

# Fetal death due to umbilical cord thrombosis in association with a swallowed amniotic string

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## ABSTRACT

We report the exceptional case of an intrauterine fetal death due to the umbilical vein thrombosis in association with a swallowed amniotic string and other pathological features of the umbilical cord. The case concerns a full-term pregnancy, the woman was just performing routinely antepartal CTG monitoring and, unexpectedly, no fetal heart beat could be recorded. An ultrasound check was immediately performed and confirmed the diagnosis of intrauterine fetal death. After labor induction a female fetus was delivered, no macroscopic external sign of malformation, except a thin fibrotic string hanging from the mouth was noticed. The following pathologic examination of fetus, placenta and umbilical cord showed an unlucky combination of adverse variables contributing all together to the fetal demise. In fact the umbilical cord presented a thrombus in the venous vessel, an absolute shortness, an eccentric insertion, and an amniotic band coiled around its origin. Finally it was confirmed the amniotic origin of the string, total length of 53,5 cm, hanging from the mouth, with its deepest part in the stomach.

Keywords: swallowed amniotic string, amniotic band syndrome, umbilical vein thrombosis, fetal hypoxia, fetal demise.

## CASE REPORT

A 28-year-old primigravida, at 40+2 weeks arrived to our observation to perform her second routine antepartal CTG monitoring; few minutes later was admitted to our Department with diagnosis of intrauterine fetal demise. The pregnancy had a regular follow-up, fetal growth around 40% percentile and no anamnestic complication emerged. The woman had no risk factors for diabetes, hypertension or other metabolic, genetic, familiar disorders. At 12-weeks gestation the woman performed the first trimester biochemical testing (blood concentration of free beta-hCG and PaPP-A) combined with the ultrasound marker of fetal Nuchal Translucency thickness for the screening for Trisomy 21, 13 and 18. Calculated risk resulted in a low risk of trisomy, therefore no prenatal invasive diagnosis was carried out. Serial ultrasound scans confirmed a regular development of the pregnancy. At 40-weeks gestation the first routine CTG monitoring was started, as suggested by our National Protocol for routine care of physiological pregnancy and it was concluded

## SOMMARIO

Noi riportiamo il caso eccezionale di una morte fetale intrauterina causata dalla trombosi della vena ombelicale in associazione con l'ingestione di una briglia amniotica ed altre anomalie del cordone ombelicale. Il caso riguarda una gravidanza a termine, la donna stava eseguendo il monitoraggio cardiocografico di routine quando inaspettatamente non venne riscontrato alcun battito fetale. Immediatamente venne eseguita una ecografia che confermò la diagnosi di morte intrauterina. Dopo l'induzione del travaglio venne partorito un feto di sesso femminile ma non venne rinvenuta nessuna anomalia macroscopica, ad eccezione di una briglia fibrosa che protrudeva dalla bocca. L'esame anatomopatologico del feto, della placenta e del cordone ombelicale mostrarono una combinazione sfortunata di variabili avverse che contribuirono tutte insieme alla morte fetale. Infatti il cordone ombelicale presentava un trombo all'interno del vaso venoso, una brevità assoluta, una inserzione eccentrica ed una banda amniotica intorno alla sua origine. Infine venne confermata l'origine amniotica della briglia, per una lunghezza totale di 53,5 cm, che fuoriusciva dalla bocca mentre la sua parte più profonda venne rinvenuta nello stomaco.

as reactive. Two days later, starting the second CTG monitoring no fetal heart beat could be recorded. An ultrasound scan was immediately performed and confirmed the diagnosis of intrauterine fetal death. The woman, shocked by the news, reported to have felt a lot of active fetal movements during the last night, so strong and frequent to remain awake whole night. Only in the late morning such movements would become weaker, softer, but always present according to her perception. After admission to our Department of Obstetrics, labour was induced with prostaglandins. Few hours later a female fetus, 2850 g, Apgar 0, was delivered presenting a suggestive thin fibrotic string protruding from her mouth for about 5 cm. No other apparent macroscopic external malformation was detected. The neonatologist tried to remove it, but the tentative failed because of the considerable resistance, as it seemed to be stuck in the throat. The pathologic examination established the amniotic origin of the string, of total length 53,5 cm, and its end was found in the stomach.

