

# CHAPTER 67

## MECONIUM ILEUS

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### Introduction

Neonatal bowel obstruction by a thick and tenacious meconium is known as meconium ileus. Meconium abnormalities cause multiple neonatal intestinal obstructive disorders of varying severity, ranging from the benign meconium plug syndrome to the complicated meconium ileus associated with cystic fibrosis (CF). The largest group of patients presenting with meconium ileus are children who suffer from CF, accounting for 75% of all caucasian patients with meconium ileus.

In the last 20 years, a specific type of meconium ileus, not associated with CF, has been described in premature neonates with very low birth weights. Meconium ileus, however, does occur in up to 20% of neonates with CF and is the earliest manifestation of the disease.

### Demographics

Cystic fibrosis is the most common serious inherited defect affecting the caucasian population. It is transmitted as an autosomal recessive condition with a 5% carrier rate and an incidence of approximately 1:2,500 live births. Hamish et al. reported the incidence of CF in live-born babies in America to be lower in blacks (1:15,000).<sup>1</sup> The cystic fibrosis transmembrane conductance regulator (CFTR) is located on the long arm of chromosome 7. The delta F508 mutation is the most common mutation among caucasians. There are, however, great differences between populations. For example, delta F508 mutation is present in 70% of CF alleles in caucasians in the United States, but accounts for only 43% in African Americans.<sup>2</sup> In the literature, a great racial variation is assumed.<sup>3</sup> Meconium ileus is reported to be a rare finding in African populations.<sup>4-7</sup>

Premature and very low birth weight (VLBW) babies can suffer from a condition resembling meconium ileus called meconium ileus equivalent (Figure 67.1). Babies at risk for meconium ileus equivalent usually can survive only in intensive care units with ventilation therapy.<sup>8</sup> Such units are rare in Africa, and this may be the reason why meconium ileus equivalent is not described in large studies of neonatal paediatric surgical care in Africa.

### Aetiology

The intraluminal obstruction in meconium ileus is due to abnormally thick and tenacious meconium (Figure 67.2). It becomes inspissated in the distal ileum, blocking the lumen. Abnormally dilated mucous glands in the distal ileum secrete mucus with a very high protein content containing an abnormal mucoprotein, which is responsible for the tenaciousness of the meconium. In VLBW babies, the problem seems to be the disproportion between the tenaciousness of normal meconium and the underdevelopment of the contractility of the bowel. In both cases, meconium is strongly attached to the wall of the distal ileum, creating pellets of white meconium in a narrow lumen.

### Classification

The clinical presentation varies depending on the type of meconium ileus. Instances of meconium ileus can be classified into uncomplicated and complicated types.



Figure 67.1: Meconium ileus in a low birth weight baby.

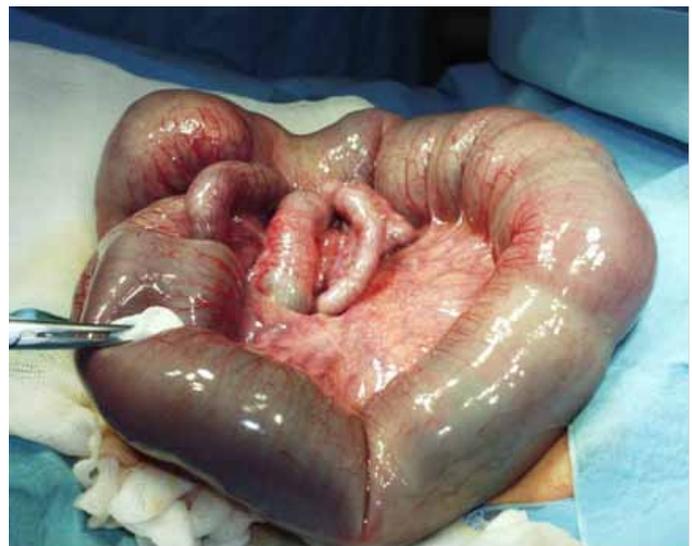


Figure 67.2: Meconium ileus with dilated ileum and contracted, obturated terminal ileum.

### Uncomplicated Form

The neonate may appear relatively normal for the first 12–18 hours of life. However, as the proximal bowel fills with air, abdominal distention, emesis (later bilious), and failure to pass meconium are noted. On examination, distended loops of intestine may be visible. Bowel sounds are present but sluggish. Mucosal plugs may be evacuated on rectal examination after withdrawal of the finger.

### Complicated Form

About half of meconium ileus patients have a complicated form, associated with volvulus, perforation, or atresia. Massive distention, tenderness, or erythema indicates the presence of complications. There are no bowel sounds; vomiting is usually bilious. The child is in a critical condition.

### Investigations

Plain abdominal radiographs show a distended intestine. Air-fluid levels may or may not be present. A “soap bubble” appearance in the right lower quadrant may be the result of air mixed with the meconium.

The initial diagnostic test is a contrast enema. In the case of a meconium ileus, it shows a microcolon. Meconium pellets in the distal ileum can also be determined. This diagnostic investigation also enables one to exclude colonic atresia and rotation anomalies. The main differential diagnosis is Hirschsprung’s disease. This will need to be excluded by suction rectal biopsies.

