common forms of POI. Manual reduction was successful in 46 cases. All 51 patients recovered after the second operation.

Conclusions Although rare, postoperative intussusception is a serious condition after abdominal procedures in infants and children. Most occur in 1 week after primary surgery, small bowel intussusception is the predominant variant of this complication. Early use of ultrasonography can confirm the diagnosis of POI. Prompt laparotomy should be considered to avoid intestinal ischemia and consequential necrosis, though the obstruction can be relieved by manual reduction in common condition.

S-74
Pseudomonas Necrotizing Proctitis in an Infant: A Case Report
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Necrotizing proctitis is a rare condition in infants with few reported cases in literature. Necrotizing fasciitis is a rapidly progressing infection presented as a severe inflammation of the fascia and soft tissue. The disease is characterized with necrosis and gangrene of the inflamed tissue. If not timely diagnosed and treated, it poses with systemic toxicity and carries significant mortality. Pseudomonas aeruginosa causing necrotizing proctitis is an exceptionally uncommon condition. We are reporting a case of a previously healthy five-month-old male presenting with necrotizing proctitis and isolates Pseudomonas aeruginosa from the anorectal tissue. The patient was admitted due to fever and absence seizure.

Two days after admission, the anus and perianal region were noted to be swollen, indurated, and with areas of ulceration and necrosis. Examination under anesthesia and rigid proctosigmoidoscopy were done. There were necrotic perianal tissues at eleven o'clock to three o'clock and at five o'clock to seven o'clock positions. There was an extension of perianal necrotic tissue immediately below the dentate line. More proximally, the mucosa looked normal. Perianal necrectomy and diverting colostomy were performed. Histopathology confirmed the presence of necrosis. The tissue culture grew Pseudomonas aeruginosa, which was sensitive to Ceftazidime and Amikacin. The patient was sent home improved. The perianal region healed rapidly well and regular anal dilatations were done. Four months post-operatively, take down of colostomy was performed. To our knowledge, this is the first reported case of a necrotizing proctitis in our country's literature.
POSTER WALK I

P-1
A Novel Stopgain Mutation of EDNRB Associated with Familial Hirschsprung’s Disease
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Hirschsprung’s disease is a genetic complex disease with 3.6% to 7.8% familial aggregation. We collected five subjects from a three generation family with three members with Hirschsprung’s disease. Under the autosomal dominant with de novo mutation model, a filtration by pathogenic prediction led to a short list of 10 variants. Among the 10 variants, the gene, EDNRB, of a stopgain mutation is highly prioritized by knowledge-based analyses. The functional effect of the truncated EDNRB was further investigated through zebra fish model generated by crisper-cas9, the mutant carrier showed an obvious slower colon peristalsis comparing with normal controls. This finding highlighted the importance of EDNRB among familial cases of Hirschsprung disease, which may further help to digest the etiology of this disease.

P-2
Results of Kimura’s Procedure (Right Colon Patch) for Extensive Aganglionosis
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[Aim of the Study] Extensive aganglionosis of Hirschsprung’s disease with a long aganglionic segment has an aspect of short bowel syndrome. Its treatment is difficult, and many patients cannot be weaned from total parenteral nutrition (TPN). At our institution, 6 patients have been treated by Kimura’s procedure with favorable outcomes.

[Methods] Six patients with extensive aganglionosis have been treated by Kimura’s procedure before Duhamel-Ikeda Z-shaped anastomosis was performed for definitive treatment. Kimura’s procedure was performed 19 days to 7 months after birth (median: 106 days), and the body weight at surgery was 2,995-7,855 g (median: 4,795 g).

[Results] The fecal volume decreased to about half after surgery, the stoma could be closed after radical operation in 4 patients, one patient is presently awaiting radical operation, and another patient had a short bowel syndrome (residual small intestine: 38 cm) and has not been weaned from TPN because the fecal volume expands when oral feeding is increased even after Kimura’s procedure.

[Conclusion] In patients with a long aganglionic segment, long-term management by TPN is required as they cannot be weaned from TPN due to the short residual small intestine, and their management is made difficult by repeated episodes of liver disorder and catheter sepsis. At our institution, the terminal ileum is also added to the patch in expectation of nutritional absorption by the terminal ileum. Kimura’s procedure has resulted in a decrease in the fecal volume and normalization of water and electrolyte abnormalities with an improved QOL.

P-3
What is the Most Common Complication after One-stage Transanal Pull-through in Infants with Hirschsprung’s Disease (HD)?
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AIM : HD is a congenital disease that could be suspected by clinical symptoms, abdominal plain X-ray, and diagnose by rectal biopsy. In 80% of cases rectosigmoid junction is involved. One stage transanal pull-through has been popular recently and may have several complications and benefits.

Methods : From 2006 to 2016, 108 infants (37girls;71 boys) with mean age 8 days (3 to 33days) that clinically suspected to HD was admitted in our center. HD proved by rectal biopsy and Swenson-like procedure was performed Transanal and Nelaton tube (12 F) was inserted in the pelvis transprineal for drainage blood or collection. From Feb 2008 in 60 cases prophylactic Hegar dilatation was performed 2 weeks after operation.

Results: Anal stricture was seen in 16 cases of all (14.8%) and (33.3%) of 48 remaining cases that this problem treated by anal dilatation in 11 cases and 5cases corrected by surgical management. We had entrocolitis in 8 cases(5.4%) that treated by medical management. In 3 cases (2.7%) we had retrocolic abscess that spontaneous drainage was done via tube drain. We did not have any anastomotic stricture after starting prophylactic anal bougination.

Conclusion: OSTAPT has many advantages , low complications and the results are excellent. It seems the most common complication is anastomotic stricture that response well to prophylactic bougination. We recommend prophylactic anal bougination with Hegar probe at about 2 weeks after operation. The results of this operation need to evaluate more in long-term.Key word: OSTAPT( one stage transanal pull through)
**Total Colectomy and Ileorectal Anastomosis with Anorectal Myotomy: A New Procedure for Treatment of Total Colonic Aganglionosis and Gastrointestinal Dysmotility**

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**Aim:** Total colonic aganglionosis present in 4-5% of cases in Hirschsprung’s disease with high surgical mortality between 13 to 23%. Many techniques have been established but we have performed State pull-through as ileo-proctostomy with long posterior myotomy in total colonic aganglionosis and sever dysmotility disorders.

**Methods:** We had 16 cases, 12 total colonic aganglionosis, 1 intestinal neuronal dysplasia (IND) and 3 chronic intestinal pseudo obstruction syndrome (CIP) from 1992 to 2016 underwent total colectomy and resection of involved small intestine. Ileorectal anastomosis with long posterior rectal myotomy was done. All patients had barium enema and rectal biopsy. Leveling ileostomy was done in 15 cases and one case had distal jejunostomy.

