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General Principles and
Newborn Surgery

Pediatric Surgery

Prem Puri
Editor

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General Principles and Newborn
Surgery

With 487 Figures and 114 Tables

 Springer

Editor

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Abstract

Meconium peritonitis (MP) is defined as aseptic, localized, or generalized peritonitis due to leakage of meconium into the peritoneal cavity due to perforation of the fetal intestine in utero. The incidence of MP is 1/30000. Even

hypotheses related with ischemic events are most popular ones; neither of them has been substantiated yet. As first meconium reaches the ileum about 16th gestational week, timing of the event also determines the pathological types of MP, as fibro-adhesive, generalized, or cystic type. Intra-abdominal calcifications, being the most pathognomonic feature of the MP, are originated from the catalytic effect of fatty meconial compounds on the precipitation of calcium salts. A grading system described by Zangheri et al. can be used for international standardization of prenatal diagnosis. Antenatal diagnosis should also include investigation for congenital anatomical or structural anomalies, cystic fibrosis, and chromosomal abnormalities. When there is prenatal suspicion of MP in a patient, postnatal diagnosis is through abdominal and/or scrotal radiographs and

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ultrasonography. The natural history of prenatally diagnosed MP is different than diagnosed postnatally since some prenatally diagnosed cases resolve spontaneously. While there is not an established treatment modality in prenatal period, postnatal treatment varies depending on the signs and symptoms of the neonate.

Keywords

Meconium · Peritonitis · Antenatal bowel perforation · Neonate

Introduction

Meconium peritonitis (MP) is defined as aseptic, localized, or generalized peritonitis due to leakage of meconium into the peritoneal cavity because of perforation of the fetal intestine in utero. Agerty was the first to successfully operate a newborn with MP. The disease was first reported by Morgagni in 1761 in “De Sedibus et Causis Morborum”; Simpson managed to find 25 cases in 1838. (Agerty 1943; Simpson 1838) MP as a term is limited to the reaction caused by perforation that is already sealed or not but before the infant is born. Peritonitis caused by postnatal gastrointestinal perforations even when there is meconium is not included in meconium peritonitis and excepted as a different group of clinical problems (Cerise and Whitehead 1969). The incidence of MP is reported as 1/30000, which might be an underestimation due to spontaneous recovery of intestinal perforation and regression of the inflammatory process without neonatal clinical manifestations (Nam et al. 2007). There are more than 2,000 cases presented in the literature with a total mortality 37.3% since the disease first reported (Table 1). While it had a mortality rate approximately 70% in 1960s, the survival rate is over 90% today in USA but still has perinatal morbidity and mortality as high as 80% in third world countries (Saleh et al. 2009; Uchida et al. 2015). Improvements in prenatal diagnostic modalities and postnatal intensive care decrease

Table 1 Mortality rate in meconium peritonitis among 2,098 cases reported in world literature

Years	Total	Survivors	Mortality (%)
Before 1952	100	8	92.0
1952–1962	102	19	81.4
1963–1968	145	51	64.8
1969–1988	752	375	50.1
1989–1995	210	150	28.6
1996–2004	374	343	8.3
2004–2016	415	370	10.8
	2,098	1,316	37.3

mortality rates below 10% in some series (Chan et al. 2005; Zangheri et al. 2007).

Etiopathogenesis

Meconium, generated in the third gestational month, is composed of amniotic fluid with bile salts, cell debris, and proteins, which have been shown to activate immune cells including macrophages. Uric acid, intestinal enzymes, cholesterol, inorganic salts, and sugar also constitute the meconium. Chemical peritonitis is triggered by lipases and bile salts spilled into the abdomen. The inflammation is mediated through phagocytosis, release of chemical mediators, and antibody-dependent cell-mediated cytotoxicity by macrophages those infiltrate into the peritoneum. TNF- α production is reported to remarkably increase when encountered with meconium, which also results in fibrin deposition and severe intra-abdominal adhesion. The progressive pro-inflammatory cytokine reaction may also enhance the inflammatory reaction (Lally et al. 1999; Rubin et al. 1996; Shyu et al. 1994). Interleukins 6 and 8 are also associated with inflammatory response syndrome in MP. Their concentrations are found to increase not only in patient’s plasma but also in the cyst or ascites just after birth in very high amount (Kanamori et al. 2012). Drainage of the cystic fluid does not suppress the inflammation.

