



PEDIATRIC SURGERY Update © **Vol. 41 No. 04 OCTOBER 2013**

Thromboprophylaxis

Thromboprophylaxis is utilized to prevent and reduce the incidence of hospital-acquired life-threatening venous thromboembolism (VTE) events such as deep venous thrombosis and pulmonary embolism. The incidence of VTE in children has increased in tertiary care centers. The presence of a central venous catheter is the most prevalent risk factor for VTE in pediatric patients. Risk factors associated with VTE include acute conditions such as major lower extremity orthopedic surgery, spinal cord injury, major trauma to lower extremity, lower extremity central venous catheter, acute infection, burns and pregnancy. Chronic conditions include obesity, estrogen containing medication, inflammatory bowel disease, nephrotic syndrome, known thrombophilia. Other risk factors include past history of previous DVT/PE or family history of VTE in first degree relatives. Adolescent above the 14 years of age with above risk factors should receive prophylaxis. Intervention for thromboprophylaxis includes early and frequent ambulation, good hydration for low risk children; mechanical prophylaxis using graduated compression antiembolic stockings or sequential pneumatic compression for moderate risk; and anticoagulant prophylaxis with enoxaparin or fractionated heparin for high risk patients. For immobile patient sequential compression is preferred. Contraindications for anticoagulation include intracranial hemorrhage, acute stroke, uncontrolled hemorrhage, coagulopathy, incomplete spinal cord injury, allergy and heparin induced thrombocytopenia. Every institution managing children at high risk should institute an algorithm of risk assessment and prophylaxis to prevent VTE. Providing thromboprophylaxis to children is cost-effective.

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Mucopolysaccharidosis

Mucopolysaccharidosis (MPS) are a group of metabolic disorders due to absence or malfunctioning of a lysosomal enzyme needed to breakdown molecules called glycosaminoglycans causing a storage lysosomal disease. Children with MPS are at high-risk for significant perioperative mortality. Excessive secretions, difficult or failed intubation, need for emergency tracheotomy and intraoperative cardiac arrest have been described in MPS patients. The most studied is MPS type I caused by a deficiency of lysosomal enzyme alpha-L-iduronidase producing accumulation of dermatan sulfate and heparan sulfate in the lysosomes. There is a spectrum of clinical disease involvement depending on age of onset, progression, cognitive involvement and organ involvement. Management of children with MPS type I include hematopoietic stem cell transplantation and recombinant human alpha-L-iduronidase enzyme replacement. Disease-related airway issues have been shown to increase the risk of transplant in MPS type I. Many deaths associated to MPS I are due to upper airway obstruction encountered during anesthetic care specially in children with the most severe phenotype. Numerous airway problems have been reported, including obscured airway landmarks owing to excess glycosaminoglycan deposition, copious thick secretions, narrow stiff airways, and difficulty oxygenating owing to glycosaminoglycan deposition within alveoli. Surgical mortality may be greater in these undiagnosed patients who are unlikely to be referred to anesthesiologists with expertise in managing difficult airways or to undergo other precautionary measures. Physicians should become familiar with the physical characteristics and surgical history that suggest MPS disorders and refer such patients to geneticists for evaluation before surgery.

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Trichilemmal Cyst

Cystic lesions of the skin in children are fairly common. Most cases are either sebaceous or pilomatrixoma cysts. Cysts where keratinization occurs without keratohyaline granules derived from the follicular isthmus of the external root sheath of

the hair follicle are called trichilemmal or pilar cysts. Trichilemmal cysts occur most commonly in the scalp due to the dense hair follicle concentration. Face, trunks, back and forehead are the other common site in that order. Trichilemmal cysts can occur as sporadic lesions or in hereditary-familial settings with autosomal dominant transmission. Though almost always benign, malignant transformation can occur rarely. They may be locally aggressive becoming large and ulcerated. Proliferating trichilemmal tumor is a solid cystic neoplasm that shows differentiation similar to that of the isthmus of the hair follicle. Trichilemmal cysts are usually a solitary intradermal or subcutaneous lesions. The cyst is lined by stratified squamous epithelium. They can grow to large sizes. Management of trichilemmal cyst consists of surgical excision. The cytologic diagnosis of pilar cysts is important because these cysts recur if incompletely excised and often undergo transformation to pilar tumors.

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