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Meconium Pseudocyst: A Rare Neonatal Surgical Emergency

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Abstract

Meconium peritonitis with formation of pseudocyst is a rare surgical emergency and can be lethal. Meconium peritonitis is defined as a sterile chemical peritonitis or a foreign body peritonitis that is caused be escape of meconium from the gastrointestinal tract into the peritoneal cavity during the fetal or perinatal period. It is an intestinal dilation of meconial content with smooth muscles in the intestinal wall connecting it to the proximal loop causing intestinal obstruction. The incidence is between 1 in 30,000 to 1 in 35,000 live births and the mortality ranges from 11% to 50%. Here, we report a case of meconium peritonitis with pseudocyst in 34weeks gestation born male baby, which was treated successfully with surgical exploration and repair. This baby had an unusual form of meconium peritonitis. Early recognition, pseudocyst resection, decortication, antibiotics, intravenous fluid supplementation and meticulous post-operative care offer the best opportunity for survival. Emergency exploratory laparotomy with excision of meconium pseudocyst with resection and anastomosis of jejuno-ileal junction was performed. Surgical outcome is extremely favourable with early antenatal ultrasonographic diagnosis, closed follow-up, appropriate imaging and timely intervention.

Keywords: Meconium pseudocyst, meconium peritonitis, computed tomography

Introduction

Meconium peritonitis with formation of pseudocyst is a rare surgical emergency and can be lethal. Meconium peritonitis is defined as a sterile chemical peritonitis or a foreign body peritonitis that is caused be escape of meconium from the gastrointestinal tract into the peritoneal cavity during the fetal or perinatal period. It is an intestinal dilation of meconial content with smooth muscles in the intestinal wall connecting it to the proximal loop causing intestinal obstruction. The incidence is between 1 in 30,000 to 1 in 35,000 live births and the mortality ranges from 11% to 50%.[1] The etiology can be due to intestinal perforation with obstruction including stenosis, volvulus, ileal atresia, extrinsic congenital bands, meconium ileus, internal hernias, duplication and Hirschsprung's disease.[2] It can also be due to intestinal perforation without obstruction, such as Meckel's diverticulum, appendicitis or any vascular insufficiency.[2] Colonic atresia and torsion of fallopian tube cyst were reported as rare causes of Meconium peritonitis. It can be classified into three pathological variants: fibrocystic, adhesive and generalised. The cystic type has meconium filled pseudocyst that may rupture in the peritoneum. Cystic fibrosis is reported to cause 25-40% of Meconium peritonitis in the western hemisphere. However, no definite cause is reported in half of the cases. Most of the cases are diagnosed postnatally, although over 20 cases have been diagnosed antenatally by ultrasound, have been reported. The pathognomic presentation is either by X-ray

abdomen or ultrasound which is characterised as Intra-abdominal calcification.[3] Some cases may improve or some cases may resolve in utero. Most literature recommend expectant management unless it is complicated. Some may require a surgical repair. The surgical indications are intestinal obstruction, continuous leakage of meconium into the peritoneal cavity resulting in progressive abdominal distension and respiratory distress or sepsis.[4]

Here, we report a case of meconium peritonitis with pseudocyst in 34weeks gestation born male baby, which was treated successfully with surgical exploration and repair.

Case Capsule

A 34weeks gestation born male baby, with birth weight 2.45kg, born to second para mother with normal vaginal delivery at a primary health care centre, brought by parents, presented to MGM emergency room with complaints of abdominal distension, unable to pass meconium and bilious vomiting since birth. Baby had immediately cried after birth. No other congenital anomaly was found immediately after birth. On examination at the time of presentation the baby was euthermic with vital signs as follows: Heart rate- 156/min, Saturation- 100% on room air, Respiratory rate- 70/min, Capillary refill time less than 3 seconds, all peripheral and central pulsations well felt, Abdominal girth of 30cm. On systemic examination: CNS- good cry, tone and activity, CVS- S1 and S2 heard, RS- Air entry bilaterally equal with no added sounds and mild tachypnea, Per abdomen- Distended and tense with shinny skin and visible dilated and prominent veins over the lower abdomen seen, Bowel sounds were hyperperistaltic. Per rectal examination showed presence of meconium in the rectum and normal anal tone. Blood investigations were Haemoglobin- 14.7 g/dl, Total leucocyte count- 6700 mm^3, Platelet- 2.01 $lacs/mm^3$, Hematocrit- 65.4%, Sodium- 130 mEq/L, Potassium- 4.4 mEq/L, Urea- 32mg/dl, Blood urea nitrogen- 15mg/dl, Creatinine- 1mg/dl, Total bilirubin- 1.2mg/dl, Direct bilirubin- 1.8mg/dl, SGOT/SGPT- 61/10 U/L, Alkaline phosphatase- 566 IU/L, Total protein- 6.1 mg/dl, albumin- 3.1 mg/dl and HIV, Hepatitis B and C viral makers were negative. Antenatal scan showed a 8.5 x 5cm cystic lesion with thin internal echoes within, in the abdomen of fetus. The baby was transferred to Neonatal Intensive Care Unit and patient was kept nil by mouth, nasogastric tube decompression, intravenous fluid and antibiotic treatment was given. Abdominal plain X-ray showed relatively a ground glass appearance with few areas of intestinal ileus. The baby had undergone plain Computed tomography (CT) of abdomen revealed a thin walled cystic lesion with air and fluid content in the anterior abdomen displacing the bowel loops posteriorly and the lesion was found to be indenting the inferior margin of right lobe of liver superiorly. It also showed moderate free fluid in perihepatic, perisplenic and interbowel pelvis region. After an appropriate diagnosis of suspected meconium pseudocyst complicated with intestinal obstruction and respiratory distress syndrome, emergency exploratory laparotomy with excision of meconium pseudocyst with resection and anastomosis of jejuno-ileal junction was performed. Intraoperatively there was a presence of meconium pseudocyst occupying the whole abdomen measuring approximately 10 x 5 x 4 cm. Excision of meconium pseudocyst done. There was a presence of jejuno-ileal perforation antenatally. Edges of jejunum and ileum were excised and anastomosis was done. Warm normal saline wash given. Corrugated rubber drain was placed in Morrison's pouch. Immediate post-operative recovery was uneventful. Post-operatively baby was shifted to Neonatal Intensive Care Unit and was kept nil by mouth with Ryle's tube decompression along with temperature recording, abdominal girth charting four hourly and monitoring of vitals done. Intravenous fluid and intravenous antibiotics were administered. Daily Ryle's tube aspirate was around 40 to 50 ml. Flatus and meconium passed on post-operative day 3. On post-operative day 10 feeding with 15ml regular formula every 4 hourly initiated and was tolerated well. The feeding was then escalated gradually up to 60ml every 4 hours on post-operative day 12. Corrugated rubber drain was removed on post-operative day 12. As a result of adequate bowel movements, the baby was discharged on post-operative day 14 after suture removal and was assessed on regular OPD follow ups.