There are many hypothesis regarding the intrauterine intestinal perforation, neither of

which has been substantiated. Segmental absence of muscular coats, absence of muscularis mucosa, vascular occlusion, and general hypoxia of the fetus in the perinatal period are the most commonly speculated ones (Lloyd 1969; Rickham 1955; Vilhena-Moraes et al. 1964). In an experimental study, it has been shown that these are consequences of MP, not the cause (Boix-Ochoa 1982). The main etiological factors are intestinal atresia, intestinal volvulus, and meconium ileus. Hirschsprung's disease, meconium plug syndrome, congenital bands, internal hernias, Meckel's diverticulum, and rectal perforation might also be other etiological factors that lead to MP through intestinal perforation. Cystic fibrosis when accompanies MP is mostly a complication of meconium ileus. Its incidence has been reported to vary between 8% and 40% (Dirkes et al. 1995). On the other hand, neonatal hypoxia or anoxia and fetal respiratory distress may lead to MP, 80% of which etiology cannot be demonstrated despite pathological findings. It's proposed that, when there is a decrease in the blood flow to the intestines as seen in hypoxic fetus, the mucin production decreases and mucosal degenerations occur which eventually lead mucosa susceptible to ischemia. Thus, the bowel wall is defenseless to proteolytic enzymes of gastrointestinal system. Ileocecal region and splenic flexure are more vulnerable to ischemia as they are less vascularized. Almost 60% of all idiopathic lesions are found at this location.

Fetus is capable of swallowing amniotic fluid at 12th gestational week, the same time with the start of bile secretion. At 16th gestational week, meconium reaches the ileum for the first time. Whether the necrosis or perforation of the intestine occurs before or after this week, it would result in intestinal atresia either solely or with meconium peritonitis. Timing of the event also determines the pathological types of MP, as fibro-adhesive, generalized, or cystic type. If the perforated area sealed off before meconium passes, there will be no spillage of meconium in to the peritoneum. Digestive enzymes leak from perforated area which causes chemical peritonitis which eventually leads to a fibroblastic reaction.

It is called fibro-adhesive type if the site of perforation is effectively sealed off. In the fibro-adhesive type of MP, intestinal obstruction by adhesive bands might be encountered. If the perforation cannot be restricted and the intestine gets more inflamed and fixed, it forms a cystic cavity formed by fixed intestinal loops and filled with meconium called cystic type. This entity restrains the inflammation from spreading to the remainder part of the abdomen. Calcium deposits at the cyst wall which is clearly be seen at prenatal and postnatal imaging. If the adhesions caused by chemical peritonitis after intestinal perforation are more fibrinous than fibrous, it is called generalized type which is usually the most common type (Fonkalstrud et al. 1966; Gugliantini et al. 1979; Kolawole et al. 1973; Lorimer and Ellis 1966). Calcified meconium is scattered throughout the peritoneal cavity. In an experimental study with rats, meconium was demonstrated to give rise to a peritoneal reaction with fibroblastic proliferation that covers the lesion, followed by foreign body granulomas and calcification (Boix-Ochoa 1982). There may be local or generalized reaction, which leads adherence of intestinal loops with a fibrous tissue that is difficult to dissect. The exact location of the perforation is hard to find due to calcifications and disseminated meconium inclusions. Patton et al. also reported systemic spreading of meconium in their studies (Patton et al. 1998).

Intra-abdominal calcifications are originated from the catalytic effect of fatty meconial compounds on the precipitation of calcium salts. Intra-abdominal calcification could not be demonstrated in MP in animals with low serum levels of calcium. Light microscopic examination of these calcifications revealed that they are in response to keratin debris (Faripor 1984). However, keratin cannot be the only source because of the presence of granulomas devoid of keratin. Since some of these granulomas resemble gouty tophi, it may be because of inflammation caused by uric acid present in meconium.

The rare entity called meconium pseudocyst should be differentiated from cystic type of

MP. In cystic type MP, the inflamed bowel loops are fixed and lead to formation of an intraperitoneal cystic cavity with a fibrous wall. On the other hand, a meconium pseudocyst does not have an epithelium, which is lost due to inflammation. It is made of dilated intestine filled with meconium that has a smooth muscle layer connecting the cyst to the normal intestine (Minato et al. 2012). The formation of a pseudocyst represents an attempted to intra-abdominal healing process to confine the perforation.

Another type of presentation is microscopic MP that is an incidental finding, most of the time (Tibboel et al. 1981). Patients mostly presented with an intestinal atresia that occurred at very early stage of gestation. Bile pigments and squamous cell remnants could be found when peritoneum viewed carefully which is a proof for perforation. The presence of collagen, calcium deposits, and giant cells surrounding meconium particles demonstrates that the event should have taken for a considerable time ago.

