



# **PEDIATRIC SURGERY Update** © **Vol. 44 No. 01 JANUARY 2015**

## **Testicular Sex Cord-Stromal Tumors**

Testicular sex cord-stromal tumors (TSCS) are very rare in children accounting for near 6-8% of all testicular neoplasms in the pediatric age. TSCS main histological types include Leydig cell tumors, Sertoli cell tumor, juvenile granulosa cell tumor and undifferentiated cell tumors. Clinically they present as a painless testicular mass with hormonal manifestation causing isosexual pseudoprecocity or estrogenic manifestation occurring in up to 20% of all cases. TSCS does not have an aggressive behavior being low-grade tumors. Malignancy is defined when the tumor has lymphatic or vascular invasion, high mitotic index and tumor necrosis. Age adjusted AFP should be negative in TSCS, otherwise it's a germ cell tumor. Leydig cell tumors represent the most common histologic type. They produce testosterone, hence precocious puberty with elevated 17-ketosteroid levels occurs. When diagnosed preop, testis-sparing enucleation may be considered because these tumors tend to follow a benign course. Sertoli cell tumors are diagnosed before one year of life, hormonal silent, occasionally producing gynecomastia due to estrogen secretion. The clinical course is usually benign in children less than five and they can also be managed with testis-sparing surgery when diagnosed appropriately. Older children should have a metastatic workup. Metastatic disease requires aggressive surgical and adjuvant therapy. Children with large cell calcifying Sertoli cell tumors are at risk for endocrine syndromes. Granulosa cell tumors occur in neonates and can be associated with ambiguous genitalia. They should be suspected in neonates with scrotal swelling, normal age-adjusted AFP level, positive inhibin and a complex cystic multiseptated hypoechoic mass in the ultrasound of the testis. They can be managed with testis-sparing surgery. Undifferentiated stromal tumors harbor malignant potential and prepubertal and postpubertal males with these tumors should undergo a metastatic evaluation.

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## Covered Cloacal Exstrophy

Covered cloacal exstrophy (CCE) is a very rare frequently misdiagnosed malformation found within the spectrum of cloacal exstrophy requiring a high index of suspicion for diagnosis. Low implantation of the umbilical cord in association with separated pubic bones and an anorectal malformation (imperforate anus) are the most common sign associated with a covered cloacal exstrophy. Besides the anorectal malformation these patients also have an absent bladder neck and short colon. Most children with CCE are females with a single clitoris. Inspection of the perineum can discover a single large orifice or four perineal orifices very close to each other. Separation of the pubic bone can be seen at the physical exam seen as two mild prominences away from the midline in the pubic area or in simple films. There is a fibrous band between the separate pubic bones. The presence of a large perineal orifice through which there is constant dribbling of urine is another important sign to establish the diagnosis. There is no abdominal wall defect present. The absence of bladder neck associated with a very small bladder will require urinary reconstruction with bladder augmentation and Mitrofanoff to keep dry urine. Those cases with short colon unable to form solid stools will need a permanent stoma reconstruction. If there is evidence of well-formed solid stools, the child can undergo a pull through procedure. During initial stoma creation no piece of colon should be left attached to the urinary tract to take advantage of its water absorptive capacity and avoid urine absorption and associated hyperchloremic acidosis. Reconstruction of the GI tract takes precedence over the urinary reconstruction. Indication for pull-through depends on successful bowel management through the stoma, which depends on the ability to form solid stools. To maximize this potential it is crucial to use all available hindgut for the initial colostomy and avoid use of colon for urologic or genital reconstruction.

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## Angiomatoid Fibrous Histiocytoma

Angiomatoid fibrous histiocytoma (AFH) is a soft tissue tumor with intermediate malignant potential that occurs primarily in children, adolescent and young adults very rarely encountered past the age of 40 years. The median age of presentation is 14 years

accounting for 0.3% of all soft tissue neoplasms. It develops as a slowly growing nodular, multinodular or cystic mass of the hypodermis or subcutis occurring most commonly in the extremity of the child. Local symptoms such as pain and tenderness are uncommon, but systemic symptoms such as anemia, fever, malaise and weight loss are occasionally encountered suggesting the production of cytokines by the tumor. The diagnosis is rarely made preoperatively. Imaging findings of AFH are as nonspecific as its histogenesis. MRI demonstrates multiple internal cystic areas, an enhancing fibrous pseudocapsule which is markedly hypointense on T1- and T2WI, and foci of susceptibility artifacts representing hemosiderin. The diagnosis of AFH is made based on histopathology and immunohistology. AFH is generally firm and circumscribed. The characteristic microscopic appearance includes distributions of ovoid to spindle cells with bland, vesicular nuclei, lymphoplasmacytic infiltrate with intervening blood-filled cystic spaces, and a fibrous pseudocapsule. Immunohistochemistry variably demonstrates positivity for desmin, CD68 and CD 99. Management of AFH is surgical resection. Wide surgical excision with clear margins and post-excisional monitoring is warranted. Most patients are free of disease after local excision with a minority developing recurrent or less commonly metastatic disease within two years after surgery. Local recurrence can also be managed with radiation therapy.

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