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Tracheomalacia

Tracheomalacia refers to a structural/functional generalized or localized weakness of the tracheal rings' support resulting in partial respiratory obstruction. Most cases are associated with esophageal atresia and as such flaccid tracheal development after external pressure from the dilated proximal blind esophageal segment has been proposed as pathogenetic mechanism. Vascular rings, prolonged ventilatory support and tracheotomy are secondary causes of tracheomalacia. Most cases develop expiratory obstruction since only the intrathoracic trachea if affected. The harsh barking cough is the most characteristic initial symptom. Nutritional problems are the result of difficulty breathing as cyanotic attacks might occur during feeding. Other incitatory elements are intercurrent respiratory infections and aspiration. Severe forms are characterized by life-threatening apneic spells, inability to extubate the airways, and episodic pneumonia. A cough and wheeze may progress to complete airway obstruction and cyanosis. Diagnosis is obtained with simple lateral thoracic films (narrow slit-like appearance), bronchoscopy during spontaneous breathing (antero-posterior narrowing in expiration), cinetracheobronchography (allows extent of tracheal collapse) or cine CT studies. Reflux must be rule out and manage aggressively. For mild to moderate symptoms no management is necessary as the child will improve with time. For severe life threatening tracheomalacia aortopexy must be undertaken. Failed aortopexy may need tracheal reinforcement with autologous cartilaginous grafts.

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Munchausen by Proxy

Munchausen by proxy (MBP) syndrome refers to a behavioral affected parent or caretaker that fabricates or induces an illness in a child and persistently seeks medical care. MBP affects children less than six years of age. The working definitions for MBP are: illness in a child simulated or produced by a parent, persistent presentation of the child for medical

care, denial of knowledge by the parent as to the etiology of the child's illness and acute symptoms and signs in the child that abates when he is separated from the instigator. The most common symptoms are: bleeding, seizures, central nervous system depression, apnea, diarrhea and vomiting. The perpetrator (mostly the mother) uses a variety of methods to obtain this means: strangulation, poisoning, laxative administration, etc. The most difficult issue is diagnostic confirmation, parental confrontation and optimal medicolegal disposition of involved children. Secret video telemetry, poisonous toxin screening and detailed past medical history collection are useful. Children might have an unnecessary diagnostic tests and operation (fundoplication, central venous catheter placement) before establishing the diagnosis of MBP. When faced with a patient with enigmatic signs and symptoms and a family with classic personality traits consider the diagnosis. Disappearance of symptoms after removal of the suspected perpetrator remains the key to diagnosis.

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Blue Rubber Bleb Nevus Syndrome

Blue rubber bleb nevus (BRBN) syndrome (also Bean's syndrome) is a rare congenital condition characterized by the presence of multiple angiomatic lesions in the skin (soft, rubbery, and compressible). They are associated with similar lesions in other organs, namely the gastrointestinal tract and oral cavity causing anemia through chronic bleeding or intussusception. Skin or endoscopic biopsy reveals the lesions to be cavernous hemangiomas. Clinically the child presents with hematemesis, melena and has multiple bluish rubber bleb-like hemangiomas over the body, in the stomach, jejunum and colon. The syndrome is likely caused by a gene mapping to chromosome 9p and shows autosomal dominant inheritance. Alpha-2a interferon therapy has been found beneficial for relieving the life-threatening consumptive coagulopathy associated with BRBN. The GI hemangiomas can be managed with intermittent laser-steroid therapy. Therapy is mainly symptomatic directed to complications.

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