**Results:** We had 13 female patients and 3 males at the age of 6 months to 5 years. Rectal biopsy of 12 Patients reported no ganglion cell, one had IND and 3 had ganglionic bowel with clinical presentation of CIP. Follow up time was 6 months to 10 years. In 3 cases, 2 weeks after initial operation myotomy from anus was performed. There were no significant complications in this group except episodes of diarrhea. Now, most of our patients are above the age of toilet training that they have voluntary bowel movement with necessity to little or no medications.

**Conclusion:** State pull-through is recommend in all cases of total colonic aganglionosis and severe dysmotility problems of colon. This technique is less difficult to perform, and avoids the complications and disadvantages of removal of the rectum and has satisfactory results.

**Evaluation of Early Outcome of One Stage Duhamel Operation without Total Mechanical Bowel Prep in Children with Hirschprung’s Disease**

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**Aim of Study:** evaluation of the early outcome of one stage Duhamel Operation without total bowel preparation in children with Hirschprung’s disease.

**Methods:** In a Case series study in 2015-2016 children consecutively admitted for elective surgery of Hirschprung’s disease were selected. Exclusion criteria were: neonates, episodes of enterocolitis, previous abdominal surgery for Hirschprung’s disease and infants with leveling colostomy. Chemical bowel prep and one or two sessions of rectal wash out by normal saline were performed in the day before the operation. Intra operatively, suspicious segment of transitional zone was resected by staplers without any fecal contamination. Intraoperative bowel prep of distal segment was done. Pull through was performed by Duhamel technique with one stapler. Post operatively, early complications were registered. In January 2015 parents were recalled and defecation assessment questionnaire were filled.

**Main Results:** 20 children were evaluated (13 boys, 7 girls). The mean age of patients was 13.28 ± 1.32 months. There was no major postoperative complication in our patients. Defecation frequency was 1-2 /day in 11 cases, 3-5 /day in 6 cases and numerous in 3 cases. Feces consistency was solid in 17 cases and Semisolid in 3 cases. Defecation control was normal in all over three years old children.

**Conclusions:** Our findings suggest that one stage Duhamel procedure without total bowel prep with one stapler is an acceptable approach for operation of Hirschprung’s disease in children.

**Hemorrhoids and Other Vascular Malformations of the Rectum in Children, the Algorithm for Diagnosis and Treatment**

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**Purpose:** Improvement of the diagnosis and treatment of hemorrhoids and other vascular malformations of the rectum in children.

**Methods:** For the period from 2013 to 2018, the retrospective study and current study of hemorrhoids and other vascular malformations of the rectum in 68 children excluded from randomized 136 children which suffering from anorectal bleeding and anal protrusion in our hospital. Data on patients’ demographics, incidence, and types, medical and surgical treatment were analyzed.

**Results:** The age of the patients was 3 months to 17 years 11 months. The incidence rates of the vascular pathology of perineum and rectum (hemorrhoids, various angiodyplasia of the colon and combined pathology - anorectal malformations and angioedema of the perineum and rectum) and non-vascular pathology were 50% (41.18%, 04.41%, and 04.41%) and 50% respectively. In the multivariate analyses, male sex was prominent in hemorrhoids, the only female was found in angiodyplasia, minimal age was 4 year in patients with hemorrhoids, depending on the pathology - their treatment and results are different.

**Conclusion:** The hemorrhoids and vascular malformations are the rare cases in children and their researches are also little. In the last years, various forms of vascular malformations in the perineum and anus were often found. The algorithm for diagnosis and treatment of hemorrhoids and other vascular malformations of the rectum in children can be established.
One-stage Laparoscopy-assisted Endorectal Pull-through for Late Presented Hirschsprung’s Disease
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Introduction
Children with late-presenting Hirschsprung’s disease (HD) are classically treated by a staged operation with enterostomy. An alternative may be one-stage laparoscopy-assisted endorectal pull-through, which has cosmetic advantages. This study describes the outcomes of children with late-presenting HD who underwent this procedure.

Methods
From January 2010 to December 2017, we retrospectively reviewed the data of patients who underwent One-stage laparoscopy-assisted endorectal pull-through older than 3yrs.

Result
Seven older (>3 years) children (four males, three females) underwent one-stage laparoscopy-assisted endorectal pull-through. A study revealed their median age was 5.3 (range, 3.2-15) years. The transitional zone was rectosigmoid junction in 3 patients, and was sigmoid colon in 4 patients. For bowel preparation, five patients required rectal irrigation under general anesthesia. The median operating time was 264min. There were no intraoperative or early post-operative complications. Patients started a diet a median of 5 days after the operation and were discharged a median of 19 days. During the median follow-up period of 9.7 months, six (85.7%) had acquired voluntary bowel movements and 28.5% had grade 1 soiling. Two (28.5%) of the patients still had constipation. The constipation was manageable with diet or laxatives in two patients.

Conclusion
One-stage laparoscopy-assisted endorectal pull-through in children with late-presenting short segment HD is feasible and safe. Rectal irrigation under general anesthesia and the use of laparoscopy and a bipolar coagulator help to overcome the technical difficulties of this procedure.

Is Barium Enema Reliable for the Diagnosis of Total Colonic Aganglionosis?
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Introduction
Children with late-presenting Hirschsprung’s disease (HD) are classically treated by a staged operation with enterostomy. An alternative may be one-stage laparoscopy-assisted endorectal pull-through, which has cosmetic advantages. This study describes the outcomes of children with late-presenting HD who underwent this procedure Barium enema is one of the diagnostic modalities for Hirschsprung’s disease, especially total colonic aganglionosis (TCA). We retrospectively reviewed the medical records of all the patients who were diagnosed as having TCA and underwent a barium enema between January 1998 and December 2016 in single center. All the tests were performed and reviewed by pediatric radiologists. Among the total 19 patients with TCA who underwent barium enema, 9 patients (47.4%) had accurate radiographic results. Eight of the 13 neonate patients (61.5%) showed typical TCA radiological findings. However, only one of the 6 patients aged >4 weeks (16.7%) had accurate radiological diagnosis. Barium enema showed low accuracy for TCA, and its diagnostic performance was better in neonatal period than in those aged >4 weeks.
Assessment of Defecation Function Beyond Infantile Period for Transanal Single-stage Endorectal Pull-through in Hirschsprung’s Disease

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(Aim of the Study) Transanal single-stage endorectal pull-through procedure (TERPT) for Hirschsprung’s disease (HD) patients has been reported favorable outcomes with a lower complication rate. Nevertheless various degrees of bowel dysfunction and fecal continence could be persisted for long time in some patients. The aim of this study is to assess long-term outcomes of TERPT, performed in infantile period, after completion of toilet training.

(Method) We reviewed 83 patients aged ≥ 4 years who underwent TERPT in infantile period after pathologic diagnosis of HD at our center from 2001 to 2013, retrospectively. Functional outcomes were investigated according to answers of Bowel Function Score (BFS), a previously validated 7-item questionnaire about bowel habit.

(Main Results) Overall BFS were similar in all investigated age group. Comparing fecal soiling and social problems between HD and normal populations, it showed a lower score at early age in HD patients, however reached to similar level at 7-year-old. Regarding a stooling frequency, it had been decreased continuously, but was not significantly different.

