Pediatric Meconium Ileus — Signs and Symptoms

Meconium originates from the Greek word “mekonion” which stands for poppy. The nomenclature attests to the tarry, black appearance of meconium which resembles raw opium preparation. Meconium ileus implies the presence of an intestinal obstruction in-utero or in neonatal period secondary to abnormally thick meconium. This article expatiates on pathophysiological aspects of meconium ileus and expounds on clinical features of the same. The article concludes with treatment, complications, and prognosis of meconium ileus.
Definition of Meconium Ileus

Meconium ileus (MI) involves an obstruction of terminal ileum by abnormally thick and sticky meconium. This disease entity is often seen in neonates with cystic fibrosis. Meconium ileus is responsible for about one-third of neonatal small-intestinal obstructions. Meconium consists of contents of the small bowel of neonates. Its chief components are succusentericus which in turn is made up of bile acids, bile salts and shredding of intestinal mucosa during intrauterine life (intestinal mucosal debris).

Evacuation of meconium within 6 hours of birth is considered normal.
Associated syndromes

Meconium, in addition to meconium ileus, is branded to cause three other associated syndromes which can be described as follows:

<table>
<thead>
<tr>
<th>Associated pathology</th>
<th>Explanation</th>
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<tbody>
<tr>
<td>Meconium ileus-equivalent syndrome</td>
<td>Often presents until early adult life; marked by recurrent intestinal obstruction and chronic constipation. Operative mortality and morbidity are unfortunately on the higher side.</td>
</tr>
<tr>
<td>Meconium peritonitis</td>
<td>Recurrent bowel obstructions secondary to healed bowel perforations and subsequent formation of adhesions. Ascites may be encountered.</td>
</tr>
<tr>
<td>Meconium plug syndrome</td>
<td>Manifesting within the first 2 days after birth, also known as functional colonic obstruction; encompasses symptoms such as vomiting, abdominal distension and inability to pass meconium normally.</td>
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**Meconium thorax**: passage of meconium in the chest through persistent congenital communications leads to meconium thorax.

**Cystic or pseudocystic meconium peritonitis**: loculated fluid collection in case of meconium peritonitis leads to a quasi-pseudocyst formation.

**Epidemiology and Classification of Meconium Ileus**

Meconium ileus accounts for about 33% of total neonatal bowel obstruction. This disease condition affects both male and female population equally and is more common in the white population. It is a rare medical condition with an incidence of 1 in 25,000 babies.

While neonates with *cystic fibrosis* invariably suffer from meconium ileus, almost 21% of patients with meconium ileus do not have cystic fibrosis.

Meconium ileus can be classified as follows:

<table>
<thead>
<tr>
<th>Type</th>
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<tbody>
<tr>
<td>Simple meconium ileus</td>
<td>Not associated with any other gastrointestinal structural anomaly</td>
</tr>
<tr>
<td>Complex meconium ileus</td>
<td>Associated with gastrointestinal diseases like atresia, perforation, and necrosis, more common in non-CF patients</td>
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**Etiopathogenesis of Meconium Ileus**

Obstruction due to inspissated meconium occurs at the level of the terminal ileum. The distal colon is narrow and contains minute pellets of stool. It is termed as “microcolon.”

Meconium ileus is often associated with cystic fibrosis, a systemic disease involving the intestine, lungs, kidney and pancreas. It occurs in about 20% of patients with cystic fibrosis as the first clinical sign.

**Cystic fibrosis** is a chronic crippling disease severely affecting the quality of life. In about 21% of patients without cystic fibrosis, meconium ileus is incriminated based on incomplete naïve nature of the interstitial cells of Cajal and myenteric plexus of Auerbach.

Cystic fibrosis is autosomal recessive in nature secondary to ΔF 508 mutation in the gene for Cystic Fibrosis Transmembrane conductance Regulator (CFTR) protein. Dysfunctional CFTR leads to the conversion of normal thin bodily secretions to inspissated tenacious nature. Thus the thick secretions choke the ducts and tubes, resulting in damage to the organs. Sweat, digestive fluids, pulmonary secretions, mucus, and semen, are all affected. Major organs affected are lungs, pancreas, intestines, and reproductive organs’
Pancreatic hydrolytic enzymes are normally responsible for the metabolism of fat and protein in the digestive tract. But as cystic fibrosis damages pancreas so the metabolism gets affected. The resultant lack of adequate digestion of fat and protein with subsequent formation of tenacious meconium material obstructing the ileum leads to meconium ileus. The meconium in cystic fibrosis has abnormally increased protein and abnormal mucous glycoprotein.

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Clinical Signs and Symptoms of Meconium Ileus

- Emesis (Vomiting), often bilious of green-colored fluid
- Failure to pass meconium within 12-24 hours, which is normally passed within the first 6 hrs. afterbirth.
- Baby is uncomfortable, restless, and averse to feed due to abdominal discomfort and distension.
- Abdominal distension, abdominal pain; sometimes loops of proximal bowel can be palpated
- Intestinal obstruction
- Ascites
- Respiratory difficulty
- Meconium peritonitis

Diagnosis and Differential Diagnosis of Meconium Ileus

Meconium ileus is a clinico-radiological diagnosis.

Clinical symptoms in a neonate with a family history of cystic fibrosis are sufficient to arouse suspicion for meconium ileus. Ancillary diagnostic tests available are as follows:

Radiological:

Abdominal X-ray: dilated bowel loops appearance without air-fluid levels is characteristic. Normal meconium is invisible on X-ray. “Soap-bubble” or “ground-glass” appearance of small air specks in meconium in the presence of associated clinical symptoms nails the diagnosis. Soap-bubble appearance by itself is not peculiar for meconium ileus and is also seen in patients with colonic atresia, Hirschsprung disease, and meconium plug syndrome. Calcified meconium flecks enclosing the peritoneal surfaces and the scrotum are encountered in meconium peritonitis.

