

## CASE REPORT

1. General Practitioner, Universidad de Ciencias Aplicadas y Ambientales U.D.C.A, Resident in Gynecology and Obstetrics Universidad Libre Cali, Colombia
2. General Practitioner, Universidad Libre Seccional Cali, Resident in Gynecology and Obstetrics, Universidad Libre Cali, Colombia
3. General Practitioner, Universidad Militar Nueva Granada; Resident in Gynecology and Obstetrics, Universidad Libre Cali, Colombia
4. Specialist in Maternal Fetal Medicine, National Institute of Perinatology, Postgraduate Professor, Universidad Libre Seccional Cali, Colombia

**Conflict of interest:** The authors declare that they have no conflicts of interest.

**Funding:** The authors declare that there is no funding for this study.

**Ethical considerations:** Informed consent obtained.

**Related technology:** No technology related to artificial intelligence was used.

**Received:** 5 February 2024

**Accepted:** 26 February 2024

**Online publication:** 30 March 2024

**Corresponding author:**

Luisa Fernanda Cano Bautista

📍 Cra. 86 25-83, Cali, Colombia. Código Postal: 764001

☎ (+57) 3232089363

✉ luca14@hotmail.com

**Cite as:** Suárez Camargo MN, Aguirre Martínez MA, Cano Bautista LF, Briceño F. Amniotic band syndrome. *Rev peru ginecol obstet.* 2024;70(1). DOI: <https://doi.org/10.31403/rpgo.v70i2603>

# Amniotic band syndrome

## Síndrome de bandas amnióticas

Mónica Natalia Suárez Camargo<sup>1</sup>, María Alejandra Aguirre Martínez<sup>2</sup>, Luisa Fernanda Cano Bautista<sup>3</sup>, Freddy Briceño<sup>4</sup>

DOI: <https://doi.org/10.31403/rpgo.v70i2603>

### ABSTRACT

Amniotic banding sequence is a congenital disorder caused by early rupture of the amnion resulting in the development of amniotic bands that become attached or entangled in the fetus and generate a spectrum of fetal anomalies (disruption, deformity and/or malformation). Its incidence varies from 1 in 1,200 to 1 in 15,000 live births and would be responsible for 1/70 fetal deaths. The prognosis depends on the extent of the defects, which can range from minor anomalies to lethal ones such as umbilical cord constriction, causing fetal death or spontaneous abortions. We present the case of a patient with prenatal diagnosis of amniotic bands with severe deformities - asymmetry of the skull, exposure of brain tissue, amelia of the upper extremity, malposition of the lower extremities - emphasizing the importance of early diagnosis of this pathology in order to define timely procedures.

**Key words:** Amniotic band syndrome, Ultrasonography, Fetoscopy, Fetal malformations

### RESUMEN

La secuencia de bandas amnióticas es un trastorno congénito dado por la rotura temprana del amnios que resulta en el desarrollo de bandas amnióticas que se adhieren o se enredan en el feto y generan un espectro de anomalías fetales (interrupción, deformación y/o malformación). Su incidencia varía de 1 de cada 1,200 a 1 de cada 15,000 nacidos vivos y sería responsable de 1/70 muertes fetales. El pronóstico depende de la extensión de los defectos, que pueden ser desde anomalías menores hasta letales como la constricción del cordón umbilical, causando muerte fetal o abortos espontáneos. Se presenta el caso de un paciente con diagnóstico prenatal de bandas amnióticas con deformidades severas -asimetría del cráneo, exposición de tejido cerebral, amelia de extremidad superior, malposición de extremidades inferiores-, haciendo énfasis en la importancia del diagnóstico temprano de esta patología para definir procedimientos oportunos.

**Palabras clave.** Síndrome de bandas amnióticas, Ultrasonografía, Fetoscopia, Malformaciones fetales

### INTRODUCTION

Amniotic band syndrome or sequence is a congenital disorder that generates a spectrum of fetal anomalies<sup>(1)</sup> (disruption, deformation and/or malformation) caused by early rupture of the amnion leading to the development of amniotic bands that adhere or become entangled in the fetus, producing either constriction of the fetal organs leading to amputation of the anatomical structure, or adhesion with adverse mechanical defects resulting in malformation or deformation<sup>(2)</sup>.