The pulmonary examination detected a massive congestion, the alveoli showed squamous cells and granules of meconium referable to acute state

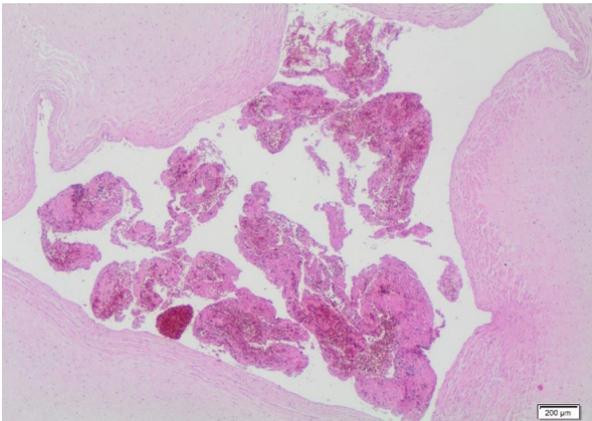
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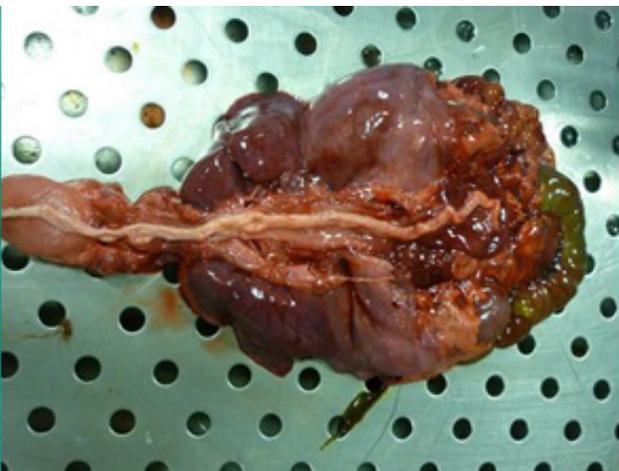
of hypoxia. The umbilical cord was characterized by an absolute shortness (28 cm), and an eccentric insertion. In addition, a mild umbilical cord coiling and an amniotic band, 135x80 mm, around its origin were described (Figure 1). The histologic examination showed a thrombus in the umbilical vein and it was considered the cause of the fetal demise (Figure 2).



**Figure 1:** The origin of the amniotic band around the umbilical cord.



**Figure 2:** Microscopic examination of umbilical vein thrombosis (H&E 200 μm).



**Figure 3:** The swallowed amniotic string (left) and the same found in the stomach (right).

Furthermore, the examination of the placenta confirmed the presence of an amniotic band and its division into two parts: one still attached to chorion surface while the other had rolled up into a string and then was swallowed by the fetus (Figure 3).

## DISCUSSION

We report a fetal demise supposed due mainly to the thrombosis of the umbilical vein, associated with a swallowed amniotic string and with an amniotic band coiling around an absolutely short umbilical cord. No sign of strangulation of the umbilical cord was anyway described by the pathologist. Although the literature has already described cases regarding umbilical vein thrombosis and the swallowing of an amniotic string by the fetus, their association in a singular case has not yet been described, more over in association with a band around the origin of an absolutely short umbilical cord. We can not establish which event first appeared, whether the thrombosis or the development of the amniotic string. However, the string and its origin, the amniotic band around the insertion of the umbilical cord, have been included in the chapter of the amniotic band syndrome.