Discussion

Meconium peritonitis was first described by Simpson in 1838.[5] Morgagni described meconium peritonitis in 1761.[5] Since then, few retrospective studies have been published to understand the natural course of

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the disease and the possible etiology of this condition. It presents in early neonatal period with vomiting, abdominal distension, and intra-abdominal calcifications. The first case diagnosed antenatally were incidental findings in women who had pelvimetry X-ray for suspected fetal macrostomia and congenital malformations.[6-7] Antenatal ultrasonographic diagnosis of meconium peritonitis was first described by Brugman and colleagues.[8]

The course of the disease ranges from spontaneous healing to rapid fatality, depending on the timing of perforation and if the perforation persists even after birth. Meconium peritonitis is a sterile chemical peritonitis induced by meconium extruding into the peritoneal cavity through an intestinal perforation, most commonly the ileum, proximal to the site of obstruction and causing irritation of the peritoneal cavity. The obstruction may be caused by stenosis, atresia, intussusception, volvulus, meconium ileus, Meckel's diverticulum, peritoneal bands. In majority of cases, the perforation is closed by the time of birth. Mortality in cases which the perforation persists until the time of birth is relatively high. However, postnatal surgical intervention to relieve the intestinal obstruction or close the perforation and restore the continuity of the intestine is usually necessary.

Since the 1980's, antenatal diagnosis by maternal ultrasonography has added to the diagnostic yield.[9-11] Antenatal diagnosis is believed to reduce the mortality rate from 50% to 11% and predict postnatal surgery in 50% of the cases with meconium pseudocyst.[12] A fetal intraperitoneal cystic lesion that shows high T1 and low T2 signals in prenatal magnetic resonance imaging is considered pathognomonic of meconium pseudocyst.[13] In our case report, the ultrasonographic findings, postnatal plain radiography, ultrasonography and plain computed tomography of abdomen, all suggested possibility of meconium pseudocyst. Embryologically, formation of meconium begins in 3rd month of gestation, intestinal peristalsis begins in 5th month of gestation and the meconium fills the entire intestine.[5]

Meconium peritonitis was noted to be higher in infants of Chinese and Malay and Indian origin. Shyu et al. and Kamala et al. reported higher incidence of meconium peritonitis in males.[14-15] In 1943, Agerty and associates were the first to recoed survival of a patient with meconium peritonitis treated surgically.[2] In 1966, Lormier and Ellis reported post-operative survival in five patients with meconium peritonitis, and tried to summarize a concept of surgical repair based on the pathologic classification.[1] Bergmans and colleagues quoted a perinatal mortality of 62 to 85% in cases of meconium peritonitis.[16] The outlook is better in cases that are diagnosed antenatally. Since then several reports of survival after surgical repair for meconium peritonitis have been published.[17-19] The main purpose of surgery is to establish intestinal continuity and to preserve at least 50% of the intestinal length along with correction of the underlying pathologic processes.

Intrauterine infections, especially parvovirus B19, have been reported to be associated with meconium peritonitis.[20-21] It was postulated that parvovirus B19 infection induced vasculitis, resulting in mesenteric vascular insufficiency, causing intestinal perforation without any antecedent intestinal obstruction. In general meconium peritonitis can be classified into three pathological variants: cystic, fibro-adhesive and generalised. In the fibro-adhesive variant, the perforation seals off before birth due to fibroblastic reaction. Subsequent intestinal obstruction is caused by adhesions. Surgical repair aims to achieve adhesiolysis and resection of non-viable intestinal segment. In cystic type, the intestinal perforation remains open. By opening the cyst and finding the perforation, the cause of the intestinal obstruction can be determined. After an adequate length of intestine is freed, the non-viable or severe entrapped can be resected, followed by exteiorization of intestine. In the generalised type, the perforation occurs in the perinatal period and results in wide dissemination of meconium in the peritoneal cavity.

Mikulicz enterostomy with early crushing of the spur appears to be the best means for re-establishing intestinal continuity. Some authors have reported successful primary anastomosis after resection of bowel in patients with cystic meconium peritonitis.[22-24] Tanaka et al recommended drainage of cyst after birth and elective surgical repair at a later stage.[25]

This baby had an unusual form of meconium peritonitis. Early recognition, pseudocyst resection, decortication, antibiotics, intravenous fluid supplementation and meticulous post-operative care offer the best opportunity for survival. In our case,

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we performed an Emergency exploratory laparotomy with excision of meconium pseudocyst with resection and anastomosis of jejuno-ileal junction. In conclusion, successful surgical treatment was done in our case and was attributed to antenatal ultrasonographic diagnosis, antenatal counselling, prophylactic antibiotics, intravenous fluid supplementation, postnatal fluid and electrolyte correction, postnatal computed tomography demonstrated meconium pseudocyst.

Conclusion

This case report demonstrates a rare incidence. Surgical outcome is extremely favourable with early antenatal ultrasonographic diagnosis, close follow-up, appropriate imaging, and timely intervention. Neonatologist and Paediatric surgeons dealing with meconium peritonitis can use the information obtained in this study for counselling parents, as the outcome is excellent if managed appropriately.

