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BMT-CVC

Bone marrow transplant (BMT) recipients are an immunocompromised group that needs multiple venous access to meet all the fluid, antibiotic and nutritional requirements during periods of intensive supportive care. All BMT children will have central venous catheters (CVC) inserted before intensive therapy is initiated. CVC are silicone-made multiple lumen catheters (Raaf, Broviac, Hickman, Leonard) inserted in the operating room under local or general anesthesia using the subclavian, external jugular, or internal jugular veins. Main reasons to remove the CVC in BMT are: end of therapy, a complication has developed, or the child dies. Complications can be divided into infectious (local or systemic), mechanical (inability to infuse or withdraw blood, accidental dislodgement, pinch-off syndrome), technical (malfunction), or thrombotic. Infection (20-40%) is the most common reason for early catheter removal in this pancytopenic population. Not all episodes of sepsis results in CVC loss since bacteremias associated with skin flora (coagulase negative staphylococci) are successfully treated with systemic antibiotics. Infections has been associated with preparation, delivery and type of solution infused via the CVC, multiple use of CVC lumens, degree of adherence to strict protocols for IV tubing and dressing changes, properties of the catheters, and host immune status of the child. Tunnel or exit site infections will need catheter removal. Occlusive episodes can be managed with urokinase administration.

Neurofibromatosis

In 1882 Von Recklinghausen described neurofibromatosis (NF) as a syndrome of multiple skin hyperpigmentation (café au lait spots) associated with subcutaneous tumors of neural origin (neurofibromas) and skeletal malformations. Some lesions are evident at birth, while others are observed in later childhood. Neurofibromas are the most common nerve tissue tumors in childhood, may occur in any part of the body where nerve fibers exist, specially subcutaneous tissue, and are histologically classified as solitary, plexiform and diffuse. Pain from pressure of a nerve in an enclosed space is the most common symptom. NF is inherited as autosomal dominant with high

penetrance and wide variability in expression. The size and number of café au lait spots tend to increase throughout puberty. NF is considered in two forms: NF I - peripheral predominance (cutaneous, visceral and skeletal, lesions can be lytic to bone, space occupying and or disfiguring), or NF II - central with CNS orbito-fascial involvement (high mortality). Complications of NF are divided into: structural (disfiguration, macrocephaly, scoliosis, congenital tibial pseudarthrosis), functional (pain, seizures, speech and intellectual deficit) and malignancy (sarcomatous degeneration). Malignant cases are more common males than females, and seen with larger symptomatic tumors. Complete excision of these lesions is often difficult and local tumor recurrence is common. Lifelong close observation is warranted in most patients.

Retroperitoneal Teratomas

Retroperitoneal teratoma is infrequent (5% of all teratomas). They are classified as mature, immature or malignant. Most patients are female, younger than six months, and asymptomatic. Clinically they present with increased abdominal girth or weight loss. An abdominal mass is commonly identified, and the presence of a tooth or a definitive bony structure in abdominal films is identified. Ultrasonography and contrast enhanced CT-Scans are of benefit in locating as well as diagnosing abdominal teratoma. Elevated alpha-feto protein level is found in malignant and some immature cases. Surgical excision is curative for the majority of cases since they are loosely attached to surrounding structures. Most children will have benign mature teratoma and show no evidence of recurrence. Retroperitoneal tumors containing high-grade immature elements should be managed with adjuvant chemotherapy.

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