Its incidence varies from 1 in 1,200 to 1 in 15,000 live births<sup>(3)</sup> and it is responsible for 1/70 fetal deaths<sup>(4)</sup>. It occurs equally in both sexes, without racial predisposition. Sporadically, although rare familial cases have been reported<sup>(5)</sup>.

The most common findings are constriction rings, digital or limb amputation in up to 80% of cases. Atypical anomalies include craniofacial defects (encephalocele, facial clefts), spinal defects and scoliosis<sup>(2)</sup>.

Prenatal diagnosis is made by ultrasound, which may show bands in the amniotic fluid as fine echogenic strands, limb constriction rings with or without distal edema, and craniofacial, chest wall or abdominal deformities, although less common<sup>(3)</sup>.

The prognosis depends on the extent of the defects, from minor anomalies to lethality as in umbilical cord constriction, causing fetal death or



spontaneous abortion. Band release by fetoscopy can prevent limb amputation and fetal death. However, the efficacy of this procedure has been difficult to evaluate because of the rarity of the condition<sup>(6)</sup>.

The case of a pregnant woman whose ultrasound showed multiple fetal malformations associated with the sequence of amniotic bands is described below.

### CASE REPORT

We present the case of an 18-year-old patient, primigravida, with no significant personal, family or toxic history. During the ultrasound evaluation at 15<sup>4/7</sup> weeks, the fetus was found to have an asymmetrical skull, frontal protrusion of brain tissue, right upper amelia and lower limbs with anomaly of position. She was admitted to the third level institution at 20<sup>3/7</sup> weeks, at which time a TORCH profile was requested (syphilis, human immunodeficiency virus, hepatitis B, toxoplasma IgG and IgM, rubella IgG, IgM and cytomegalovirus IgG and IgM), with negative results. The anatomical detail ultrasound by perinatology reported amniotic band involving lower and upper limbs (Figure 1A and 1B), skull with exposure of the encephalic mass (Figure 2A and 2B), agenesis of the right upper limb (Figure 3).

The patient received counseling by the psychology service, with the decision to perform voluntary termination of pregnancy. Genetic study was not requested at the patient's petition. No birth data was obtained.

### DISCUSSION

Amniotic banding sequence is a rare congenital condition of multifactorial origin involving genetic, infectious or environmental processes, being a challenge to determine its exact cause<sup>(7)</sup>. Two hypotheses have been formulated on the formation of amniotic bands: according to the extrinsic model theory there is rupture of the amnion without disruption of the chorion, generating oligohydramnios; the fetus would pass into the extraembryonic coelom and connect with the mesoderm on the chorionic surface, resulting in intertwining of fetal parts and cutaneous abrasions; in Streeter's intrinsic model theory, the anomalies and fibrous bands have a common origin due to early disturbance of the development of the germinal disc of the embryo<sup>(8)</sup>.

There are multiple risk factors for the development of the sequence of amniotic bands, among which the consumption of psychoactive substances such as cocaine -which is a potent vasoconstrictor and affects uteroplacental flow-, tobacco and carbon monoxide stand out<sup>(8)</sup>. In

FIGURE 1. A. OBSTETRIC ULTRASOUND SHOWING RIGHT LOWER LIMB WITH AMNIOTIC BAND LESION AT THE LEVEL OF THE DISTAL AND MIDDLE THIRD WITH INVOLVEMENT OF THE TOES. B. 3D ULTRASOUND VIEW SHOWING DEFORMITY IN THE RIGHT FOOT AND CONSTRICTION RING (BAND) IN THE DISTAL THIRD OF THE IPSILATERAL LEG.