Clinical Findings and Diagnosis

Antenatal and postnatal ultrasounds are the primary investigation modalities for diagnosis of MP. Prenatal ultrasonography not only provides an accurate diagnosis of the disease but also estimates the severity of it and determines the need for intervention (Fig. 1). Intraabdominal calcifications use to appear early, so prenatal diagnosis of

MP during pregnancy is feasible (Blumental et al. 1982; Bowen et al. 1984; Dunne et al. 1983; Garb and Riseborough 1980). A grading system described by Zangheri et al. can be used for international standardization of prenatal diagnosis by using the information related with fetal intra-abdominal calcifications, fetal ascites, pseudocysts, and bowel dilatations (Zangheri et al. 2007) (Table 2). Patients with a score greater than 1 have a high risk for urgent neonatal surgery, while the ones with 0 can be delivered at term without any complication. Antenatal diagnosis should also include investigation for congenital anatomical or structural anomalies, cystic fibrosis, and chromosomal abnormalities (Chan et al. 2005; Nam et al. 2007). Detection of cystic fibrosis is done by screening for the most common gene mutations and sweat chloride test and also is recommended screening for congenital infections including herpes simplex virus, cytomegalovirus, parvovirus B19, and toxoplasmosis (Cassacia et al. 2003).

Fetal magnetic resonance imaging (MRI) provides additional data for the diagnosis of MP (Fig. 2) besides ultrasonography (57.1% vs. 42%), even it is not needed in all cases (Chan et al. 2005). However, it might help assessment of any accompanying disorders, which is crucial due to its repercussions in the immediate postoperative period. Even though ultrasonography is sensitive, there are many diseases for the differential diagnosis of cystic masses such as ovarian cyst, duplication cyst, or mesenteric cyst that is difficult

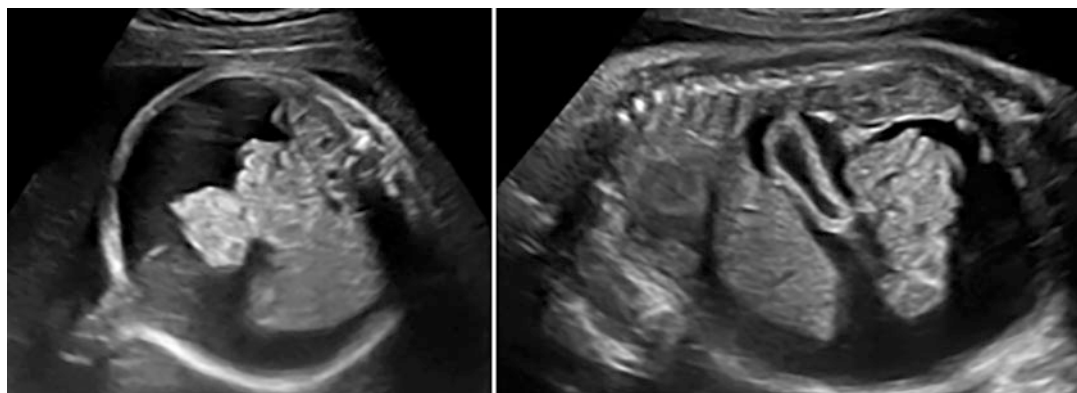


Fig. 1 Prenatal ultrasonography showing ascites with collapsed intestines

Table 2 Zangheri's grading system for meconium peritonitis

Score		
0		Intraabdominal calcification
1	A	Intraabdominal calcification with ascites
	B	Intraabdominal calcification with pseudocyst
	C	Intraabdominal calcification with bowel dilatation
2		Intraabdominal calcification with any of two associated findings
3		Intraabdominal calcification with all associated findings

to diagnose (Aydin 2016; Degnan et al. 2010). Limitations of prenatal ultrasonography in detection of calcifications are an ultrasonographic entity. (Zangheri et al. 2007) Fetal MRI can be a useful tool for description of the exact pathology and comorbidities.

When there is prenatal suspicion of MP in a patient, postnatal diagnosis is through abdominal and/or scrotal radiographs and ultrasonography. In patients without prenatal suspicion or diagnosis, the diagnosis of MP in the postnatal period is based on clinical presentation and radiological findings of intestinal obstruction. A newborn with abdominal distension is the very first sign, which is present at the birth or develops soon after. Bilious vomiting and stop in meconium pass can be other signs of intestinal obstruction. When the abdominal distention is severe, it may lead to respiratory distress. Abdominal radiographs might reveal pneumoperitoneum or calcifications in the peritoneal cavity, which is pathognomonic (Fig. 3) (Miller et al. 1988; Smith and Clatworthy 1961). In some cases, calcifications may also be seen at scrotum mimicking scrotal mass. Hyper-echogenic concretions with posterior acoustic shadowing on ultrasounds confirm calcifications, while cysts or ascites could be other findings (Fig. 4).

