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Polydactylism

Polydactylism (accessory finger or toe) is one of the most common congenital anomaly of hands or feet in children. The prevalence of polydactylism is almost two cases for every each 1000 born alive newborns. Familial occurrence is associated in almost 20% of cases with variable gene penetrance. Polydactylism of the hand is more common than in the foot. In the hand the ulnar accessory finger predominates. A few cases undergo traumatic intrauterine amputation, a condition known as rudimentary polydactylism. Different genes are involved in the pathomorphogenesis of postaxial polydactylism. Polydactyly may be preaxial (medial, thumb side) or tibial (hallux-side), postaxial (lateral) or ulnar (side of the little finger or toe), and central (middle fingers or toes). The duplication may appear at the distal and medial phalanges or at the whole digit. Surgical amputation of the affected finger or toe is indicated for cosmetic reasons or for functional disturbances in wearing shoes respectively. Careful clinical and radiographic evaluation should be made prior to treatment to achieve good functional and cosmetic results. Polydactylism can be diagnosed prenatally and when isolated is associated with good perinatal outcome. Primary suture ligation of accessory digits in infancy can be associated with later development of neuroma in the stump. Identification and high transection of the accessory digital nerve is essential in the treatment of pedunculated supernumerary digits.

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Postsplenectomy Guidelines

Removal of the spleen in children is considered necessary in some hemolytic diseases and trauma. Hemolytic disease where splenectomy is therapeutic includes idiopathic thrombocytopenic purpura, congenital spherocytosis and Sickle cell disease. Removal of the spleen with its attendant immunologic shortness can cause overwhelming sepsis. Overwhelming postsplenectomy infection is a fulminant process that carries a poor prognosis.The risk of serious infection is highest among young children, in immunologically

compromised individuals and in the first few years after splenectomy. Current guidelines for children undergoing elective splenectomy include immunization for pneumococcus, meningococcus and hemophilus at least two weeks prior to the procedure. After emergency splenectomy the immunization protocol is the same, except that the amnestic response of the host is lower. Another guideline for splenectomized children includes the use of continuous antibiotic prophylaxis (penicillin) until the age of twelve years. Compliance with this antibiotic regimen is poor. It is recommended that parents be advised to bring the postsplenectomy child to the hospital anytime an illness or fever develops that might require an immediate loading dose of an appropriate antibiotic.

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Splenic Artery Embolization

Splenic artery embolization as an alternative for splenectomy is a minimally invasive technique that has been utilized for the past 25 years. The procedure is done under local anesthesia, sedation or general anesthesia. Partial splenic artery embolization has been used in cases of thalassemia major to reduce transfusion requirements with variable results. Also in children with secondary hypersplenism or massive spleens due to portal hypertension, variceal bleeding (portal vein thrombosis, biliary atresia and biliary cirrhosis) or myelodysplastic disorders. The leukopenia and thrombocytopenia is corrected temporarily and variceal hemorrhage is ameliorated in most cases. Objective is to embolize at least two-third of the splenic circulation. After splenic artery embolization the child develops prolonged fever, pain in the left hypochondrium, leukocytosis and ileus. If the embolization causes total shutdown of the arterial circulation the child might develop a subcapsular hematoma with effusion, abscess formation or need for open splenectomy. Embolization allows for safe surgical splenectomy. Long term follow-up shows evidence of partial splenic regeneration. Partial splenic embolization is a useful method for reducing serum bilirubin concentrations in patients with hypersplenism following the Kasai procedure for biliary atresia, has also been found safe and effective minimally invasive treatment for patients with bleeding from a blocked distal splenorenal shunt, and as therapy for post-traumatic splenic artery pseudoaneurysm.

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