Meconium Ileus

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Algorithmic Approach

A. Meconium ileus is characterized by newborn bowel obstruction secondary to inspissated meconium impacted within the distal small bowel. Over 75% of children with meconium ileus will have cystic fibrosis [1]. Meconium ileus is the initial presentation of cystic fibrosis in 15–27% of affected individuals [1]. There may be a positive family history leading to increased suspicion of the diagnosis. Prenatal diagnosis may be suggested in the setting of sonographic evidence of polyhydramnios, dilated bowel, a calcified mass, or echogenic intra-abdominal cyst. These findings are variable and nonspecific [1, 2]. Postnatally, the history of early-onset abdominal distension, bilious emesis, and delayed passage of meconium, with palpable loops of bowel and/or an abdominal mass, is highly suggestive of meconium ileus [2, 3]. Malrotation with volvulus should be ruled out in any neonate presenting with bilious emesis.

B. Infants with uncomplicated meconium ileus present with progressive abdominal distension in the first 24–48 h of life. Abdominal exam reveals a distended but soft abdomen with palpable, doughy bowel loops. Abdominal x-rays in uncomplicated meconium ileus demonstrate loops of dilated bowel typically without air-fluid levels with a classic "ground glass" or "soap bubble" appearance [3]. Infants with complicated meconium ileus due to atresia, volvulus, meconium peritonitis, or meconium pseudocyst often present with abdominal distension at birth [1]. This may be associated with signs of fetal distress and peritonitis. Abdominal x-rays in complicated meconium ileus classically demonstrate calcifications suggestive of in utero perforation with a possibility of meconium peritonitis with or without a well-formed pseudocyst [3].

C. Contrast enema in meconium ileus will demonstrate a microcolon secondary to disuse with outlining of meconium pellets in the distal small bowel. Use of a water-soluble hyperosmolar contrast agent such as Gastrografin® can be diagnostic and therapeutic [1–3].

D. Initial treatment of uncomplicated meconium ileus is non-surgical with solubilizing enemas. Hyperosmolar enemas are effective but can cause significant fluid shifting and dehydration; thus concurrent IV fluid administration is required [1–3]. Gastrografin® is the most commonly used agent [3]. The addition of N-acetylcysteine can be helpful as it has been shown to aid in the dissolution of inspis-

