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Neck Adenopathy

Cervical adenopathies are the most common neck masses identified in children. Though they may occur in any location, the anterior cervical chain position is the most common presentation. Lymph node enlargement is usually preceded by a viral upper respiratory infection Nodes are soft, mildly tender and usually decrease in size after a short course of antibiotics. Acute suppurative adenitis occur during early childhood (six months to three years of age) most commonly in submandibular and deep cervical nodes preceded by pharyngitis. Nodes develop erythema, swelling and cellulitis. Infecting agents include staph aureus and group A streptococcus. Other less common organisms are fungi, atypical mycobacteria and Bartonella species. Antibiotics and drainage are curative. Chronic adenitis refer to nodes that persist enlarged for more than six-weeks. Tonsillar nodes (solitary, non-tender, mobile and soft) are most commonly affected by a reactive hyperplasia process and can be observed. Work-up should include complete blood count, ESR, skin (PPD) and Mono test, chest film and ultrasound looking for the perfusion patterns of affected lymph nodes. Indications for biopsy of chronic lymphadenopathies include older children (> eight years), rapid growth, nodes greater than two centimeters, supraclavicular position, cluster - hard - fixed non-tender characteristic, an abnormal chest film, history of malignancy or very anxious parents. Biopsy is done of the largest node under general anesthesia as an outpatient procedure.

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Familial Adenomatous Polyposis

Familial history of colon cancer is an important indicator of future risk for colorectal cancer. The more extensive and closer the affected relatives, the greater the risk. Highest risk is found in Familial Adenomatous Polyposis (FAP). Once the risk is appreciated screening for the disease must take place. This involves genetic analysis for members of syndrome

families along with lower gastrointestinal endoscopy for the rest as polyps can occur throughout the gastrointestinal tract. FAP is a genetic (autosomal dominant) premalignant condition that will ultimately manifest with the development of colorectal carcinoma. FAP has been linked to germline mutations of the adenomatous polyposis coli (APC) gene. Initial presentation can be rectal bleeding. Besides multiple adenomatous polyps of the colon predisposing to malignancy at an early age, a variety of extra colonic manifestations are associated with this condition. Once the diagnosis of FAP is established endoscopic surveillance should be instituted. All polyps should be subjected to histopathological exam to determine presence of adenomatous epithelium. With the presence of dysplastic changes total colectomy with the creation of an ileorectal or ileoanal (pouch) anastomosis is recommended. Almost one-third of cases develops adenomas in the ileal pouch after proctocolectomy. Baseline small bowel enteroscopy should be done at the time of surgery and in the postop period in children with FAP and juvenile polyposis. With duodenal polyps enteroscopy should be done at the time of surgery. Biopsy and/or excision of larger polyps should be done as they may harbor a carcinoma.

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Denys-Drash Syndrome

The Denys-Drash Syndromes (DDS), first described in 1967, encompass the combination of male pseudohermaphroditism (XY gonadal dysgenesis), early onset glomerulopathy caused by diffuse mesangial sclerosis, Wilms' tumor, and constitutional mutations in the WT1 suppressor gene. WT1 gene, found in 11p13 chromosomal region, expresses a regulated transcription factor of the zinc-finger family proteins restricted to the genitourinary system, spleen, dorsal mesentery of the intestines, muscles, central nervous system and mesothelium. Mutations in the WT1 gene have been found in less than 10% of Wilms tumors specimens examined and in greater than 95% of DDS patients. Most of the mutations described are dominant missense mutations. Wilms tumor associated with

DDS occurs at a younger age. Since DDS patients eventually go into end stage renal failure, the diagnosis of DDS should be suspected in any child with Wilms tumor that develops renal failure. Patients with Wilms' tumor and aniridia or genitourinary malformation should be followed closely throughout life for signs of renal failure.

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