



# **PEDIATRIC SURGERY UPDATE ©**

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### **MI-like Syndrome**

Meconium-associated intestinal obstruction during the neonatal period can be the result of an intrauterine perforation (meconium peritonitis), a distal colonic plug (meconium plug syndrome) or intraluminal tenacious meconium associated to decreased pancreatic enzyme (trypsin) activity from cystic fibrosis (meconium ileus). Although most cases of meconium ileus (MI) are associated to mucoviscidosis, 10-20% are sweat-test and genetic marker negative. This is what we call meconium ileus-like syndrome. Most babies with MI-like syndrome are markedly premature (22 to 30 weeks) with very low birth weight. Postnatal factors associated are patent ductus arteriosus, hyaline membrane disease, and intraventricular hemorrhage. The syndrome is the result of a combination of intestinal hypoperfusion and dysmotility due to hemodynamic abnormalities and immaturity of the myenteric plexus of the ileum and colon respectively. Clinically, they have absent stooling during the first two weeks of life, diffuse abdominal distension, retrograde peristalsis, meconium with a high content of albumin, obstruction in the distal ileum, and microcolon. Medical management consists of a gastrograffin enema (is diagnostic and therapeutic), oral gastrograffin, and 10% acetylcysteine (mucomyst) as enteral cleansing agent. Surgery is indicated for progressive clinical deterioration or development of pneumoperitoneum. Follow-up shows no gastrointestinal or pulmonary dysfunction.

### **Meckel's**

Meckel's diverticulum (MD), the pathologic structure resulting from persistence of the embryonic vitelline duct (yolk stalk), is the most prevalent congenital anomaly of the GI tract. MD can be the cause of: gastrointestinal bleeding (most common complication), obstruction, inflammation and umbilical discharge in children and 50% occur within the first two years of life. Diagnosis depends on clinical presentation. Rectal bleeding from MD is painless, minimal, recurrent, and can be identified using 99mTc- pertechnetate scan; contrasts studies are unreliable. Persistent bleeding requires arteriography or laparotomy if the scan is negative. Obstruction secondary to intussusception, herniation or volvulus presents with findings of fulminant, acute small bowel

obstruction, is diagnosed by clinical findings and contrast enema studies. The MD is seldom diagnosed preop. Diverticulitis or perforation is clinically indistinguishable from appendicitis. Mucosal polyps or fecal umbilical discharge can be caused by MD. Overall, complications of Meckel's are managed by simple diverticulectomy or resection with anastomosis. Laparoscopy can confirm the diagnosis and allow resection of symptomatic cases. Removal of asymptomatic Meckel's identified incidentally should be considered if upon palpation there is questionable heterotopic (gastric or pancreatic) mucosa (thick and firm consistency) present.

### **Nesidioblastosis**

Persistence of early fetal cells leading to neonatal hyperinsulinism characterized by severe hypoglycemia can be caused by Nesidioblastosis, one of the islet cell dysmaturity syndrome. Diagnosis rests on four criteria: the presence of increased insulin levels during hypoglycemia, low urinary excretion of ketone bodies, the need to infuse more than 15/mg/kg/min glucose to maintain normal serum levels, and a positive response to glucagon. Seizures, cyanosis, and central respiratory disturbances are the presenting symptoms of hypoglycemia in most cases, and prompt diagnosis avoids the sequelae of mental retardation. Subtotal (95%) pancreatectomy should be done early in these patients if hypoglycemia cannot be controlled with medical therapy (diazoxide) or side effects of therapy are documented. Pancreatic function is not seriously impaired in most patients after subtotal pancreatectomy. Avoiding the need of pancreatectomy in selected patients by long-term use of octreotide (somatostatin analog) may be possible.

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