Received: 22 January 2018 | Revised: 6 August 2018 | Accepted: 13 November 2018

DOI: 10.1002/pd.5392

ORIGINAL ARTICLE

WILEY PRENATAL DIAGNOSIS

Prenatal diagnosis of antenatal midgut volvulus: Specific ultrasound features

Caroline Bartholmot MD[1] | Jean-Michel Faure MD[1] | Frederic Grosjean MD[2] | Alain Couture MD[3] | Dominique Forgues MD[4] | Florent Fuchs MD, PhD[1,5] | Olivier Prodhomme MD, MSc[3]

[1] Department of Obstetrics and Gynecology, CHU Montpellier, Montpellier, France

[2] Department of Obstetrics and Gynecology, Nimes University Hospital Center, Nîmes, France

[3] Department of Pediatric Imaging, CHU Montpellier, Montpellier, France

[4] Department of Abdominal and Urologic Surgery, CHU Montpellier, Hôpital Lapeyronie, Montpellier, France

[5] Inserm, CESP Centre for Research in Epidemiology and Population Health, U1018, Reproduction and Child Development, Villejuif, France

Correspondence
Dr Florent Fuchs, Department of Obstetrics and Gynecology, CHU de Montpellier, Hopital Arnaud de Villeneuve, 371, av. du Doyen Gaston Giraud, 34295 Montpellier Cedex 5, France.
Email: f-fuchs@chu-montpellier.fr

Abstract

Objective: To assess specific, direct, and indirect prenatal ultrasound features in cases of fetal midgut volvulus.

Methods: Retrospective case series of neonatal volvulus, based on postnatal and prenatal imaging findings that occurred from 2006–2017. Prenatal and postnatal signs including the specific "whirlpool sign" were computed. Postnatal volvulus was confirmed by pathology examination after surgery or neonatal autopsy.

Results: Thirteen cases of midgut volvulus were identified. Though not a specific sign, a decrease in active fetal movements was reported in eight patients (61.5%). The prenatal whirlpool sign was directly seen in 10 cases, while an indirect but suggestive sign, a fluid-filled level within the dilated loops, was present in five cases. No intestinal malrotation was observed. Pregnancy outcomes were two terminations of pregnancy, both associated with cystic fibrosis, one early neonatal death, three prenatal spontaneous regressions, and seven favorable outcomes after neonatal surgery with resection of midgut atresia.

Conclusions: Identification of the whirlpool sign or of a fluid-filled level within the dilated loops improves the accuracy of ultrasound findings for suspected volvulus. In the absence of total volvulus (in cases of intestinal malrotation) or association with cystic fibrosis, the prognosis appears good.

1 | INTRODUCTION

Intestinal or midgut volvulus is a rare condition in which intestinal loops are twisted around the axis of the pedicle of the superior mesenteric artery. It is a surgical emergency in the postnatal period, with the child's survival and functional prognosis dependent on the delay until treatment.[1] The most frequent cause of postnatal midgut volvulus is intestinal malrotation with total volvulus of the small bowel.[2] Other causes have also been reported, including cystic fibrosis, gastrointestinal duplication, and the presence of either a cyst or a tumor mass. The principal postnatal ultrasound sign is the "whirlpool sign,"[3] also called the whirl sign, which represents the intestinal loops winding around the superior mesenteric artery.

Prenatal diagnosis of this type of malformation can be difficult in the absence of specific signs.[4] That is, a prenatal whirlpool sign is only sometimes visible, and no other sign has been described in the literature[5] to date.

The aim of this work is to describe and specify the direct and indirect ultrasound signs of prenatal diagnosis of fetal midgut volvulus based on ultrasound data of the largest retrospective case series thus far published.

2 | MATERIAL AND METHODS

This study received approval by our local ethics research committee under the notification number 2017_CLER-MTP_10–04.

The first two authors contributed equally to this work.
The last two authors contributed equally to this work.

This retrospective descriptive study reviewed all cases of fetal midgut volvulus diagnosed between 2006 and 2017 at the multidisciplinary prenatal diagnosis centers of the university hospital centers of Montpellier and Nîmes, in France.

The study includes all cases in which midgut volvulus was prenatally suspected after ultrasound. All ultrasound scans were performed with a GE Voluson E8-expert apparatus (GE Medical Systems®, Milwaukee, WI, USA) equipped with a with real-time 4D abdominal transducer supporting a bandwidth of 2–8 MHz (RAB 4–8-D). The examination was performed transabdominally in two-dimensional (2D, also known as B) mode and color Doppler mode. Additionally, three-dimensional ultrasound was used to assess the relationship between the midgut volvulus and other abdominal organs by performing multiplanar reconstruction of the abdominal cavity.

The ultrasound diagnosis of midgut volvulus was defined by the presence of direct or indirect signs of volvulus. The direct sign was considered to be the whirlpool sign, which depicts loops of the small intestine more or less dilated, winding in a spiral form in 2D mode and corresponding in color Doppler to a particular coiled configuration of the superior mesenteric vessels, with the vein winding around an axis centered on the artery. Indirect signs included the presence of a differential fluid-filled level in a dilated intestinal loop or voluminous intestinal dilatation, polyhydramnios, a meconium pseudocyst or ascites, or signs of meconium peritonitis with gastrointestinal calcifications. Other ultrasound features that we looked for were the relative position of the mesenteric vessels, hyperechoic intestines, and ability to visualize the gall bladder. In addition to US/Doppler studies, mothers were offered to undergo a complementary fetal magnetic resonance imaging (MRI), where indicated (data not shown).

Management of mothers whose fetuses were suspected of having a midgut volvulus consisted of weekly ultrasound examination and fetal heart rate monitoring. All mothers were routinely offered a prenatal genetic counseling with parental testing for the 30 most frequent mutations of cystic fibrosis[6] (currently in existence in our country) and if necessary, complemented by amniocentesis for fetal karyotyping and screening of cystic fibrosis. Decision to deliver the fetus was not based on strict intestinal dilatation cutoff but was more frequently based on a bundle of clinical and sonographic arguments according to gestational age.

