

Amniotic band syndrome with sacral agenesis and umbilical cord entrapment: A case report emphasizing the value of evaluation of umbilical cord

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ABSTRACT

Amniotic band syndrome is a rare congenital disorder caused by entrapment of fetal parts by fibrous amniotic bands in utero. The congenital anomalies seen in this syndrome vary widely and defects may be isolated or multiple and do not follow a specific pattern. Asymmetric distribution of defects is the hallmark of this syndrome. The diagnosis is difficult to make on ultrasound and relies on identification of amniotic bands. We report a case of amniotic band syndrome with sacral agenesis diagnosed on routine antenatal ultrasound scan in the second offspring of a recently diagnosed diabetic mother. The associated features were entrapment of umbilical cord, caudal adhesions and lower limb anomalies. Medical termination of pregnancy was done and all the fetal anomalies as well as umbilical cord abnormalities were confirmed. The importance of meticulous scanning to evaluate for amniotic bands and the umbilical cord in addition to the fetal structures is emphasized.

CASE REPORT

CASE REPORT

A 30 year old gravida 2 para 1 female was referred to our hospital for a routine antenatal ultrasound examination at a gestation age of 17 weeks 3 days by last menstrual period. The patient had a non-consanguineous pregnancy. The first child was a healthy five year old male from an uneventful full term normal gestation. In the present gestation she was diagnosed with uncontrolled diabetes mellitus by a routine blood test at 13 weeks. There was no history of recent infection, invasive procedure, maternal trauma or prenatal exposure to radiation or teratogenic drugs. No family history of congenital malformations was elicited.

Ultrasound examination revealed a single intrauterine gestation with fetal biometry corresponding to 17 weeks gestational age. A large anterior abdominal wall defect was noted with exteriorization of liver, stomach and bowel (Fig. 1). Multiple thin and linear amniotic bands were identified (Fig. 2). One band was seen to trap the umbilical cord in its entire length and was seen looping around the left thigh of the fetus (Fig. 3, 4). The umbilical cord was very short and demonstrated two vessels (a single umbilical artery and an umbilical vein) (Fig. 5). Another amniotic band was seen encircling the thoraco-abdominal junction resulting in a focal constriction (Fig. 1A).

The right lower extremity was absent and the left lower extremity revealed clubfoot (Fig. 6). Bilateral upper limb bones were normal. There was a rotational deformity of the lumbar spine and non-visualization of the sacrum (Fig. 7, 8). The cervical and thoracic spine appeared normal. Thoracic structures including the heart were normal. Intracranial structures and facial contents also appeared normal. There was no cleft lip or cleft palate. The fetus was examined for about 30 minutes, during which period no movements of the left lower extremity were observed suggesting restricted movements of the left lower limb by the amniotic band, resulting in akinesia. The placenta was carefully examined and found to be normal. Liquor volume was appropriate.

After parental counselling, medical termination was performed by labor induction using misoprostol. The patient delivered a stillborn male child weighing 250 gram, with all the fetal malformations as observed in the antenatal scan (Fig. 9, 10). Radiograph of the fetus confirmed rotational scoliosis of the lumbar spine and sacral agenesis (Fig. 11). The umbilical cord was confirmed to be very short and measured 10 cm in length and was seen entrapped by the amniotic band around the left thigh (Fig. 12). The umbilical cord had a single umbilical artery. No placental abnormalities were found.

DISCUSSION

Amniotic band syndrome (ABS) is a rare fetal malformation which ranges from mild deformities to severe anomalies incompatible with post natal life [1].

Etiology and demographics

ABS is a rare, sporadic condition with an incidence of 1: 11,200 births and no sex prediction [2, 3]. Antenatal diagnosis of this syndrome is difficult because the amniotic bands may be very thin and difficult to identify on ultrasound, unless a thorough search is performed. The exact cause of this polymorphic abnormality is yet unknown. Various etiological factors hypothesized are amniocentesis, collagen vascular disease of mother, deformed uterus and drugs as cocaine and mifepristone [3]. Various theories have been postulated to explain the pathogenesis. Amniotic disruption theory by Torpin et al is the most widely accepted theory. This theory suggests that amniotic band syndrome occurs when amnion ruptures before 12 weeks of gestation resulting in chorionic side of amnion emanating numerous mesoblastic fibrous strings which entrap fetal parts [3, 4]. As fetus develops, the amniotic bands can trap extremities causing immobilization, constriction or amputation of structure [3]. Early insult results in facial clefts and brain defects, while late insult (after 45 days gestation) results in limb involvement without facial clefting or CNS involvement, as seen in our case [4].