The amniotic bands are generally in relationship with many different anomalies: craniofacial (encephalocele, exencephaly, clefts), body wall (parts of abdominal or thoracic organs can herniate through the body wall defect and into the amniotic cavity), limb (amputation, syndactyly, clubfoot, constriction rings with possible development of distal lymphedema, hand deformities), visceral (lung hypoplasia), spinal defects, scoliosis, ambiguous genitalia and umbilical cord strangulation<sup>(1)</sup>.

In addition to this wide spectrum of congenital anomalies related to the amniotic band syndrome we should also include the possible formation of an

amniotic string, like a rolled up amniotic membrane.

The pathogenesis of this syndrome remains unclear; some authors attribute it to an early amnion rupture<sup>(2)</sup>, considering two possibilities: on the one hand a significant abdominal trauma during pregnancy, on the other hand a possible relationship between an invasive procedure, such as amniocentesis, fetoscopy and the rupture of the amnion (known as "pseudoamniotic band syndrome"<sup>(3-4)</sup>); others refer it to a vascular disruption<sup>(5)</sup> or a genetic defect of the embryo<sup>(6-7)</sup>. Most cases are sporadic but some authors suggest a multifactorial or polygenic cause<sup>(8)</sup>.

This last hypothesis is supported by the recurrence risk of amniotic band syndrome in monozygotic twins. After having analyzed some cases in literature reporting a swallowed amniotic string by the fetus, we can affirm that this condition occurs very seldom and it is not easy to predict the fetal prognosis. In the literature, Torpin et al reported a case of a normal male infant with a fibrous string extending from his mouth. The doctors removed it, length of 94 cm, from the gastrointestinal tract while its end was still attached to placental disc<sup>(8)</sup>. In this case the newborn had a favorable outcome<sup>(9)</sup>. Torpin tried to explain this situation referring to a premature rupture of the amnion and illustrated three possibilities<sup>(9)</sup>. In the first case the amnion may remain a sac or roll up into a string with its end free or attached to the chorionic surface<sup>(9)</sup>.

The second possibility considers that the rupture of amnion may generate sheets or strands and in the third case some strings are already detached while the amnion separates it self from the chorionic wall<sup>(9)</sup>.

The first hypothesis seems to fit perfectly to our clinical case, as like the detached part of the amniotic band, completely free from its origin, had rolled up into a string. Although it remains difficult to establish the fetal outcome we suggest that the isolated presence of a swallowed amniotic string, does not necessarily determinate an adverse fetal outcome.

In order to emphasize this hypothesis we have also considered beside Torpin's case, the experiences of Gramling<sup>(9)</sup>, Jahoda and Schaller<sup>(10)</sup>, which also assisted at the birth of healthy children with a strand protruding from their mouth.

In spite of these positive experiences, Boughizane et al reported a swallowed amniotic string by a fetus at term and its adverse outcome<sup>(11)</sup>. In this case the fetal death was due to the strangulation of the umbilical cord. Also in this case the amniotic string, 90 cm, was swallowed by the fetus into the small intestine<sup>(11)</sup>.

The strangulation of the umbilical cord by an amniotic band occurs in 10% of the amniotic band sequence cases and reports a high mortality rate<sup>(1,12-15)</sup>. Even if in our case there was not detected a

strangulation of the umbilical cord, the presence of an amniotic band around the insertion of the umbilical cord, the shortness and the mild coiling of it self, may have led to a slowdown or in the end to a blood flow stasis in the fetal circulation. This condition would have than facilitated the occurrence of the thrombus in the umbilical vein. Although the etiology of umbilical cord thrombosis is not exactly known, Heifetz presented a possible explanation: a short cord can stretch the vessel, than the vasoconstriction favorites the thrombosis<sup>(16)</sup>. Umbilical cord vessels thrombosis (TUV) is a rare event, Heifetz et al reported the following incidence: 1:1300 deliveries, 1:1000 perinatal autopsies and 1:250 high-risk pregnancies<sup>(6)</sup>.

