



CLINICAL PRESENTATION OF NEONATAL MECONIUM ILEUS IN NORTH INDIA

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ABSTRACT

Introduction: Meconium Ileus (MI) is one of the causes of intestinal obstruction. Meconium obstruction poses a great challenge not only in recognition and diagnosis but also in the management. Cystic fibrosis has been associated with MI, with the incidence ranging from 15–40%.

Aim: To evaluate the clinical presentation, diagnosis, surgical management, postoperative care, and outcome in neonates with meconium ileus.

Materials and Methods: This study was done retrospectively in the department of Pediatric surgery and Pediatric, PGIMS Rohtak to include all patients who presented with meconium ileus in pediatric surgery department over a period of 15 years i.e. from 2000 to 2015. Neonatal birth characteristics, surgical findings, postnatal management and outcomes were reviewed from the data collected from department records.

Results: From 2000 to 2015, 30 neonates were managed for meconium ileus. Simple, uncomplicated meconium ileus occurred in 12 infants (40%) and complicated meconium ileus occurred in 18 (60%). Thirty neonates presented with complications of MI including ileal atresia type III in 17, adhesions in 10, and giant pseudomeconium cyst in 10, volvulus in 3 and duplication cyst in 1. Ileal atresia was the commonest complication (41.46%).

Conclusion: Early recognition in antenatal period and early surgical management along with good NICU care will help in reducing the postoperative morbidity and mortality.

KEYWORDS : Meconium ileus, neonates, intestinal obstruction

INTRODUCTION

Meconium ileus was first described by Landsteiner in 1905. Till 1942, there was not even a single case report in which a patient with severe obstruction from this cause could be saved.¹

Meconium ileus is the condition characterized by extremely viscid, protein-rich inspissated meconium causing terminal ileal obstruction which accounts for approximately 20% of all the neonatal intestinal obstructions.² Although meconium ileus rarely occur in otherwise normal children, majority of these patients have association with cystic fibrosis (CF). Meconium ileus is the presenting symptom in about 10% to 15% of cystic fibrosis patients.³

Meconium ileus can be classified into two types: uncomplicated cases in which a simple intraluminal ileal obstruction occurs and complicated cases which are related to the mechanical complications caused by meconium bolus. The heavy meconium-filled segment of bowel just proximal to the narrow obstructed segment filled with intraluminal concentrations may twist, producing a volvulus. The volvulus may result in ischemic necrosis, causing intestinal atresia or perforation with extravasation of meconium into the peritoneal cavity and development of a pseudocyst or giant cystic meconium peritonitis.⁴

Meconium leakage into the abdominal cavity causes an inflammatory response by stimulating peritoneal macrophages followed by intestinal obstruction in severe cases. The estimated incidence of meconium peritonitis is 1:30,000. Meconium peritonitis is a cause of substantial perinatal morbidity and mortality which is as high as 80% in the third world healthcare setting. Thus early recognition of the underlying etiology, pathophysiology as well as the specific perinatal management is the

prerequisite for optimizing the postnatal outcome.⁵

MATERIAL AND METHODS

STUDY DESIGN

This retrospective study was carried out in the Paediatric and Pediatric surgery department of Pt. Bhagwat Dayal Sharma PGIMS, Rohtak over a period of 15 years from 2000 to 2015. Data was collected from patients' hospital records which was analysed for age at presentation, sex, gestational age, clinical presentation, diagnosis, surgical procedure performed and their outcome. The patients satisfying the following criteria were included : (1) clinical presentation with decreased frequency of defecation associated with abdominal distention and bilious or non-bilious vomiting and (2) X-ray findings of abdomen suggestive of distended small bowel loops without air-fluid levels or pneumatosis and a normal-sized colon or microcolon with multiple filling defects on contrast enemas.(3)

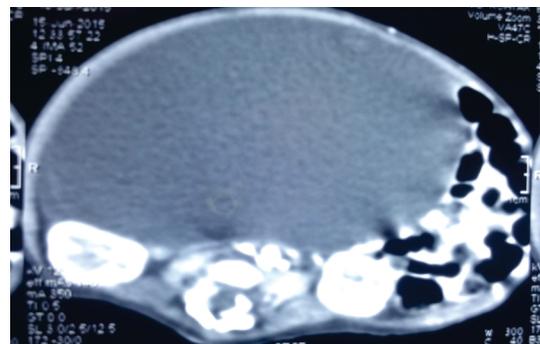


Fig.1- CT Scan abdomen showing giant pseudomeconium cyst

Newborns with meconium obstruction in other parts of the intestine, neonates with meconium plug syndrome, and newborns with other mechanical or functional causes of bowel obstruction were excluded.

