



PEDIATRIC SURGERY Update*

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Parastomal Hernia

Parastomal hernia (PH) is an incisional hernia that occurs within the surrounding of a stoma where abdominal content, typically bowel or omentum, protrude between the skin and bowel stomal edge surrounded by a hernial sac. Parastomal hernia is a common complication of various type of stomas. It can progress asymptomatic, resulting in an abdominal deformity, but it can lead to bowel incarceration and strangulation needing urgent surgery. In infants and young children, the most common indications for performing a stoma include necrotizing enterocolitis, Hirschsprung's disease and anorectal malformations. In adolescent children the indication is intractable functional constipation, intestinal pseudo-obstruction, and inflammatory bowel disease. An ostomy prolapse in children is more common than parastomal hernia in children. Prolapse is more common after loop than end enterostomies. Patients with gastrointestinal motility disorders have a higher complication rate and more severe complications in comparison to the children without gastrointestinal motility disorders. In adults the two most common conditions associated with stoma construction include colorectal cancer and inflammatory bowel disease. It is believed that 30 to 50% of stoma will develop a parastomal hernia, and one-third of these cases will need surgical correction. End (colon) ostomies have a higher probability of developing a parastomal hernia than loop (ileum) ostomies. Risk factors associated with developing a parastomal hernia include age above 60 years, obesity, diabetes, tobacco consumption, systemic and local infection, COPD, steroid therapy, inflammatory bowel disease and cancer. The incidence of parastomal hernia as a recurrence after corrective surgery is lower when using mesh for the repair. Diagnosis of a parastomal hernia is by physical examination. In the vast majority of cases the only clinical symptom is a deformity of the abdominal wall around the stoma. Parastomal hernia can be overlooked in obese patients. The use of ultrasound, CT-Scan or MRI increases the diagnostic accuracy. Indications for surgical management of parastomal hernia are limited to those with severe symptoms and complications of bowel obstruction occurring in 30% of all hernia patients. Indications for surgical management include incarceration, strangulation, obstruction, parastomal fistula, perforation, ischemia, recurrent symptoms of obstruction, difficulty maintaining collection device, hernia-related pain, and problems with irrigation of the stoma. Several methods utilized for corrective surgery of a parastomal hernia include open transposition of the stoma, use of mesh reinforcement, or repair using minimally invasive technique. Transposition is associated with a lower risk of hernia recurrence when compared with local reconstruction. When mesh is utilized, it can be placed superficially (onlay technique), pre-peritoneally (sublay technique) or intraperitoneally (inlay technique). Laparoscopic technique relies on intraperitoneally implanted prosthetics. Reinforcement of

the abdominal wall with prosthetic material is the method of choice since it promises good results and low incidence of complications and recurrences over long periods of time.

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Juvenile Granulosa Ovarian Tumor

Granulosa cell tumors of the ovary are rare benign ovarian sex cord-stromal tumors. Granulosa cell tumors are divided into a juvenile granulosa cell and adult granulosa cell variety tumor. Ovarian tumors account for approximately 1% of all tumors in children and adolescent. Juvenile granulosa cell tumor (JGCT) accounts for 67% of sex cord-stromal tumor in the pediatric population and approximately 5-12% of all ovarian neoplasms in children. Nearly half of the patients are diagnosed in the first decade of life with a median age of presentation of 7 years. The clinical significance of JGCT is due to its estrogen secreting properties resulting in pseudo precocious puberty without ovulation. More than 80% of patients present with symptoms of precocious puberty including increased pubic hair, vaginal bleeding, breast enlargement and advanced bone age. In older ages and adolescents JGCT causes other manifestations such as hirsutism, abnormal uterine bleeding and abdominal discomfort. There is a high level of sex hormones and suppressed gonadotropin level in this condition. JGCT secretes estradiol due to the presence of theca cells that secrete androstenedione which is subsequently converted to estradiol by the granulosa cells. Inhibin A & B which are synthesized by the granulosa cells are also elevated supporting the diagnosis. A pelvic mass is usually present. The triad of a palpable adnexal mass, elevated serum estradiol and absent or decreased gonadotropin is almost diagnostic of JGCT. JGCT are usually large (averaging 12 cm) and in most cases limited to the ovary. Under ultrasound granulosa tumors are solid and cystic or mainly solid with a spongiform appearance with the solid portion being heterogeneous in echogenicity. On MRI the solid component is typically isodense and enhances. Fluid-wave levels within the cystic component represent areas of hemorrhage. Granulosa cell tumors of the ovary rarely calcify or spread to the peritoneum, unlike epithelial neoplasm. JGCT are typically unilateral and confined to the affected ovary at diagnosis. Hence, most cases (>90%) are diagnosed with FIGO Stage 1 which respond well to unilateral salpingo-oophorectomy. Surgery should be performed in this age group with unilateral oophorectomy only for stage 1. There is no role

