تاریخچه تکاملی بیماری هیرشپرونگ و مالفورماسیون ARM

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شرح این بیماری به قرن 17 میلادی برمی‌گردد یعنی در سال 1691 که فردریک رایش Fredrick Ruysch آناتومیست آلمانی دختربچه 5 ساله‌ای را که به علت انسداد روده فوت نموده بود شرح داد. در سال 1800 بانتینی یک بچه با مگاکولون را معرفی کرد و در سال 1886 هارالد هیرشپرونگ از دانمارک در شهر برلین اولین شرح بالینی مگاکولون را ارائه کرد. جناب هیرشپرونگ متخصص بیماری‌های کودکان و پاتولوژیست بیمارستان کودکان ملکه لوئیز کپنهاک ـ دانمارک بود. ایشان در سال 1887 دو مورد از این بیماری تکاملی را شرح داد که بعدها به نام وی بیماری هیرشپرونگ خوانده شد. تصور ایشان این بود که بیماری به علت اتساع کولون ایجاد می‌شود در حالی که فیزیوپاتولوژی و اتیولوژی آن چیز دیگری بوده است. عنوان مقاله آقای هیرشپرونگ این بود «یبوست در نوزادی به علت اتساع و هیپرتروفی کولون» تا شروع قرن بیستم دانسته‌ها در مورد بیماری هیرشپرونگ همین مطالب بود و بیماران گرفتار به این بیماری اکثرا می‌مردند که علت مرگ سوءتغذیه شدید یا انتروکولیت بوده است و جراحانی که این بچه‌ها را عمل می‌کردند معمولاً قسمت گشاد روده را بای پاس با یا بدون آناستوموز برمی‌داشتند. در سال 1901 تی‌تل (Tittel) متوجه شد که قسمت دیستال کولون درگیر بیماری است یعنی این قسمت فاقد گانگلیون سل است و از این زمان به بعد مقالات گوناگونی در این زمینه منتشر و نبود گانگلیون سل را پاتوگنومونیک بیماری معرفی کردند. به هر حال تا اواخر دهه 1940 میلادی هنوز درک کاملی از اتیولوژی و فیزیوپاتولوژی بیماری هیرشپرونگ وجود نداشت. Ehrenpreis در سال 1946 گفت اتساع کولون ثانوی به انسداد دیستال آن است و آگانگلیونوزیس را سبب مگاکولون تکاملی دانست، وایت هاوس و کرنوهان با بررسی بیماران خود و مرور مقالات منتشر شده اعلام کردند که آگانگلیونوزیس دیستال کولون یا رکتوم علت انسداد فونکسیونل روده است. بین سال‌های 1946 و 1948 سه گروه از محققین غربی گزارش کردند که نبود گانگلیون سل در شبکه میانتریک عضلانی و زیرمخاطی عامل اولیه بیماری است. سونسون و بیل در سال 1948 اولین کسانی بودند که گفتند بیوپسی تمام ضخامت رکتوم برای تشخیص قطعی و طرح درمانی لازم است و طرح درمانی آن‌ها حذف روده بدون گانگلیون یا رکتوسیگموئیدکتومی و جایگزین کردن قسمت طبیعی روده پروگزیمال به جای آن بوده است و این عمل امروز هم انجام می‌شود. در طول 50 سال بعد آگاهی پزشکان در مورد بیماری باعث شد که امروز بجای تشخیص دیرهنگام بیماری در سال‌های کودکی، بیماری هیرشپرونگ را در بیش از 90 درصدر نوزادان در دوره نوزادی تشخیص بدهند و مرگ و میر بیماری را از 70 درصد سال 1966 به یک درصد در سال 2000 برسانند. در سال‌های بعد تکنیک‌های تشخیص و درمانی گوناگونی ابداع شد و تحولی عظیم در درمان و عاقبت این بیماران رخ داد که تکنیک‌های با تهاجم کمتر با لاپاراسکوپی و عمل یک مرحله‌ای ترانس آنال در دوره نوزادی یا بعد از آن از مهمترین این پیشرفت‌ها می‌باشد. ولی به یاد داشته باشیم بزرگانی که نام برده شدند همراه با رهباین دوحامل، سوآوه، راویچ، پرم پوری و دیگران پایه‌گزار این پیشرفت‌ها بودند.

Results of Surgical Repair of Hirschsprung's Disease in Mofid Children's Hospital

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Purpose:

Considerable controversy exists regarding the optimal surgical technique for the treatment of Hirschsprung's Disease, We collected results of pull-through in last 7 years ago, to evaluate differences in surgical preferences among pediatric surgeons in our center and compare the consequences. We want to compare our results with another centers for identify our pitfalls.

Material & Method:

From 1385 until 1392 we had 193 patients with Hirschsprung's Disease that operated primary in our center. Referral patients with complication removed from our study. We provided Hirschsprung's Disease sheet and collected all information from the files. Then we called and requested them for visit. Results: We had 193 patients with mean age 20 months (10 days-168 month) for pullthrough procedure. 75% were male. 50% were identified in infancy period. 9% had associated anomalies that most common was down syndrome (5.1%). length of involvement was, Ultra short 11(5.7%), Rectosigmoid 139(72%), Descending colon 8(4.1%), transverse colon17(8.8%) ,ascending colon 1(0.5%), Total colonic 8(4.1%), Ileum 2(1%), Jejunum1(0.5%).Type of repair was Duhamel 6 (3.1%), Soave 32 (16.6%), Swenson 58(30.6%), Transanal 83(43%) ,State 5(2.6%), Myectomy Transanal 2(1%). 85(44.1%) patients underwent single-staged repair. 71(36.8%) patients in 2 stages and 25 patients (13 %) in 3 stages. Fallow up duration was between 1-8 years. Complications as early or late were seen in 75(39%) of patients that included: Entrocolitis in 20(10%), pelvic Abscess formation in 2(1%) cases, Fecal incontinence in 8(4%), anastomic Stricture 24(12%), Constipation 13(6.5%), fecal peritonitis 3(1.5%), Urinary retention 3(1.5%), obstruction 4(2%) ,colostomy prolapsus 4(2%), late entrocutaneous fistula followed abscess formation 3(1.5 %) ,colovesical fistula 1(0.5%), Massive perianal abscess formation and perianal fistula in 2(0.5%) and anastomosis in aganglionic segment in 1(0.5%). Frozen section biopsy was performed in 137 patients (71%), that frozen and permanent reports were different in 13 cases (6.7%).3(1.5%) patients expired. For 2 cases with entrocolitis colostomy established. And for 2 cases with fecal soiling Malone operation was done. Rectal stricture management include: anal dilatation program in 9 patients (4.7%), redo pull through 3(1.6%), y-v Anoplasty 3(1.6%), anal dilatation program under general anestesiain were done for others. 2 cases with perianal abscess formation led to colostomy insersion.

Conclusion: The only way to improve results is to gain more experience, assess the complications met, and learn from other.

Posterior Anorectal Myectomy for treatment of intractable chronic constipation in children

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Background:

Many children with constipation fail to respond with conventional medical therapy. Surgery can produce a good result in dysfunction of the colon secondary to aganglionosis. However, its role in treating idiopathic constipation is more controversial.

Patients and Methods:

A consecutive series of 44 patients with chronic idiopathic intractable constipation were included in this study. All children were investigated by barium enema and anorectal manometry. Due to inadequate response to medical therapy, all of these patients were selected for internal sphincter myomectomy. Patients were followed-up from 3 to 12 months.

Results:

Short-term (3 months) and long-term (6 months) follow-up was available for all patients. The histology examinations showed normal ganglion cells in 32, hypoganglionosis in eight and aganglionosis in four patients. In short-term, regular bowel habits, without the need for laxatives or low dose drugs were recorded in 35 patients (79.5%). Overall there was an improvement in 68.2% of the children after 6 months follow-up. There was not any correlation between histopathological findings, duration of symptoms, age and sex of operation and response to myectomy.

Conclusion:

Anorectal myectomy is an effective procedure in patients with intractable idiopathic constipation. It relieves symptoms in 68.2% of patients with chronic refractory constipation.

Comparison between two Methods of early feeding and conventional feeding in patients who underwent TERPT procedure due to classic hirschsprung's disease

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Introduction:

Trans anal endorectal pull through is a new method for one stage operation in patients with classic HD. Reduction in admission duration and drug administration are some benefits of this method in comparison with the other two stages methods. Early feeding after surgery is recommended in many GI operations with no additive complication. In this study we evaluated the complications of this surgery after two methods of early feeding and conventional feeding.

Materials and method:

All patients who underwent TERPT procedure due to classic HD from April 2016 to April 2017 in Children Medical Center, were interred in this study. Patients were divided in two groups randomly. In the case group oral feeding started 24 hours after surgery and in the control group oral feeding started 3-5 days after surgery. All patients were evaluated about infection, stenosis, abdominal distention, fever, obstruction, peritonitis, abscess and leak.

Results:

Infection was occurred 0% in case group and 5.88% in control group. Stenosis was occurred 20% in case group and 11.76% in control group. Abdominal distention was occurred 30% in case group and 17.64% in control group. There was no report of obstruction, peritonitis, abscess and leak in two groups. There were no meaningful differences between infection, stenosis, abdominal distention, fever, obstruction, peritonitis, abscess and leak which was occurred in two case and control group.

Conclusion:

We recommend early oral feeding after TERPT in patients with classic HD for reduction in admission duration and reduction in drug administration, with no additive complication.

Comparison of Outcomes of two Methods of Surgery (J-pouch and ileoanal anastomosis) in Children with total colon aganglonosis in Hirschsprung's Disease

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Abstract

Background:

Hirschsprung disease is commonly diagnosed in early childhood by aganglion areas in rectal biopsy. Diagnosed infants usually undergo a primary procedure and the definitive surgical treatment is usually performed several months later. Different surgical methods have been proposed for its treatment, but the detailed outcome of each method should be further investigated. Thus, we aimed to retrospectively assess the outcomes between two surgical methods for consistency of gastro-intestinal continuity, including J-Pouch and ileoanal anastomosis, in total colectomy procedure in our center.

Materials and methods:

In this study, we retrospectively assessed all children undergoing total colectomy after primary ileostomy in Children's Medical Center Hospital, Tehran, Iran, from 1994 to 2012. In this center, gastro-intestinal continuity was provided by J-pouch procedure from 1994 to 2003, and by ileoanal anastomosis from 2003 to 2012. In the second method, 0.1 mg/kg/dose loperamide was started after the first surgery (ileostomy) and was increased until the skin around ileostomy was just like the intact skin around colostomy with no significant inflammation. Data including demographic characteristics, need for re-ileostomy, duration of hospitalization, duration of NPO after surgery, and amount of loperamide were recorded, and compared between the two groups. Postoperative short-term complications were also recorded and compared. During the three-year follow-up period, all patients were assessed for soiling, manometric results, and fecal continence.

Results:

Among 37 patients undergoing total colectomy, 48.6% underwent J-pouch procedure (group 1) and 51.4% ileoanal anastomosis (group 2). In general, 54.1% were female and 45.9% were male. Mean hospitalization time was significantly lower in the second group (P=0.000). Mean NPO time was 7.06±2.55 days in the first group and 3.63±0.49 days in the second group (P=0.000). The rate of enterocolitis and mean surgical duration were significantly higher in the first group (P=0.001, and 0.000). None of the patients reported any fecal incontinence or constipation after surgery in both groups. Other postoperative complications had no statistically difference regarding leak, peri-anal inflammation, number of defecations, soiling, anastomosis stricture, need for re-ileostomy, pelvic abscess, peritonitis, and adhesion band.

Conclusion:

As the results of the present study indicated, the surgical method of ileoanal anastomosis has significant advantages to J-pouch procedure, including less hospitalization time, surgical duration and NPO duration, no cases of enterocolitis, fecal incontinence or constipation, which, in general, indicates that ileoanal anastomosis is a better method than J-pouch.

Keywords: Hirschsprung Disease; total colon aganglionosis

Ileal perforation as first presentation of Hirschsprung disease

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Abstract

A rare case of ileal perforation, as a fatal initial presentation of total colonic aganglionosis (TCA) in infancy is reported. A 8-week-old girl, was brought to the emergency department with symptoms of complicated intestinal obstruction. She looked ill, was lethargic, markedly dehydrated and had a severely distended abdomen.

An abdominal X-ray revealed multiple air fluid levels seen in a distended small intestine. During exploratory laparotomy the ileum was massively dilated with distal segment perforation.

Ileal perforation repair was performed. A totally collapsed microcolon was identified. Biopsies were taken from the high rectum, sigmoid and hepatic flexure. Appendectomy and ileostomy were performed.

All biopsies, as well as the appendix, showed absence of ganglion cells. Despite this procedure the patient progressively deteriorated and later died due to sepsis.

Ileal perforation in infants is a rare, but potentially fatal initial presentation of TCA. Early detection is essential to prevent life-threatening complications.

Colonic Perforation in Neonate

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Abstract:

Neonatal colonic perforation is a rarely seen condition. Plain abdominal radiography of a 24-hour newborn consulted for vomiting and bloody stool revealed the presence of subdiaphragmatic free air, which necessitated surgical exploration.

Transverse colonic perforation was detected during the exploration, and subsequently, a colostomy and appendectomy were performed. The postoperative follow-up period was uneventful.

Necrotizing enterocolitis, Hirschsprung disease, and mechanical obstruction are some of the causes of colonic perforation during the neonatal period.

Now, we are explained a case of colonic perforation in an asphyctic newborn delivered after prolonged labor.

Key Words: Hirschsprung, necrotizing enterocolitis, colonic perforation, neonatal, perinatal asphyxia

Total Colectomy and Ileorectal Anastomosis with Anorectal Myotomy A New Procedure for Treatment of Total Colonic Aganglionosis and Gastrointestinal Dysmotility

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Background:

Total colonic aganglionosis is present in 4-5% of the cases in hirschsprung's disease with high surgical mortality between 13 to 23%. Many techniques with several advantages and disadvantages were established. We have performed State pull-through as ileoproctostomy with long posterior myotomy in total colonic aganglionosis and sever dysmotility disorders.

Methods:

16 cases, 12 total colonic aganglionosis, 1intestinal neuronal dysplasia (IND) and 3 chronic intestinal pseudo obstruction syndrome (CIP) from 1992 to 2016 underwent total colectomy and resection of part of involved small intestine and ileorctal anastomosis in one layer with 4/0 vicryl with long posterior rectal myotomy. All patients had barium enema and rectal biopsy. Leveling ileostomy was done in 12 cases of which one had distal jejunostomy. 3 of 16 children, proximal diverting loop ileostomy had been established. 3cases, 2 weeks after initial operation, myotomy from anus, performed.

Results:

13 female patients and 3 male at the age of 6 months to years. Rectal biopsy of10 Patients reported no ganglion cell of which one had extended aganglionosis to distal jejunum, one had IND and two had ganglionic bowel with clinical presentation of CIP. Follow up time was 6months to 10 year. There were no significant complication in this group of patients except episodes of diarrhea and severe dehydration that need hospitalization and hydration. All have acceptable bowel function following operation (2-6 times a day). Now, 6 children of 16 patients are above the age of toilet training have voluntary bowel movement with little or no medication (Leopromid).

Conclusions:

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

Evaluation of Calretinin immunohistochemistry staining pattern in hirschprung disease

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Background:

Hirschprung disease (HD) is a congenital intestinal motility disorder characterized by the absence of ganglion cells and presence of abnormal hypertrophic verve bundles at the intestinal wall in routine haematoxylin and eosin (H&E) staining. So far various apecific and immunohistochemical stains were used to facilitate the diagnosis of the disease. The aim of this study was evaluation of immunohistochemical staining pattern with calretinin marker in nerve fibers and ganglion cells of intestinal wall in patients with HD.

Methods:

30 patients with histopathologic diagnosis of HD and 20 patients that underwent colectomy for another reason (as control group) were selected, and 80 blocks of full tickness paraffin- embedded intestinal specimens (30 blocks of ganglionic segments, 30 blocks of aganglionic segments and 20 blocks of control group) were obtained. Stained slides by IHC method for calretinin marker were observed separately by two pathologists and discrepancies were reviewed in a common session to get the final result.

Results:

Nerve fibers and ganglion cells in ganglionic segments showed positive immunoreactivity for calretinin without any false positive reaction and nerve fibers in aganglionic segments were not immunoreactive for this marker exept to cases (6.7%) of false negative with immunopositivity in nerve fibers of muscularis properia. Sensitivity of this method for demonstration of ganglion cells in full thickness specimens of intestinal wall was 93.3%, specificity was 100%, positive predictive value was 100% and negative predictive value was 93.8%.

Conclusions:

Immunohistochemistry with calretinin is a reliable method for demonstration of ganglion cells in intestinal wall and in combination with routine H&E stain will facilitate and accelerate the diagnosis of HD and eliminate the need for serial sections to find small and immature ganglion cells especially in neonates.

Keywords: Hirschsprung disease, immunohistochemistry, calretinin

Hirschsprung disease: A 10 year experience at Aliasghar hospital

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From 2013-2018, 43 children with diagnosis of hirschsprung disease underwent operation, 16 was male, 27 were male, median age was 2 month.

Approximatly 30% of patients underwent operation in the newborn period. About 50% of operation performed in two stages and 50% in one stage. All patient underwent Swenson procedure and posterior myectomy was performed in 7 patients (16.3%), 21% of patient were long segment in diagnosis. The mean follow up was 1year (2 week until 7 year)

After operation, the most common complication was chronic constipation(16%), five patient developed soiling and five patient enterocolitis pre op enterocolitis was nine patient (20%).

Reoperation was required in only one child (2%).