(Conclusions) Functional outcomes of TERPT performed in infantile period, after completing toilet training are similar to that of normal population. In most of cases, uncomfortable symptoms had been diminished and functions improved with age. But in some patients, poor functional status had been persisted. Further studies would be needed.

Treatment and Outcome of Long-segment and Total Colonic Aganglionosis

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(Aim of the study) To assess the postoperative outcomes in patients with long-segment and total colonic aganglionosis(TCA).

Method: From 2008 to 2017, the clinical course and therapeutic results of 7 patients were retrospective reviewed: 4 with aganglionosis extends proximal to sigmoid(long-segment disease) and 3 with TCA. Results: In long-segment disease, 2 patients underwent a primary transanal endorectal pull-through(TERPT) and 2 underwent abdominal-assisted pullthrough. Two of TCA patients were treated by Martin procedure and 1 underwent two operations; the first was TERPT and the second was total colectomy with colo-ileal anastomosis. Residual aganglionosis was found to be the cause of re-operation. The average age at diagnosis and surgery were 44.85(13-150) and 68.8(75-180) days. The follow-up period was 7-81 months. Internal sphincter achalasia occurred in one patient which could be treated with anorectal myectomy. Five patients had postoperative enterocolitis and 3 in 5(60%) had associated anastomosis stricture which rapidly improved by routine dilatation. Perianal excoriation was detected in 5 patients since early postoperative period but improved with time corresponded to continuity decrease in stool frequency during the first six months after surgery. At 1 year follow-up, almost patients had acceptable bowel function with 2-4 stools per day. All patients above 4 years of age achieved a voluntary bowel movement without or occasionally need medication.
Conclusion: Abnormalities of bowel function and enterocolitis are common after definitive surgery for long-segment and TCA. The awareness of postoperative complication is necessary and will lead to prevention and early management.

P-11
Malone Antegrade Continence Enema in Treatment of Fecal Incontinence and Chronic Constipation --- A Single Center Experience
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Purpose To patients with incontinence or chronic constipation, Malone antegrade continence enema is an excellent option of treatment. We want to analysis the benefit, the change of life, and the complications to those who received the operation.

Method This study analyses a single medical center for patients with defecation incontinence or chronic constipation due to congenital or acquired diseases. A total of seven adults and five children received Malone surgery for the above reasons during 2010 to 2017.

Result Two of the five children were treated with Malone surgery for chronic constipation after Duhamel procedure for Hirschsprung’s disease. The other three were due to incontinence after the surgery of imperforate anus. Seven adults received the operation due to fecal incontinence secondary to traumatic spinal cord injury. All patients satisfied with the improvement of life after surgery. Two children resisted treatment because of pain during the insertion of the tube, and they became accustomed to it after one year of use. An adult had a stoma dislocation and re-operation one month after the procedure. Seven patient had over-growth granuloma around the stoma, which caused stoma obstruction, and received re-operation in average three months. Three of the seven patients even need to keep the tube at the stoma.

Conclusions For patients with incontinence, the biggest problem is the embarrassment caused by defecation odor, affecting interpersonal relationships, and even social exclusion. According to the result of our study, patients had a good improvement in their life after the surgery, and acceptable to complications.

P-12
Rectal Capillary Hemangioma: An Uncommon Cause of Rectal Bleeding in Children
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Introduction: Hemangioma of the rectosigmoid is less frequent vascular malformation. Most common clinical presentation is painless rectal bleeding, which can be acute or chronic. Most cases were cavernous subtype, but capillary subtype was rarely reported. Colonoscopy is the most important examination for confirming the diagnosis. The mainstay of treatment is surgical resection. Propranolol is benefit for treatment hemangioma but for rectal capillary hemangioma is less previously reported.

Aim: To report the case of capillary hemangioma at the rectum and its alternative treatment.

Method: Case report

Result: A 14 years old boy who presented with recurrent passage of the bright red blood after defecation for 1 year. The colonoscopy found circumferential mass with ulceration at rectum and pathological diagnosis was capillary hemangioma. He was treated with propranolol for 8 months and laxative drug for prevent constipation, after that he did not pass bloody of stool and no anemia. Follow up colonoscopy showed decrease in size of the rectal mass.

Conclusion: Capillary hemangioma of the rectum was rarely reported and the mainstay of treatment of rectosigmoid hemangioma is surgical resection, but the operation is difficult to perform and may have unpleasant complications. Propranolol is one of alternative treatment that can reduce clinical and lesions.
**P-13**

**Intestinal Plexiform Schwannoma: A Case Report**
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**Aim of the study:** Plexiform schwannoma (PS) is a benign tumour of the peripheral nerve sheath with Schwann cells arranged in a multinodular growth pattern (plexiform). It is typically found in the skin while visceral localization is exceptional. Reported cases of visceral PS are less than 15 in both adults and children. We herein discuss the case of a 17 year-old girl with mesenteric PS resected by video-assisted laparoscopy.

**Case report:** The patient came to our attention for abdominal pain, rectal bleeding, fever and back pain. She had always been well except for the last few months (recurrent abdominal pain imputed to constipation and treated with stool softeners). Laboratory tests were unremarkable. The abdominal US and MRI showed an irregular 3.5 cm abdominal mass in the epigastric region. The Tc-99m-labeled octreotide scintigraphy was negative. A laparoscopy was performed (one transumbilical lens and two operative trocars) showing a tough mesenteric mass of the first jejunal loops. The affected bowel was exteriorized through the umbilicus and the mass was resected together with the adjacent intestine (total length 10 cm). A termino-terminal anastomosis was performed at the end. There were no post-operative complications. Histology revealed PS. One year after surgery MRI was normal and the girl was asymptomatic.

**Conclusions:** Although benign schwannomas rarely occur in the gastrointestinal tract, we suggest that PS should be considered as differential diagnosis for any patients with an abdominal mass. Given the possible association with neurofibromatosis, the finding of PS should stimulate an examination for other manifestations of this disorder.

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**P-14**

**Toilet Training and Toilet Refusal Syndrome: A Cross _ Sectional Study in Iranian Children**
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**Aim of the study** Aim of this study was to evaluate the pattern of toilet training (TT) and toilet refusal syndrome (TRS) in Iranian children.

**Methods** In a cross _ sectional study mothers of 60 children attending colorectal follow up Clinic of Isfahan University Children hospital were directly interviewed. Questionnaire were filled out containing item on demographic data , the parent’s view , applied method and TRS rate, toilet phobia and the age at which TT was accomplished in children aged 2 to 14 years.

**Results** Mean initiation and completion ages were 19.83± 7.24 and 26.18± 8.96 months respectively. The duration of TT was 23.86± 29.53 weeks. Fifty eight percent who used of parents intensive approach and 39.2 percent who used child oriented approach. There was no assisted infant toilet training approach in our study. There was no correlation between the levels of education of mother with TRS. TF was seen in 3.3 percent of children. Punishment was used in 6% of children. There was no correlation between mother education and father applying punishment for training .

