



Hemolytic-Uremic Syndrome

Hemolytic-Uremic Syndrome (HUS) refers to the constellation of signs and symptoms that occurs after enteric infection with the verotoxin-producing bacterium *Escherichia coli* O157:H7. The organisms are carried in the intestines of cattle; partially cooked contaminated hamburger is the single most common vector. *E. coli* produces a potent cytotoxin that gains access to the circulation, is taken up by glycolipid receptors on intestinal and glomerular endothelial cells causing cell death. Usually the HUS triad consists of an microangiopathic hemolytic anemia, thrombocytopenia and acute renal failure. HUS is the most common cause of acute renal failure in infants and young children. The prodromal phase of HUS starts with fever, vomiting, crampy abdominal pain and bloody diarrhea. In a few occasions the child will develop signs of peritoneal irritation mimicking an acute abdomen. After the prodromal phase the child develops pallor, severe anemia, petechia, oliguria, edema, hypertension and electrolyte disturbances. Due to the associated inflammatory enterocolitis simple abdominal films might show thumbprinting indicative of edema and intestinal mucosal hemorrhage. Treatment of HUS is supportive, but plasma exchange may be useful in selected high-risk subsets. Antibiotics have not yielded any benefit. Plasma infusions and plasma exchange appear to be efficacious. Dialysis is necessary in many children. Surgical complications associated with HUS include bowel perforation (pneumoperitoneum), intussusception or later development of intestinal strictures.

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Giant Pigmented Nevus

Giant Pigmented Nevus (GPN) refers to a congenital melanocytic nevi characterized by a diameter of 20 cm or greater in adulthood, or a lesion occupying greater than 2% of the body surface area of an infant or child. Generally, melanocytic nevi is solitary, but smaller satellite lesions may be present within a GPN. The mode of inheritance of GPN is probably

multifactorial with a 2:1 female predominance. The lesion is typically brown pigmented, varies in size and shape, contains hair and can be found anywhere on the face, neck, trunk or extremity of the child. GNP extensively involving the extremities can result in reduced growth of the affected limb. GPN has a higher incidence of malignant transformation (melanoma formation) due to the increase number of nevus cells at risk for such transformation. Lifetime incidence of malignant transformation is four to 6%. This capacity of GPN to developed into malignant melanoma is the main reason for opting for prophylactic surgical excision as treatment of choice. Tissues expansion is the primary modality for excision and reconstruction in the face and scalp since it can be started early in life and repeated as required. For the trunk excision with abdominoplasty or skin graft is preferred. In the extremity excision and skin grafting is best option. Dermabrasion or laser therapy cannot ensure complete removal of nevus cells.

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Alveolar Capillary Dysplasia

Alveolar capillary dysplasia (ACD), also known as acinar dysplasia, is a rare cause of severe and irreversible pulmonary hypertension that usually presents with acute onset respiratory failure during the first 48 hours of life. ACD is characterized by muscularization of pulmonary arterioles, deficient number of alveolar units and pulmonary capillary vessels with thickened interalveolar septa. ACD also shows abnormally immature parenchymal development in the lungs. The primary pulmonary vascular anomaly is likely to be a failure of fetal lung vascularization dating from the second trimester of fetal life. ACD is uniformly fatal due to inefficient gas exchange. Usual presentation is a term neonate, normal at delivery that develops sudden hypoxia, respiratory acidosis and hypotension within 48 hours of life. Associated anomalies include cardiac, intestinal and GU anomalies. Management consists of mechanical ventilation followed by high frequency ventilation or ECMO. A lung biopsy establishes the histologic diagnosis of this uniformly fatal entity. Open lung biopsy may prevent from using costly, invasive and probably ineffective procedures such as ECMO.

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8 PSU 1993-2005
ISSN 1089-7739