CHAPTER 23

Meconium Ileus

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INTRODUCTION

Meconium ileus is the earliest clinical manifestation of cystic fibrosis (CF) and occurs in 8–10% of patients with CF at birth. The clinical features are mainly due to the presence of abnormal, inspissated and viscid mucus of intestinal origin. In neonates affected by this condition, the impacted meconium produces an intraluminal obstruction occurring in the midileum, leading to a progressive distension. About 40% of patients with meconium ileus are complicated by intestinal volvolus, atresia, gangrene and necrosis, perforation, peritonitis with abdominal calcifications and, finally, meconial pseudocyst.

More recently relief of the intestinal obstruction has been obtained by irrigating the intestine with various solutions such as normal saline, 1% *N*-acetylcysteine, hyperosmolar Gastrografin enema, surfactant, or DNase. With respect to different types of surgical and medical efforts, the survival rate at 1 year increased from 10 to 90%, and the operative mortality is drastically decreased to 15–23% of the treated newborn.

Meconium ileus accounts for 9–33% of all neonatal intestinal obstructions (300 new cases in Italy each year), with an incidence of 1:2500 newborns, representing the third most common cause of neonatal small bowel obstruction after ileal and duodeno-jejunal atresia and malrotation. Polyhydramnios is the most frequent feature observed in prenatal diagnosis of complicated forms of meconium ileus. The presence of fetal hyperechogenic bowel on the ultrasound, associated with dilated bowel and/or ascites could be indicative of an intestinal obstruction. A family history of CF is clearly evident in almost 25% of these patients. Meconium ileus is uncommon in premature infants (5–12%), and associated congenital anomalies are rare.

In the meconium ileus, the intestine shows different aspects if we consider the *proximal,* the *middle* and the *distal* ileum. In the first portion, nearly normal evidence is present, with a progressive dilatation at the mid-portion borderline. In the *proximal ileum*, the content has a semi-liquid consistency and is not yet viscous. A marked and severe dilatation of the *middle ileum* is always seen: the intestine contains thick, dark green and putty-like meconium, firmly adherent to the walls. The intestinal obstruction causing a hyper peristalsis is responsible for the congestion and hypertrophy of the walls. The *distal ileum* is full of concretions called "rabbit pellets", grey stained and with a typical beaded appearance. This small bowel condition is responsible for a narrow, empty and small colon, never used, which is called *microcolon.*

Main symptoms include abdominal distension (96%), bilious vomiting (50%) and delayed passage of meconium (36%). From a clinical point of view, it is possible to recognize two different conditions: a simple, uncomplicated and non-surgical type, and a complicated, severe type, with a mortality of at least 25% of all cases. In the first type (58%), signs and symptoms of a distal ileal obstruction are seen not later than 48 h after birth: generalized abdominal distension with dilated and visible as well as palpable loops of bowel, bilious vomiting, no stools and narrowing of the anus and rectum, with only a dense and rubber-like grey meconium sticking to the anal wall. In the second type (42%), the neonate represents a surgical emergency, which must be treated within 24 h of birth, when the signs of a hypovolemic shock or sepsis are not well established. A fetus with complex meconium ileus is at increased risk of postnatal bowel obstruction and perforation. A usual X-ray image of fine, granular soap-bubble ("Singleton's sign") or ground-glass appearance ("Neuhauser's sign") is due to a dense meconium mixed with air, typical of the distal ileum; this picture is usually located in the mid-abdomen or in the right iliac fossa. When the meconium ileus is complicated, the abdominal radiograph may show calcification as a result of meconium peritonitis due to a fetal perforation of the intestine. A double-bubble image or air-fluid levels can be seen when a secondary ileal atresia (single or double) is the final bowel remodelling after a complete volvulus associated with a severe ischaemic damage. If the intestinal perforation occurs early in the antenatal period, the X-ray appearance of a round rim of calcification underlines a meconium pseudocyst.