**Conclusions** The intensive method of training is more popular in our country. The age of initiation and completion is similar to other country. Child punishment should be addressed in non _ urban family.

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**P-15**

**Evaluation of Predicting Factors of Reducibility of Intussusceptions with Barium Enema in Children**
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**Aim of study:** The aim of this Study is evaluation of the predicting factors of reducibility with barium enema in children with intussusception.

**Methods:** In a Case series study in 2015 children consecutively admitted with intussusception diagnosis were selected. Exclusion criteria were: clinical symptoms of peritonitis, free air in plain x-ray and shock. Data such as age, sex, fever, leukocytosis, free fluid in sonography, time of symptom appearance and positive bloody stool were collected with checklist. Then barium enema was performed. The relationship between levels of success of treatment with check list data was analyzed.

**Main Results:** 45 children were evaluated (35 boys, 10 girls). The mean age of patients was 2.48 ± 1.27 years. There was a relationship between Positive blood stool and free fluid in sonography with unsuccessful barium enema treatment.

**Conclusions:** In attention to, the bloody stool and free fluid in sonography reduce successfulness of Barium enema. Our recommendation is to attempt not more than one session in barium enema if there are signs of positive blood stool and free fluid in sonography.
P-16

A Way of Aiding Surgeon-parents Communication: The Parents Associations
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Background 15 years ago we helped the parents form an association where they would get together and solve some of their common problems after the anorectal malformation surgery. As the population is small they had relatively few members and began accepting as members all the parents with children after any of the bowel disease procedures. Has this association been a success? Should we support it or should we improve the surgeons communication and information to the parents? Can the nursing staff that work with the surgeons be of any help?

Method All the members of the association were sent questionnaires about their hospital stay and their experience of the help they got from other members.

Results 96 questionnaires were sent. They returned 77% of the questionnaires posted with the following answers:

- The surgeon has explained the anomaly my child has excellently 72%, well 14%, below expectation 6% and 8% did not grade it but commented that the explanation was to technical and too much detail of the procedure but not enough of what happens later at home. The internet helped in 60% but left them more confused in 37%, only 3% did not consult the internet.
- The nursing staff prepared us for the necessary procedures at home excellently in 37%, well in 25%, adequately in 27%, inappropriately in 11%.

Conclusions The association contributes greatly to the parents feeling of wellbeing when they take care of their child after surgery for bowel disorders.

P-17

Single-port Laparoscopic Assisted Bianchi Surgery for Low Cryptorchidism Accompanied with Anomalies of Processus Vaginalis
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Objective To describe the value of single-port laparoscopic assisted Bianchi surgery in the treatment of low cryptorchidism accompanied with anomalies of processus vaginalis.

Methods We made a scrotal incision of the affected side to enter the tunica vaginalis. Then the tunia vaginalis was opened to expose the testis for understanding size of the testis and development of the spermatic cord vessels and the vas deferens. The processus vaginalis was cut to the high position for double ligation with silk thread.

Results The operation time was 20-45 min (mean, 39.9 min). The amount of bleeding was 1-2 ml. The children had mild pain, slight scrotal swelling, quick recovery and no need for antibiotics after operation. After 6 h, a liquid diet was started and the patients were discharged 2-3 days after operation without any incision infection. The 33 cases were followed up for 0.5-6 months (mean, 4.8 months). All the testes remained in satisfactory position, with good appearance, left and right scrotum symmetry, no testicular atrophy, testicular retraction, hydrocele, inguinal hernia or other complications.

Conclusions Single-port laparoscopic assisted Bianchi surgery for children with anomalies of processus vaginalis accompanied with low cryptorchidism has advantages of easy cryptorchidopexy and high ligation of hernial sac.

P-18

Air Test as a Simple Method of Screening for Hirschsprung’s Disease
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Background: The rectoanal inhibitory reflex (RAIR) is usually examined by anorectal manometry (ARM) to diagnose Hirschsprung’s disease (HD). We developed a novel technique called “air test,” which examines the presence of RAIR by the infusion of air into the rectum during contrast enema. The aim of this study was to present the technique and the diagnostic accuracy of the air test.

Methods: This study enrolled children consulted for chronic constipation (CC) between January 2012 and December 2016 for whom the air test was performed. The test was conducted during contrast enema under fluoroscopic observation using 20-50 ml injections of air into the rectum. The demographics, results of the air test and additional examinations, as well as the outcomes of subsequent treatments were analyzed retrospectively.

Results: The air test was conducted in 179 patients (median: 3 years, range: 0-14 years), and was positive in 150 and negative in 29 cases. Of the 29 patients with negative results, four were diagnosed with HD by rectal suction biopsy (RSB). Of the remaining 25 patients, RSB was conducted in seven and HD was excluded in all cases. In all 150 patients with positive air test results, CC was adequately controlled with conservative treatment.

Conclusions: The air test can be used as a new modality that is performed simultaneously with contrast enema as a non-invasive screening method for HD.
Accuracy of Calretinin for Diagnosis of Hirschsprung’s Disease
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Aim of the Study: To study accuracy of calretinin stain (highlighted ganglion cells, and/or nerve trunks, nerve fibrils) for diagnosis of Hirschsprung’s disease compare with gold standard method (Hematoxylin and Eosin; H&E stain).

Methods: A prospective double-blind diagnostic study, collected data from February 2015 to February 2017 was done. Hirschsprung’s patients who underwent Transanal endorectal pull through (TERPT) surgery were included. The pulled through specimens were identified into 3 zones, ganglionic zone, transitional zone and aganglionic zone which randomly sent to pathologist. Each specimen was stained with H&E and calretinin. Sensitivity, specificity, agreement and Kappa analysis were done.

Main Results: 40 Hirschsprung’s patients (120 specimens) were included for analysis. There were 72 males (72.5%) and 11 females (27.5%) with average age at surgery of 8.15 months. From 120 specimens, we found that there were 94 specimens showed no ganglion cells in H&E stain and 96 negative stain of calretinin (aganglionic bowel). 26 specimens in H&E stain showed ganglion cells and 24 specimens in calretinin stain were positive stained consistent with ganglionic bowel. The analysis represented Sensitivity 92.3%, Specificity 100%, agreement 98.3% and Kappa 0.949 (95% confidence interval 0.880-1.000, P<0.001).

Conclusions: Calretinin stain was found to be as accurate as H&E stain and could be used for diagnosis of Hirschsprung’s disease. In rectal suctional biopsy specimen which muscular layer not included, calretinin also can be used with high sensitivity and specificity.

Long-term Outcomes of Surgical Treatment for Hirschsprung’s Disease
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Objective: Recently, surgical treatment for Hirschsprung’s disease (HD) has greatly improved. However, some patients still suffer from numerous defecation problems. We assessed the long-term outcomes of HD patients after surgery.

