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Prepubertal Testicular Tumors

Testicular tumors presenting before puberty in male patients occur very rarely and very distinct from the adult counterpart. Prepubertal testicular tumors are usually of one histologic type. The most common prepubertal testicular tumor is yolk sac tumor followed by teratoma. Median age at surgery is 17 months and most children present with a solid scrotal painless mass. This must be followed by Ultrasound and tumor markers. The most common type of malignant tumor is yolk sac (62%) and the most common benign tumor is a mature teratoma (23%). Yolk sac histology is more common in Asian/Pacific population, as compared with white American where teratoma predominates. In yolk sac tumor the epithelial lining of the cysts and tubercles secretes high concentration of alpha-fetoprotein (AFP), a tumor marker. AFP is both important in the diagnosis and follow-up of such tumors, especially for recurrence after treatment. Elevation of AFP can also occur in benign teratomas and normal infants up to the age of two months. Most children with yolk sac tumors have clinical stage I disease, with age at presentation of 15 months and absence of metastatic disease. Clinical stage I disease is managed with radical orchiectomy using an inguinal approach. Yolk sac tumors metastasized through lymphatics and blood borne to lymph nodes and lung. Stage II and higher stages are managed with retroperitoneal lymph node dissection, adjuvant chemotherapy and radiotherapy. In cases of teratoma, the age at operation is 12 months, and immature teratomas present earlier in life than matured teratomas. No child with a prepubertal testicular teratoma showed metastatic disease, regardless of the presence or absence of testicular-sparing surgery. It is proposed that any child with ultrasound showing salvageable normal testicular parenchyma and normal AFP should be managed with testis-sparing surgery.

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Prenatal Diagnosis Esophageal Atresia

Esophageal atresia (EA) with or without tracheoesophageal fistula is the most common congenital anomaly of the esophagus in children. Prenatal diagnosis of EA relies on the indirect findings in routine sonography at 16-20 weeks gestation of a small or non-visualized stomach bubble in conjunction with subsequent maternal polyhydramnios after 24 weeks. These US findings are nonspecific and can be transient in nature creating unnecessary anxiety in the expectant mother and relatives. Two other more direct signs of the US diagnosis of EA include a dilated proximal esophageal pouch (upper pouch sign) and failure to visualize the entire thoracic esophagus. Using all these findings in US the rate of diagnosing EA prenatally occurs one-third of the cases. Most cases of EA that are diagnosed prenatally are the long-gap esophageal variety without tracheoesophageal fistula which occur with an overall incidence of 8%. The most common variant of EA with distal tracheoesophageal fistula is missed most of the time since it decompresses through the fistulous tract into the stomach and filling it preventing the development of polyhydramnios. Prenatal diagnosis of EA gives us the opportunity for optimal perinatal management with delivery of the fetus in a tertiary care pediatric center with expertise in esophageal surgery. Fetal MRI and biochemistry of the amniotic fluid can help confirm the diagnosis since gamma glutamyl transpeptidase (GGTP) and alpha-protein (AFP) in amniotic fluid are elevated in EA. The dynamic sequence of MRI during fetal swallowing is needed to increase the diagnostic yield. Since 50% of EA children have an associated malformation these must be looked carefully to correlate with the diagnosis of VACTERL, CHARGE, abnormal karyotype or other associated syndromes dictating prognosis.

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Aberrant Subclavian Artery

Aberrant subclavian artery, also known as arteria lusoria, is the most common form of aortic arch vascular anomaly. It results from regression of the right 4th aortic arch between the carotid and subclavian arteries. The right subclavian artery usually persists as a branch from the descending aorta distal to the takeoff of the left subclavian artery

and coursing posterior to the esophagus, though it can also pass between the esophagus and the trachea or even anterior to the trachea very rarely. Patients with an aberrant subclavian artery can develop symptoms which include dysphagia, cough, stridor, regurgitation, asphyxia induced by feeding, a globus sensation (or lump in the throat), failure to thrive and thoracic pain, though most patients with this anomaly remain asymptomatic throughout their lifetime. Infants can present with respiratory symptoms due to dysphagia and aspiration of food particles. Some asymptomatic patients can elicit symptoms during exercise. Barium swallow imaging shows a characteristic diagonal posterior compression defect at the level of the 3rd and 4th vertebrae which is diagnostic. CT or MRI angiography and transthoracic echocardiogram confirms the diagnosis. Symptomatic children should undergo surgical management. The goal of operative repair is relieving the symptoms and restores circulation. This is done by reimplanting the aberrant subclavian artery into the ascending aorta or the right common carotid artery directly through a right thoracotomy in children. Anatomic variations that are associated with an aberrant subclavian artery include abnormal origin of the right vertebral artery from the aorta or from the right common carotid artery, the presence of a c common carotid trunk, a right-sided thoracic duct and a nonrecurrent laryngeal nerve.

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