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Meconium Obstruction of Prematurity

Meconium obstruction of prematurity (MOP) is an specific type of meconium obstruction described in premature neonates with very low birth weight (< 1500 g) or extremely low birthweight (< 1000 g). This obstruction occurs in infants with particular risk factors, affects the ileum and colon, and is not associated with cystic fibrosis. Ileal obstruction by meconium in premature, low birth weight babies is a distinct clinical entity. MOP is caused by inspissated meconium in the colon and/or terminal ileum resulting in mechanical bowel obstruction. The etiology is thought to be a combination of the highly viscid meconium of prematurity and the poor motility of the premature bowel. Delayed maturity of interstitial cells of Cajal has been suggested as a cause of distal ileal meconium obstruction combined with increased viscosity of exocrine secretions. This condition is not related to mucoviscidosis or Hirschsprung's disease and is commonly seen in extremely low-birth weight infants. MOP is similar to meconium plug syndrome or small left colon syndrome. Guidelines for the diagnosis of MOP in low-birth weight babies include the following characteristics: 1) severe prematurity and low birth weight; 2) presence of at least one risk factor such as high-risk pregnancy, maternal diabetes, cesarean delivery, or maternal magnesium sulfate administration (magnesium depress smooth muscle cells of the bowel); 3) low-grade obstruction; 4) benign systemic and abdominal examination; and 5) distended loops of bowel without air-fluid levels. A contrast colon study is not essential to establish the diagnosis unless there is therapeutic intent. Likewise testing for cystic fibrosis is not indicated. The presentation of this disorder occurs around 10-14 days of life and involves abdominal distension, bile-stained vomiting, and intestinal perforation. MOP is very severe in extremely low births infants where their bowel can perforate easily. Plain X-rays film of the abdomen shows multiple dilated loops of bowel without pneumatosis and without air-fluid level. The diagnosis may only be confirmed by intervention resulting in the passage of meconium plugs or by contrast radiology. The goal of treatment is to evacuate the tenacious meconium by stimulating peristalsis and reducing its viscosity. Once the obstruction is release there will be no recurrence. In extremely premature infants with delayed passage of meconium glycerin suppositories or saline irrigation are regularly utilized in the NICU to help evacuate meconium. Initial management of MOP includes using a diluted Gastrograffin enema which is diagnostic, therapeutic and the gold standard. The higher osmolarity of Gastrograffin and need of radiological suite transport to small babies has been questioned. If the contrast successfully refluxes through the ileocecal valve the distal ileum can be seen full of impacted meconium. The success rate depends heavily on the extent of colon and ileum filling which is less in MOP as compared with mucoviscidosis from cystic fibrosis. When adding Tween-80 to the Gastrograffin enema it was more

efficient, but this media might be toxic. Gastrograffin can be absorbed into the bloodstream causing tissue dehydration. Gastrograffin enema is inappropriate for hemodynamically unstable patients complicated with bowel obstruction. Iopamidol, a hydrophilic contrast medium used mainly for angiography is less invasive of capillary epithelial cells with an osmotic pressure of 300-600 mOsm which is significantly lower than Gastrograffin. If perforation or absorption into the bloodstream occurs, Iopamidol is less invasive. The procedure which is performed in the incubator using ultrasound guided hydrostatic enema has been utilized with good results. The failure rate of Iopamidol is associated with delaying management. Surgical intervention is considered for patients who develop rapid abdominal distension that is at risk of perforation. The procedure should consist of enterostomy, irrigation and manual evacuation of the impacted meconium. In cases of bowel perforation minimal resection and primary anastomosis is favored.

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Melanotic Neuroectodermal Tumor of Infancy

Melanotic Neuroectodermal Tumor of Infancy (MNTI) is a very rare tumor found in infants mostly involving the bones of the jaw. There are around 500 cases reported worldwide, with most cases from USA followed by India, Germany, and Brazil. MNTI is a pigmented neoplasm which arises from neural crest cells. NMTI typically occurs in infants younger than one year of age with a slight male predilection. Many cases are associated with an increase of urinary vanillylmandelic acid secretion (40%). This tumor has many names, such as congenital melanocarcinoma, pigmented ameloblastoma, retinal anlage tumor, pigmented epulis, melanotic epithelial odontoma or melanotic progenome. Mean age of presentation is 4 to 5 months. A few congenital and prenatal cases have been reported. MNTI is a benign tumor which is locally aggressive with rapid onset and very fast growth rate. Infants presents with a rapidly growing, painless, firm non ulcerated mass with a blue or black discoloration (bluish pigmentation), affecting the craniofacial region in 90% of cases. The maxilla as the most common site of involvement followed by skull and mandible. The mean size for MNTI is 3.5 cm, but lesions can attain a size of 20 cm. Local invasion by the tumor can lead to bony destruction, tooth displacement, and feeding difficulties. Only 3% of these tumors are frankly malignant with just a few producing metastases. Metastatic spread has been documented to lymph nodes and the central nervous system. Extent of