Intraoperative Sonographic Guided Pull-through Anorectoplasty:
A Novel Procedure for Imperforate Anus and Rectourethral Fistula: A Clinical Trial Study

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Abstract

Background: Optimal surgical management of the neonate with imperforate anus (IA) depends on determining accurate location of sphincter muscle complex, pouch of rectum and rectourinary fistula. We aimed to investigate a novel minimally invasive technique of anorectoplasty assisted by intraoperative sonography pull-through for repair of anorectal malformation and rectourinary fistula.
Materials and Methods: Eight male patients with anorectal malformation and IA underwent formation of a diverting colostomy within 48 hours after birth. These patients had anorectoplasty about 6-8 weeks postoperatively. A urinary catheter was inserted per urethra into the bladder and sonography of perineum was carried out under general anesthesia. The neoanus was reconstructed by suturing the pulled-through anorectum to the anal sphincter muscle complex and the skin using absorbable 4/0 sutures.
Results: In all cases, the distance of pouch of rectum to the skin was 15-18 mm and entrance of the guide wire to fistula was 6-7 mm. Of eight patients 6 had removal of urinary catheter after two days, and 2 patients after 10 days because the fistula was not closed. The median range of hospital stay was 2.12 days and follow- up was 30 days. We calibrate all of patients with size 12F dilators. Patients were followed up at one week and one month postoperatively, and all had defecation frequency of 3-5 times a day. There were no complications.
Conclusion: Intra-operative sonography guided pulled-through anorectoplasty (ISPA) is a novel and safe technique for surgical treatment of IA and rectourethral fistula. ISPA is a minimally invasive approach, which preserves the external anal sphincter muscle complex with good functional outcome.

Key Words: Anorectal malformations, Children, Clinical Trial Study, Sonography

Application of laparoscopic pull-through for Hirschsprung's disease

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Abstract

3 infants with Hirschsprung's disease referred to the hospital (rectal biopsy showed myenteric plexus without Gcell). Patients were lower than 1 year old (4, 6, 11 months). All patients were candidate of laparoscopic pull-through. To do Laparoscopy, all cases were placed in supine and head low position. Subsequently, first access was placed in the umbilical zone and two ports were inserted in the right and left quadrants. If needed, accessory grasper was used. After gross observation of large intestine, the TZ region was detected. After that, levelling biopsy of distal and proximal TZ were performed. Definite region of TZ was diagnosed. Colectomy was performed distal to TZ region and proximal intestine to TZ was prepared for pull-through. After completion of the mentioned process, laparoscopy ended. Patients were placed in lithotomy position and prepared intestine was trans anal pull-through. In this technique, multiple biopsy sampling were not required which led to cost reduction. Accurate intestinal observation and dissection limited the possibility of ischemia. Less potential for future adhesive intestinal obstruction, better cosmetic result and less damage to sphingter were other advantageous of this technique.

Application of modified laparoscopic appendectomy

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Abstract

Background: Appendicitis is the most acute surgical conditions in children. The choice of surgical procedure to treat these patients is a subject of debate. Laparoscopic appendectomy is a standard technique to treat these patients. This technique is performed in two ways including extra and intra corporal. Both of them have their own advantages and disadvantageous. In this study, we aimed to use a modified laparoscopic appendectomy which is the combination of both techniques. We tried to apply a minimally invasive procedure with lower cost than conventional laparoscopic methods.

Method: In this study, total of 100 patients with appendicitis, ranging in age between 2 to 16 years entered the study. All patients underwent modified laparoscopic appendectomy. Laparoscopic 2-port technique was used. The ports were selected in two different sizes 3 and 5mm. One of these ports was used to pass the camera. The role of another port was to guide the grasper and the suture during the procedure. In addition, stab wound was used to pass the second grasper. Dissection, meso and base of appendix were done by intracorporal method. Then, in extra corporal, the modified suture (designed by authors) and guided to the abdomen by grasper and meso and base of appendix were ligated. At the end stage, appendectomy was done and appendix was placed in the bag and was taken out by 5mm port. After the procedure, tissue samples were sent to pathology laboratory for further investigation. All patients were followed up after the procedure for possible complications. The data were analyzed by SPSS version 16.

Results: Of 100 patients, 38 were female and 62 were male. The histopathological examinations showed appendicitis in 92 patients, diverticulitis in 2 patients, perforation ileum in 1 patient, ovarian portion in 1 patient, and appendicitis without specific manifestations in 4 patients. The duration of surgery was between 20 to 80 min and patients discharged after 24 to 48 h. One month after surgery, infection was reported just in one case. No abscess observed in patients.

Conclusion: This modified technique was identified as safe as other conventional techniques and was considerably more cost-effective than other procedures. In addition, better cosmetic result was obtained. Minimal complications and Low incidence of infection in patients were other attractive points of this technique. Duration of surgery and hospital stay also were reasonable. This procedure had the capacity to investigate other pathologic examinations. So it can be used as an alternative method in appendectomy surgeries.

Key words: Modified laparoscopy, appendectomy, Pediatric

Preoperative Trans-Perineal Sonographic Findings in Children with Imperforate Anus for Detection of Anal Sphincter Muscle Complex and the Anal Canal Pathway: A Pilot Study

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Abstract

Background:
The exact anus reconstruction is the critical in patients with imperforate anus which is related to the correct diagnosis of sphincter complex.

Objectives:
The aim of this study is exact investigation of the perineal region for ultrasound detection of place and pathway of sphincter muscle complex.

Patients and Methods:
This descriptive cross-sectional study was performed at Mashhad University of medical sciences during 2016. Transperineal sonography was done in ten patients (6-12 week age, 8 male and 2 female) with imperforate anus.

Results:
The shortest distance between rectal pouch and skin was between 8 to 20 mm, but the distance between rectal pouch and skin via the anal sphincter path was longer (11 to 23 mm). The multi-layer view of anal dimple was seen in all patients except one. It had a curved and occasionally parasagittal path and it is more eccentric than muscle complex. The thickness of anal muscle sphincter complex could be seen in all patients with 2- 3.6 mm, occasionally asymmetric.

Conclusion:
The multi-layer view of anal dimple and the anal sphincter complex are the two important sonographic findings, which can better differentiated the level of anal malformation and act as an indicator for the location of this procedure.

Keywords: Ultrasound, Imperforate anus (IA), Child

Short outcomes of Laparoscopic Herniotomy

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Abstract

Introduction:

Inguinal hernia repair is one of the most frequently performed pediatric surgical operations. Open herniotomy is its standard treatment. Recently, many centers routinely perform laparoscopic hernia repair in children. Reported advantages of laparoscopic hernia repair include excellent visual exposure, minimal dissection, less complications, comparable recurrence rates, and improved cosmetic results compared with the traditional open approach. In this study, we aimed to perform children with hernia by laparoscopic herniotomy method and followed up them.

Method:

In this cohort study, total of 82 patients with hernia entered the study. The age range of patients was between 1-14 years. All participants underwent laparoscopic herniotomy. In this technique, umbilial port was used to pass the camera. Exploration of pelvic and observation of two performed inguinal canal were done by two stab wound. Then, dissection of sac from lateral to medial was performed. At the end, resection of the hernia sac was done.

Results:

The study population comprised 22(26.8%) girls and 58 (70.7%) boys. The defect was left-sided in 10 (12.1%), right-sided in 31 (37.8%), and bilateral in 41 (50%), and 2 (2%) patients without hernia. In this technique, surgeon was able to view lateral side. After follow up, there was low injury to vas and vessels. A few scars were observed.

Conclusion:

This technique was a minimally invasive method and did not need to do dissection. As a result, if the hernia diagnosis was not accurate, there would be low injury and scar to the body.

Key words: Hernia, Herniotomy, Laparoscopy

Soiling Management in Patients with habitual constipation

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Background:

Constipation is an extremely common problem in the pediatric population. Most constipated pediatric patients can be treated with simple measures such as dietary changes and laxatives, but some have severe constipation and need more aggressive treatments. Most have functional (idiopathic) constipation, which has a wide spectrum of severity.

Medical treatment with enemas, laxatives, and medications has traditionally been used for patients with soiling, with varying degrees of success. Bowel management with an organized protocol can have a dramatic impact on a patient with fecal incontinence. Likewise, bowel management in a patient with overflow pseudoincontinence can treat the impaction, avoid constipation, and promote the conditions needed for fecal continence.

Material and Method:

It's a prospective study on 12 patients from 2014-2018. Patients with soiling due to habitual constipation confirmed either by history, physical exam or barium enema and older than 3 included. For their management if there was a fecal impaction disimpaction performed and after precise parents training the patients put on laxative and bowel management program. It was performed daily for a month then one other day for another month and twice a week in the third month.

Results:

There were 8 girls and 4 boys with mean age 5.4years old. In this study all patients had a good response and after 3 months they were clean with normal defecation.

Conclusion:

Bowel management program although first introduced for fecal incontinence in ARM patients it can also be a good way to control soiling in patients with intractable constipation in those who don't response to routine medical management.

Neonatal colorectal disease: Necrotizing Enterocolitis

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Necrotizing enterocolitis is a frequently serious form of neonatal intestinal injury that characterized by ulceration and necrosis of the small bowel and colon. The lesions resemble acute ischemic damage.The lesions can be patchy or diffuse and in general, may affect any part of the GI tract. Terminal ileum, right colon (the proximal colon more commonly than the distal colon), and stomach are preferred sites of involvement. Transmural necrosis histologically occurs and may be associated with gas cysts within the bowel wall as pneumatosis intestinalis. The affected segment of colon is dilated, hemorrhagic, and necrotic with fibrinous exudates and adhesions on serosal surface. Therefore infants surviving the acute episode may later develop fibrous strictures.

One or more perforations are commonly identified during this stage. The incidence of NEC is on the rise, with 1% to 5% of infants in NICUs admitted (5% to 10% of all very-low-birth-weight (<1500 g) infants) and 2% to 22% of all premature infants, usually during the first 12 weeks of life. NEC generally affects low-birth-weight neonate because prematurity is the greatest risk factor for it. Although many investigators think NEC is a part of the spectrum of ischemia, its cause is probably multifactorial. A combination of ischemia, oral feeding, and pathogenic organisms are considered as the etiology for the presence of NEC. An increasing evidence supports a role for infectious agents in the pathogenesis of the lesion in as many as 23% of patients, bacterial organisms are isolated from blood cultures (such as E. coli, Klebsiella, Enterococcus, and coagulase-negative Staphylococcus) .Some of others risk factors are umbilical artery catheters insertion, perinatal asphyxia, respiratory distress syndrome, and persistent patent ductus arteriosus .

Signs and symptoms of NEC are nonspecific. Bradycardia, lethargy, vomiting, and abdominal distention are some of the early signs. In some cases, infants show discoloration of the abdominal wall or a palpable abdominal mass. Most common laboratory abnormalities are metabolic acidosis, thrombocytopenia, and leukocytosis.

An absolute neutrophil count of less than 1.5×10^9 cells per liter and progressive thrombocytopenia (that correlate with severe tissue injury and necrosis) are associated with a poor prognosis.

Severe infectious colitis and Hirschsprung disease – associated enterocolitis is the most differential diagnosis of NEC. Treatment of the NEC in the most patient are medical and includes bowel rest, abdominal decompression with a gastric tube, monitoring of fluid and electrolyte balances and administration of broad-spectrum antibiotics. Surgery is indicated in cases of bowel perforation or instances of medical therapy failure.

Hirschsprung disease as a complex genetic disease

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Hirschsprung disease (HSCR), a developmental disorder, occurs due to abnormal enteric nervous system development, which results in absence of ganglion cells of the entire gut. HSCR is categorized as a short segment, long segment, and total colonic and total intestinal aganglionosis. The incidence of familial history in HSCR cases is reported 5 to 20%. Other congenital abnormalities are seen in 30% of HSCR cases. The Mendelian inheritance (either dominant or recessive with decreased penetrance) was shown in familial and syndromic cases. In 70% of the cases, HSCR happens a sporadic with a non-Mendelian type of inheritance and the etiology of these are very complex with many genetic and Environmental Factors.

There are well-known associations with an increased risk of HSCR that include Down's syndrome, dominant sensorineural deafness, Waardenburg syndrome, neurofibromatosis, neuroblastoma, Phaeochromocytoma, the MEN Type IIB syndrome, and other abnormalities. Chromosomal anomalies also are found in up to 12% of HSCR cases and Down syndrome exist in about 2–10% of the HSCR cases.

HSCR as a complex genetic disease is associated with the complex cellular mechanisms in ENS development (i.e., proliferation, migration, controlled differentiation). The defects in many genes can increase HSCR risk with the activity of cell surface receptors (RET, EDNRB), extracellular ligands (GDNF, NRTN, EDN3), and transcription factors (SOX10, PHOX2B, ZFHX1B), as well as specific chromosomal anomalies (trisomy 21).

The RET gene is the first gene associated with HSCR, a proto-oncogene located on chromosome 10. The germline variants in RET accounts for 50% of familial and about 15–20% of sporadic cases. The genetic background and modifying factors are described to influence length segment without Ganglion cells. The second gene, EDNRB, was related to HSCR and found in association with Waardenburg Syndrome. EDNRB also has epistasis interaction with RET.

HSCR is described with genetic heterogeneity and, Up to date, deleterious mutations of more than 16 genes have been identified with the disease. The other HSCR genes, the exception of the RET gene, only have small penetrance. The identified genes and variants implicated in HSCR development have been explaining approximately 25 to 30% of all cases. Therefore, there are unidentified genes or unrecognized variants in known genes related to disease risk, need to be investigated. Whole exome sequencing (WES), a powerful tool for identifying variants in novel disease-associated genes, can be used to detect genetic defects in HSCR cases.

Key words: Hirschsprung, Genetic Complex disorder, WES

The Results of Two-stage Soave Boley Procedure in Hirschsprung's Disease in a 18 Year Period

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Background:

Several surgical techniques have been introduced for the treatment of Hirschsprung Disease, including total or partial resection of aganglionic segment and pull-through the ganglionic segment in consequence. In the current study the results of two-stage Soave-Boley procedure has been scrutinized.

Material and Method:

Medical records of 62 patients (46 male and 26 female) who were undergone surgery for Hirschsprung Disease, was evaluated. Following criteria were assessed ;age, gender, aganglionic length, primary surgical treatment, patients` age at the time of colostomy, surgical steps, immediate and late surgical complications followed by surgery, cause of mortality and pattern of bowel habit followed by surgery.

Results:

61 patients were undergone colostomy or end ileostomy of distal ganglionic segment as the first stage of treatment. Soave - Boley technique was applied in 54 patients. In 32 patients (52.4%) Soave - Boley procedure was accompanied with appendectomy and ceccostomy and in 22 patients (36%) Soave - Boley as a pure procedure without appendectomy or ceccostomy was undertaken. Satisfactory results were determined in 90.1% of patients and acceptable bowel habit was recorded. Immediate post-operative complications were noted in 9(7.14%) patients while 14(22.9%) of patients revealed late complications. Mortality rate was 9.4% (3patients). The mortalities were due to septicemia followed by ileostomy placement, severe entrocolitis 6 months after the surgical procedure and sudden death in the fourth post-operative day.

Conclusion:

By eliminating one stage of surgical procedure, better results and fewer complications might be encountered. Soave-Boley technique might be preferred due to lack of high cost stapler demand.

Finally, Two-stage Soave-Boley technique is considered as a cost-effective and appropriate method of treatment in Hirschsprung Disease.

Laparoscopic-assisted Pull Through and Anorectoplasty: A Single-center Experience

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Abstract

Aim:

To analyze the postoperative outcome and associated complications of laparoscopic-assisted anorectal pull-through (LAARP) for high anorectal malformations (ARMs) practiced at our institute.

Materials and Methods:

A retrospective study was done to analyze the results for LAARP procedure done for high anorectal malformations (ARMs) from 2013 to 2017. A total of 4 male patients had undergone LAARP. Staged procedure was done in all patients. All neonates with high ARMs underwent sigmoid loop colostomy at birth, LAARP at 2-4 months of age and colostomy closure after 8 weeks. The patients were followed up with clinical evaluation and continence scoring.

Results:

The complications were mucosal prolapse(2), anal stenosis, misslocation of neoanus(2) and transient neurogenic bladder (urinary retention). The progress has been satisfactory and weight gain is adequate but patients had some degree of either incontinence(1) or constipation(2) on follow-up.

Conclusion:

LAARP provides excellent visualization of the rectal fistula and surrounding structures. It is minimally invasive and leaves small abdominal and perineal wounds. Early postoperative recovery, early ambulation, and decreased pain to the patient are seen in LAARP patients. In our study, only one patient is beyond 4 years of age and his social continence is not good. Long-term follow-up is essential for evaluation of final results.

Keywords: Anorectal malformations, laparoscopic-assisted anorectal pull-through, rectovesical fistula

What is the Most Common Complication after One-Stage Transanal Pull-Through in Infants with Hirschsprung's Disease (HD)?

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Introduction:

HD is a relatively common congenital disease that could be suspected by clinical symptoms, abdominal plain X-ray, and finally diagnosed by rectal biopsy. In 80% cases rectosigmoid junction is involved. One stage trans-anal pull-through (OSTAPT) procedure has been popular recently and may have several complications.

Methods and Patients:

From 2006 to 2016, 180 infants (62 girls, 118 boys) with mean age 8 days (3 to 33 days) and clinically suspected to HD was admitted in our center. HD proved by rectal biopsy. All patients after full bowel preparation and rectal washed out were candidate for OSTAPT operation. A Swenson-like procedure was performed and the anastomosis was done between the well blood supply ganglionic colon and the rectum at 1 cm above dentate line. Interrupted suture with 5.0 Vicryle was used. Nelaton tube (12 F) inserted in the pelvis via transprineal for drainage of blood or collection. From Feb 2008 in 30 cases prophylactic Hegar dilatation was performed 2 weeks after operation.

Results:

Anal stricture was seen in 28 cases (14%) that treated by anal dilation in 20 cases and 8 cases corrected by surgical management. Entrocolitis in 8 cases (5%) that treated by medical management. In 3 cases retrocolic abscess that spontaneous drainage via tube drain. There were any anastomotic strictures after starting prophylactic anal bouginage.

Conclusion:

One stage TAPT has many advantages, low complications and the results is excellent. It seems the most common complications is anastomotic stricture that response well to prophylactic bouginage. We recommend prophylactic anal bouginage with Hegar probe at 2 weeks after operation. The result of this operation need to evaluate more in long-term.

Defecation Disorders after Surgery for Hirschsprung's Disease in Children

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Background:

Hirschsprung's disease (HD) is a congenital disease of the intestinal nervous system characterized by absence of ganglionic cells in distal colon followed by functional obstruction. After corrective surgery, the majority of children with HD develop defecation disorders such as soiling, constipation, fecal incontinence, and/or enterocolitis. The aim of this investigation was to determine the prevalence, diagnoses, therapies, and 6-month clinical outcomes in children with HD after corrective surgery.

Methods and Materials:

In this cross-sectional study performed at pediatric surgery ward in Tabriz Children's Hospital, prevalence, diagnosis and treatment of defecation disorders after surgery for HD were studied. First, defecation pattern was determined in 230 HD patients who had undergone surgery within previous ten years. Later, thirty patients with severe defecation disorders were recruited. Diagnostic and therapeutic procedures were performed. Clinical outcome was evaluated after six months.

