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Total Colonic Aganglionosis

Hirschsprung's disease (HD) is characterized by bowel obstruction. Diagnosis is confirmed by finding aganglionosis of the inner-myenteric plexus with absent calretinin staining in rectal biopsy. Development results from abnormalities in the colonization of the enteric nervous system with faulty neuroblast migration during fetal life. Most cases involve aganglionosis of rectosigmoid.

Total colonic aganglionosis (TCA), with or without distal small bowel involvement, occurs in 8% (5-15%) of all HD cases, with a 1:1 gender involvement. Within this group, those with a transitional zone within 5 cm of the ileocecal valve are TCA, a transitional zone of more than five cm from ileocecal valve to 20 cm from ligament of Treitz is called small intestinal HD, and a transitional zone within 20 cm of the ligament of Treitz is recognized as total intestinal HD.

Digestive autonomy may be achieved in children who have at least 80 cm of remnant ganglionic small bowel. TCA involvement has significant morbidity and mortality than short segment HD requiring more complex medical and operative management. Improvement in management is associated with earlier diagnosis of TCA, earlier management of enterocolitis and improvement in supportive care.

TCA is regarded as a genetic, sex-modified, multifactorial condition with a variable severity and incomplete penetrance of a number of genes. Contrary to other belief, the use of the appendix for the diagnosis of TCA has a 100% sensitivity, specificity, positive predictive value, and negative predictive value, but should not be used as the sole diagnostic indicator of TCA, as the absence of calretinin corroborates the diagnosis. The appendix should not be removed completely, just the distal 1 cm, in case is needed later for bowel management (Malone).

The surgical treatment of TCA has two goals: remove the aganglionic bowel, and to provide children with a good quality of life, which is reflected in an acceptable frequency of bowel movements, fecal continence, and no symptoms of enterocolitis. Most children with TCA are managed with a diverting ileostomy with ganglion cells from diagnosis. These children with ileostomy struggle with growth and fluid losses and benefit from a comprehensive nutritional evaluation and optimization before a pull-through.

Prior to pull-through, the consistency of stool should thicken to an applesauce consistency. This can occur between 9 and 12 months after the ileostomy. Pectin and water-soluble fiber can increase the consistency and volume output. Loperamide is also utilized to increase consistency since it increases gastrointestinal transit time by reducing activity in both the

longitudinal and circular muscle fibers, as well as reducing secretions of gastric acid, bile and pancreatic enzymes thus reducing the luminal fluid content.

Proceeding with the pull-through for TCA in HD once the child is growing well and the stools have started to thicken is the best recommendation, the timing of which usually occurs sometimes between six and 18 months of life. Once ready for pull-through, the surgery is generally performed without a covering stoma, unless anatomic or technical decisions favor a proximal diversion.

There is no consensus on the preferred procedure to perform in TCA. Long term TCA children may only achieve a good outcome in 50-60% of cases. Reconstruction procedure for TCA encompass two groups: those that retain various lengths of aganglionic colon as a form of patch to improve absorptive capacities (Kimura, Martin, and Duhamel), and procedures that use small bowel ganglionic bowel such as the straight ileoanal (Swenson, Soave, transanal or laparoscopic) or J-pouches with short limb (4-5 cm) pull-through.

The most commonly performed surgical reconstructions in TCA are J-pouch with ileoanal anastomosis, straight ileoanal anastomosis and Duhamel. Contemporary experts' recommendations support straight ileoanal pull-through as a preferred conduit, which should be diverted if constructed in the newborn period. The Duhamel procedure has been popular for TCA so long as a spur is avoided to avoid postoperative obstructive symptoms.

The residual aganglionic segment of the rectum can result in constipation/fecalomas and incontinence. Recurrent or chronic enterocolitis and the need for intrasphincteric botulinum injections are less common after the Duhamel operation. The Martin procedure with a long side-to-side anastomosis of ganglionic ileum to aganglionic colon has been abandoned due to technical difficulty, a high complication rate and the need for multiple operations. The same has happened with the Kimura colonic patch procedure. The Swenson procedure leaves the least amount of aganglionic bowel behind but due to the low dissection, the sphincters can be damaged causing incontinence, or damage to other structures such as vagina and urethra.

Pena suggests an approach for TCA consisting of colectomy with straight ileoanal anastomosis and ileostomy at presentation in the newborn period. Closure of ileostomy is performed when the child is toilet trained for urine and willing to tolerate rectal irrigations. A covering enterostomy significantly reduced the incidence of postoperative complications such as anastomotic leak. The most frequent early complication among patients without a protective stoma is perineal rash.