Nevertheless, Avagliano et al noted, in their retrospective study, a considerable discrepancy of TUV incidence: 1:10 in stillbirth, 90 times higher than Heifetz's study<sup>(17)</sup>. This difference was attributed to many causes: the analysis included only stillbirth, the average gestational age was different, pregnancies with malformations or genetic abnormalities were excluded and also other factors in relationship with the histologic examination were not considered<sup>(17)</sup>. Heifetz's analysis reported 52 cases of thrombosis occurring 85% in the umbilical vein and 15% in the umbilical arterial<sup>(16)</sup>. Benirschke et al, also confirmed this more remarkable frequency in the vein than in the arterial umbilical vessels<sup>(18)</sup>. Generally the presence of TUV is associated with high risk of perinatal morbidity and mortality<sup>(16, 19-21)</sup>. Even if Heifetz's study noted that arterial umbilical thrombosis caused a worse fetal outcome than umbilical vein thrombosis<sup>(16)</sup>. The definition of the onset time of umbilical cord thrombosis, if it occurs before or after fetal death, is crucial to establish a connection between the thrombus and the clinical complications of the pregnancy<sup>(22)</sup>. The hypothesis that the thrombus determined a severe fetal hypoxia is supported by the presence of many movements felt by the woman before the fetal death and this is what happened in our clinical case. The same event was described in a twin gestation, in which a fetus died due to umbilical vein thrombosis<sup>(23)</sup>. We agree with the conclusion of Baxi et al, the strong fetal activity might be related to the increasing of fetal distress due to the decrease of blood flow in the fetal circulation.

## CONCLUSION

We have reported a singular case of fetal demise characterized by the simultaneous presence of a swallowed amniotic string, hanging from the mouth and ingurgitated up to stomach, and the thrombosis of the umbilical vein, in addition to an amniotic band around a short umbilical cord.

In the literature were reported cases of swallowed amniotic strings and umbilical vessels thrombosis but their association in a single case was until now not described. Although we can not establish nor which event occurred first, neither the singular role played by every adverse factor, the fetal demise was probably related to the umbilical vein thrombosis. We emphasize this speculation considering the strong fetal activity (signal of fetal distress), reported by the woman during the night before the detection of the fetal death and the anomalies of the umbilical cord. As reported in Heifetz' study<sup>(16)</sup> the presence of umbilical vein thrombosis not associated with other cord anomalies might have a better outcome. In fact in our experience as well as in Baxi's case, also a shortness and a peripheral insertion of the umbilical cord were detected. For this reason, we emphasize that these factors (the shortness, the eccentric insertion and the amniotic band surrounding the origin) may have

been determinant contributory causes of the poor fetal outcome.

They probably accentuate the reduction of blood flow, favouring the obstruction of the vessels causing the formation of blood clots and events chain up to an acute hypoxemic event.

The presence of the amniotic band probably played a supplementary role in facilitating this process especially if we consider its origin, but we have also to admit that a single presence of an amniotic string swallowed by the fetus is not directly involved with the fetal death.

The contemporary coexistence of all these adverse features encourages the network among science, philosophy and statistics, suggesting that also for the complex, imponderable, only partially known variables of the prenatal world applies the inevitable concept of the "unlucky fetus".