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FIGURE 1- X ray abdomen suggestive of ground glass appearance with intestinal ileus

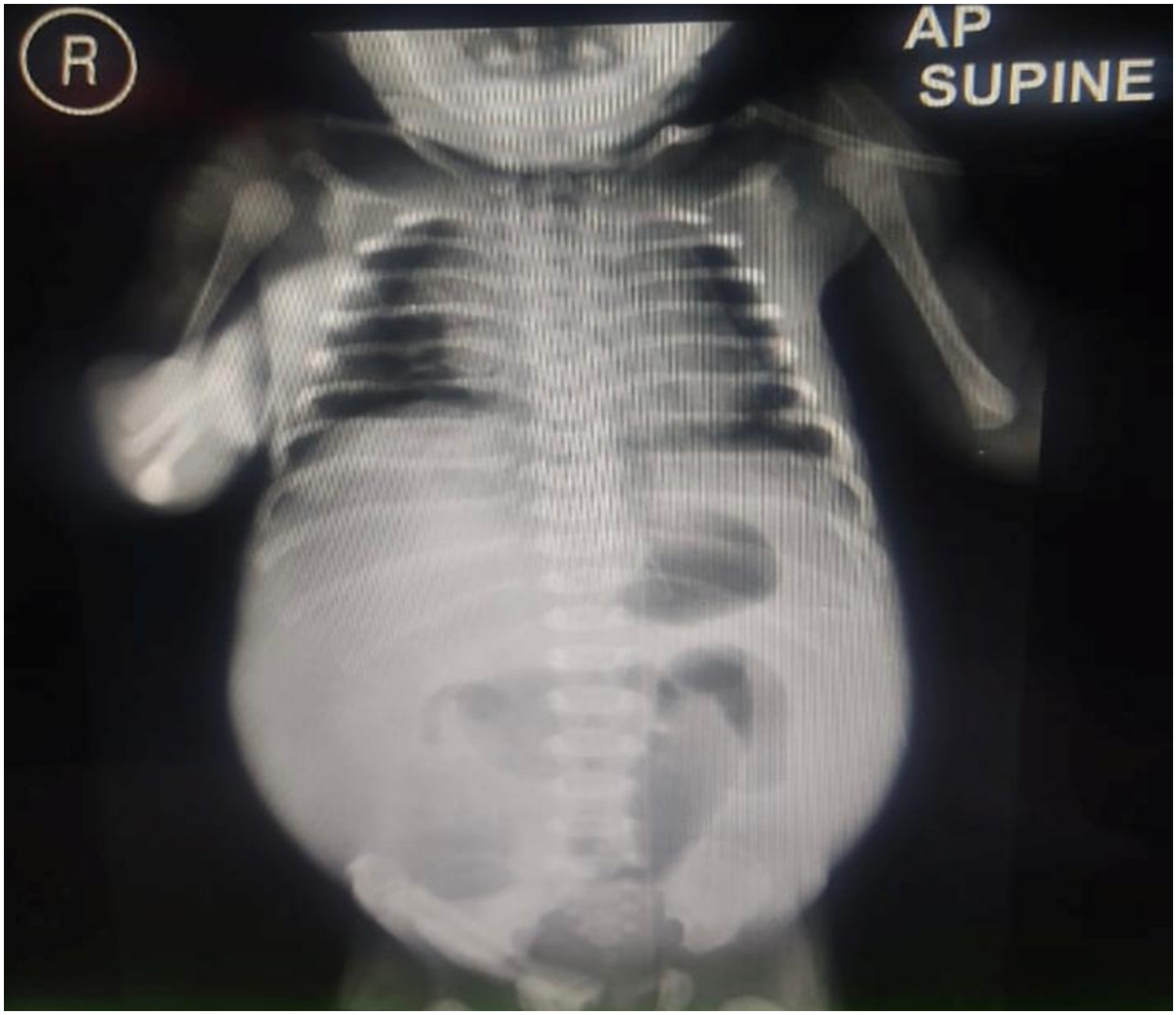

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FIGURE 2a- Saggital CT image reveals a large cystic lesion measuring 6.7cm craniocaudally with air fluid level within, posterior displacement of bowel loops noted

