



PEDIATRIC SURGERY Update © **Vol. 40 No. 03 MARCH 2013**

Malignant Pancreatic Tumors

Malignant tumors originating primarily in the pancreas occur rarely in children. They mostly arise from the exocrine cells of the pancreas, as most endocrine pancreatic tumors are benign. Malignant pancreatic tumors comprise in order of frequency: solid pseudopapillary tumor, pancreatoblastoma, ductal adenocarcinoma, acinar cell carcinoma and a few malignant endocrine tumors. Pancreatic tumors in children most commonly present with a palpable mass or abdominal pain. Jaundice occurs less frequent than in adult. Primary imaging includes ultrasound, CT-Scan, MRI. This can be followed by surgery or fine-needle image-guided biopsy to establish a histological diagnosis and plan further therapy. This second option of biopsy is used when the child present with distant metastasis or an unresectable primary tumor. The two most common malignant pancreatic tumors are the solid pseudopapillary tumor (SPT) and the pancreatoblastoma. Pancreatoblastoma is a more commonly found in the first decade of life and is a more aggressive tumor, while SPT is more common in females during adolescent years and carries a better overall prognosis. About one third pancreaticoblastoma cases are metastatic at the time of diagnosis. These tumors can be responsive to chemotherapy and radiation. Preoperative chemotherapy can successfully decrease tumor size and allow a more complete surgical resection. Adequate management of malignant pancreatic tumor is complete surgical resection. On multivariate analysis, histologic type is the only factor that significantly predicts survival. Patients with poorly differentiated carcinoma shows the worst survival probability.

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Rectovestibular Fistula with Normal Anus

Rectovestibular fistula with a normal anus is a rare anorectal malformation affecting females patients. It is also known as anovestibular fistula or H-type rectovestibular fistula. Is more commonly found in Asia. Most cases present with early infant history of fecal discharge through an external opening in the posterior vestibule of the genitalia. A few will develop a perineal vulvar abscess before the fistula becomes visible. Anal stenosis has been associated with this condition. The embryology of this fistula is thought to represent a persisting cloacal canal or an interruption of the dorsal part of the embryonal cloacal membrane by an isolated defect. The fistula uniformly extends from the vestibular fourchette to the anterior wall of the rectum one to 3 cm above the dentate line. Contrast enemas and endoscopic studies are not very useful as diagnostic aid. The diagnosis is best confirmed during an exam under anesthesia passing a small probe from the vestibule to the internal rectal orifice. Management of rectovestibular fistula has included perineal repair, vestibuloanal pull-through, anterior perineal anorectoplasty, fistulectomy and limited posterior sagittal anorectoplasty. To avoid a colostomy bowel preparation and systemic antibiotics are necessary. Most common complications are recurrence and wound dehiscence. Fistula recurrence occurs when blood perfusion in the local rectal tissue is poor and separation of the divided ends of the fistula fails. It is important to close the fistula orifice using healthy anterior rectal wall.

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Duodenum Inversum

Duodenum inversum refers to a congenital anomaly where the third portion of the duodenum is located to the right of the second portion or above the duodenal bulb. In other words the third portion instead of continuing leftward toward Treitz, reverses direction and travels in a superior posterior track prior to crossing the midline above the pancreas. Most cases occur in male adults. Duodenum inversum is associated with nonspecific symptoms such as epigastric discomfort, nausea, distension or duodenal obstruction. Other associated conditions include cholelithiasis, pancreatitis and annular pancreas. The condition can mimic mesenteric artery syndrome. Diagnosis is confirmed with UGIS. Management of duodenum inversum without obstruction is conservative (antacids, antispasmodics). With duodenal obstruction surgery may be required. The

obstruction is due to fibrotic bands. Lysis can be performed laparoscopically.

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