The final diagnosis of volvulus was confirmed in the postnatal period by pediatric surgery during an intervention for neonatal intestinal obstruction or during the fetal autopsy in case of intrauterine fetal demise, postnatal fetal demise, or termination of pregnancy (TOP). According to French laws, a TOP for fetal reasons is allowed in case of "a fetal malformation of a particular gravity, recognized as incurable at the time of diagnosis." TOP can be performed up to birth. When postnatal diagnosis (surgery or autopsy) was not available, the diagnosis of volvulus was made from a comprehensive analysis of the file by a group review of the prenatal imaging, based on our growing experience (in particular with cases undergoing spontaneous regression). For cases with spontaneous regression, a pediatric radiologist reanalyzed images of prenatal MRI. The same radiological team also performed, in such cases, postnatal imaging.

The exclusion criteria for our study included the absence of relevant prenatal imaging and the absence of a definitive pathologic diagnosis after surgery or autopsy.

What's already known about this topic?

Prenatal diagnosis of midgut volvulus is a rare condition that remains hard to confirm because of the lack of specific ultrasound features described in literature. However this pathology is a surgical emergency in the postnatal period, with the child's survival and functional prognosis dependent on the delay until postnatal treatment is undertaken

What does this study add?

The presence of a whirlpool sign and/or of intestinal dilatation with a fluid-filled level in the dilated loops improves the accuracy of ultrasound and Doppler findings for volvulus. Prenatal information to the parents must then be reassuring but prudent, in view of the mostly favorable prognosis with a possible spontaneous resolution. The identification of a volvulus allows for close prenatal monitoring and referral to postnatal management in a specialized pediatric center.

The clinical and paraclinical data collected included characteristics of the mother (age and parity), the pregnancy (first trimester ultrasound data, serum marker results for trisomy 21 screening, singleton or multiple pregnancy), any malformation (gestational age at diagnosis, circumstances of discovery, description of ultrasound images and their changes over time), and characteristics of the delivery (pregnancy outcome and gestational age at birth, pediatric surgery data) or results of the fetal autopsy.

For the purpose of the study, each patient was contacted by phone to ascertain the status of the living children who had midgut volvulus *in utero*. The data are reported as mean (m ± SD) or numbers and percentages (n[%]).

3 | RESULTS

The study included 13 cases of fetal midgut volvulus during the study period. Characteristics of the patients, the pregnancy, and the delivery are presented in Table 1, case by case. Midgut volvulus was diagnosed most often during the third trimester, with peaks at 27 (n = 5) and 32 weeks of gestation (n = 6). The clinical context at diagnosis was either during a routine ultrasound examination (6/13 [46%]) or occurred through a sonographic examination performed because of a significant reduction in active fetal movements (8/13 [61.5%]).

Ultrasound features of the 13 cases of midgut volvulus and their outcomes are presented in Table 2. The whirlpool sign was visualized for 10 fetuses (77%) in 2D and color Doppler modes (Figures 1 and 2). In the other three cases, indirect signs resulted in suspicion of volvulus: meconium pseudocyst, n = 4 (30.7%); ascites and peritoneal calcifications, n = 3 (23%); and intestinal dilatation, n = 10 (77%), all

TABLE 1 Characteristics of the 13 cases of midgut volvulus

Case	1	2	3	4	5	6 and 7 (MCDA twins)	8	9 (Twin B of a DCDA Twin Pregnancy)	10	11	12	13	TOTAL (m ± SD or n[%])
Year	2006	2010	2012	2012	2012	2013	2014	2014	2014	2015	2017	2017	
Patient age (years)	32	21	31	29	30	40	34	37	36	28	29	27	31.17 ± 5.1
Parity (including this pregnancy)	3	2	2	1	1	2	3	1	3	3	1	1	1.9 ± 0.9
Gestational age at diagnosis (weeks)	27.7	26.6	34	27	28.9	32.9	33	22.2	31	31	22	27	28.6 ± 4
Decreased active fetal movements	-	1	1	1	1	1	-	-	1	1	-	1	8/13 (61.5%)
Tested for cystic fibrosis mutation	-	1	-	1	1	1	-	1	1	1	-	1	8/13 (61.5%)
Amniocentesis	1	1	1	1	1	1	-	-	-	1	-	1	8/13 (61.5%)
Cystic fibrosis	1	-	-	1	-	-	-	-	-	-	-	-	2/13 (15%)
Intestinal atresia	-	1	1	-	1	-	1	-	-	1	1	-	6/13 (46%)
Outcome	TOP	Birth	Birth	TOP	Birth	Birth (2)	Birth	Birth	Birth	Birth	Birth	Birth	TOP (15%); birth (85%)
Gestational age at outcome	28.3	37.3	35.9	31	38	33.2	35	37.7	38.6	34.1	36	40	35.4
Mode of delivery	V	V	CD	V	CD	V/V	V	CD	CD	CD	V	V	V 8/13 (61.5%); CD 5/13 (38.5%)
Reason for delivery	-	S/P	E	-	S/P	S/P	S/P	S/P	S/P	E	I	S/P	
Reason for CD	-	-	NR	-	Breach	-	-	Breach	Two previous CDs	NR	-	-	
Spontaneous resolution	-	-	-	-	-	-/Yes	-	Yes	-	-	-	Yes	
Day of postnatal surgery	-	Day 3	Day1	-	Day 1	Day 1/-	Day 1	-	Day 1	Day1	Day 1	-	

Abbreviations: CD, cesarean delivery; DCDA, dichorionic, diamniotic; E, emergency; I, induction for increasing digestive dilatation; MCDA, monochorionic diamniotic; NR, nonreassuring fetal heart beat; S/P, spontaneous/planned; TOP: termination of pregnancy; V, vaginal delivery.