Results:

Defecation pattern was normal in 65% of the patients. In 21% of the cases, defecation disorders were mild and negligible, with no need of treatment. In 13% (30 patients), the pattern was impaired. Soiling, constipation, fecal incontinence and enterocolitis were the postoperative disorders. Of 30 patients with defecation disorders, 18 children (60%) and 12 patients (40%) had undergone multi-stage and TOSEPT surgical procedures, respectively. Defecation disorder was developed in 25.3% and 7.7% of the patients underwent multi-stage and transanal one-stage endorectal pull-through (TOSEPT) surgical procedures, respectively. Twelve patients and 10 children were treated with reoperation and medical therapy, respectively. Clinical outcome was excellent in five patients (16.7%), good in 15 patients (50%), fair in 8 patients (26.7%), and poor in 2 patients (6.6%).

Conclusion:

Majority of the children with HD and postoperative defecation disorders have a favorable long-term clinical outcome when treated with minimally invasive surgical methods such as TOSEPT. Moreover, postoperative defecation disorders can be successfully treated using surgical procedures, medical therapy, as well as teaching both parents and their children.

One stage transanal endorectal (primary) pull through, a new technique in hirschsprungs disease and comparison with traditional staged techniques

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Backgrand:

During the past decade the authors have advanced the use of primary pull through for Hirschsprung disease in newborn and preliminary results have suggested excellent outcome. In this study we tried to compare short term and long term complications of the two procedures to provide a guide for choosing the safer and more effective approach.

Methods:

This study was planed case control clinical trial and all the HD cases treated during seven year period in Al-Zahra university hospital, Isfahan, Iran, 30 patients underwent one-stage approach, but the remaining 48 were treated via a multiple – stage approach. Short term and long term complications were compared between the two groups using t-test.

Result:

In staged pull through 56.2% of the patients have early complications includes; anastomotic dehiscence (8.3%), perineal excoriation (10.4%), pelvic infection (6.3%), early intestinal obstruction (18.8%), prolonged ileus (8.3%), but in primary pull through 3.3% of the cases have only prolonged ileus (P=0.001) and late complications in staged and primary groups were respectively, anastomotic stricture 8.3% vs. 3.3% (P=0.015), anastomotic retraction 2.1% vs. 3.3% (P=0.02), enterocolitis 27.1% vs. 6.6 (P<0.05) rectal prolaps and intestinal obstruction 2.7% and 16.7% vs. zero percent in primary pull through. In primary pull through 16.7% of the patients had a transient fecal incontinency during 14-60 days after surgery (mean 25.8± 19.3).

Conclusion:

Performance of a primary pull through for hirschsprung disease in the newborn is an excellent option, especially with an earlier diagnosis it can be an improved strategy, which will bring a better prognosis for HD patients.

Key words: Hirschsprung disease, Primary endorectal pull through

Botox Injection for Patients with Internal Anal Sphincter Achalasia Persistent to Posterior Internal Anal Sphincter Myectomy

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Background and Aim:

In studies, the gold standard for treatment internal anal sphincter achalasia (IASA) is considered posterior internal anal sphincter myectomy (ISM).We present our results of Botox injection treatment (BIT) in patients with non-relaxing internal anal sphincter after posterior internal anal sphincter myectomy.

Patients and methods:

The medical records of 35 patients with internal anal sphincter achalasia(IASA) managed by posterior internal anal sphincter myectomy during 2011-2015 were inspected. All patients presented with intractable constipation with or without soiling. Before posterior myectomy, all patients underwent barium enema and anorectal manometry. IASA was defined as the absence of rectoanal inhibitory reflex with normal rectal biopsies. In14 patients (8 males) with Mean ages 95 month (60-128) symptoms persisted. All of them had Constipation Grade3, Resistance to laxatives and diet and 5 had degrees of soiling. Botox injection 20U/kg was performed in general anesthesia in four quadrants into the intersphincteric groove. Laxative after injection continued in all cases.

Results:

Patients were followed for 2 years later (range 18–26 month). No intraoperative complications happened. In one patient, transient soiling occurred for 2 weeks after BIT.12 patients had improvement in bowel function more than 6 months,2 had improvement for less than 6 month.12 of 14 patient had normal bowel function after BIT with a P-value of less than .05 considered significant. Bowel Function required continued use of laxatives in 4 cases but 10 cases remained on small doses of laxatives. Although all patients were resistant to use of laxatives, they were able to use laxatives for having normal bowel function after BIT.5 patient had soiling before injection (4 occasionally and 1 every day without social problem) but after injection 4 patient had improvement and 1 patient had soiling occasionally and with a P value of less than .05 considered significant. No patients required enema after botox injection. There was no need to another injection after 2 year follow up.

Conclusions:

Intrasphincteric botulinum toxin is a safe and less-invasive for symptomatic internal sphincter hypertonicity after posterior myectomy.But it needs more long-term follow-up

Keywords: Hirschsprung's disease, Internal sphincter achalasia, Botox

Reoperations in ARM; Experience in 52 cases

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ARM are represented by a wide spectrum of defects. On the good side of the spectrum, those include mal formations that can be repaired with relatively easy technics and can obtain excellent functional results making the patient basically a normal individual that can enjoy a normal life. On the other hand on the bad side of the spectrum, one can find complex defects that make the functional outcomes rather somber. In these later cases it is almost impossible to restore normal bowel function, urinary function and sexual function. Between these two there are many types of malformation with different prognosis.

Pediatric surgeons must be serious in their philosophy that all patients with ARM should be clean of stool and urine in the underwear after the age of three, either with adequately reconstructed benign malformation or with malformations with bad functional prognosis. The patients are maintained artificially clean of stool (by BMP) and dry of urine (by CIC) through the native urethra or through a neourethra (continent diversion).

In Bahrami Children's Hospital we have 52 cases of reoperations during 20 years' experience in the treatment of the ARM. Fecal incontinence was the chief compliant in all of the patients. Physical examination revealed anal stenosis in32of them, while the normal caliber anus was found in others. The 2nd abnormality was mislocate anus which was found in 28 cases. Cloacal type was in 18cases in which only anorectoplasty had been performed elsewhere, but the UG sinus was intact and abnormal. Short sacrum was noted in 40 cases. Although we have not classified the patients as Pena, but their approach to treatment of these patients have more success.

However it is unacceptable to the patients with good prognosis to receive a bad surgical technique that destroys important structures and mechanisms of bowel and urinary control. Unfortunately it is something that happens more often than desired.

We feel that more morbidity occur in repair of anorectal malformation. In Pena‘s experience from 2032 cases over the30 years 478 cases were reoperations. 153 of them were to attempt to regain bowel control, 325 of them was for failed repair due to serious complications. From 909 male cases 223 was reoperations (93 for improve bowel control and 130 for failed repair. From 1123 female cases 60 were for bowel control and 195 for other reasons.

Unfortunately, we believe that the reoperations for anorectal malformations are not very reproducible. The surgeon must be open- minded to forget many of the traditional anatomic concepts. In addition the surgeon must be very gentle, careful and delicate. ARM patients are born with a rectum, sometimes a vagina and sometimes a urethra located in abnormal anatomic position. These structures are attached together and they share common walls without a plan of dissection. The separation of these structures is a mandatory step and is a surgical challenge. We have seen more complications when they have been attempted through a laparoscope.

Finally the wide spectrum of anatomic variations found in these patients is not well known by the majority of pediatric surgeons. In this paper the etiology of failed anorectoplasty is discussed and post-operative results are presented.

Bowel Management Program (BMP) on fecal incontinence in children (case series Study)

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Back ground:

Fecal incontinence is one of the most disturbing and psychologically distressing problems affecting children and cause poor self image, relationships, and unhappiness at school. In this study, we report the efficacy of Bowel Management Program (BMP) on fecal incontinence in children.

Method:

This is a case series study that reports the results of BMP on children with fecal incontinence. BMP was used for children with fecal incontinence that was referred to colorectal clinic since May 2017. BMP was trained to parents by a skilled nurse. Enema was done every night at a same time. The goals of BMP are to complete emptying the colon and being clean for next 24 hours. Patients were followed every week and feedbacks were received via social media. If the goals were achieved, enema was maintained with lower volume for the next 7 days.

Results:

Seven patients underwent BMP. Three patients were boy and the average age was 7.6 years (5- 10 years). Three had neuro spinal disorders (myelomenningocele and seizure), and the other had anorectal malformation and Hirschsprung disease. One child did not keep on the enemas and was excluded the BMP study. 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 one patient that was during enema and soothed with calming down the flow of serum. No other complication was seen.

Conclusion:

BMP should be considered as a non- invasive and acceptable way to conserve children with fecal incontinence and help them coming back to society.

Key words: Fecal incontinence, children, bowel management program

Urologic problems in ARM; Experience in 65 cases

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Urologic problems in pts with ARM represent a very important source of morbidity. In fact it is more likely for a child with an ARM to die from the urologic problem rather than from GI or any other associated defect. The incidence has been estimated to vary from 25 to 85%. The majority of reports inform of an incidence around 30-50%. The frequency of association increases directly proportional to the height of the defect and the complexity of malformation.

In addition, pts with ARM should be followed on a long-term basis, monitoring their kidney function and the anatomy of the urinary tract, because we know that many of them have anatomic or functional problems with a tendency to deteriorate.

All newborn babies with ARM should have a kidney ultrasound, within the 1st 24 h of life and before any surgical intervention. Female babies, particularly those with cloaca must have pelvic ultrasound to rule out the presence of Hydrocolpos. A male baby with normal kidney ultrasound and normal sacrum and no evidence of tethered cord and normal urination can be operated on without any further urologic tests. If the ultrasound shows hydro nephrosis further urologic tests including VCUG is necessary. In newborn with tethered cord and or abnormal sacrum (SR< 0.4), he is a nephrosis and mega ureter must disappear or improve. If these changes were not evident, the ureterostomy, nephrostomy, vesicostomy must be considered. In a small group of newborn with cloaca there is a very narrow common channel or almost atresia. This interferes with empting of bladder. After drainage of Hydrocolpos if the ultrasound show full bladder, it is justify doing the vesicostomy.

In cases with massive VUR and mega ureter and hydro nephrosis and presence of poor sacrum and tethered cord, vesicostomy is indicated. The newborn baby should not take to operating room before urologic work up. The most common urologic abnormalities in male patients with ARM are:

1-Absent Kidney 2-Urethral problem

The other urologic problems associate with ARM are:

1-Bifid scrotum 2-Hypospadias 3-Ectopic ureters in males 4-Ectopic Vas Deference 5-Ectopic veru montanum 6- Megalo urethra 7-UVJO and UPJO 8-Neurogenic Bladder 9- Post operative problems 10-Sexual Problems 11- Tethered cord.

We studied the charts of 65 randomized patients with ARM include 35male and 30 female. Four male (11.4%) and five (16%) female had unilateral non-function kidney (absent, dysplastic, MCDK). Three of 35male (8.5%) and five of 30 female (6.6%) had hydro nephrosis and mega ureter. The cause of hydro nephrosis and mega ureter was massive reflux or uvjo due to primary or the pressure of hydro metrocolpus to bladder base. The prevalence of vur was studied in these patients. Five of 35 male patients (14%) and six of 30 female patients (20%) had vur. These 65 patients evaluated or tethered cord in neonatal period by ultrasound. Sixteen of 65 patients (25%) had positive signs on ultrasound.

New method of transanal pull through operation in patients with hirschsprungs disease

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Email: psrg.sums@yahoo.comBackground

Hirschsprungs disease is an enteric nervous system dysfunction. Although, several types of pull through operations exist for this condition, each has its own pros and cons. We improved on this method by some essential modifications to conserve the symmetry of sphincter and also made the operation easier withless associated complications. Methods: During the period of month between January 2011 and February 2012 we performed a new method of transanal soave pull through after obtaining parental consent and ethical approval on 50 cases of Hirschsprungs disease. Mucosectomy was started 1.5 cm above the dentate lineas in classic soave. Once frozen biopsy showed ganglioncells the mucosal dissection and normal caliber colon reached, the dissection was stopped and a four quadrant myotomy on soave cuff done. The mucosal cuff was resected and full thickness anastomosis done above dentate line over a rectal tube. All patients started feeding on the second day of operation. Results: The patient were followed-up post-operatively withendorectal sonography and anorectal manometry todetect the intactness of external and internal sphincter;this was followed by an incontinence score. The mean age was 4.5 years (range 1-3 years). Internal sphincter pressure ranged from 29.6 ± 6.7 mmHg before intervention to 37.5 ± 6.5 mmHg after intervention and48.4 ± 8.3 mmHg 6-months after study ($P < 0.0001$). Defecation pattern score changed from 7.4 ± 1.9 to 6.1 ± 1.4, 6 month after study ($P = 0.002$). Early obstruction occurred in 20 of them, but it wasrelieved by rectal dilatation for 2-month. Three cases developed anal stenosis that underwent reoperation. There was no anastomotic leak or peritonitis.

Conclusion:

By avoiding opening the peritoneal cavity and saving the configuration of sphincter complex, appears to have fewer complications.

An introduction to Bowel Management program: Cincinnati Children's colorectal center experience

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Introduction:

Fecal incontinence is common in patients operated on for anorectal malformations or those children who suffered from severe retractable constipation. Treatment with enemas, laxatives, and medications are often given by clinicians in an indiscriminate manner and without a demonstrated benefit. Patients and parents who are struggling with childhood soiling and incontinence ate always frustrated and hopeless. The quality of life becomes poor and these patients usually suffer from physical and psychological impacts. In 2014, I was in colorectal center, Cincinnati Children's, and I listened to a presentation by Alberto Pena on the topic of Bowel Management. It was the first time I ever heard the words "bowel management" and these two words gave me hope that our children could be kept and remain artificially clean especially when I got more familiar with their fantastic bowel management program and its surprising remarkable results. In this article I decide to share this unique experience with the hope that we have a way to improve the quality of life for our children. This article will introduce a systematic diagnostic approach and bowel management program that is developed for patients suffering from fecal incontinence.

Material and method:

We applied the protocol in our retractable cases of fecal incontinence after precluding all correctable causes.

Results:

Data from 20 patients were reviewed and we observed a significantly higher failure rate among those who started the program as an outpatient plan.

Conclusion:

We concluded that we have to admit the patient and guardians for training and the bowel management program is not applicable except in the way it is running originally which means development of an independent unit with a fixed well trained nurse practitioner.

Key words: Fecal incontinence; bowel management; anorectal malformations

BMP in Patients with ARM Experience in 36 cases

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Fecal incontinence is a serious problem that provokes social segregation and psychological sequel. Patients with ARM frequently suffer from fecal incontinence despite the efforts of pediatric surgeon. At least 25%- 30% of patients with ARM suffer from fecal incontinence. In addition another 30% will suffer from other functional defecation disorders such as constipation, occasional soiling, and fecal incontinence during periods of diarrhea.

Medical management with enemas, laxative, and medication has been attempted in the past with varying success. These treatments are often given without a specific rationale and in an indiscriminative manner. There are patients who were referred to a psychiatrist when the surgeon felt that the treatment had been technically correct and the incontinence must have a psychological basis.

In Pena's experience 348 patients with fecal incontinence due to ARM were managed. They introduced their conclusions as follow: there are different types of fecal incontinence that depend on the original malformation and original operation. Proper evaluation and implementation of their treatment is necessary. Laxatives, medications to modify colonic motility, enemas, and colonic irrigations were found to play a role in the management of these patients. The impact on a patient's quality of life with bowel management is perhaps more significant than that of surgery itself.

In Pena's experience 348 patients seen due to fecal incontinence after anorectoplasty elsewhere.

The clinical and radiologic evaluation revealed different types of incontinence.

Group 1 included 147 pts who were considered candidates for reoperation

Group 2 included 172 pts who had no potential for bowel control and therefore candidates for bowel management.

Group 2A: 44 pts with incontinence and constipation: daily large enema only

Group 2 B; 128 pts with incontinence and tendency to diarrhea

Group 3; 29 pts with pseudo incontinence. This pts have original defect with good prognosis, good sphincters, good sacrum, and a well located rectum. They suffer from severe constipation, mega sigmoid, chronic fecal impaction, and overflow pseudo incontinence and were treated with laxatives or sigmoid resection.

Bowel management was successful in 93%of pts in the constipation group (2A) and 88% in the diarrhea group (2B). 93% of pts in group 3 became fecally continent. Bowel management consisting of enemas, laxatives' and medications is successful when administered in an organized manner. It is vital to determine the type of fecal incontinence from which the pts suffer and to target their treatment accordingly. In my country Iran we have great challenges with the parents in accepting the BMP for treatment of fecal problems including incontinence and constipation related to ARM.

We suggested the BMP to36 patients with treated ARM and fecal incontinence and constipation. Twenty of them had short sacrum. Six of the patients had experience of abdomino-perineal pull- through and six other had PSARP and four other had cloacal malformation. BMP was successful in those who performed the job properly. The parents were satisfied and the patients were clean under this program. There is not the same prescription to all of patients. The BMP is job of try and error. You must use different formula including the different amount of normal saline and fluid soap and glycerin and laxative to achieve satisfactory results. We have never used the phosphate in our experience.

Evaluation of the quality of life (QOL) in school- aged children who underwent Bowel Management Program for fecal incontinence

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Background:

Fecal incontinence is one of the most annoying and emotionally stressful complications in school-aged patients.

Purpose:

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.

Method and materials:

School- aged 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. The assessment`s validity and cronbach`s alpha are 0.84 and 0.82 respectively. The data was estimated by the SPSS 16 through the descriptive statistics.

Results:

A total of 6 school- aged children with fecal incontinence underwent BMP. Three were male and the mean age was 9.1± 1.9 years. The quality of life was in moderate level and the total score of QOL was 66.71± 19.24. The highest score (78.33± 16.33) was related to emotional aspects and the poor function was in school with the score of 61.88± 2.9. The social aspect was in high level (74.16± 23.75).

Conclusion:

Fecal incontinence disturbs all aspects of quality of life in children and BMP can improve these aspects.