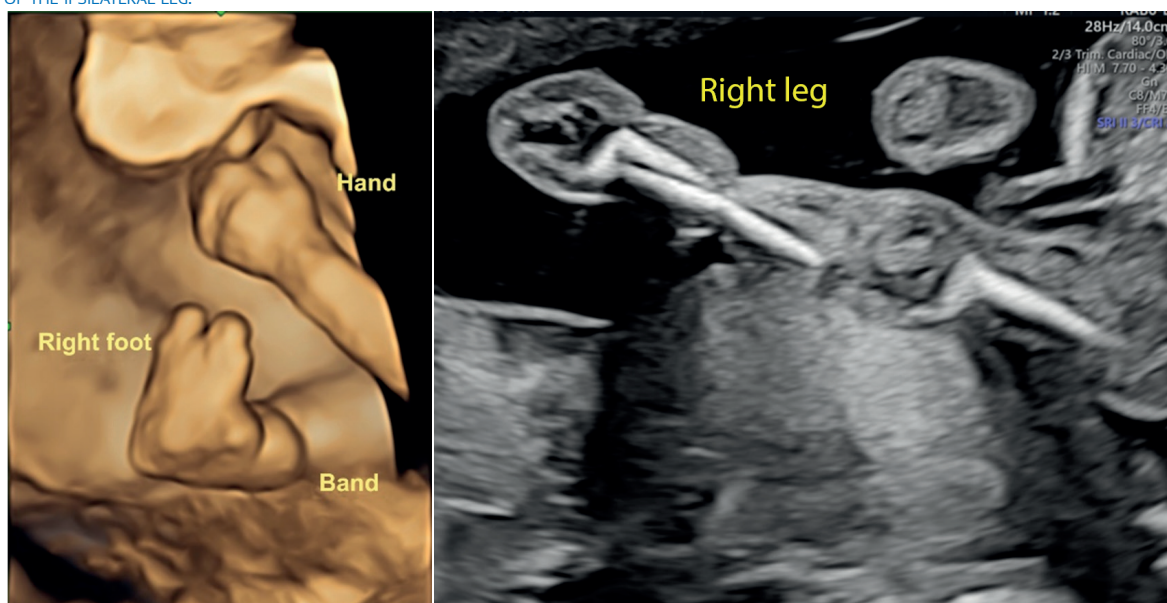
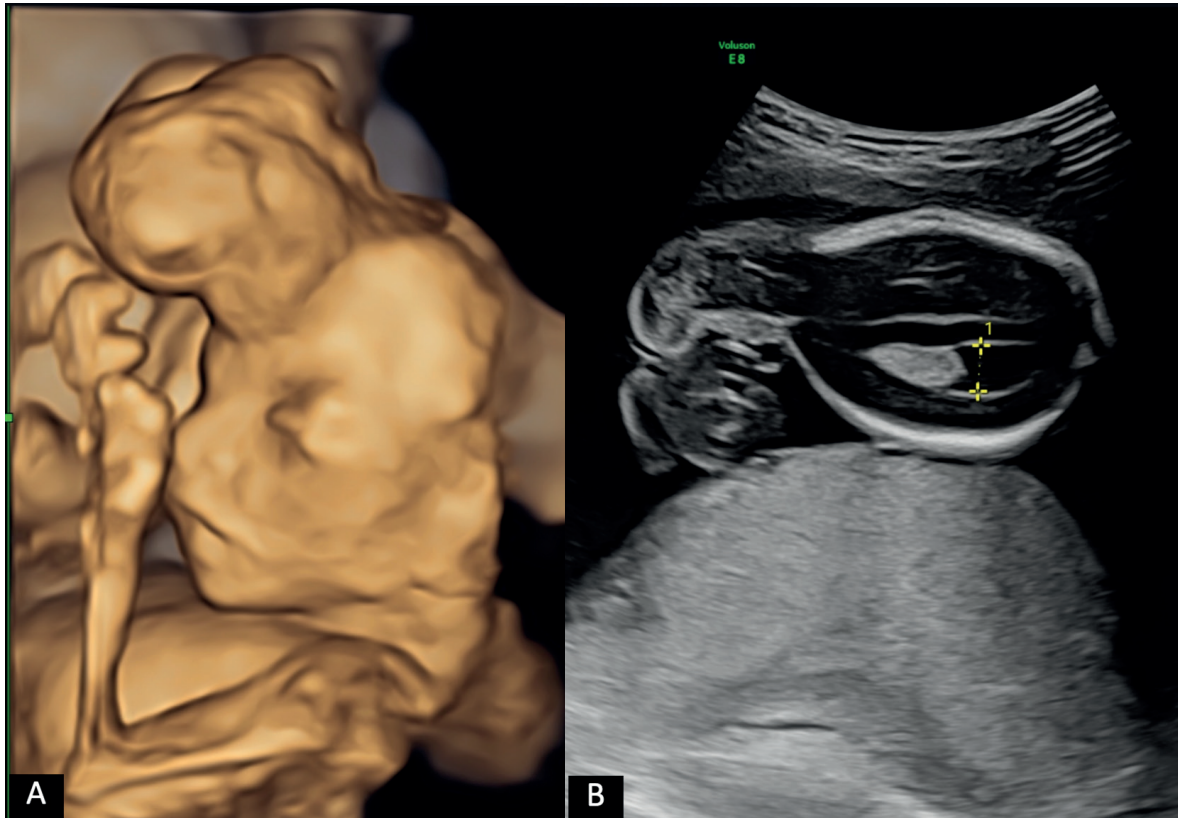




