

CASES REPORT

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Absence of Wharton's jelly Ausencia de gelatina de Wharton

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ABSTRACT

Absence of Wharton's jelly is a rare entity in which the umbilical cord vessels are partially or completely separated from the umbilical cord. It is associated with important fetal morbidity and mortality. Since its first report in 1961, only 12 cases have been documented to date. We present two cases of this entity. The first one occurred in a 21-year-old primigesta whose ultrasound study showed a nuchal cord and oligohydramnios. A 2,620 g male neonate was born and died a few hours later. The second case was in a 31-year-old multigesta with a history of HIV positivity and abortion. She presented with a 375 g male fetus with left renal agenesis and abdominal and cerebral hemorrhage. In these cases, a 9 cm and 7.8 cm segment of umbilical artery was found, respectively, separated from the umbilical cord.

Key words: Umbilical cord, anomaly, Wharton jelly, absence, Stillbirth

RESUMEN

La ausencia de la gelatina de Wharton es una entidad rara que consiste en que los vasos del cordón umbilical se separan del cordón parcial o totalmente. Está relacionada con importante morbimortalidad fetal. Desde su primera comunicación en 1961 a la fecha solo se han documentado 12 casos. Presentamos dos casos de esta entidad. El primero ocurrió en una primigesta de 21 años en cuyo estudio de ultrasonido se halló circular de cordón y oligohidramnios. Nació un neonato masculino de 2,620 g que falleció a las pocas horas. El segundo caso fue en una multigesta de 31 años con antecedente de positividad a VIH y aborto. Ella presentó un óbito fetal, masculino, de 375 g, con agenesia renal izquierda y hemorragia abdominal y cerebral. En estos casos se halló segmento de arteria umbilical de 9 cm y 7,8 cm, respectivamente, separado del cordón umbilical.

Palabras clave. Cordón umbilical, anomalía, Gelatina de Wharton, ausencia, Mortinato

INTRODUCTION

The umbilical cord (UC) is responsible for maternal-fetal blood flow, and its various abnormalities are associated with significant perinatal morbidity and mortality⁽¹⁾. These are accidents such as prolapsed or circular cord or true knot, structural anomalies of length, thickness, implantation and torsion, and inflammatory processes, vascular anomalies, embryonic remains, tumors or alterations in Wharton's jelly (WJ)⁽²⁾.

Absence of Wharton's jelly (AWJ) is an alteration of the umbilical cord in which one or two of its vessels, mainly arteries, are totally separated from the UC in a segmental or total manner. The first case was reported by Bergman in 1961⁽³⁾, who described it as mucoïd degeneration of Wharton's jelly. Thomson⁽⁴⁾, in 1996, called it 'linear disruption of the umbilical cord' and Laberrere⁽⁵⁾, in 1985, named it 'absence of Wharton's jelly'.

Wharton's jelly is a specialized mesenchymal tissue that surrounds and protects the vessels of the umbilical cord. Its 'absence' or decrease is associated with an increased risk of intrauterine growth restriction, fetal distress in labor or perinatal morbidity and mortality⁽⁶⁾.

Wharton's jelly is constituted of a tissue derived from the extraembryonic mesoblast⁽⁷⁾ formed by an extracellular matrix rich in mucopolysaccharides, particularly hyaluronic acid and chondroitin sulfate⁽⁸⁾. Within it are found myofibroblasts⁽⁹⁾ that have contractile function⁽¹⁰⁾, some mast cells or macrophages, intercellular bundles of collagen type I, III and VI⁽⁸⁾ and laminin, distributed in a delicate network of microfibrils⁽⁹⁾. It does not contain lymphatic vessels.

The WJ regulates the turgor of the umbilical cord and surrounds the fetal vessels replacing the adventitia, protecting them from compression, twisting or bending⁽¹⁰⁾ that may occur during fetal movements. Its main function is the protection of the umbilical vessels, neutralizing the influence of external pressure on the blood flow between the placenta and the fetus.

REPORT OF CASES

Case 1: A 21-year-old primigesta arrived at the emergency room in labor and with symptoms of preeclampsia. An ultrasound showed simple circular cord and oligohydramnios. The laboring woman gave birth to a 2,620 g male infant with Apgar 1 at one minute and 2 at 10 minutes. He was taken to the ICU and died a few hours later; no necropsy was performed. The placenta

weighed 450 g, measured 18 x 14.5 x 1.5 cm and the umbilical cord 66 x 1.6 cm. At 20 cm from the placental insertion, the umbilical artery was observed separated at a length of 9 cm. In addition, it showed a small ulcer of 0.9 x 0.4 cm and near the fetal insertion, two foci of hematomas; these were observed at the time of delivery (Figure 1).

