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Brunner Gland Adenoma

Brunner gland adenoma (BGA), also known as an hamartoma, is a rare duodenal lesion comprising not more than 5% of benign duodenal lesions. Most Brunner gland adenomas are of small size without causing significant symptoms. When BGA grows, they can cause obstruction or bleeding. Brunner glands are submucosal mucin-secreting glands predominantly located in the posterior wall of the duodenal bulb and second portion of the duodenum segment and progressively decrease in size and number in the distal potions of the proximal bowel. Brunner's glands secrete an alkaline fluid composed of viscous mucin which protects the duodenal epithelium from acid chyme of the stomach. BGA are rarely larger than 2 cm in asymptomatic individuals, while larger than 5 cm in symptomatic patients. Brunner's gland could be classified into three types based on size: type 1 (diffuse nodular hyperplasia) confined to the mucosa with multiple sessile projections occupying most of the duodenum; type 2 (circumscript nodular hyperplasia), found in the bulb duodenum and usually smaller than 1 cm; and type 3 (Brunner's gland adenoma) stemmed with sized 1-2 cm, generally without clinical manifestations. Etiology of BGA is unknown. It is believed that gastric hypersecretion of acid results in hyperplasia of Brunner's gland resulting in adenoma formation and excrescence toward the bowel lumen. Others believe the loss of the alkaline protection of the exocrine pancreas leads to compensatory hyperplasia of Brunner's gland with increased production of mucus and alkali. Helicobacter infection has also been found culprit of BGA, though a clear relationship has not been met. It is believed BGA are hamartomas. The word adenoma might be a misnomer, since the mass is not a true neoplasm, but rather a hamartomatous or hyperplastic collection of mature glands with no known potential for malignant transformation. Patients with BGA are usually asymptomatic, or developed symptoms of nausea, vomiting, bloating, dyspepsia, vague abdominal pain, melena, or hematemesis. Pancreatitis (ampullary lesions), intussusception and diarrhea have also been reported. Those in the pylorus often presents with epigastric pain, dyspnea, or melena, whereas those in the posterior wall of duodenum often presents with postprandial fullness. When the adenoma grows it can cause symptoms of obstruction or gastrointestinal bleeding. Chronic bleeding with ulceration is found in most symptomatic patients. On rare occasion BGA can cause gastric outlet obstruction. The most common laboratory finding in symptomatic patients is anemia. Abdominal imaging (Ultrasound/CT-Scan/MRI) or endoscopy can detect the lesion. Upper endoscopy localizes the lesion and biopsy can provide the diagnosis of a BGA. Endoscopic biopsy shows involvement of the mucosa and submucosa layers without deeper extension, variable echogenicity, and cystic changes within the lesion. Cytologically, Brunner's gland shows loose clusters of flat, two-dimensional cells with minimal overlapping or atypia showing

abundant, finely granular, and vacuolated cytoplasm. In most cases, diagnosis is confirmed after endoscopic or surgical resection. Endoscopic resection is recommended to avoid developing symptoms with time. When endoscopic resection is not possible, surgical resection is indicated. Recurrence rate after both modalities of management is very low. BGA have been reported very rarely in the pediatric age and can be associated with surgical repair of duodenal atresia. BGA are benign and the prognosis after endoscopic or surgical management is good.

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Hirschsprung's Associated Enterocolitis (HAEC)

Hirschsprung's disease (HD) occurs in approximately one in 5000 live births with most babies presenting with failure to pass meconium in the first 24 hours of life. Failure to recognize HD early in infancy place them at high risk of developing Hirschsprung's associated enterocolitis (HAEC). HAEC is a serious life-threatening inflammatory complication of HD. HAEC occurs preoperatively in 6-26% of cases, and post pull-through surgery in 5-42%. HAEC is histologically characterized by cryptitis, the appearance of neutrophils in intestinal crypts. They progress to crypt abscess, mucosal ulceration and fibrinopurulent debris. In severe cases ischemia, transmural necrosis and perforation can occur leading to shock and hypoperfusion. Pathophysiologic mechanism associated with HAEC includes partial mechanical obstruction (bacterial translocation), unbalanced microflora (dysbiosis), insufficient immunoglobulin secretion, abnormal mucin production (impaired mucosal barrier), and dysfunction of the enteric nervous system. Mortality of HAEC ranges between 1% and 10%. Several factors contribute to an increased risk of HAEC, these include family history of HD, Down syndrome, long-segment HD involvement, obstruction from any cause (retained aganglionosis, transitional zone pull-through, dysmotility following pull-through, anastomotic stricture, twist in the pull-through or tight muscular cuff following surgery), and prior episodes of HAEC. Some authors believe no patient or clinical characteristic is associated with the risk of postoperative HAEC. The risk of developing HAEC increases with the length of the aganglionic segment and delay in diagnosis. The diagnosis is suspected when a child with constipation develops abdominal distension, pain, vomiting, explosive watery diarrhea, fever, lethargy, rectal bleeding, and shock. Initial images consist of plain radiographs which can demonstrate a cutoff sign in the rectosigmoid colon with absent distal air, dilated proximal bowel loops, air fluid levels,