Methods: Between 1997 and 2016, 41 HD patients were treated at our institute. Fourteen of these patients were followed for over 5 years. We reviewed their sex, age at the time of surgery, range of aganglionosis, surgical method, number of defecations per day, and presence or absence of developmental disorders using information from their medical records.

Results: At the time of a 5-year follow-up examination after surgery, the patients defecated less than 4 times per day, on an average. Patients who underwent surgery using the Duhamel-Ikeda method had fewer defecations per day (mean, 1.38 times per day) than patients who underwent surgery using the laparoscope-assisted transanal endorectal pull-through method (mean, 2.22 times per day). Patients with short aganglionosis (mean, 1.76 times per day) defecated less frequently than those with long aganglionosis (mean, 2.33 times per day). And patients with developmental disorders (mean, 2.67 times per day) defecated more frequently than those without developmental disorders (mean, 1.83 times per day).

Conclusion: Many HD patients had successfully acquired good defecation habits at 5 years after their surgery. The range of aganglionosis and the surgical methods affected the acquisition of good defecation habits. How developmental disorders influence the acquisition of good defecation habits or defecation problems after surgery remains unclear.

Transanal Endorectal Pull-through for Hirschsprung’s Disease in the Neonate and Early Infant
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The recent introduction of minimally invasive pull-through techniques may accelerate the primary definitive operation to be performed at further earlier period. We ascertain the feasibility and safety of totally transanal pull-through in the neonatal period.

The operation starts with a circumferential mucosal incision 5 mm above the dentate line. This is important to prevent loss of sensation, which may predispose the child to long-term problems with incontinence. The transanal submucosal
dissection was extended above the extramuscular plane. An incision was then made in the rectal muscle posteriorly to join the dissection from above. The muscle was split posteriorly down to the proposed anastomotic line to accommodate the ganglionated colon to be pulled through the sleeve. A single-layered, full-thickness anastomosis is created with interrupted, mono-filament absorbable sutures. The sutures should include a generous bite of the pull-through colon, as well as substantial bite of underlying muscle and a small bite of distal mucosa. A care must be taken not to include the dentate line in the sutures, and may compromise later continence.

Both the frequency of stools and the perineal excoriation usually settle down within several weeks to months postoperatively. Transanal endorectal pull-through in neonatal patients is as feasible and safe as in older children. However, temporary postoperative skin rash occurs more frequently in neonatal patients, and postoperative dilatations are required more often than in older children.

Sunday 7th October

POSTER WALK III

P-22

Thorough Retrorectal Dissection and Multiple Suture Fixation without Using Mesh for Adolescent Rectal Prolapse

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Introduction: In primary repair of rectal prolapse in children, the trend is to limit retro-rectal dissection to above the peritoneal reflection, localize rectopexy sutures to the sacral promontory, and use mesh. We present an intraoperative video recording of our thorough retro-rectal dissection and fixation technique without mesh.

Case report: A 15-year-old male presented to our department with severe recurrent full-thickness rectal prolapse of 10-40cm with every bowel motion for 6 months. As there was no obvious improvement with conservative management, primary surgical repair was planned. Preoperative barium enema showed prolapse was caused by anal verge inversion. With the patient supine and under general anesthesia, four ports were used to mobilize and fixate the rectum. After inspection of the extremely floppy rectosigmoid, tension was applied with a grasper, the peritoneum incised on the right side of the rectum starting from the peritoneal reflection to the sacral promontory, and blunt dissection deep into the pelvis up to the levator ani muscle layer to prevent nerve injury and preserve vascularity. A total of 7 polypropylene sutures (4-0 Proline) were used to fix the seromuscular posterior wall of the rectum to the median raphe (levator plate) of the levator ani muscle, as well as the presacral fascia, and the periosteum of the sacral promontory. All were tied extracorporeally. Operative time was 230 minutes. Stool softeners and daily enemas were continued for one month after surgery to minimize excessive straining.

Conclusion: At 2 years follow-up, there was no evidence of prolapse and no complications.

P-23

A Follow-up Study about Clinical Effect of Secondary Megacolectomy for Postoperative Recurrent Constipation in Children with Anal Atresia

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Objective: To evaluate the clinical effect of secondary megacolectomy for postoperative recurrent constipation in children with anal atresia.

Methods: 32 patients with recurrent constipation after anoplasty for anal atresia underwent secondary megacolectomy. The clinical data were analyzed retrospectively, and the postoperative bowel movements were evaluated by Krickenbeck classification.

Results: 32 patients were originally in intermediate and low malformation, including 11 cases of perineal fistula, 13 cases of vestibular fistula, 6 cases of rectourethral bulbar fistula and 2 cases of imperforate anus without fistula, aged 5~13 years old. 9 cases of 11 patients with perineal fistula underwent transperineal anus posterior cut-back, and the other 2 cases performed modified Pena procedure. 11 of 13 cases of vestibular fistula had transperineal transposition, and the other 2 cases had modified Pena procedure. 6 cases of rectourethral bulbar fistula and 2 cases of imperforate anus without fistula underwent modified Pena procedure. All patients had grade 3 constipation and distension of the distal intestines, in which 26 cases involved rectum and the other 6 cases involved rectum and sigmoid colon. Among 32 patients after secondary megacolectomy, 3 cases had incision infection, 2 cases had adhesive intestinal obstruction, and none had anastomotic leakage. 11 cases had autonomic defecation, 3 cases had fecal feces (all grade 1), and 18 cases had constipation (5 cases of grade 1, 11 cases of grade 2 and 2 cases of grade 3).

Conclusions: Secondary megacolectomy may evidently improve the bowel control of children with repeated constipation after anoplasty for anal atresia.

P-24

Anorectal Malformation with Fistula Exiting at the Penile Skin Associated with Bifid Scrotum and Minor Penoescrotal Transposition

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Anorectal malformations (ARM) have a worldwide incidence of 1-5 in 10,000 live births. ARM with recto-perineal fistulas is one of the most common anorectal malformations in male patients. Two-thirds of the ARM has an association with other congenital malformations; genitourinary anomalies are the most common (28-50%), but those with fistula without urethral communication are rare. Bifid scrotum (BE) refers to the deformity in which the labioscrotal folds are completely or partially separated without a median raphe. It is often associated with proximal hypospadias, being rare in the isolated presentation. Almost 15% is associated with an ARM. Surgical treatment depends on the severity of penoscrotal transposition and the association with hypospadias. Many surgical techniques have been described for the treatment of this anomaly. The purpose of this work is to present the association of two surgical techniques (posterior sagittal anorectoplasty and scrotoplasty with omega flaps) for the treatment of a 3-month-old patient with anorectal malformation and bifid scrotum with mild penoscrotal transposition (previously treated with colostomy) with good aesthetic results and functional continence without constipation to date.