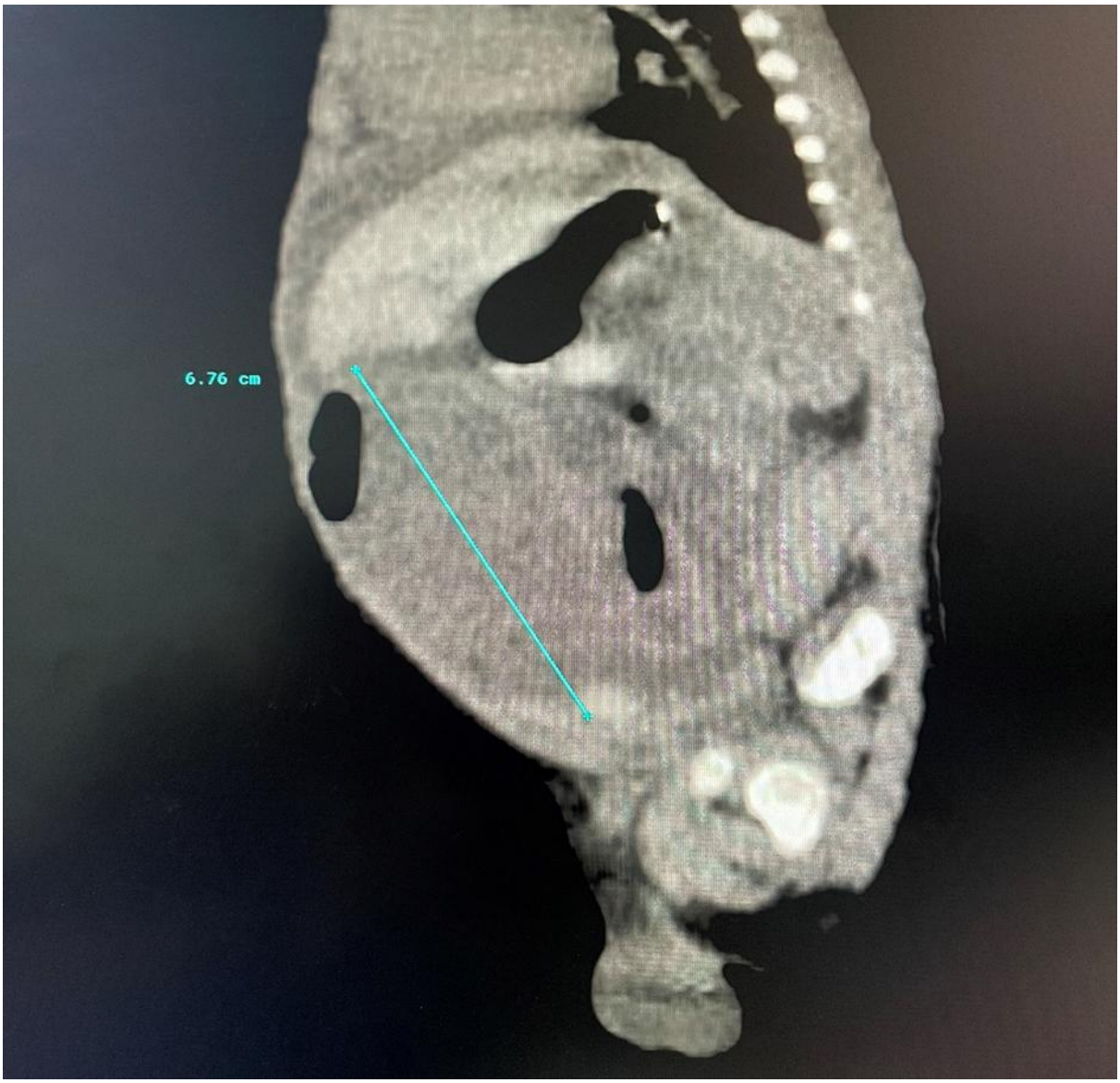

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FIGURE 2b- Axial CT image reveals a large well defined thin walled cystic lesion in the anterior abdomen measuring 11.7x4.2cm (Transverse x Anteroposterior)

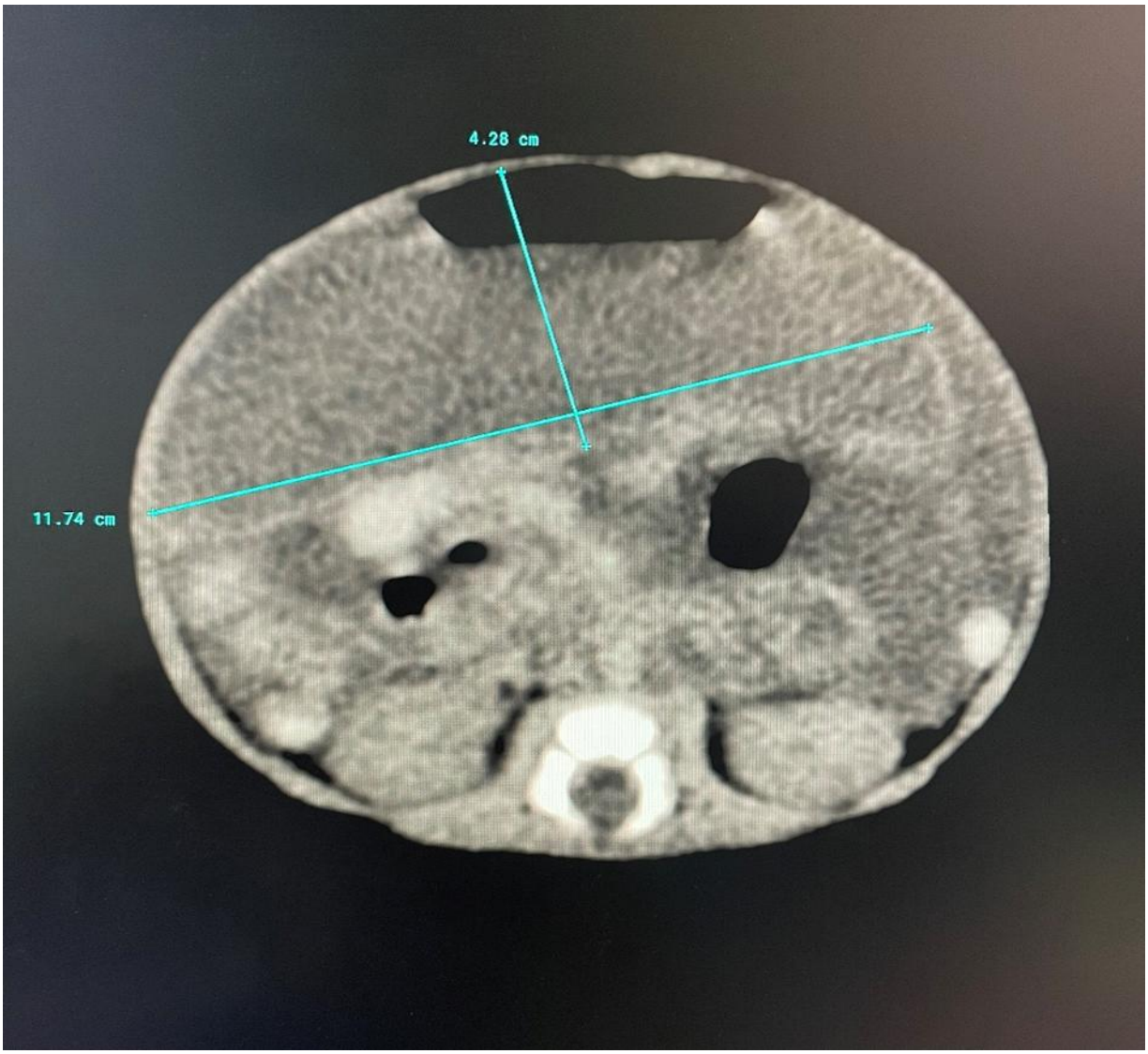

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FIGURE 3- Preoperative image: Distended abdomen with dilated and prominent veins