Van Alen proposed the vascular disruption theory that states that this malformation may be caused by events that affect the blood supply to various organs during embryogenesis which may result in abnormal development of structures or damage to preformed structures [5]. This theory has been questioned by various investigators and cannot fully

explain all the abnormalities seen in this condition [6, 7]. Endogenous theory was described by Streeter in 1930, suggesting that these abnormalities were caused by imperfect histogenesis [8]. However, some of the cases of amniotic band syndrome have shown geographical and temporal clustering and cannot be explained by this theory. Differences in regard to the exact etiology and pathogenesis still persist.

Clinical and Imaging findings

Fetal malformations encountered in this syndrome broadly fall into three main categories: limb defects, craniofacial defects, and visceral defects [1]. There is a wide range of abnormalities encountered in this syndrome and no two cases are identical. In our case, the fetus had multiple amniotic bands with visceral and limb anomalies and a short umbilical cord with single umbilical artery. Short umbilical cord is a common finding in amniotic cord syndrome, possibly due to limited fetal movements and available fetal space [9]. Short cord has not been reported in early aborted cases of the amniotic band syndrome, suggesting it is a secondary feature [9]. A normal umbilical cord has two umbilical arteries and one umbilical vein, surrounded by Wharton's Jelly [10]. Umbilical cord with only single umbilical artery (SUA) is associated with increased risk of renal and cardiovascular anomalies. A less common association of SUA has been described in cases of ABS, as also seen in our case [9]. A topographic correlation of the presence of SUA has been suggested with regional location of amniotic adhesions in ABS [9]. In their study of 48 cases, Belinda et al. reported the presence of finding of SUA in 64% cases of ABS with caudal adhesions and caudal malformations, however SUA was not seen in any case of pure cephalo-thoracic malformations [9]. A proposed mechanism for this specifically regional association of SUA with caudal adhesions is the exposure of umbilical cord in the absence of a covering abdominal wall resulting in disruption of one umbilical artery [9].

Although ultrasound is the mainstay of prenatal imaging diagnosis of ABS, fetal magnetic resonance imaging is a complementary imaging modality in cases where fetal surgery is contemplated, given its large field of view and high soft tissue contrast, since it provides excellent 3-D anatomical detail of the entire pregnancy [11]

Treatment and prognosis

Management options are limited. In-utero surgical correction may be attempted if there is threatened limb amputation or constriction of the umbilical cord. Fetoscopic laser release of amniotic bands has been successfully performed for threatened limb amputation [12]. In less severe cases with digital abnormalities, cleft lip or cleft palate, surgical correction can be performed after birth. As in our case, extensive abnormalities detected on antenatal ultrasound entail parental counselling and termination of pregnancy.

There is an increased risk of premature rupture of membranes, prematurity and low birth weight. Prognosis depends on the degree of malformation and location and severity of the constricting bands. Bands encircling the head and the umbilical cord can be concerning for fetal demise, with umbilical cord constriction by amniotic bands seen in 10% of

cases [11]. Entrapment of the umbilical cord may be a precursor to cord strangulation and subsequent derangement of vascular supply to the fetus resulting in fetal demise [13]. Therefore, in a case with unusual distribution of fetal defects, careful evaluation for amniotic bands should be done. It is also critical to evaluate the entire length of umbilical cord for entrapment and use color Doppler for evaluation of blood supply to the fetus in these cases.

Differential diagnoses

Amniotic bands should not be mistaken for chorioamniotic separation that is normally seen early in pregnancy and after amniocentesis. Another mimic of amniotic bands is amniotic sheets. Amniotic sheets develop from uterine synechiae and do not have any attachment to the fetus or umbilical cord. Unlike the membranes in ABS which are composed only of amnion, the membranes in this condition comprise of two layers of amnion and chorion and hence amniotic sheets are thicker and layered. Furthermore, amniotic sheets are not associated with fetal anomalies [14]. Therefore, when linear structures are seen in the amniotic cavity, it is important to differentiate these two conditions by looking for associated fetal anomalies and for the attachment of the membranes.