TABLE 2 Ultrasound features of the 13 cases of midgut volvulus

	1	2	3	4	5	6 and 7 (twins)	8	9	10	11	12	13	TOTAL
Year	2006	2010	2012	2012	2012	2013	2014	2014	2014	2015	2017	2017	
Gestational age at diagnosis (weeks)	27.7	26.6	34	27	28.9	32.9	33	22.2	31	31	22	27	
Whirlpool sign: 2D and Doppler	1	1	1	1	1	-	-	1	1	1	1	1	10/13 (77%)
Fluid-meconial level	1	-	1	-	1	-	1	-	-	-	-	1	5/13 (38%)
Bowel loop dilatation	1	1	1	-	1	1 and 1	1	1	-	1	-	-	8/13 (61.5%)
Meconium peritonitis: calcifications, ascites, pseudocysts	1	1	1	-	1	1 and 1	1	-	-	1	1	-	9/13 (69%)
Normal position of mesenteric vessels	Yes	Yes	Yes	Yes	Yes	Yes x2	Yes	Yes	Yes	Yes	Yes	Yes	13/13 (100%)
Outcomes	TOP	Birth	Birth	TOP	Birth	Birth x2	Birth	Birth	Birth	Birth	Birth	Birth	TOP: 2/13 (15%); birth 11/13 (85%)

Abbreviation: TOP, termination of pregnancy..

confirmed postnatally by pediatric surgery or fetal autopsy, both with a pathologic examination (Figures 3 and 4). Retrospectively, fluid-filled levels were seen clearly in dilated small bowel loops in five fetuses (38.5%) (Figure 5). This sign consists of a horizontal level separating an anechoic part superiorly and an echoic part inferiorly (meconium and/or blood content). Neither the whirlpool sign nor the fluid-filled levels in dilated bowel loops were found without volvulus and especially in bowel atresia without volvulus. No prenatal ultrasound images showed malposition of the mesenteric vessels with intestinal malrotation.

The causes of the cases of volvulus presented here were (a) suspected idiopathic mesenteric fusion in a monochorionic diamniotic twin pregnancy affecting both fetuses (cases #6 and #7), (b) two cases associated with cystic fibrosis confirmed genetically before birth (cases #1 and #4), and (c) nine cases of idiopathic volvulus without predisposing factors (cases #2, #3, #5, #8, #9, #10, #11, #12, and #13). Among those 13 cases, three fetuses (23%) experienced spontaneous prenatal regression of the whirlpool sign on both 2D and color Doppler ultrasound (cases #7, #9, and #13).

Six children (46%) were born preterm (cases #3, #6, #7, #8, #11, and #12), three of them spontaneously, because of tocolytic failure (cases #6, #7, and #8) and, in three other cases, because of an induction of labor for increased bowel dilatation (#12) or emergency cesarean delivery for fetal heart rate decelerations (#3 and #11).

Among the 13 cases, eight (61.5%) newborns required immediate pediatric surgical management (cases #2, #3, #5, #6, #8, #10, #11, #12). The diagnosis was confirmed for all eight, including four newborns with direct positive signs of volvulus and four other newborns with indirect secondary signs of this episode (small bowel atresia, peritonitis, meconium pseudocyst, and intestinal perforation) (Figure 6 and 7). For all of cases, surgery consisted of a resection and anastomosis of the midgut. One child (case #3) died during the immediate neonatal period despite intensive resuscitation. Termination of pregnancy was performed for two fetuses (cases #1 and #4) (15.5%) because of the association with cystic fibrosis known to be of particular gravity when prenatally diagnosed (often severe mutations). [7-9]

4 | DISCUSSION

Prenatally diagnosed midgut volvulus remains a rare entity, seldom described in the literature.[10,11] This article presents the largest retrospective series of these cases yet published. The most relevant prenatal ultrasound signs for this disease were the whirlpool sign (77%) and a fluid-filled level in dilated intestinal loops (38.5%).

We observed two peaks of diagnosis: the first around 27 weeks of gestation, and the other around 32 weeks. Our data are consistent with the literature showing that the onset of volvulus usually starts around 27 weeks, when the ultrasound signs generally become apparent.[12] Beyond ultrasound monitoring, clinical monitoring of active fetal movements shows that their decline also appears to be a marker of the onset of volvulus. Among 13 cases, eight (61.5%) mothers reported a reduction in these movements. Because this sign is very unspecific and difficult to quantity or identify objectively, depending as it does on the mother's personal evaluation, it is rarely mentioned

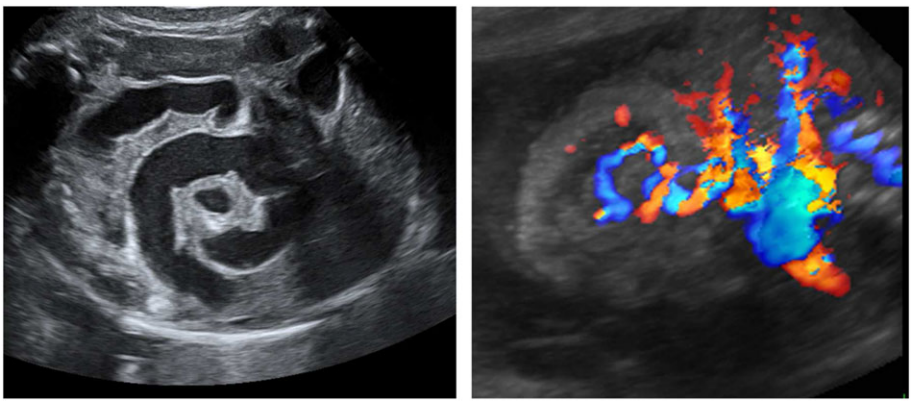

FIGURE 1 Case #2, 26 weeks of gestation. Prenatal midgut volvulus: Ultrasound features: whirlpool sign (axial ultrasound image in 2D and color Doppler) [Colour figure can be viewed at wileyonlinelibrary.com]