pneumatosis intestinalis, "sawtooth' appearance with irregular intestinal lining, or pneumoperitoneum from bowel perforation. Barium enema and colonoscopy are contraindicated in the acute setting due to the risk of perforation, while CT-Scans are of little value in the diagnosis and treatment of HAEC. Chronic HAEC symptoms include persistent diarrhea, soiling, intermittent abdominal distension, and failure to thrive. Should this occur after surgical management mechanical obstruction from aganglionosis should be suspected and confirmed with rectal biopsy. The diagnosis of HD is made histologically by the absence of ganglion cells, presence of nerve hypertrophy and absent calretinin immunohistochemistry. For prophylactic prevention of HAEC routine rectal irrigation or diverting colostomy is indicated in selected patients. Rectal irrigations reduce fecal stasis and bacterial load, limiting colon distension. Children with HD and severe congenital heart disease should be diverted to avoid HAEC. Probiotics management has controversial results as a preventive measure. The clinical suspicion and severity of HAEC have been graded based on history, physical examination, imaging studies and laboratory findings similar to Bell's criteria for neonatal enterocolitis. Four or more of 16 criteria seen in the Table below are diagnostic. Such score helps make the correct diagnosis of HAEC. Existing scoring systems perform poorly in identifying episodes of HAEC, resulting in significant underdiagnosis. Management of HAEC includes fluid resuscitation using isotonic solutions, broad spectrum intravenous antibiotics, decompression of the gastrointestinal tract and bowel rest. Systemic antibiotics are used empirically in HAEC, with metronidazole typically chosen to managed anaerobes. Those children with severe sepsis and acutely ill (Grade III) benefit with admission to the intensive care unit some of them needing vasopressor therapy and ventilatory support. Rectal washouts with warm saline are the mainstem of management and they should be performed two to four times daily at a rate of 10-20 ml/kg of weight each time or until the effluent is clear. In newborns presenting with severe HAEC, shock and sepsis immediate diversion (leveling colostomy) should be considered. Other risk factor associated with the need of diversion includes delayed presentation, co-morbid conditions, presence of HAEC or multiple risk factors for HAEC in the preoperative period. Diversion improves patient symptoms but does not resolve HAEC development later in life. HAEC can also occur in the postoperative period after pull-through definitive surgery. Rectal washouts are also effective to manage postoperative HAEC. There is a trend toward a higher incidence of enterocolitis in the primary endorectal pull-through group as compared with those with a two-stage approach. Postoperative HAEC can occur more than 18 months after definitive surgery. The postoperative risk of developing HAEC after definitive surgery is highest in those operated with Swenson and Duhamel approach (20%), and lowest with Soave procedure (11%). Children with recurrent HAEC, occurring in 2-33% of patients, should be evaluated for anatomic or pathologic causes of obstruction. A water-soluble contrast enema can identify any mechanical cause of obstruction. Should the suspicion of persistent aganglionosis be considered a rectal biopsy should be performed. Risk factors for recurrent HAEC includes preoperative HAEC, history of central nervous system infection and congenital chromosome anomalies and does not include placement of an ostomy prior to pull-through and congenital cardiac anomalies. Recurrent postoperative HAEC does not have an impact on mortality. Children with previous episodes of HAEC are more likely to develop subsequent episodes.

Criterias used to score HAEC* - 4 or more are diagnostic

Distended abdomen
Diarrhea with explosive stools Diarrhea with foul smelling stools Lethargy
Explosive discharge of gas and stools on rectal exam
Fever
Dilated loops of bowel an X-ray films
Leukocytosis
Decreased peripheral perfusion
Multiple air-fluid levels
Previous history of suspected enterocolitis
Left shift on complete blood count
Diarrhea with bloody stools
Cutoff sign in rectosigmoid colon
Sawtooth appearance with irregular mucosal lining
Pneumatosis intestinalis