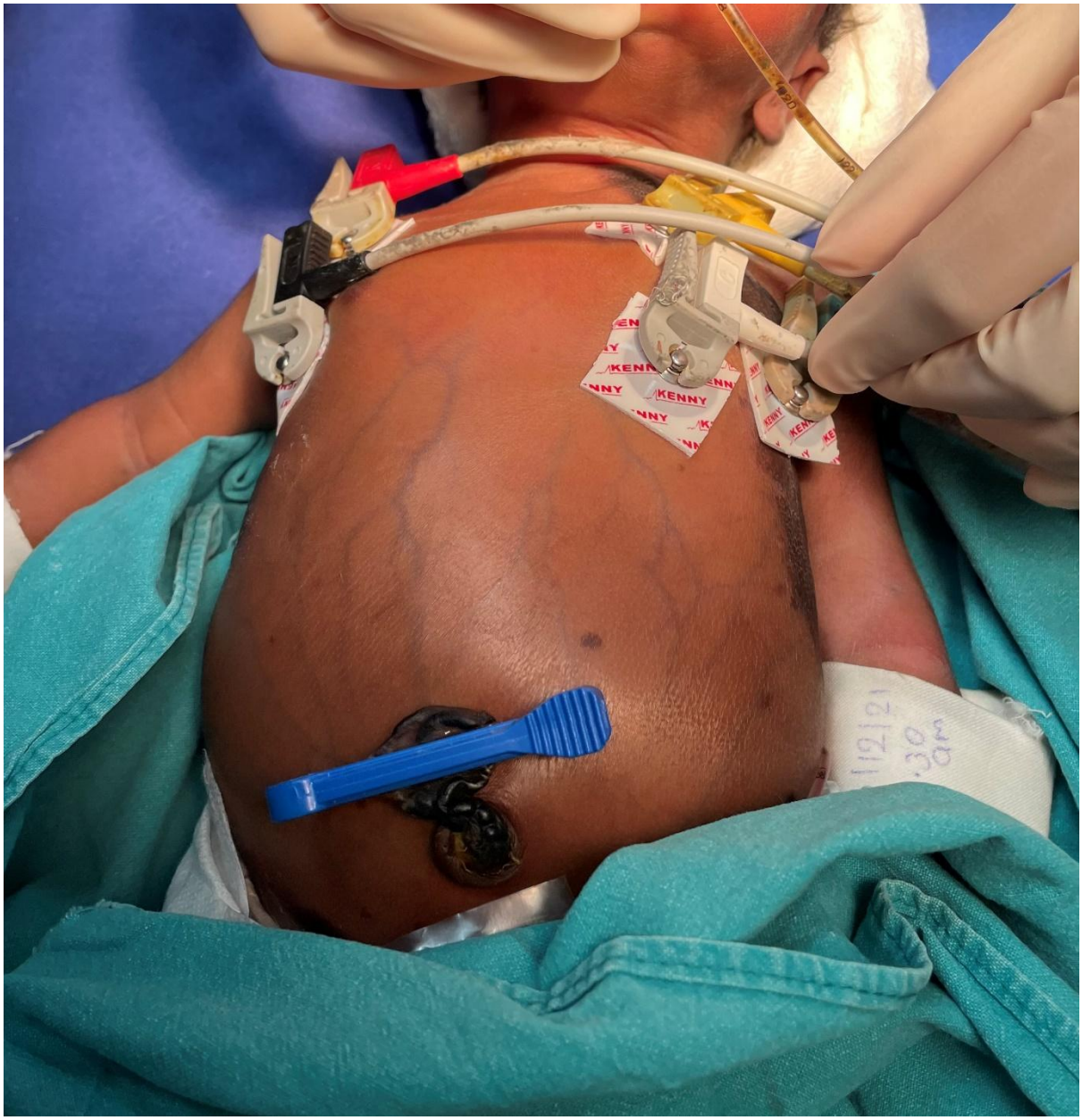

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FIGURE 4- Intraoperatively, anterior part of cyst wall with drainage of meconium

