

MECONIUM DISEASE WITH SURGICAL IMPLICATIONS

Eugen S. Boia¹, Radu E. Iacob¹, Marioara Boia²

REZUMAT

Obiective: Patologia meconială este specifică perioadei neonatale și este cauza a jumătate din obstrucțiile intestinale la nou-născut. Scopul acestui studiu este de a prezenta experiența Clinicii de Chirurgie Pediatrică Timișoara în tratamentul cazurilor internate în perioada 1993 - 2002.

Material și metodă: Lucrarea se bazează pe studiul retrospectiv al cazurilor cu patologii meconială tratate în clinica noastră în ultimii 10 ani. Datele necesare elaborării studiului au fost obținute prin analiza foilor de observație, a rezultatelor examenelor radiologice, protocoalelor operatorii și a protocoalelor de necropsie la cazurile cu evoluție fatală.

Rezultate: În perioada menționată au fost tratați în clinică 16 nou-născuți: 5 cazuri de peritonită meconială, 5 cazuri de ileus meconial și 6 nou-născuți cu sindrom de dop meconial. Afecțiunea este mai frecventă în mediul urban, sexul masculin fiind mai des interesat (62%); tratamentul chirurgical a fost aplicat în 10 cazuri (cu peritonită meconială și ileus meconial) iar 6 cazuri au beneficiat de tratament medical (sindrom de dop meconial). În 4 cazuri evoluția a fost fatală.

Concluzii: Patologia meconială este adesea implicată în obstrucțiile intestinale neonatale. Interesează mai frecvent sexul masculin iar diagnosticul este dificil de stabilit. Evoluția postoperatorie nu este întotdeauna favorabilă.

Cuvinte cheie: patologie meconială, nou-născut

ABSTRACT

Objective: The meconium disease is one of the neonatal specific diseases, which is responsible for half the cases of bowel obstruction at children. The aim of the study is to present the experience of the pediatric surgeons from the Department of Pediatric Surgery Timisoara, in the treatment of cases of hospitalized meconium disease since 1993 to 2002.

Material and Methods: This work is based on a retrospective study of cases with meconium disease treated in our clinic in the last 10 years. Data necessary for this study have been obtained from: registers analyses, X-ray analyses, surgery protocols and necropsy papers in fatal cases.

Results: 16 new-born children suffering of meconium disease were hospitalized during the studied period: 5 cases of meconium peritonitis, 5 cases of meconium ileus and 6 cases of meconium plug syndrome. The disease is more frequent in urban areas and 62% of patients were male; surgery was necessary in 10 cases (meconium peritonitis and meconium ileus) and medical treatment in 6 cases; 4 cases had fatal evolution.

Conclusions: The meconium disease is seldom observed in neonatal obstructions. It appears more often in males and the diagnosis is difficult to establish. The postoperative evolution is not always good.

Key Words: meconium disease, new-born

INTRODUCTION

The meconium disease is one of the neonatal specific diseases, which is responsible for half the cases of bowel obstruction in children. It has several types: meconium ileus, meconium peritonitis and meconium plug syndrome.^{1,2} Meconium ileus (MI) is caused by inspissated meconium that blocks the intestine and in most of the cases is associated with cystic fibrosis - CF. CF is a systemic illness and is transmitted as an autosomal

recessive trait. The first description of pathologic changes in CF was made in 1905 by Landsteiner. In 1953, Gross reported a successful relief of obstruction after resection and a Mikulicz's enterostomy.²⁻⁴ In 1969, Noblett introduced the hyperosmolar enema (Gastrografin) in treatment of simple meconium ileus and meconium plug syndrome.⁵

Positive diagnosis is based on: clinical findings - picture of a bowel obstruction; sweat test - positive above 60 mEq/L²; genetic tests (10-20% are sweat-test and genetic marker negative - this is what we call meconium ileus-like syndrome); contrast enemas;⁶ abdominal X-ray often indicated distended loops of intestine (some are enormously dilated and others are only moderately distended) and may mimic ileal atresia or other forms of obstruction. Abdominal films that demonstrate pneumoperitoneum or calcifications indicate perforation with meconium peritonitis.

¹ Pediatric Surgery and Orthopedics Clinic, Timisoara, ² Neonatology and Health Care Clinic, Timisoara

Correspondence to:
Eugen S. Boia, MD, PhD, M. cel Batran, 17, ap.1, Timisoara, Romania
Tel: 0740164030; E-mail: eemboia@rdslink.ro

Received for publication: Sep. 22, 2003. Revised: Apr. 26, 2004.

Meconium ileus must be differentiated from other causes of congenital bowel obstruction like Hirschsprung`s disease, atresia and stenosis, annular pancreas, malrotation, duplication cyst, neonatal small left colon syndrome, neoplasia, hypothyroidism and other rarer causes.^{7,8}

Treatment: medical (nonoperative) treatment, in cases of simple meconium ileus and meconium plug syndrome, consists of repeated hyperosmolar enema and adequate hydration; surgical treatment is indicated in cases of meconium peritonitis or severe meconium ileus.^{5,6,9,10}

MATERIAL AND METHODS

The aim of the study is to present the experience of the pediatric surgeons from the Department of Pediatric Surgery Timisoara, in the treatment of cases of hospitalized meconium disease since 1993 to 2002.