RESULTS

A total of 30 newborns were included in the study. Male-female ratio was 0.8:1(13:17). Birth weight ranged from 1.75 to 2.5 kg (mean=2.12kg). Mean age of presentation was 4.4 days. The patient's characteristics are shown in Table I.

Table I Patient's characteristics

Patients	N=30
Gender (M:F)	0.8:1
Birth weight(kg)	2.2(1.75-2.50)
Mean age of presentation(days)	4.4(1-20)

Simple, uncomplicated meconium ileus was found in 12 infants (40%) while complicated meconium ileus in 18 infants (60%). Thirty neonates presented with complications of MI including ileal atresia type III in 17, adhesions in 10, giant pseudomeconium cyst in 10, volvulus in 3 and duplication cyst in 1. Intestinal atresia which was present in 17 patients was the commonest complication (41.46%) as shown below in fig 2.

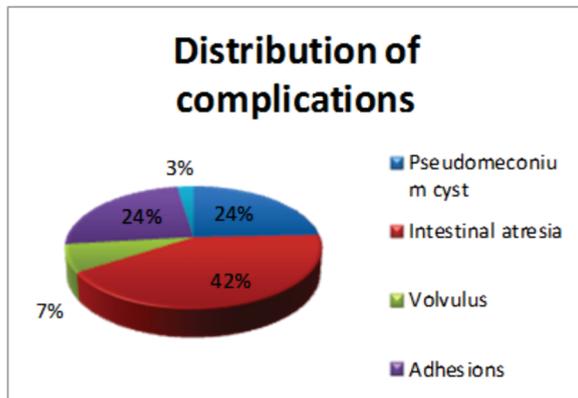


Fig.2.- Distribution of complications of meconium ileus

Abdominal roentgenograms showed peritoneal calcifications, variable-sized bowel loops and mass effect. Barium enema showed an unused microcolon. Exploratory laparotomy was done through supraumbilical transverse muscle cutting incision. Gut was examined for the complications. Simple uncomplicated meconium ileus was managed by Bishop-Koop ileostomy. While in cases of complicated meconium ileus, Cyst was excised, adhesions were lysed and end to back/end to end ileo-ileal anastomosis was done in 18 neonates. Patients were transferred to NICU for postoperative management. Oral feeds were started once the patients passed stools and were discharged at 10-15 days of life. Patients were followed up to detect CF. Postoperative survival rate was found to be around 60-70%.

DISCUSSION

Various types of meconium obstruction have been described since the first report by Clatworthy Jr et al. in 1956.⁶ Meconium obstruction commonly occurs in premature newborns but there are no established diagnostic criteria. The incidence of meconium obstruction has been increasing owing to better survival and live births of extremely premature neonates with better medical care over the past 20 years.^{7,8} The term Meconium related ileus (MRI) includes both meconium plug syndrome (MPS) and meconium ileus disease.^{6,9} MPS is a functional obstruction in the small intestine or proximal right colon caused by impaired meconium excretion. MPS seems to be associated with immature intestinal function and

dysmotility in premature infants, with a multifactorial etiology. Milla explained that a more mature migrating motor complex of the intestine was evident only after 34-35 weeks of gestation, while Yoo et al proposed that a delayed maturity of interstitial cells of Cajal could lead to meconium obstruction.^{10,11}

Meconium ileus needs to be differentiated from Long segment Hirschsprung disease. Meconium has high albumin content so thick inspissated meconium pellets are formed in MI in the distal segment which differentiates from long segment Hirschsprung on gross examination and confirmatory differentiation is done on biopsy. Approximately 10% to 15% of patients with CF present with meconium ileus (MI). Approximately 50% of these neonates present with a simple uncomplicated obturation intestinal obstruction while the remaining patients present with complications of MI, including volvulus, gangrene, atresia, and perforation, which may result in giant cystic meconium peritonitis.⁴

In our study, simple uncomplicated meconium ileus was found in 12 infants (40%) while complicated meconium ileus in 18 infants (60%). These thirty neonates presented with complications of MI, including volvulus in 3, ileal atresia type III in 17, adhesions in 10, and giant pseudomeconium cyst in 10 and duplication cyst in 1. Thus, Intestinal atresia was the most common presentation (41.46%). Atresia are due to failure of recanalisation or due to intrauterine vascular catastrophe due to volvulus, duplication and intrauterine intussusception resulting in atresia of different types.¹² Nam et al reported 31 MP patients who had intestinal atresia in 14 patients (45%), volvulus in 2 patients (6%), and uncertain ileal perforation in 10 patients (32%). Kamata et al reported 20 MP patients who had intestinal atresia in 18 patients (90%) and meconium ileus in 2 patients (10%).¹³