The same diagnostic methods are used in premature babies, although due to the prematurity, recognition of the problem may be delayed.

## Treatment

### Nonoperative Treatment

Uncomplicated meconium ileus may be successfully treated nonoperatively. All patients require standard supportive care:

- oral gastric tube decompression;
- intravenous fluids to replace deficits and counteract ongoing losses; and
- meticulous attention to the acid-base balance.

In uncomplicated cases, a gastrografin enema (dilution 3:1) is the treatment of choice.<sup>9</sup> It can be accompanied by use of N-acetylcysteine/saline (1:5) in several enemas and in addition to the oral tube. The effect of this enema is to draw large volumes of fluid into the lumen. Therefore, the child must be well-rehydrated prior to this procedure. The child’s pulse rate and urine output have to be carefully monitored. This procedure is successful in more than 50% of affected children. The patient should evacuate spontaneously over the next 6–8 hours. If the patient fails to evacuate or if a complicated meconium ileus is present, a surgical procedure must be carried out.<sup>7</sup>

### Operative Treatment

Operative treatment is indicated when the nonoperative treatment fails or is associated with complications such as perforation or in the complicated type of meconium ileus. In all cases, a supraumbilical transverse incision is used.

Three procedures can be used:

1. enterostomy and decompression;
2. resection and stoma formation; or
3. resection and anastomosis.

### Enterostomy

Enterostomy is performed by opening the bowel on the antimesenteric border proximally where the dilated bowel tapers down. A size 10 catheter is pulled upwards, and the sticky meconium is washed out by using gastrografin or a 1:5 solution of acetylcysteine/saline. Patience is required during this procedure; it takes a considerable amount of time because one has to work carefully. Further, the distal plugs have to be washed out. When the small bowel is empty, the enterostomy can be closed in the usual way.<sup>10</sup>

### Resection

Resection and formation of a stoma is the most common form of management. Several forms of stoma are possible, with the most wide-

spread being the Bishop-Koop type anastomosis (Figure 67.3).<sup>11</sup> The most distended part of the ileum is resected. An end-to-side anastomosis is constructed about 3 cm distal to the resection margin. The open end is brought out as a stoma and sutured to the skin. In critical cases, a double-barrel stoma may be the best option.

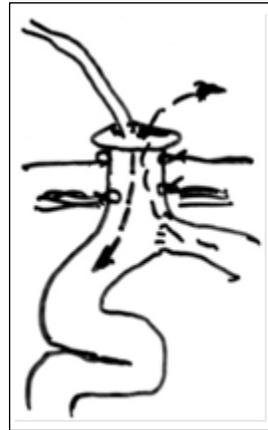


Figure 67.3: Bishop-Koop anastomosis for irrigation.

## Postoperative Complications

General supportive care is provided as after any major laparotomy. The oral gastric tube is left in place until bowel function returns. Use of N-acetylcysteine via the oral tube, or a stoma, or by enema may further aid passage. Oral feeding is started with pancreatic enzyme supplementation.

CF is confirmed or ruled out by determining the sweat chloride level. Close attention has to be given to pulmonary care in CF children.

Weaning is often not a problem. Extended physiotherapy and specific attention to pulmonary infections and general growth of the child are decisive for the quality of the child’s further life.

In some cases, the Bishop-Koop anastomosis closes by itself. In other cases, special attention has to be paid to clearance of the obstructed segment. After this, closure of the enterostomy can be carried out. In CF, distal ileum obstruction tends to recur, so special care has to be taken to ensure normal bowel movements.

## Prognosis

The prognosis for children suffering from CF has improved in the developed countries due to neonatal care, general nutrition, treatment of pulmonary infections, and specific antibiotics.

Neonates with very low birth weight require a specific type of treatment. Their survival depends on intensive neonatal care with ventilation, broad spectrum antibiotics, and a specific enteral nutrition.

## Ethical Issues

In developed countries, CF is a genetic-based diagnosis. Specific care has to be given to children with CF. This has enabled the life-span of CF sufferers to be extended to 30 to 40 years, whereas historically in Europe these children died at about age 20. One of the main procedures that has made this improvement possible is the gastrografin enema developed by Helen Noblett.<sup>9</sup> In combination with this, use of N-acetylcysteine often makes it possible to avoid having to perform a laparotomy. If the combination fails, surgery and enterostomy have to be carried out.<sup>2</sup>

## Evidence-Based Research

The condition is uncommon, and relevant studies of surgical treatment with a significant number of patients are not available.

## Key Summary Points

1. Meconium ileus is very rare in Africa.
2. Among Caucasians, 75% of meconium ileus is associated with cystic fibrosis.
3. A rare variant of meconium ileus not associated with cystic fibrosis has been described in premature very low birth weight babies.
4. The patients present as uncomplicated or complicated meconium ileus.
5. The treatment of uncomplicated cases is nonoperative by gastrografin enema.
6. In complicated and failed nonoperative treatment cases, operative intervention is required.
7. The prognosis of children with meconium ileus has improved with advances in supportive care in the developed countries.

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