Figure 23.1, 23.2

The first step of the treatment includes a nasogastric tube decompression, antibiotic prophylaxis, and correction of dehydration, electrolytes and hypothermia.

A contrast enema with water-soluble and hyper- or iso-osmolar contrast is the medical treatment of choice and mucosal safe, for uncomplicated cases. A recent study that used various enema solutions administered in a mouse model showed that surfactant and Gastrografin were the most efficacious for the in vivo relief of constipation in comparison with perflubron, Tween-80, Golytely, DNase, *N*-acetylcysteine and Viokase. Intestinal mucosal damage was absent and viscosity had been significantly reduced in vitro.

The enema evacuation should be obtained under fluoroscopic control, with a gentle and progressive increasing of the intraluminal pressure, thus avoiding unexpected fractures of the colon. A correct procedure prevents leakage of the contrast medium by taping the buttocks as well as the catheter dislocation. If the contrast medium fails to progress into dilated small bowel loops, the presence of an acquired atresia is definite and the radiologist must stop the examination because of a high risk of perforation. Of those submitted to this procedure, 50% of neonates benefit from enema alone over the next 48 h, without any additional treatment; in some cases, a second enema may be used with a complete evacuation of the meconium filling the ileal loops. Acetylcysteine administered by mouth is useful and helps to relieve the obstruction. Radiographs are taken at 3, 6, 12, 24 and 48 h with the aim of evaluating progression and possible complications. At this time feeding is begun. Hypovolemic shock and early perforation are around the corner, but an appropriate and meticulous procedure can avoid these complications.