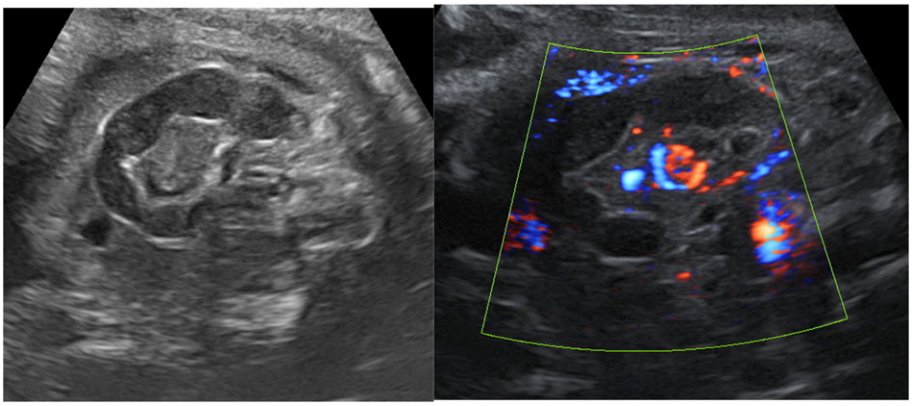

FIGURE 2 Case #10, 32 weeks of gestation. Prenatal midgut volvulus: Ultrasound features: whirlpool sign (axial ultrasound image in 2D and color Doppler). With color Doppler, mesenteric vessels appeared as a vascular pellet at the center of the spiraled intestinal loops [Colour figure can be viewed at wileyonlinelibrary.com]

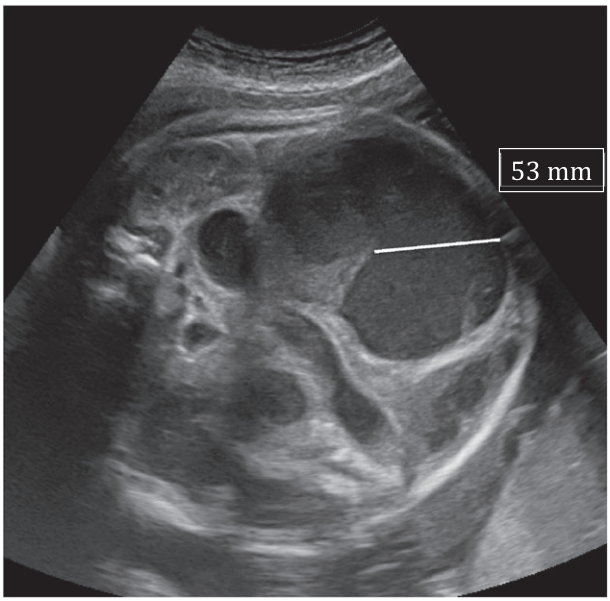

FIGURE 3 Case #9, 22 weeks of gestation. Prenatal midgut volvulus: Ultrasound features: whirlpool sign associated with massive intestinal dilatation at 22 weeks (axial ultrasound image)

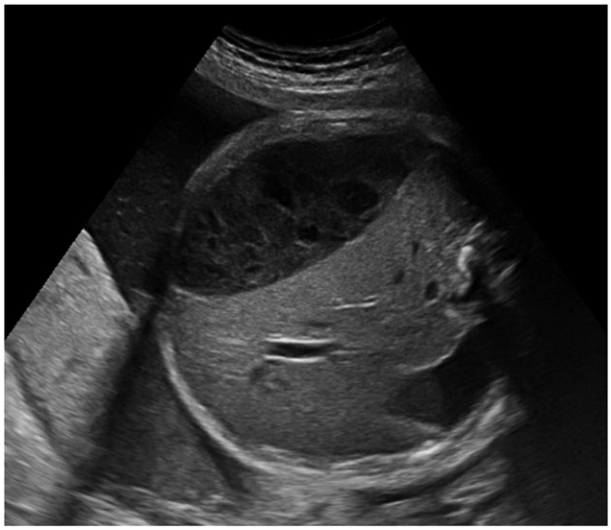

FIGURE 4 Case #12, 35 weeks of gestation. Prenatal midgut volvulus: Ultrasound features: wide meconium pseudo cyst (axial ultrasound image)

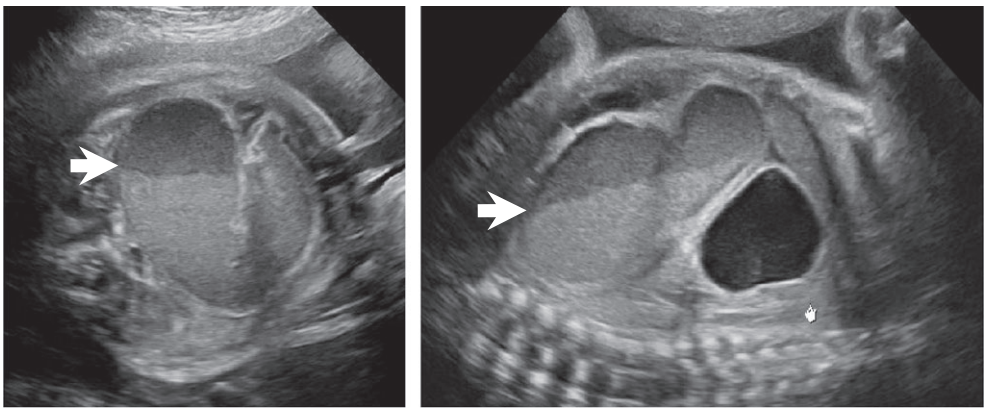

FIGURE 5 Case #3, 34 weeks of gestation. Prenatal midgut volvulus: Ultrasound features: fluid-filled level (→) (axial and parasagittal ultrasound images)

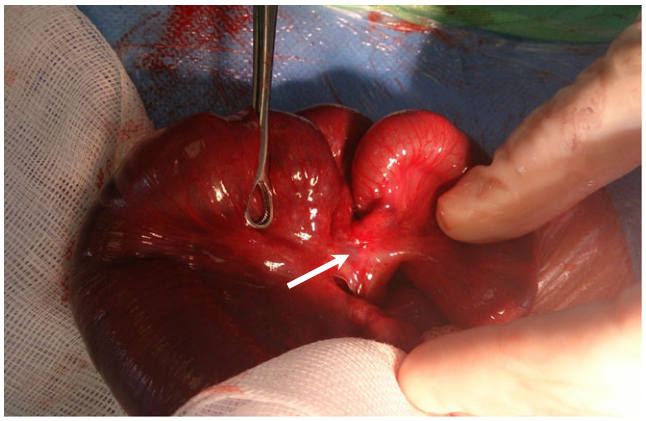

FIGURE 6 Case #6, 33 weeks of gestation. Monochorionic diamniotic twin pregnancy with a volvulus diagnosed in both fetuses. Spontaneous delivery at 33 weeks (tocolysis failure). Surgical confirmation of the volvulus only in twin A with an abnormal mesenteric fusion (→arrow) [Colour figure can be viewed at wileyonlinelibrary.com]

