



# **PEDIATRIC SURGERY Update** ©

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### **Gardner Fibroma**

Gardner fibromas are superficial, poorly circumscribed benign soft tissue tumors and sometimes painful lesions which consist mainly of thick haphazardly arranged collagen bundles with few interspersed spindle cells of fibroblast type. Males and females are equally affected. They are associated with Gardner syndrome and may be the first manifestation of the disease. Gardner fibromas are precursors lesion of desmoid tumors. Both Gardner fibromas and desmoid tumors are associated with Gardner syndrome. Gardner syndrome is an autosomal dominant inherited disease characterized by the triad of gastrointestinal adenomatous polyps, multiple osteomas and mesenchymal tumors of the skin and soft tissue. Gardner syndrome and Familial adenomatous polyposis (FAP) are considered to be variants of the same disease. Both are caused by mutation in the APC gene located on chromosome 5q21. The cutaneous and bone abnormalities precede development of polyposis with approximately 10 years. Other cutaneous manifestations of Gardner syndrome include neurofibromas, lipomas, leiomyomas and pigmented skin lesions. The diagnosis of Gardner syndrome can be made by genetic testing or by colonoscopy showing multiple polyps. The most common sites affected by Gardner fibromas include the back, paraspinal region and chest wall though they may appear in any part of the body. Gardner fibromas are most commonly observed in the first decade of life. The management of a Gardner fibroma consists of wide total local excision something that can be technically demanding or even impossible because of the poor circumscription or anatomic boundaries this type of tumor is characterized. Prediction of Gardner fibroma behavior is impossible, but in general 5-10% resolve spontaneously, 30% undergo cycles of progression and resolution and 50% remain stable after diagnosis, but 10% progress rapidly growing to massive sizes and infiltrating adjacent tissue. The development of a Gardner fibroma raise sentinel suspicion of Gardner syndrome or FAP which should be diagnosed by endoscopy or genetic testing.

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## Renal Rhabdoid Tumor

Renal rhabdoid tumors (RRT) are highly malignant neoplasms first described in the kidney of young children. Rhabdoid tumors also arise in extra-renal sites such as soft tissue and central nervous system. RRT have a common genetic abnormality, namely the mutation or deletion of the SMARCB1/hSNF5/INI-1 gene located at chromosome 22q11.2. A concomitant brain tumor is present in almost 21% of children with a RRT. The exact cell type of derivation of Rhabdoid tumors of the kidney remains unknown though a possible origin has been ascribed to primitive cells located in the renal medulla. Histologically RRT cells are arranged as diffuse sheets or as alveolar or trabecular pattern. RRT can occur sporadically or as part of hereditary cancer syndrome known as Rhabdoid Tumor Predisposition Syndrome. Prognosis of malignant rhabdoid tumor of the kidney is related to age at time of diagnosis and stage of disease and not to the location of the tumor. The younger the child with RRT the less probable is his long-term survival. Older than age 1.5 years at diagnosis regardless of anatomic location has higher overall survival. RRT is commonly diagnosed between 0-3 years of age with a peak incidence between age of 10-18 months. It is also associated with extensive metastasis at time of diagnosis and as mentioned before can have synchronous brain tumor. There is no standard management for RRT and the prognosis is very poor with overall survival of 15-36% though more recent treatment regimens using surgery, radiotherapy, high dose chemotherapy and autologous stem cell rescue may increase survival to 66.7% with a median follow up of almost two years. Surgical treatment of RRT follows that used in Wilms tumor. Biopsy of the primary tumor is usually not carried out prior to removal to avoid rupture of the tumor capsule and spillage of tumor cells. Germline analysis is recommended for individuals of all ages with rhabdoid tumors and prenatal diagnosis can be performed in families with a known SMARCB1 alteration. Surveillance guidelines for patients with germline mutation have been developed.

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## Endocarditis Prophylaxis

Infective endocarditis is a rare potentially life threatening infection of the heart valve or endocardium. Congenital heart disease (CHD) and palliative or corrective surgery associated to the structural defect are the two most common predisposing conditions for the development of infective endocarditis. Cyanotic CHD, left-sided lesions and endocardial cushion defects are associated with increase risk of infective endocarditis in children. The risk of developing infective endocarditis is substantially elevated during the six months postoperative period of cardiac surgery and in children less than three years of age. Lesions associated with cyanosis at birth (dextrotransposition of great arteries and tetralogy of Fallot) has the highest incidence of infective endocarditis. Infective endocarditis is less common in right sided than left sided lesion. Endothelialization of prosthetic material introduced by cardiac surgery occurs within six months following the procedure. The most common organism covered during endocarditis prophylaxis is *Streptococcus viridans* a commensal bacteria that populate the skin, oral, gastrointestinal and respiratory mucosa. The American Heart Association guidelines recommend providing prophylactic antibiotic coverage for endocarditis to children who are to undergo a procedure including dental which has an unrepaired cyanotic CHD including palliative shunts and conduits, completely repaired CHD with prosthetic material or device (whether placed by surgery or catheter intervention) during the first six months after the procedure. Also prophylaxis is recommended for repaired CHD with residual defect at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization) and cardiac transplantation recipients who develop cardiac valvulopathy. Any child with CHD undergoing a general surgery or dental procedure should be evaluated by its pediatric cardiologist in seek of advice regarding endocarditis prophylaxis. Procedures that require prophylaxis include dental including manipulation of gingival tissue or perforation of oral mucosa, tonsillectomy and adenoidectomy. Endocarditis prophylaxis is no longer needed for gastrointestinal and genitourinary procedures since they all receive broad spectrum prophylaxis by nature of the clean contaminated classification.

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