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Rectal Atresia

Colon and rectal atresia are very rare forms of bowel atresia, a congenital anomaly that results in complete blockade of the intestinal lumen. Rectal atresia (RA) is a congenital anomaly characterized by a blind-ending rectum. It is believed to arise because of an intrauterine vascular accident during fetal development. Prenatal environmental risk factors suggested to cause increased risk for developing rectal atresia include paternal smoking, maternal overweight obesity, and diabetes. The anal opening is anatomically normally positioned, reason why RA is considered separate from an anorectal malformation. Rectal stenosis refers to an anomaly where the rectum is narrowed but maintains a lumen that connects the anus and the more proximal colon. Rectal atresia occurs in 0.3-1.2% of all anorectal malformations with a male to female predominance of 7:3. RA is divided into five types. Type I consist of rectal stenosis, with type I-A being a stenotic segment, and I-B being a web with a hole. Type II there is rectal atresia with a septal defect. Type III, being the most common type, involves rectal atresia with a fibrous cord between the two ends. In Type IV rectal atresia there is a gap between the two segments. Type V is subdivided into three, with V-A being rectal atresia with stenosis, type V-B multiple rectal atresia, and type V-C has thickened Houston's valve and multiple rectal stenosis. In rectal atresia the pelvic structures are well developed. The anal canal, external sphincter, internal sphincter, and genitals are normal and well-developed. The proximal rectal pouch usually does not have fistulous connection with the urinary or genital tract. The diagnosis of rectal atresia is suspected when a temperature probe is unable to be inserted through the rectum. A rectal exam confirms the diagnosis of a blind ending rectum. Inability to insert a firm rectal tube, thermometer, or blunt Hegar dilator for more than 1 to 3 cm (mean 2 cm) from the anal skin is a clear indication of this malformation. The baby develops progressive abdominal distension, bilious vomiting, and failure to pass meconium. The diagnosis of a rectal stenosis is delayed since bowel movement are present. A contrast study is needed to diagnose a rectal stenosis showing a string of contrast passing into a larger proximal dilated rectum. It is recommended to perform an echocardiogram, spinal and renal ultrasound, along with MRI of the spine to check for associated malformations. Differential diagnosis of RA includes Hirschsprung disease, small bowel atresia, colonic atresia, small left colon syndrome, neuronal intestinal dysplasia, meconium plug and meconium ileus. A presacral mass is the most common associated condition with rectal atresia (30%). They could represent a presacral teratoma or anterior sacral meningocele. The mass should be resected during the operative procedure to repair the rectal atresia. Tethered cord syndrome can also be associated with RA. Management include diversion colostomy followed by planned repair. A distal colostogram before reconstruction is critical to confirm

both the distance between the distal rectal pouch and the anal canal, as well as to verify the absence of a fistula with the urinary or genital tract. The goal of reconstruction is to achieve continence and normal bowel functionality by preserving the anal canal and sphincter mechanism. Two of the most common utilized approaches include posterior sagittal anorectoplasty (PSARP), or an endorectal-transanal pull through approach. With an PSARP a posterior sagittal incision is performed from coccyx to the posterior edge of the anus. The proximal bowel is dissected, mobilized and an anastomosis is performed with the distal rectum. Preserving the anterior anal canal avoids damage to the anterior rectal wall, dentate line, and sphincters. Regular dilatations will be needed before colostomy closure. In the transanal approach a submucosal dissection is done proximally in the atretic distal rectum, the proximal rectum mobilized and pull-through creating a new anastomosis. Alternatively, an endoscope can be utilized proximally into the rectum through the colostomy and the light utilized to guide the resection and dissection of the proximal rectum. The prognosis of repair of rectal atresia is good with adequate continence and good bowel control in most cases.

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Santulli Enterostomy

In 1961, Santulli and Blanc introduced a side to end anastomosis with a proximal end enterostomy to effectively decompress the proximal bowel and permit early instillation of nutrients into the distal bowel in babies born with intestinal atresia. In the Santulli enterostomy, the proximal afferent bowel is fashioned into a stoma and anastomosed side-to-end into the distal efferent bowel. As a critical fact, the intestinal segment from the anastomosis to the skin (distal end of the afferent bowel) is constructed no more than six cm to avoid a blind loop after closure. The distal bowel is checked for patency with a water-soluble contrast enema prior to closure of the ileostomy. Bowel continuity occurs relatively rapid, can be monitored while the enterostomy acts as a safety valve in the case of distal or anastomotic obstruction. The Santulli procedure was initially described for management of intestinal atresia as a promising alternative given the high rate of complications, such s malfunction or leakage, seen in primary anastomotic attempts. Avoiding complete diversion of the distal bowel (small intestine and colon) is highlighted to further enhance intestinal motility and colonic function. The choice of using the Santulli enterostomy is made by the surgeon either because of a lasting discrepancy (greater than four to one) between bowel

