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Haddad Syndrome

Haddad syndrome is a rare disorder considered a neurocristopathy, a set of disease processes characterized by maldevelopment of the neural crests. Neurocristopathies are a group of diverse disorders resulting from defective growth, differentiation, and migration of the neural crest cells. Children with Haddad syndrome present with the combination of congenital central hypoventilation syndrome (also known as Ondine=s curse), and Hirschsprung=s disease (HD). Almost 10% of these cases of Hirschsprung=s disease have total intestinal aganglionosis. The initial clinical manifestation in the neonatal period is apnea of no identifiable cause followed by constipation or bowel obstruction. Strong clinical suspicion, rectal biopsy and genetic mutation detection makes the diagnosis of Haddad syndrome. Other associated features includes ophthalmic abnormalities, esophageal dysmotility, sensorineural hearing loss, neural crest tumors and signs and symptoms of autonomic nervous system dysfunction. A genetic basis for Haddad syndrome has been suggested associated with a mutations detection rate above 90% in chromosome 4p12 PHOX2B gene. Inheritance is autosomal dominant. Management consist of tracheotomy, home ventilatory support, TPN, proximal decompressive ostomy and long small bowel myectomy-myotomy. The prognosis is poor specially in underdeveloped countries.

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Disc Cell Battery Ingestion

Preschool children and toddlers enjoy taking things from their hand to their mouth. This includes disc, button or coin cell batteries with more than 3000 coin cell battery ingestion reported yearly in the United States. Button batteries are being used with increasing frequency in a variety of devices including hearing aids, watches and

calculators. Most of these ingested foreign bodies will pass the gastrointestinal tract without causing harm, but a few will produce a very serious complication. Such rare complications include esophageal perforation & stricture, aortoesophageal fistula, gastric perforation, tracheoesophageal fistula and vocal cord paralysis. The tissue damage that result from contact with charged battery is a chemical burn caused by production of sodium hydroxide (cathode) and hydrochloric acid (anode) generated from electric current passing through physiologic electrolyte solution. The alkaline burn with liquefaction necrosis, fat saponification and inflammatory cell infiltration causes the most severe histologic injury. It's not caused by the content of the battery or pressure necrosis changes. Coin cell batteries differ from coin currency in simple x-rays. If the battery impacts in the esophagus or hypopharynx, emergency endoscopic management is necessary. Once in the stomach, the battery will usually pass through the gastrointestinal tract without long-term complications. Its passage can be monitored with serial radiographs.

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Proteus Syndrome

Proteus syndrome (PS) is a rare congenital hamartomatous syndrome that causes sporadic overgrowth of multiple tissues in a patchy or mosaic pattern. The overgrowth can involve skin, subcutaneous tissue, connective tissue (including bone), the central nervous system, and viscera. Complications of PS include progressive skeletal deformities, plantar gigantism of the hands and feet, invasive lipomas, benign and malignant tumors, and deep venous thrombosis with pulmonary embolism. The name Proteus comes from a Greek mythical sea god who was able to change his body form freely. The disease process is not usually apparent at birth but develops rapidly in childhood. Common manifestations include macrodactyly, vertebral abnormalities, asymmetric limb overgrowth and length discrepancy, hyperostosis, abnormal and asymmetric fat distribution, asymmetric muscle development, connective-tissue nevi, and vascular malformations. Diagnosis and management of the disease depend heavily on clinical evaluation and imaging using strict criteria. Histopathological features of lesions resected from children with PS predominantly include hamartomatous mixed connective tissue lesions, benign neoplasms such as lipomas, and lymphatic-rich vascular malformations. Potential complications such as difficult intubation, pulmonary hypertension, and pulmonary thromboembolism necessitates careful preoperative and

anesthetic preparation.

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