Case 2: A 31-year-old multigesta woman G=6, P4014 with a history of HIV positivity and one miscarriage, arrived at the emergency room with symptoms of imminent abortion and delivered an aborted fetus. The placenta weighed 175 g, measured 11 x 9 x 2 cm and the umbilical cord 23 x 1 cm. At 13 cm from the distal end there was a 7.8 cm segment with separation of an artery (absence of WJ). The male fetus weighed 375 g and at necropsy left renal agenesis and hemorrhage of the abdominal and cerebral cavity were found.

FIGURE 1. CASE 1. 1) ABSENCE OF WHARTON'S JELLY. 2) HEMATOMAS OF THE UMBILICAL CORD. 3) ULCER IN THE UMBILICAL CORD.

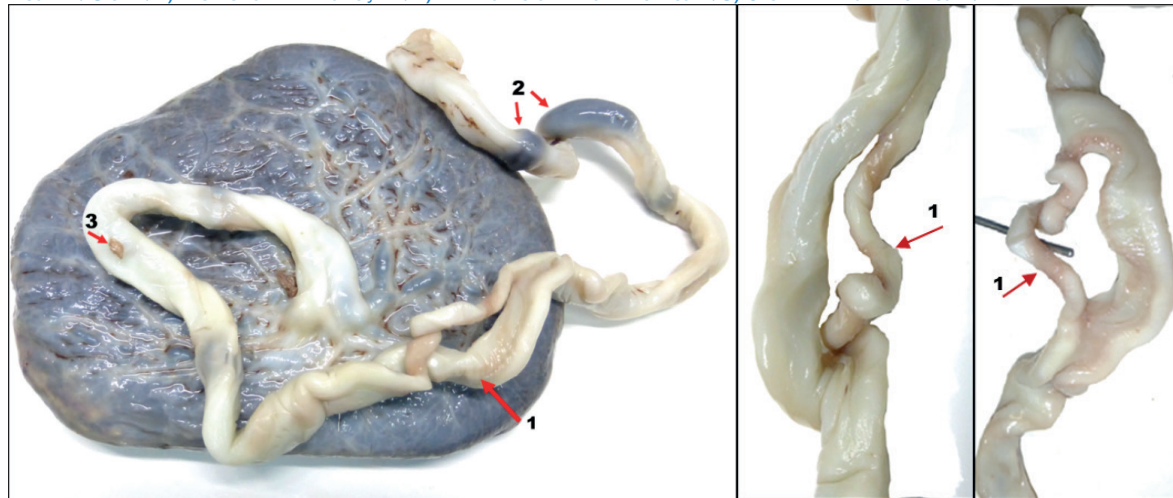
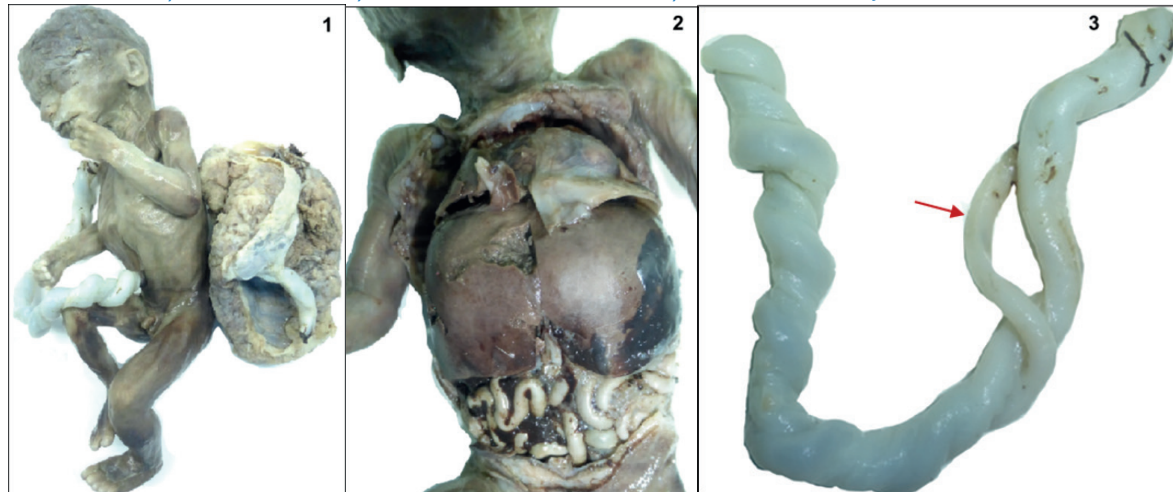


FIGURE 2. CASE 2. 1) FETUS AND PLACENTA. 2) FETUS WITH ABDOMINAL HEMORRHAGE. 3) ABSENCE OF WHARTON'S JELLY.