Medical Causes of Chronic Constipation in Children

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Functional constipation is responsible for more than 95 percent of cases of constipation in healthy children one year and older, and is particularly common among preschool-age children. Functional constipation describes persistently difficult, infrequent, or seemingly incomplete defecation, without evidence of a primary anatomic or biochemical cause. This definition is operationalized by the "Rome IV" diagnostic criteria, which require at least two of six symptoms describing stool frequency, hardness, size, fecal incontinence, or volitional stool retention, with the stipulation that organic causes of constipation are excluded by a thorough evaluation.

The extent of the diagnostic evaluation to exclude organic causes is not specifically defined. In most cases, organic causes of constipation can be excluded on the basis of a careful history and physical examination. Focused laboratory and radiographic testing should be performed for children with atypical features (warning signs of possible organic constipation), or for those who fail to respond to a well-conceived and carefully administered intervention program, including disimpaction, frequent and effective use of laxatives, and behavioral management for at least six months.

Most important medical causes of functional constipation are:

A- Functional causes:behavioral constipation-Dietary habits-Genetics-Infant dyschezia
B- B-medical causes: Cow's milk intolerance-Slow transit constipation-Cystic fibrosis mild or moderate anterior displacement of the anus-Celiac disease-Internal anal sphincter achalasia-Dyssynergic defecation-Infantile botulism-Lead poisoning-Hypothyroidism-Chronic intestinal pseudo obstruction-Neurologic disorders-Extremely low birth weight Multiple endocrine neoplasia type 2

Evaluation - Evaluation of a child with constipation relies primarily on a focused history and physical examination; further testing is performed if the initial evaluation raises concern for an organic cause of constipation. The history should focus on features that suggest functional constipation and Acute and Chronic alarm signs.

Physical exam; General and perineum exam is very important. Notice to the "Rome IV" criteria is fundamental in patients suspicious to behavioral constipation.

Imaging;

Abdominal radiograph – A plain abdominal radiograph is not indicated for the routine evaluation of functional constipation.

Barium enema – A barium enema provides supportive evidence for Hirschsprung disease in children with features suggestive of this disorder. The study should be performed "unprepped".

Spine radiographs – Plain films of the lumbosacral spine should be performed for children with evidence of spinal dysraphism or neurological impairment of the perianal area or lower extremities.

Laboratory tests;

Celiac screening – For children with failure to thrive or recurrent abdominal pain, perform a complete blood count and serologic screening for celiac disease.

Urine analysis and culture – For children with a history of rectosigmoid impaction, especially in association with encopresis, perform a urine analysis and urine culture.

Thyroid stimulating hormone – For children with impaired linear growth and depressed reflexes, or those with a history of central nervous system disease, we suggest screening for hypothyroidism.

Electrolytes and calcium – For children at risk for electrolyte disturbances we suggest measuring serum concentrations of electrolytes and calcium.

Blood lead level – Screening for lead toxicity should be performed in children with risk factors. Screening recommendations vary by community.

Motility testing - Motility testing is typically considered in patients who have no obvious organic cause of constipation and who fail to respond to vigorous treatment of functional constipation.

Colon transit studies:

This study is generally reserved for the secondary evaluation of selected patients in whom the diagnosis is unclear despite a thorough initial evaluation and trials of treatment.

•To help distinguish between retentive and nonretentive fecal incontinence.

•To identify children with abnormally slow movement of food residue through the colon, a condition referred to as "slow-transit" constipation.

• To identify children with stool expulsion disorders, suggesting outlet obstruction.

Anorectal manometry;

The test is performed mainly in children with intractable constipation that restricts their lifestyle, or when there is suspicion of internal anal sphincter achalasia, or Hirschsprung disease.

Anorectal manometry also can identify patients with dyssynergic defecation, which is a functional disorder characterized by the incomplete evacuation of fecal material from the rectum due to paradoxical contraction or failure to relax pelvic floor muscles when straining to defecate.

Comparison of botulinium toxin injection anposterior anorectal myectomy in treatmentof internal anal sphincter achalasia

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Background:
We compared the efficacy of intra-sphincteric botulinum toxin (BT) injection and posterioranorectal myectomy (PARM) for the treatment of internal anal sphincter achalasia (IASA).

Methods:
Twenty eight of 120 patients (23%) with chronic constipation, who were referred to our clinic from September 2005 to December 2006, were evaluated. Patients had an absence of rectoanal inhibitory reflex on anorectal manometry (ARM) and showed no transitional region on barium enema. Fourteen patients each underwent rectal biopsy, and were treated with either in trasphincteric BT injection (Group I) or PARM (Group II). Nine patients were excluded because of absent ganglion cellson histology or positive acetylcholinesterase staining (AChE). The remaining 19 patients were followedup. All patients underwent ARM and constipation severity score (CSS) assessment 2 weeks before, and 1 and 6 months after the treatment. Patients were followed up telephonically at 12 months after treatment.

Results:
Clinically good response was seen after 12 months in 3 patients each in Groups I and II. The median values of resting rectal pressure in Group I before and 6 months after BT injection were 60 mmHg and 40 mmHg ($P<0.0001$), respectively, while in Group II the corresponding values were 60 mmHg and 45 mmHg ($P<0.0001$), respectively. Compared to pre-treatment, median CSS improved in both Group I (14 to 13) and Group II (16 to 14) at 6 months after treatment ($P<0.0001$ for both). However, there was no difference in resting rectal pressure and CSS between the groups. Three patients in Group II developed local abscess, postoperatively.

Conclusions:
BT injection has a similar efficacy as compared with PARM for the treatment of IASA, is less invasive and, is also associated with fewer complications

HD & Anorectal Manometry

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Background:

The most frequently performed motility test in infants and children is ARM Constipation is a common problem in infants and children, but the majority does not need to be screened with ARM.

Some indications for ARM testing include:

1-HD in patients who have been chronically constipated

2-laxative dependent or laxative unresponsive,

3- In inflants who have delayed passage of the meconium stool at birth or

4- Prolonged neonatal jaundice

5-failure to thrive

6-In children who have a gush of stool

7-The evaluation of fecal incontinence that is found in children in postoperation states and in those suffering from meningomyelocele, tethered cord and various other types of spinal cord dysfunction

Most pediatric patients tolerate the test well. Again, water – perfused systems delivering less than 1 ml water permin and a catheter tip 5 mm or less in used most frequency owing to their cost-effectiveness compared with solid – state manometry.

Hirschsprung's disease:

The primary indication for ARM is to rule out HD in infants and children, although ARM is also used in the evaluation of fecal incontinence. In the absence of ganglion cells, there is no normal reflex relaxation of the IAS when, the rectum is distended with air. The presence of the rectoanal inhibitory reflex (RAIR) – an intrinsic reflex mediated by the myenteric plexus – rules out HD.

ARM is also used in pediatric surgical cases to assess the candidacy for anorectal myectomy or a second pull through operation for HD.

Researchers:

Have observed the value of repeat ARM in the workup for HD. In a study of 56 patients with a history of delayed passage of meconium, 95 studies were performed. Anorectal manometry was done at weekly intervals for up to a month; ater, at which time suction rectal biopsy was added in conjunction withan indeteminant or equivocal RAIR. The authors concluded that, although the procedure could provide false-positive results, no false negatives were observed. Five cases of HD were diagnosed by manometry and confirmed by rectal biopsy.

Anorectal manometry procedure

ARM is performed in the follows steps. An enema the night before the test is recommended preparation. Some patients may need a 'cleanout' in the 2-3 days beforehand to assure at least a minimally empty rectal vaulton arrival for ARM.

Sedation

Toddlers and very anxious children may be sedated with oral midazolam.

Functional assessment

In older children, afull functional assessment of the anorectum starts with a pressure profile of the length of the anal sphincter. ARM provides information about the strength of the muscles forming the anal sphincter.

Sensory are measured to determine:

1- The volume of air first sensed, usually correlating with the perception of flatus
2-The volume at which the patient first senses the urge to defecate
3- The maximal volume tolerated, matching the urgent sensation to pass a stool.

Risks of Anorectal Manometry is unlikely to cause any painAnorectal manometry is a safe, low risk procedure and .Complications are rare.

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Familial Hirschsprung Disease

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Introduction:

Hirschsprung disease (HD) is created by the congenital absence of ganglion cells in the distal colon and rectum. It impresses the rectum and part of the sigmoid colon in patients, but it also can involve more proximal segments, the entire colon, or most of the small bowel. There is an increased risk for HD among patients with trisomy 21, and the number of other genetic syndromes.

● In all patients with HD, and particularly those with syndromic HD, genitourinary anomalies, hearing loss, and visual impairment are common. Congenital heart disease is common with syndromic HD.

● The majority of patients with HD are diagnosed in the neonatal period with symptoms of distal intestinal obstruction, including bilious emesis, gross abdominal distension, and failure to pass meconium in the first 48 hours of life. Patients with less intense HD may not be diagnosed until later in infancy or childhood. Such patients have a history of chronic constipation.

Result:

In this study, we follow up four families, with familial HD disease. Each family had two or three HD disease. There was no familial marriage. There was at least one TCA case in each family and we did not find any familial relationship between their parents. There were no accompanying anomalies, including GUT, visual and hearing impairment, CHD and ARM. There was no relationship between mother`s age at the time of birth and the incidence of disease. They were from 4 different geographic regions, Kashan, cum, Zanjan, and Ahwaz. All patients were examined and treated during the first month of birth. Except for a patient who mentions fecal incontinence at night, and a patient has diarrhea, another problem is not reported during the study.

Conclusion:

We must consider the familial HD disease in any case of HD disease who is referred to us, as in our study except one familial case, all three other familial cases did not mention us about the familial disease and we found it in family history taking from their parents.

Study of Sclerotherapy in children with prolapse of rectum

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Back ground:

Treatment of prolapse of rectum should be directed to the underlying cause, they usually respond to conservative treatment. Patients who present with a prolapsed rectum should undergo prompt manual reduction. Sclerotherapy is a good initial procedure. There are controversy about inject sclerosing agents.

Surgical treatment is reserved for patients who do not improve with conservative management or patients with complicated rectal prolapse (eg, recurrent rectal prolapse, painful episodes, ulceration, rectal bleeding).

Aid:

Is report of our experience in sclerotherapy treatment of prolapse of rectum with D/W50%.

Method: we respectively studied included the files of cases with idiopathic rectal prolapse in a 5 years treated by injection of D/W50% sclerotherapy we excluded others cases as rectal malformation and Hirschproung disease.

Our approach usually was as fallow, the patient is placed in the lithotomy position under general anesthesia. A 20-gauge spinal needle is introduced through the anal mucosa via a proctoscope or is externally introduced 2-3 cm from the anal margin at 3points contain (12.5 cc to 25 CC at 9, and 6 O'clock and 25 CC at 6 O'clock) with a guiding finger in the anal canal, to a point several centimeters above the dentate line. The sclerosant is circumferentially injected into the submucosal and perirectal space as the needle is withdrawn. To prevent, necrosis, bleeding, or stenosis, care should be taken to avoid injecting the sclerosing agent into the mucosa. The patients are discharged the same day with simple analgesics and stool softeners.

Result:

Total admitted patients were 27 patients every year as fallow: four pts. in 2013, six in 2014 seven 2015, and eleven 2016 male / female ratio was 19.8 and age mid age was 2years and 3mounth (between 1years and.11mounth to 14 y. and 10m. most patients had not any problem post sclerotherapy. 2 patients need to second injection 2 patients complicated: one with small perianal abscess and other with pre sacral other 13 Y old male with w eight; 70 KG that treated by local abscess drainage.

Conclusion:

Although he success rates and complications of the treatment reported in the literature differ for each sclerosing agent but Injection sclerotherapy with D/W50% is a good initial procedure < is cheaper and less invasive than surgery and has a success rates range from 90-100%.

Key Words: Sclerotherapy, children, prolapse of rectum

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Challenges in Transitional health care for patients with Hirschsprung disease and anorectal malformations: A Systematic Review

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Background:

Anorectal malformations and hirschsprung are one of the most common congenital intestinal anomalies affecting newborns that are a relatively common condition managed by pediatric surgeons that is a chronic condition associated with long-term morbidity. Despite advances in neonatal care and surgical techniques, many patients with a history of ARM and HD are affected by long-term challenges involving bowel ‘bladder and sexual dysfunction, and psychosocial issues. These outcomes or challenges are additionally exacerbated by the lack of a structured transition of care from the pediatric to the adult setting. We compared their health-related quality of life. Although follow-up is performed in most pediatric patients, transfer to adult health care is often problematic. This study assesses transitional care with the help of questionnaires in consultation with adult patients.

Methods:

A systematic review was searched for that described of Pub Med, Embassy, science direct and Cochrane Library and performed to evaluate current best practices for chronic illnesses of childhood with residual symptoms or need for medical care into adulthood.

Results:

Systematic review revealed improved results in transition programs as determined by patient follow-up, medication adherence, patient and family satisfaction through the use of multidisciplinary teams. Passive fecal incontinence was reported by 7/27, other defecator problems, including urge incontinence and incomplete evacuation in 17/27 and anal or abdominal pain reported by 9/27. Quality of life was lower than a matched population. Only 13/27 returned for repeat assessment at 1 year; however, a further 8 reported that that their problems had resolved. In those attending follow-up, negative thoughts and feelings about their condition had decreased and one more patient was fully continent. There was no change in quality of life, bowel function or pain recorded. Twelve out of thirteen patients reported that they had found the transitional clinic satisfactory.

Conclusion:

A significant number of hirschsprung disease and anorectal malformations patients experience bowel problems many years after definite surgery. The transitional outpatient clinic provides care adapted to the needs and wishes of adult HD and ARM patients. It is a novel addition to quality of care of patients with complex congenital disorders.

Key words: Transitional health care, Hirschsprung disease, Anorectal malformation, quality of life

Botulinium toxin, as bridge to transanal pullthrough in neonate with Hirschsprungs disease

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Background:

The aim of this study is to find easier way of home care while obviating the colostomybefore single stage pull through operation.

Methods:

From August 2005 to December 2006, eight cases of neonatal Hirschsprung disease were treated. Mean age 4.5 (2-6) day/old with absent anorectalinhibitory reflex, rectosigmiod disease in Barium enema, positive Acetylcholine esterase (Ache) staining, good response to rectal washout. They underwent botulinium toxin injection (5 unit /kg/quadrant) in fourquadrant intrasphincteric. They were followed until pull through operation in 8-10 weeks post injection.

Results:

Four of 8 (50%) cases only needed rectal washout for three to five days post injection until pullthrough operation, two had decrease in number of rectal washouts /day and the remaining two underwentcolostomy five days post injection because of no response.

Conclusion:

Botulinium toxin injection canhelp in palliative care in patients with Hirschsprung disease who are waiting for colostomy or definitivepullthrough. It gives an option of eaiser home care for these patients

Report of the First cases of laparoscopic pull-through anorectoplasty for persistent cloaca; Low in vasive and very simple method

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Purpose:

Persistent cloaca is the most severe form of anorectal malformation (ARM) encountered in girls and its usual treatment is one of the most complex and complicated pediatric surgeries. In the usual procedure, the common channel is divided into three sections with a relatively large incision in perineum, and the rectum, vagina and urethra are formed. The aim of this study was to describe the surgical technique and initial outcomes of laparoscopic anorectal pull-through for persistent cloaca.

Materials and Methods:

loaparoscopic rectal pull-through was performed for 2 patients with persistent cloaca. The patient ages were 6 and 9 months.

Surgical methods:

The surgery was performed in four steps: In the first stage, colostomy was performed. In the second stage, an abdominal pull-through and anorectoplasty was performed in a laparoscopic manner, in which the distal rectum is isolated from the common urogenital channel similar to the cases of high imperforate anus. In the third stage colostomy was closed. At the fourth stage at the age of four the common duct will be divided into two sections, the vagina and the urethra.

Results:

Laparoscopic rectal pull-through was successfully performed in 2 patients. Operative time was 150 and 200 minutes (mean, 175 minutes). The length of the common urogenital channel was 3.5 cm in the first and 40 mm in the second patient. There were no intraoperative deaths or complications.

The first patient suffered mucosal prolepsis 2 months and anal stricture 2 years after surgery that treated surgically. The 2ed patient suffered wound dehiscence 1 week after surgery that treated successfully by debridement and repair.

Conclusion:

Laparoscopic rectal pull-through is a feasible, effective, and less traumatic approach for anorectoplasty in patients with persistent cloaca.

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Comparison between Two Rectopexy Techniques, Mesh Rectopexy and Open Posterior Sacral Rectopexy

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Background:

Rectal prolapse is a common issue in children. It causes severe distress in parents. Rarely, it need surgical intervention to solve this problem. In this survey, we compare two method of perineal rectopexy for treatment of full thickness rectal prolapse.

Methods:

This clinical trial was done from April 2010 to march 2017 in our institute. After failure of medical and conservative management for at least 12 months, patients candidate for surgical intervention. Excluding factors were consisted previous anorectal surgery, any predisposing medical illness, solitary rectal ulcer, mucosal polyp, colorectal tumor and malnutrition. Patient consecutively were selected for open posterior rectopexy and mesh (gauze) rectopexy. Patients were followed for more than one year and rate of side effects and recurrence were compared between two groups. Data were collected and compared for statistical analysis by SPSS version 16 and P value less than 0.05 was considered as valid.

Results:

In this study we evaluated 71 patients that seven of them were excluded because excluding factors. We had 32 patients in each group. 54 (84%) of them were male. In each group we had 5 (16%) female. Mean age was 59.5±1 months in all patients and mean weight was 17.6±6.9 kilograms. There were no statistically differences in age and weight between two groups. Duration between occurrence of prolapse and time of surgery were 22.4±0.6 months in open posterior approach and 18±0.5 in mesh group. Mean operation time was 44.7±16.4 and 29.7±8.8 minutes consecutively (p value 0.0001). Mean duration of hospitalizations were 2.40+ 0.59 versus 2.88±1.07 (p value = 0.046). In open perineal approach, there were 8(25%) patients with recurrent episodes of rectal prolapse in one year postoperative period and 3(9.4%) patients in other group. None of patient need to re operation and all of them were managed by conservative methods. Bowel function were changed and showed better results after surgery in both groups.

Conclusion:

Mesh rectopexy shows better post operative results, less operation time and post operative episodes of rectal prolapse but has more complications in early post operative period.

A case report of complete colonic duplication

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Introduction:

Complete colonic duplication is a very rare anomaly with different presentation according to its location and size. Complete colorectal duplication can occur in 15% of GI duplication. Only 15% of them are rectal and there are very few reports of rectal prolapse or rectocele.

