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ABSTRACT Introduction: Meconium Ileus is the obstruction of the terminal ileum by abnormally tenacious meconium. Postnatal clinical signs such as abdominal distension, failure to pass meconium and eventual bilious vomiting, usually evolve within 24-48 hours. Can also be diagnosed antenatally on maternal ultrasound scan of the abdomen. Objective: To report a case of meconium ileus presenting as acute intestinal obstruction. Clinical Case: A late preterm (36 weeks), appropriate for gestational age (AGA) (2500 grams), female child, delivered via emergency caesarean section, who presented at 8 hours of life with respiratory distress and further developed abdominal distension with failure to pass meconium since birth. Conclusion: A hyperosmolar contrast enema is both diagnostic & therapeutic.

KEYWORDS : Meconium Ileus, Cystic Fibrosis; Hirschsprung's Disease; Contrast Enema.

INTRODUCTION:

Meconium ileus (MI) is impaction of inspissated meconium in the distal small bowel. It accounts for up to 30% of cases of neonatal intestinal obstruction.¹ In neonates, MI is one of the earliest manifestations in patients with cystic fibrosis (CF). It presents in up to 20% of patients with CF. If left untreated, the prognosis is poor.²

We report a case of meconium ileus with intestinal obstruction in a late preterm AGA baby and review the clinical presentation, radiological features and treatment of this condition.

Clinical Case:

A late preterm (36 weeks), AGA (2500 grams), female child, delivered via emergency caesarean section, indication being preterm with non-reassuring fetal heart rate and breech presentation. Baby was born to a 29-year-old primigravida mother. She had regular antenatal check-up, had taken iron, calcium and folic acid supplements during pregnancy.

Baby was admitted in NICU of Jaipur Golden Hospital at 8th hour of life with Compliant of respiratory distress. Baby was started on tube feeds at admission, oxygen through nasal prongs via blender and empiric antibiotic coverage. Baby was gradually weaned to room air at 11th hour of life. Antibiotics were stopped after blood culture reported no growth.

At 48 hours of life, baby did not pass meconium, mild abdominal distension was noted with dilated abdominal veins, sluggish bowel sounds. Anal opening was patent. Increase in abdominal girth (AG) compared to baseline. Abdominal x-ray done was suggestive of obstruction (dilated bowel loops with few air fluid levels).



Figure 1: X-ray suggestive of dilated bowel loops with few air fluid levels.

Ultrasound scan of the abdomen was done, which showed multiple dilated air-fluid filled bowel loops suggestive of acute intestinal obstruction. Pediatric surgeon reference was done. On examination, abdomen was distended, skin over abdomen was stretched & shiny, dilated bowel loops & peristaltic movements were visible. AG increased to 36 cm. Tympanic note heard on percussion. Sluggish bowel sounds heard on auscultation. Conservative management was advised by pediatric surgeon. At 72 hours of life, the baby passed thick meconium pellets like stool.





Figure 2: Abdomen is distended, skin over abdomen is stretched & shiny, dilated bowel loops.

Figure 3: Thick, inspissated mucoid material.

On day 4 of life, baby didn't pass meconium, abdominal distension increased to 37 cm. A gastrograffin dye study was done under supervision of pediatric surgeon, dye reached till ceacum. After the procedure the baby passed meconium. Gradually the abdominal distension decreased and paladai feeds were restarted on day 6 of life. Gradually the feeds were built up and iv fluid stopped. Baby tolerated feeds well & bowel output was normal. The diagnosis of meconium ileus leading to intestinal obstruction was made retrospectively. Further work up for CF was advised but parents declined.

The baby was again admitted after 2 weeks with complaints of poor feeding. During the course of hospital stay the baby's bowel output was normal.



Fig4: Gastrograffin Dye Study

DISCUSSION:

About 20% of cases with CF have Meconium Ileus, whereas about 80-

90% of cases with Meconium Ileus have associated CF. It is seen in isolation too, particularly in low birth weight (LBW) infants (mostly due to gut immaturity) It is divided into 2 categories: simple & complex Meconium Ileus. In the latter the obstruction is complicated by associated gastrointestinal pathology, such as atresia, necrosis, and perforation.²

CF Transmembrane Conductance Regulator (CFTR) within the small intestine is responsible for both Cl and HCO³ excreation. Abnormal CFTR creates an acidic and dehydrated environment resulting in thick, dehydrated mucus with elevated levels of stool albumin, increased mineral content, and protein-bound carbohydrates. All these factors result in viscid meconium that eventually leads to physical obstruction of the terminal ileum.3 CF gene is located on long arm of chromosome 7. More than 1900 CFTR polymorphisms have been reported. Most common mutation being F508del.4

Prenatally, serial antenatal ultrasound scans may detect echogenic, dilated bowel in uncomplicated cases; while various findings (ascites, cysts, calcification, etc.) are found with complications.³ Erect x-ray abdomen shows dilated small bowel loops proximal to the impaction. Classically, there is paucity or absence of air-fluid levels & a "bubbly" appearance of the distended intestinal loops.⁵

In simple cases, a hyperosmolar contrast enema is both diagnostic & therapeutic: the microcolon, & narrow terminal ileum with filling defects (meconium pellets) & a post procedure evacuation of the pallets with gradual relief of obstruction are typical. In complicated cases- ascites, calcification, large soft-tissue densities, extraluminal air, and pneumoperitoneum are seen in combination. Surgical options include enterotomy & lavage, various stomas to facilitate diversion (end/loop/chimney-Bishop-Koop, Santulli) & gut irrigation. N-acetyl cysteine (NAC), a mucolytic, has been administered at various stages of the management for lysing & easing the expulsion of the inspissated meconium.

Today, early and late survival rates for both simple and complex Meconium Ileus are consistently reported over 80%.

Although Meconium Ileus can be associated with other etiologies, it is most offen associated with CF, making it necessary to screen neonates with Meconium Ileus for CF.⁷ The newborn screen is based on abnormally elevated levels of immunoreactive trypsinogen (IRT) detected in a dried blood spot on the Guthrie card.8 However, confirmation of the diagnosis of CF must be made using the gold standard sweat test or genetics.5

Neonates with a presumed diagnosis of functional immaturity of the colon whose symptoms do not resolve after therapeutic enema should be considered for colonic biopsy to look for Hirschsprung disease.¹

CONCLUSION:

Intestinal obstruction accounts for about 33 % of all NICU admissions. Of which around 30 % cases are of Meconium Ileus. Postnatal clinical signs such as abdominal distension, failure to pass meconium and eventual bilious vomiting, usually evolve within 24-48 hours. The prognosis for infants presenting with both simple and complicated meconium ileus has improved with the advancement of both nonoperative and operative treatments, along with good nutritional support and better treatment of bacterial infections. In simple cases, a hyperosmolar contrast enema is both diagnostic & therapeutic. In complicated Meconium Ileus or cases where hyperosmolar enema is unsuccessful, prompt surgical management is required.

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