



# **PEDIATRIC SURGERY UPDATE ©**

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### **ACC**

Adreno-cortical carcinoma (ACC) is a rare potentially fatal tumor during childhood. It occurs with significant frequency in Southern Brazil. Virilization alone (pubic and/or body hair, clitoris or penis enlargement, and adult voice), or associated to Cushing's syndrome from corticosteroid hypersecretion is the most prominent feature. Feminizing tumors are uncommon and nonfunctional tumors rare. Most children are females with a median age of four years. Other signs include: abdominal mass, hypertension, seizures, and weight loss. Features associated with malignancy included high levels of 17-keto steroids, tumor weight greater than 100 g, tumor size greater than 200 cm<sup>3</sup>, and histologic evidence of diffuse growth pattern, vascular invasion, and tumor cell necrosis. CT Scan is the single most important modality in assessing primary and metastatic disease at diagnosis, and plays an important role in defining the extent of the primary lesion preoperatively. Early diagnosis and aggressive surgical extirpation may lead to an increased survival of children with ACC. An anterior thoracoabdominal approach is favored for suspected malignancies with en bloc resection. There is no evidence that adjuvant therapy provides any additional benefit. The prognosis is age related and dependent on the resectability of the tumor. Children with small adrenocortical tumors have an excellent prognosis with surgery as sole therapy.

### **Pheochromocytoma**

Pheochromocytoma is a rare APUD (amine precursor uptake decarboxylate) tumor in children. A well differentiated, functionally mature neoplasm that originates mainly from the adrenal medulla, other times from the sympathetic chain, the paraganglia or organ of Zuckerkandl. Most patients are males, with an average age of 9-11 years showing sustained hypertension (80%); 24% of these tumors are bilateral, 15% multiple and 15% extra-adrenal in location. Other signs are: headache, sweating, blurred vision, fatigue, palpitations, and anorexia. An increase familial incidence as a single Mendelian dominance is also noted in the pediatric age group. Affected patients should be screened for MEN 2. The most accurate method of diagnosis is catecholamines determination in blood and urine (VMA). CT-Scan will localize most tumors,

and the yield can be increased with the complementary use of MIBG (meta-iodo-benzylguanidine), an adrenergic tissue localizing agent, particularly in extra-adrenal or metastatic lesions. Management consists of preoperative control of hypertension with alpha adrenergic blocking agents followed by transabdominal surgical resection under strict intra-operative monitoring. The high percentage of children with bilateral, extra-adrenal, and multiple tumors warrants the need for long-term follow-up. Metastatic well-differentiated tumors are better managed with radioactive MIBG. In the malignant variety chemotherapy should be used for residual disease after surgery and for metastatic disease.

### **Pilomatrixoma**

Pilomatrixoma, also known as calcifying epithelioma of Malherbe, is a benign tumor of the skin originating from hair sheath cells. Most cases occur in children less than ten-years of age with a 2:1 female to male ratio. Presents clinically as asymptomatic superficial slow-growing dermal or subcutaneous mass most commonly located in the head and neck or upper extremity. Histology shows two characteristic epithelial cells: the basophilic and ghost cells, and calcification is usually present (these features can help the diagnosis when using FNA cytology). Pilomatrixoma can be confused with an epidermal inclusion cyst, lymph node, or calcified hematoma. The “faceted stone sign” (palpation of a tumor with a faceted rocky consistency) is evident. Management consists of local excision, and recurrences are very rare. Prognosis after removal is excellent. Multiple familial pilomatrixomas could be considered a cutaneous marker of Gardner syndrome.

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