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## Gardner Syndrome

Gardner=s syndrome refers to a group of children born with familial adenomatous (multiple) polyposis and significant extracolonic manifestations. Familial adenomatous polyposis is an autosomal dominant disorder originating from a germline alteration of the adenomatous polyposis coli gene in the long arm of chromosome 5. The most significant extracolonic manifestation of Gardner=s syndrome consists of soft tissue (desmoid) tumors and osteomas. Bowel cancer develops in one-third of patients with Gardner=s syndrome from malignant degeneration of the adenomatous polyps. Desmoid tumors are typically benign but locally aggressive slow-growing tumors that surround and compress adjacent vascular structures and viscera. Affected children are asymptomatic until they manifest rectal bleeding or multiple soft/hard tissue tumors. Osteomas appear in the mandible causing dental abnormalities (odontomas, cementomas, cysts, supernumerary teeth). Surveillance by colonoscopy is imperative in affected family members. Development of a subcutaneous fibroma single or multiple that recurs as a desmoid tumor is a sentinel event identifying children with Gardner=s syndrome. Management of Gardner=s syndrome consists of excision of the soft/hard tissue tumor and removal of the affected colon (proctocolectomy) with preservation of the sphincteric muscle mechanism. Sulindac has been reported to produce drug-induced complete regression of colonic adenomas in Gardner=s syndrome.

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## Turcot Syndrome

Turcot (glioma-polyposis) syndrome refers to the presence of multiple adenomatous polyposis coli associated with glioblastoma multiforme, medulloblastoma, or glioma developing during the pediatric teens. The polyposis in Turcot is associated with a low number of polyps, large polyps over three cm in diameter, and complication by colonic cancer occurring during the second or third decades of life. Turcot syndrome is determined by an autosomal gene with pleiotropic effect and variable expressivity. Children with Turcot syndrome can develop multiple regions of congenital hypertrophy of the retinal pigment epithelium with areas of surrounding hypopigmentation in the fundi of both eyes, a fact which can help in the diagnosis. Two types of Turcot syndrome have been identified: Type I, also known as true Turcot syndrome (autosomal recessive) with less than 100 intestinal polyps, large size and apt to transform to the malignant tumor. Brain tumor is mainly diagnosed as glioblastoma or astrocytoma and mismatch repair genes might be involved. Type II with FAP-associated type (autosomal dominant) predisposing to medulloblastoma. Management in both cases is surgical.

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## Splenic Abscess

Splenic abscess is an uncommon event identified during the pediatric age. Nevertheless, is a potentially fatal disorder if not diagnosed and managed in a timely fashion. Children harboring a splenic abscess present with fever, leukocytosis and left upper quadrant abdominal pain. Simple chest films could be associated with a left pleural effusion or basal atelectasis. Most children with splenic abscess have an associated predisposing medical condition such as sickle cell disease, immune deficiency (HIV), leukemia, aplastic anemia, perforated bowel, typhoid fever, endocarditis, otitis media, appendicitis or trauma. Staphylococci, Salmonella and Escherichia coli are the most common etiologic agents in single abscess, while Candida species predominates in multiple splenic abscess. Many children are septic before the diagnosis is made. Blood cultures are seldom positive. Diagnosis is made with abdominal ultrasound or CT-Scan. Initial management incorporating the strategy of preserving the spleen consists of CT-guided percutaneous drainage and antibiotics. This combined approach is effective in two-thirds of cases. If the child does not improve, splenectomy follows. Children with immune deficiency and splenic

abscess have rapid resolution of symptoms with immediate splenectomy. Some cases with splenic abscess will completely resolve with antibiotics alone.

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