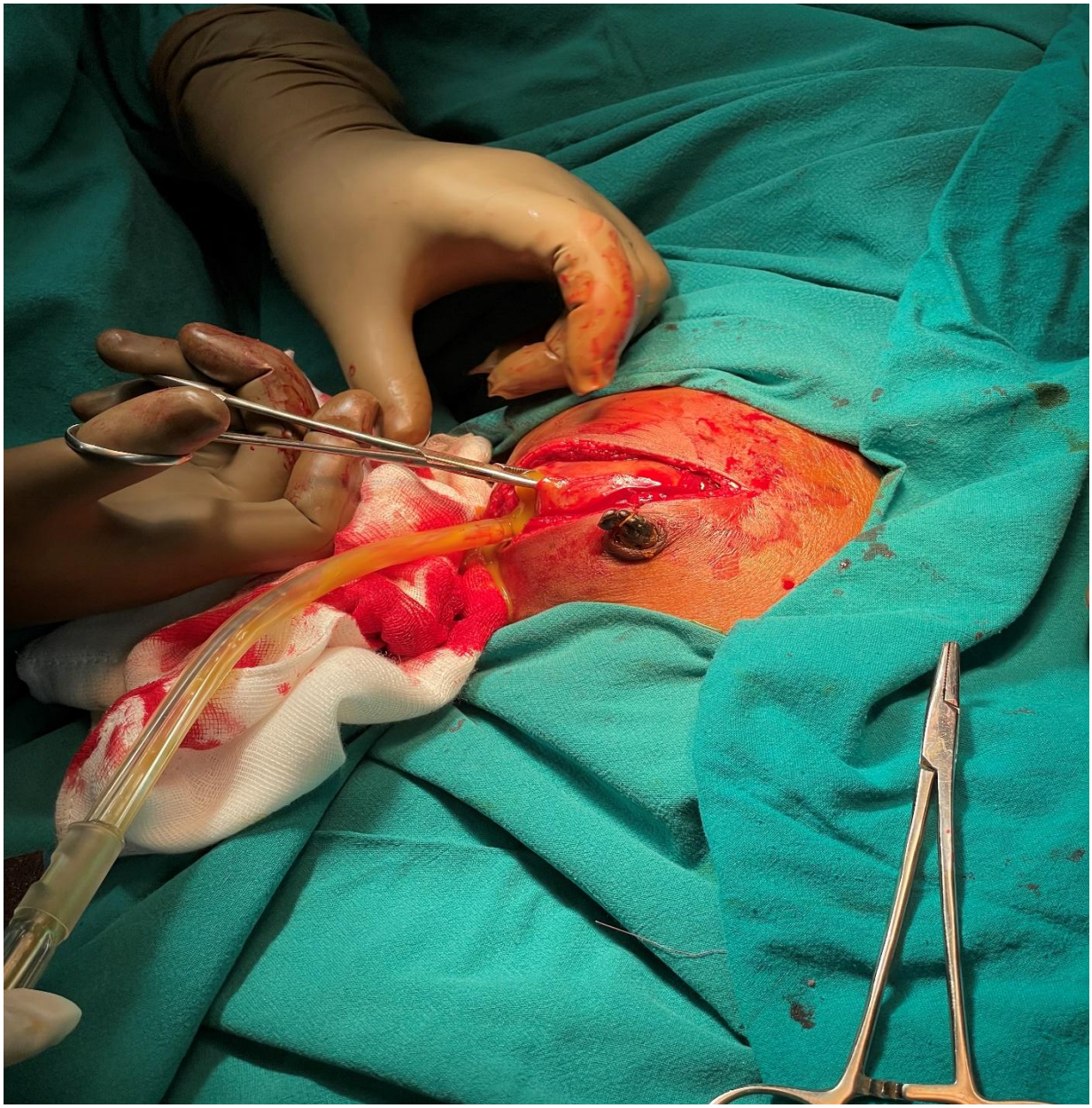

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FIGURE 5- Excised part of pseudocyst wall with proximal end of bowel (Jejunum) opening

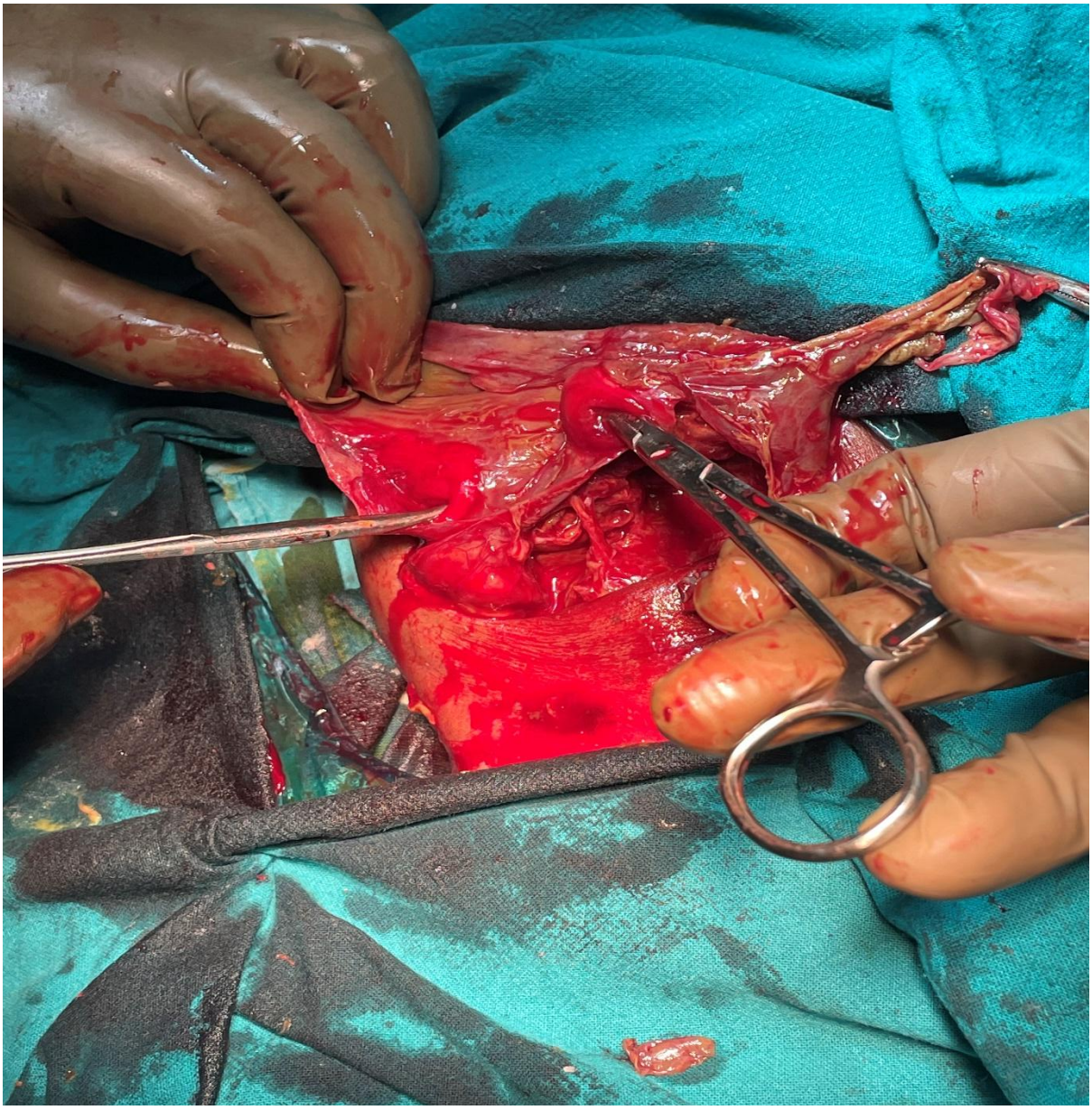

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FIGURE 6- Jejuno-ileal ends due to antenatal bowel perforation

