PEDIATRIC SURGERY *Update* ©
Vol. 21 No. 01 JULY 2003

Bilateral Wilms Tumor

Synchronous bilateral Wilms tumor, also termed Stage V Wilms disease, occurs in approximately 5% of all cases of Wilms tumor in children. Definitive progress has been made during the past twenty years in diagnosis and management of bilateral Wilms tumor with marked improvement in prognosis. Diagnosis of bilaterality can be confirmed either most commonly during initial imaging or rarely while performing exploratory laparotomy. Some of the better prognostic factors associated with bilateral Wilms tumor are a patient age less than three years at diagnosis, lower stage of the most advanced lesions, favorable histology and negative nodal involvement. Most children (96%) with stage V tumors have favorable histology. Currently, management of bilateral Wilms tumor entails initial biopsy of both tumor masses, staging of lymph node metastasis followed by preoperative chemotherapy. Cytoreductive chemotherapy before surgical resection reduces tumor burden and permits more renal preservation procedures. Jointly, renal salvage procedures (partial nephrectomy and enucleation) have been recommended to conserve renal parenchyma. This has facilitated the use of parenchymal-sparing operations, with the potential advantage of decreasing the incidence of end-stage renal disease. Patients with inoperable tumors and extensive intravascular tumor extension can also benefit from this approach. Long-term follow-up reveals a group of children who develops insidious renal compromise when more than 70% parenchyma is compromise. Bilateral Wilms tumor with unfavorable histology is associated with poor prognosis.

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Crohn's Disease

Crohn's (terminal ileitis) is a chronic, transmural inflammatory bowel disease most frequently involving the terminal ileum and proximal colon that adversely affect growth and sexual maturation in children. Incidence is growing and etiology is undetermined. Diarrhea, abdominal pain, failure to thrive and weight loss are the most frequent clinical feature. Diagnosis is established by colonoscopy or imaging studies (CT-Scan). Initial management is medical and consists of azulfidine or 5-amino salicylic acid preparations, local and systemic steroids, metronidazole, immunosuppressives, and enteral and/or parenteral nutrition. Indication for surgery is limited to complications of the disease process and includes failure of medical therapy, perforation, abscess, severe malabsorption and growth retardation, persistent bowel obstruction, fistulas (entero-enteric and entero-urinary) and strictures. Surgery can be accomplished using limited resection and anastomosis or stricturoplasty. Best long-term results after surgery occurs in children with disease confine to the small bowel and ileocecal region. Diffuse ileocolonic involvement (Panenteritis), preoperative use of 6-MP, and colonic involvement is associated with early relapse. Early relapse after surgery is also seen after failure of medical therapy independent of disease location as the sole indication for surgery and in children undergoing resection within one year of the onset of symptoms.

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Amniotic Band Syndrome

Amniotic or constricting band syndrome (ABS) refers to progressive intrauterine amputation of fingers or limbs associated with a wide spectrum of congenital anomalies involving the trunk and craniofacial region. Incidence is one in 15,000 livebirths. Most amputations occur in the upper limb. In the hand, digital amputations are most common in the index, middle, and ring fingers, whereas in the foot, amputations of the hallux are most often noted. The congenital (intrauterine) band, a product of rupture amnion, produces compression and chronic ischemia of the affected limb. Multiple anomalies are

associated with most ABS cases such as orbital defects, lid anomalies, lacrimal outflow obstruction, ocular malformations, waist constriction, clubfoot, fascial cleft, cleft palate and lips. Follow-up ultrasound exams have afforded the opportunity of observing the in utero process of limb strangulation and subsequent spontaneous lysis of an amniotic band in a few cases. Management depends on clinical findings at birth. Prognosis depends on the severity of the abnormalities and the involvement of internal organs.

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ISSN 1089-7739