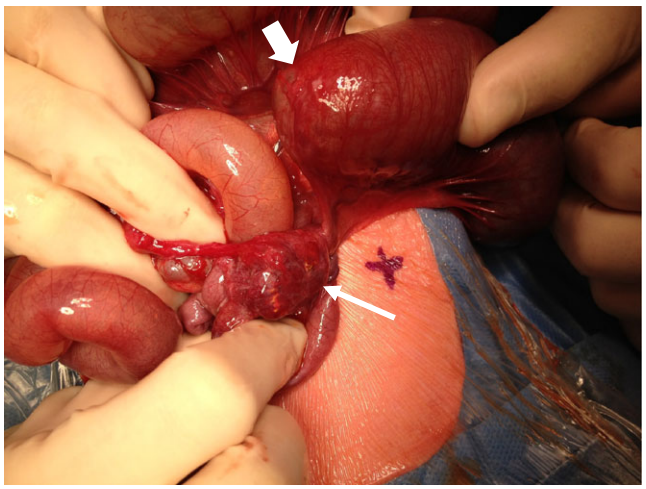

FIGURE 7 Case #5, diagnosis at 29 weeks of gestation. Planned cesarean delivery at 38 weeks for breech presentation. Surgical confirmation of the volvulus with an atresia (dilatation of the proximal gut, large arrow), and a small distal gut (thin arrow). There was no short midgut [Colour figure can be viewed at wileyonlinelibrary.com]

in the literature.[13,14] However, in our case series, more than 50% of patients in this situation reported a decrease in fetal movements, which doesn't seems to be accidental. It appears that at the moment of the volvulus and/or the secondary intestinal perforation, the fetus, stunned by the pain, limits its movements. Finally, the two periods of gestation during which signs of volvulus were observed also corresponded to the periods following routine ultrasound screening during pregnancy in France. This might be considered a bias in terms of the discovery of the diagnosis even though some of these ultrasound examinations were justified by a reduction in active fetal movements.

The whirlpool sign described postnatally has thus also been reported before birth. Nonetheless, their descriptions differ. After birth, Pracros et al described a mesenteric mass resulting from a spiral involving the mesenteric vessels and the strangulated loops.[3] Before birth, the whirlpool sign corresponds to the circular winding of the loops involved in the volvulus mechanism around one another. This sign was found in 77% of our cases, as well as by Yoo et al[11] and by Has and Gunay.[10] The visualization of the whirlpool sign in 2D ultrasound thus defines a positive diagnosis of intestinal volvulus. Nonetheless, its appearance differed according to both gestational age at diagnosis and the degree of dilatation of the intestinal loop. It was easy to visualize at an earlier gestational age and when the dilated loop was only moderate (Figures 1 and 2). On the other hand, this whirlpool sign was no longer visible at the stage of meconium peritonitis. In the presence of more extensive intestinal dilatation, it was necessary to follow the path of the loops to see how they were coiled or to use 3D volume acquisition of the fetal abdomen with multiplanar reconstruction (Figure 8). Visualization of the whirlpool sign in color Doppler mode was identical to that in 2D, varying similarly by gestational age at diagnosis and degree of the dilated loop. It took the appearance of a vascular pellet at the center of the spiraled intestinal loops during an isolated visualization of their winding (Figure 2). The appearance of superior mesenteric vessels winding in a spiral around their axis was definitively identified in only five cases in our series (Figure 2). We were not able to determine conclusively the direction of the rotation—clockwise or counterclockwise—of the wrapping around the vascular pedicle. The direction has been described in the postnatal period and appears decisive for the diagnosis of intestinal volvulus.[15] Although the diagnostic performance of the whirlpool sign

