Amniotic band syndrome
Síndrome de bandas amnióticas

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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 severas deformidades - 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 (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.

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

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.

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.

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.
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(6).

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 4/7 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 3/7 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(7). 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(8).

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(8). In
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In 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\(^9\). 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\(^8\).

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\(^7\). 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\(^9\).

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)\(^10\). 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\(^\text{11}\). 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 2. A. 3D ultrasound showing encephalic mass exposure and anomalies at the level of the left upper limb. B. Discontinuity of the bony table.](image-url)
mobility and the morphology of the extremities, in order to approach the diagnosis\(^\text{(10)}\).

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\(^\text{(12)}\).

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\%\(^\text{(13)}\).

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\(^\text{(7,14)}\).

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\(^\text{(10)}\). 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\%\(^\text{(17)}\). 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 Houser and collaborators point out\(^\text{(18)}\).

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\(^\text{(19)}\).

For severe alterations, such as central nervous system involvement including the exencephaly found in this case, postnatal mortality is up to 100\%\(^\text{(20,21)}\).

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


