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Dermoid Cysts

Dermoid cysts are among the commonest epithelial tissue malformation found in children. Histologically these cysts are subclassified into epidermoid, dermoid cysts and teratomas. Dermoid cysts occur most commonly among regions of embryologic fusion such as the top lateral corner of the eyebrows (angular dermoid), in the midline of the neck (confusing with thyroglossal duct cysts), over the midline of the nose, in the temporo-parietal and suprasternal area. Dermoids are non-tender, mobile, translucent, slowly growing tumors composed of keratin, hair follicles and sebaceous glands within a thin wall that contains epithelial elements. Midline dermoids are entrapped epithelium of branchial arch origin at the time of fetal midline fusion. Occasionally the cyst rests on the dura (intracranial extension) or on the orbital fascia causing bone erosion seen in x-rays. Intraglossal dermoids have also been described. Infection is the most common complication and is caused by repeated local trauma. Those with intracranial extension are at risk of aseptic meningitis. History and physical exam are the most important element in the evaluation of neck masses in children. Complete surgical excision when the diagnosis is made is the treatment of choice.

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Inoperable Wilms' Tumor

On occasion Wilms tumors (Nephroblastoma) grow to such massive size that primary surgical excision poses a high risk of complications and mortality to the child. This type of tumor is either found unable to be resected during surgical exploration or judge inoperable by clinical evaluation or imaging studies. The inoperable criterias most commonly utilized are: huge size of the tumor, involvement of adjacent vital structures, intra-caval involvement above the level of the hepatic veins and atrial tumor extension. This factors significantly increase the risk of surgical morbidity, principally hemorrhage, during initial

nephrectomy. Solely relying on imaging studies for staging and deciding inoperability can lead to inaccurate decision. In such cases it is recommended to do initial exploration to determine operability and obtain tumor biopsy including suspicious lymph nodes or other metastatic foci. Preop chemotherapy has been found to almost always ease surgical resection by reducing tumor size decreasing the incidence of tumor rupture. Histologic patterns are still recognized after preop chemotherapy, though there can be initial biopsy sampling errors. Failure of tumor reduction can be followed by radiation therapy. Inoperability as a criterion is an adverse prognostic factor independent of stage and results in loss of important staging information. Once there is evidence of tumor size reduction and evidence of vena cava tumor regression definite resection should be completed.

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Ivemark's Syndrome

First described in 1959, Ivemark's syndrome refers to a uniformly fatal renal-hepatic-pancreatic dysplasia of unknown origin affecting infants in the first six-months of life. Prenatal diagnosis has been made as early as the 18 weeks of fetal development. An autosomal recessive pattern of inheritance has been proposed in this syndrome. The renal malformation consists of cystic dysplasia characterized by disturbance in glomerular differentiation, delay in tubular differentiation and abnormal expression of epithelial markers in glomeruli and tubules (multicystic kidneys). Hepatic abnormality consists of enlarged portal areas and elongated biliary ducts with a tendency to perilobular fibrosis. The intrahepatic ductal dilatation resembles Caroli's disease. The pancreatic abnormality consists of dilated duct, cysts and fibrosis with diminution of parenchymal tissue. Clinically the child develops during the neonatal period cholestatic jaundice followed by renal and pancreatic failure. Ivemark syndrome should not be confused with the asplenia-cardiac anomaly syndrome also bearing the same eponym. Ultrasound findings in the three affected organs suggest the diagnosis that unfortunately will be confirmed at autopsy. Management is supportive.

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