



## CASE REPORT

# Intrauterine midgut volvulus without malrotation: Diagnosis from the 'coffee bean sign'

Jun Seok Park, Seong Jae Cha, Beom Gyu Kim, Yong Seok Kim, Yoo Shin Choi, In Taik Chang, Gwang Jun Kim, Woo Seok Lee, Gi Hyeon Kim

Jun Seok Park, Seong Jae Cha, Beom Gyu Kim, Yong Seok Kim, Yoo Shin Choi, In Taik Chang, Department of Surgery, College of Medicine, Chung-Ang University, Seoul 156-755, Korea

Gwang Jun Kim, Woo Seok Lee, Department of Obstetrics and Gynecology, College of Medicine, Chung-Ang University, Korea  
Gi Hyeon Kim, Department of Radiology, College of Medicine, Chung-Ang University, Seoul 156-755, Korea

Correspondence to: Seong Jae Cha, MD, Professor, Department of Surgery, College of Medicine, Chung-Ang University, 224-1 Heukseok-Dong, Dongjak-Gu, Seoul 156-755, Korea. [jwas@naver.com](mailto:jwas@naver.com)

Telephone: +82-2-62991548 Fax: +82-2-8247869

Received: October 30, 2007 Revised: December 9, 2007

## Abstract

Fetal midgut volvulus is quite rare, and most cases are associated with abnormalities of intestinal rotation or fixation. We report a case of midgut volvulus without malrotation, associated with a meconium pellet, during the gestation period. This 2.79 kg, 33-wk infant was born *via* a spontaneous vaginal delivery caused by preterm labor. Prenatal ultrasound showed dilated bowel loops with the appearance of a 'coffee bean sign'. This patient had an unusual presentation with a distended abdomen showing skin discoloration. An emergency laparotomy revealed a midgut volvulus and a twisted small bowel, caused by complicated meconium ileus. Such nonspecific prenatal radiological signs and a low index of suspicion of a volvulus during gestation might delay appropriate surgical management and result in ischemic necrosis of the bowel. Preterm labor, specific prenatal sonographic findings (for example, the coffee bean sign) and bluish discoloration of the abdominal wall could suggest intrauterine midgut volvulus requiring prompt surgical intervention.

© 2008 WJG. All rights reserved.

**Key words:** Midgut volvulus; Coffee bean sign; Meconium ileus

**Peer reviewers:** Damian Casadesus Rodriguez, MD, PhD, Calixto Garcia University Hospital, J and University, Vedado, Havana City, Cuba; Takayuki Yamamoto, MD, Inflammatory Bowel Disease Center, Yokkaichi Social Insurance Hospital, 10-8 Hazuyamacho, Yokkaichi 510-0016, Japan; Luigi Bonavina, Professor, Department of Surgery, Policlinico San Donato, University of Milano, via Morandi 30, Milano 20097, Italy

Park JS, Cha SJ, Kim BG, Kim YS, Choi YS, Chang IT, Kim GJ, Lee WS, Kim GH. Intrauterine midgut volvulus without malrotation: Diagnosis from the 'coffee bean sign'. *World J Gastroenterol* 2008; 14(9): 1456-1458 Available from: URL: <http://www.wjgnet.com/1007-9327/14/1456.asp> DOI: <http://dx.doi.org/10.3748/wjg.14.1456>

## INTRODUCTION

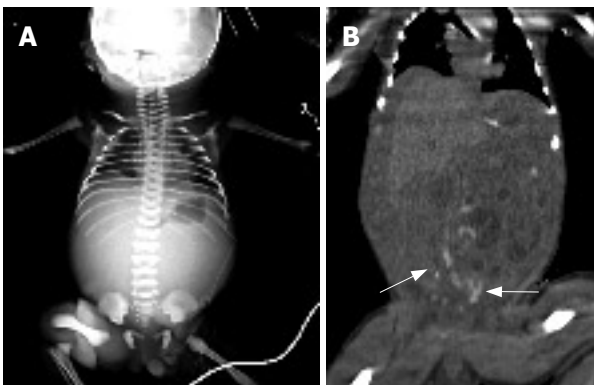
Midgut volvulus is a condition in which the small bowel or proximal colon twists around the superior mesenteric artery. This condition most commonly presents during the first year of life and has high rates of morbidity and mortality<sup>[1,2]</sup>. Midgut volvulus without malrotation is an extremely rare surgical condition, which may also occur during gestation<sup>[3-6]</sup>. We recently encountered an unusual case in which intrauterine volvulus occurred with prenatal meconium pellets. Emergent prenatal ultrasonography revealed the presence of the 'coffee bean sign' in this fetus. The patient required resection of a significant amount of necrotic small bowel and treatment with intra-operative saline irrigation. The patient also needed postoperative gastrografin enema due to persistent meconium ileus. Fortunately, the patient survived and has continued to thrive without parenteral nutrition. We discuss the pathogenesis of intrauterine midgut volvulus associated with complicated meconium ileus and the issues surrounding their emergent diagnosis and management.