segments, or when the aspect of the distal bowel part of the bowel is not deemed satisfactory enough to perform an anastomosis. The Santulli enterostomy can be used for intestinal atresia, meconium ileus, midgut volvulus, necrotizing enterocolitis, multiple intestinal atresia, and colonic atresia. The Santulli enterostomy can be used to managed NEC even in very small premature infants with good overall results. It enables rapid access of the intestinal contents into the distal bowel and may promote enteral feeding and early stomal closure enabling preservation of sufficient intestinal length to avoid short bowel syndrome. The onset of stools passing through the anus after the Santulli procedure is noted approximately ten days after construction. Once anal stooling is passing, and the proximal effluent of the stoma decreases the Santulli enterostomy can be closed. Closure of the Santulli enterostomy is simple and requires very little additional resection of bowel. Though Santulli originally suggested extraperitoneal closure under local anesthesia, most of these closures are performed under general anesthesia. In meconium ileus the Santulli enterostomy has a better effect on ileostomy output as compared to loop ileostomy method. The rate of surgical complications and hospitalizations are significant lower in Santulli ileostomy as compared to loop ileostomy. The Santulli ileostomy also gives best cosmetic results with minimal complications and better that loop ileostomy method for management of uncomplicated meconium ileus. The Santulli enterostomy avoids the non-use of the distal bowel, especially the colon, restoring the enterohepatic circulation, preserving intestinal microbiota, avoiding diversion colitis, and reducing the risk of cholestasis, sodium depletion and metabolic acidosis seen in short bowel syndrome. The Santulli enterostomy can be used as a first-choice surgery any time there is risk of primary anastomotic disruption, or to manage a complicated former double enterostomy. The Santulli enterostomy has also been utilized in adult's patients for complicated hernias with intestinal necrosis and resection, complicated colorectal surgery that needs diverting enterostomy, mesenteric ischemic disorders which cannot be managed by primary anastomosis, trauma patients with intestinal discontinuity that needs a diverting enterostomy and even patients with gynecologic malignancies with spread to the gastrointestinal tract and need revision. Major intraabdominal contamination that will put the anastomosis in jeopardy or critically ill patients who will not be able to tolerate a longer anesthesia may be contraindications for using the Santulli enterostomy procedure. Recently, the Santulli procedure was utilized effectively in symptomatic children with immature ganglion cells proven by full-thickness pathological biopsy.

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Hirschsprung's Disease and IBD

Hirschsprung's disease (HD) results from the absence of ganglion cell in the distal bowel described most commonly as classic if the rectosigmoid is affected to total colonic aganglionosis in very few cases (10%). Most diagnostic work-up and removal of aganglionic segment with pull-through reconstruction in children with HD is undertaken within the first year of life. Following surgical correction of HD some children (5-42%) develop an associated enterocolitis (HAEC) characterized by intestinal inflammation resulting in abdominal distension, fever, diarrhea, and sepsis. Inflammatory bowel disease (IBD) refers to describe conditions such as Crohn disease and ulcerative colitis causing symptoms that include diarrhea, pain, obstruction, weight loss, bloody stool, and fistula formation. Several reported cases have raised the possibility that HD and IBD may coexist, having a similar etiology and representing a spectrum of intestinal inflammatory disease in children. HAEC and IBD have similar clinical presentation including diarrhea, hematochezia, and abdominal pain. Both conditions are characterized by an abnormal intestinal mucosal barrier function. Less than 100 children with HD associated with IBD have been reported. Mean age at diagnosis of IBD was 7.7 years, most were males (73%), long-segment disease and total colonic aganglionosis predominated in 86% of all patients, with Duhamel procedure leading the cases (84%). Most IBD cases associated with HD were Crohn disease. An increasing length of aganglionosis is associated with a higher risk of HAEC. Individuals with HD have an increased risk to be diagnosed with IBD later in life, though the underlying cause is unknown. In Canada, being diagnosed with HD resulted in a 12-fold increased risk of subsequently being diagnosed with IBD. Persons with IBD were 24-40 times more likely to have had HD than matched control. Age at IBD onset is similar in individuals with HD and the general population with a median age of 21 years. The diagnosis of IBD in children with HD is difficult to confirm resulting in a delay. Crohn disease is the most common subtype of IBD found within this association. HD is more common in persons with trisomy 21, and trisomy 21 is a risk factor for developing HAEC. Trisomy 21 is not typically associated with IBD. IBD can emerge in more than 2% of children with HD and is more frequently classified as Crohn's disease than ulcerative colitis. Post-HD IBD is 3-fold more common in males. Management of IBD in these children is surgical in 30% and medical in the rest. Medical management include a combination of biologic agents (infliximab), methotrexate and steroids with mixed results. Genetics studies have identified nine hub genes associated with the co-occurrence of HD and Crohn disease. Among these genes, CXCL10 secreted by immune and non-immune cells is significantly higher in the Crohn disease and aganglionic segment of HD. Targeting CXCL10 could be an attractive approach to managing IBD. CXCL10 can be a potential biomarker for the development of Crohn disease in HD patients. These hub genes and diagnostic models will be beneficial to the prevention and diagnosis of postoperative concurrent Crohn disease after HD and provide a theoretical basis for the molecular mechanism of HD and Crohn disease co-occurrence.

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***Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP**
Professor of Pediatric Surgery, UPR - School of Medicine, UCC School of Medicine & Ponce School of Medicine.
Director - Pediatric Surgery, San Jorge Children's & Woman Hospital.
Postal Address: P.O. Box 10426, San Juan, Puerto Rico USA 00922-0426.
Tel (787) 340-1868 E-mail: *pediatricsurgerypr@gmail.net*
Internet: pedsurgeryupdate.com

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