Figure 23.1

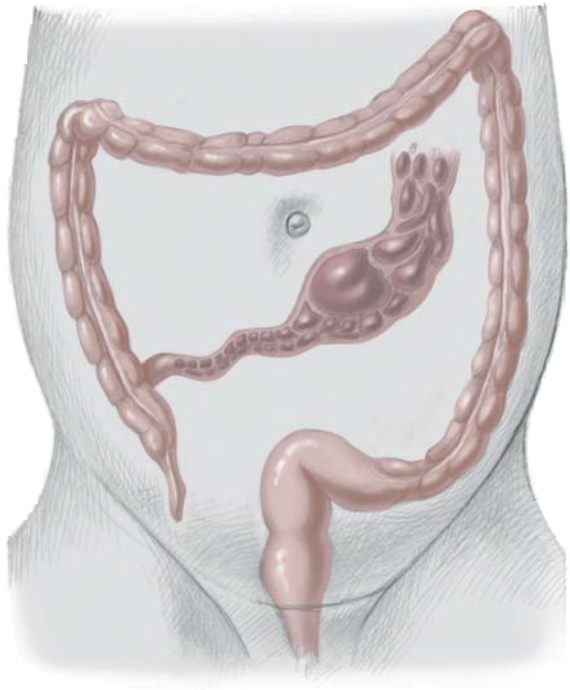


Figure 23.2

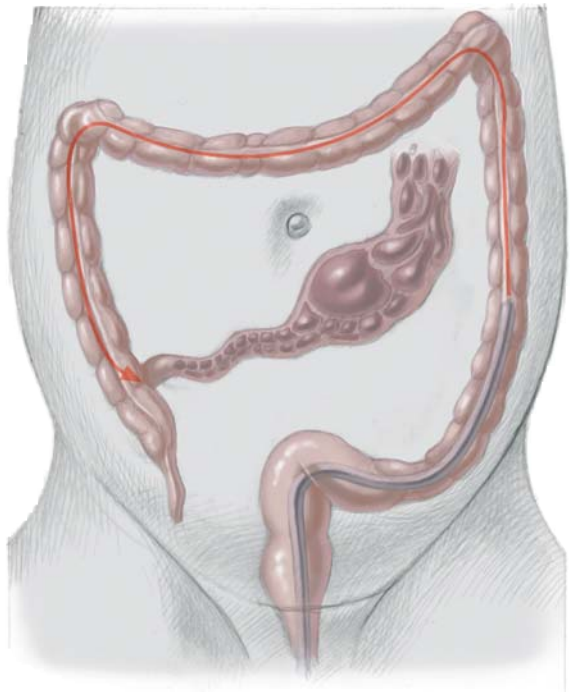


Figure 23.3

Only 6% to 10% of uncomplicated forms fail the nonoperative management using a water-soluble contrast enema. If no significant difference in intestinal diameters and no microcolon is present, a limited enterotomy and repeated warm saline irrigations through a smooth catheter provide for the best result. Meconium discharge may be manually supported, using an enterotomy placed in the dilated hypertrophic ileum. The catheter is two-way directed with care, clearing the small as well as the large bowel. After exclusion of an atretic intestinal segment, the enterostomy is closed by single interrupted seromuscular stitches.

Figure 23.4, 23.5

In contrast, approximately half of neonates with meconium ileus cannot be treated adequately with irrigations and/or present additionally an intestinal obstruction complicated by neonatal intestinal perforation or ileal atresia secondary after intrauterine perforation. They always require a surgical procedure such as resection of the dilated meconium-filled ileum and ileal anastomosis (as shown in Chap. 22). Additionally, complicated cases of meconium ileus are seen in newborns with an extreme difference in diameters of the proximal and distal ileum, and a significant microcolon. In those cases, the double-tube ileostomy technique according to Rehbein has proved to be an effective treatment and avoids the secondary laparotomy for stoma closure.

A horizontal predominantly right-sided laparotomy, approximately 2 cm below the umbilicus, is performed. A small transverse incision into the enlarged ileum is performed, approximately 5–7 cm in front of the narrow part filled with stool pellets. Four stay sutures are inserted into the margins of the incision. If an atresia exists, the atretic part is resected and the thickened meconium from the proximal part and the grey stool pellets from the distal part are evacuated by numerous irrigations with warm saline through a 5–8 Ch feeding tube supporting the manoeuvre by gentle manual forward and backward manipulation. Once all intestinal contents are evacuated, a 10 Ch feeding tube with larger cut openings is inserted through the enterotomy in the proximal non-dilated ileum, and a second 5 Ch tube is inserted into the distal narrow ileum or microcolon in a T-tube fashion.

The enterotomy is closed around the tubes and is tightly fixed to the appropriate part of the ventral abdominal wall – in a similar fashion as in a gastrostomy. Both tubes are carefully fixed with nonabsorbable sutures to the skin. If a bowel segment had to be resected, the stoma for the tubes is best situated approximately 5 cm in front of the anastomosis, and the small tube is passing the anastomosis far into the narrow intestine. Post-operatively the large tube serves for suctioning and evacuation of the intestinal contents; the small distal tube serves for constant irrigation with increasing amounts of fluid (first saline, later pre-digested milk), thereby promoting a rapid enlargement of the ileum and the microcolon. As soon as the evacuations through the large tube become less and normal bowel movements occur, it is indicative that most of the intestinal contents are passing by distally. The double tubes can then be simply removed. We have treated our patients since the 1980s by this double-tube method successfully, and cutaneous enterostomy is no longer performed.