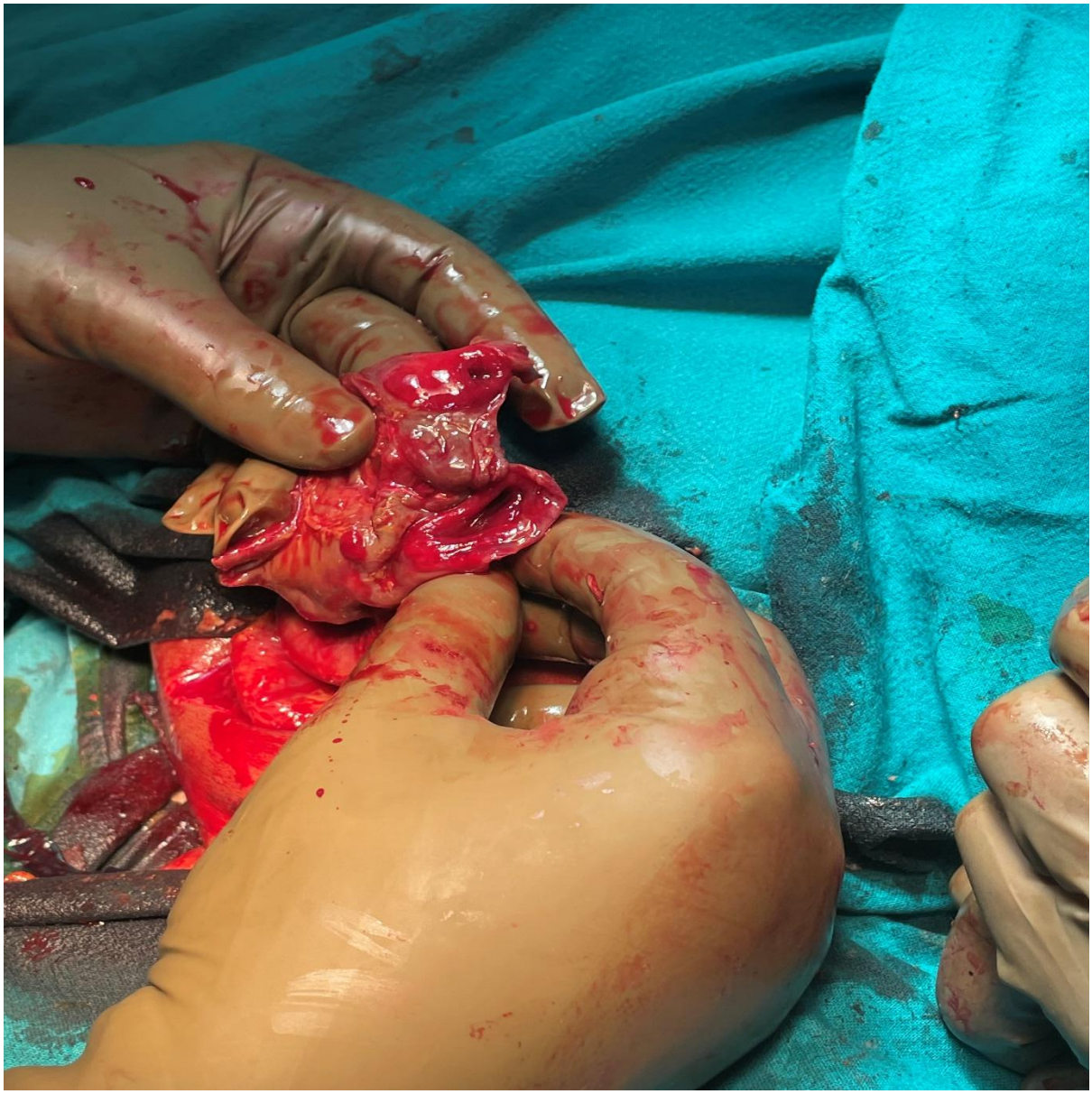

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FIGURE 7- Jejuno-ileal anastomosis done