The unique finding in our case was identification of an additional finding of sacral agenesis. In our case the mother was diagnosed with diabetes during an antenatal blood test at 13 weeks of gestation. A great confounding factor of overt diabetes is present in our case and sacral agenesis is the commonest complication of maternal diabetes, occurring nearly 200 times more frequently in diabetic mothers, hence sacral agenesis in the present case seems more attributable to maternal diabetes rather than as an association of ABS [15]. The teratogenic mechanism of sacral agenesis in diabetes is not fully understood. Welch and Aterman postulated that sacral agenesis was a result of exposure of a genetically predisposed embryo to some factor in the uterus, such as insulin, antibodies to insulin, or some abnormality of carbohydrate metabolism [16].

To conclude, ABS is a rare disorder with a wide range of fetal abnormalities, which vary in severity and may involve different organs. Our case revealed sacral agenesis in addition to other findings of amniotic band syndrome, which was a unique finding probably attributable to overt diabetes in the mother.

TEACHING POINT

On antenatal ultrasound evaluation of a fetus with asymmetric distribution of defects, meticulous evaluation for the presence and location of amniotic bands should be performed to confidently diagnose amniotic band syndrome. Thorough assessment of the entire length of umbilical cord is critical to exclude cord entrapment and strangulation, which is vital for prognostication and management of these cases.

REFERENCES

1. Burton D J, Filly RA. Sonographic diagnosis of the amniotic band syndrome. *AJR Am J Roentgenol.*1991 Mar; 156(3):555-8. PMID: 1899757.
2. Chen HE, Chen CP, Hsu CY et al: Typical body wall defect associated with craniofacial anomalies and amniotic bands diagnosed in early pregnancy. *Taiwan J Obstet Gynecol.* 2007 Sep; 46(3):286-7. PMID: 17962113.
3. Cignini P, Giorlandino C, Padula F et al. Epidemiology and risk factors of amniotic band syndrome, or ADAM sequence. *J Prenat Med.* 2012 Oct; 6(4):59-63. PMID: 23272276.
4. Oliemen Peterside, James E Omietimi, Oyedeji O Adeyemi: Amniotic band syndrome: a report of two cases and review of the literature. *IOSR Journal of Dental and Medical Sciences;* 5: Mar.- Apr. 2013
5. Van Allen MI. Fetal vascular disruptions: mechanisms and some resulting birth defects. *Pediatr Ann.* 1981 Jun; 10(6):219-33. PMID: 7254912.
6. Werler MM, Bosco JL, Shapira SK: National Birth Defects Prevention Study. Maternal vasoactive exposures, amniotic bands, and terminal transverse limb defects. *Birth Defects Res A Clin Mol Teratol.* 2009 Jan; 85(1):52-7. PMID: 19067400.
7. Davies BR, Giménez-Scherer JA. Comparison of the amniotic band disruption complex with acardiac twins does not support its vascular origin. *Fetal Pediatr Pathol.* 2007 Mar-Apr; 26 (2):87-99. PMID: 17701691.
8. Streeter GL. Focal deficiencies in fetal tissues and their relation to intrauterine amputations. *Contrib Embryol Carnegie Inst* 1930; 22:1-44.
9. Davies BR, Giménez-Scherer JA, Hernández-Sierra JF. Fetal amniotic adhesions. Their topographic concordance with regionally clustered malformations. *Arch Med Res.* 2001 Jan-Feb; 32(1): 48-61. PMID: 11282181.
10. De Laat MW, Franx A, van Alderen ED, Nikkels PG, Visser GH. The umbilical coiling index, a review of the literature. *J Matern Fetal Neonatal Med.* 2005 Feb;17(2):93-100. Review. PMID: 16076615.
11. Neuman J, Calvo-Garcia MA, Kline-Fath BM, Bitters C, Merrow AC, Guimaraes CV, Lim FY. Prenatal imaging of amniotic band sequence: utility and role of fetal MRI as an adjunct to prenatal US. *Pediatr Radiol.* 2012 May;42(5):544-51. PMID: 22134536.
12. Keswani SG, Johnson MP, Adzick NS et al. In utero limb salvage: fetoscopic release of amniotic bands for threatened limb amputation. *J Pediatr Surg.* 2003 Jun; 38(6):848-51. PMID: 12778379.