The natural history of prenatally diagnosed MP is different than diagnosed postnatally since some prenatally diagnosed cases resolve spontaneously. Because the bacterial overgrowth at meconium begins just after birth, immediate and proper diagnosis is fundamental for the

prognosis of neonate. Twenty-four percent of patients' cultures were found to be positive at first 12 h, while 86% were at 72 h. Patients operated between 36 and 48 h of life have a four times higher mortality rate than those operated during postnatal 24 h, while after 48 h, the mortality rates vary between 80% and 91% (Boix-Ochoa 1982; Tibboel and Molenaar 1984). While immediate diagnosis and surgery if needed is crucial, termination of pregnancy is unnecessary (Wang et al. 2008). Early detection of the disease is not associated with poor outcome. In a study, only 22% of fetuses with prenatally diagnosed MP required surgery with 11% mortality (Dirkes et al. 1995). However, an elective preterm delivery by cesarean section at 35th gestational week could be recommended to prevent disease progression and enable an early intervention (Saleh et al. 2009).

MP might be presented also as an inguinal or scrotal mass. These patients do not present any relevant history related with MP. Many of them present with unilateral hydrocele at birth. Scrotal, as intraabdominal, calcifications could be demonstrated by radiological imaging being pathognomonic for MP (Cook 1978; Gunn et al. 1978; Heydenrych and Marcus 1976). Nodules in these patients mostly regress spontaneously and do not require surgery.

Differential diagnosis should be done very precisely. Intra-abdominal calcifications may also be seen in cases of multiple intestinal atresia, colonic atresia, Hirschsprung's disease, high anorectal malformations, and cloacal anomalies. On the other hand, adrenal and liver calcifications can also be identified with cytomegalovirus, parvovirus, or hepatoblastoma (Aydin et al. 2013; Boix-Ochoa 1982; Bowen et al. 1984; Sciarrone et al. 2016).

Meconium periorchitis first reported in 1953 is the result of MP and spillage of meconium through open processus vaginalis. This entity can be diagnosed by ultrasonography during intra-uterine life, but mostly incidentally at the postnatal period. A detailed examination during fetal life is crucial to prevent any unnecessary orchiectomy in the neonate. Surgical excision might determine