P-25
Laparoscopic-assisted Anorectal Pull-through for High Imperforate Anus with Obscure Fistula in a Male
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After posterior sagittal anorectoplasty was reported by Pena about thirty-six years ago, this procedure has become a standard rule for patients with imperforate anus. Based on Pena’s description, a long incision from coccyx to perineal body is made with adequate separation of levators and external sphincter to facilitate dealing with fistula and putting rectum at the center of muscle complex and also within the sphincter. However, levators and external sphincter are not original appearance after destruction and reconstruction. Laparotomy is often necessary for identifying the fistula and repair when high type of imperforate anus is confirmed. Georgeson et al. first described laparoscopic assisted anorectal pull-through for high imperforate anus, and many pediatric surgeons have proved its benefits (smaller wounds, shorter hospital stay, lower complication rate, etc.) recently. In Taiwan, there are still few surgeons who have experience to perform laparoscopic assisted anorectal pull-through for imperforate anus. With clear vision and appropriate instruments, we successfully perform the technique for a male infant with obscure fistula in high-pressure distal colostography. There is no complication after six-month follow-up.
**P-26**

A Pitfall in the Diagnosis of Anorectal Malformation without Fistula: Usefulness of Ultrasonography with Lower Abdominal Compression

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**BACKGROUND:** To avoid unnecessary colostomy of anorectal malformation (ARM), we determine the type of ARM by ultrasonography. We report useful ultrasonography diagnosis with lower abdominal compression for a case with ARM without fistula.

**CASE:** A male neonate born at 35 weeks gestation (birth weight 2286 g) was suspected intermediate type of ARM because 20mm fistula-like tract from rectal pouch toward perineum was confirmed by ultrasonography at 6 hours after birth. However, fistula-like tract dilated toward 2.3 mm from perineum by ultrasonography with lower abdominal compression at 24 hours after birth, and we diagnosed the patient with low type of ARM. Transperineal anoplasty was performed on the 1st day of life. The postoperative course was uneventful.

**CONCLUSION:** The standard of our diagnosis with low type of ARM is rectal pouch-perineum distance of less than 10 mm by transperineal ultrasonography. Usual transperineal ultrasonography can lead to a misdiagnosis on the rectal pouch level due to an excessive contraction of the puborectal muscle complex. Our case documents the importance of ultrasonography with lower abdominal compression.

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**P-27**

A Rare Pediatric Case of Capillary Hemangioma of the Transverse Colon

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Hemangiomas are benign tumors commonly seen as head/neck lesions in children, occurring rarely in the gastrointestinal tract (GIT). We report the case of an 11-year-old girl with a rare capillary hemangioma of the transverse colon. She was referred to our hospital for management of gross hematochezia. On presentation, vital signs were stable with minor abdominal tenderness; hemoglobin was 5.7g/dL. She was admitted for blood transfusion and investigations. Abdominal ultrasound and computed tomography (CT) failed to identify any focal GIT lesions; a Meckel’s scan was negative; capsule endoscopy excluded small intestine pathology. Colonoscopy identified a 15mm mass in the colon that was not biopsied thoroughly because of its friable, hypervascular appearance. CT colonography confirmed the mass was in the middle of the transverse colon. Because the mass was sessile, depth of invasion unknown, and possibly malignant, open resection through a 5cm midline incision was chosen over endoscopic resection. The lesion was easily identified because of serosal hypervascularization. Intraoperative histopathology of a nearby enlarged mesenteric lymph node showed no signs of malignancy. Partial resection of the transverse colon including the lesion, and end-to-end anastomosis were performed without additional lymph node clearance. Histopathology of the resected specimen revealed vascular proliferation with a single layer of capillary endothelium in the submucosal layer of the colon. The postoperative course was uneventful. While the treatment was straightforward, our case documents the importance of awareness of rarer potential causes of massive hemorrhage in children, such as GIT capillary hemangioma, for expediting diagnosis and treatment.

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**P-28**

Efficacy of Bowel Management Program (BMP) on Fecal Incontinence in Children: A Team Work Approach

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In this study, we report the efficacy of Bowel Management Program (BMP) on fecal incontinence in children.

**Methods** This is a case series study that reports the results of BMP on children with fecal incontinence. BMP was trained to parents by a skilled team (Pediatric surgeon, pediatric psychiatrist, research assistant, registered nurse and education nurses). Patients were followed every week and feedbacks were received via social media.

**Main Results** Twelve patients underwent BMP. Five patients were boy and the average age was 8.5 years (5-10 years). Five had neuro spinal disorders (myelomeningocele and sizer), and the other had anorectal malformation and Hirschsprung’s disease. Two children did not keep on the enemas and was excluded the BMP study, so underwent the other treatments. In all of the cases, emptying the colon was happened about half an hour after the enema and they were clean for next day till the time of enema. Abdominal pain was seen in two patient that was during enema and softened with calming down the flow of serum. No other complication was seen.

**Conclusions** BMP should be considered as a team work, non-invasive and acceptable way to conserve children with fecal incontinence and help them coming back to society.
P-29

Congenital Perianal Lipoma Detected by Prenatal Ultrasound: A Case Report
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Congenital perineal lipoma, including perianal lesions, has been rarely reported, and perioperative strategies vary depending on its location and accompanying congenital anomalies. We herein report a case of congenital perianal lipoma first detected by prenatal ultrasound. A female neonate was referred to us for the evaluation of a perianal mass. She had been considered to be male prenatally because fetal ultrasound showed a perineal mass similar to a scrotum and penis. A postnatal examination revealed an appropriate-for-age neonate with a soft round mass 1.5 cm in diameter just left of the anal verge. She passed urine and stool smoothly, and contrast enema confirmed no anorectal malformation. Magnetic resonance imaging (MRI) showed that the lesion had a signal intensity consistent with fat located close to the anal sphincter, and no spinal anomaly (e.g. spina bifida) was identified. We excised the lesion (pathologically confirmed to be lipoma) simply at two months old, taking care to avoid damaging the anal sphincter using a muscle stimulator. She has been doing well with good bowel movement and satisfactory cosmetic results for a follow-up period of three months. Among the reported cases of perineal lipoma, our literature search revealed only one that was diagnosed prenatally, with others suspected based on ambiguous genitalia. Perineal lipoma is often accompanied by anorectal anomaly. A thorough physical examination after birth, MRI and contrast enema are crucial for planning the surgical strategy. A muscle stimulator is therefore useful for preserving the anal sphincter during resection.

P-30

The Related Factors of Postoperative Hirschsprung’s Associated Enterocolitis
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Aim of the study Hirschsprung’s associated enterocolitis (HAEC) is the most common complication after the operation of Hirschsprung’s disease (HD) and potentially fatal complication. The aim of this study was to determine the related factors of HAEC after the operation of HD.

Methods All HD patients, aged 0-15 years that were operated on 2004 to 2017 were reviewed. The demographic data, rectosigmoid ratio, preoperative bowel preparation, operative procedure and HAEC were recorded.

