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Idiopathic Thrombocytopenic Purpura

Idiopathic thrombocytopenic purpura (ITP) is probably the most frequent cause of persistent thrombocytopenia in children. Antedated by a viral (usually respiratory) infection, ITP is characterized by acute hemorraghic manifestations such as petechia, purpura, and mucous membrane bleeding (epistaxis). Platelet number and survival time are reduced significantly causing prolonged bleeding time. This is caused by rapid destruction of the reticuloendothelial system (spleen, bone marrow, and liver) due to IgG binding to platelets (autoinmune). Acute ITP regresses spontaneously within 3 to 6 months in 80% of cases. Initial management may consist of short steroid course and/or immunoglobulin. Chronic ITP sets itself if thrombocytopenia persists longer than six months. When this occurs, most cases are asymptomatic except during intercurrent infectious process. Platelet counts below 50,000/mm3, failure of medical therapy, long-term child restriction, or persistent bleeding may need splenectomy. Complete clinical cure is obtained in 70-80% of instances following splenectomy with a low operative risk. Recurrent symptoms may be caused by a missed accessory spleen. Laparoscopic splenectomy is a safe, cost-effective procedure with reduced hospital stay, faster recovery, better cosmetic results and least trauma. Isolated splenic thrombocytolysis and hyperplasia of megakaryocytopoiesis and of splenic follicles after splenectomy correlated with better stable remission and platelet counts. The response to splenectomy has been found to correlate directly with the response to immunoglobulin therapy. Intermittent immunoglobulin therapy with observation is a reasonable alternative until spontaneous remission occurs.

Malone Stoma

This safe and highly effective novel technique was introduced by PS Malone in 1990. Consist of building a continent appendicocecostomy through which the cecum is intermittently catheterized for administration of an antegrade enema to manage fecal soiling, incontinence, and even intractable constipation if nonsurgical management has failed. This includes children with anorectal malformations, neuropathic etiology (spina bifida, spinal cord injury),

and sphincteric trauma. Originally described as reversing the appendix and reimplanting it into the cecum with a submucous tunnel to diminish reflux, the procedure has undergone several modifications such as orthotopic cecal imbrication, the fixation of the ileocecal region at the inner side of the abdominal wall after creation of an appendicocutaneous catheterizable stoma, laparoscopic approach, tubularized cecal or ileal flap in those without an appendix, and the use of a button. The child will catheterize once or twice a day washing his distal colon. Most families are satisfied since the child spends more time in normal activities and school. A level of commitment by the child and family is needed. Children with colonic motility disorders and incontinence may not beneficiate from this procedure. The stoma might break, stenosed, closed, or bleed in a few cases. If the child is socially continent with a regimen of enemas, suppositories or washouts, he should continue that way.

RET Oncogene

The RET proto-oncogene is a protein tyrosine kinase gene (Ret protein) expressed in the cells derived from the neural crest. Germline mutations in the RET gene have been associated with neuroblastoma, pheochromocytoma, multiple endocrine neoplasia (MEN) 2, familial medullary thyroid carcinoma (MTC), radiation-induced thyroid papillary carcinoma, and recently Hirschsprung's disease. The Ret protein might have a critical role in the embryogenesis of the enteric nervous system. RET analysis is a suitable method to detect asymptomatic children with MEN at risk to develop MTC allowing us to consider thyroidectomy at a very early stage of neoplasm development (C-cell hyperplasia) or prophylactically.

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