- Prenatal ultrasonography: non-specific changes of intestinal obstruction can often be detected.
- Contrast enema: other associated meconium syndromes can be ruled out.
- Barium enema: terminal ileum obstruction with distal “microcolon” can be precisely looked for.

Sweat testing: for diagnosis of cystic fibrosis when suspected.
Differential diagnosis for neonatal intestinal obstruction

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<tr>
<td>Anorectal malformations</td>
<td>Absent anus or fistula may be present on external examination. Requires detailed workup SOS surgical intervention.</td>
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<tr>
<td><strong>Hirschsprung disease</strong></td>
<td>The most common cause of large bowel obstruction in neonates; it is the culmination of abnormal innervations of the bowel extending proximally from the internal anal sphincter.</td>
</tr>
<tr>
<td>Meconium plug syndrome</td>
<td>It causes colonic obstruction.</td>
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<tr>
<td>Electrolyte abnormality</td>
<td>Hypokalemia, hypocalcemia, hypothyroidism can simulate an intestinal obstruction.</td>
</tr>
<tr>
<td>Sepsis</td>
<td>Necrotizing enterocolitis (NEC) is a grave differential of meconium ileus leading to intestinal obstruction. It is typically seen in preterm infants. Late soap bubble pattern after 12-18 hours of life secondary to peristaltic migration of air in the colon is suggestive of meconium plug syndrome. However, soap bubble appearance in the colon as early as 12 hours should raise suspicion for NEC.</td>
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<tr>
<td>Maternal causes</td>
<td>Magnesium administered as part of eclampsia treatment protocol can simulate an intestinal obstruction.</td>
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Treatment for Meconium Ileus

The treatment strategy of meconium ileus begins with **conservative management** as the first line of action, followed by **surgical intervention** if the former fails.

Conservative management

This is often advocated as the first line of action in simple, uncomplicated, stable neonates. IV (intravenous) fluid line is set. Nothing is given orally. Nasogastric (NG) tube is put in the stomach through the nose to evacuate bile and air.

**Gastrografin** has fallen out of favor in view of rare but well-documented cases of fulminant colitis and dehydration being reported. Dilution of gastrografin to about 3:1 is required. Safer alternatives are now available. Hypaque, Omnipaque is typically used.

Surgical intervention

An **exploratory laparotomy** is typically performed in the case of complicated patients with associated atresia, perforation or necrosis. A **double-barrel ileostomy** is typically created and repeated **N-acetyl cysteine lavages** to clear off the meconium and is performed through the ileostomy. **Primary anastomosis** is attempted in the absence of peritonitis as per the surgeon’s decision.

Complications of Meconium Ileus

- Ileal atresia or stenosis of ileum- narrowing and obstruction of ileum
- Ileal perforation- rupture of ileum due to impaction and infection, resulting in meconium peritonitis
- Meconium peritonitis as a result of perforation- it can be cystic or noncystic
- Ischemia of bowels
- Bowel infarction- the destruction of bowels due to a reduced blood supply
- Volvulus- twisting of loops of intestines around itself and supporting mesenteric tissue culminating in a serious medical emergency
- Giant cystic meconium peritonitis
Prognosis for Meconium Ileus Patients

Meconium ileus is a **curable** entity. Once treated adequately, there is no relapse. Survival at 1 year is about 90% in patients with simple meconium ileus. Further prognosis depends on the probable presence of underlying systemic disease such as **cystic fibrosis**.

The treatment then concentrates on **enzyme replacement** for augmentation of the nutritional status and **control of recurrent pulmonary infections**. Future prospects include gene transfer therapies and chemical modulation of the epithelial cell abnormality.

**Summary**

Meconium is a normal intestinal content of the neonate consisting of succus entericus, bile substances and denuded intestinal mucosa. It is passed at about 6-12 hours after birth by a normal neonate. Inspissated, tenacious meconium causing intestinal obstruction at the level of terminal ileum results in meconium ileus. Meconium ileus is often associated with **cystic fibrosis** and is often the first clinical presentation of the same.

A neonate with meconium ileus presents with **small bowel obstruction** with abdominal distension, pain and inability to pass meconium. An X-ray picture of “ground-glass” appearance is characteristic. The distal colon appears small and is termed as “**microcolon**.”

The treatment of meconium ileus begins with **gastrografin enema supplemented with N-acetylcysteine**. In the case of enema failure, complicated MI or unstable vitals, **surgical intervention** steps in.

**Review Questions**

The correct answers can be found below the references.

1. **Which of the following statements is true?**
   - A. Normal meconium is yellowish in color.
   - B. A normal neonate passes meconium in-utero.
   - C. A normal neonate passes meconium in the first 6-12 hours after birth.
   - D. Meconium is secondary to ingestion of toxic substances by the mother.

2. **Meconium ileus is associated with which systemic illness?**
   - A. Noonan syndrome
   - B. Hirschsprung’s disease
   - C. Wilm’s tumor
   - D. Cystic fibrosis

3. **Which of the following statements is false?**
   - A. Meconium ileus always requires surgical management
   - B. Meconium ileus with perforation is treated surgically
   - C. Conservative management involves use of a gastrografin enema
   - D. Surgical management involves the use of a double-barrel ileostomy
References


Nelson’s textbook of Pediatrics.

Correct answers: 1C, 2D, 3A

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