Classic eggshell calcifications and an elective surgical approach in giant cystic meconium peritonitis (GCMP): a case report

Erik R. Barthel\textsuperscript{1}, Allison L. Speer\textsuperscript{1}, Daniel E. Levin\textsuperscript{1}, Bindi J. Naik-Mathuria\textsuperscript{2}, Tracy C. Grikscheit\textsuperscript{1}\ast

Address:
\textsuperscript{1} Children's Hospital Los Angeles
Division of Pediatric Surgery
4650 Sunset Blvd Mailstop 35
Los Angeles CA 90027
USA

\textsuperscript{2} Pediatric Surgery
Texas Children's Clinical Care Center
6701 Fannin St., 8th Floor
Houston, TX 77030

Email:
\texttt{ebarthel@chla.usc.edu}
\texttt{aspeer@chla.usc.edu}
\texttt{dalevin@chla.usc.edu}
\texttt{bnaik@texaschildrens.org}
\texttt{tgrikscheit@chla.usc.edu}\ast

\ast Author for correspondence.
Abstract
Introduction: Along the spectrum of meconium peritonitis, giant cystic meconium peritonitis (GCMP) is relatively rare. These patients often present with nonspecific physical findings such as distension and emesis. However, plain abdominal films remain invaluable for identifying the characteristic calcifications seen with meconium pseudocyst, and large eggshell calcifications are pathognomonic for the giant cystic subtype.

Case presentation: We present classic plain x-ray findings and an intraoperative image of a premature low birth weight male infant treated for GCMP with a staged procedure involving peritoneal drainage, ostomy creation and closure.

Conclusion: Pediatric surgeons have a range of potential therapeutic approaches for GCMP. A delay of definitive surgical management in the setting of massive abdominal soiling is a safe and acceptable strategy if adequate temporizing drainage is performed in the early perinatal period.

Introduction
Meconium pseudocyst was first described by Carl von Rokitansky and others in the German-language literature in the 19th century [1,2]. Reports of meconium peritonitis appear in the English literature beginning in the early 20th century [3,4]. Infants may present with failure to pass meconium, abdominal distension, emesis, and symptoms of peritonitis [5]. Meconium pseudocyst occurs as a result of in utero or postnatal intestinal perforation. This perforation is often secondary to ischemia or obstruction and although meconium is sterile, chemical irritation of the peritoneum can ensue [6]. As the body attempts to wall off the source of inflammation, dystrophic calcification occurs, resulting in the classic eggshell calcifications seen on abdominal radiography demonstrated in Fig. 1 [7].

Case presentation
A 2.2 kg 31 1/7-weeks gestation boy was born vaginally to a 21 year-old G1P0 woman after prolonged rupture of membranes. He had a prenatal history of dilated bowel loops and hydrops on ultrasonography, as well as fetal anemia requiring intrauterine blood transfusion 18 days prior to delivery. The patient was intubated at 11 minutes of life for poor respiratory effort. Cord arterial blood gas on transfer to NICU revealed a pH of 7.32, pCO2 50, pO2 20. His abdomen was markedly distended, exhibited progressive erythema, and radiographs showed dilated bowel loops and significant, well-demarcated bilateral calcifications (Fig. 1, arrows).

The patient was diagnosed with giant meconium pseudocyst, a walled-off collection of meconium that can cause neonatal peritonitis, which is distinguished from other forms of meconium peritonitis by the persistence of a communication between the digestive lumen and the peritoneal cavity leading to the entire abdomen becoming filled with meconium [8]. On the second day of life, he was taken to the operating room, placed in the supine position, and exploratory laparotomy was performed. At surgery extensive soiling and inflammation precluded identification of intestinal
loops for safe stoma formation. Bilateral Penrose drains were therefore placed as a temporizing measure to allow decompression of meconium from the abdomen.

Although the patient’s sepsis improved with source control, he remained obstructed with high nasogastric output. He also had persistent impairment of his pulmonary function from the obstruction and abdominal distension. Therefore, he was taken for a second operation one month later. In this procedure, an extensive lysis of adhesions was performed, revealing a meconium cyst remnant (Fig. 2). Distal microcolon was also visualized, and an ileostomy was brought out at this time. Feeds were started on postoperative day 11 and were slowly advanced over the next four weeks to goal. The patient was discharged and continued to gain weight appropriately at home. He was brought back in for elective ileostomy closure at six months of life.

Discussion
Outcomes for neonatal meconium peritonitis have improved over time, with relatively high mortality rates reported from the 1950s improving to a survival rate of about 90% with modern advances in pediatric surgery and perinatology [9]. Surgery remains the definitive treatment, but there is a lack of standardization in the literature. Multiple drainage procedures, stoma creation and final ostomy closure are still common [10]. Of note, a recent retrospective series of 38 patients demonstrated improved survival with primary anastomosis, except for low birth weight infants [9]. Our patient had extensive soilage and adhesions that required a temporary drainage procedure, and definitive stoma creation was delayed for one month. A similar strategy has been reported previously [11]. These authors presented two cases in which they placed external drains under ultrasound guidance, and then waited 16 and 20 days, respectively, before proceeding to laparotomy. They elected to perform a primary ileal anastomosis at surgery in both patients. However, we chose to bring out a stoma at delayed laparotomy given the patient’s prolonged NICU stay and sepsis course. While our choice of stoma creation, rather than primary anastomosis, is consistent with the recommendations of ref. [9] given the birth weight of 2.2 kg, we stress that our decision to start with a temporary drainage procedure prior to definitive surgery is more consistent with the initial management by the authors of [11].

Conclusion
A classic appearance of large calcifications on plain abdominal films in the setting of a distended and distressed neonate should place giant cystic meconium peritonitis (GCMP) high upon the differential diagnosis. Initial peritoneal drainage with a delay in definitive surgical management is a reasonable approach in the setting of massive meconium soiling of the abdominal cavity that prohibits the safe identification of bowel loops for stoma creation. At reoperation, bringing out a stoma is a safe option, especially for low birth weight infants.
Abbreviations
GCMP – giant cystic meconium peritonitis

Consent
Written informed consent was obtained from this minor patient’s parents for publication of this case report and the accompanying images. A de-identified copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that they have no competing interests.

Authors’ contributions
ERB, ALS, and DEL performed the literature search and wrote the manuscript. BJN-M and TG performed the surgeries, provided intraoperative images and critically reviewed the manuscript prior to submission. All authors have read and approved the final manuscript.

Acknowledgements
The authors thank the Children’s Hospital Los Angeles Division of Pediatric Surgery for educational and logistical support. ERB and TCG are supported by the California Institute for Regenerative Medicine, grant numbers TG2-01168 and RN2-00946-1, respectively. ALS is supported by a Society of University Surgeons Ethicon Scholarship.

Figure legend
Figure 1. A: Plain abdominal radiograph showing bilateral eggshell calcifications consistent with giant cystic meconium peritonitis (GCMP, arrows). B: Photograph taken at the patient’s second operation, where the peritoneal drain was removed and an ileostomy was brought out. The remnant of the original meconium pseudocyst can be seen at the bottom of the figure (arrow).

References