Main Results Twenty-four HD patients were operated and it was found that 13 (54.17%) of the patients had postoperative HAEC. There were 16 patients (66.67%) who had preoperative HAEC, 11 patients (45.83%) had both pre and postoperative HAEC and 9 patients (37.50%) had presented HAEC in the series. The patients that pathological lesion limited to the sigmoid colon had HAEC 61.11% unlike long segment HD and total colonic aganglionosis cases in which 33.3% presented HAEC episodes. All the patients had reverse rectosigmoid ratio. There were no related in postoperative HAEC and preoperative bowel preparation. The patients which LBW were related to HAEC (P=0.012). The operative age that is less than 4 months old were related to lower HAEC. All the patients who had anastomosis complications had HAEC (3 patients).

Conclusion HAEC remains to be the most common problem of postoperative HD. The factors that related to postoperative HAEC are LBW, preoperative HAEC and anastomosis complication. Although there is no standard preventive method for HAEC, early detection and early operation can reduce risk of postoperative HAEC.

P-31

Retrospective Analysis of Hirschsprung’s Disease in Infants Less Than 3 Months: A Single Center Study
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Objective: To investigate the efficacy of Hirschsprung’s disease (HD) in infants less than 3 months who were underwent transanal pull-through procedure(TAPT).

Methods: From July 2011 to January 2018, 41 infants less than 3 months diagnosed as HD were underwent TAPT procedure or laparoscopic-assisted TAPT procedure in our department. Clinical data, perioperative conditions, postoperative complication, postoperative anal function evaluated by Wingspread score and barium enema were collected.

Results: Age at the time of surgery was (47.59±16.28) d. All the cases completed single-stage operation procedure successfully without reoperation and death case. Length of resected bowel was (14.12±2.87) cm. In short term days, no complications such as anastomotic leak, interlayer infection and pelvic infection occurred, 10 cases of perianal erosion and 8 cases of intestinal colitis were cured after treatment. In long-term days, 3 cases with mild fecal contamination were improved after defecation management. All cases grow-up well and well nourished.

Conclusions: It is suitable for infants less than 3 months to undergo TAPT procedure, has the advantages of safe, less complications, well short-term and long-term efficacy.
P-32
Obstetric Considerations in Patients with Anorectal Malformations
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Aim of Study: The purpose of this study is to review two cases of pregnancy in patients with history of anorectal malformation (ARM).

Methods: Cases were identified from a single institution.

Results: Patient #1 is a 30-year-old female presenting at 24 weeks gestation with history of colostomy, posterior sagittal anorectoplasty (PSARP) and colostomy closure for rectovestibular fistula. Pregnancy also complicated by congenital heart disease, pulmonary hypertension, chronic hypertension, and developmental delay. She was managed by Maternal Fetal Medicine (MFM) who preferred assisted vaginal delivery due to complex cardiac history and high peri-operative risks. Given an adequately reconstructed perineal body, we supported trial of vaginal delivery with antepartum MRI to screen for tethered cord prior to epidural placement. She was ultimately induced preterm at 28 weeks gestation due to worsening pre-eclampsia and had a forcesps assisted vaginal delivery without injury to the anus.

Patient #2 is a 24-year-old female with history of rectovestibular fistula treated with colostomy, PSARP, and colostomy closure. For management of fecal incontinence, she ultimately had a Malone procedure. She presented for consultation at 29 weeks gestation with minor prolapse of her Malone which will be reassessed postpartum. We supported trial of vaginal delivery which will be scheduled at 37 weeks due to fetal growth restriction.

Conclusions: Multidisciplinary care in conjunction with MFM is instrumental in providing care during pregnancy for patients with ARM. Evaluation of the perineum and consideration of medical co-morbidities is essential before advising mode of delivery.

P-33
Transverse-loop Colostomy in Phased Treatment of Congenital Anorectal Malformation
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Purpose To investigate the advantages and disadvantages of transverse-loop colostomy in the phased treatment of congenital anorectal malformation (ARM).

Methods: A retrospective study was carried out on 62 congenital ARM children. All the children underwent protective transverse-loop colostomy as phased surgery. The clinical manifestations and results of related auxiliary examinations were analyzed.

Results: ① Before anorectoplasty and colostomy closure, only 21 cases and 22 cases of mild anemia occurred respectively in the totally 62 cases. ② Serum albumin examination showed that mild hypoproteinemia occurred in 5 cases and 1 case respectively before anorectoplasty and colostomy closure. ③ Electrolyte examination showed there were 7 cases of mild electrolyte disorder before anorectoplasty, and 6 of them were recovered before the colostomy closure. There was no hyperchloremic acidosis happened. ④ Urine routine results indicated that abnormal WBC in urine was found in 19 children before anorectoplasty and 4 children after fistula repair. Only 1 case experienced obvious symptoms of urinary tract infection. ⑤ High pressure distal colostography was executed in 57 cases. The distance from the distal rectum to anal crypt was 34.63±6.01 mm measured in the high-pressure distal colostogram, and 37.33±6.17 mm measured during operation (P>0.05). ⑥ All cases had no severe bowel retraction, stricture or prolapse stoma in the follow-up period.

Conclusion: Transverse-loop colostomy can be used in the phased treatment for congenital ARM children. It is simple, and causes few complications and exerts no serious adverse effects on nutritional status and growth and development in the children.

P-34
Two Case Reports of the Botulinum Toxin A Injection for Chronic Severe Constipation
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We report two cases of chronic severe constipation with megacolon who received botulinum toxin A injection to the internal anal shincter. Case 1 is a 4-year-old boy. He had the autism spectrum disorder and mild intellectual impairment. He felt difficulty in passing stools from 3 years old. Physical and radiological examinations revealed massive fecal impaction and abdominal distension. The symptom didn’t improve after fecal disimpaction, and medical treatments of glycerin enema and medications. We tried the treatment of botulinum toxin A injection. After three times of injection at 6 months intervals, he gradually showed easy defecation with glycerin enema and medications. Case 2 is a 5-year-old boy. He had attention-deficit/hyperactivity disorder. He showed fecal impaction and overflow incontinence in the first outpatient visit.
He underwent fecal disimpaction and glycerin enema treatment. We performed botulinum toxin A injection once. One year after the treatment, he became to defecate everyday with no medication. The results of botulinum toxin A treatment revealed to shorten the duration of treatment in two children with chronic severe constipation. The botulinum toxin A relaxed the sphincter contraction and helped easy defecation and toilet training for the children with fecal difficulty.

**P-35**

One-stage Single Incision Laparoscopic-assisted Total Proctocolectomy with Ileal J-Pouch Anorectal Line Anastomosis in Pediatric Familial Adenomatous Polyposis

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**AIM OF STUDY** Total proctocolectomy with ileal pouch anal anastomosis (IPAA) is the operative procedure of choice for familial adenomatous polyposis (FAP) patients. We report herewith our operative experience and outcome of a case of one-stage single incision laparoscopic-assisted total proctocolectomy with IPAA at “anorectal line” in a 14-year-old girl with FAP.