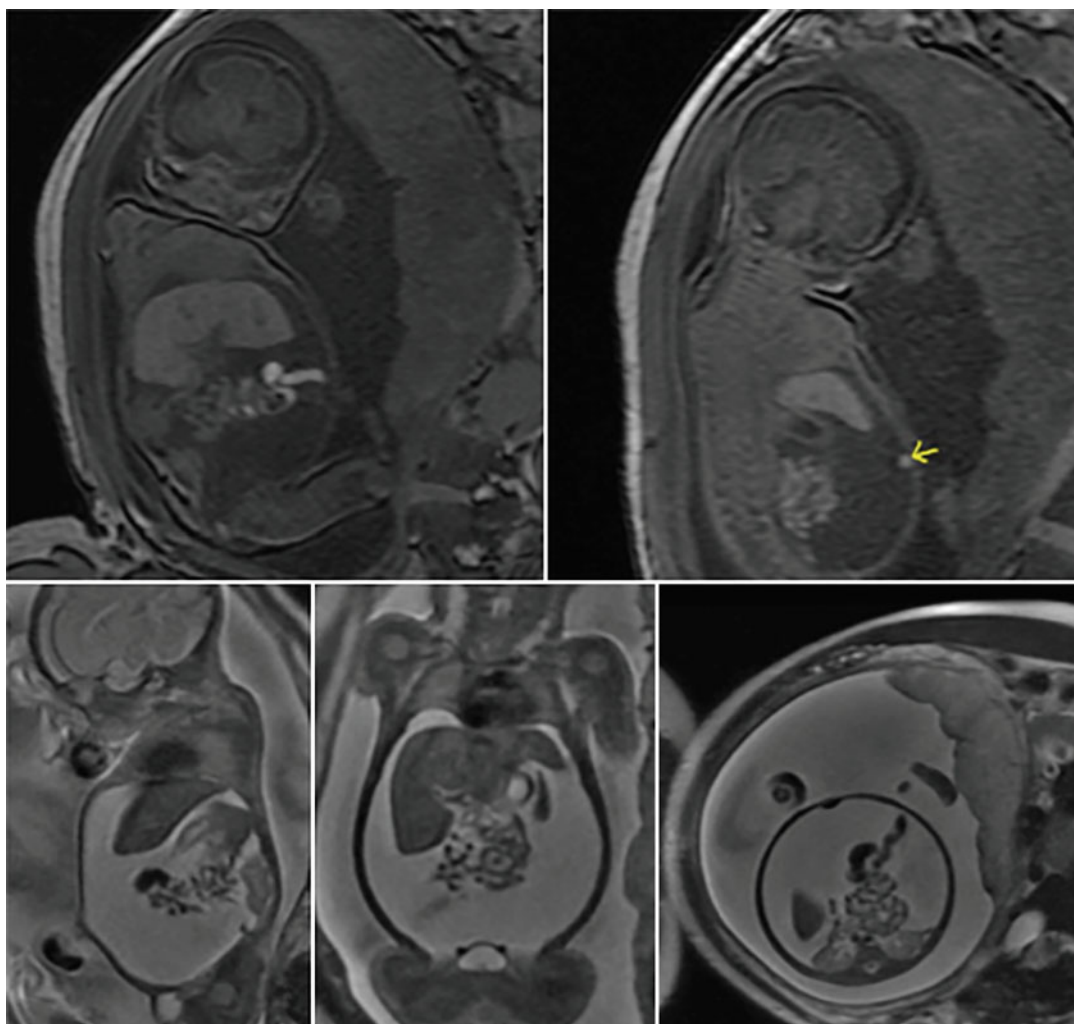


Fig. 2 Fetal MRI. Abnormal meconium dilated small bowel loop in mid-abdomen extending to the left abdominal wall in association with septation. Also shows complex ascites and small area of calcification (*arrow*)

the diagnosis but is not necessary (Alanbuki et al. 2013; Regev et al. 2009).

Treatment

There is not an established treatment modality in prenatal period. There are some occasional trials such as injection of urinary trypsin inhibitor into fetal abdomen to reduce meconium-induced chemical peritonitis and avoid a postnatal surgery or fetal paracentesis to diminish intraabdominal pressure to improve mesenteric vascular supply

and remove inflammatory debris (Izumi et al. 2011; Taba et al. 2010). Although there are many prenatally diagnosed cases with intraabdominal calcifications, only very few require postnatal surgery. Associated findings could be a good marker to decide who is a candidate for surgery and select them for delivery in tertiary neonatal surgical centers (Zerhouni et al. 2013).

While intestinal obstruction and/or perforation with MP is a definitive indication for surgery (Fig. 4), meconium peritonitis without any obstruction or perforation findings is just a

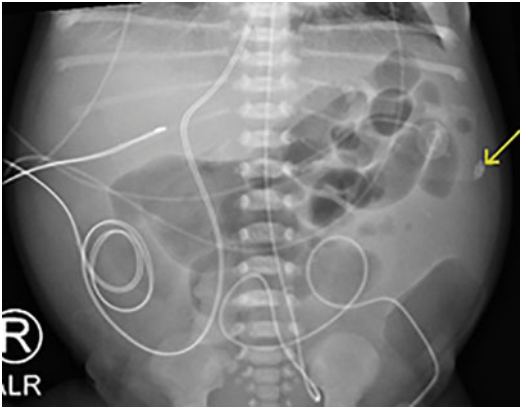


Fig. 3 Postnatal abdominal plain x-ray of a baby diagnosed as meconium peritonitis showing calcification in the abdomen (*arrow*)

candidate for follow-up. These children should be observed at least for 48 h without enteral feeding with appropriate antibiotic coverage. Oral gradually progressing feedings could be started to the ones without any clinical findings after this follow-up period.

For the ones that require surgery, all precautions should be taken to decrease morbidity and mortality such as monitoring vital signs, control of temperature to prevent heat loss in the operating theater, and prophylactic antibiotic. Even if the perforation is visible, the operative management should be condensed on intestinal resection and end-to-end anastomosis of the segments without trying to repair primarily. In localized or