FIGURE 2. A. 3D ULTRASOUND SHOWING ENCEPHALIC MASS EXPOSURE AND ANOMALIES AT THE LEVEL OF THE LEFT UPPER LIMB. B. DISCONTINUITY OF THE BONY TABLE.



the case presented, the patient denied the use of psychoactive substances, alcohol or tobacco. Other associated risk factors are abortion attempts, uterine trauma, bicornuate uterus, procedures such as intrauterine device removal, chorionic villus biopsy or amniocentesis, cerclage or fetoscopy<sup>(8)</sup>. A low socioeconomic and educational level and few prenatal controls were identified for the case. Most cases are not hereditary, although cases of familial recurrence have been published. There was no family history of the problem in the patient.

Regarding the clinical presentation, this sequence encompasses a wide spectrum of manifestations that can range from malformations or amputations in extremities, skull, face, trunk and abdomen, presented in isolation or in any combination, which will determine the prognosis<sup>(8)</sup>.

Lamrissi et al. in 2022 mentioned a 21-year-old patient with a 20-week gestation whose ultrasound showed a single fetus with caudal regression type III, sacral agenesis associated with spina bifida, myelomeningocele, scoliosis,

omphalocele of the liver, spleen, intestine and stomach secondary to the presence of an amniotic band<sup>(7)</sup>. On the other hand, Hoa Pan and collaborators in 2023 contrasted the severity of the lesions in a case of a 38-week fetus with ultrasound scans within normal limits, but at birth with the presence of amputation of the fourth and fifth fingers of the left hand and partial syndactyly, in addition to constriction of the third finger by a fibrous band and severe deformity of the left lower extremity<sup>(9)</sup>.

Ultrasound diagnosis varies depending on the affected part. It can be evidenced as a simple constriction ring, up to severe morphological and functional alterations (craniofacial, trunk, extremities and other areas)<sup>(10)</sup>. Ushakov and Lia published in a series of 28 cases the 4 types of characteristics that may be present for the diagnosis of this entity, such as: amniotic net, division of the amnion, amnion connections, difficulty for fetal mobilization<sup>(11)</sup>. However, in some cases it is not possible to visualize the echoes generated by the bands, especially in early gestations, which makes its diagnosis difficult. Taking this into account, it is important to evaluate both the



FIGURE 3. ULTRASOUND IN 3D VIEW SHOWING RIGHT UPPER LIMB AGENESIS.



mobility and the morphology of the extremities, in order to approach the diagnosis<sup>(10)</sup>.