Case report:

A 2 year old boy with abdominal protrution, chronic constipation, difficulty in defecation, and rectocele(a bulging covered by mucosa) since 6 months old. In phynsical exam abdomen was soft with a palpable pelvic mass with doughy consistency and in rectal exam prerectal mass with soft consistency was palpated.

Imaging study:

Chest and abdominal X-ray, barium enema, ultrasound, VCUG, DMSA: Dilated bowel loops pushed anteriorly and cervical hemi-vertebra, absence of right kidney, pushed bladder anteriorly and superiorly.

Operative finding:

Complete colorectal duplication with blind posterior tube and accumulation of large amount of stool which was evacuated after resection of blind ended posterior tube and the 2 end of duplicated colon were fenestrated to each other and opened to common anal canal.

Conclusion: Side to side total colorectal tubular duplication may present as pelvic mass, mucosal prolapse (rectocele), and chronic constipation and may be associated with urinary and vertebral anomalies and treatment is simple resection of distal common wall and fenestration of 2 duplicated colon to each other.

The H-type anorectal malformations in Tabriz Pediatric Hospital Surgery ward during 2006 to 2017

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Background:

Congenital anorectal abnormalities are one of the most common abnormalities that are classified according to certain characteristics. This classification has been recently applied and is based on a well-considered therapeutic approach. What is obviously seen in all of these types and divisions is the shape and loation of the anus. But in some cases, despite the normal location and the size of the anus, there is an abnormal association between the gastrointestinal tract and the vagina that has not been addressed in reference books (H type rectovaginal fistula).

Methods:

Between 2006 and 2017, 165 girls with anorectal anomalies were admitted to Tabriz Children's Hospital. Some of these patients experienced rectal fistula in the vagina without having any problem in the location and size of the anus or any history of manipulating and procedures in the anorectal and genitalia. The diagnostic and therapeutic measures taken in the above patients and the results of their treatment and follow up will be discussed in detail.

Results:

Number of all patients were six and the patients' presenting symptom of passage of stool per vagina were also six and in one case accompanied by labial abscess. There was associated anorectal stenosis in 1 patient .There were no associated anomaly or morbidity and also all patients had normal sphincter tone. The remaining 5 patients had normal anal openings. The fistulas were all demonstrated on direct inspection under anesthesia. The fistula was located in the vestibule (5), vestibule and labia (1). four patients had been operated on three times previously using a diverting colostomy then perineal repair and reparing the stoma and two of them operated twice which was a diverting colostomy and reparing it which means spontaneously closure of fistula. In 3 cases, a posterior sagittal approach was used with mobilization of rectum and dividing and repairing fistula tract on both sides then repairing and reinforcing prineal body. One case went on fistulectomy with intact prineal body. After our repairs, the patients have been followed up till the time of presentation and we have seen no recurrences.

Conclusions:

In addition to vaginal passage of stool, an H-type fistula should be suspected when there is a labial abscess in an infant, and an associated anal stenosis must be checked for. Direct inspection is the key, with a careful look in the vestibule. Giving time after diverting colostomy may lead to spontaneous closure in some cases and the essential technical point for repair is to get healthy anterior rectal wall to cover the area of fistula on the posterior vagina accompanied by reinforcing the prineal body.

Prospective follow up of children with anorectal malformation: our center experience until 10 years of age

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Abstract

Purpose:
Longitudinal follow-up of bowel function in children with anorectal malformations (ARMs) as they grow, to determine the anorectal function problems and help to resolve them somedeal.

Material and Methods:
This study included 262 patients with ARM that operated in our center between 2006 until 2013.Patients that definitive reconstruction was performed in another center and underwent reoperation in this center excluded. Also children that expired or did not come for visit removed. Bowel function was prospectively evaluated by using a structured questionnaire that asked from their parents. Additional bowel treatment with enemas and stool softeners and use of diapers were recorded.

Results:
Interviews were completed with 242 children, age ranging from 3 to 10 years. 37.7% of patients had constipation. 32.5% Grade 1 Manageable by changes in diet, 54.3% Grade 2 Requires laxative and 13.2% Grade 3 Resistant to laxatives and diet. 18.6% of patients had fecal soiling, 31.5%.

Grade 1: Occasionally (once or twice per week), 24%
Grade 2: Every day, no social problem and 44.5%
Grade 3: Constant, social problem.

Conclusion:
In the present study there were many bowel function problems in ARM children, that needs additional attention to achieve them more near to level of healthy children. Pediatric surgeons who do the definitive surgery on anorectal malformations should don't lose contact with the patients as they become adults .These patients have many great troubles in adolescence.

Key words:
Anorectal malformation; Postoperative complications, Bowel functional outcome; fecal incontinence

Surgical Management of Ulcerative in Children and Young Adult

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Introduction:

Ulcerative colitis (UC) in children and young adult is frequently severe and medical treatment-refractory. There is an assumption that patients treated with 3-stage procedures for acute phase and active ulcerative colitis are undergoing a safer surgical approach and thus spared the complications associated with a 2-stage procedure.

Material and method:

Retrospective analysis of 15 patients who underwent 2 and 3-stage ileal pouch-anal anastomosis (IPAA) surgery for active ulcerative colitis due to failure of medical management over a 14 year period (2000 to 2014). The mean follow-up was 4 years (range, 2 month to 14 years).

Results:

Fifteen patients were evaluated in this study. Mean age was 10 years (4 months to 20 years). Mean time of medical therapy before surgery was 3.8 years. In 14 operated patients the indications of surgery were severe anemia, rectal bleeding, abdominal pain and very bad general condition irresponsive to medical therapy and in 1 case colon perforation during medical therapy and colonoscopy. In7 cases the surgical operation was two stages; Total proctocolectomy+ Endorectal ileoanal anastomosis+ loop ileostomy of which 4 had j pouch ileoanal and 3 straight ileoanal followed by closure of ileostomy after a period of 1 month in 6 cases and after 2 years in one case.

In 7 cases operation was three stages:

1- Total colectomy+ Hartman+ End Ileostomy

2- Total proctectomy, J pouch Ileo-anal anastomosis with stapler, loop Ileostomy

3- Closure of Ileostomy. 2 cases after first stage are waiting for the rest stages. Post-operative complications were as follows: in 2 stages 4 patients had complication of which 3 early septic complication (wound infection, pelvic abscess, bowel perforation and peritonitis), 3 late complication (delayed wound healing, entero-cutaneous fistula, perianal fistula and incontinency). In 3 stages, 3 cases had early complications wound infection, pelvic abscess and DVT.

Conclusion:

1- Chronic UC, long-term steroid therapy, Malnourishment, Suppress immune system and Urgent surgery are high risk of complications of primary surgery even with protective ileostomy.

2- Stage operation provides an optimal situation with less complication in acute phase of UC patients.

50% Glucose Injection in Presacral Space for Rectal Prolapse in Children

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Abstract

Aim of study:

A wide variety of sclerosing agents have been used in rectal submucosa in treatment of rectal prolapse in children. We have used 50% glucose in presacral space for the first time in the treatment. The aim of this study is to review the results of a 50% glucose injection in presacral space.

Methods:

In this study we included children who failed to respond to conservative treatment. The outcome OF 50% glucose injection sclerotherapy and the presence of complications were investigated. Under general anesthesia, the patient was placed in the lithotomy position. The left index finger was inserted into the rectum to control the position of the needle. A 20-gauge spinal needle was introduced through the perianal skin and was advanced. The 50% glucose was slowly injected through presacral space into, the right perirectal area the left perirectal area and posterior to the rectum at3points. The injection was continued until 5-6 ml of 50% glucose were injected in each quadrant.

Results:

A total of 15 children with complete rectal prolapse aged from4to 7 years, were treated between 2012 until 2015.Conservative treatment had previously failed in all patients. All of them were cured after one injections without any recurrence.Only one patient led to presacral abscess that underwent derange. There were no other complications. No fecal soiling was seen.

Conclusions:

The success rates and complications of the treatment reported in the literature differ for each sclerosing agent. Injection sclerotherapy by 50% glucose for treatment rectal prolapse in children is a simple and effective treatment.

Key words: 50% glucose, rectal prolapse, sclerotherapy

Limb Loss Caused by Thrombosis led to the Diagnosis of Ulcerative Colitis (case report)

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Arterial thrombosis associated with ulcerative colitis usually occurs in the postoperative period with a good response to anticoagulant therapy and embolectomy. Our patient was a 14-yearold girl with ulcerative colitis who presented with bilateral pulsless extremities, which did not respond to medical treatment and embolectomy. Subsequent colectomy did not save her limbs. The repeated thrombosis caused gangrene of extremities in the below knee region leading to bilateral amputation. Thrombosis can be the first presentation of ulcerative colitis

Severe Food Protein-Induced Enterocolitis Syndrome to Cow's Milk in Infants

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Abstract:

Cow's milk is the most common cause of food-protein-induced enterocolitis syndrome (FPIES). The aim of this study was to examine the clinical features and treatment outcomes of infants with severe FPIES to cow's milk.

We reviewed infants ¤12 months of age who were hospitalized and diagnosed with severe FPIES to cow's milk between 2015 and 2017 in Bahrami children's hospital in Tehran.

Patients' clinical features, feeding patterns, laboratory tests, and treatment outcomes were reviewed. A total of 5 infants met the inclusion criteria. All infants presented with diarrhea, edema, and hypoalbuminemia. Other main clinical manifestations included regurgitation/vomiting, skin rashes, low-grade fever, bloody and/or mucous stools, abdominal distention, and FTT.

They had clinical remission with resolution of diarrhea and significant increase of serum albumin after elimination of cow's milk protein (CMP) from the diet. The majority of infants developed tolerance to the CMP challenge test after 12 months of avoidance. In conclusion, we reported the clinical experience of 5 infants with severe FPIES to cow's milk, which resulted in malnutrition, hypoproteinemia, and FTT. Prompt treatment with CMP-free formula is effective and leads to clinical remission of FPIES in infants.

Keywords: cow's milk protein allergy; food-protein-induced enterocolitis syndrome; infants

A Medical-Therapy Resistance Constipation due to Internal Rectal Prolapse in Children (Diagnosis and treatment)

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Introduction:

Constipation is a prevalent disease in children which some of them need surgery procedures. One type of medical therapy resistance constipation has special symptoms and will occur due to internal rectal mucosal prolapse. This can cause constipation with painful fecal discharge and perspiration.

Treatment will be offered in two ways: with injection of Sclerozing agents in submucosal area or by Excision.

Methods and Procedures:

From 02/20/2016 to 02/19/2018, 48 patients with internal rectal prolapse were treated.46 of them have been cured with Sclerozing agent injection whereas 2 patients underwent surgery. one patient from injection group was not cured therefore, underwent prolapse Excision surgery.

Results:

From 46 total injected patients, 28 were fully cured, 9 patients were healed relatively so they had been fully recovered with food diet in 3 cases procedures were not efficient enough while 5 other cases are still under investigation. 3 patients whom underwent surgery were cured.

Conclusion:

Internal rectal prolapse is one of the medical-resist constipations and is considered as Idiopathic. Physicians intend to cure it with medicines. Symptoms of this disease are special and it can be cured by rectal submucosal injection of Sclerozing agent or surgery.

The 10-year study of colorectal polyps in children aged under 14 At Alzahra teaching Hospital in 2004-2014

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Introduction:

Intestinal polyp, especially juvenile polyp IS an Important gastrointestinal tract disease in children.lt is more frequent in patients with a positive family history since malignancy IS possible in this disease, diagnosis based on t1imcal and paraclinical signs and consequently treatment sec m Indispensable

Materials and methods:

Based on a descriptive, cross sectional study some 200 children aged under 14 who were suffering from colorectal polyps and visiting Alzahra teaching hospital during 2004-2014 and subjected to medical care were included in the study.

The information available on file Included clinical signs on visit, diagnosis; pathology reports, treatment and positive family history were collected and analyzed using SPSS 11 whereupon indices for central tendency and, distribution were reported.

Results:

Analysis of the data, from 200 case studies reveal ad that pathologically R5% suffered from Juvenile polyps, 2% from tubular adenoma, 1 % from tubular adenoma. 1 % from Villous adenoma. The mean and standard deviation of the patients' ages were 5.6 ± 3.33 on diagnosis.

55% of the patients were male and 45% were female. 5.7% of the cases had a positive family history 85% suffered from rectal bleeding, 31 % from rectal prolapus and 17% from constipation. 4% from diarrhea. 4% from abdominal pain and 1% from G.I.T obstructive signs .53% were diagnosed by rectosigmoidoscopy, 18% by colonoscopy, 2% by Ba enema and 27% by physical and rectal examination Pathological examinations revealed no malignancy, familial adenomatous polyp or inflammatory mass. 91 % of the patients were treated by endoscopyandpolypectomy and 6% by cauterization and 3% (6 patients) by laparotomy.

Conclusion:

In this study juvenile polyp was the most common type of polyp In GIT. Further, endoscopic methods were used in diagnosis and treatment which were consistent with other similar studies.

However clinical sign of patients were more sever at the outset than in other studies, which can be due to lack of parents knowledge and attention or due to lack of parents knowledge and attention or due to lack of attention on the part of medical doctors. Since polyps can lead to certain type of malignancies at older ages. Their timely diagnosis and treatment at well as screening of patients with a positive family history can play an effective role in the prevention of polyp related disease.

Key words: Colorectal polyp, Juvenile polyp, Children

Submucosal Ethyl Alcohol Injection for Management of Prolonged Rectal Prolapse in Pediatrics; A Single Center Experience

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Background:

Rectal prolapse is a benign and self-limited condition that causes considerable anxiety for the child and his family according to severity of the disease. Conservative management often is associated with improved symptoms; but in some cases the conditions is refractory. Injection of sclerosing agents could be a treatment strategy. The aim of the current study was to evaluate the effects of submucosal injection of ethyl alcohol on prolonged rectal prolapse.

Methods:

This cross-sectional study was conducted in pediatric surgery department of Namazi hospital, a tertiary healthcare center affiliated with Shiraz University of Medical Sciences, southern, Iran during a 4-year period from 2012 to 2016. We included those pediatric (<18 years) patients with prolonged rectal prolapse (>8 weeks) who were irresponsive to the conservative therapy. Around 1.5-2 mL of alcohol was linearly injected in three sites (two laterals and one posterior). We followed the patients for 6 months and the outcome was evaluated.

Results:

Overall we included a total number 164 patients with mean age of 4.66 ± 1.78 years. There were 87 (53.1%) male and 77 (46.6%) female among the patient with mean duration of prolapse of 5.6 ± 2.8 months. Overall, 157 (95.7%) responded to the single injection while 3 (1.8%) required second injection. The rate of non-responders was 2.5%. the complication included fecal soilage in 22 (13.4%) and pain in 18 (10.9%). There was no infection and no recurrence after the procedure.

Conclusions:

Submucosal injection of 96% ethyl alcohol is effective for management of prolonged rectal prolapse in children. This is a safe and applicable procedure which could be used in those irresponsive to conservative therapy.

Key Words: Rectal Prolapse; Ethyl alcohol; Submucosal Injection; Outcome

Pediatric Ingestion magnet foreign body with multiple enteric fistulas

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Background:

Ingestion of foreign bodies is a common pediatric problem, with more than 100.000 cases occurring each year. The vast majority of pediatric ingestions are accidental. Foreign body ingestion is a worldwide phenomenon, but the type of foreign body ingested may also vary by geographic region. In the United States, the most common pediatric foreign bodies ingested are coins, followed by a variety of other objects, including toys, toy parts, sharp objects, batteries, bones, and food. Increasing popularity of strong magnets as toys has led to their ingestion by children, putting them at risk of potentially harmful gastrointestinal tract injuries.

Management of foreign body ingestions varies based on the object ingested, its location, and the patient's age and size.

Aim is report a case of magnet and metal ingestion.

Case Report A 19 months old boy referred with Obstipation, nausea and vomiting to our Hospital. Her family, unaware of this ingestion, during work ups and abdominal x-ray revealed a metal density in cecum position so prepared for operation. During operation there was distended small bowel loops and jejunojejunal fistula in two sites. Metal foreign bodies consist of a magnet and screw bring out via appendix base and fistula sites repaired.

Conclusion:

Most foreign bodies or only single magnate with observation can be evacuated without difficulty. Although rare, multple magnets or amagnet with a iron particle that reach the intestinal tract may cause complications such as intestinal fistula formation, perforation, volvulus or appendicitis there fore requred timely and switable surgical intervention

Key words: Children, ingestion foreign bodies, magnate

Exclusive Breastfeeding effect on perianal abscess in infants, 2014-2017

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Background:

Perianal abscess is a relatively common disease during infancy. Many studies have been conducted on breastfeeding benefits but so far, there has been little research on the relationship between breastfeeding and perianal abscess. The purpose of this study was to investigate the relationship between breastfeeding and perianal abscess.

Methods:

In this case-control study, the cases were selected from infants who referred to the Pediatric center of excellence Tehran university of medical sciences with a perianal abscess from February 2014 to February 2017. We had 50 samples in case group and 100 samples in control group, consist of infants. The control group was the infants who came to vaccination center.

Results:

In this study, 78% of the case group and 66% of the control group, 10% of the case group and 12% of the control group, and 12% of the case group and 22% of the control group had exclusive breast feeding, exclusive formula feeding and combined feeding in the first six months of life, respectively. Without considering the confounding variables, P value was 0.274 for the relationship between exclusive breastfeeding. Finally, multi-variable logistic regression analysis was performed to eliminate the confounding variables, with a P value of 0.19 for exclusive breastfeeding, 0.05 for exclusive formula feeding and for 0.9 for combined feeding.

Conclusion:

The results of this study showed that breast milk does not have protective effect against perianal abscess in infants, directly. However; there is a significant correlation between exclusive formula feeding and the perianal abscess formation.

Currarino Triad: What Pediatric surgeons need to know?

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Introduction:

Currarino syndrome (CS) was first described by Currarino et al. as a congenital disorder characterized by the triad of anorectal malformation (ARM), sacrococcygeal defect, and presacral mass. In most cases of Currarino syndrome, presentation occurs in infancy or childhood. Symptoms such as intractable constipation and bowel obstruction in infancy are frequently associated with this condition. In its classic form, the anorectal anomaly consists of a very characteristic anal stenosis which is funnel-shaped up to the dentate line. The only mandatory clinical feature for diagnosis of CS is the sacral anomaly.

