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ORIGINAL ARTICLE

Management of fetal extraperitoneal rectal perforation: a case series and review of the literature

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Abstract

Purpose Fetal extraperitoneal rectal perforation (FERP) is an extremely rare entity. The objective of this report is to review the available literature on this condition and to add our experience with four additional cases managed at our institution.

Methods A literature search was performed for journal articles addressing this condition. Management strategies and outcomes were then analysed, together with additional information provided from retrospective record review of four cases managed at our institution.

Results A total of 18 patients were identified and included, 14 from the literature and 4 from our records. Initial investigations varied between authors with contrast enema being the most frequently performed study (7/18). All patients were treated with faecal diversion via colostomy formation. Exploratory laparotomy was performed in 6 cases, perineal debridement and washout in 9 cases while drains were left in situ in 14 cases. At 3–6 months of age, a distal contrast study was performed before closure of colostomy.

Conclusion Although FERP is a rarely encountered clinical condition, timely recognition and appropriate management can result in good outcomes. Diagnosis can be achieved based on clinical and abdominal X-ray features alone. General management principles involve a diverting colostomy and extended drainage with closure of the colostomy 3–6 months later.

Keywords Fetal extraperitoneal rectal perforation · Diverting colostomy · Meconium cyst

Introduction

Fetal extraperitoneal rectal perforation (FERP) is an extremely rare entity [1–4]. At present, the general management principles include faecal diversion via a sigmoid colostomy and drainage of the meconium-filled cavity. The role of debridement of the cavity is debated [2, 3], as is the need for an exploratory laparotomy or laparoscopy at the time of colostomy formation [2, 4]. Debate also exists about the need for pre-operative workup. This report reviews the investigative and management strategies previously adopted and how they have evolved, adding our experience with an additional four cases that were managed at our institution.

Materials and methods

A retrospective review of the medical records of four cases of FERP that presented at Chris Hani Baragwanath Academic Hospital (CHBAH) and Charlotte Maxeke Johannesburg Academic Hospital (CMJAH) over the past 5 years was performed.

A review of the literature was also performed in order to identify the described investigative and management strategies for this condition, how they have evolved over time and how the approach used in our institutions compares to the globally accepted interventions. A PubMed search was performed using the term ‘fetal extraperitoneal rectal perforation’. Only human subjects were included. All articles meeting these criteria were reviewed with no date restriction. Articles in which the diagnosis of FERP were incorrect or in which the management strategies were not clearly explained were excluded.

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Results

Case series

Case 1

A 3 days old term boy was referred to our institution with enlarged genitalia, a left dark discoloured buttock mass and meconium draining from a sinus on the left hemi-scrotum (Fig. 1). Abdominal X-ray showed normal gas distribution with absence of gas in the rectum and an air-fluid level in the left scrotum/buttock (Fig. 2). His white cell count and C-reactive protein were within normal limits. A divided descending colostomy with distal mucous fistula was fashioned on day 3 of life via a left iliac fossa incision and the extraperitoneal meconium collections were widely drained. Intra-operatively, no intraperitoneal extension was found.

His post-operative course was complicated by an intracranial bleed, recurrent seizures and apnoea episodes due to a congenital intracranial anomaly identified on computed tomography. Despite these medical complications, his post-operative surgical plan proceeded without incident. He underwent a distal pressure-augmented colostogram at 5 months of life which demonstrated a normal rectum without any residual fistula. Closure of colostomy was then performed and the patient was subsequently discharged from follow up without complications.

Case 2

A term girl, appropriate for gestational age, was referred to our institution on day one of life with a right labium majus mass and meconium draining from a sinus located medial to the mass (Fig. 3).

A divided colostomy with distal mucous fistula was fashioned on day 2 of life via a left iliac fossa incision with wide drainage of the extraperitoneal meconium collections. She had an uneventful post-operative course. On subsequent follow ups, a significant reduction in size of the mass and closure of the fistula were observed.

A later distal colostogram showed a persistent recto-labial fistula. A modified Swenson-type pull through with fat pad was then used to repair the fistula [5]. Histological examination of the resected bowel ruled out the presence of Hirschsprung disease. The colostomy has been subsequently closed and the child is thriving. She has not yet reached the age of toilet-training but reports the sensation of needing to defecate.

