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Slow Transit Constipation

Chronic idiopathic constipation is a distressing problem in children associated with poor appetite, recurrent abdominal pain and irritability. Two types of constipation have been described: slow transit constipation (STC) associated with slow nuclear transit study of the colon, and functional fecal retention (FFR) associated with normal transit but a delay of anorectal release in the presence of ganglion cells. STC children characteristically demonstrate delayed passage of the first meconium stool beyond 24 hrs of age, symptoms of severe constipation within a year, or treatment-resistant encopresis at 2-3 years, irregular bowel motion, colicky abdominal pain, frequent uncontrollable soiling and softer stools as compared with FFR children who pass hard, infrequent stools. Transverse colon elongation is more common in SCT, whereas sigmoid colon elongation is not more common in FFR.The correlation between STC and deficiency of substance P, an excitatory neuropeptide, in the myenteric axons of the bowel wall implies that this substance is involved in the cause of the dysmotility. Management is different for both conditions. In FFR, transit as far as the rectosigmoid is normal, thus once the rectum is emptied (with enemas or rectal disimpaction) and behavioral modification is instituted the condition improves. FFR will not improve with colonic resections. With STC long-term management is warranted These children do not benefit from high fiber diet, but can be managed with laxatives and/or enemas. Those with STC that does not improve will need appendicostomy, colostomy or colonic resection. Quality of life is significant lower in children with STC as compared with normal children with both physical and psychosocial functioning score reduced.

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Intramuscular Hemangioma

The most common type of deep soft-tissue hemangioma occurs within the intramuscular compartment. Most intramuscular hemangiomas (IH) arise within skeletal muscle and occurs within the lower extremity during the first three decades of life. There is a slight female predominance. They are benign lesions histologically classified into capillary, cavernous or mixed types. The most common symptom is pain in the affected muscle region associated with a mass, absent cutaneous changes and neurologic symptoms due to nerve impingement. The most common involved muscle with IH is the quadriceps femoris muscle. Though US and CT-Scans are routinely performed, MRI remains as the imaging of preference for diagnosis (high-signal intensity on T2-weighted images) and extent of local involvement. In MRI the cavernous type is predominantly lobulated and the capillary serpiginous with the mixed type being a combination of both. Angiography can help plan surgical approach for larger lesions and even use preoperative embolization to reduce blood loss. Intramuscular cavernous hemangiomas do not undergo spontaneous regression and may be locally destructive because pressure is exerted on neighboring structures. Management of IH consists of medical and surgical modality. A trial of propranolol should be tried in small children. Surgery is indicated if there is accelerated growth, intractable pain, functional impairment, local skin necrosis, cosmetic deformity or suspicion of malignancy. To reduce recurrences, surgery consists of wide excision whenever possible avoiding permanent deformity or functional impairment.

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Branchiogenic Carcinoma

Branchiogenic carcinoma refers to a squamous cell carcinoma that arises from second branchial cleft cysts. To be labeled as branchiogenic carcinoma strict diagnostic criteria must be met: 1- the location of the tumor in the anatomic region of the branchial cleft cyst or sinus, 2- the histologic appearance of the tumor is consistent with tissue present in a branchial vestige, 3- there is no other evidence of another primary malignancy after exhaustive work-up (including nasopharyngoscopy, laryngoscopy, bronchoscopy and esophagoscopy), and 4- the histologic identification of transition from the normal squamous epithelium of the cyst to carcinoma. The tumor may be papillary within the

cyst and will always be accompanied by a significant lymphoid component. Most cases of neck cyst harboring squamous cell carcinoma are metastatic disease from a primary in the head and neck region, and not primary branchiogenic carcinoma. Management of branchiogenic carcinoma includes complete surgical removal with modified radical neck dissection. Postoperative irradiation to the ipsilateral neck is recommended. Chemotherapy is utilized when invasion to surrounding tissue is present.

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