Figure 23.3

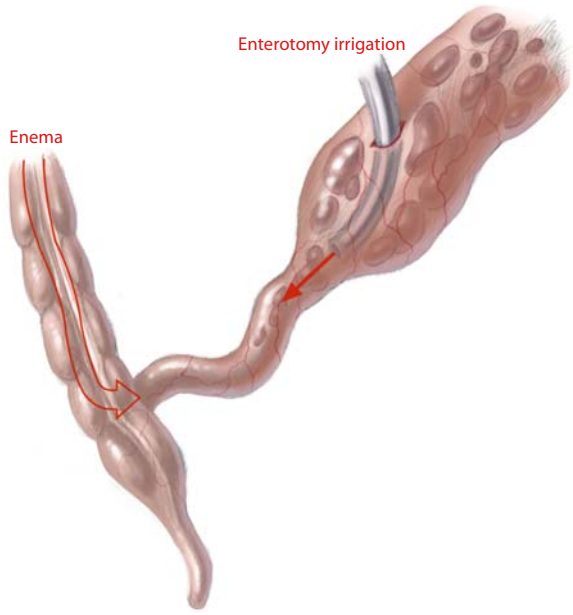


Figure 23.4

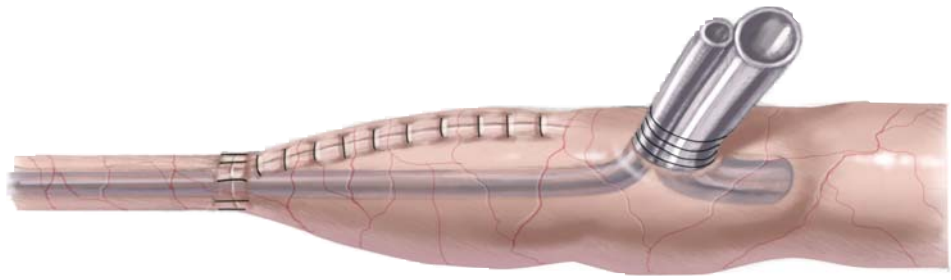


Figure 23.5

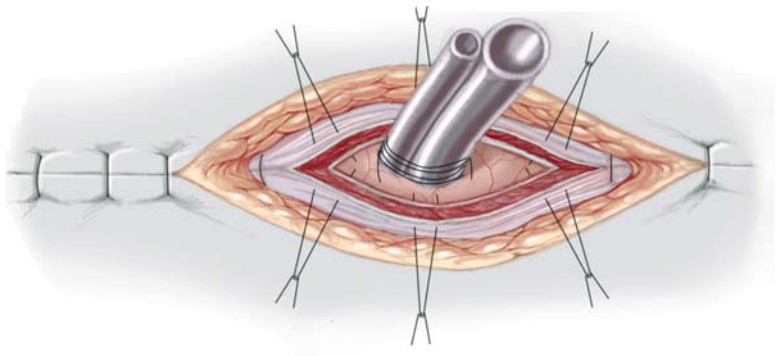


Figure 23.6–23.9

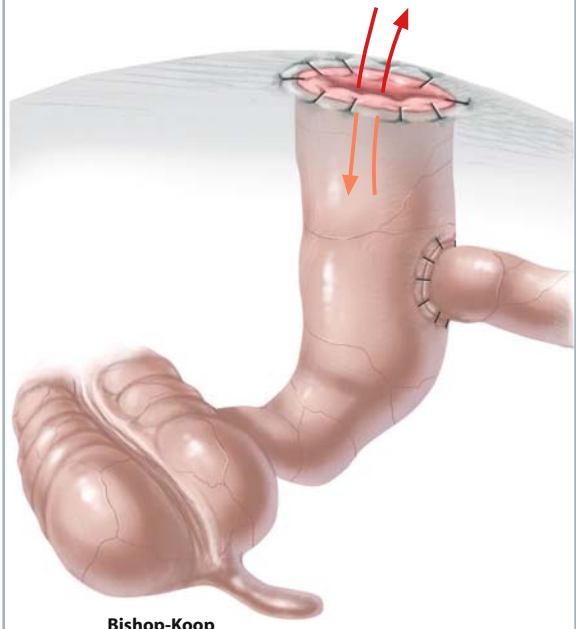
Different surgical techniques have been developed in the past consisting of a resection of the enlarged bowel segment and temporary decompression by means of a distal or proximal enterostomy. The most simple form is a double-barrelled ileostomy according to Mikulicz, with the two loops brought out side-to-side. This solution is quick and avoids an intra-abdominal anastomosis. More technical alternatives have been described thereafter: a distal ileostomy with end-to-side ileal anastomosis (Bishop-Koop) has been called "distal chimney enterostomy". This procedure consists of a Roux-en-Y anastomosis between the end of the proximal segment and the side of the distal segment, at least 3 to 5 cm from the open end. The open limb of the distal segment is used as an ileostomy. A variation of this technique has been described, using an angulating proximal segment, which is obliquely anastomosed with the distal stump. Proximal chimney enterostomy, the so-called Santulli procedure, consists of a proximal ileostomy with end-to-side ileal anastomosis. The end of the distal limb is anastomosed to the side of the proximal limb, the end of which is used as the enterostomy. This technique should facilitate irrigation as well as decompression of the proximal small bowel.