Postoperative care must consider an aggressive approach to perineal skin care (cyanoacrylate-based liquids), optimization of stool content output with pectin/water-soluble fiber, avoidance of foods with high sugar content, sodium supplementation, and use of loperamide for frequent stools. Enterocolitis is overall the most common postop complication, followed by fecal incontinence and soiling. Over time, children with TCA or long-segment aganglionosis do not suffer from worse fecal incontinence in general. A

difference in stool consistency may underlie the association between liquid fecal incontinence and TCA and constipation in children with rectosigmoid aganglionosis. General quality of life is comparable on reaching adulthood in HD patients.

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Vaginal Reconstruction

Absence of the vagina in children is rare. Vaginal reconstruction in children is needed in patients with congenital agenesis of the vagina (aplasia of Mullerian ducts associated with the 46 XX Mayer-Rokitansky-Küster-Hauser syndrome), complete androgen insensitivity syndrome (46 XY testicular feminizing syndrome), congenital adrenogenital syndrome with an absent or hypoplastic vagina, after pelvic tumors, or posttraumatic injury. In all these cases, vaginal replacement is needed.

The proposed motivation for performing feminizing genitoplasty procedures earlier in life is to produce the best psychological outcome for the child by helping them to avoid stigma, optimizing future self-esteem and sexual satisfaction, and reducing psychological risk. The most common condition needing vaginal reconstruction is congenital adrenal hyperplasia (40%) with a median age of 2.4 years and almost 75% performed in patients under age 12.

Recent studies demonstrate a low 30-day postoperative complication (6%), readmission (7.9%), and reoperation (4%) rate after vaginoplasty in children. Surgically assigning anatomical gender before a patient has the capacity to make informed decisions regarding their own gender identity can be detrimental to psychological well-being. The goal of vaginal reconstruction surgery is a technique with very low morbidity and complications as well as no mortality. Its aim is the creation of a vagina that requires a simple and non-psychologically traumatic nursing that gives the possibility to perform coitus without pain, discomfort, and embarrassing secretions.

In children, basically three types of surgical procedures have been developed for vaginal reconstruction: the inlay skin-graft technique, the use of grafts (peritoneum, bladder buccal mucosa, amnion), and replacement using the bowel (colon or ileum). The split-thickness

skin graft is indicated only in cases of small reconstructions of the distal vaginal tract since it requires lubrication, prolonged vaginal dilations, or sexual intercourses to maintain patency. Using this technique contracture, shortening, bleeding, and dyspareunia are frequently found.

The amnion graft or the pelvic peritoneum graft should be avoided in children since those tissues are very fragile and require vaginal stenting and dilatations. Some nonsurgical techniques to create an artificial vagina, such as the Abbe-McIndoe procedure (perineal cleavage covered with a skin graft) or simply mechanical dilatations, are difficult to apply in preadolescent children due to the long period of vaginal dilatations, and in most cases, subsequent surgical management is needed.

Most of these cases need a well-motivated patient. These procedures are complicated by a high grade of vaginal stenosis, flap loss, graft shortening, unsightly donor site scars, the possibility of hair in the vagina, deficient lubrication, and the potential for squamous cell carcinoma in the graft. With paucity of local tissues, vaginal reconstruction may require the utilization of donor material.

Autologous buccal mucosa is an excellent graft material because of its robust vascularity and elasticity. With excellent color and texture matching to genital and vaginal skin, buccal mucosa generates moist, hairless, non-keratinized neovaginal mucosa. The tissue is readily available, and the donor site scar is completely hidden inside the mouth.

Autologous buccal mucosa vaginoplasty also avoids abdominal surgery with no risk for bowel obstruction, prolapse, or anatomic leak. Autologous buccal mucosa can be utilized in distal vaginal augmentation in patients with distal vaginal agenesis or urogenital sinus vaginal reconstruction as well as offer a novel technique for total neovagina creation in cases of vaginal agenesis, the secondary repair of vaginal stenosis after previous vaginoplasty, as well as for the creation of the external genitalia when tissues are lacking.

Buccal mucosa can be used to augment a foreshortened vagina, create a total neovagina, or in the repair of a vaginal stricture after prior vaginoplasty. Also, the use of buccal mucosal graft coupled with intravaginal wound vacuum therapy offers a promising new approach resulting in an excellent engraftment rate.