## CASE REPORT

A 27-year-old pregnant woman was referred to our clinic at 33 wk of gestation because of fetal intestinal dilatation found on sonography, and the onset of preterm labor. Routine examinations at a local clinic had revealed a dilated intestine in the fetus, but this had improved spontaneously four weeks earlier. Transabdominal ultrasound on referral showed a segment of markedly dilated fetal intestine, which suggested a closed loop obstruction (Figure 1). Fetal ultrasound measurements were appropriate for the gestational age, and the Doppler indices were normal. No ascites was seen in the fetal abdomen. As it was possible the bowel obstruction would be complicated by intestinal necrosis, preterm delivery was considered beneficial. At 33 wk and two days, a male infant was delivered transvaginally. The patient weighed 2690 g and had Apgar scores of 8 and 9 at 1 and 5 min, respectively.



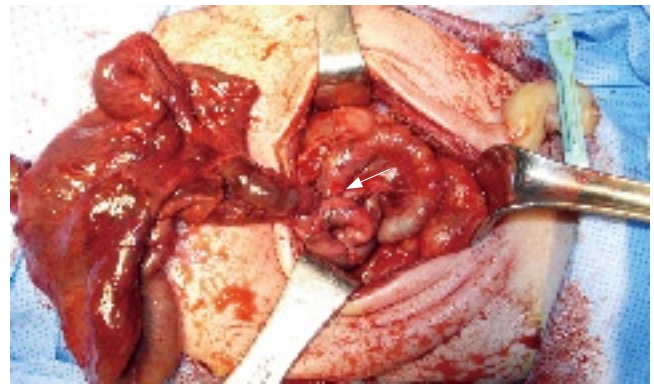
**Figure 1** A fetal sonogram showing dilated bowel loops with the appearance of a 'coffee bean sign'. No ascites was seen in the fetal abdomen.



**Figure 2** **A:** Pre-operative infantogram showing a gas shadow only in the stomach, with an absence of any distal gas shadow; **B:** Unenhanced abdominal CT showed meconium (arrow) in the distal small bowel, with mild fluid distension of the proximal small bowel.

The abdomen was distended markedly and there was a bluish skin discoloration on the periumbilical abdominal skin. Nasogastric aspiration recovered 10 mL of bilious material. A rectal examination was normal. Initial laboratory values included a white blood cell count of 26 000/mm<sup>3</sup>, a platelet count of 416 000/mm<sup>3</sup> and prothrombin time of 11 s. Blood gas values from the umbilical artery were pH 7.14, PO<sub>2</sub> 51.3 mmHg and PCO<sub>2</sub> 60.44 mmHg. A plain supine abdominal radiograph did not demonstrate bowel gas, except in the stomach. An emergency computer tomography (CT) scan performed without contrast media revealed marked intestinal dilatation mainly in the left abdomen, and a large amount of hemorrhagic ascites (Figure 2).

Exploration revealed a volvulus of the small bowel with extensive necrosis extending from 40 cm distal to the ligament of Treitz to 15 cm proximal to the ileocecal valve. The distended segment of intestine was twisted at the level of the narrow meconium filled distal ileum (Figure 3). There was no intestinal malrotation, mesenteric defect or atresia. After detorsion of the midgut volvulus, the thick meconium of distal ileum was irrigated by instilling saline with an 8-Fr rubber catheter. The involved loop was then resected and an end-to-oblique anastomosis was constructed between the dilated proximal and smaller



**Figure 3** On laparotomy, the infant was found to have a midgut volvulus with necrosis and perforation of the small bowel. The small bowel was found to be twisted at the level of the narrow meconium-filled distal ileum (arrow).

distal bowels by manual anastomosis. On the fourth day after surgery, the patient required treatment with a gastrografin enema to loosen the persisting meconium ileus. A presumptive diagnosis of cystic fibrosis was made postoperatively, based on meconium ileus. There was no family history. To date, the baby has been screened, but all results have been negative. The infant made a remarkable recovery and was able to tolerate enteral feeding with a steady weight gain by four months after surgery.

## DISCUSSION

Midgut volvulus is a surgical emergency frequently encountered in neonates. Most cases of volvulus in infants and fetus are associated with intestinal malrotation or congenital anomalies, such as omphalocele, gastroschisis, intestinal atresia or an annular pancreas<sup>[7]</sup>. On the other hand, the etiology of volvulus without malrotation is unknown, and associated anomalies are rare<sup>[5]</sup>. Several studies have shown that the absence of a segment of small bowel muscle or a mesenteric defect might be associated with this condition<sup>[8-11]</sup>. There is no clear explanation for the cause of this event in our patient, because laparotomy did not reveal any intestinal malrotation or congenital mesenteric anomalies. However, we suggest the fetal midgut volvulus and preterm labor was caused by the complicated meconium ileus. Intestinal volvulus might occur when the distended segment of the small bowel becomes twisted at the level of the narrow pellet-filled distal ileum. Gestational volvulus can result in ischemic necrosis, leading to fetal stress, which might activate the release of both adrenal and hypothalamic stress hormones. These might enhance placental, decidual and amniochorionic corticotrophin-releasing hormone release, while premature rupture of fetal membranes and preterm labor can be mediated by placental and membrane prostanoid release<sup>[12,13]</sup>.

