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Cloacal Exstrophy

Cloacal exstrophy is the most severe presentation of a ventral abdominal wall defect. Formerly a fatal disorder it has yield to a higher survival during the past years with improvement in quality of life. The incidence is between one in 200-400,000 live births. Premature rupture of the cloacal membrane before descend of the urorectal septum is the most plausible explanation of the defect. The anomaly consists of a hypogastric omphalocele, two lateral hemibladders joined to a central strip of exstrophied intestinal epithelium (ileocecal plate) through which ileum prolapses, imperforate anus, and ambiguous genitalia (see figure). Other associated anomalies are cardiac, orthopedic (equinovarus), and neurological (tethered cord syndrome, meningocele). Prenatal sonographic diagnosis has been reported. Radiological evaluation should include plain films of chest, spine and ultrasound of urinary tract. Optimal reconstruction centers initially around closure of the omphalocele, approximation of the symphysis pubis (iliac osteotomies may be needed with late repairs), establishment of intestinal continuity preserving all hindgut bowel present with colostomy, and functional closure of the bladder. Infants with rudimentary genitalia are assigned the female gender, early gonadectomy is advised for genetic males. During the preschool years reconstructions focus on urologic (intermittent catheterization) and fecal (pull-through) continence. Vaginal construction will be done later in life.

Burkitt's

Burkitt's lymphoma (BL) is a highly malignant tumor first described during the late 50's in African children (jaw), endemic in nature, and composed of undifferentiated lympho-reticular cells with uniform appearance. The American BL variety is non-endemic, mostly attacks children between 8-12 years of age, predominantly (>75%) with abdominal disease such as unexplained mass, pain, or intussusception. The head and neck region follows. The tumor can appear as a localized, diffuse (multifocal, non-resectable) or metastatic abdominal mass (bone marrow and CNS). It's considered the fastest growing tumor in humans with a doubling time around 12-24 hrs. Chemotherapy is the

primary treatment modality due to its effectiveness in rapidly proliferating cells. The role of surgery is to establish the diagnosis (using open biopsy), stage the tumor, remove localized disease, relieve intestinal obstruction and provide vascular access. Complete resection whenever possible offers the patient improved survival. Is more readily accomplished in patients with localized bowel involvement operated on an emergency basis due to acute abdominal symptoms. The only predictor of event free survival is extent of abdominal disease at diagnosis. Debulking (cytoreductive) procedures increases morbidity and delays initiation of chemotherapy worsening prognosis. Extensive tumors should be managed with minimal procedure and immediate chemotherapy (a/o radiotherapy). Bone marrow and CNS involvement are ominous prognostic signs.

Liver FNH

Focal Nodular Hyperplasia (FNH) is a benign liver tumors found in children. Most are female (80%) in their teen or childbearing age, asymptomatic or with non-tender mass on routine exam. Liver function tests are usually normal. Have no malignant potential but should be differentiated by imaging or biopsy from a liver cell adenoma. Is not a life threatening lesion except in women taking oral contraception that may develop hemorrhage. Diagnostic imaging is a CT showing a well circumscribe mass of low density, arteriogram a hypervascular mass, and normal uptake on liver nuclear scan. Laparoscopically guided needle or open biopsy should be done for diagnosis. Histology describes nodular aggregates of normal hepatocytes with areas of intranodular bile duct proliferation. Asymptomatic lesions can be follow-up with ultrasound and resected if they enlarged or become symptomatic. Prognosis is excellent even in tumors left behind.

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