Case 3

A 5 days old term male was referred to our department with an enlarging right buttock mass (Fig. 4). There was no

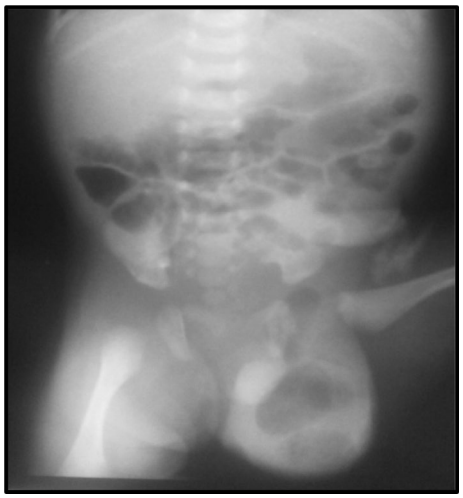

Fig. 2 Abdominopelvic radiograph showing air in the mass of the left buttock/scrotum in Case 1

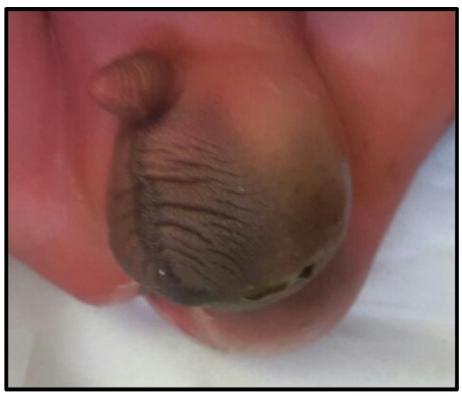

Fig. 1 Left buttock and scrotal mass with sinus on the left hemi-scrotum in Case 1

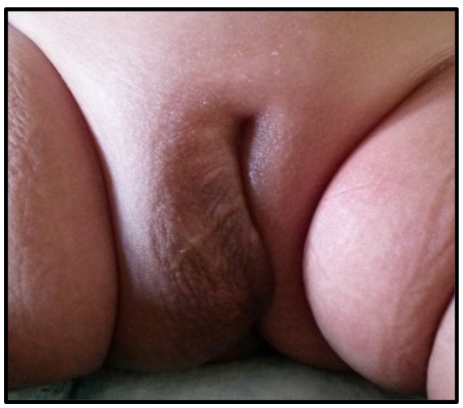

Fig. 3 Enlarged right labium majus in Case 2

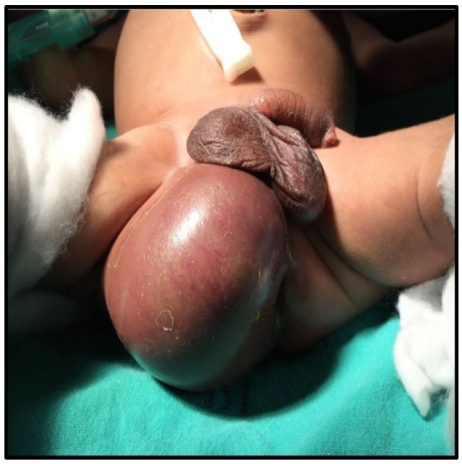

Fig. 4 Right buttock mass in Case 3

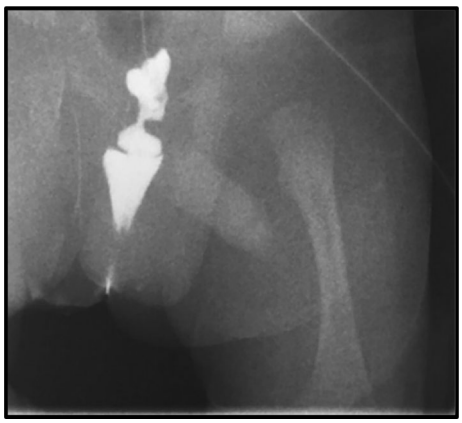

Fig. 5 Distal colostogram showing rectal stricture in Case 3

evidence of a draining sinus and the baby was passing stool normally through the anus.

He was taken for an examination under anaesthesia and was found to have a meconium-filled cyst of the right buttock. The decision was made to fashion a divided sigmoid colostomy with distal mucous fistula, evacuate the meconium and leave a pencil drain in the cavity.

