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

INTRODUCTION
The umbilical cord (UC) is responsible for maternal-fetal blood flow, and its various abnormalities are associated with significant perinatal morbidity and mortality\textsuperscript{(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)\textsuperscript{(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\textsuperscript{(3)}, who described it as mucoid degeneration of Wharton's jelly. Thomson\textsuperscript{(4)}, in 1996, called it 'linear disruption of the umbilical cord' and Laberrere\textsuperscript{(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\textsuperscript{(6)}.

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

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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.
**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>
<thead>
<tr>
<th>Author and year</th>
<th>N°</th>
<th>Parity</th>
<th>Age</th>
<th>Antecedent</th>
<th>Delivery</th>
<th>Weight</th>
<th>Weight/ Sex</th>
<th>Result</th>
<th>Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bergman(3) 1961</td>
<td>1</td>
<td>G1</td>
<td>25</td>
<td>-</td>
<td>Vaginal</td>
<td>NS</td>
<td>2.270 g/NS</td>
<td>Stillbirth</td>
<td>Meconium aspiration Intrauterine asphyxia</td>
</tr>
<tr>
<td>Laberrere(5) 1985</td>
<td>3</td>
<td>G1 25</td>
<td>-</td>
<td>C-section</td>
<td>560 g</td>
<td>3.220 g/M</td>
<td>Deceased newborn</td>
<td>Meconium aspiration</td>
<td></td>
</tr>
<tr>
<td>Thomson(4) 1996</td>
<td>1</td>
<td>NS 25</td>
<td>-</td>
<td>Vaginal</td>
<td>515 g</td>
<td>3.450 g/F</td>
<td>Disabled newborn</td>
<td>Meconophly Psychomotor delay</td>
<td></td>
</tr>
<tr>
<td>Kulkarni(11) 2007</td>
<td>1</td>
<td>G1 19</td>
<td>HIV</td>
<td>Vaginal</td>
<td>500 g</td>
<td>2.500 g/M</td>
<td>Morbid child</td>
<td>Patent vitelline duct</td>
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</tr>
<tr>
<td>Oliveira(6) 2014</td>
<td>1</td>
<td>G1 22</td>
<td>-</td>
<td>Vaginal</td>
<td>385 g</td>
<td>NS/NS</td>
<td>Stillbirth</td>
<td>Acute funisitis Chorioamnionitis 3</td>
<td></td>
</tr>
<tr>
<td>Cole(10) 2016</td>
<td>1</td>
<td>Multiparous</td>
<td>31</td>
<td>NS</td>
<td>C-section</td>
<td>NS</td>
<td>3.285 g/M</td>
<td>Healthy child</td>
<td>Fetal distress</td>
</tr>
<tr>
<td>Trivedi(9) 2020</td>
<td>1</td>
<td>G3P(1) 22</td>
<td>C-section Neonatal death</td>
<td>C-section</td>
<td>350 g</td>
<td>2.000 g/M</td>
<td>Deceased newborn</td>
<td>Neonatal ICU</td>
<td></td>
</tr>
<tr>
<td>Murphy(8) 2020</td>
<td>1</td>
<td>G1 42</td>
<td>IVF</td>
<td>C-section</td>
<td>739 g</td>
<td>3.680 g/F</td>
<td>Live newborn</td>
<td>Neonatal ICU Meconium aspiration</td>
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<tr>
<td>Botezatu(14) 2021</td>
<td>1</td>
<td>G1 29</td>
<td>-</td>
<td>C-section</td>
<td>500 g</td>
<td>3.500 g/M</td>
<td>Live newborn</td>
<td>Abnormal fetal cardiac tracing and prolonged late deceleration</td>
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<tr>
<td>Wade(6) 2022</td>
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<td>G1 34</td>
<td>-</td>
<td>C-section</td>
<td>350 g</td>
<td>3.150 g/M</td>
<td>Live newborn</td>
<td>Nuchal cord Oligohydramnios</td>
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<tr>
<td>Romero (2023)</td>
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<td>G1 21</td>
<td>-</td>
<td>Vaginal</td>
<td>450 g</td>
<td>2.620 g/M</td>
<td>Deceased newborn</td>
<td>Nuchal cord Oligohydramnios Neonatal ICU</td>
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<td>Romero (2023)</td>
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<td>G6 P4014</td>
<td>31</td>
<td>HIV, abortion</td>
<td>Vaginal</td>
<td>175 g</td>
<td>375 g/M</td>
<td>Stillbirth</td>
<td>Left renal agenesis. Abdominal and cerebral hemorrhage</td>
</tr>
</tbody>
</table>

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


