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Fibrosarcoma

Fibrosarcoma, a malignant tumor of fibrous tissue, is a unusual neoplasms in infants and children. It comprises 10% of all soft-tissue sarcomas in children and the most common during the neonatal period. Anaplastic spindle-shaped cells arranged in a herringbone pattern with abnormal mitosis, nuclear pleomorphism and increase basophilia are characteristic pathological findings. Two peaks in the incidence are affected: infants and young children under the age of five (congenital variety) and patients between 10 and 15 years of age. A slight male predominance is reported. Clinically the child presents with a rapidly enlarging mass or swelling in the soft tissue. Sites of origin in order of decreasing frequency are: the extremity, head and neck, trunk, and retroperitoneum. Most important prognostic factors are: site of origin, and extent of disease at diagnosis. Fibrosarcoma is a locally aggressive tumor. Tumors in infants have a more benign course. Extremity lesions have a low rate of recurrence and better prognosis. Axial tumors have a high rate of metastasis and death from disease. Adequate biopsy is necessary for diagnosis. Mainstay of therapy consists of surgical resection by wide local excision or amputation for neurovascular bundle involvement or development of a dysfunctional extremity. Chemotherapy (VAC) is a useful adjunct for unresectable metastatic or primary disease and to alter the surgical approach to bulky tumors. Overall survival is 80% at five years.

CAH

Congenital adrenal hyperplasia (CAH) involves a functional defect in any of the five enzymatic steps required for cortisol synthesis, most commonly 21- (involved in 90-95% of cases) and 11-hydroxylase level. This primary genetic defect transmitted as autosomal recessive impairs the ability of the adrenal cortex to synthesize cortisol causing increase feedback secretion of ACTH and adreno-cortical hyperplasia of the gland. Increase output of steroids proximal to the block (androgenic precursors) causes virilization in affected males and females. Its more severe form is associated with aldosterone deficiency and life-threatening salt wasting. Female pseudohermaphrodite due to virilizing CAH is the most frequent form of intersexuality found. The phenotypic picture

varies from mild clitoral enlargement alone to complete masculinization of the urethral meatus at the tip of the penis. Prenatal diagnosis (southern blotting of DNA) is based on finding the disease gene on the short arm of chromosome 6. Likewise management in the mother (dexamethasone) is started empirically until the affected status is known by chorionic villus sampling. After birth management consists of cut-back or flap vaginoplasty with clitoral recession at 3-6 months of age. Children with high vaginal entry proximal to the urethra external sphincter can undergo early one-stage reconstruction at 8-12 months of age. Long term surgical results of female children show adequate sexual identification, reproduction, intellectual functioning and acceptable genitalia.

Microgastria

Microgastria is a rare congenital anomaly of arrested foregut development resulting in a small stomach with minimal reservoir capacity. Frequently associated to: malrotation, asplenia, cardio-pulmonary, and limb anomalies. Clinically the infant develops feeding intolerance: postprandial vomiting, gastro-esophageal reflux, aspiration pneumonia, and failure to thrive. Diagnosis is made by UGIS showing a small saccular or tubular midline stomach, proximally dilated esophagus, and incompetent cardia. Management consists of small frequent feeding in less severe cases to creation of a double lumen Roux-en-Y jejunal pouch (Hunt-Lawrence Pouch) for food storage in symptomatic children. This procedure (see figure) provides a pouch for food intake, improves nutritional status, ease drainage and prevents alkaline reflux esophagitis. Vitamin's B-12 supplement is needed due to decrease production of Intrinsic factor.

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