13. Lurie S, Feinstein M, Mamet Y. Umbilical cord strangulation by an amniotic band resulting in a stillbirth. *J Obstet Gynaecol Res.* 2008 Apr; 34(2):255-7. PMID: 18412792.
14. Özkavukcu E, Haliloğlu N. Gray-scale and color Doppler US findings of amniotic sheets. *Diagn Interv Radiol.* 2012 May-Jun; 18(3):298-302. PMID: 21975666.
15. Mills JL. Malformations in infants of diabetic mothers. *Teratology* 25:385-94.1982. *Birth Defects Res A Clin Mol Teratol.* 2010 Oct; 88(10):769-78. PMID: 20973049.
16. Al Kaissi A, Klaushofer K, Grill F. Caudal regression syndrome and popliteal webbing in connection with maternal diabetes mellitus: a case report and literature review. *Cases J.* 2008 Dec 19; 1(1):407. PMID: 19099562.



FIGURES

Figure 1 (bottom): Male fetus at 17 weeks of gestation with amniotic band syndrome.

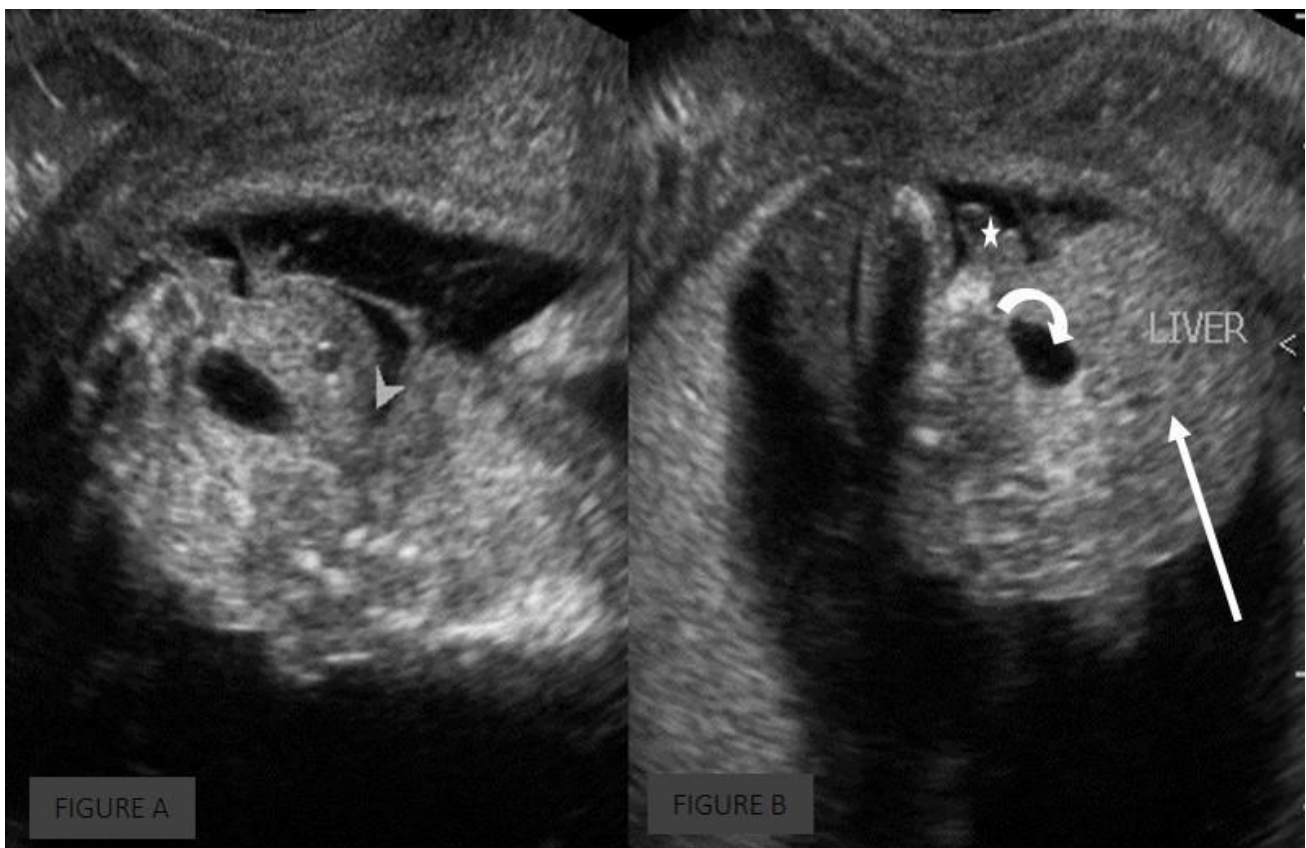
Technique: Antenatal sonogram performed on Philips HD7 ultrasound machine with a transvaginal array transducer (4-8MHz).