Additional diagnostic aids, such as fetoscopy, allow to confirm the diagnosis and generate timely interventions in order to eliminate the area of constriction, reestablishing blood flow and thus avoiding the risk of limb amputation or death due to umbilical cord constriction<sup>(12)</sup>.

Gueneuc et al. in 2018 reported a series of 5 cases of fetuses with amniotic band syndrome in limbs and lysis of the bands by laser dissection by fetoscopy, finding in control ultrasound

scans the presence of free limb movements and decreased edema, with a success rate of 75%<sup>(13)</sup>.

After making the diagnosis, it is important to know the involvement of the fetus, indicating the type and location of the anomalies, since the outcome in the fetus in its perinatal state or the incompatibility with extrauterine life depends on this, as occurs when there are craniofacial defects or severe visceral anomalies. Meanwhile, isolated limb malformations can be corrected with surgical management intrauterine or in neonatal life<sup>(7,14)</sup>.

On the other hand, cases have been documented between 1992 and 2018 where intrauterine deaths occur in 74% of cases of amniotic banding sequence, secondary to compression or constriction of the umbilical cord<sup>(10)</sup>. In general, this sequence has been found in one out of every 70 stillbirths according to different case series, as described by Kaloisek in 1988 and Glass in 2010.

The main prognostic factor is the restoration of perfusion, preserving the function of the affected limb in 50%<sup>(17)</sup>. The Doppler examination of the limb plays an important role and, according to the flow compromise (the lower the flow, the lower the possibility of saving the limb), invasive procedures are avoided, due to the high risk of complications such as premature rupture of membranes, as Huser and collaborators point out<sup>(18)</sup>.

Ronderos and collaborators, in 2006, report a successful case of lysis of adhesions in the right lower extremity at 28 weeks of gestation through fetoscopy, with complete reestablishment of the flow in the extremity and adequate functionality at two years of life<sup>(19)</sup>.

For severe alterations, such as central nervous system involvement including the exencephaly found in this case, postnatal mortality is up to 100%<sup>(20,21)</sup>.

In conclusion, amniotic band syndrome is an uncommon pathology, with a wide spectrum of clinical presentation that influences the severity of the picture, which makes its early diagnosis important in order to perform timely interventions.