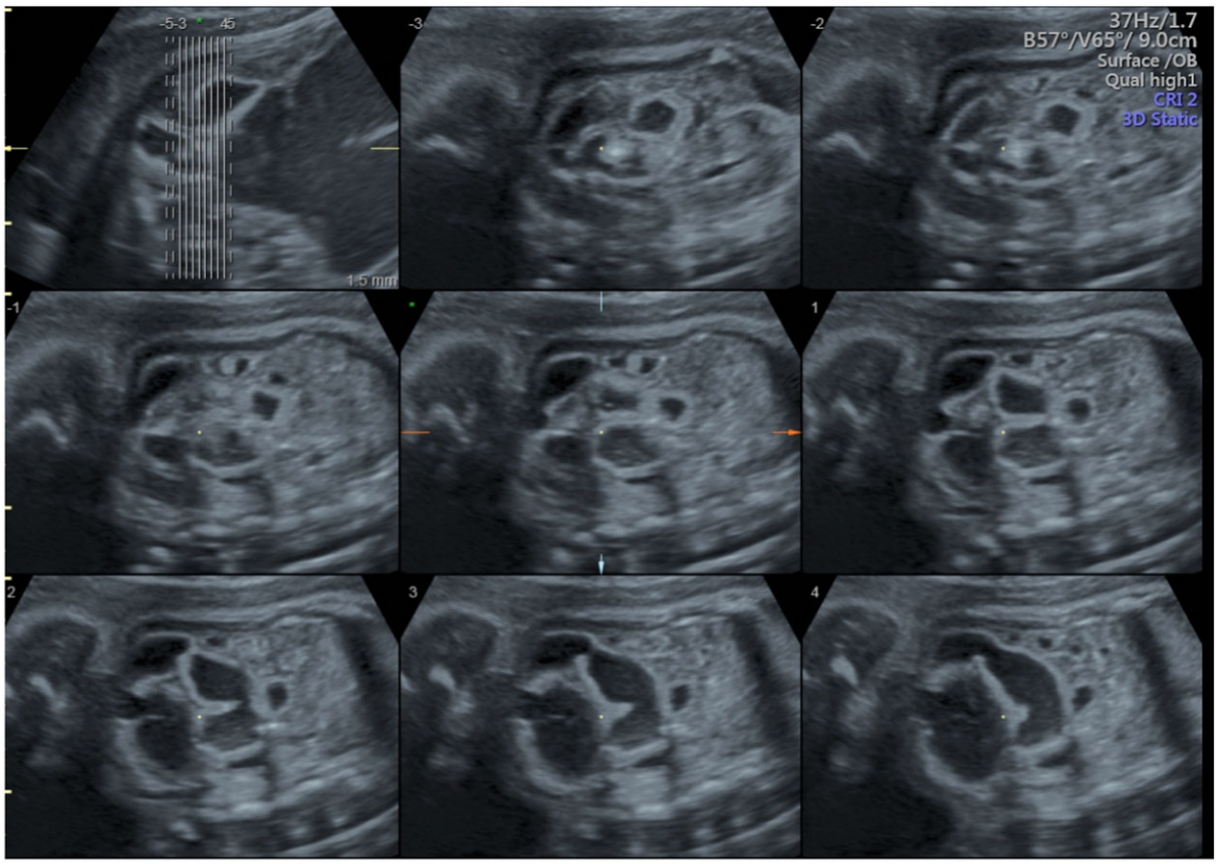

FIGURE 8 Case #4, 27 weeks of gestation. Prenatal midgut volvulus: Ultrasound features: whirlpool sign in 3D mode and multiple plane examination [Colour figure can be viewed at wileyonlinelibrary.com]

has not been evaluated before birth, its postnatal sensitivity and specificity are 89% and 92%,[3,12] respectively.

Another ultrasound sign described for the first time in this work is the existence of a fluid-filled level within a dilated loop of the small bowel (Figure 5). This sign was present in 38.5% of the cases in our series. Although not mentioned in the prenatal literature, it is nonetheless known by pediatric radiologists as a sign of intestinal distress and evidence of a loop without peristalsis.[16] It corresponds to an echogenic material below, probably meconium, with more or less blood content, with an anechoic fluid above. Both peristalsis and fluid-filled level needs to be analyzed together. It is generally located in the loop just above the obstacle, which is the most distended. On the other hand, this sign is never present in intestinal atresia without volvulus. When midgut volvulus occurs *in utero*, it can be complicated by ischemic necrosis of the loops because of its associated vascular effects.[17-21]

Another postnatal sign of midgut volvulus reported in literature is the coffee bean sign.[20] It has been described in postnatal radiology, essentially in volvulus of the sigmoid and solely when it involves the volvulus of a single loop twisted on itself. The pathophysiology of prenatal volvulus, although segmental, involves several loops of the small intestine. We therefore did not observe this sign in our prenatal series. Contrary to what has been reported in the literature, our series included no cases of total volvulus.[18,22]

Volvulus is frequently associated with atresia of the small intestine, which can be the cause or the mechanical consequence of midgut volvulus.[23,24] In our series, for the cases diagnosed at the earliest stage, volvulus was clearly shown without any evident or substantial intestinal dilatation. It was only secondary, in the cases that did not regress, that this dilatation appeared, being associated with the atresia due to the volvulus. This initial absence of intestinal dilatation suggests that for some of these cases, the volvulus caused the atresia and not vice versa. The final lack of atresia in the cases of regression also demonstrates that the cause of the volvulus in these situations was not related to a primary atresia, because it cannot regress spontaneously.

Three of our cases of volvulus, however, did spontaneously regress, with no complication observed during the ultrasound follow-up. For these three cases, the ultrasound image was identical, characterized by an isolated whirlpool signs of small-bowel loops, not significantly dilated, and winding around a central pellet of the mesenteric vascular pedicle. There was neither significant intestinal dilatation nor any sign of meconium peritonitis. The follow-up ultrasound performed 2 weeks after the diagnosis showed the complete disappearance of the whirlpool sign in all three cases. All three of these newborns had normal postnatal clinical examinations and normal abdominal ultrasound findings. We debated for these three cases the possibility of a segmental volvulus that regressed spontaneously or a false-positive whirlpool sign. Several spontaneous resolutions *in utero* have been reported.[25-27] In our series, one of these cases was diagnosed in a dichorinic diamniotic twin pregnancy only in twin B (case #9). A complementary MRI was performed at 23 weeks and was confident with the diagnosis of a volvulus in twin B, as diagnosed with ultrasound, and assessed the normality of the digestive tract of twin A

(Figure 9). The follow-up during pregnancy showed a spontaneous resolution of twin B volvulus, which was confirmed after birth with postnatal imaging.

After birth, the most frequent cause of total volvulus is intestinal malrotation on an incomplete common mesentery. We systematically looked for malrotation in our series by studying the relative positions of the mesenteric superior vessels. This study took place in 2D ultrasound and was sometimes completed by a color Doppler examination. Because the vascular pedicle was wrapped by the volvulus, the study of the relative positions of the mesenteric vessels could not be performed in the usual plane. The mesenteric artery and vein could be visualized only on an axial section of the abdomen between the plane through the left renal vein and that of the aortic source of the superior mesenteric artery (Figure 10). Accordingly, the relative position of the vessels had to be defined before the wrapping of the pedicle began. For all of the fetuses studied, the vein was normally positioned on