Findings: Figure A. Transvaginal grayscale sonogram of the fetus in the sagittal plane reveals presence of amniotic band at thoraco-abdominal junction anteriorly causing a constriction (arrowhead). B. Transvaginal grayscale sonogram of the fetal abdomen in the axial plane demonstrates exteriorisation of liver (long arrow) and stomach (circular arrow). Asterisk in Figure B denotes herniated bowel.

Figure 2 (top): Male fetus at 17 weeks of gestation with amniotic band syndrome.

Technique: Antenatal sonogram performed on Philips HD7 ultrasound machine with a transvaginal array transducer (4-8MHz).

Findings: Transvaginal grayscale sonogram of the fetus in the sagittal plane depicting the location of amniotic bands at thoracic-abdominal junction anteriorly (arrows).



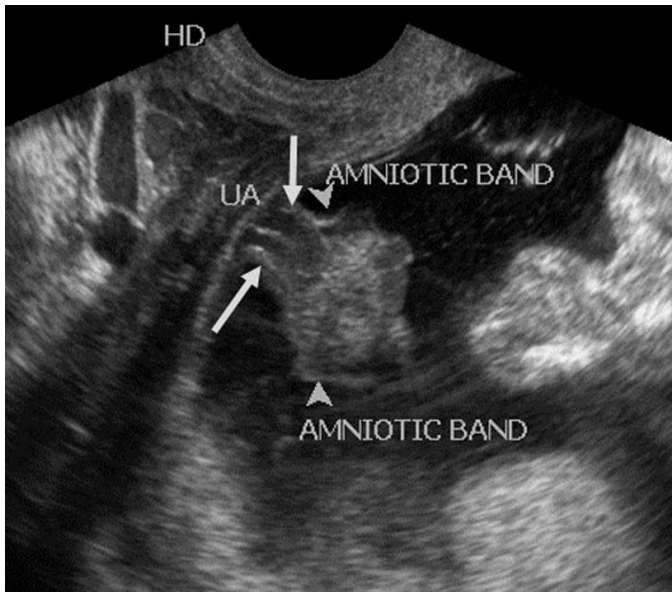


Figure 3: Male fetus at 17 weeks of gestation with amniotic band syndrome.

Technique: Antenatal sonogram performed on Philips HD7 ultrasound machine with a transvaginal array transducer (4-8MHz).

Findings: Transvaginal sonogram depicting the entrapment of the two vessel umbilical cord (white arrows) by the amniotic bands (white arrowheads). Grayscale ultrasound image of the umbilical cord proximal to its placental end demonstrates well the amniotic bands engulfing the umbilical cord and trapping it.