## REFERENCES

1. Gică N, Apostol LM, Gică C, Huruță I, Vayna AM, Panaitescu AM, Gana N. Amniotic Band Syndrome-Prenatal Diagnosis. *Diagnostics (Basel)*. 2023 Dec 23;14(1):34. doi: 10.3390/diagnostics14010034
2. Singh AP, Gorla SR. Amniotic Band Syndrome. 2022 Dec 11. In: *Stat Pearls* [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK545283/>. PMID: 31424867.
3. Gandhi M, Rac MWF, McKinney J. Amniotic Band Sequence. *Am J Obstet Gynecol*. 2019 Dec;221(6):B5-B6. doi: 10.1016/j.ajog.2019.09.020
4. Munisteri MK, Gonzalez-Brown V, Villazana-Kretzer D. Amniotic band syndrome: A case series. *J Case Rep Images Obstet Gynecol* 2022;8(1):33-8. doi: 10.5348/100122Z08MM2022CS
5. Nardoza LM, Araujo Júnior E, Caetano AC, Moron AF. Prenatal diagnosis of amniotic band syndrome in the third trimester of pregnancy using 3D ultrasound. *J Clin Imaging Sci*. 2012;2:22. doi: 10.4103/2156-7514.95436
6. Minella C, Costantino B, Ruano R, Koch A, Weingertner AS, Favre R, Sananes N. Fetoscopic Release of Amniotic Band Syndrome: An Update. *J Ultrasound Med*. 2021 May;40(5):1039-48. doi: 10.1002/jum.15480
7. Lamrissi A, Moorabbin M, Ouajih O, Jalal M, Fichtali K, Bouhya S. Amniotic band syndrome: A case report. *Int J Surg Case Rep*. 2022 Jun;95:107096. doi: 10.1016/j.ijscr.2022.107096
8. Alfaro Y, Rodríguez LF, Domínguez MM, Molineros Gallón LF, Pintar AX, Portillo-Miño JD. Síndrome de bandas amnióticas: reporte de caso y revisión de literatura. *Pediatría*. 2022;54(Suplemento 1):14-7. doi: 10.14295/rp.v54iSuplemento1.351
9. Phan TH, Thi Nguyen PT, Nguyen PN, Pham HH, Ngo QD, La Nguyen PT. Amniotic band syndrome leading to severe malformations of the newborn: a case report at Tu Du Hospital, Vietnam, and literature review. *Ann Med Surg (London)*. 2023 Mar 24;85(3):592-7. doi: 10.1097/MS9.000000000000263
10. Niu Z, Meng H, Zhang X, Ouyang Y, Zhang Y, Wu X. Two case reports: Early detection of amniotic band syndrome by adhesion between hand and umbilical cord at 11 to 14 weeks' gestation. *Medicine (Baltimore)*. 2019 Dec;98(50):e18302. doi: 10.1097/MD.00000000000018302
11. Ushakov F, Lia C. Amniotic band syndrome: first trimester diagnosis and classification. *Ultrasound Obstet Gynecol*. 2017;50:186. doi: 10.1002/uog.18098
12. Richter J, Wergeland H, DeKoninck P, De Catte L, Deprest JA. Fetoscopic release of an amniotic band with risk of amputation: case report and review of the literature. *Fetal Diagn Ther*. 2012;31(2):134-7. doi: 10.1159/000335026
13. Gueneuc A, Chalouhi GE, Borali D, Mediouni I, Stirnemann J, Ville Y. Fetoscopic Release of Amniotic Bands Causing Limb Constriction: Case Series and Review of the Literature. *Fetal Diagn Ther*. 2019;46(4):246-56. doi: 10.1159/000495505
14. López-Muñoz E, Becerra-Solano LE. An update on amniotic bands sequence. *Archivos Argentinos de Pediatría*. 2018 Jun 1;116(3). doi: 10.5546/aap.2018.eng.e409
15. Glass JM. Fetal decapitation associated with amniotic bands. *J Diagn Med Sonogr*. 2010;26:32-4. doi: 10.1177/8756479309344740
16. Kalousek DK, Bamforth S. Amnion rupture sequence in previable fetuses. *Soy J Med Genet* 1988;31:63-73. doi: 10.1002/ajmg.1320310110
17. Javadian P, Shamshirsaz AA, Haeri S, Ruano R, Ramin SM, Cass D, Olutoye OO, Belfort MA. Perinatal outcome after fetoscopic release of amniotic bands: a single-center experience and review of the literature. *Ultrasound Obstet Gynecol*. 2013 Oct;42(4):449-55. doi: 10.1002/uog.12510
18. Hüsler MR, Wilson RD, Horii SC, Bebbington MW, Adzick NS, Johnson MP. When is fetoscopic release of amniotic bands indicated? Review of outcome of cases treated in utero and selection criteria for fetal surgery. *Prenat Diagn*. 2009 May;29(5):457-63. doi: 10.1002/pd.2222
19. Ronderos-Dumit D, Briceño F, Navarro H, Sanchez N. Endoscopic release of limb constriction rings in utero. *Fetal Diagn Ther*. 2006;21(3):255-8. doi: 10.1159/000091351
20. Ostia-Garza PJ, Luna-Ríos E, Plaza-Benhumea L. Exencefalia: diagnóstico prenatal y reporte de caso. *Perinatol Reprod. Hum*. 2022 Jun;36(1):25-9. doi: 10.24875/per.20000024
21. Sobana M, Halim D, Faried A, Riestiano BE, Pajan HF, Islam AA, Dariansyah AD, Parenrengi MA. Amniotic band syndrome with CNS involvement: a pediatric neurosurgeon's dilemma-a case series and literature review. *Childs Nerv Syst*. 2023 Aug;39(8):2003-17. doi: 10.1007/s00381-023-06008-w