DISCUSSION

Absence of Wharton's jelly is a very rare event. Kulkarni⁽¹¹⁾ in an Indian hospital between 1965-2006 found one case in 7,000 deliveries per year on average. A familial relationship has not been reported and its recurrence must be very small⁽⁴⁾. The etiology is unknown, but several hypotheses are postulated, such as mucoid degeneration of the WJ around the vessels⁽³⁾, incomplete fusion of the amniotic sheath and umbilical mesenchyme during early development⁽⁴⁾, hypoplasia of the amniotic membrane with secondary loss of the WJ^(5,6), a more severe form of UC cyst formation⁽⁴⁾ or the result of proteolytic digestion of the amniotic membrane around the cord by meconium enzymes, as meconium-stained amniotic fluid has been found in many cases⁽⁴⁾.

To date, 12 cases have been published, and with our two cases, this would make 14 cases. Table

1 summarizes the maternal, fetal and placental characteristics.

In 2021, Botezatu⁽¹⁴⁾ compiled 10 cases of AWJ, but in his publication he incorporated one published by Damasceno⁽¹⁶⁾, who exposed images of the case and described it as 'a marked reduction of the diameter in a segment of 1 cm in length'. This does not correspond to AWJ, but to a constriction of the UC seen in macerated fetuses. Mainly for this reason it has not been considered in our review.

In these 14 cases, maternal age ranged from 19 to 42 years, 9 were primigravidae, 2 multiparous had a history of previous perinatal mortality, 50% had cesarean section and two had a history of positive HIV. In the case of the fetuses, 10 were male, one was unknown, 8 (67%) had perinatal mortality, 3 were underweight, 3 were born with congenital malformations and one case was the product of in vitro fertilization.

TABLE 1. MATERNAL AND FETAL CHARACTERISTICS OF 14 CASES OF WHARTON'S JELLY ABSENCE.

Author and year	N°	Mother				Placenta	Fetus or neonate			
		Parity	Age	Antecedent	Delivery		Weight	Weight/ Sex	Result	Findings
Bergman ⁽³⁾ 1961	1	GI	25	-	Vaginal	NS	2270 g/NS	Stillbirth	Meconium aspiration Intrauterine asphyxia	
Laberrere ⁽⁵⁾ 1985	3	GI	25	-	C-section	560 g	3.220 g/M	Deceased newborn	Meconium aspiration	
		GI	30	-	C-section	500 g	4.100 g/M	Deceased newborn	Meconium aspiration	
		G2P0	25	-	Vaginal	405 g	2.920 g/M	Stillbirth	Meconium-stained amnion	
Thomson ⁽⁴⁾ 1996	1	NS	NS	NS	Vaginal	515 g	3.450 g/F	Disabled girl	Microcephaly Psychomotor delay	
Kulkarni ⁽¹¹⁾ 2007	1	GI	19	HIV+	Vaginal	500 g	2.500 g/M	Morbid child	Patent vitelline duct	
Oliveira ⁽³⁾ 2014	1	GI	22	-	Vaginal	385 g	NS/NS	Stillbirth	Acute funisitis Chorioamnionitis 3	
Cole ⁽¹⁰⁾ 2016	1	Multiparous	31	NS	C-section	NS	3.285 g/M	Healthy child	Fetal distress	
Trivedi ⁽⁶⁾ 2020	1	G3P1+1	22	C-section Neonatal death	C-section	350 g	2.000 g/M	Deceased newborn	Neonatal ICU	
Murphy ⁽¹²⁾ 2020	1	GI	42	IVF	C-section	739 g	3.680 g/F	Live newborn	Neonatal ICU Meconium aspiration	
Botezatu ⁽¹⁴⁾ 2021	1	GI	29	-	C-section	NS	3.500 g/M	Live newborn	Abnormal fetal cardiac tracing and prolonged late deceleration	
Wade ⁽¹⁵⁾ 2022	1	GI	34	-	C-section	NS	3.150g/M	Live newborn	Nuchal cord Oligohydramnios	
Romero 2023	2	GI	21	-	Vaginal	450 g	2.620 g/M	Deceased newborn	Nuchal cord Oligohydramnios Neonatal ICU	
		G6 P4O14	31	HIV +, abortion	Vaginal	175 g	375 g/M	Stillbirth	Left renal agenesis. Abdominal and cerebral hemorrhage	

NS: not specified, M= male, F= female, IVF= in vitro fertilization



Studies on Wharton's jelly are scarce in the literature. It represents an open field of research to formulate correlations with obstetric problems, as well as the establishment of pathogenic mechanisms for some groups of stillbirths⁽¹³⁾.

