



PEDIATRIC SURGERY Update © **Vol. 30 No. 03 MARCH 2008**

Cutaneous Mucinosis

Frequently pediatric surgeons are asked to evaluate a skin disorder in children. Most cases are evaluated and managed by dermatologists, but in a few patients due to the nature of the lesion or age an excisional or incisional biopsy is required to establish the histologic diagnosis. Such is the case of juvenile cutaneous mucinosis, a very rare disorder of skin found in children. Characteristically cutaneous mucinosis is a nontender hypopigmented plaque-like raised papula which can appear in the trunk or extremity singly or with several other associated lesions. Cutaneous mucinosis are a heterogeneous group of diseases in which mucin accumulates in the skin or within the hair follicle. Mucin is a gelatinous substance composed of glycosaminoglycans, especially hyaluronic acid and dermatan sulfate bound to small quantities of chondroitin sulfate and heparin sulfate. Two groups are identified: the primary cutaneous mucinosis in which the mucin deposit is a distinctive histopathologic feature that manifests as a clinically specific lesion, and the diseases associated with histopathologic mucin deposition as an additional finding (secondary) such as myxedema, lupus erythematosus or scleroderma. The primary juvenile variety occurs in children characterized by an early age of onset, the presence of plaques and nodules in a characteristic distribution, and rapid onset followed by spontaneous resolution of the lesions within a period of weeks to months.

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Werdnig-Hoffmann Disease

Spinal muscular atrophy is a common autosomal recessive neuromuscular disorder characterized by degeneration of motor neurons of the spinal cord. Werdnig-Hoffmann disease (WHD), the second most common neuromuscular disease in childhood, is a type of spinal muscular atrophy. Werdnig-Hoffmann is subdivided into two groups on the basis of a combination of age of onset, milestones of development and age of survival. Type I has an

acute onset before six months of life and the progression of disease is severe with a more uniform poor prognosis and early death. They showed generalized hypotonia, abnormal respiration and could not sit without support. Type II onset of the disease is between the age of six and 18 months and progression is slower. A gene termed 'survival of motor neuron' (SMN) has been recognized as the disease-causing gene. SMN encodes a protein located within a novel nuclear structure and interacts with RNA-binding proteins. Pre- or postnatal diagnosis is made with this genetic testing. There is no effective therapy for WHD. Management consists of preventing or treating the complications of severe weakness, such as restrictive lung disease, poor nutrition, orthopedic deformities, immobility, and psychosocial problems. Tracheostomy, gastrostomy and fundoplication with non-invasive mechanical ventilation can help prolong life in WHD.

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Treadmill Injury

Injury is the leading cause of preventable death and disability in childhood and early adulthood. Most of us have a treadmill, or jogging machine in our house to stay fit. Treadmills have been found to account for an increase in the number of injuries in our children. Approximately 25,000 children with a median age of 2.5 years are injured on exercise equipment each year. Jogging machines can cause friction burns, abrasions, blunt trauma, or even amputation. Most of these injuries will require surgical intervention and rehabilitation. Almost all injuries are friction injury due to contact with the moving treadmill belts. Friction injuries that convert into a full thickness burn needing skin grafting surgery. The upper extremity is more commonly affected than the lower extremity with almost 75% of cases involving the palmar aspect of the hand. Most common mechanism is when the machine is in use by an adult and a curious toddler comes toward the running treadmill. Other cases are the older child who has the height to reach and activate the machine sustaining injury when they fall. Prevention modalities include additional manufacture safety features, warning labels, and parental education. Parent supervision during use of this type of machine is paramount. Recommendations include limiting children access, facing the treadmill toward the open room, use a back mirror, and avoiding the use of headsets while on the treadmill.

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