## REFERENCES

- 1) Barros M, Gorgal G, Machado A.P, Ramalho C, Matias A, Montenegro N: **Revisiting Amniotic Band Sequence: A Wide Spectrum of Manifestations**. Fetal Diagn Ther 2014;35:51-56.
- 2) Torpin R: **Amniochorionic mesoblastic fibrous strings and amniotic bands: associated constricting fetal malformations or fetal death**. Am J Obstet Gynecol 1965;91:65-75.
- 3) Winer N, Salomon LJ, Essaoui M, et al. **Pseudoamniotic band syndrome: a rare complication of monochorionic twins with fetofetal transfusion syndrome treated by laser coagulation**. Am J Obstet Gynecol 2008; 198:393.e1.
- 4) Rujiwetpongstorn J, Tongsong T. **Amniotic band syndrome following septostomy in management of twin-twin transfusion syndrome: a case report**. J Perinatol 2008; 28:377.
- 5) Van Allen MI, Curry C, Gallagher L: **Limb body wall complex. I. Pathogenesis**. Am J Med Genet 1987;28:529-548.
- 6) Hunter AG, Seaver LH, Stevenson RE: **Limb body wall defects. Is there a defensible hypothesis and can it explain all the associated anomalies?** Am J Med Genet A 2011;155A:2045-2059.
- 7) Streeter G: **Focal deficiencies in fetal tissues and their relation to intrauterine amputation**. Contrib Embryol 1930;22:3-44.
- 8) Maligner G, Rosen N, Achiron R, Zakut H: **Pierre Robin sequence associated with amniotic band syndrome ultrasonographic diagnosis and pathogenesis**. Prenat Diagn 1987;7:455-459.
- 9) Torpin R, Goodman L, Gramling ZW: **Amnion string swallowed by the fetus**. Am J Obstet Gynecol 1964 Nov 15;90:829-31.
- 10) Jahoda E, Schaller A: **Amniotic thread swallowed by the fetus**. Zentralblatt für Gynäkologie 1972;94:1229-1232.
- 11) Boughizane S, Zhioua F, Jedoui A, Kattech R, Gargoubi N, Srasra M, Ben Romdhane K, Meriah S: **Swallowing of an amniotic string by a fetus at term**. J Gynecol Obstet Biol Reprod 1993;22(4):409-10.
- 12) Chatzigeorgiou K, Theodoridis T, Efstratiou I, Athanasiadis A, Zepiridis L, Tzeveleki F, Tarlatzis B: **Strangulation of the umbilical cord by an amnion band - a rare cause of intrauterine demise: a case report**. Cases J 2009;2:9108.
- 13) Lurie S, Feinstein M, Mamet Y: **Umbilical cord strangulation by an amniotic band resulting in a stillbirth**. J Obstet Gynaecol Res 2008;34:255-257.
- 14) Heifetz SA: **Strangulation of the umbilical cord by amniotic bands: report of 6 cases and literature review**. Pediatr Pathol 1984;2:285-304.
- 15) Larciprete G, Montagnoli C, Fusco P: **Severe Fetal Distress and Umbilical Cord Strangulation**. Case Reports in Medicine; 2011:645487.
- 16) Heifetz SA: **Thrombosis of the umbilical cord: Analysis of 52 cases and literature review**. Pediatr Pathol 1988;8:37-54.

- 17) Avagliano L, Marconi AM, Candiani M, Barbera A, Bulfamante G: **Thrombosis of the umbilical vessels revisited. An observation study of 317 consecutive autopsies at a single institution.** *Human Pathology* 2010;41:971-979.
- 18) Benirschke K, Kaufmann P, Baergen RN. **Pathology of the human placenta.** 5th ed. Springer;2006. P. 426-8.
- 19) Singh V, Khanum S, Singh M. **Umbilical cord lesions in early intrauterine fetal demise.** *Arch Pathol Lab Med* 2003;127:850-3.
- 20) Byrd L, Wells S, Mayers F. **Umbilical cord thrombosis - a cause of intrauterine demise?** *J Obstet Gynaecol* 2000;20:52.
- 21) Sato Y, Benirschke K. **Umbilical arterial thrombosis with vascular wall necrosis: clinicopathologic findings of 11 cases.** *Placenta* 2006;27:715-8.
- 22) Parast MM, Crum CP, Boyd TK. **Placental histologic criteria for umbilical blood flow restriction in unexplained stillbirth.** *Human Pathology* 2008;39:948-53.
- 23) Baxi LV, Daftary A, Loucopoulos A. **Single fetal demise in a twin gestation: Umbilical vein thrombosis.** *Gynecol Obstet Iizvest* 1998; 46: 266-267.
- Oncol* 21:504-509, 1995, 18.