A distal colostogram performed prior to closure of colostomy showed a rectal stricture at the peritoneal reflection (Fig. 5). At examination under anaesthesia, the stricture could be calibrated with a size twelve Hegar and the decision was made to reverse the colostomy and manage the stricture with anal dilatations. By 6 months post-colostomy closure the child was thriving with resolution of the stricture and without signs of constipation. He was subsequently discharged from follow up.

Fig. 6 Large scrotal mass extending posterior to the perineum in Case 4

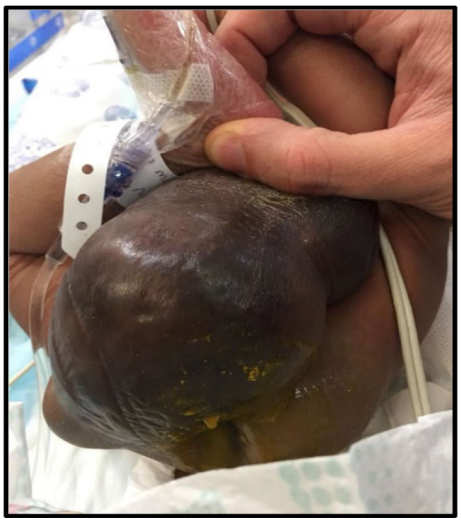

Fig. 7 Meconium visibly draining from the mass post-aspiration in Case 4

Case 4

A 1 day old term male presented with a soft, fluctuant, non-tender, darkly discoloured mass involving the scrotum and extending posterior to the perineum (Fig. 6). Aspiration of the mass demonstrated the presence of meconium (Fig. 7). Abdominal X-ray demonstrated minimally distended loops of bowel and no air-fluid levels in the scrotum. A contrast enema was performed due to suspicion of rectal duplication. This demonstrated a connection between the bowel and scrotal mass (Fig. 8).

A Hartmann colostomy and drainage of the perineal cyst was performed on day one of life. The colostomy was

performed via a transverse supraumbilical incision while drainage was accomplished through incision of the mass in the midline, decompression and washout, followed by insertion of a pencil drain in the scrotum.

The child was discharged on day eleven post-surgery. He has subsequently attended follow up at our colorectal clinic and is doing well. A rectal suction biopsy excluded the presence of Hirschsprung disease. The mass has significantly reduced in size, the repeat contrast enema shows no signs of persistent fistula and he is due for colostomy closure.

Table 1 summarises the information in terms of demographic, clinical features, initial investigations, acute management and outcomes in the four patients managed at our institution.

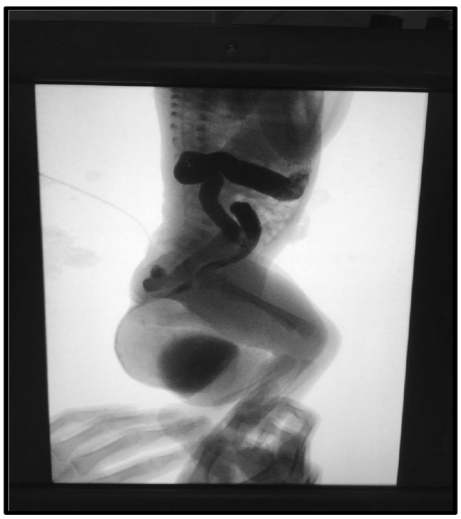

Fig. 8 Pre-operative contrast enema demonstrating contrast in the scroto-perineal mass in Case 4

Literature review

We found 17 reports of FERP in the literature. Three reports were excluded as they did not meet the criteria: two of them because the site of perforation was unclear [6, 7] and one of them because of insufficient information regarding the clinical presentation and management [8].

Fourteen cases were therefore included in the review. Six were male and eight were female. All but two of these cases were born at term. Table 2 summarises the information in terms of demographic, clinical features, initial investigations, acute management and outcomes.