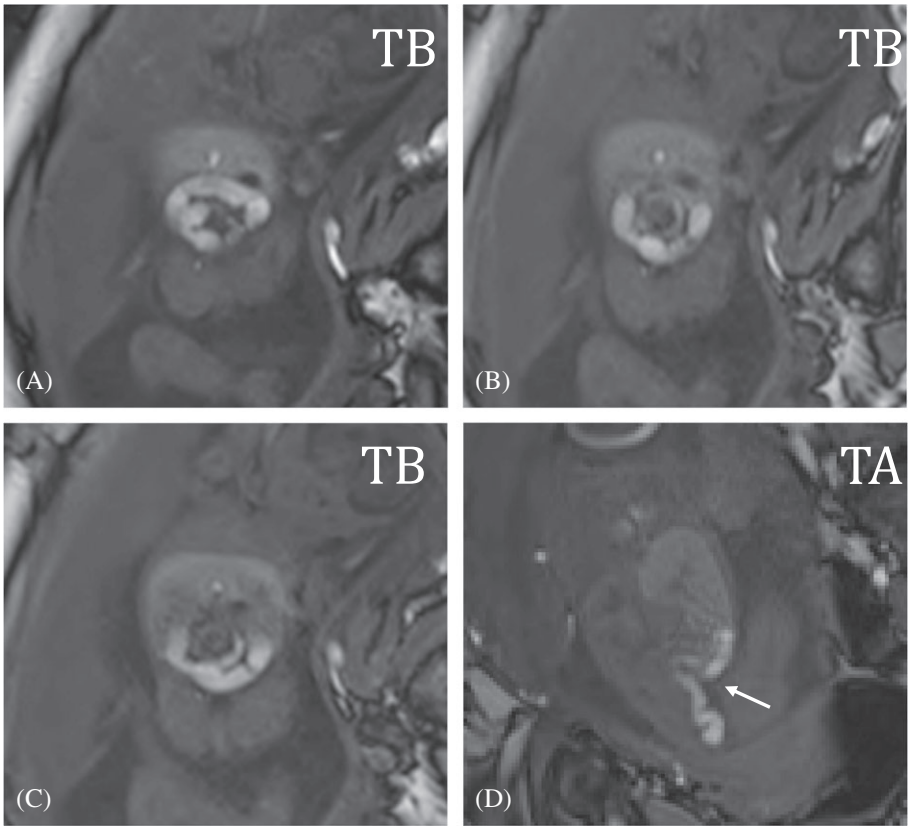

FIGURE 9 Case #9, 22–23 weeks of gestation. A, B, C, Dichorionic diamniotic twin pregnancy, twin B (case #9). MRI T1 weighted sequence coronal plane showing the volvulus of few bowel loops with hypersignal content (due to meconial and/or haemorrhagic content). D, Twin A (unaffected). Normal meconium pattern in the sigmoid and the rectum (→arrow) for comparison.

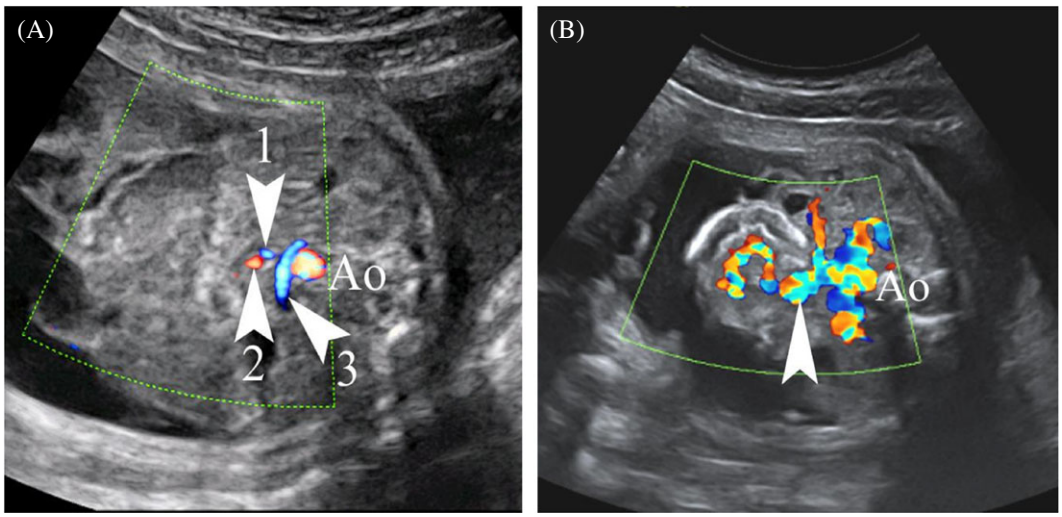

FIGURE 10 Case #5, 29 weeks of gestation. The mesenteric artery (1 and arrow on image B) and vein (2) could be visualized only on an axial section of the abdomen between the plane through the left renal vein (3) and that of the aortic (Ao) source of the superior mesenteric artery [Colour figure can be viewed at wileyonlinelibrary.com]

the right edge of the mesenteric artery. None of the cases with postnatal confirmation of the ultrasound findings in our series included any case of intestinal malrotation with total volvulus (no postnatal confirmation for one of the two cases that ended with TOP because of cystic fibrosis).

Weekly ultrasound scans should be performed to monitor the increase in the volume of the intestinal loops and screen for serious forms. The only case in our series with a postnatal complication (case #3) was the one with a large gastric and duodenal dilatation (with a fluid-filled level). Accordingly, an increase in the volume of the dilated loops, with or without a meconium pseudocyst (Figure 4), can call for a more reserved prognosis, which must be provided to the parents, and birth may be scheduled for early surgical management.

Our study has several limitations. On the one hand, analysis of the cases was retrospective, with a review of all files from 2006 through 2017. The ultrasound examinations were analyzed based on the regular images recorded in the department database, and we were unable to visualize any dynamic sequences except for the video loops that had been made.

Several strengths of this work must nonetheless be mentioned. This is only the second series of prenatal diagnosis of intestinal volvulus thus far reported in the literature. The first series did not describe new ultrasound sign and reported only one case with a certain prenatal diagnosis.[5] Our series included the largest number of cases in the literature and considered all of the potential ultrasound signs, especially the specific signs that could lead to diagnostic ultrasound clusters of volvulus.

5 | CONCLUSION

Prenatal diagnosis of fetal midgut volvulus can be made by using 2D and color Doppler ultrasound. Our diagnostic performance with prenatal ultrasound improved progressively with the demonstration of diagnostic ultrasound clusters of volvulus: the whirlpool sign (77%) and intestinal dilatation with a fluid-filled level in the dilated loops (38.5%). Prenatal volvulus is more often diagnosed at two peaks of gestational age of 27 weeks and 32 weeks, sometimes induced by maternal perception of a decrease of fetal movements. The discovery of prenatal volvulus requires ruling out an association with cystic fibrosis (two cases of the 13 in our series, all ended with TOP) and looking for intestinal malrotation (0/13). Besides these two situations, substantial dilatation and/or a voluminous meconium pseudocyst requires to give to the parents a reserved prognosis (1/13). Outside those situations, prenatal information to the parents must then be reassuring but cautious because of the mostly favorable prognosis (10/11 (91%)[TOP excepted]) with a possible spontaneous resolution (3/13). The identification of volvulus allows close prenatal monitoring and referral to postnatal management in a specialized pediatric center.