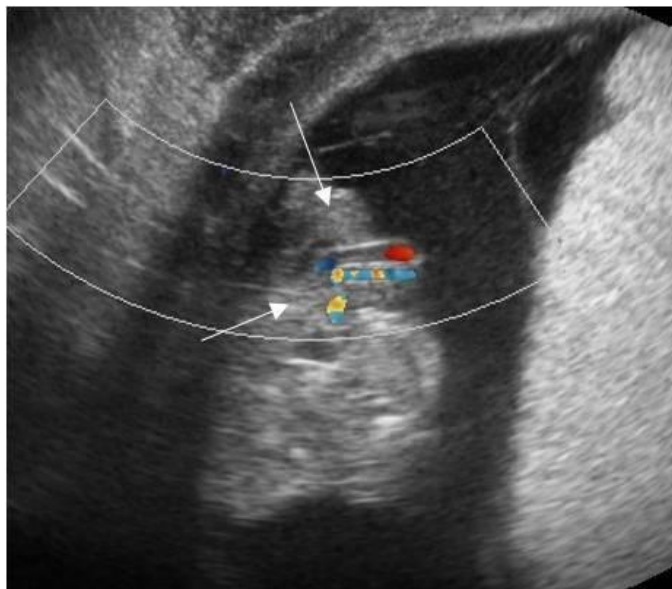


Figure 4: Male fetus at 17 weeks of gestation with amniotic band syndrome.

Technique: Antenatal sonogram performed on Philips HD7 ultrasound machine with a transvaginal array transducer (4-8MHz).

Findings: Transvaginal color Doppler scan of the umbilical cord proximal to its placental end demonstrates well the entrapment of the two vessel umbilical cord (in color) by the amniotic bands (thin arrows).

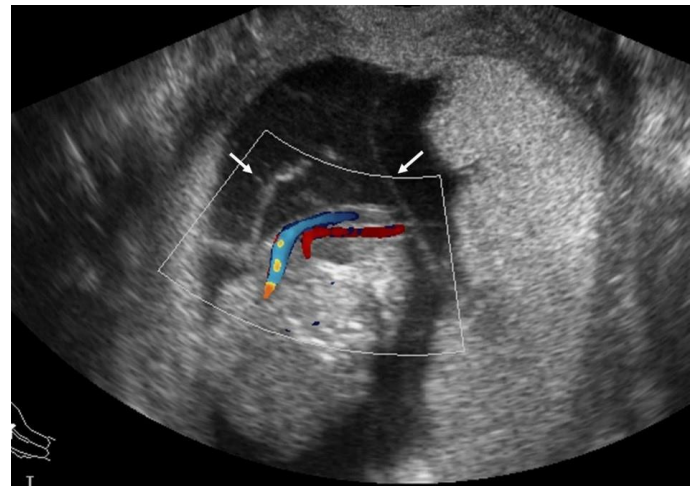


Figure 5: Male fetus at 17 weeks of gestation with amniotic band syndrome.

Technique: Antenatal sonogram performed on Philips HD7 ultrasound machine with a transvaginal array transducer (4-8MHz).

Findings: Transvaginal color Doppler scan of umbilical cord proximal to its placental end depicting umbilical cord with a single umbilical artery (in red) and an umbilical vein (in blue). Amniotic bands are well seen (white arrows).



Figure 6: Male fetus at 17 weeks of gestation with amniotic band syndrome.

Technique: Antenatal sonogram performed on Philips HD7 ultrasound machine with a transvaginal array transducer (4-8MHz).

Findings: Transvaginal grayscale sonogram of the left leg of the fetus in longitudinal plane. The left foot depicts clubfoot deformity (single arrowhead). The left tibia and fibula in long axes (double arrowheads). The right lower extremity was not visualized.



Figure 7: Male fetus at 17 weeks of gestation with amniotic band syndrome.

Technique: Antenatal sonogram performed on Philips HD7 ultrasound machine with a transvaginal array transducer (4-8MHz).

Findings: Transvaginal grayscale sonogram of the fetus in sagittal plane showing anterior angulation of the spine (thin long arrow). The lumbar spine shows an area of apparent defect (short arrows) in the mid sagittal plane, suggestive of rotational deformity of lumbar spine. H denotes fetal heart.

