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Thalassemia

Thalassemia, also known as Mediterranean or Cooley=s anemia, is a genetically determined heterogeneous group of hemoglobinopathies affecting the synthesis of hemoglobin alpha and/or beta-globin chains. This autosomal dominant illness can occur in major (homozygous), intermediate or minor (heterozygous) varieties. A defect in the synthesis of hemoglobin subunits results in the accumulation of intracellular particles in the red blood cell contributing to early destruction by the spleen. The anemia of thalassemia is an inborn error of metabolism causing large amounts of fetal hemoglobin to be produced, instead of adult hemoglobin. With the block in iron metabolism, large deposits of iron occur throughout the body. Heterozygous (minor) thalassemia child is asymptomatic, while the major or intermediate thalassemia child develops severe chronic anemia, jaundice, hepatosplenomegaly, frontal bossing and growth retardation. With continued hemolysis come gallbladder pigment stones (25%), hypersplenism and splenic infarcts. Splenectomy is palliative and indicated for the management of chronically transfused patients in order to increase red blood cell survival and decrease transfusion requirements. Cardiac iron overload is the most frequent cause of death from chronic transfusion therapy. After splenectomy, a high incidence of sepsis and portal vein thrombosis (hypercoagulable state) has been reported. Partial splenectomy reduces transfusion requirements for a limited amount of time due to regrowth of the splenic remnant.

References:

1- Cohen AR, Galanello R, Pennell DJ, Cunningham MJ, Vichinsky E: Thalassemia. Hematology (Am Soc Hematol Educ Program). 14-34, 2004
2- Fujita F, Lyass S, Otsuka K, Giordano L, Rosenbaum DL, Khalili TM, Phillips EH: Portal vein thrombosis following splenectomy: identification of risk factors. Am Surg 69(11):951-6, 2003
3- Al-Salem AH, Nasserulla Z: Splenectomy for children with thalassemia. Int Surg 87(4):269-73, 2002
4- al-Salem AH, al-Dabbous I, Bhamidibati P: The role of partial splenectomy in children with thalassemia. Eur J Pediatr Surg 8(6):334-8, 1998
5- Laopodis V, Kritikos E, Rizzoti L, Stefanidis P, Klonaris P, Tzardis P: Laparoscopic splenectomy in beta-thalassemia major patients. Advantages and disadvantages. Surg Endosc 12(7):944-7, 1998
6- Skarsgard E, Doski J, Jaksic T, Wesson D, Shandling B, Ein S, Babyn P, Heiss K, Hu X: Thrombosis of the portal venous system after splenectomy for pediatric hematologic disease. J Pediatr Surg 28(9):1109-12, 1993

Latex Allergy

Allergy to natural rubber latex is an increasing common condition in both children and health care workers. Almost 20 to 40% of children with spina bifida are allergic to latex. Other affected persons are health care workers, latex industry workers, immune

compromised individuals, children with bladder exstrophy, anorectal anomalies, and persons with positive risk factors such as multiple surgical procedures. Chemical additives in latex gloves can cause an irritant or allergic contact dermatitis. Latex proteins are responsible for most of the immediate IgE-mediated hypersensitivity allergic reactions. Symptoms range from rhinitis, conjunctivitis and urticaria to intraoperative anaphylaxis and death. Skin prick testing with natural rubber latex and glove tests are safe diagnostic procedure. In children with spina bifida significant and independent risk factors identified for latex sensitization are multiple interventions and higher levels of total serum IgE. The only currently available treatment is complete avoidance of latex. For children with known history of latex allergy premedication with antihistamines and steroids is in order.

References:

1- Birmingham PK, Suresh S: Latex allergy in children: diagnosis and management. Indian J Pediatr 66(5):717-24, 1999
2- Degenhardt P, Golla S, Wahn F, Niggemann B: Latex allergy in pediatric surgery is dependent on repeated operations in the first year of life. J Pediatr Surg 36(10):1535-9, 2001
3- Zucker-Pinchoff B, Stadtmauer GJ: Latex allergy. Mt Sinai J Med 69(1-2):88-95, 2002
4- Cremer R, Kleine-Diepenbruck U, Hering F, Holschneider AM: Reduction of latex sensitization in spina bifida patients by a primary prophylaxis programme (five years experience). Eur J Pediatr Surg 12 Suppl 1:S19-21, 2002
5- Sparta G, Kemper MJ, Gerber AC, Goetschel P, Neuhaus TJ: Latex allergy in children with urological malformation and chronic renal failure. J Urol 171(4):1647-9, 2004
6- Kimata H: Latex allergy in infants younger than 1 year. Clin Exp Allergy 34(12):1910-5, 2004

Hermansky-Pudlak Colitis

Hermansky-Pudlak syndrome is a triad of tyrosine-positive oculocutaneous albinism, platelet dysfunction, and the deposition of an abnormal ceroid-like pigment in the tissues. Ceroid lipofuscin is accumulated in lysosomes organelles. Children with Hermansky-Pudlak syndrome can suffer from pulmonary fibrosis, renal failure, and cardiomyopathy besides other complications. All identified affected patients in the northwest Puerto Rico are homozygous for a 16-bp duplication in exon 15 of a recently cloned gene. This duplication is associated with a broad range of pigmentation and an increased risk of restrictive lung disease in adults. Several families with the syndrome rarely suffer from granulomatous colitis, a unique type of inflammatory bowel disease with clinical features suggestive of idiopathic ulcerative colitis and pathologic features suggestive of Crohn's disease with perineal and perirectal involvement. Diagnosis is established with endoscopy. Management is similar to cases with inflammatory bowel disease directed toward pathogenetic mechanisms. Corticosteroids, sulphasalazine and the new salicylates, the immunosuppressants azathioprine, 6-MP and, more recently, cyclosporin and metronidazole have become the accepted and standard forms of treatment. Some cases are refractory to medical treatment needing segmental resection of the affected bowel.

References:

1- Sherman A, Genuth L, Hazzi CG, Balthazar EJ, Schinella RA: Perirectal abscess in the Hermansky-Pudlak syndrome. Am J Gastroenterol. 84(5):552-6, 1989

2- Witkop CJ, Nunez Babcock M, Rao GH, Gaudier F, Summers CG, Shanahan F, Harmon KR, Townsend D, Sedano HO, King RA, et al: Albinism and Hermansky-Pudlak syndrome in Puerto Rico. Bol Asoc Med P R. 82(8):333-9, 1990
3- Schinella RA, Greco MA, Cobert BL, Denmark LW, Cox RP: Hermansky-Pudlak syndrome with granulomatous colitis. Ann Intern Med. 92(1):20-3, 1980
4- Gahl WA, Brantly M, Kaiser-Kupfer MI, Iwata F, Hazelwood S, Shotelersuk V, Duffy LF, Kuehl EM, Troendle J, Bernardini I: Genetic defects and clinical characteristics of patients with a form of oculocutaneous albinism (Hermansky-Pudlak syndrome). N Engl J Med. 338(18):1258-64, 1998
5- Sandberg-Gertzen H, Eid R, Jarnerot G: Hermansky-Pudlak syndrome with colitis and pulmonary fibrosis. Scand J Gastroenterol. 34(10):1055-6, 1999
6- Mahadeo R, Markowitz J, Fisher S, Daum F: Hermansky-Pudlak syndrome with granulomatous colitis in children. J Pediatr. 118(6):904-6, 1991

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