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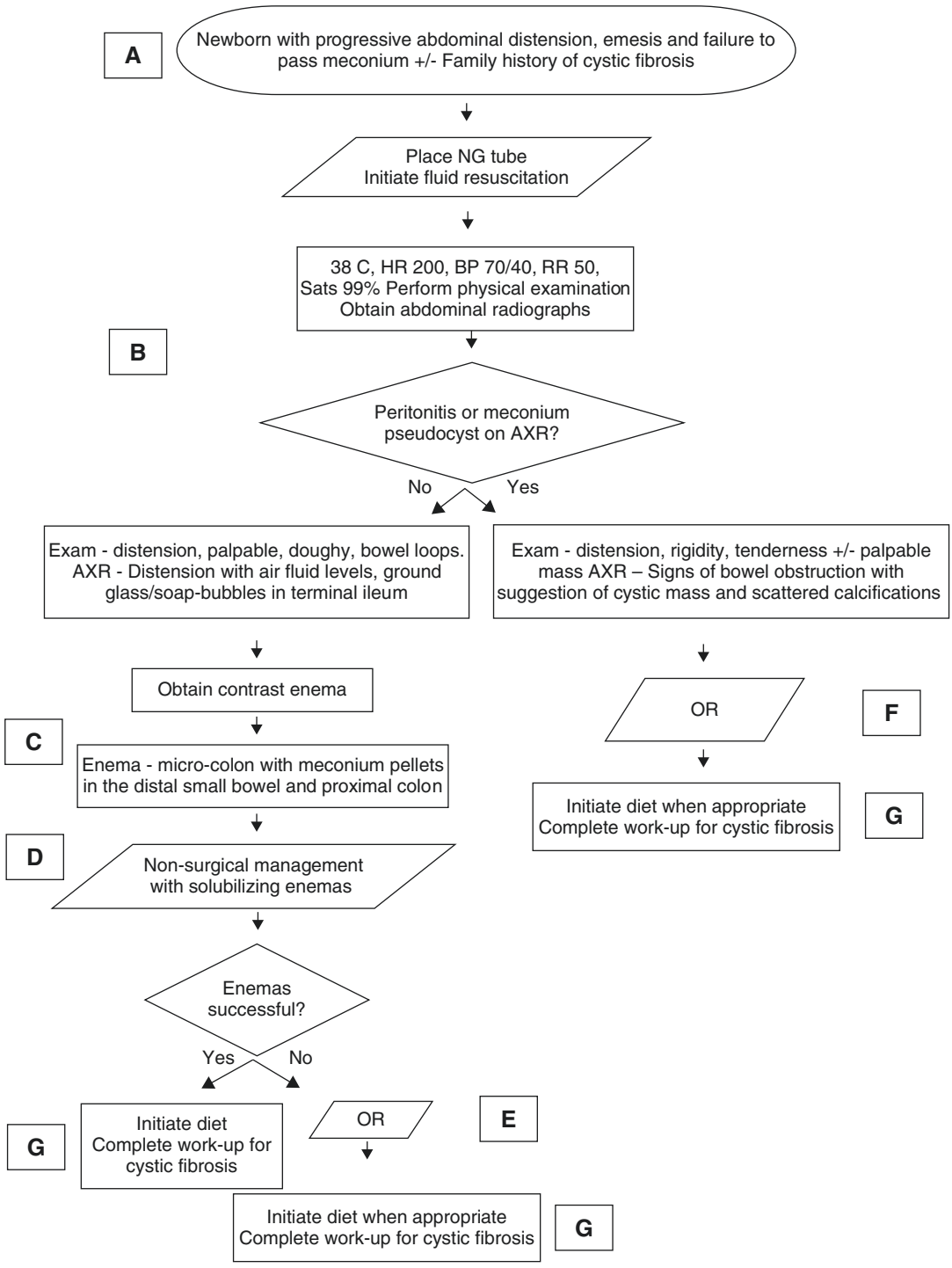
sated meconium in animal studies [2]. Enemas can be repeated until the meconium obstruction clears as long as the infant remains clinically well without worsening obstruction or signs of intestinal compromise [1, 2].

E. Operation is required for infants with uncomplicated meconium ileus that fails to improve with solubilizing enemas to evacuate the inspissated meconium and relieve their obstruction before complications develop. A wide variety of surgical options exist including enterotomy with irrigation or stoma creation. Classically described stomas for meconium ileus include the Mikulicz double-barreled stoma, Bishop-Koop (distal "chimney") stoma, or Santulli (proximal "chimney") stoma [1–3]. Resection is rarely required in uncomplicated disease.

F. Complicated meconium ileus is managed operatively. If volvulus is present, the bowel should be detorted and assessed for viability. If viable, the distal bowel is cleaned of meconium as described earlier. If the bowel is nonviable, then resection should be completed after which the distal bowel is cleaned of meconium and a decision is made regarding stoma formation versus anastomosis. If an atresia is present, the proximal dilated segment is resected, the distal bowel cleaned of meconium, and a decision made regarding stoma formation versus anastomosis [1–3]. In cases of perforation with meconium peritonitis or a meconium pseudocyst, abdominal washout with debridement of the cyst followed by resection and stoma formation is preferred [3].

G. A diet may started once the infant is stabilized and stooling. Work-up for cystic fibrosis should be initiated. Diagnostic modalities include genetic analysis and sweat chloride testing [1, 2]. Genetic testing assesses for an array of causative mutations, with the most common being ΔF508 [1]. Sweat chloride testing can be done after 48 h of life; however, it may be difficult to obtain the volume for sweat needed in children less than 2 weeks of age [1, 2]. Long-term complications of meconium ileus associated with cystic fibrosis include failure to thrive, pancreatic insufficiency, constipation, rectal prolapse, and distal intestinal obstruction syndrome (DIOS) also known as meconium ileus equivalent syndrome [1, 3].



Algorithm 128.1

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