Meconium ileus was considered to be a fatal condition till 1948 when Hiatt and Wilson introduced enterotomy and saline irrigation of the obstructing pellets.¹⁴ Mikulicz enterostomy was introduced by Gross in 1953 which successfully relieved the obstruction.¹⁵ Later on in 1957, Bishop and Koop described resection and end-to-side anastomosis with a distal ostomy to allow postoperative irrigation.¹⁶ Then in 1961, Santulli and Blanc reported resection with side-to-end anastomosis and proximal ostomy.¹⁷

Similarly we too in our study performed Bishop-Koop ileostomy in 12 neonates who presented with simple uncomplicated meconium ileus while Resection with end to back ileo-ileal anastomosis in 18 neonates who presented with atresia, volvulus, giant cystic meconium peritonitis and perforation, with postoperative survival rate of 60-70%.

CONCLUSION

The survival of MI patients has drastically improved because of early recognition in antenatal period, early surgical management and better postoperative NICU care. Irrespective of the type of MI, survival has improved, independent of the surgical procedure.

DECLARATIONS

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REFERENCES

1. Verma A, Rattan KN, Yadav R. Neonatal Intestinal Obstruction: A 15 Year Experience in a Tertiary Care Hospital. J Clin Diagn Res 2016;10(2):10-13.
2. Caniano DA, Beaver BL. Meconium ileus: a fifteen-year experience with forty-two neonates. Surgery 1987;102:699-703.
3. Saleh N, Geipel A, Gembruch U, Heep A, Heydweiller A, Bartmann P, Franz AR, et al. Prenatal diagnosis and postnatal management of meconium Peritonitis. J. Perinat. Med

- 2009;37:535–538.
4. Fuchs JR, Langer JC. Long-term outcome after neonatal meconium obstruction. *Pediatrics* 1998;101:1-6.
 5. Rescorla FJ, Grosfeld JL, West KJ, Vane DW. Changing patterns of treatment and survival in neonates with meconium ileus. *Arch Surg* 1989;142:837-40.
 6. Clatworthy HW Jr, Howard WH, Lloyd J. The meconium plug syndrome. *Surgery* 1956;39:131–142.
 7. Dimmitt RA, Moss RL. Meconium diseases in infants with very low birth weight. *Semin Pediatr Surg* 2000;9:79–83.
 8. Coppola CP. Meconium plug syndrome and meconium ileus. In: Coppola CP, Kennedy AP Jr, Scorpio RJ, editors. *Pediatric surgery: diagnosis and treatment*. Cham (CH): Springer International Publishing; 2014:183-5.
 9. Rickham PP, Boeckman CR. Neonatal meconium obstruction in the absence of mucoviscidosis. *Am J Surg* 1965;109:173–77.
 10. Milla PJ. Development of intestinal structure and function in neonatal gastroenterology. In: Tanner MS, Stocks RJ, editors. *Neonatal gastroenterology: contemporary issues*. Newcastle upon Tyne(UK):Intercept; 1984. 1-20.
 11. Yoo SY, Jung SH, Eom M, Kim IH, Han A. Delayed maturation of interstitial cells of Cajal in meconium obstruction. *J Pediatr Surg* 2002;37:1758–1761.
 12. Rowe, et al. *Essentials of paediatric surgery*. Mosby year book, Inc (1995).
 13. Uchida K, Koike Y, Matsushita K, Nagano Y, Hashimoto K, Otake K, Inoue M, et al. Meconium peritonitis: Prenatal diagnosis of a rare entity and postnatal management. *Intractable Rare Dis Res* 2015:1-5.
 14. Hiatt RB, Wilson PE. Celiac syndrome: therapy of meconium ileus: report of eight cases with a review of the literature. *Surg Gynecol Obstet* 1948;87:317-27.
 15. Gross RE. *The Surgery of Infancy and Childhood*. Philadelphia, Pa:WB Saunders Co; 1953:175-91.
 16. Bishop HC, Koop CE. Management of meconium ileus: resection, Rouxen- Y anastomosis and ileostomy irrigation with pancreatic enzymes. *Ann Surg* 1957;145:410-14.
 17. Santulli TV, Blanc WA. Congenital atresia of the intestine: pathogenesis and treatment. *Ann Surg* 1961;154:939-48.