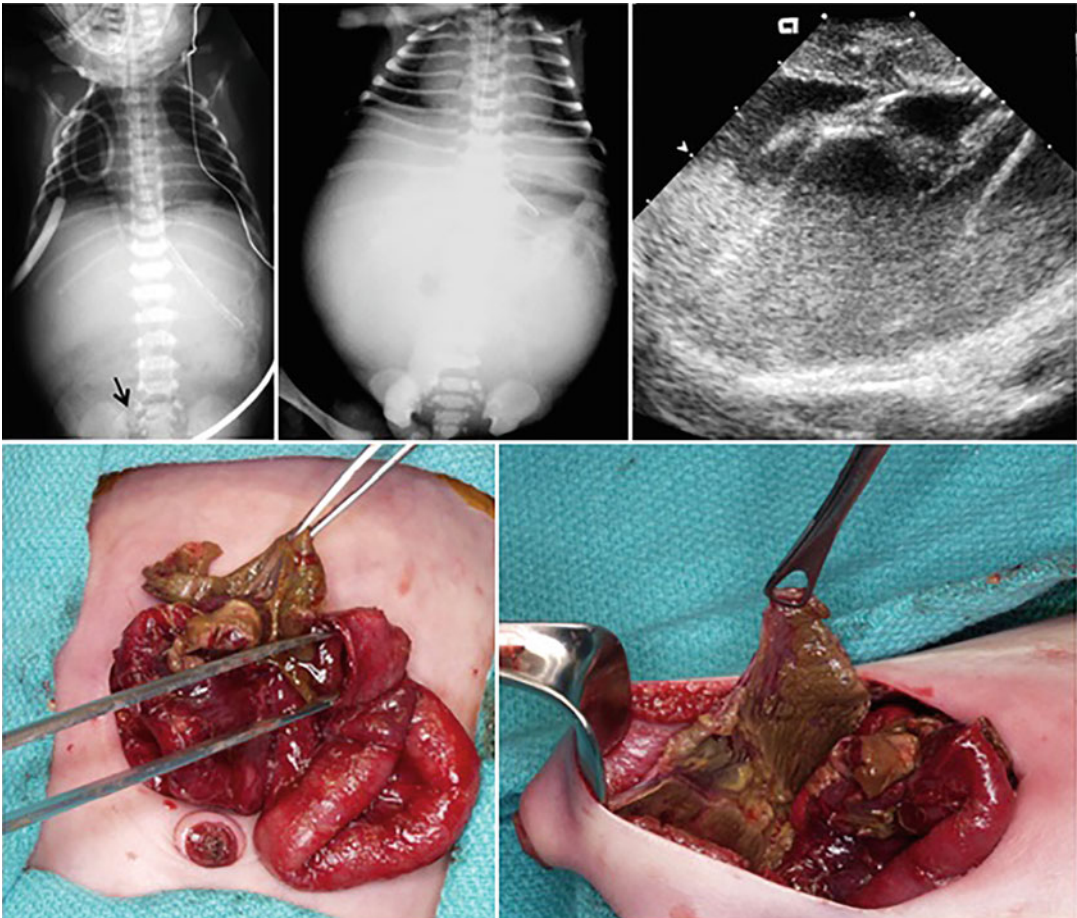


Fig. 4 Postnatal abdominal plain X-ray and US of a baby diagnosed as meconium peritonitis showing calcification in the abdomen (*arrow*) and ascites. Surgical appearance of

the bowel perforation with meconial intestinal and peritoneal staining

generalized type of MP, attempts for the lysis of adhesions should only be done in purpose to discover the perforation if possible or just relieve major obstruction due to the possibility of spontaneous relief of fibro-adhesive peritonitis in 8–14 days. After determination of etiology, surgical technique decision should rely on the general status of the patient and discordance of the intestinal calibers. Either end-to-end anastomosis according the Louw's technique or two-stage operation of Rehbein, temporary enterostomy and delayed reconstruction of intestinal integrity might be chosen. Most of the patients with MP could be treated with primary anastomosis but the non-stable low birth weight infants (Louw 1967; Miyake et al. 2011; Rickham 1955). In patients with cystic type MP, a two-stage operation should be done. Maintaining enteral and parenteral nutrition and allowing transportation of contents from proximal intestine to distal is necessary between two stages. Overall two-stage operations have a lower mortality rate than one-stage operation (7.2% vs 22%) (Boix-Ochoa 1982). They also have some advantages compared to one-stage operations such as: time of recovery is shorter, they do not have the risk of dehiscence of anastomosis due to infection, fibro-adhesive bands disappear by the time of second operation, calibration of the intestines becomes suitable for anastomosis, and there will be enough time for neuroendocrine system maturation.

Another treatment modality is cyst drainage and late laparotomy that might be chosen in patients not suitable for immediate operation but require intervention due to mass effect of the cyst. It should be accompanied by supportive decompression treatment, parenteral nutrition, and broad spectrum antibiotics. This modality is safe and effective in decompression of gastrointestinal tract and prevention of sepsis (Tanaka et al. 1993). However, in case of meconium pseudocyst, resection of the cyst and re-anastomosis of the segments should be the preferred treatment (Minato et al. 2012).

Complications

As most of the abdominal surgeries, ileus due to adhesions is the most frequent complication of MP. Anastomotic leakage, necrosis of ostomy, enterocutaneous fistula, respiratory problems related with cystic fibrosis, and sepsis are some other complications reported (Boix-Ochoa 1982). Gentle and delicate operative manipulation is required to reduce the mortality and morbidity in these cases.

