



# **PEDIATRIC SURGERY UPDATE ©**

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### **Multiple Endocrine Neoplasia**

Multiple endocrine neoplasia (MEN) refers to a familial (autosomic dominant) disorders involving several endocrine glands with hyperplasia or tumor. Two patterns are recognized: MEN 1 which comprises tumors of the pituitary, parathyroid, and pancreatic islets and MEN 2 which occurs in two forms: MEN 2A includes medullary carcinoma of the thyroid (MCT), pheochromocytoma and hyperparathyroidism, and MEN 2B with MCT, pheochromocytoma, a marfanoid habitus and neuroma phenotype. The most constant (100%) and life threatening feature of MEN 2 is MCT (usually multicentric and bilateral). Calcitonin is secreted by C cells in abnormally increase amount both in C-cell hyperplasia (precursor of MCT) and MCT. The diagnosis of MCT relies on chemical elevation of basal or stimulated (pentagastrin) calcitonin levels. The MEN 2B gene mutation (exon 16) has been located to a region of chromosome 10 that contains the ret proto-oncogene (same region for the genes for MEN 2A and familial MCT). Direct DNA testing has established the place of prophylactic surgical therapy in this familial cancer syndrome before the development of biochemical or clinical disease. Management for MCT should include total thyroidectomy, and excision of suspicious lymph nodes in the central and lateral compartments of the neck. Thyroidectomy done for elevated chemical marker has a higher rate of curability than when the diagnosis is made clinically (palpable node). After thyroidectomy the child should be yearly followed monitoring plasma levels of calcitonin and carcinoembryonic antigen to detect tumor recurrence, and catecholamines assays for pheochromocytoma.

### **Neonatal Testicular Torsion**

Neonatal testicular torsion (NTT) is a rare condition occurring most commonly during the neonatal period or before birth. The baby presents with a red, swollen scrotum the product of an extra-vaginal torsion of the spermatic cord. The diagnosis (and exclusion of other pathological conditions) is done using color Doppler ultrasound examination revealing lack of intra testicular blood flow on the affected side and normal flow within the contralateral testis (unless the condition arises bilaterally). It has been suggested that the cause of

testicular regression syndrome is antenatal torsion of the testis, others believe in vascular injury secondary to birth trauma as the most likely cause. Treatment requires surgical exploration. Controversy exists concerning the timing of exploration as well as the need for contralateral orchiopexy, since some reports suggest that the contralateral testicle is not a risk for torsion. Therapy of the ipsilateral testicle is determined by operative findings. While testicular salvage is nil (subsequent atrophy is the rule), surgical intervention is necessary for any hope of testicular preservation.

### **Intestinal Leiomyosarcomas**

Intestinal leiomyosarcomas are very rare gastrointestinal tract (smooth muscle cells) sarcomas in children. Most cases have been diagnosed during the first decade of life, almost 50% in newborns, with a slight female predominance. Microscopic appearance consists of spindle cells with blunt-ended oval nuclei, mitotic figures, anaplasia and bizarre cell forms. Clinically they present with symptoms of intestinal obstruction (intussusception) and/or perforation, other times with abdominal pain or lower GI bleeding. They are relatively small, confined and evenly distributed along jejunum, ileum and colon. Therapy consists of radical surgical resection with effort placed on removal of an adequate margin of normal tissue, even if adjacent organs are in consideration. Adjuvant chemotherapy should be considered for lesions with incomplete or questionable margins of resection. Metastasis occurs via the bloodstream and mainly to the lungs. Poor prognosis is associated to high-grade differentiation and inadequate resection, not cytometric DNA ploidy.

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