The enterostomies can be closed by an end-to-end anastomosis when uninhibited passage of intestinal contents is established, mostly between 7 and 12 days after.

Figure 23.6

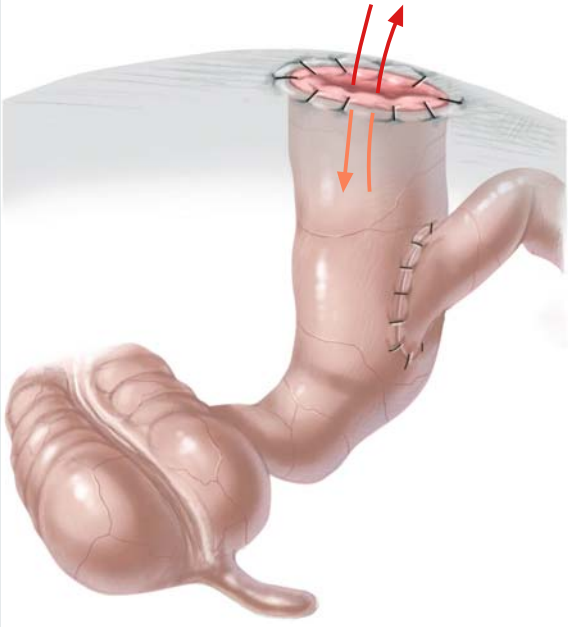
Mikulicz

Figure 23.7



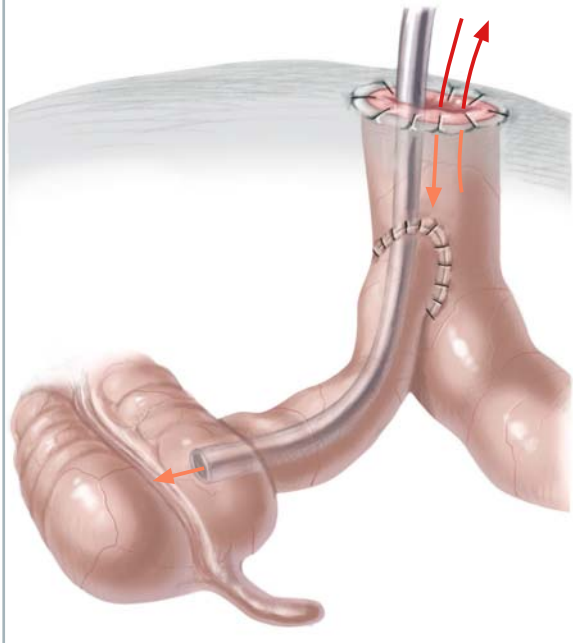
Bishop-Koop

Figure 23.8



modified Bishop-Koop

Figure 23.9



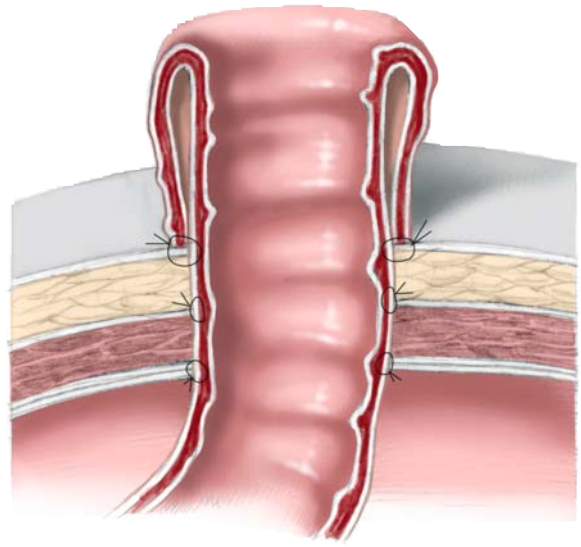
Santulli

Figure 23.10

When a terminal ileostomy or colostomy is desired in neonates and/or children, the preferred method used is the nipple-valve system formation. This is an easy technical procedure and avoids any kind of infiltration or stricture or retraction of the neostoma.

A 2- to 3-cm-long intestinal segment is used with a serosal surface free of fat and with a good vascular supply. Seromuscular wall is sutured to the fascia in four quadrants. Next the stitch is taken through the skin seromuscular wall and edge of the stoma, thereby creating the nipple. Four to six sutures are normally needed depending upon the size of the stoma.

Figure 23.10



CONCLUSION

Generally, the lesser the extend of to bowel resection the earlier is the recovery of peristalsis.

Actually, bowel resection with primary anastomosis has been proved to be as well effective and safe as stoma formation, but is associated with a reduced length of initial hospital stay. Complications include pulmonary infections, which is the most important one with an incidence of at least 8 to 10%. Anastomotic leakage occurs for different reasons: a technical mistake, an insufficient blood supply, or an unrecognized distal obstruction. Delayed recovery of peristalsis is another frequently observed complication and is due to an abnormal stretching of the intestinal walls during the fetal life. Total parenteral nutrition is the support of choice and a central venous catheter is mandatory in these situations.

