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Prune Belly Syndrome

Prune belly syndrome (PBS) is a rare malformation occurring one in 40,000 live births affecting almost exclusively males (95%), and characterized by deficiency of the abdominal muscles, malformations of the urinary tract and bilateral cryptorchidism. The protruding hypoplastic abdominal wall looks like a dried prune. The pathogenesis of PBS is nor fully understood. Malformations of the urinary tract in PBS are due to dysplasia of the smooth muscle of the renal pelvis, ureters and prostatic part of the urethra. Three clinical manifestations are characterized in PBS: non-viable oliguric form due to severe kidney dysplasia, a serious form consisting of marked renal dysplasia with megaureters, mega-vesicles and progressive renal failure or the more favorable form with moderate renal dysplasia and different degrees of ureters and bladder enlargement. Diagnosis can be made prenatally and clinically. Orthopedic deformities (hip dysplasia, missing extremity, club feet) are the second most common associated malformation. Anomalies of the GI tract (malrotation, volvulus, atresia) occur in almost 30% of PBS. Those with oligohydramnios develop pulmonary hypoplasia. Prenatal vesicoamniotic shunt for urinary obstruction can prevent pulmonary hypoplasia and renal dysplasia. The associated nephropathy is partly dysplastic and obstructive. Cryptorchidism is present in almost all cases, with favorable histology. Bladder capacity is enlarged with detrusor muscle thickened and presence of vesicoureteral reflux. Ureters are elongated, dilated with inefficient peristalsis. Treatment of PBS encompasses preserving kidney function with temporary urinary diversion and subsequent surgical reconstruction, reimplantation of dilated ureters, orchidopexy and abdominoplasty. Timing of repair is controversial and should be tailored on an individual basis following a conservative approach. Abdominoplasty and orchiopexy have both physiological and improved quality of life benefits. Renal failure is main cause of death.

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Poland Syndrome

Poland syndrome (PS) is a spectrum of congenital chest wall deformities sporadic in occurrence characterized by chest wall hypoplasia. This is caused by absence of the pectoralis major, pectoralis minor, serratus anterior, rectus abdominis and latissimus dorsi muscle. Other associated conditions include athelia or amastia, nipple deformities, limb deformities (syndactylism, brachydactyly), absent axillary hair and limited subcutaneous fat. Severe (complex) cases include thoracic cage anomalies, most frequently involving ribs II–V. The disease may be inherited as an autosomal-dominant trait. Clinical manifestations of PS are extremely variable and rarely are all the features recognized in one individual. The right side is more commonly affected and is present in males 70% of the time. The etiology of Poland syndrome is unknown but might include abnormal migration of the embryonic tissues forming the pectoralis muscle, hypoplasia of the subclavian artery or a traumatic event in utero. Poland syndrome can occur in varying degrees with mild hypoplasia to total aplasia of muscles, ribs and cartilage. Surgical repair varies according to the extent of Poland syndrome, age and sex of the patient. In girls chest wall reconstruction should precede breast reconstruction. In the complex forms chest wall reconstruction has traditionally been advocated with the use of contralateral, autologous rib grafts stabilized with mesh. Recently the vertical expandable prosthetic titanium rib expander has been reported to stabilize the chest wall after rib grafting. Additional stages of reconstruction include expanders, musculocutaneous flaps, breast implants, nipple and areolar reconstruction and fat grafting. For symmetry reasons surgery to the contralateral breast can be considered.

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Liposarcoma

Adipose tumors comprise 5% of all soft-tissue neoplasm in children. Two-third are simple lipomas, 30% lipoblastoma and the rest rare liposarcomas. Liposarcomas occur most commonly in the 3rd to 7th decade of life with a slight predominance in males. They are extremely unusual in children younger than 10 years. In children liposarcoma occurs most commonly in the lower extremity, most have tumors> 5 cm at initial

presentation and metastatic disease at the time of initial diagnosis is uncommon. Clinically they present as nontender, slow and progressively growing soft-tissue mass. The myxoid variant is the most common histologic variant of liposarcoma in children. Histologic grade is one of the most important predictors of outcome, with low-grade myxoid tumors having significantly improved survival rates compared to the round-cell, pleomorphic, and dedifferentiated subtypes. Complete surgical resection remains the mainstay of local therapy, but adjuvant radiation therapy is effective at controlling microscopic residual disease after surgical resection. Myxoid tumors are radiosensitive and pre-, intra- and postoperative radiation approach have been effective therapy. The role of chemotherapy for treatment of pediatric liposarcoma is not well established except a role in facilitating tumor resection in patients with unresectable disease. The overall prognosis of myxoid liposarcoma is excellent with surgical treatment alone. The pleomorphic subtype portends a poorer prognosis.

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