Conclusion and Future Directions

In conclusion, current survival of the patient is up to 100% in some centers with the improvements in the surgical techniques and postnatal care of the neonates. Even the exact mechanism of MP is still unknown; with the advances in antenatal treatment modalities, early diagnosis of the patients is more frequent. As CF is one of the main known causes of MP, improvements in diagnosis and treatment of CF would decrease the incidence of MP and improve the outcome.

Cross-References

- ▶ [Anorectal Malformations](#)
- ▶ [Colonic and Rectal Atresias](#)
- ▶ [Hirschsprung's Disease](#)
- ▶ [Jejunioileal Atresia and Stenosis](#)
- ▶ [Malrotation](#)
- ▶ [Meconium Ileus](#)

References

- Agerty HA. A case of perforation of the ileum in newborn infant with operation and recovery. *J Pediatr.* 1943;22:233.
- Alanbuki AH, Bandi A, Blackford N. Meconium periorchitis: a case report and literature review. *Can Urol Assoc J.* 2013;7(7–8):495–8.
- Aydin E. Rapidly growing intraabdominal cyst in a newborn: intestinal duplication. *APSP J Case Rep.* 2016;7:18.

- Aydin E, Emre S, Adaletli I, et al. Incidentally diagnosed adrenal calcification. *J Turk Assoc Pediatr Surg.* 2013;2-3:89-92.
- Blumental DH, Rushovich AM, Williams RK, et al. Prenatal sonographic findings of meconium peritonitis with pathologic correlation. *J Clin Ultrasound.* 1982;10:350-2.
- Boix-Ochoa J. Patología quirúrgica del meconio. *Med Esp.* 1982;81:30-51.
- Bowen A, Mazer J, Zarabi M, et al. Cystic meconium peritonitis: ultrasonographic features. *Pediatr Radiol.* 1984;14:18-22.
- Cassacia G, Trucchi A, Nahom A, et al. The impact of cystic fibrosis on neonatal intestinal obstruction: the need for prenatal/neonatal screening. *Pediatr Surg Int.* 2003;19:75-8.
- Cerise EJ, Whitehead W. Meconium peritonitis. *Am Surg.* 1969;35:389.
- Chan KL, Tang MH, Tse HY, et al. Meconium peritonitis: prenatal diagnosis, postnatal management and outcome. *Prenat Diagn.* 2005;25:676-82.
- Cook PL. Calcified meconium in the newborn. *Clin Radiol.* 1978;29:541-6.
- Degnan AJ, Bulas DI, Sze RW. Ileal atresia with meconium peritonitis: fetal MRI evaluation. *J Radiol Case Rep.* 2010;4(3):15-8. <https://doi.org/10.3941/jrcr.v4i3.356>.
- Dirkes K, Crombleholme TM, Craigo SD, et al. The natural history of meconium peritonitis diagnosed in utero. *J Pediatr Surg.* 1995;30:979-82.
- Dunne M, Haney P, Sun CCJ. Sonographic features of bowel perforation and calcific meconium peritonitis in utero. *Pediatr Radiol.* 1983;13:231-3.
- Faripor F. Origin of calcification in healed meconium peritonitis. *Med Hypotheses.* 1984;14:51-6.
- Fonkalstrud EW, Ellis DG, Clatworthy Jr HW. Neonatal peritonitis. *J Pediatr Surg.* 1966;1:227-39.
- Garb M, Riseborough J. Meconium peritonitis presenting as fetal ascites on ultrasound. *Br J Radiol.* 1980;53:602-4.
- Gugliantini P, Caione P, Rivosecchi M, et al. Intestinal perforation in newborn following intrauterine meconium peritonitis. *Pediatr Radiol.* 1979;8:113-5.
- Gunn LC, Ghionzoli OG, Gardner HG. Healed meconium peritonitis presenting as a reducible scrotal mass. *J Pediatr.* 1978;92:847.
- Heydenrych JJ, Marcus PB. Meconium granulomas of the tunica vaginalis. *J Urol.* 1976;115:596-8.
- Izumi Y, Sato Y, Kakui K, et al. Prenatal treatment of meconium peritonitis with urinary trypsin inhibitor. *Ultrasound Obstet Gynecol.* 2011;37(3):366-8.
- Kanamori Y, Terawaki K, Takayasu H, et al. Interleukin 6 and interleukin 8 play important roles in systemic inflammatory response syndrome of meconium peritonitis. *Surg Today.* 2012;42(5):431-4.
- Kolawole TM, Bankole MA, Olurin EO, et al. Meconium peritonitis presenting as giant cysts in neonates. *Br J Radiol.* 1973;46:964-7.
- Lally KP, Mehall JR, Xue H, et al. Meconium stimulate a pro-inflammatory response in peritoneal macrophages: for meconium peritonitis. *J Pediatr Surg.* 1999;34:214-7.
- Lloyd JR. The etiology of gastrointestinal perforations in the newborn. *J Pediatr Surg.* 1969;3:77.
- Lorimer WS, Ellis DG. Meconium peritonitis. *Surgery.* 1966;60:470-5.
- Louw MB. Resection and end to end anastomosis in the management of atresia and stenosis of small bowel. *Surgery.* 1967;62:940.
- Miller JP, Smith SD, Newman B, et al. Neonatal abdominal calcification: is it always meconium peritonitis? *J Pediatr Surg.* 1988;23:555-6.
- Minato M, Tadao O, Hisayuki M, et al. Meconium pseudocyst with particular pathologic findings: a case report and review of the literature. *J Pediatr Surg.* 2012;47(4):e9-12. <https://doi.org/10.1016/j.jpedsurg.2011.11.050>.
- Miyake H, Urushihara N, Fukumoto K, et al. Primary anastomosis for meconium peritonitis: first choice of treatment. *J Pediatr Surg.* 2011;46(12):2327-31.
- Nam SH, Kim SC, Kim DY, et al. Experience with meconium peritonitis. *J Pediatr Surg.* 2007;42:1822-5.
- Patton WL, Lutz AM, Willmann JK, et al. Systemic spread of meconium peritonitis. *Pediatr Radiol.* 1998;28:714-6.
- Regev RH, Markovich O, Arnon S, et al. Meconium periorchitis: intrauterine diagnosis and neonatal outcome: case reports and review of the literature. *J Perinatol.* 2009;29(8):585-7.
- Rickham PP. Peritonitis of the neonatal period. *Arch Dis Child.* 1955;30:23.
- Rubin BK, Tomkiewicz RP, Patrinos ME, et al. The surface and transport properties of meconium and reconstituted meconium solutions. *Pediatr Res.* 1996;40:834-8.
- Saleh N, Annegret G, Ulrich G, et al. Prenatal diagnosis and postnatal management of meconium peritonitis. *J Perinat Med.* 2009;37(5):535-8. <https://doi.org/10.1515/JPM.2009.097>.
- Sciarrone A, Teruzzi E, Pertusio A, et al. Fetal midgut volvulus: report of eight cases. *J Matern Fetal Neonatal Med.* 2016;29(8):1322-7.
- Shyu MK, Chen CD, Hsieh FJ, et al. Intrauterine intervention in a case of recurrent meconium peritonitis. *Prenat Diagn.* 1994;14:993-5.
- Simpson JY. Peritonitis in the fetus in utero. *Edinb Med Surg J.* 1838;15:390.
- Smith B, Clatworthy HW. Meconium peritonitis: prognostic significance. *Pediatrics.* 1961;27:967.
- Taba R, Yamakawa M, Harada S, et al. A case of massive meconium peritonitis in utero successfully managed by planned cardiopulmonary resuscitation of the newborn. *Adv Neonatal Care.* 2010;10(6):307-10.
- Tanaka K, Hashizume K, Kawarasaki H, et al. Elective surgery for cystic meconium peritonitis: report of two cases. *J Pediatr Surg.* 1993;28:960-1.