Case Report:

14 months old girl referred with constipation to our Hospital and admitted with diagnose of ARM and candidate for surgery. Pre operation work ups such as abdominopelvic sonography and cardiologist consult done which were normal. During operation after release of rectum for anoplasty there was a fibrotic mass in persacral region so resected and sent for pathology.

Conclusion:

The Currarino triad belongs to the group of persistent neurenteric malformations. Currarino syndrome (CS) was first reported by Currarino et al. in 1981 as a disorder comprising three pathological conditions; anorectal malformation, a sacral defect and a presacral mass. CS is an autosomal dominant inherited disorder caused by a mutation in the HLXB9 homeobox gene located on chromosome 7q36, some have thought it to be necessary to perform genetic studies for the accurate diagnosis of CS. Over 300 cases of Currarino syndrome (CS) have been reported in the literature but the incidence of the CS is unknown. CS related lesions are present in 56.9% of their families.

The incidence of abdominal symptoms with CS in infancy is low (16%) because the anorectal malformation in patients with CS is an anal stenosis (75%), which causes constipation, which is a common complaint in this syndrome. Nearly half of the CS patients with anal stenosis the anorectal malformation may not be diagnosed until adulthood because their constipation is mild.

The presacral tumor may be an anterior meningocele (68%), a benign teratoma (18%), an enteric cyst, a dermoid cyst, a lipoma, a leimyosarcoma, or, rarely as in our patient, a hamartoma.

If anorectal stenosis is identified in a child, the possibility of the other 2 components of the triad should be considered. Pelvic ultrasound scan, computed tomography (CT) scan, and pelvic x-rays should be performed. Computed tomographic myelography and magnetic resonance imaging (MRI) are the supplementary imaging modalities of choice for confirming the diagnosis and clarifying the extent of anomalies.

Treatment differs according to CS type and accompanying anomalies. It can be done with first colostomy then correction of ARM and resection of mass and then colostomy closure. Some investigators suggest one stage operation of ARM correction after colostomy closure that may be hazardous due to risk of infection.

Classification of Anorectal malformations

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Introduction:

Anorectal malformations (ARM) comprise a wide spectrum anomalies of the anorectal system, urogenital system, sacral spine and perineal musculature. The extent of anomalies in these four components decides the type of anorectal malformation.

Gender variations in the type of malformations must also be clearly defined before primary workup and management plan is drawn.

Based on the anatomy, various classifications have been proposed to define the pathology of these anorectal anomalies. The earliest classification dates back to 1953 when Gross proposed a simple differentiation based on the levator muscle (Fig.1), i.e. supralevator – for those above the levator ani or infralevator anomalies, for those below the levator ani.

With advancement in understanding of the pathology of the malformations, a need was felt to define these lesions more appropriately. During a meeting to celebrate in the Melbourne Royal Children's Hospital, a new International classification was proposed in 1970 as shown in table 1

In 1984, during a conference on Ano-rectal malformations organized by Prof. D. Stephens and Prof. D. Smith Wingspread, Wisconsin, another classification was proposed. This classification also included the special groups in cloacal and rare malformations as shown in Table 2.

By the early 1980's, several other rare anomalies such as perineal groove, H type of anorectal anomalies, pouch colon, rectal ectasia, rectal atresia, etc. were introduced and documented which were not included in the Wingspread's classification. Thus, in 1995 Pena introduced a disparate classification system as shown in Table 3.

May 2005, 21 years after the Wingsrpead classification saw the Krickenbeck meeting organized by Professor Alex Holschneider from Cologne, Germany. The goal of the meeting was to develop international criteria for treatment and develop a uniform scoring system for comparable follow-ups. The Pena's classification was modified as per the type of fistula and included rare variants as shown in table 4

With recent researches in the pathogenesis of anorectal malformations, the previous theories have been discarded. While in the past, defects in lateral fusion were thought to be causative, there is evidence from animal models and from detailed study of human fetuses with major anomalies that a deficiency in the dorsal component of the cloacal membrane and the adjacent dorsal cloaca is causative. A subsequent malfunction of the primitive streak and tail bud in the early development phase around 3-4 weeks has been proposed (yet to be clearly defined) as causation for associated anomalies of the pelvic floor.

The histological analysis of specimens from human fetuses with non-viable malformations revealed the following findings:

1. Primarily, the maldevelopment affects the anal canal and rectum is secondarily affected.
2. There is ventral displacement of anal canal which opens either on the perineum or forms a fistula to urogenital tract.
3. Those malformations in which a fistula is not demonstrated, a rudimentary partly regressed connection is found on histology.
4. In those with fistula from rectum to urogenital structures, there is a gradual transition of the anal mucosa to urogenital mucosa.
5. In proximal fistulae, the development of trigone of bladder, the upper urethra and the urethral sphincter is also abnormal in males whereas in females, vaginal development is inappropriate causing a urogenital sinus caudal to mesonephric ducts (as seen in persistent cloaca).
6. With deficient anal canal, the striated muscles of the perineum often have abnormal configuration. The longitudinal fibers of external anal sphincter are concentrated medially, the bulbospongiosus muscle is displaced medially in high lesions and the puborectalis sling, the external urethral sphincter and ischiocavernous muscles are variably affected depending on the severity of the lesion.
7. The likelihood of associated abnormalities in the development of pelvis, perineum, bladder, ureters, phallus etc. were proportional to the length of agenesis as measured from the actual anal site.

A thorough clinical assessment (substantiated with radiological assessment when needed) is essential for accurately classifying the malformation as the choice of surgical treatment is largely dependent on the extent of the anomaly.

The important aspects in history and clinical examination are listed in tabular form as shown in Table 5.

Assessment of the type of anomaly often needs radiological assistance in the form of x-rays or ultrasonography. Few associated anomalies also need to be investigated at the time of birth, especially the genitourinary and cardiac lesions.

The timing and method of radiological investigations are tabulated as in Table 6 as follows.

Anorectal malformations present with a high incidence of associated anomalies. The anomalies are presented in a tabular form in the decreasing order of frequency as shown in table 7.

New Classification of Anorectal malformation (ARM)

We have two types of classifications of Anorectal malformation.

Type I

Nonsyndromic ARM

Male:

1. Recto-Prineal Fistula
2. Recto Urethro-bulbar fistula
3. Recto-urethro-prostatic fistula
4. Recto bladder neck fistula
5. Imperforated Anus without fistula

Female:

- Complex and unusual defects:
    1. Recto-prineal fistula
    2. Recto vestibular fistula
    3. Cloacal with short common channel < 3cm
    4. Cloacal with long common channel > 3cm
    5. Imperforated Anus without fistula

- Complex and unusual sever defects
    1. Cloacal extrophy
    2. Covered cloacal extra
    3. Posterior cloaca
    4. ARM associated with presacral mass
    5. Rectal atresia
    6. Clocal with long common chnnel>3cm
    7. Hshape fistula (recto vaginal)
    8. Rectal duplication

Type II

Syndromic ARM

1. VACTERL: Vertebral anomalies, anal atresia, cardiac malformations,tracheo easophageal fistula, renal anomalies and limb anomalies
2. MURCS: Mullerian duct aplasia, renal aplasia, and Cervico- thoracic somite dysplasia
3. OEIS: Omphalocele, extrophy bladder, imperforate anus and spinal defects
4. Axial Mesodermal dysplasia
5. Townes- Brocks syndrome
6. Sirenomelia- caudal- regression Syndrome
7. Pallister- Killian Syndrome
8. Cat-eye-Syndrome (CES)
9. Reiger- syndrome
10. FG - Syndrome
11. Curarino - syndrome

Table 1: International classification of Anorectal anomalies

TYPE		MALE		FEMALE	
High (Supra levator)	Anorectal agenesis	Without fistula		Without fistula	
		With fistula	Rectovesical Rectourethral	With fistula	Rectovesical Rectocloacal Rectovaginal
	Rectal atresia				
Intermediate	Anal agenesis	Without fistula		Without fistula	
		With fistula	Rectobulbar	With fistula	Rectovaginal - low
					Rectovestibular
Low (Trans levator)	At normal site	Covered anus - complete Anal stenosis			
	At perineal site	Anterior perineal anus Anocutaneous fistula - covered anus (incomplete)			
	At vulvar site			Vulvar anus Anovulvar fistula Anovestibular fistula	

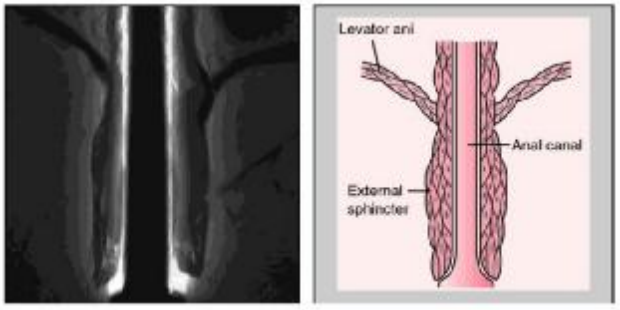

Figure 1: Diagrammatic representation of levator ani

Table 2: Wingspread classification

	Boys	Girls
High	Anorectal agenesis • With Rectovesical fistula • Without fistula • Rectal atresia	Anorectal agenesis • With rectovaginal fistula • Without fistula • Rectal atresia
Intermediate	• Rectobulbar urethral fistula • Anal agenesis without fistula	• Rectovestibular fistula • Rectovaginal fistula • Anal agenesis without fistula
Low	• Anocutaneous fistula • Anal stenosis	• Anovestibular fistula • Anocutaneous fistula • Anal stenosis
Rare malformations		

Table 3: Pena's classification of ARM [6]

		WITH FISTULA	WITHOUT FISTULA
NON-SYNDROMIC ARM	Rectoperineal malformations	Imperforate anus with recto-urethral fistula • Recto-urethral bulbar fistula • Rectourethral prostatic fistula • Bladder neck fistula	Cloacal malformations with a short common channel (<3cm)
	Imperforate anus in female	Rectovestibual fistula Rectovaginal fistula Cloacal malformation	Cloacal malformations with a long common channel (>3cm)
			H-shaped fistula (rectovaginal)
			Rectal duplication
SYNDROMIC ARM	VACTERL	Pallister-Hall syndrome	Townes-Brock syndrome
	MURCS	Lowe syndrome	Ulnar –mammary syndrome
	OEIS	Heterotaxia	Okihiro syndrome
	Axial mesodermal dysplasia	FG syndrome	Reiger syndrome
	Klippel Feil syndrome	X Linked mental retardation	Hirschsprung's disease
	Sirenomelia-caudal regression	Ciliopathies	Feingold syndrome
	Trisomy 21,13,18	Fraser syndrome	Kabuki syndrome
	Pallister-Killian syndrome	MIDAS syndrome	Opitz BBB/G syndrome
	Cat-eye syndrome	Christian syndrome	Johanson-Blizzard syndrome
	Parental unidisomy 16	Carnarino syndrome	Spondylocostal dysostosis
	Deletion 22q11 syndrome	Baiker-Gerold syndrome	Short rib-polydactyly syndrome

Table 4: Krickenbeck classification of ARM [7]

MAJOR CLINICAL GROUPS	RARE /REGIONAL VARIANTS
Perineal (cutaneous) fistula Rectourethral fistula/atresia/stenosis Bulbar fistula Prostatic fistula Rectovesical fistula Vestibular fistula Cloaca ARM's with no fistula Anal stenosis	Pouch colon atresia/stenosis Rectal atresia/stenosis Rectovaginal fistula H-type fistula others

Table 5: Clinical features in ARM in newborn at birth

		BOYS	GIRLS
History	Perineum – Pelvic floor	Absent/present anus	
		Specks of meconium in anal region	
		Failure to pass meconium	
		Meconuria	Passing meconium through introitus
		Flaturia	
	External Genitalia	Normal or abnormal	
	Abdomen	Vomiting	
		Distension	
		Distension with visible peristalsis	
	Others	Any other abnormality	
		Family history	
Examination	Pelvic floor	Absence or presence of anal opening	
		Position of anus – normal or anteposed	
		Bulge in perineum on crying or straining	
		Anal dimple	
		Shape of buttocks	
		Anal reflex	
		Perineal groove	
		Bucket handle deformity	
		Meconium or mucus running up the median scrotal raphe	
	Genitals	Phallus – Normal or hypospadiac	Appearance of external genitalia + labia normal or shortened (cloaca)
		Meconium staining at urethral meatus	Number of openings in vestibule • Single opening – cloaca • 2 openings – rectovaginal fistula/rectovestibular fistula with absent vagina • 3 openings – anovestibular fistula
		Testis descended/undescended	
		Any other abnormality	
	Abdomen	Large visible loop occupying more than half of abdomen	
		Palpable kidney/any other palpable lump – solid or cystic	
			Hydrocolpos – palpable lump in lower abdomen
	Lumbo-sacral spine	Occult or obvious spinal dysraphism	
		Absent sacral vertebrae of variable levels	
	Other associated anomalies – as described below		

Table 6: Radiological assessment in newborns with Anorectal malformations

TYPE OF TEST	TIMING		INTERPRETATION
	BOYS	GIRLS	
Plain X-Ray Abdomen Erect	At birth/at the time of prone cross table lateral (PCTL) xray (10)	If massive distension of abdomen and failure to pass meconium	• Multiple dilated bowel loops with air-fluid levels and absent rectal gas • Large dilated loop with A-F level (pouch colon)
X-ray lumbo-sacral spine -	At time of PCTL	At birth/at time of pre-operative work-up	• Measure sacral ratios • Sacral defects • Hemivertebrae • Presacral masses
PCTL X-ray	12-18 hours after birth or later if presentation is after 18 hours	Not done	Presence of rectal gas shadow : • Low anomalies – below the M line • Intermediate anomalies – above the M line and below the PC line • High anomalies – above the PC line Other features : • Air in bladder • Beaking of terminal rectal pouch (fistula)
Invertogram [11]	Obsolete	Not done	
Ultrasonography	Abdomen	Abdomen and Pelvis	• Urological anomalies especially hydroureteronephrosis (VUR), Hydronephrosis, absent kidney, etc. • Presence or absence of uterus/ovaries in females as well as s/o hydro/hydrometrocolpos
	Cardiac – 2D Echo - Congenital Cardiac anomalies		
	Spine – screening for occult spinal malformations		
MRI – abdomen and pelvis [12,13,14]	• Delineates the level of anomaly • Provides information about the fistula • Pelvic floor musculature – puborectalis sling, external anal sphincter anatomy is clearly defined • Anomalies of spine, spinal cord, urogenital system can be simultaneously diagnosed		

Table 7: Associated anomalies in ARM

SYSTEM	TYPE OF ANOMALY	FREQUENCY
Urinary	Vesico-ureteric reflux Hydronephrosis Renal agenesis Renal dysplasia	50%
Genital	Vaginal septum	50%
	Uterine didelphys/Bicornuate uterus	35%
	Cryptorchidism	3-19%
	Vaginal duplication/vaginal agenesis/absent ovary	
Vertebral [15]	Lumbosacral anomalies Tethered cord Cord lipomas Syringohydromyelia	30-35%
Cardiovascular	VSD Tetralogy of Fallot Transposition of great vessels Hypoplastic left heart syndrome	12-22%
Gastrointestinal [16]	Tracheo-esophageal fistula	10%
	Duodenal obstruction	
	Malrotation	
	Hirschsprung's disease	
Curarino triad [17,18]	- Sacral defect + presacral mass + imperforate anus	> 350 cases reported in literature
Other anomalies	As listed in Pena's classification	Rare

Cloacal Malformations: Diagnosis, classification & Surgical management

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The diagnosis of a cloaca is a clinical one. This defect should be suspected in a female born with imperforate anus and small-looking genitalia. Careful separation of the labia discloses a single perineal orifice. The length of the common channel varies from 1-7 cm, and is very important for operative and prognostic implications. A common channel of less than 3 cm usually means that the defect can be repaired with a posterior sagittal operation without opening the abdomen. Common channels longer than 3 cm are more complex, mobilization of the vagina is often difficult, and some form of vaginal replacement may be needed during the definitive repair. The average length of the common channel 4.7 cm for patients that required a laparotomy and 2.3 cm for those those do not. Vaginal reconstruction involved a vaginal pull-through, vaginal flap, vaginal switch, and vaginal replacement with rectum, ileum, and colon: Current surgical techniques for cloacal reconstruction are posterior sagittal anorecto vagino urethroplasty (PSARVUP) and posterior sagittal anorectoplasty (PSARP) with totalurogenital mobilization (TUM). Before the introduction of the total urogenital mobilization Complications included vaginal stricture or atresia, urethral strictures, and urethro-vaginal fistula. About half of total evaluated patients are continent of urine and others remain dry with intermittent catheterization through their native urethra and through a Mitrofanoff-type of conduit. High percentage of the patients with a common channel longer than 3 cm require intermittent catheterization compared with when their common channel was shorter than 3 cm. Sixty percent of all cases have voluntary bowel movements (28% of them never soiled, and 72% soiled occasionally). Forty percent are fecally incontinent but remain clean when subjected to a bowel management program. One thirth of patients born with hydrocolpos If not treated correctly during the neonatal period or the surgeons failed to drain the dilated vaginas, which interfered with the drainage of the ureters and provoked urinary tract infections, pyocolpos, and/or vaginal perforation. In patients the colostomy was created too distally, it interfered with the pull-through. patients suffered from colostomy prolapse All of these patients required a colostomy, revision before the main repair.some surgeons divids persistent cloac into 2 distinct groups of patients: group A those with a common channel shorter than 3 cm (62%) and group B a common channel longer than 3 cm (38%). The separation of these groups has important therapeutic and prognostic implications. Group A patients can be repaired posterior sagittally with a reproducible, relatively short operation. Because they represent the majority of patients, that most well-trained pediatric surgeons can repair these type of malformations, and the prognosis is good. Those with a common channel longer than 3 cm usually require a laparotomy and have a much higher incidence of associated urologic problems.

A new technique experience for urogenital management

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Introduction:

Genital ambiguous is a very common phenomen in disorders of Sex development (DSD). Advances in the knowledge of genital. Anatomy in DSD have enabled the development and improvement of various surgical techniques.

Mobilization of the urogenital sinus first described by Pena, has become incorporated by most surgeons and has mostly replaced older techniques. Concerns have been raised about the effect of this operation on urinary continence.

TUM and PUM are the most common vaginoplasty surgeries for patients with CAH, urogenital sinus, Cloaca anomalies.