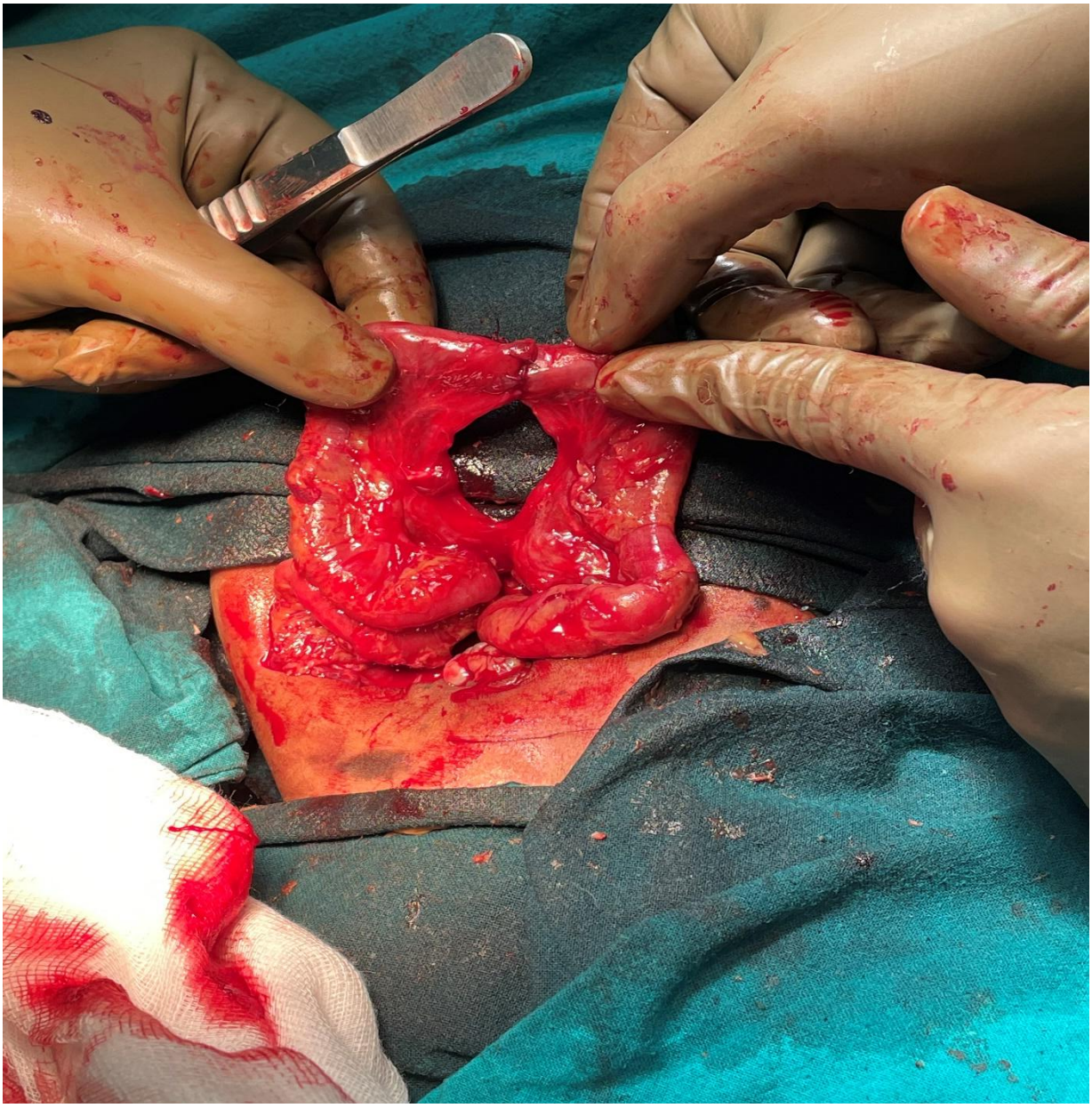

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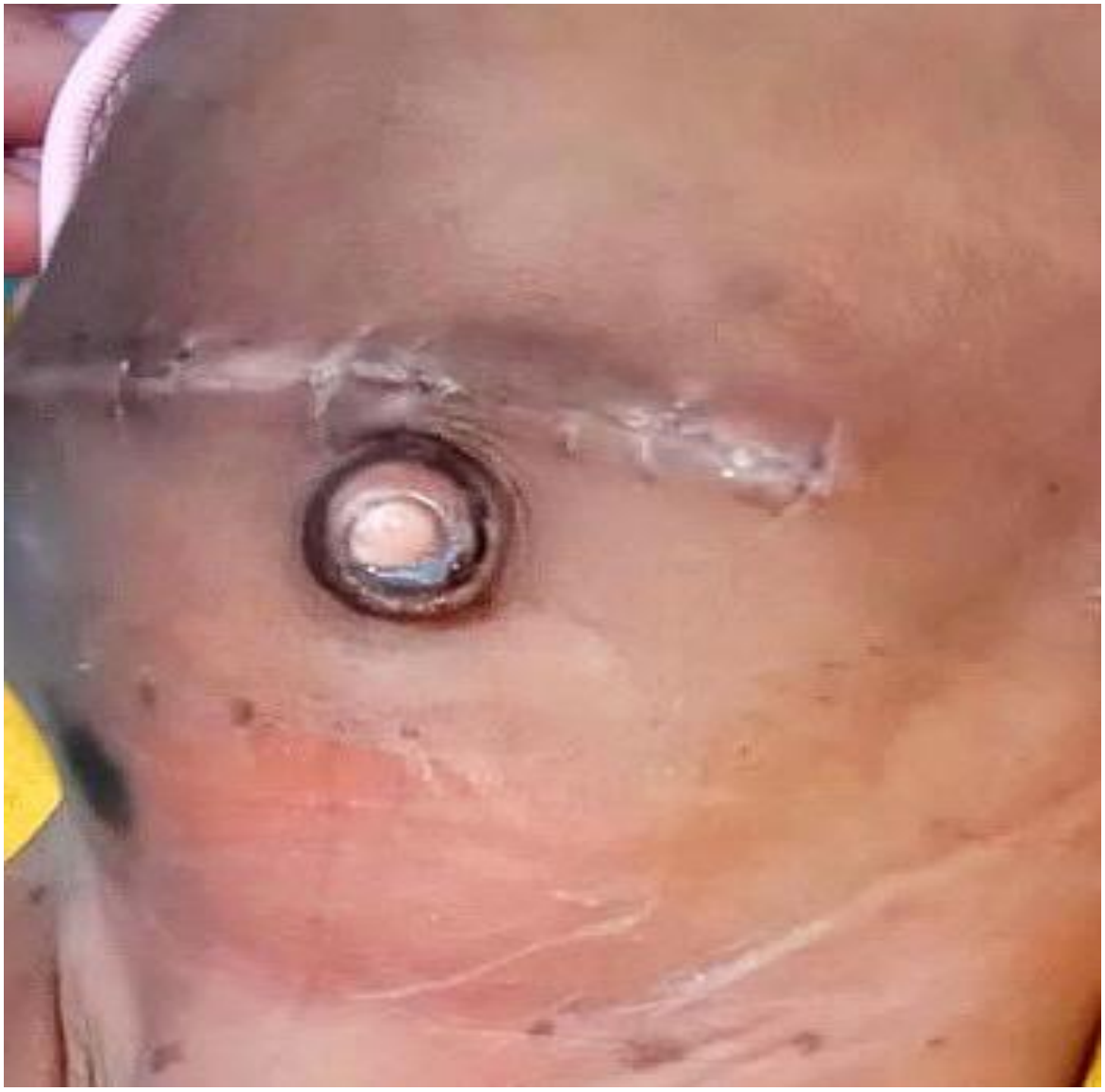
FIGURE 8- Post-operative healed scar



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