Data necessary for this study have been obtained from: registers analyses, history of the family, sweat test results, X-ray analyses, surgery protocols and necropsy papers in fatal cases.

Study of:

- The incidence of the disease;
- Sex ratio;
- Possibilities of early diagnosis;
- The results of the treatment to new-born suffering of meconium disease.

Table 1. Types of meconium disease

Type of disease	Cases
meconium peritonitis	5 (31,25%)
meconium ileus	5 (31,25%)
meconium plug syndrome	6 (37,5%)

The distribution of cases indicates a predominance of the incidence to males (10:6).

Distribution according to the environment:

- Urban area - 11 cases (68.75%);
- Rural area - 5 cases (31.25%).

Prematurity was found in 3 cases and malformations were associated in 4 cases (bowel atresias in 3 cases of meconium peritonitis and laparoschizis in 1 case of cystic fibrosis).

Diagnosis:

- History of family data: cystic fibrosis was found in 1 case - the first born deceased at 3 years old;
- The sweat test was positive in two cases of meconium ileus;
- The simple abdominal x-ray:

1. Disparity in the size of the loops (unlike the uniform dilatation of the loops found in the simple atresias - see Figure 1)

2. A soap bubble appearance in the lumen representing a mixture of air and meconium (Fig. 2) and the water-soluble contrast enema - a microcolon aspect. (Fig. 3)



Figure 1. Dilated bowel loops without air-fluid levels

Differential diagnosis excluded other causes of neonatal bowel obstructions such as Hirschsprung`s disease, atresia and stenosis, annular pancreas, malrotation, duplication cyst, neonatal small left colon syndrome, neoplasia, hypothyroidism and other rarer causes.



Figure 2. Large dilated loop of bowel containing meconium and gas : soap bubble sign



Figure 3. Barium enema examination (microcolon aspect; the filling defects in the ileum represents concretions of the meconium)

Treatment:

Surgery has been used in 10 cases:

- Resection of the dilated ileum with Mikulicz enterostomy in 8 cases; (see Fig. 4-7)
 - Explorative laparotomy with drainage in 2 cases.
- Postoperative care in Intensive Care Unit:
- Antibiotic coverage;
 - Continued and persistent washouts;
 - Vigorous respiratory care including postural drainage and chest clapping.

End-to-end anastomosis was performed after 3 or 4 weeks.

In the cases with meconium plug syndrome the repeated hyperosmolar enema and adequate hydration has been used with good results and without any complications.



Figure 4. Dilated proximal intestine with distal ileum containing pellets of meconium



Figure 5. Resection of a dilated segment of ileum; thick meconium removed during surgery



Figure 6. Resected dilated segment of ileum

Evolution:

- Good postoperative evolution in 6 cases;
- 4 cases with fatal evolution (septic shock, cardio-respiratory failure, disseminated intravascular coagulation);
- Late complications related to mucoviscidosis.



Figure 7. Meconium ileus with distal perforation - the sigmoid colon

CONCLUSIONS

1. The meconium disease is seldom observed in neonatal obstructions.
2. It appears more often in males.
3. The diagnosis is difficult to establish.
4. The postoperative evolution is not always good.

REFERENCES

1. Lloyd DA. Meconium ileus. In: Welch KJ, Randolph JG, et al. (Editors) *Pediatric Surgery, Year Book*, Chicago, 1986, p. 849-58.
2. Boia E, Boia M. *Urgențe chirurgicale neonatale*, Timisoara 1995, Ed. POPA'S ART.
3. Santulli TV. Meconium ileus, In Holder TM, Ashcraft KW (editors) *Pediatric Surgery*, WB Saunders, Philadelphia, 1990, p. 356-73.
4. O'Neill JA, Grosfeld JL, Boles ET, et al. Surgical treatment of meconium ileus. *Am J Surg* 1970;119:99-105.
5. Noblett HR. Treatment of uncomplicated meconium ileus by Gastrografin enema: A preliminary report. *J Pediatr Surg* 1969;4:190-7.
6. Lennon J, Cavaleria M. Meconium diseases in infants with very low birth weight. *Semin Pediatr Surg* 2000;9(2):79-83.
7. Hajivassiliou CA. Intestinal obstruction in neonatal/pediatric surgery. *Semin Pediatr Surg* 2003;12(4):241-53.
8. Burge D, Drewett M. Meconium plug obstruction. *Pediatr Surg Int* 2004;20(2):108-10.
9. Mascarenhas MR. Treatment of gastrointestinal problems in cystic fibrosis. *Curr Treat Options Gastroenterol* 2003;6(5):427-41.
10. Ein S, Shandling B, Reilly B, et al. Bowel perforation with nonoperative treatment of meconium ileus. *J Pediatr Surg* 1987;12:146-7.