In conclusion, AWJ is a poorly recognized entity and the shortening of the time between one publication and another is observed. A careful study of the umbilical cord should be performed in the prenatal ultrasound to avoid adverse fetal outcomes, and in the pathology to perform studies to clarify the unknowns of this entity.

REFERENCIAS BIBLIOGRÁFICAS

1. Tantbirojn P, Saleemuddin A, Sirois K, Crum CP, Boyd TK, Tworoger S, Parast MM. Gross abnormalities of the umbilical cord: related placental histology and clinical significance. *Placenta*. 2009;30(12):1083-8. DOI: 10.1016/j.placenta.2009.09.005
2. Stanek J. Association of coexisting morphological umbilical cord abnormality and clinical cord compromise with hypoxic and thrombotic placental histology. *Virchows Arch*. 2016;468(6):723-32. DOI: 10.1007/s00428-016-1921-1
3. Bergman P, Lundin P, Malmstrom T. Mucoid degeneration of Wharton's jelly: An umbilical cord anomaly threatening fetal life. *Acta Obstet Gynecol Scand*. 1961;40:372-8. DOI: 10.3109/00016346109159935
4. Thomson LL, Hoo JJ. Linear disruption of umbilical cord: a rare anomaly of the cord associated with acute fetal distress and perinatal death/profound psychomotor retardation. *Am J Med Genetics*. 1996;62:348-9. DOI: 10.1002/(SICI)1096-8628(19960424)62:4<348::AID-AJMG5>3.0.CO;2-N
5. Labarrere C, Sebastiani M, Siminovich M, Torassa E, Althabe O. Absence of Wharton's jelly around the umbilical arteries: An unusual cause of perinatal mortality. *Placenta*. 1985;555-9. DOI: 10.1016/s0143-4004(85)80010-2
6. Trivedi S, Ratanoo L, Purohit S, Rastogi P. Absence of Wharton's jelly: an association with feto-maternal morbidity. *Int J Reprod Contracept Obstet Gynecol*. 2020 Mar;9(3):1318-20. DOI: <https://doi.org/10.18203/2320-1770.ijrcog20200926>
7. Benirschke K, Kauffmann P, Baergen RA. *Pathology of Human Placenta*. Fifth edition. 2006: pag. 381. Springer New York, NY. DOI: <https://doi.org/10.1007/b137920>
8. Nanaev AK, Kohnen G, Milovanov AP, Domogatsky SP, Kaufmann P. Stromal differentiation and architecture of the human umbilical cord. *Placenta*. 1997;18(1):53-64. DOI: 10.1016/s0143-4004(97)90071-0
9. Takechi K, Kuwabane Y, Mizuna M. Ultrastructural and Immunohistochemical Studies of Wharton's Jelly Umbilical Cord Cells. *Placenta*. 1993;14:235-45.
10. Cole J, Israfil-Bayli F. Wharton's jelly: The significance of absence. *J Obstet Gynaecol*. 2016 May;36(4):500-1. DOI: 10.3109/01443615.2015.1094041
11. Kulkarni M, Matadh P, Achok C, Pradeep N, Avinash T, Kulkarni A. Absence of Wharton's Jelly around the umbilical arteries. *Indian J Pediatr*. 2007;74(8):787-9. DOI: 10.1007/s12098-007-0142-7
12. Murphy SJ, Deegan N, O'Leary BD, McParland P. absence of Wharton's jelly. *BMJ Case Rep* 2020;13:e237222. doi:10.1136/bcr-2020-237222
13. Oliveira CC, Duflath RM, Coelho KR. Absence of Wharton's jelly: case report. *J Bras Patol Med Lab*. 2014;50(6):452-5.
14. Botezatu R.; Raduteanu S, Ciobanu AM, Gica N, Peltecu G, Panaitescu AM. Absence of Wharton's Jelly at the Abdominal Site of the Umbilical Cord Insertion. *Rare Case Report and Review of the Literature*. *Medicina*. 2021;57:1268. <https://doi.org/10.3390/medicina57111268>
15. Wade M, Gueye M, Mbodji A, Ndiaye MD. Absence of Wharton's jelly around an umbilical artery. *Int J Reprod Contracept Obstet Gynecol*. 2022;11:259-61. DOI: <https://dx.doi.org/10.18203/2320-1770.ijrcog20215115>
16. Damasceno EB, de Lima PP. Wharton's jelly absence: a possible cause of stillbirth. *Autops Case Rep*. 2013 Dec 31;3(4):43-7. doi: 10.4322/acr.2013.038. PMID: 28584806; PMCID: PMC5453660.