We reviewed the urinary continence outcomes of children who underwent TUM or PUM at our institution. Our aim was to emphasize the importance of joint mobilization of urogenital sinus after sepration from the rectum.This maneuver avoids sepration of urinary tract from genital tract and reduces oprating time by more than 60 %. The functional and cosmetic results are also excellent.

Methods:

We retrospectively evaluate the short-term surgical results of feminizing genitoplasty with total mobilization of the urogenital sinus in patients with Cloaca and CAH. Review of medical records of all patients undergoing feminizing genitoplasty with mobilization of the urogenital sinus.

We evaluated the rates of complications from surgery and of urinary incontinence, as well as cosmetic results, according to the opinion of the surgeon and the family.

Results:

A total of 8 patients were included in the study. The mean age at surgery was 51months. CAH was diagnosed in six patients and Cloaca in the other two. The vagina was separated from the urethra, with suitable distance in all cases. No patient had urinary incontinence after surgery. The mean follow-up of patients was. 20 months (3-56 months). In all cases, surgeons recorded being satisfied with the aesthetic result of post-surgical genitalia. The family was recorded as satisfied with the aesthetic result of the genitalia after surgery. In every case, there was no need for a second surgical procedure.

Conclusion:

The total mobilization of the urogenital sinus is a surgical advace and feasible and safe technique. The technique permits good cosmetic results, and urinary incontinence is absent.

Colonic Atresia – A Case Report

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Background:

Colonic atresia (CA) is a rare cause of congenital bowel obstruction, accounting for 5−15% of all cases of bowel atresia in newborns. Various incidences had been reported, ranging from 1 in 1500 to 1 in 66.000 live births.

CA presents from birth with typical features of obstruction: abdominal distention, bilious vomiting and failure to pass meconium. Plain X-rays typically show dilated loops of proximal bowel. The diagnosis is made following contrast enema studies showing a narrowed distal colon coming to an abrupt halt at the level of the atresia.

Operative management of CA has been modified over the years. Earlier studies recommended resection with primary anastomosis for atresia proximal to the splenic flexure, and colostomy with delayed anastomosis for atresia distal to this point. In recent years surgical technique, whether to proceed with ostomy in all cases or to individualize the operative procedure to the location of the atresia, and the condition of the patient are still controversial.

Although, primary anastomosis may be technically difficult because of the very large discrepancy between the sizes of the distal and proximal bowels, and the loss of length associated with resection of the dilated proximal colon, some authors have advocated the use of resection and primary anastomosis, regardless of the location of the atresia.

Case presentation:

Our patient was a neonate who was diagnosed as colonic atresia trough contrast enema and clinical presentation. A type l atresia of descending colon was found at laparotomy.

Food Allergy and Surgical Gastrointestinal Diseases

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Several studies have reported an association between surgical gastrointestinal disease (such as intestinal obstruction or perforation achalasia) and food allergy which the most cited one is cow's milk protein allergy.

Eosinophilic esophagitis, gastroenteritis (EGID), and colitis, refer to a spectrum of clinical diseases that present with variable degrees of infiltration of the gastrointestinal tract by eosinophils in the absence of other known causes of tissue eosinophilia. EGID more recently has become well recognized and is going to be a growing problem.

EGID is a heterogeneous disease with respect to its clinical presentation. Clinical findings may reflect the extent, location and depth of the eosinophil infiltration in the digestive organs. All parts of the digestive tract may be affected; esophagus to rectum, even eosinophilic infiltration of the bile ducts has been reported previously. The presentation of Eosinophilic colitis, as with EGE, tends to depend on which intestinal layer is most affected by the eosinophilic infiltration.

EoE is described as a constellation of esophageal dysfunction, notably dysphagia, with an eosinophil-predominant infiltrate seen on histology Food impactions are common and may be the presenting symptom. Other symptoms include reflux, retrosternal pain, and abdominal pain in adults.Symptoms that are unresponsive to medical or surgical treatments for gastroesophageal reflux disease (GERD) EC (eosinophilic colitis) may have similar features to that of colon carcinoma on barium enema, and care must be taken with radiographic interpretation of this test, especially in the elderly population.

Patients most commonly report abdominal pain but may also have nausea, vomiting, poor appetite, weight loss, and diarrhea at the time of diagnosis. Patients with EGE may present with varying clinical symptoms based on location and the depth of eosinophilic infiltration of the bowel wall. The classification previously described by Klein et al. divides EGE into three distinct types: mucosal, muscular, and subserosal.

The mucosal subtype is the most commonly reported form of EGE possibly due to the ease of obtaining mucosal tissue samples during routine endoscopy, patients often present with vague, non-specific symptoms of abdominal pain, nausea, diarrhea, and vomiting which may mimic other gastrointestinal diseases. Some patients may present with more severe symptoms related to blood loss in stools, iron deficiency anemia, malabsorption, or a protein-losing enteropathy.

The muscular subtype, the second most commonly reported form, is characterized by infiltration of eosinophils predominantly in the muscle layer, causing bowel wall thickening and, in turn, symptoms of

intestinal obstruction. Gastric outlet obstruction, caused by distal stomach and pyloric submucosal infiltration, has been reported in the adult population. In infants, it may mimic pyloric stenosis. In patients with EC affecting the left colon, obstructive colo-colonic intussusception has been reported.

Serosal form or eosinophilic ascitis: is the rarest presentationof EGE in which eosinophil-rich inflammatory infiltrate permeates all layers of the digestive wall, reaching the serosal cover and causing the appearance of eosinophilic ascitis. There are reports of some EGE patients presenting with intestinal perforation.

Patients mostly have positive skin allergen test to a variety of food substances without any anaphylactic reaction. The immune response to various food allergens in the setting of EGE is considered to be a delayed type of hypersensitivity reaction. Eosinophils are the key cells implicated in the pathogenesis of EGE and EC.

Food protein-induced enterocolitis syndrome (FPIES) is another entity in children which is a noneimmunoglobulin E (IgE)-mediated allergic disorder triggered by the ingestion of food, that mimics surgical condition and usually presents within the first 6 to 12 months of life with some nonspecific symptoms along a spectrum of severity: repeated debilitating vomiting is typical and often accompanied by diarrhea, and in more severe cases hypotension can develop, manifesting as signs of shock. The symptoms resolve once the offending food protein has been removed from the diet and reoccur on re-exposure.

Gynecologic Concerns in patients Born with Anorectal Malformations: Experience in 32 cases

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In newborn with anorectal malformation, determination of gynecologic abnormalities is not urgent because bowel and urologic problems can be serious or even life threatening. Delineating the reproductive anatomy can be very important to prevent problems in the newborn period or during the puberty.

The internal female genitals: uterus, fallopian tubes and upper part of vagina develop simultaneous with urologic and GI systems. The anomalies of GI and urologic systems could also affect the re productive system.

The mullerian ducts exist on either side of the embryo and migrate down into the pelvis. The ducts meet tog hater in the midline and fuse to form one uterus. In patients with an abnormality in development, one hemi uterus with an attached fallopian tube may develop in each side. Canalization occurs to create the central portion of the uterus, the cervical canals and the vagina. Finally, the endometrium grows, which complete uterine development. The menses requires an open tract to avoid blockage of flow. At the time of puberty and menarche, the patency of the reproductive structures should be evaluated.

Sixty percent of female patients with cloaca have some degree of septation (division or separation) of the uterus and/or the vagina. It can be minimal such as a partial septum or a partial division within the vagina, or it can be much more significant with a duplicated vagina and double uterus and cervix (two hemi uteri).

Thirty percent of patients with cloaca suffer from Hydrocolpos. Most of patients with Hydrocolpos have duplicate mullerian systems (two hemi uteri and two hemi vagina). This septation disorder may be partial or total, symmetric or asymmetric. In the asymmetric type there is unilateral atresia of the mullerian system. When this goes unrecognized, it may produce accumulation of menstrual fluid at the age of puberty as well as retrograde menstruation.

Genital tract anomalies are less common in conditions such as perineal and vestibular fistula. Vaginal septa occur in patients with perineal and vestibular fistula in about three percent of cases. These septa may be removed during initial surgery.

In our experience there are 32 cases of cloacal type of ARM in all of the internal genitalia is abnormal. Twenty of these have 1-3cm common Chanel. In these we repaired the anomaly using the posterior sagittal approach and TUM without the opening the abdomen. Two cases had no uterus and

vagina. Until puberty the patients have no gynecologic problem. If the patients develops pubic and axillary hair and the breast start to develop but no menstruation, one must suspect that there is some sort of obstruction or absence of one or both of the mullerian structures.4 patients of our series required emergency laparotomy due to trapped menstrual blood in the peritoneum which formed pseudocysts. Now it is our routine to check the mullerian duct patency by 3fr feeding tube and injection of saline to confirm it comes out through the vagina. Tubo-ovarian abscess and tubal pregnancy are reported in these patients.

Vaginal septum with two hemi-uterus occurred in 2% of perineal fistula and 6% of vestibular fistula and 60% of cloaca. Malignancy in cloacal remnant and ulcerative colitis in neo-vagina have been reported.

It is imperative to accurately determine the reproductive anatomy. The initial time to accomplish this is during the definitive surgical repair. The reproductive structures can be visualized and the patency confirmed if the surgical repair requires an incision on the abdomen. If the child has a colostomy, the colostomy closure is the next opportunity to be able to assess the internal gynecologic structures.

The time between breast development and the onset of the periods is the ideal time for evaluation. It is easiest to consider the reproductive concerns chronologically in a girl or young woman" life. Hydrocolpos is more likely to occur in patients with a complex cloacal anomaly. The absence of menses can occur in a significant number of patients with cloacal anomalies due to the absence of a uterus or an under-developed uterus. It is recommended to undergo an ultrasound of the pelvis within about 6 months after onset of breast development. The patency of the reproductive tract is essential for menstrual blood to drain.

Gynecologic concerns are particularly important at several intervals: Definition of reproductive anatomy provides the key to appropriate counseling and preparation.

ACE in Patients with ARM: Experience in 26 cases

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The bowel management program, consisting mainly the administration of enemas and treatment with specific medication and specific diet, greatly changes the quality of life of many children with ARM. Giving an enema to a small child is a relatively easy task. In older children who needs enema for a long time or on a permanent basis, it becomes logistically more difficult. Taking the advantage of gravity, the enema in a teenager is very messy. Sometimes the patient squat on the tub to give themselves the enema. Teenager usually need to be assisted by another person to receive enema.

Malone, Ransley and Kiely had a great idea of creating a mechanism for the administration of enema in an ante grade fashion. They thought that an enema could be given through an orifice created in the abdominal wall, connected to the appendix, which allowed the passage of a small catheter directly into the colon to give the enema awhile the patient is sitting on the toilet. The original idea of Malone, Ransley and Kiely included a technique consisting of dividing the base of the appendix, rotating the appendix 180, and implanting it in the wall of the cecum, in a submucosal fashion, to avoid leakage of stool through the appendix. In retrospect, this represent a complex maneuver that we now believe it is unnecessary. The basic idea of Malone and Kiely is still extremely valuable, but now we know that we do not have to go through all the surgical maneuvers they described. They also advocated the opening of the orifice in the right lower quadrant, which something that we do not do at the present time: we prefer the umbilicus.

Our goal is to perform an operation that will allow the patient to have an active life, play sports, swim and not to be embarrassed of an orifice that rather looks like a colostomy or to have to hide a plastic device protruding from the abdomen.

We think that we should use the appendix as much as possible. Yet about 30% of the patients in whom we plan to do this operation have no appendix because it had been removed in the past. In these cases we make the appendix from a flap of the colonic wall or small bowel wall (Neo-appendicostomy). Sometimes we obligated to create the new appendix from the ascending colon near the cecum but away from the ileocecal valve.

It is very important to know the appendix has not be severed. Some radiologists believe that an ultrasound is a reliable study about the presence or absence of appendix. A CT scan is more reliable. Patient who have appendix, does not need colonic preparation to do the procedure. The creation of neo appendix from colon require full bowel preparation.

Continent appendicostomy and neo-appendicostmy have two main complications. Stricture of small stoma and leakage of the stool. The incidence of strictures in Pena series was 18%. Creation of triangular skin flap have resolved the problem. Leakage of stool from stoma is seen in 2.9% of cases despite the plication. In cases without plication the incidence of leak is 29% (Pena's experience).

We have experience of 26 cases of ACE including 18 appendicostomy and 8 cases of neo- appendicostomy (ascending colonic flap) in our patients. We have not leakage from the stomas which we have leave at RLQ of abdominal wall. We have not used the umbilicus as a stoma of ACE in our patients. In all of our patients we have plicated the colon wall around the appendix or neo-appendix to prevent the leakage. Those patients who do the enema from these stomas, all are satisfied from the job.

Anal Posterior Transposition in Patients with Anterior Ectopic Anus; A Novel Technique

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Background:

Anorectal malformations (ARM) are rare birth defects concerning the anus and rectum. Approximately 1 in 2500 to 1 in 5000 new born babies are affected. Anterior ectopic anus (AEA) is a common cause of constipation in children and a rare form of ARM. In the current study we present the outcome of our novel technique of anus posterior transposition for patients with AEA.

Methods:

This cross-sectional study was conducted in pediatric surgery department of Namazi Hospital; a tertiary healthcare center affiliated with Shiraz University of Medical Sciences, southern, Iran during a 4-year period from 2011 to 2016. We included those pediatric (<18 years) patients with AEA who underwent surgery in our center during the study period. The surgical procedure included performing a longitudinal skin incision and horizontal closure leading to posterior transposition of the anus without need for rectal displacement. The complication and outcome was evaluated after 1-year follow-up.

Results:

Overall we included a total number 5 patients with mean age of 19.8 ± 7.9 months. There were 3 (60.0%) male and 2 (40.0%) female among the patients. The operation duration was 22.3 ± 5.9 minutes and the bleeding was 15.3 ± 3.6 mL. None of the patients experienced surgery complication and only 1 (20.0%) developed postoperative infection which was treated conservatively. None of the patients developed anal stenosis and constipation after a 1-years follow-up. All the patients had normal stool diameter.

Conclusions:

Posterior transposition of the anus is a safe and feasible procedure for those with AEA. This is a simple procedure being associated with acceptable outcome and minimal complications.

Keywords: Anterior Ectopic Anus (AEA); Posterior Transposition; Outcome

Intestinal dysmotility after surgery in Hirschsprung disease

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Objective:

To describe the case of an infant with Hirschsprung disease presenting as total colonic aganglionosis, which, after surgical resection of the aganglionic segment persisted with irreversible functional intestinal obstruction; discuss the difficulties in managing this form of congenital aganglionosis and discuss a plausible pathogenetic mechanism for this case.

Case description:

The diagnosis of Hirschsprung disease presenting as total colonic aganglionosis was established in a 2-month-old infant, after an episode of enterocolitis, hypovolemic shock and severe malnutrition. After colonic resection, the patient did not recover intestinal motor function that would allow enteral feeding.

Postoperative examination of reminant ileum showed the presence of ganglionic plexus and a reduced number of interstitial cells of Cajal in the proximal bowel segments. At 12 months, the patient remains dependent on total parenteral nutrition.

Comments:

Hirschsprung disease presenting as total colonic aganglionosis has clinical and surgical characteristics that differentiate it from the classic forms, complicating the diagnosis and the clinical and surgical management. The postoperative course may be associated with permanent morbidity due to intestinal dysmotility. The numerical reduction or alteration of neural connections in the interstitial cells of Cajal may represent a possible physio pathological basis for the condition.

Key Words: Infant; Hirschsprung disease; Gastrointestinal motility

Anatomical and Physiological Analysis of Internal and External Anal Sphincter after Anorectal Anomaly Correction; A Single Center Experience

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Background:

Fecal incontinence is a common complication of anorectoplasty in patients with imperforated anus. Complete rectal displacement and sphincter muscle dysfunction are the most common causes of fecal incontinency in these patients. Currently, several methods are being used for determining the anal sphincter function. The aim of the current study was to the anatomical and physiologic status of the anal sphincter in those with imperforated anus undergoing anorectoplasty.

Methods:

This cross-sectional study was conducted during a 4-year period from 2009 to 2013 in pediatric surgery department of Namazi Hospital of Shiraz University of Medical Sciences. We included all the patients with imperforated anus who underwent three-stage anorectoplasty surgery. The patients were called and visited in the clinic at least 5 years after the surgery. They were evaluated regarding the rectal tone utilizing the Kelly scoring system, rectal manometry, endorectal sonography and magnetic resonance imaging (MRI) at least 6 months after the surgery.

Results:

Overall we included a total number of 20 patients with imperforated anus who underwent anorectoplasty during the study period. The mean age of the patients was 8.15 ± 1.63 years at the time of inclusion. The mean Kelly score was 2.85 ± 1.84. About 40% of the patients had poor sphincter function according to the Kelly score while 35% had moderate and 25% good anal sphincter function. The mean ASP and ARP was found to be 41.06 ± 19.11 and 9.31 ± 6.16 mmHg, respectively. Based on the Strack score of the sonography, 15% had good sphincter function and 25% had moderate function. In MRI, 80% of the patients had poor and moderate sphincter function. We found that anal manometry had the highest sensitivity and specificity in diagnosis of the anal dysfunction.

Conclusions:

The results of the current study demonstrate that anal sphincter manometry has the highest diagnostic accuracy in evaluating the anal sphincter dysfunction in those with imperforated anus undergoing anorectoplasty.

Key words: Imperforated anus; Rectal manometry; Endorectal sonography; Magnetic resonance imaging (MRI); Diagnostic Accuracy

Outcome of Anterior Sagittal Anorectoplasty in Females with Perineal or Vestibular Fistula; A Single Center Experience from Iran

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Background:

Anterior sagittal anorectoplasty is a standardized operative treatment for females with congenital rectoperineal or vestibular fistula. However, the short- and long-term outcomes are not well-established. The aim of the current study was to evaluate the short- and long-term surgical outcome of anterior sagittal anorectoplasty in patients with congenital rectoperineal or vestibular fistula.

Methods:

This cross-sectional study was conducted in pediatric surgery department of Namazi hospital, a tertiary healthcare center affiliated with Shiraz University of Medical Sciences, southern, Iran during a 4-year period from 2012 to 2016. We included those female pediatric patients with congenital rectoperineal or vestibular fistulas who underwent anterior sagittal anorectoplasty. The medical records were reviewed to obtain the demographic information and the outcome variables.