All 14 cases presented with a mass of the perineum, genitals or buttock. In 5 (37%) the mass was mainly on the left, in 3 (21%) it was mainly on the right, in 3 (21%) it was bilateral while in the remaining 3 (21%) it was in the midline. Three cases (21%) demonstrated drainage of meconium

Table 1 Information in terms of demographic, clinical features, initial investigations, acute management and outcomes for the four patients with fetal extraperitoneal rectal perforation managed at our institution

Case	Demographics	Clinical features	Pre-operative imaging	Acute Management				Outcomes
				Laparotomy/laparoscopy	Stoma	Debridement	Drainage	
1	Male, term, day 3	Enlarged genitalia Left buttock mass with skin discolouration Meconium draining from left scrotal mass	AXR		X		X	Spontaneous fistula closure
2	Female, term, day 1	Discoloured right labium majus mass Meconium draining from medial side of mass	AXR		X		X	Persistent fistula requiring closure with modified Swenson-type pull through
3	Male, term, day 5	Enlarging discoloured right buttock mass	AXR		X		X	Spontaneous fistula closure Rectal stricture treated with dilatations
4	Male, term, day 1	Discoloured mass involving scrotum and perineal body	AXR Contrast enema	X	X		X	Spontaneous fistula closure

AXR abdominal radiograph

Table 2 Information in terms of demographic, clinical features, initial investigations, acute management and outcomes for the 14 patients with fetal extraperitoneal rectal perforation encountered in the literature

Author	Gender, gestational age, age at presentation	Clinical features	Pre-operative imaging	Acute Management				Outcomes
				Laparotomy/laparoscopy	Stoma	Debridement	Drainage	
Misudo et al. [10]	Female, postterm, day 3	Indurated left buttock mass Abdominal distension on day 5 due to urinary retention	Pelvic U/S Voiding cystogram Contrast enema Needle biopsy Sweat test	X	Not performed at initial surgery	X	X	Persistent fistula post-laparotomy. Required stoma
Sundararajan et al. [3]	Female, term, day 1	Cystic right buttock swelling Bluish discolouration of skin Subsequent rupture with meconium drainage	AXR Contrast enema Fistulogram Abdominal U/S	X	X		X	Spontaneous fistula closure
Pitcher et al. [1]	Female, term, day 5	Cystic left buttock mass Signs of peritonitis/pneumoperitoneum	Contrast enema	X	X		X	Spontaneous fistula closure
	Female, preterm, day 1	Cystic left buttock mass Signs of peritonitis/pneumoperitoneum	Contrast enema, AXR	X	X		X	Spontaneous fistula closure
	Male, term, day 4	Cystic right anterior perineal mass Tracking of gas into thigh			X	X	X	Spontaneous fistula closure
	Male, term, day 2	Cystic buttock mass			X	X	X	Spontaneous fistula closure
	Female, term, day 1	Very large cystic left anterior perineal mass Subsequent rupture			X	X	X	Spontaneous fistula closure
	Male, term, day 14	Cystic posterior midline mass, aplasia cutis	Contrast enema		X			Persistent fistula requiring closure with posterior sagittal approach
	Female, term, day 2	Enlarging discoloured cystic mass in sacral region Area of necrotic skin	AXR		X	X	X	Spontaneous fistula closure

Table 2 (continued)

Author	Gender, gestational age, age at presentation	Clinical features	Pre-operative imaging	Acute Management				Outcomes
				Laparotomy/laparoscopy	Stoma	Debridement	Drainage	
	Male, term, day 1	Gaseous mass of right perineum and scrotum			X	X		Spontaneous fistula closure
	Male, term, day 1	Bilateral buttock masses Abdominal distension		X	X			Spontaneous fistula closure Rectal stricture treated with dilatations
Tan et al. [9]	Female, late preterm, day 1	Left labioperineal swelling Discoloured skin	AXR Perineal U/S Pelvic x-ray Pelvic CT Pelvic MRI	Duodenoduodenostomy for duodenal atresia (day 1)	Delayed—day 5			Excision of mass via posterior sagittal approach (13 weeks)
Ibrahim et al. [4]	Female, late preterm, 1 day pre-delivery/day 1	Mass between fetal thighs on antenatal U/S Cystic swelling of perineum and buttocks Brownish discharge from left buttock Thick, mildly hyperpigmented skin	Perineal U/S Spinal U/S AXR Contrast enema	X	X	X	X	Spontaneous fistula closure
Garge et al. [2]	Male, term	Abdominal distension Bilateral fluctuant buttock swellings involving scrotum with skin discolouration	AXR Abdominal U/S Scrotal U/S Perineal U/S	X	X		X	Spontaneous fistula closure

U/S ultrasound, *AXR* abdominal radiograph, *CT* computerised tomography, *MRI* magnetic resonance imaging

from the mass and three cases (21%) had signs of peritoneal contamination.

