Meconium Ileus

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

Meconium ileus is the congenital (baby is born with it) blockage of intestine contents due to abnormally thick meconium at the point where small bowel connects to large bowel. Blockage happens because the thick meconium does not allow intestinal fluid to go through. Alternatively, blockage can cause the intestine to rupture, twist or not be in continuity (one normal length of intestine).

Meconium ileus is seen mostly in two populations:

- Cystic fibrosis (CF) patients: 10-33% infants diagnosed with meconium ileus have a family history of CF. 10-20% of CF patients will have associated meconium ileus, and this may be the only sign of CF at birth. The abnormal gene which causes CF is responsible for causing the meconium to be thicker compared to babies without CF, which increases risk of meconium ileus.
- Preterm infants: Medications to slow down labor have been associated with meconium ileus.

Signs and Symptoms - "What symptoms will my child have?"

In utero, or when fetus is in mother's womb, an ultrasound can see "too much" fluid within the uterus known as polyhydramnios, which can be a sign of fetus intestinal blockage or obstruction.

Early signs: Inability for baby to tolerate feedings, vomiting, swollen/enlargened belly, and no passage of meconium in the first 24-48 hours after birth.

Late signs: Discoloration of the skin of the belly, inconsolable crying due to belly pain, fever, green vomiting. Baby may have low blood pressure and a high heart rate if they are dehydrated or if there is a hole in the intestine due to thinning of the wall because the small intestine was so dilated.

Conditions that mimic this condition: intestinal atresia, malrotation, Hirschsprung disease, incarcerated (stuck) groin hernia.

Diagnosis - "What tests are done to find out what my child has?"

Physical Exam: Babies with meconium ileus may have large bellies. They will not pass stool in the first 24-48 hours. Babies may have pain when touching their bellies, but that is typically a later finding if there is thinning or a hole in the intestine. Babies may lose more weight after birth than the expected weight loss.

Blood tests: Blood will be sent to check for the CF gene. Basic lab tests may be performed to look at the electrolytes in the blood if there is concern for dehydration.

Sweat test: Sweat is collected and tested for concentration of chloride to determine whether someone has CF. This is not typically a test done for babies, as they do not generate the sweat needed for the test.

Abdominal x-ray: Abdominal x-rays are performed to evaluate causes of blockage symptoms and causes of stool not passing in the normal time frame. On x-rays for meconium ileus, the small intestine is typically dilated and filled with air (from baby swallowing air). There may be a "soap bubble" appearance of the x-ray if air intermixes with the thick meconium.

Contrast enema: Water-soluble contrast is instilled through a rubber tube placed gently in the anal opening, and several x-rays are taken to evaluate the large intestine and where the blockage may be. Sometimes, the contrast mixes with the thick stool and allows the stool to be evacuated. The contrast enema can also see a "micro" colon, or when the large intestine is smaller than normal large intestine, due to no passage of intestinal contents through the colon while baby was in the uterus.

Treatment - "What will be done to make my child better?"

Non-operative management

- If babies are showing signs of obstruction/blockage, a tube may be placed into their nose or mouth down into their stomach to remove stomach contents.
- An IV will be placed and IV fluids will be given to rehydrate babies. Sometimes, a special IV needs to be placed to give the baby nutrition (Total Parental Nutrition - TPN).
- If the contrast enema shows signs of meconium ileus without complications from the blockage (such as intestinal twist, rupture, or blind ends in the intestine) medical attempts are made to thin the meconium. Water soluble contrast is placed through a catheter into the anus and injected into the colon. This contrast pulls water into the intestine to dissolve the thick meconium and allow it to pass.

- The infant will have x-rays performed to observe progress. If the meconium does not pass completely the first time, but the infant remains stable, the enema can be repeated safely, and after exam by a physician. This strategy is successful 22-35% of the time.
    - Risk: Risk of this treatment is perforation or a hole in the colon. This would require the baby to be taken to the operating room for repair.
- Another option is placing an special medication (N-acetylcysteine) through the tube in the nose to help thin the meconium.

Surgery

- Surgery is needed if an infant fails to pass meconium with the enemas and the bowel remains blocked. Surgery is also required if there are complications resulting from meconium ileus such as intestinal rupture, twist, or discontinuity.
- The goal of surgery is to relieve the intestine of blockage. If an infection exists, the goal is also to control the infection (for instance, repair a hole in the intestine). What is done in the operating room depends on many factors including how sick baby is, size of the baby, and damage to the intestine. Options may include simple flushing of the intestine, cutting out the affected intestine and putting it back together, or creation of a stoma/ostomy. The surgeon will make the decision of what is the best and safest alternative for the baby while in the operating room.

Preoperative preparation: IV fluids and antibiotics.

Postoperative care: Infants are managed in the neonatal intensive care unit. The infant may need the support of a breathing machine (ventilator). It will take days or sometimes weeks for the intestine to gain full function so nutrition will have to be provided by vein (TPN).

Risks: Risks of operation include anesthesia, bleeding, postoperative infection, damage to internal structures which may require further operation, and recurrence of meconium ileus.

- Complications can include failure to thrive or the pancreas and lungs not working like they should due to CF.

Benefits: The bowel obstruction from meconium ileus and accompanying complications make the infant very ill. Often surgery is needed to relieve the blockage and help control any existing infection.

Home Care - "What do I need to do once my child goes home?"

Diet: The infant's formula will be determined by many factors such as whether or not CF is present and the length of intestine remaining if any needed to be removed in surgery.

Activity: Activity should be normal.

Wound care: By the time the baby is released, s/he should be able to have regular baths. If a stoma remains, you will be trained in stoma care prior to discharge.

When to call a doctor: Not able to keep down feeds, vomiting (in particular, green or bright yellow vomiting), swelling of the belly, no bowel movements, belly pain, redness or drainage from wound.

If the infant is diagnosed with CF, they will be referred to a specialized group of doctors (lung and gut specialists) to address all aspects of CF care including dietary supplements, exposure restrictions, lung care.

Long Term Outcomes - "Are there future conditions to worry about?"

The outcome for meconium ileus treatment has improved to almost 100% survival. If the child required an operation but does not have CF, there is a lifetime risk of intestinal blockage from scarring of about 10-15%. Children with CF are at a much higher likelihood of having pulmonary and gastrointestinal complications as they get older.

Updated: 11/2021
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