MNTI is performed with dental radiographies, CT-Scan and MRI, though imaging is seldom diagnostic and tissue biopsy is needed. On imaging the tumor presents as a well-demarcated radiolucent lytic lesion within bone that may have features concerning for local destruction. CT-Scan shows a hyperdense mass with bone remodeling and expansion. Pathology shows a characteristic biphasic cell distribution of large epithelioid melanogenic cells and small primitive neuroblastic cells with scattered melanin pigment. The diagnosis is further confirmed using immunohistological stains since both cell types are positive for vimentin and neuron-specific enolase. Immunohistochemical exam can reveal signs of possible aggressive growth behavior. The differential diagnosis of MNTI includes other small round blue cell tumors of childhood, especially neuroblastoma, Ewing sarcoma, alveolar rhabdomyosarcoma, malignant melanoma, and lymphoma. Management of MNTI consist of surgical excision with a 2-5 mm healthy margin of tissue during removal. Complete surgical excision is curative. Recurrences is due to multicentric growth and incomplete surgical excision. Recurrence can be fatal especially when involving the central nervous system or other vital structures. Predominance of a neuroblast-like component and an inconspicuous large cell component were also associated with an aggressive course and high risk of local recurrence. Infants receiving a diagnosis within the first 2 months of life were more likely to have recurrence within 6 months and a shorter disease-free survival. Infants with a diagnosis at age 4.5 months or older had minimal risk of recurrence. Neoadjuvant therapy (chemotherapy and/or radiotherapy) is usually reserved for inoperable tumors, involvement of the central nervous system and other vital structures, or when clear surgical margins are not obtainable. Due to the high recurrence rate of 15-27%, is imperative five year of follow-up.

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Gastric Pneumatosis

Pneumatosis intestinalis is the result of gas infiltrating into the wall of the bowel. It can be detected as a radiological finding, or intraoperatively as the result of an underlying pathological process. Gastric pneumatosis (GP), also known as interstitial emphysema of the stomach, is a rare and primarily radiological diagnosis that can occur in children and adults. It has been seen in preterm and term babies, along with infants up to one year of age. The condition can be noninfectious (gastric pneumatosis) or infectious (emphysematous gastritis). Gastric pneumatosis is caused by a disruption of the gastric mucosa, which results in air dissecting into the stomach wall. This disruption results in

air dissecting into the stomach wall. Four mechanisms can be identified, often acting concurrently, including local or systemic hypoperfusion with gastric ischemia; spontaneous or iatrogenic disruption of the gastric mucosa; intramural infection by gas-producing organisms; and dissection of mediastinal air toward the stomach. In children this disruption typically results from gastric outlet obstruction caused by pyloric stenosis, duodenal atresia or stenosis, duodenal ulcers, malrotation, or tumors. It can also occur from protracted vomiting, instrumentation or tracking of air through the mediastinum or the pneumothorax. Pyloric stenosis is the most common cause of gastric pneumatosis in young infants. Pneumatosis of the stomach likely results from the increased intragastric pressure associated with pyloric hypertrophy and gastric outlet obstruction. The mechanical damage theory is the most common theory regarding the pathophysiology of gastric pneumatosis in infants. It involves proximal gastrointestinal obstruction typically at the pylorus or duodenum, leading to gastric dilatation and elevated gastric pressure. Chronically elevated gastric pressure accompanied by forceful vomiting causes transient pressure peaks leading to gastric mucosal tears which allows air to dissect into the submucosal space. Emphysematous gastritis refers to mucosal disruption caused by gas-forming bacteria invasion. Causes in infants include necrotizing enterocolitis, caustic ingestion, recent abdominal surgery, or gastroenteritis. Pneumatosis intestinalis and specifically gastric pneumatosis are uncommon but potentially dangerous conditions in the burn-injured patient. Risk factors for burn patient include low-flow state, distension and recent trauma or instrumentation. In adults, infection with gas forming organism (Escherichia coli, Proteus, Clostridium welchii and Staphylococcal aureus), gastric outlet obstruction, and instrumentation are the most common cause of gastric pneumatosis. Radiographically the gastric pneumatosis appears linear, cystic, or as small, clustered bubbles in simple KUB films. CT Scan is more sensitive and can further identify the area of pneumatosis in context of portal venous gas. Management of gastric pneumatosis should consist immediate decompression of the stomach with a nasogastric tube. This is followed by a period of stabilization with broad-spectrum antibiotics to reduce bacterial translocation and managing the cause of the pneumatosis. In cases of pyloric stenosis, a pyloromyotomy is performed after preoperative stabilization and correction of electrolytes imbalances. In cases of duodenal obstruction causing gastric pneumatosis management consist of correction of electrolytes disturbances and fluid imbalances, gastric decompression, and subsequent surgical correction of the duodenal obstruction. Clinicians must use the clinical picture in combinations with radiographic evidence to distinguished between gastric emphysema and emphysematous gastritis because there are significant prognostic differences among these pathologies. The mortality for gastric pneumatosis is 41% in adults and 6% in children.

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