In pediatric patients, isolated intestinal segments provide an ideal tissue for an artificial vagina creation. The sigmoid segment is the segment of choice as excellent tissue for vaginal replacement, with the ileal bowel when the sigmoid is not available. Ileum produces more abundant and less lubricating secretions than the sigmoid segment and is also more fragile causing bleeding after intercourse.

The sigmoid colon is easy to mobilize, offers a more appropriate size vaginal canal, and permits obtaining an adequate length without problems. The usual length utilized is 10-12 cm of sigmoid colon with good functional results and low risk of diversion colitis. In colovaginoplasty, the secretions are initially excessive, but after two to 3 months they

normalized. Colonic mucosa is more resistant to trauma and a mould is not needed to maintain patency.

Using the colon, the possibility of end-to-end anastomosis between the neovagina and the uterus in cases of a patent uterus is possible. The ideal surgical time for vaginal reconstruction is after puberty.

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Low-Pressure Pneumoperitoneum

Over time, there has been a substantial increase in the utilization of laparoscopy as a surgical approach in pediatric patients. Carbon dioxide (CO2) serves as the insufflation gas, and the absorption of CO2 can lead to hypercapnia. The establishment and maintenance of a stable pneumoperitoneum play a crucial role in minimally invasive surgery, as it is imperative for creating adequate operative space to safely manipulate instruments.

The induction of hypercapnia through CO2 insufflation results in an elevation of cerebral blood volume and cerebral blood flow velocity. Elevated intraabdominal pressure during CO2 pneumoperitoneum contributes to increased intracranial pressure. CO2 insufflation during laparoscopy has both mechanical and pharmacological effects on the cardiorespiratory system. Hypercapnia, resulting from CO2 absorption into peritoneal blood vessels, induces an increase in heart rate (HR) and mean arterial pressure (MAP) through heightened release of catecholamines.

Even after desufflation, both HR and MAP remain elevated due to a delay in the reduction of CO2 levels. Hypercapnia induces moderate vasodilation in most tissues and marked vasodilation in the brain. However, CO2 pneumoperitoneum may lead to side effects such as pulmonary hypertension, hypercapnia, and acidosis. To mitigate these effects, patients are hyperventilated.

Hemodynamic changes observed during pneumoperitoneum with pressures exceeding 8 mmHg in children include increased heart rate and blood pressure, along with decreased

cardiac index (CI). CO2 insufflation, with the operating table in the reverse-Trendelenburg position while maintaining constant minute-ventilation, results in significant increases in CI, HR, MAP, and peak inspiratory pressure (PIP). The increase in CI is primarily due to an elevated HR. These effects are influenced by both the intraabdominal pressure (IAP) magnitude and the neurohumoral effect induced by hypercapnia.

An IAP above 10 mmHg in children results in decreased venous return, left ventricular preload, left ventricular cardiac output, and aortic blood flow. Additionally, it leads to changes in pulmonary function due to diaphragm elevation, reduced functional residual capacity, and increased alveolar dead space.

Low-pressure pneumoperitoneum (LPP) during laparoscopy has been introduced to minimize intraoperative hemodynamic changes and neuroendocrine stress reactions. LPP (below 10 mmHg), facilitated by deep neuromuscular blockade, is considered safe and feasible while offering physiological and clinical benefits.

These advantages include lower postoperative pain scores, reduced incidence of infectious complications, and decreased opioid consumption. Patients operated at LPP exhibit lower surgical site hypoxia and inflammation markers, along with less impaired early postoperative cytokine production. LPP results in less tissue hypoxia, lower circulating tissue damage markers, and a less impaired postoperative innate cytokine production capacity.

Increased IAP during laparoscopy predominantly affects the cardiovascular and pulmonary systems. The primary impact of LPP lies in postoperative pain and analgesic consumption. Post-laparoscopic pain can be categorized into three components: referred shoulder pain, superficial or incisional wound pain, and deep intra-abdominal pain.

Different types of pain may correspond to distinct etiologies, with referred pain often attributed to CO2-induced diaphragm or phrenic nerve irritation. Deep intraabdominal pain is primarily caused by bowel traction, abdominal wall stretch, and compression of intraabdominal organs. Symptoms attributable to pneumoperitoneum pressure may also contribute.

While the use of LPP may significantly increase operative time due to reduced surgeon visibility, its implementation is vital in addressing postoperative pain and promoting optimal patient outcomes.

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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.
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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