Meconium ileus may be an early indication of a more severe phenotype of CF. This was suggested by the significantly lower pulmonary function found in children with a history of meconium ileus compared to age- and sex-matched children who did not have meconium ileus. In this view, patients without CF demonstrated better growth and functional status, and had a lower incidence of lung diseases as well as gastrointestinal problems. It is also possible that patients affected by meconium ileus have a more severe form of CF that, from a pathological point of view, consists of a more severe compromise of mucous glands

The complicated forms are susceptible to a higher number of long-term surgical complications, including especially small bowel obstructions (of adhesive origin) and blind loop syndromes (30% of the cases). These long-term problems are more commonly ascribed to a higher incidence of peritonitis and small bowel ischaemia. Nearly all patients undergoing enterotomy and irrigation do not manifest long-term surgical morbidity. There is general agreement that resection and stomas could be avoided with a more meticulous attention to care and clearing the intestinal walls from the more dense concretions.

Long-term complications in neonates affected by uncomplicated meconium ileus who were not operatively treated are rarely seen, and only mild and transient complications have been observed in newborns treated with minor surgical procedures, such as enterotomies and irrigations. However, meconium ileus did not directly affect long-term functional or social status in children with CF, and the quality of life is mainly correlated with the extension of lung lesions, which is the most limiting factor with respect to the school, social life and physical activity.

Survival of neonates with meconium ileus has improved over the last two decades because of neonatal intensive care, improved surgical technique and medical treatment. In general, an overall immediate survival of 90% is achieved using the modern protocols and nearly all deaths are pertinent to the adolescents. Only few children die because liver and or septic complications. Deaths are mainly due to sepsis, primary or secondary to pulmonary interstitial emphysema, or due to aspiration pneumonia. In a large series reported and analysed by Fuchs, only one child died because of the meconium ileus itself.

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