The most frequently mentioned pre-operative imaging was contrast enema, performed in six patients (43%). Abdominal and pelvic radiographs were mentioned in five cases (37%), ultrasonography was performed in five patients (37%) while computerised tomography and magnetic resonance imaging scans were performed in one patient.

All cases were managed with a divided colostomy to achieve faecal diversion. In one case this was not performed at the initial operation, which involved only exploratory laparotomy with pelvic debridement and placing of a retrorectal drain. Two days later faeces was discharging from the site of the drain and the child underwent a relook laparotomy with stoma creation.

In seven patients (50%) the stoma appears to have been fashioned through an oblique left iliac fossa incision. In six cases (43%) the stoma was fashioned following a formal exploratory laparotomy and one patient underwent a pre-operative laparoscopic exploration. In six of these seven cases (86%) exploration was performed in order to rule out peritoneal contamination while in one it was due to concomitant urinary retention.

For 3 of the 14 patients (21%), no information regarding the treatment of the meconium-filled mass was described. Drainage of the cavity was described in 9 of the 14 cases (64%), whereas debridement was described in 7 of the 14 cases (50%) with 6 patients undergoing both procedures. In 1 patient the decision was made not to drain or debride the mass, but this resulted in persistence of the mass by 13 weeks of age, at which time it was excised through a posterior sagittal approach.

All 14 cases in the literature underwent a contrast enema prior to closure of colostomy. The colostogram demonstrated a spontaneous closure of the fistula in 13 out of 14 patients (93%). A persistent fistula was identified in only one patient and this was closed via a posterior sagittal approach before stoma reversal. Another patient developed a rectal stricture which was demonstrated at contrast enema and treated conservatively with anal dilatations prior to reversal.

Discussion

Fetal extraperitoneal rectal perforation is an extremely rare entity with few reported cases in the literature. The term FERP was coined by Pitcher et al. in 2008 in an article which added nine cases to the relatively limited body of information about the condition [1, 9].

While the exact aetiology and pathogenesis of this condition has not yet been determined, three theories have been proposed: local ischaemia of the rectal wall late in gestation, forceful peristalsis against a distal obstruction and herniation of the supralevator rectum through the pelvic floor with resultant strangulation and perforation [1-4, 9].

Despite the uncertainty regarding the exact pathogenesis of FERP, it presents with typical clinical features. These features include a large perineal, buttock or genital swelling often with skin discolouration secondary to underlying meconium. The mass is usually deviated to one side rather than being located in the midline. It is usually soft to palpation and non-tender. A fistula draining meconium can also be observed [4]. Differential diagnoses include a sacrococcygeal teratoma, rectal duplication cyst/diverticula, ischiorectal abscess and imperforate anus [1, 3, 9].

A sacrococcygeal teratoma usually does not display skin discolouration and would not discharge meconium on rupturing [3]. Rupture of a rectal duplication cyst or diverticula may drain meconium but if a mass is present it is usually palpable on per rectum examination. An ischiorectal abscess is unusual in a newborn infant and would present with tenderness, warmth, erythema and drainage of pus instead of meconium [3]. An imperforate anus is easily ruled out in FERP cases by the presence of a normally-sited anus [3].

The clinical features of FERP are so typical that some authors believe that faecal diversion and drainage of the cyst should be instituted without the need for further investigations [4]. On the other hand, some authors suggest that the rarity of the condition makes diagnosis on clinical grounds alone difficult [2] and therefore recommend that pre-operative investigations be performed.

It is not easy to understand exactly how many of the authors requested an abdominal or pelvic radiograph before the stoma was fashioned but we believe that it is far more than the five cases reported. All the children in our series underwent an abdominal radiograph. Suggestive features on radiograph include paucity of gas in the bowel as well as presence of air-fluid levels or calcifications in the mass [2, 4, 9]. Pneumoperitoneum may also be observed [1].