- Tibboel D, Molenaar JC. Meconium peritonitis. A retrospective, prognostic analysis of 69 patients. *Z Kinderchir.* 1984;39:25–8.
- Tibboel D, Gaillard JL, Molenaar JC. The ‘microscopic’ type of meconium peritonitis. *Z Kinderchir.* 1981;34:9–16.
- Uchida K, Yuhki K, Kohei M, et al. Meconium peritonitis: prenatal diagnosis of a rare entity and postnatal management. *Intractable Rare Dis Res.* 2015;4(2):93–7. <https://doi.org/10.5582/irdr.2015.01011>.
- Vilhena-Moraes R, Cappellano G, Mattosinho Franca LC, et al. Peritonite meconial. *Rev Paul Med.* 1964;65:231.
- Wang CN, Chang SD, Chao AS, et al. Meconium peritonitis in utero – the value of prenatal diagnosis in determining neonatal outcome. *Taiwan J Obstet Gynecol.* 2008;47(4):391–6.
- Zangheri G, Andreani M, Ciriello E, et al. Fetal intra-abdominal calcifications from meconium peritonitis: sonographic predictors of postnatal surgery. *Prenat Diagn.* 2007;27:960–3.
- Zerhouni S, Mayer C, Skarsgard ED. Can we select fetuses with intra-abdominal calcification for delivery in neonatal surgical centers? *J Pediatr Surg.* 2013;48(5):946–50.