**CASE** The patient had intussusception of sigmoid colon and referred to our hospital. Polyposis with adenocarcinoma at sigmoid colon was found under colonoscopy. The patient had glioblastoma a year ago and was, after all, diagnosed as Turcot syndrome. At surgery, the patient was in the lithotomy position and pneumoperitoneum was created using the open method through the single 3cm incision at umbilicus. A single multi-channel umbilical port allowed us to perform enough mobilization and exteriorization of the total colon. This technique also facilitated a safe oncologic practice, with the removal of 15 lymph nodes and a full length of colon. The ileal J-pouch was created extracorporeally and pulled-through, and the transanal mucosectomy with hand-sewn anastomosis at “anorectal line” was performed. Instead of two-stage proctocolectomy with ileostomy, transanastomotic fecal drainage tube was inserted until postoperative day (POD) 10. The patient was discharged on POD37 after testing initial adjuvant chemotherapy. Short-term postoperative complications including anastomotic leakage, stenosis and wound infection were not occurred. The patient has now 5-6 bowel movements/24 hours and good fecal control.

**CONCLUSIONS** One-stage single incision laparoscopic-assisted total proctocolectomy with ileal J-pouch anorectal line anastomosis is technically feasible, and provides good continence and cosmesis.

**P-36**

A Case of Covered Anus Complete Presenting with Fecal Incontinence Claiming a Redo Operation 5 Years after Cut-back Anoplasty

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Cut-back anoplasty is widely applied in Japan to neonates of anorectal malformations especially for the low anomalies. We present a male patient born with a low type of imperforate anus. Meconium was seen through under the skin from the dorsal end of the scrotal raphe to the anal pit. Anoplasty was performed on the next day of birth by the cut-back procedure. Postoperative diagnosis was covered anus complete. Short-term postoperative course was uneventful. As the patient grew up, toilet training appeared to be unsatisfactory. In spite of fecal control by probiotics, he presented with the increasing frequency of fecal soiling at the age of five. The anal opening was found to have malpositioned anteriorly along the perineal raphe close to the scrotum with residual mucous membranous tissue in between. Electrical stimulator showed the center of sphincter muscle contraction 3 cm backward from the anal opening. A second operation was performed referring to Okada et al. who reported the effectiveness of anterior sagittal anorectoplasty as a redo operation for postoperative fecal incontinence (JPS, 1993). During the operation, the contractions in the sphincter and the levator ani muscles were confirmed to be well preserved. It was assumed that fecal incontinence in this case was because the primary anus had not exactly been constructed inside the sphincter muscle. Discussion is to be made on cut-back anoplasty mainly about its long-term complications.

**P-37**

Transrectal Approach for Excision of Vaginal Germ Cell Tumour with Pedicled Rectal Mucosal Patch for Posterior Wall Vaginoplasty

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**Background:** In pediatric vaginal tumours, there is very little tissue to spare if one contemplates organ preservation.

**Case:** An 18 month old girl presented with profuse vaginal bleeding of 2 weeks duration. A globular mass was felt just anterior to rectum 5x4 cm, rectal mucosa was free. Ultrasonography depicted a mass with mixed echogenicity 3.8 x 2.9x 2.7 cm. On MRI, mass was 4 x3.7x 2.8cm, involving the upper vaginal canal, cervix and lower uterus. Alpha feto protein was raised to 4244ng/dl. After 6 cycles of chemotherapy, lesion was excised in prone position with a transrectal approach. The anterior anal wall was incised for 2cm. The rectal mucosa was then lifted off the tumour. The tumour was involving the anterior rectal wall serosa. It was excised along with the posterior vaginal wall. The vaginal wall was approximated for postoperative fecal incontinence (JPS, 1993). During the operation, the contractions in the sphincter and the levator ani muscles were confirmed to be well preserved. It was assumed that fecal incontinence in this case was because the primary anus had not exactly been constructed inside the sphincter muscle. Discussion is to be made on cut-back anoplasty mainly about its long-term complications.
Secondary suturing of the vagina and deep perineum was done. At follow up after 9 months, the rectal mucosa had taken up well as vaginoplasty.

**Conclusion:** Pedicled rectal mucosa flap was successfully used for partial vaginoplasty.

**P-38**

**A Rare Case of Mature Cystic Colonic Teratoma**

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**AIM OF THE STUDY** We would like to report an unusual case of mature cystic teratoma (MCT) of the left colon presenting with intestinal obstruction in a syndromic patient with low-anorectal malformation (ARM).

**CASE REPORT** The boy was born at term with multiple malformations: low ARM with perineal fistula corrected on the third day of life without colostomy in a low-income Country, situs ambiguous, dextrocardia, double outlet right ventricle with pulmonary stenosis and anterior aorta. At two years he moved to our Country to perform urgent cardiac surgery. In the meanwhile he developed intestinal obstruction. We thought about intestinal stenosis with fecal impaction. The Angio-CT showed an oval-shaped abdominal "mass" but it was not decisive in defining its nature. At bowel enema the contrast means stopped in the descending colon. Once again, the hypothesis of a stenosis could not be ruled out. During laparotomy we identified a colonic cystic mass removed with colon resection. Histology showed MCT and follow-up was uneventful.

**CONCLUSIONS** MCT rarely occurs outside the sacrococcygeal-gonadal location. They are frequently associated with ARM in Currarino Syndrome: the mass is located in front of the sacrum and causes anal stenosis. Nevertheless, in the reported patient, MCT did not arise from the spine but from the left colon. Paediatric cases of colonic MCT have never been reported. Affected adults can develop constipation and/or intestinal obstruction. When the diagnosis is not clear, an open approach is to be preferred to remove the mass in both symptomatic and asymptomatic patients.

**P-39**

**Perianal Suppuration in Infants: Amenable to Nonoperative Management in Selected Cases**

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**Introduction** Perianal abscess is commonly encountered in daily pediatric practice. Although the most frequent approach is drainage with/without fistulotomy, its relative advantage and the place of conservative measures have not been established. This study sought to evaluate outcomes of conservative management of selected cases of perianal abscesses in infants.

**Methods** Data were retrospectively collected for medically managed patients (age <24 months) with perianal abscesses attending a tertiary pediatric medical center in 2014-2018.

**Results** The cohort included 19 patients, all male, of mean age 8.4 months at symptom onset. Twelve were being managed for the first time; the rest had undergone 1-4 previous drainage procedures under anesthesia (n=4), spontaneous drainage (n=1), or antibiotic treatment (n=2). Mean length of care in the community prior to presentation was 3 days, during which 5 patients received antibiotics. Indications for conservative management included spontaneous drainage into the anal canal (n=8) or perianal skin (n=4) and phlegmonous infiltrate without obvious fluctuation (n=7). On diagnosis, intravenous antibiotics were started in 18 patients and oral antibiotics in one. Average hospitalization time was 39 hours. Three patients failed treatment and required surgical drainage. Of the 16 patients who improved with conservative management, 2 were lost to follow-up; mean follow-up time in the remainder was 22.4 months. Three patients had a single recurrent episode that was managed conservatively. A fistula-in-ano developed in one patient and healed during follow-up.

**Conclusions** Perianal abscesses are amenable to conservative management in selected cases. Avoiding surgical intervention is advantageous, especially considering the high recurrence rate.