Results:

Overall we included a total number 24 patients with mean age of 5.38 ± 2.23 years. The operation duration was 68.3 ± 15.3 minutes. Wound infection occurred in 3 (12.5%) patients. A total number of 19 (79%) patients had voluntary bowel movements after the operation. Only 6 (25.0%) patients had soiling once or twice per week. The constipation resistant to medical therapy was reported in 9 (37.5%) patients. Mucosal prolapse occurred in 3 patients (12.5%). There was no recurrence of fistula, anal stenosis or anterior displacement of the neorectum.

Conclusions:

The results of the current study demonstrate that anterior sagittal anorectoplasty is an effective and safe option for patients with anorectal malformation and congenital vestibular fistula.

Key Words: Anorectal fistula; Rectoperineal fistula; Anterior sagittal anorectoplasty; Recurrence; Outcome

Surgical treatment of high confluence urogenital sinus using the posterior sagittal transrectal approach: a useful strategy to optimize exposure and outcome

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Introduction:

Urogenital sinus (UGS) is a complex, challenging, and controversial surgical issue for the pediatric surgeon. It consists of a persistent communication of vagina and urinary tract join together as a single common channel (CC), reaching the perineum. It can be an isolated anomaly or associated with congenital adrenal hyperplasia [CAH], or cloacal anomaly. CAH with high vaginal confluence is a rare congenital anomaly, observed in only about 5% of patients CAH. Reconstruction of the vagina and external genitalia in such patients is quite challenging. Many surgeons believe that children with such a malformation should undergo staged or delayed reconstruction, so that vaginoplasty is done when the child is older. Early vaginoplasty thought to be difficult due to patient size and poor visualization, short vagina and difficult dissection. The posterior sagittal approach has been beneficial for acquiring exposure to high urogenital sinus anomalies while it requires splitting of the rectum and temporary colostomy. We report our experience and results of our patients' series with high confluence CAH who are approached by postero-sagittal technique.

Material and method:

We operated 7 patients with very high confluence CAH (1 trigonal opening, 4 bladder neck opening, 2 proximal urethra opening) via postero-sagittal approach during last 3 years. Mean common channel length was 5.7 mm. 3 cases underwent upfront colostomy while protective colostomy was done at the beginning of genitoplasty. Those who underwent single stage operation received an intensive bowel preparation from 3 days before surgery. All cases underwent primary cystoscopy for surgical planning. Conjugated estrogen ointment was prescribed for two cases about a month before surgery. Operation protocol consisted of colostomy diversion in supine position, cystoscopy and placement of intra vaginal and vesical Foley catheters in lithotomy position and finally definitive postero- sagittal dissection was performed in prone position. Anus and rectum were dissected in the mid line and bladder neck was approached posteriorly. Vaginal poach was identified by the guide of indwelled catheter and opened posteriorly. Vaginal opening to urogenital tract was identified and vagina detached from UG tract keeping a safe margin from bladder neck. Total vaginal mobilization was performed and we entered the pelvic cavity and release the uterus also and pull through the utero-vaginal complex into the perineum. The rectum and anus were repaired in anatomic layers and an external genitalia was reconstructed in the conventional manner in the lithotomy position.

Results:

Patients were followed postoperatively and colostomy closure was performed a month after feminizing surgery. Patients were followed bimonthly to assess the results specially rate of vaginal stenosis. In this article we will share our lessons learned during applying this new method.

Conclusion:

Postero-sagittal approach provide a generous exposure to common channel and as we approach posteriorly, the bladder neck remains safe but as the aggressive dissection and multiple operation is needed, we suggest this approach just for very high confluence cases that are not correctable with TUM or trans perineal (anterior approach).

Key words: Urogenital sinus; Postero-sagittal; vaginoplasty

Antegrade Colonic Enema Procedure for Fecal Incontinence

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Background:

One of the major problems in pediatric surgery patients is fecal incontinence. After evaluating these patients, most of them are intractable to conservative bowel management. ACE Procedure has been recommended in these patients. Here we present the efficacy of the procedure in 21 patients with fecal incontinence.

Methods:

The chart of 21 patients with fecal incontinence who had underwent ACE procedures were reviewed. There were 6 patients with anorectal malformation, 4 with sacral agenesis, 3 Hirschsprung disease (post pull through) and 8 with intractable constipation and soilage. 9 patients had multiple operations. All of these patients had undergone evaluation with anorectal manometry, endorectal ultrasound and rectal mucosal biopsy. Bowel management were performed with some improvement. After discussing with the parents, ACE was recommended for these patients.

Results:

There were 7 girls and 14 boys. The age at ACE operation were 6-14 years. Mean age of was 9.2 year. Four patients had Laparotomy and 17 underwent Laparoscopy. Appendix was used in 19 patients, tubes were made from cecum in one patient and transverse colon in another. Umbilicus was used in 15 patients and in 6 patients the exit was in RLQ. Two patients developed adhesion band obstruction post operatively which responded to conservative treatment. Two patients had stricture at site of appendicostomy and needed re-operation for stricturoplasty. The patients were called and 10 patients answered. The follow-up was 1-5 years in these ten patients. Nine patients were happy with the operation, but one was unhappy and stopped irrigation. 5 patients did the irrigation once a day and were clean with no soilage. Two patients performed the irrigation every other day. Two patients still had occasional soilage. One patient had occasional leakage from the umbilicus. Two patients had stopped the irrigation and were well continent.

Conclusions:

Laparoscopy ACE procedure is simple and effective. Educating how to irrigate through ACE is very important. The complication and failure rates are low. Most of the patients do not need any anti-reflux operation and simple appendicostomy through umbilicus is effective.

Key Words: Antegrade colonic edema; Fecal incontinence; Pediatric surgery

Outcome of Anorectal Malformations Surgery during 10 Years

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Background & Purpose:

Anorectal malformations are among the most common disorders of children. The prevalence is 1 over 5000 alive birth. These anomalies are accompanied other congenital disorders in 50 to 60% of cases. In this study, we evaluated the early and late outcome of surgical treatment of patients with imperforate anus during 10 years.

Materials& Methods:

In a cross- sectional study, children referred to Al-Zahra hospital for imperforate anus who treated by surgical intervention were evaluated. Data were collected by hospital data sheet and re-examination of patients. In some cases follow up done by dialing the parents.

Results:

One hundred forty two patients were studied (83 boys, 59 girls). In 72.5 % of patients imperforate anus was high type. 69% of patients had fistula. Mortality rate of patients was 15.5% (22 cases) .All of expired cases had high type anomaly.

The most common type of operation was PSARP (40. 8%). Constipation was the most common early complication (20.4%).

Rectal prolepses were the most rare complication (4.6%). Constipation (12.5%) and incontinence (10.5%) were most common late complications. Late complications were more common in high type imperforate anus.

Conclusion:

Mortality and late complications were more common in high type imperforate anus, in-patient with associated anomalies and in them with major procedures.

Key Words: Imperforate anus, associated anomalies, Complication

Hirschsprung disease in adult

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Abstract:

Adult Hirschsprung disease (HD) is a rare motor disorder of the gut that is frequently misdiagnosed as refractory constipation.

The primary pathogenic defect in adult HD is identical to that seen in infancy or childhood, and is characterized by the total absence of intramural ganglion cells of the submucosal (Meissner) and myenteric (Auerbach) neural plexuses in the affected segment of the bowel.

94% of HD cases are diagnosed before the patient reaches 5 years of age, however, on rare occasion, mild cases of HD may go undiagnosed until he or she reaches adulthood.

In this study, we describe 2 cases of adult HD with a history of longstanding recurrent constipation, relieved by laxatives with progressive abdominal distention, colicky pain or acute intestinal obstruction. Barium enema or CT scan revealed a grossly distended proximal large colon with fecal retention. Intraoperative frozen section biopsy was performed in all cases and showed aganglionosis of the stenotic segment and a normal distal rectum. The modified one-stage Martin-Duhamel or Rehbein's procedure is a feasible surgical option for treating cases of adult HD involving a segment or the entire bowel.

In all cases, patient symptoms were completely resolved after pull-through and there were no complications arising immediately post-surgery or at one-year follow-up.

Adult HD should be considered in the differential diagnosis of cases where adult patients present with chronic constipation or even acute intestinal obstruction.

Keywords: Adult, constipation, treatment, Hirschsprung disease

The diagnostic value of serum amylase in prediction of perforated appendicitis in children

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Background:

Recent studies indicate increased serum amylase in children with acute appendicitis. The objective of the current study was to determine the diagnostic accuracy of raised serum amylase level in prediction of perforated appendicitis in children with acute appendicitis.

Methods:

Children aged 3 to 15 years who were admitted and underwent appendectomy due to acute appendicitis in two referral university hospitals from 2012 to 2013 were included. Venous samples were obtained and serum amylase, lipase, ALT (alanine aminotransferase), AST (aspartate aminotransferase), and alkaline phosphatase were assayed.

Results:

There were 61 children with the diagnosis of acute appendicitis. Of this, 18 cases had perforated appendicitis and 43 had non-perforated appendicitis. Mean (±SD) serum amylase level was 69.2 (±28.9) mg/dL in perforated group and 29.9 (±11) mg/dL in non-perforated group ($P< 0.001$). At serum level of 46 mg/dL, serum amylase had a sensitivity of 89% and specificity of 100% in prediction of acute perforated appendicitis.

Conclusions:

The serum amylase level in perforated appendicitis was significantly higher compared to patients with non-perforated appendicitis. Serum amylase has good diagnostic value in prediction of perforated appendicitis.

Management of Hydrocolpos in cloacal Malformations

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Hydrocolpos in patients with persistent cloaca must be diagnosed and treated early in life for increasing the index of suspicion for hydrocolpos in patients with cloaca and to describe approaches for its treatment with the hope that errors in the management of hydrocolpos can be avoided. We reviewed papers of patients diagnosed with cloaca and management. Emphasis was placed on evaluating for the presence of hydrocolpos, type of drainage, and complications related to the persistence of the hydrocolpos. About one thired of patients have Hydrocolpos. If the hydrocolpos do not draine, Complications includes: multiple urinary tract infections (8), hydrocolpos infection (7), sepsis (7), failure to thrive (6), ruptured hydrocolpos (4), and development of hydronephrosis in previously normal kidneys. The hydrocolpos persisted or reaccumulated if the first modality be: vesicostomy, intermittent perineal catheterization, single aspiration, or plasty of the perineal orifice. Proper drainage of the hydrocolpos alone, with no urologic intervention, dramatically improved the hydronephrosis in some cases. Effective drainage of the hydrocolpos at birth is tube vaginostomy, tubeless vaginostomy, catheter placed and left in the vagina through cystoscopy. Preferred approach is a transabdominal indwelling vaginostomy tube at present that can be do with laparoscopic, cystoscopic- and colposcopic-assisted vaginostomy tube placement. The drainage of the hydrocolpos alone may dramatically improve the hydronephrosis, and therefore, suggested that only after the hydrocolpos is drained should a urological intervention be contemplated. I described vaginostomy tube placement with laparoscopic, cystoscopic- and colposcopic-assisted method.

Congenital Pouch Colon, A Case Report

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Abstract:

Congenital pouch colon (CPC) is an unusual abnormality in which a pouch-like dilatation of a shortened colon is associated with an anorectal malformation (ARM) as a rare variant. It is categorized into four subtypes (Types I–IV) based on the length of normal colon proximal to the colonic pouch. In males, the pouch usually terminates in a colovesical fistula just proximal to the bladder neck .For all subtypes of CPC, it is preferable to preserve a segment of the pouch by fashioning a narrow colonic tube for pull-through, the technique known as coloplasty or tubular colorraphy.

Case presentation:

A 16-hours-old newborn male, weighing 3180 g, who admitted in NICU presented to us with the complaints of absent anal opening and abdominal distension. He was delivered by cesarian section in 37 weeks of pregnancy because of preeclampsia. He was brought to our hospital and worked up as ARM. The general condition of the child was good, abdominal distension was present. There was no history of vomiting. The perineum was flat and he had hypospadias(distal shaft). There was no other spinal or muscular malformations. Echocardiogram was normal, ultrasonography of the abdomen showed bilateral normal kidneys and left ureter was dilated. X-ray of abdomen showed dilated colon (its diameter was 90 mm), indicating high ARM. With diagnosis of high ARM we did exploratory laparotomy with low midline incision. Intraoperatively, we found one dilated segments of the colon (pouch Typ II).

Ileum opens into a normal cecum that opens into pouch colon (9 cm diameter) The terminal ileum was located in left upper quadrant and distal closed pouch was near the bladder . The appendix was seen; tenea was absent on the dilated pouch. ladd` s procedure, ileostomy and appendectomy were done.

After13 month when patient was gained weight adequatly,we did coloplasty by using stapler and then pullthrough and anorectoplasty with this new colon was performed. Two month later distal cologram was done and ostomy was closed and no complication occured but we can not assess fecal continency because of child`s age.

Avoidance and Management of Stomal Complications

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Introduction:

The construction of an intestinal stoma is fraught with complications and should not be considered a trivial undertaking. Serious complications requiring immediate reoperations can occur, as can minor problems that will subject the patient to daily and nightly distress. Intestinal stomas undoubtedly will dramatically change lifestyles; patients will experience physiologic and psychologic detriment with stoma-related problems, however minor they may seem. There are estimated to be approximately 120,000 people at any one time with stomas in the UK .Approx 1000 Ireland per yearAverage 175 new stomas each year at Beaumont Hospital. In general, postoperative stomal complications are a relatively frequent source of morbidity. Early complications are considered those that present within 30 days of surgery; late complications occur after 30 days.

Common complications:

Common complications include poor stoma siting, high output, skin irritation, ischemia, retraction, parastomal hernia (PH), and prolapse. Surgeons should be cognizant of these complications before, during, and after stoma creation, and adequate measures should be taken to avoid them. In this review, the authors highlight these often seen problems and discuss management and prevention strategies.

Goal of good stoma management:

To maintain healthy peristomal skin integrity. Skin around the stoma should be clean, dry and intact with no significant difference between peristomal skin and the remainder of the healthy abdominal skin.

Risk Factors:

Many risk factors that predispose a patient to develop complications have been proposed, including patient-, operation-, and disease-specific issues. Commonly reported patient-specific parameters include age, gender, body mass index (BMI) and nutritional status.

Preoperative Siting/Poor Siting:

A poorly sited stoma usually does not manifest its true degree of morbidity on patients.When stomas are placed in unfavorable locations, ill fit, leakage of effluent and gas, skin irritation, trauma, and poor visualization of the stoma can result. The stoma should be brought through the rectus abdominis muscle.

Patient Education:

The evaluation of the patient in consultation with a(wound ostomy continence)WOC nurse can significantly benefit the patient. Ideally, education, marking, and discussion of expectations should be performed preoperatively, as the involvement of skilled WOC nursing has been shown to be beneficial.

An adequate budget for education and dedicated patient education personnel are necessary for effective perioperative teaching and counseling of ostomy patients.

Conclusion:

In today’s health-care system, patients who require a stoma need access to outpatient preoperative and postoperative training and emotional support.

How Can Physical Therapy Help For Fecal Incontinence after an Anorectal Repair Surgery?

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Background:

Fecal incontinence is recurrent uncontrolled passage of gas, liquid, or solid stool and also passive incontinence, urge incontinence or faecal seepage. 30% - 56% patients have significant fecal soiling after surgery. Pelvic Floor Muscles (PFM) Support the abdominal & pelvic contents and Control bowel & bladder function and also Counteract changes in abdominal pressure and Maintain continence. So it can be an effective part in treatment of incontinence.

Methods:

Physiotherapy can help FI patients after anorectal repair surgeries by Biofeedback, Electromyography (EMG), Peri-anal or Intra-anal, Electrical Stimulation (ES) and Pelvic Floor Muscles Training (PFMT). Aims of Physiotherapy Management for Faecal Incontinence include Improvement of strength & endurance of pelvic floor muscles, Educating coordination of pelvic floor muscles, Improvment of control of sphincters, Training of faecal-continence function, Improvement awareness, Bowel habit re-education, Life-style modification, coping strategies and skin care, Psychological and emotional support, Improvement social life and quality of life.

Results:

Improvement in Overall Bowel Function and patient satisfaction would be reached if the patient and parents cooperated with therapist.

Conclusions:

Improvement was maintained with home-based program.

Outcome of Children operated for congenital anorectal malformations: A prospective single center study

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Background:

Anorectal malformations (ARM) occur in approximately 1 in 5000 live births and affects males and females almost equally. Operative correction of pediatric ARM is of potential clinical interest; however, long-term outcome of patients in respect to probable complications requires precise follow up and surveillance. The aim of the present study was to assess the outcomes of children undergoing surgical correction of ARMs.

Methods:

In a prospective follow-up study, we wanted to assess occurrences of incontinence, constipation, soiling, abdominal distension, diarrhea, stenosis, dilated sphincter and failure to thrive (FTT) in ARM patients. In addition, management of these conditions has been discussed. Reoperations have also been reviewed. The primary outcome of the study was determination of occurrence of incontinence at follow-up visits. Secondary outcomes were occurrence of constipation, anal stenosis, soiling, abdominal distension, dilated sphincter, diarrhea and FTT at follow-up visits. In addition, the decision of research team on patients at follow-up visits was considered as secondary outcomes.

Results:

Two hundred ninety patients were studied. Of the study patients, 174 children (60.4%) were boys and 114 children (39.6%) were girls. Mean age of boys was 4.8 ± 2.0 years of age and mean age of girls was 5.0 ± 2.0 years of age. The mean follow-up period of patients was 39.5 ± 29.1 months. During the study follow-up period, 63 patients (21.7%) had complications. The most common complication was constipation. It was present in 21 patients (33.3%). Soiling, incontinence, dilated sphincter, FTT, stenosis, abdominal distension and diarrhea were present in 21 (33.3%), 11 (17.5%), 9 (14.3%), 6 (9.5%), 6 (9.5%), 5 (7.9%), 3 (4.8%) and 2 (3.3%) patients respectively.

Conclusions:

We found that the commonest complications following ARM surgery are constipation, soiling, incontinence, dilated sphincter, FTT, stenosis, distension and diarrhea. The overall complication rate was 21.7%. 7.2 % and 3.1% of study population experienced constipation and incontinency respectively. 3.1% of study population required reoperation. We revealed that outcome of surgical correction of ARMs is considerably good and complication rates are acceptable. Continence rate was acceptable.