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Budd-Chiari Syndrome

Obstruction to the hepatic venous outflow tract is commonly known as Budd-Chiari Syndrome. The Budd-Chiari syndrome (BCS) in children can be the result of a congenital or acquired web in the inferior vena cava, a thrombotic, inflammatory, neoplastic process or an hypercoagulable state (antithrombin 3 deficiency). Hepatic venous outflow obstruction produces hepatic dysfunction producing abdominal pain, ascites, jaundice, hepatosplenomegaly, portal hypertension and cirrhosis. The factors that influence management of the BCS include the state of hepatic dysfunction, type of presentation (acute or chronic), how much venous occlusion is present and the presence of collateral circulation. Pulsed Doppler ultrasound, venography and liver biopsy are very helpful in diagnosis. Management of BCS in children has included use of anticoagulation, thrombolytic therapy, angioplasty with or without stenting, transjugular intrahepatic portosystemic shunts and surgical portosystemic shunts. This last choice has fewer options in the face of liver transplantation and does not improve survival. The combination of thrombolytic therapy and balloon angioplasty is the best option in the acute setting of BCS or during the first four weeks after development of the syndrome. Late or chronic presentation with established hepatic cirrhosis and portal hypertension sequelae is best managed with liver transplantation. Early diagnosis offers the best possible chance of cure.

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Pyoderma Gangrenosum

Pyoderma gangrenosum is a rare, poorly understood, ulcerative skin disorder that occurs in

all age groups. A systemic disorder such as inflammatory bowel disease (e.g., ulcerative colitis, Crohn's disease), malignancy or juvenile rheumatoid arthritis is usually associated in almost three-fourth of cases seen in children. Pyoderma Gangrenosum (PG) often begins as a small pustule, but results in localized skin destruction and ulceration which is characterized by an expanding ulceration with undermined violaceous borders. PG characteristically involves ulceration in the buttocks, thighs and perianal area while sparing the legs. Lower legs are the most commonly affected sites in adults. Infants appear to have an unusual distribution of perianal and genital lesions not often described in other age groups. The distribution of lesions in children is similar, often involving the lower extremities, but pyoderma gangrenosum of the head and face appears to be more common in children. Infants may have ulcers in genital and perianal areas. An altered immune response could be the origin of PG. Diagnosis is established by biopsy. Histologically, lymphocytic and/or leukocytoclastic vasculitis is present in most of the biopsy specimens obtained from the borders of the lesions. The most frequently prescribed treatment for children is systemic corticosteroids, which generally are very effective.

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Sertoli-Leydig Ovarian Tumors

Sertoli-Leydig cell ovarian tumors are rare androgen producing tumors causing masculinization in most girls. A few are nonfunctional tumors. Sertoli-Leydig cell tumors used to be called arrhenoblastoma or androblastomas. One-third of all Sertoli-Leydig cell tumors (SLCT) occurs in children. Most SLCT are unilateral. Histologic diagnosis depends on the presence of heterologous endodermal and mesenchymal components. The androgenic effect of the tumor causes accelerated somatic growth and amenorrhea in prepubertal girls. Postpubertal girls develops irregular menstrual cycles, hirsutism and masculinization. Most affected children usually present with a pelvic mass. Testosterone and alpha-fetoprotein produced by the tumor are used as genetic tumor markers. Diagnosis is usually done by ultrasound or CT-Scan in association with the masculinizing clinical picture. Management consists of unilateral salpingo-oophorectomy. Poorly differentiated tumors might need adjuvant chemotherapy and radiotherapy. Prognosis correlates most meaningfully with the stage and degree of differentiation of the tumor. High-stage tumors are all clinically malignant.

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