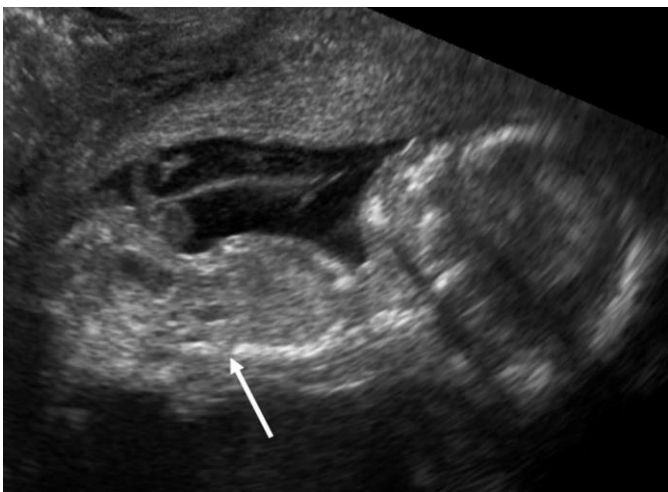


Figure 8: Male fetus at 17 weeks of gestation with amniotic band syndrome.

Technique: Antenatal sonogram performed on Philips HD7 ultrasound machine with a transvaginal array transducer (4-8MHz).

Findings: Transvaginal grayscale sonogram of the fetus in sagittal plane demonstrates abrupt interruption of fetal spine at the lumbar level (arrow).



Figure 9: Aborted fetus with amniotic band syndrome.

Findings: Close up picture of aborted fetus in supine position confirming the antenatal findings of left clubfoot (thin arrow), the absence of right lower extremity, exteriorization of liver (bold arrow) and bowel (asterisk). The placenta is seen to the left of the aborted fetus in the picture.



Figure 10: Aborted fetus with amniotic band syndrome.

Findings: Close up picture of aborted fetus in prone position confirming the rotational deformity (thin arrow) of the lumbar spine and sacral agenesis (bold arrow). The placenta is seen to the left of the aborted fetus in the picture.



Figure 12: Aborted fetus with amniotic band syndrome. Findings: Close up picture of left lower extremity of the aborted fetus confirming the entrapment of the umbilical cord (bold arrow) by the amniotic band (thin arrow) which is wrapped around the left thigh of the fetus.

Figure 11: Aborted fetus with amniotic band syndrome. Findings: Plain X ray of the aborted fetus in supine position depicting rotational deformity of the lumbar vertebral bodies (thick arrow) and evidence of sacral agenesis (thin arrow).

Etiology	Entrapment of fetal parts by disrupted amnion
Synonyms	<ul style="list-style-type: none"> • Amniotic band syndrome • ADAM (Amniotic deformity, adhesion, mutilation) • Amniotic disruption complex • Streeter bands • Congenital constriction bands
Incidence	1 in 11,200 births
Gender predilection	None
Diagnostic Modality	Ultrasound
Ultrasound features	<ul style="list-style-type: none"> • Multiple fetal defects in asymmetric distribution along with amniotic bands • Extremity defects • Craniofacial abnormalities • Anterior abdominal wall defects • Chest wall defects • Scoliosis
Increased incidence of	Premature rupture of membranes, prematurity, low birth weight
Imaging pearl	Always consider amniotic band syndrome in cases with asymmetric fetal defects and evaluate for amniotic bands and assess umbilical cord

Table 1: Summary table of the key aspects and imaging findings associated with amniotic band syndrome.

Amniotic Sheet	<ul style="list-style-type: none"> • Composed of two layers of chorion and amnion • No associated fetal anomalies • No attachment to fetus or umbilical cord
Chorioamniotic separation	<ul style="list-style-type: none"> • Normal in early pregnancy • No fetal part entrapment or associated fetal anomalies • May occur after amniocentesis
Gastroschisis	<ul style="list-style-type: none"> • Anterior abdominal wall defect to the right of midline • Not covered with membrane • Free floating bowel loops • Cardiac and genitourinary anomalies are commonly associated but not limb anomalies • No amniotic bands
Omphalocele	<ul style="list-style-type: none"> • Midline anterior abdominal wall defect covered by peritoneum • Umbilical cord inserts over the defect • Contains bowel and/or liver • No amniotic bands

Table 2: Differential diagnosis table for amniotic band syndrome.

ABBREVIATIONS

ABS = Amniotic band syndrome
SUA = Single umbilical artery

KEYWORDS

Amniotic band syndrome; sacral agenesis; ultrasound; umbilical cord entrapment; amniotic sheet

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