for simple ovarian cystectomy. Staging should include peritoneal cytology, exploratory laparotomy, and unilateral salpingo-oophorectomy. Bilateral ovarian involvement is uncommon in stage 1 tumors and wedge biopsy is not recommended. Prognostic factors include the size of the tumor, degree of nuclear atypia and mitotic activity. Tumor rupture is not a negative prognostic factor. Serum estradiol, CA-125 and inhibin B may be used for follow-up postoperatively. Precocious puberty changes subside, and physiologic puberty occurs at the normal expected age in all cases after tumor removal. Advance disease might need cytoreductive surgery followed by combination aggressive chemotherapy. Lymph node involvement is a rare phenomenon in sex-cord stromal tumors.

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Duct of Luschka

Ducts of Luschka are subvesical accessory biliary ducts located in the gallbladder fossa. They branch from the right hepatic or common hepatic duct, are not accompanied by artery or vein as other bile ducts draining liver segments (a portal triad is absent). Ducts of Luschka are small, less than 1-2 mm in diameter, usually originating from the lower aspect of the right hepatic lobe running along the gallbladder fossa and liver parenchyma. They do not open into the gallbladder. Drainage may be into intrahepatic or extrahepatic biliary ducts. Ducts of Luschka should be differentiated from hepatocystic ducts which are aberrant ducts that could drain a significant amount of liver parenchyma into the gallbladder or cystic duct. The reported prevalence of duct of Luschka is 4%. Injury to the ducts of Luschka during laparoscopic or open cholecystectomy can cause postoperative bile leak and peritonitis. Most injuries to the duct of Luschka occur after ligation and division of the cystic artery and cystic duct while dissecting the gallbladder from the liver fossa. These ducts can also be injured during liver resection, liver transplantation and interventional radiological procedures. In very rare occasions the ducts of Luschka can be identified intraoperatively. Patients with bile leaks have variable clinical course presenting with mild abdominal pain, tenderness, fever, or biliary peritonitis with sepsis. There is mild elevation of serum bilirubin and alkaline phosphatase. Timing of presentation of such leaks is usually within the first postoperative week. The patient with duct of Luschka leaks will develop a fluid collection (biloma) diagnosed by US or CT-Scan. A percutaneous drainage is usually

necessary to drain the bile leak. Performing a fistulogram through the draining catheter will demonstrate a communication with the biliary tree. Though a HIDA scan will demonstrate a bile leak, it cannot give an anatomical impression of where the bile leak is coming. MRCP can diagnose a leaking duct of Luschka. ERCP is the standard mode of diagnosing a duct of Luschka leak. ERCP can also be therapeutic by reducing intrabiliary pressure with sphincterotomy and endobiliary stent placement. The management of a duct of Luschka leak depends on the clinical condition of the patient. Asymptomatic patients with a low output leak can be managed with simple drainage. Spontaneous resolution of the leak may occur because accessory ducts do not drain a significant portion of the liver. Should the leak produce a higher output of bile, ERCP with sphincterotomy, stenting or nasobiliary tube placement should be in order. Patients with severe symptoms and those where the leak persists despite endoscopic treatment should be reexplored and ligation of the leaking duct performed.

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