* J Pediatr Surg. 53(4):708-717, 2018

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Transanal Pull-Through

In 1998, De la Torre introduced a new one-stage surgical technique to manage Hirschsprung's disease (HD) where rectal mucosectomy, aganglionic segment colectomy, posterior myectomy, and normoganglionic colon pull-through is performed through the

anus. HD aganglionosis affects the rectosigmoid (classic HD; 85%), long-segment (10%) and total aganglionosis (5%) of the colon with sporadic cases affecting the proximal small bowel. The transanal pull-through (TAPT) is mainly design for children with classic HD and uses a prone approach, though it can also be performed in supine position. Advantages of the TAPT include abdominal or intraperitoneal bowel opening is not necessary unless the child has a previous leveling colostomy, risk of adhesion is decreased, excellent cosmetic results, earlier full oral feedings, shorter hospital stay, less costs, less pain, reduced operating time, endorectal dissection preserves the anorectal sphincters and their blood supply along with innervation causing less damage to fecal and urinary incontinence. Should the ganglionic bowel need further dissection proximally laparoscopy can provide this mobilization. The procedure can be performed in newborns with potential benefits of avoiding a colostomy and establishing colonic continuity early in life improving continence results. A stoma in HD is considered appropriate in children with bowel perforation (usually cecal), severe malnutrition, severe enterocolitis, very dilated proximal bowel, total colonic aganglionosis or lack of adequate pathological support. Should the surgeon choose to wait beyond the neonatal period for final correction of HD the risk of developing enterocolitis should be minimized by ensuring adequate decompression of the proximal dilated bowel by appropriate daily irrigations, administration of prophylactic metronidazole or probiotics, and use of breast feeding or elemental infant formula. Using laparoscopy concomitantly with the transanal approach can help in the proximal bowel dissection, distal bowel dissection and obtaining a seromuscular biopsy for frozen section to determine the ganglionic bowel to be pull-through. A frozen full-thickness biopsy helps the pathologist see both submucosa and myenteric plexus in look for ganglion cell and nerve hypertrophy minimizing error. Whether using a supine or prone position for the procedure depends on the certainty that you are dealing with classic HD and proximal mobilization will not be required. The prone position has the advantage that mesenteric vessels can be send and controlled more effectively than in the supine or lithotomy position. The transanal dissection can be started 0.5-1 cm above dentate line in neonates and 1-2 cm above the dentate line in older children as the transitional epithelium must not be damaged to avoid loss of sensation and incontinence. Dissection on the outside of the rectum, as that used during laparoscopic or open Swenson procedures, may increase the risk of injury to pelvic nerves and vessels, and to the prostate, urethra, and vagina. Comparison between complete TAPT and laparoscopic-assisted TAPT do not differ between rates of major complications, including leaks, strictures, enterocolitis, fecal incontinence, postoperative obstructive symptoms, and mean length of stay. Comparison between the Duhamel procedure and the TAPT have shown they are similar in respect postoperative fecal incontinence and operation time, but the Duhamel is associated with longer hospital stay and lower rate of enterocolitis. When performing the TAPT in neonates, a higher incidence of enterocolitis has been reported due to increased risk of sphincter spasm and anastomotic strictures. Excising the entire posterior rectal muscle cuff (myectomy) has been effective in reducing the incidence of enterocolitis. Current guidelines suggest doing the TAPT between two and three months of age if the child is growing well and the bowel is sufficiently decompressed. Whether performing the procedure totally transanally or laparoscopy assisted does not affect long-term bowel function. More than 20% of patients develop at least one complication within the 30-days following a TAPT. Older age at time of surgery increases the risk of developing a

postoperative complication. They include anastomotic leakage, abdominal abscess, and anastomotic strictures. Ischemia and increased tension on the colon anastomosis play an important role in leakage and stricture. This is one reason to consider perioperative diverting stoma in older patients. Older age at time of surgery, laparotomy-assisted and long segment disease increases the risk of developing a postoperative complication. Long term problems with pull-through surgery for HD include obstructive symptoms (30%), persistent constipation, soiling, enterocolitis and descending aganglionic bowel. Persistent bowel obstructive problems might be caused by mechanical obstruction, recurrent or acquired aganglionosis, disordered motility in the proximal colon, nonrelaxation of the internal anal sphincter, or stool-holding behavior. Mechanical obstruction might also be caused by an anastomotic stricture, twisted descended pull-through colon, or rolling of a long muscular cuff left behind. They are managed by sequential dilatations or redo pull-though surgery. Soiling can be caused by damage sphincter function (incontinence), abnormal sensation and pseudo-incontinence. Anorectal manometry is indicated to determine the cause. Soiling not associated with constipation is caused by true fecal incontinence and does not improve with time. Thus, to preserve the anal canal and avoid sphincter damage are of vital importance during the TAPT. Enterocolitis is managed with bowel rest, colonic irrigations, and systemic antibiotics. Most children have an excellent quality of life going into adulthood after TAPT. Besides the management of HD, the TAPT can be utilized for children born with rectal atresia, for severe chronic idiopathic constipation associated with megarectosigmoid, idiopathic rectal prolapse, and children with rectal prolapse after anorectal malformation correction.

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