CONFLICT OF INTEREST

The authors report no conflict of interest with this work.

ORCID

Florent Fuchs https://orcid.org/0000-0003-3530-6328

REFERENCES

1. Raherison R, Grosos C, Lemale J, et al. Prenatal intestinal volvulus: a life-threatening event with good long-term outcome. *Arch Pediatr*. 2012;19(4):361-367.
2. Crisera CA, Ginsburg HB, Gittes GK. Fetal midgut volvulus presenting at term. *J Pediatr Surg*. 1999;34(8):1280-1281.
3. Pracros JP, Sann L, Genin G, et al. Ultrasound diagnosis of midgut volvulus: the "whirlpool" sign. *Pediatr Radiol*. 1992;22(1):18-20.
4. Valladares E, Rodriguez D, Vela A, Cabre S, Lailla JM. Meconium pseudocyst secondary to ileum volvulus perforation without peritoneal calcification: a case report. *J Med Case Reports*. 2010;4(1):292.
5. Sciarrone A, Teruzzi E, Pertusio A, et al. Fetal midgut volvulus: report of eight cases. *J Matern Fetal Neonatal Med*. 2016;29(8):1322-1327.
6. Durand M, Coste K, Martin A, et al. Fetal midgut volvulus as a sign for cystic fibrosis. *Prenat Diagn*. 2008;28(10):973-974.
7. Takacs ZF, Meier CM, Solomayer EF, Gortner L, Meyberg-Solomayer G. Prenatal diagnosis and management of an intestinal volvulus with meconium ileus and peritonitis. *Arch Gynecol Obstet*. 2014;290(2):385-387.
8. Tonni G, Grisolia G, Granese R, et al. Prenatal diagnosis of gastric and small bowel atresia: a case series and review of the literature. *J Matern Fetal Neonatal Med*. 2016;29(17):2753-2761.
9. Scotet V, De Braekeleer M, Audrezet MP, et al. Prenatal detection of cystic fibrosis by ultrasonography: a retrospective study of more than 346 000 pregnancies. *J Med Genet*. 2002;39(6):443-448.
10. Has R, Gunay S. 'Whirlpool' sign in the prenatal diagnosis of intestinal volvulus. *Ultrasound Obstet Gynecol*. 2002;20(3):307-308.
11. Yoo SJ, Park KW, Cho SY, Sim JS, Hhan KS. Definitive diagnosis of intestinal volvulus in utero. *Ultrasound Obstet Gynecol*. 1999;13(3):200-203.
12. Chao HC, Kong MS, Chen JY, Lin SJ, Lin JN. Sonographic features related to volvulus in neonatal intestinal malrotation. *J Ultrasound Med*. 2000;19(6):371-376.
13. Sammour RN, Leibovitz Z, Degani S, Shapiro I, Ohel G. Prenatal diagnosis of small-bowel volvulus using 3-dimensional Doppler sonography. *J Ultrasound Med*. 2008;27(11):1655-1661.
14. Uerpairojkit B, Charoenvidhya D, Tanawattanacharoen S, Manotaya S, Wacharaprechanont T, Tannirandorn Y. Fetal intestinal volvulus: a clinico-sonographic finding. *Ultrasound Obstet Gynecol*. 2001;18(2):186-187.
15. Shimanuki Y, Aihara T, Takano H, et al. Clockwise whirlpool sign at color Doppler US: an objective and definite sign of midgut volvulus. *Radiology*. 1996;199(1):261-264.
16. Cozza S, Ferrari FS, Stefani P, et al. Ileal occlusion with strangulation: importance of ultrasonography findings of the dilated loop with intraluminal fluid-fluid resulting from sedimentation. *Radiol Med*. 1996;92(4):394-397.
17. Kornacki J, Czarnecka M, Blaszczynski M, et al. Congenital midgut volvulus associated with fetal anemia. *Fetal Diagn Ther*. 2010;28(2):119-122.
18. Miyakoshi K, Ishimoto H, Tanigaki S, et al. Prenatal diagnosis of midgut volvulus by sonography and magnetic resonance imaging. *Am J Perinatol*. 2001;18(08):447-450.
19. Noreldeen SA, Hodgett SG, Venkat-Raman N. Midgut volvulus with hemorrhagic ascites: a rare cause of fetal anemia. *Ultrasound Obstet Gynecol*. 2008;31(3):352-354.
20. Park JS, Cha SJ, Kim BG, et al. Intrauterine midgut volvulus without malrotation: diagnosis from the 'coffee bean sign. *World J Gastroenterol*. 2008;14(9):1456-1458.

21. Usmani SS, Kenigsberg K. Intrauterine volvulus without malrotation. *J Pediatr Surg*. 1991;26(12):1409-1410.

22. Miyakoshi K, Tanaka M, Miyazaki T, Yoshimura Y. Prenatal ultrasound diagnosis of small-bowel torsion. *Obstet Gynecol*. 1998;91(5 Pt 2):802-803.

23. Mercado MG, Bulas DI, Chandra R. Prenatal diagnosis and management of congenital volvulus. *Pediatr Radiol*. 1993;23(8):601-602.

24. Yu W, Ailu C, Bing W. Sonographic diagnosis of fetal intestinal volvulus with ileal atresia: a case report. *J Clin Ultrasound*. 2013;41(4):255-257.

25. 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(25):1519-1521.

26. Heydanus R, Spaargaren MC, Wladimiroff JW. Prenatal ultrasonic diagnosis of obstructive bowel disease: a retrospective analysis. *Prenat Diagn*. 1994;14(11):1035-1041.

27. Weissman A, Goldstein I. Prenatal sonographic diagnosis and clinical management of small bowel obstruction. *Am J Perinatol*. 1993;10(3):215-216.

How to cite this article: Bartholmot C, Faure J-M, Grosjean F, et al. Prenatal diagnosis of antenatal midgut volvulus: Specific ultrasound features. *Prenatal Diagnosis*. 2019;39:16–25. https://doi.org/10.1002/pd.5392