Previous reports have described cases with midgut volvulus in which the fetal sonograms showed intestinal dilatation, a discrete cystic or solid abdominal mass, ascites, peritoneal calcification, polyhydramnios and, typically, the whirlpool or snail sign<sup>[14,15]</sup>. Unfortunately, our patient did not show any definitive sonographic sign of midgut

volvulus. Instead, retrospective analysis of the patient's prenatal sonographic imaging revealed a coffee bean sign, which is a specific indicator of sigmoid volvulus in adult patients. Attention should always be paid to the risk of a midgut volvulus with such prenatal sonographic findings.

The nonenhanced CT scan performed after spontaneous vaginal delivery showed excessive hemorrhagic ascites, which was invisible during prenatal sonography. This suggests the bowel necrosis and meconium peritonitis may have developed rapidly during the spontaneous vaginal delivery. Rapid emergency Cesarean section or accelerated delivery should be considered for such expectant mothers who have a history of recurrent closed loop obstruction in the fetus, and who present with acute preterm labor, because the symptoms might occur perinatally with rapid progression to gangrene.

Delays in diagnosis are likely in such cases, as physicians tend to doubt or not suspect the possibility of intrauterine volvulus because it is so rare. Therefore, close prenatal monitoring is necessary if there is any suspicion of typical sonographic signs in the fetus. The adoption of more prompt delivery methods with exploration, avoidance of unnecessary special studies and appropriate postnatal intervention are all essential to reduce the likely morbidity and mortality of intrauterine volvulus associated with complicated meconium ileus.

## REFERENCES

- 1 **Torres AM**, Ziegler MM. Malrotation of the intestine. *World J Surg* 1993; **17**: 326-331
- 2 **Andrassy RJ**, Mahour GH. Malrotation of the midgut in infants and children: a 25-year review. *Arch Surg* 1981; **116**: 158-160
- 3 **Pellerin D**, Bertin P. Primary postnatal volvulus of the small intestine. *Ann Chir Infant* 1972; **13**: 83-94
- 4 **Yadav K**, Nayar PM, Patel RV, Das GC. Volvulus neonatorum without malrotation. *J Indian Med Assoc* 1987; **85**: 16-19
- 5 **Usmani SS**, Kenigsberg K. Intrauterine volvulus without malrotation. *J Pediatr Surg* 1991; **26**: 1409-1410
- 6 **De Felice C**, Massafra C, Centini G, Di Maggio G, Tota G, Bracci R. Relationship between intrauterine midgut volvulus without malrotation and preterm delivery. *Acta Obstet Gynecol Scand* 1997; **76**: 386
- 7 **Crisera CA**, Ginsburg HB, Gittes GK. Fetal midgut volvulus presenting at term. *J Pediatr Surg* 1999; **34**: 1280-1281
- 8 **Molvarec A**, Babinszki A, Kovacs K, Toth F, Szalay J. Intrauterine intestinal obstruction due to fetal midgut volvulus: a report of two cases. *Fetal Diagn Ther* 2007; **22**: 38-40
- 9 **Black PR**, Mueller D, Crow J, Morris RC, Husain AN. Mesenteric defects as a cause of intestinal volvulus without malrotation and as the possible primary etiology of intestinal atresia. *J Pediatr Surg* 1994; **29**: 1339-1343
- 10 **Morikawa N**, Namba S, Fujii Y, Sato Y, Fukuba K. Intrauterine volvulus without malrotation associated with segmental absence of small intestinal musculature. *J Pediatr Surg* 1999; **34**: 1549-1551
- 11 **Cascio S**, Tien AS, Agarwal P, Tan HL. Dorsal mesenteric agenesis without small bowel atresia: a rare cause of midgut volvulus in children. *J Pediatr Surg* 2006; **41**: E5-E7
- 12 **McLean M**, Bisits A, Davies J, Woods R, Lowry P, Smith R. A placental clock controlling the length of human pregnancy. *Nat Med* 1995; **1**: 460-463
- 13 **Wolfe CD**, Patel SP, Linton EA, Campbell EA, Anderson J, Dornhorst A, Lowry PJ, Jones MT. Plasma corticotrophin-releasing factor (CRF) in abnormal pregnancy. *Br J Obstet Gynaecol* 1988; **95**: 1003-1006
- 14 **Baxi LV**, Yeh MN, Blanc WA, Schullinger JN. Antepartum diagnosis and management of in utero intestinal volvulus with perforation. *N Engl J Med* 1983; **308**: 1519-1521
- 15 **Mercado MG**, Bulas DI, Chandra R. Prenatal diagnosis and management of congenital volvulus. *Pediatr Radiol* 1993; **23**: 601-602

S- Editor Zhu LH L- Editor McGowan D E- Editor Liu Y