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H-type Rectovestibular Fistula

H-type rectovestibular fistulas, also called congenital rectovestibular fistula with a normal anus, is a rare condition encountered in Western female infants, except in Asia where is encountered more commonly. Clinically the anus is in a normal position while the child discharges fecal material or flatus from the vestibule or vagina. Most of these fistulas extend from the posterior vestibular fourchette to the anterior wall of the rectum located below the level of the levator muscles and one to three cm above the dentate line. No patient has history of perineal abscess or cellulitis in the congenital variety. Some cases are associated with a stenotic or anteriorly displace anal canal. Müllerian duct structure agenesis (Mayer-Rokitansky syndrome) has been associated with this type of fistula. The fistula is lined by squamous epithelium and most have a small diameter (1-5 mm). Management consists of resection of the fistulous tract preferably at the age of three years when the perineal body is better developed. Several procedures have been reported in the literature such as simple fistula resection, the vestibuloanal pull-through, transperineal closure of the fistula, posterior sagittal anorectal approach and a transanal approach. Most of the procedures can be performed without the need of a protective colostomy, but a good bowel preparation is needed. Complications include wound infection and fistula recurrence with reoperation in 50% of the cases.

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Rhabdoid Tumor

Rhabdoid tumor is a rare, very aggressive malignancy identified in fetus and young children. Rhabdoid tumors (RT) can occur within the kidney, central nervous system and extrarenal in location. Rhabdoid tumors are characterized by early metastasis and high mortality rate. Most cases are identified in the perinatal period during the first year of life. The tumor metastasize to multiple sites such as skin, placenta, bones, lungs, lymph nodes,

brain and liver. Extrarenal RT occurs more commonly in the perinatal period, presents as a mass lesion in head/neck, skin tumor nodules or metastatic disease, has a bad prognosis. Renal RT occurs primarily in infants and newborns, presents as an abdominal mass, fever and hematuria, with high-tumor stage. Most have metastasis at diagnosis with presence of CNS lesions. Survival increases with increasing age at diagnosis. Central nervous system RT occurs primarily in infants in the posterior fossa presenting as an intracranial mass and hydrocephalus, characterized by extensive brain invasion, recurrence and short survival. For cure an aggressive surgical approach to achieve total gross resection of the tumor is needed along with adjuvant chemo- and radiotherapy. Patients with localized disease and complete surgical resection are most likely to survive long-term.

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Thoracic Duct Ligation

The lymphatics of the lower extremity and lower torso join with those of the mesentery of the bowel to form the cisterna chyli. The cisterna chyli lies in the lumbar prevertebral plane behind the aorta and inferior vena cava. Confluents of the cisterna chyli create the thoracic duct which ascends in the right thoracic prevertebral plane, medial to the azygos vein and behind the esophagus and aorta. The thoracic duct is a tubular structure one to 3 mm in diameter. It crosses from the right side of the chest to the left at the level of the fourth or fifth thoracic vertebrae and it usually empties into the left jugulo-subclavian junction. The accessory thoracic duct empties in a similar fashion on the right side. Either contrast or scintigraphic lymphangiography can help delineate the anatomic variants identified in almost 40% of human. Direct contrast lymphangiography is considered the best technique to delineate the anatomy of aberrant lymphatics channels. Postsurgical, traumatic, congenital or spontaneous chylothorax that does not respond to medical management with drainage, medium-chain triglyceride, or TPN might need ligation of the thoracic duct. Low thoracic duct ligation is a reliable means of control of postoperative chylothorax. Ligation can be performed through either right chest thoracotomy or video-assisted thoracoscopy.

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