According to the literature review, a contrast enema was the most frequently performed investigation in patients with FERP. On the contrast enema, displacement and narrowing of the rectum can be seen, as well as a possible fistula communicating with the mass [4, 10]. Ultrasound of the perineum and pelvis may show a cystic mass with echogenic foci which displaces the rectum or bladder [2-4, 10].

More invasive imaging techniques such as computerised tomography and magnetic resonance imaging scans were only performed pre-diversion in one of the fourteen cases encountered in the literature. The MRI identified a small locule of air within the extradural spinal canal, however no neuroenteric communication was identified during mass excision [9].

Based on the literature review and on our experience, we believe it is not necessary to perform any investigations

other than a simple abdominal x-ray in the face of the typical clinical features of FERP. Requesting additional investigations may in fact delay the treatment thus allowing progression of the disease with increased morbidity. Furthermore the findings do not alter the next step in management, namely faecal diversion [3, 4].

Once a diagnosis of FERP is deemed likely, timely surgical intervention is indicated. Faecal diversion via descending/sigmoid colostomy is generally accepted as the standard management strategy for FERP. All cases reviewed in the literature as well as all of our patients were managed with a diverting colostomy. The need for a divided colostomy with a distal mucous fistula rather than a loop or Hartman's colostomy is debatable. The spontaneous closure of the FERP in the majority of cases and the low incidence of rectal strictures indicate that a distal mucus fistula is probably not needed as access to the rectum is still possible through a patent anal opening.

While diverting colostomy is undoubtedly part of the management of cases of FERP, differences arise between author recommendations in terms of the need for a formal laparotomy or diagnostic laparoscopy at the time the stoma is fashioned. However, since peritoneal contamination was only found in the cases in which there were clinical or radiological signs of peritonitis, we believe that unless the clinical picture is suggestive of peritoneal involvement, an exploratory laparotomy is not indicated and a left flank incision is sufficient to fashion a colostomy and explore the abdominal cavity.

Drainage of the meconium-filled cavity also needs to be a standard of care in patients with FERP. In fact, the only meconium-filled mass which required surgical excision was the one that was initially treated conservatively and not drained [9].

Formal debridement of the meconium-filled mass is also a matter of debate. On the one hand it is believed that if the mass is properly drained, debridement is unnecessary as meconium is sterile and it is limited to extraperitoneal tissues [3]. Furthermore, avoiding debridement removes the risk of iatrogenic injury to pelvic nerves and surrounding structures [10]. On the other hand, despite being sterile, extraperitoneal meconium may cause an intense inflammatory reaction which can be minimised by debridement of affected tissues [10]. Our recommendation is that perineal debridement should not be performed if the FERP has been recognised and managed with a diverting colostomy and drainage of the meconium-filled cavity before the child is fed and meconium becomes unsterile.

Completion of management involves closure of the colostomy. This should be performed at 3–6 months of age after a distal colostogram has confirmed closure of the fistula and absence of distal obstruction. If obstruction is still present, this should be relieved and any residual fistula repaired. In our experience, we used a Swensen-type pull through with fat pad to close a persistent fistula. A routine rectal biopsy to exclude Hirschsprung disease before stoma closure has not been performed or discussed by any of the authors. In our series we performed a rectal suction biopsy in one of the patients and histological examination of resected bowel in another and both showed normal ganglion cells with absence of hypertrophic nerves. As 'forceful peristalsis against a distal obstruction' is one of the postulated theories for FERP, we believe that Hirschsprung disease should be kept in mind and possibly excluded in all patients before reversal of the stoma.

Conclusion

Fetal extraperitoneal rectal perforation is an extremely rare condition with a typical set of clinical features which allow for early recognition. It is our belief that the only necessary investigation prior to intervention is a standard abdominal radiograph to rule out intraperitoneal involvement. Surgical management should include faecal diversion with drainage of the meconium-filled mass. We believe that exploratory laparotomy/laparoscopy is not necessary unless there are clinical or radiological signs of peritoneal involvement. Perineal debridement is not indicated unless the meconium cyst is contaminated. A distal colostogram at 3–6 months of age confirms spontaneous closure of the fistula in the majority of cases. Only rare cases need a further operation to close the persistent fistula prior to reversal of the stoma. A rectal biopsy should be considered to rule out Hirschsprung disease as a cause of distal obstruction in patients with fetal extraperitoneal rectal perforation.

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