

Spontaneous rupture of the intertwin membrane in a monochorionic diamniotic pregnancy leading to umbilical cord entanglement

Rotura da membrana amniótica entre os fetos numa gravidez gemelar monocoriónica diamniótica com entrelaçar dos cordões umbilicais

Maria Flores-Casteleiro¹, Vera Trocado², Carla Monteiro¹

Unidade Local de Saúde de Braga e Unidade Local de Saúde do Alto Minho

Abstract

Monochorionic monoamniotic twin pregnancies are associated with the highest complication rates, including selective fetal growth restriction and fetal death due to hemodynamic imbalances and possibly umbilical cord entanglement. Monochorionic diamniotic pregnancies typically don't face this last risk. This case reports a rare instance of umbilical cord entanglement in a monochorionic diamniotic twin pregnancy following spontaneous rupture of the intertwin membrane, with one fetus diagnosed with fetal growth restriction. Spontaneous septostomy in such pregnancies, though rare, can result in complications typically seen in monochorionic monoamniotic twins, highlighting the need for careful monitoring and management.

Keywords: Monochorionic diamniotic; Umbilical cord entanglement; Rupture intertwin membrane.

Resumo

A gravidez gemelar monocoriónica monoamniótica associa-se a elevada taxa de complicações, incluindo restrição de crescimento fetal seletiva e morte fetal, devido a desequilíbrios hemodinâmicos e, possivelmente, ao complexo entrelaçar dos cordões umbilicais. A gravidez monocoriónica diamniótica, tipicamente, não apresenta este último risco. O caso relata o entrelaçar dos cordões umbilicais fetais numa gravidez monocoriónica diamniótica, após rotura espontânea da membrana amniótica entre os fetos, com diagnóstico de restrição de crescimento fetal seletiva. A rotura da membrana entre os fetos, apesar de rara, pode resultar em complicações tipicamente observadas em fetos monocoriónicos monoamnióticos, salientando a necessidade de uma monitorização e abordagem cuidadosa.

Palavras-chave: Monocoriónica diamniótica; Entrelaçar dos cordões umbilicais; Rotura da membrana entre fetos.

Monochorionic monoamniotic (MCMA) twin pregnancies have the highest complication rates. In structurally normal pregnancies, fetal deaths often result from hemodynamic imbalances due to extensive placental anastomoses and possibly tight

1. M.D., Serviço de Ginecologia e Obstetria, Unidade Local de Saúde de Braga, Braga, Portugal.

2. M.D., Unidade Local de Saúde do Alto Minho, Viana do Castelo, Portugal; Life and Health Sciences Research Institute, School of Medicine of the University of Minho, Braga, Portugal; ICVS/3Bs – Associated Laboratory, Braga/Guimarães, Portugal.

umbilical cord entanglement (UCE)¹. This can lead to severe complications, including selective fetal growth restriction (sFGR) or fetal death, from umbilical vessel pressure and impaired blood flow². The monochorionic diamniotic (MCDA) twins, with an intertwin dividing membrane, are usually not at risk of UCE. This case highlights intertwin UCE in an MCDA twin pregnancy with preterm rupture of membrane (PROM) and sFGR.

A 32-year-old gravida 2 para 1 with no significant medical history, had a 12-week ultrasound revealing a MCDA twin pregnancy. At 22 weeks, anatomy scans showed normal findings, but fetus B had sFGR and an image of apparent intertwining UCE was visualized (Figure 1), although the intertwin membrane was partially visualized. The woman remained under hospital surveillance with serial ultrasounds. At 25 weeks she had amniotic fluid leakage with a diagnosis of PROM, fetal maturation and antibiotic therapy were initiated, and an ultrasound was performed: fetus A had an estimated fetal weight (EFW) in the 22,8th percentile, with normal umbilical artery (UA) doppler and abnormal middle cerebral artery (MCA) doppler; while fetus B had sFGR (EFW 1,5th percentile), with anomalous UA and MCA dopplers. Both had visible bladders, normal amniotic fluid and the membrane was partially visible. Fetal surveillance included thrice-daily non-stress tests (NSTs) and doppler evaluation every 48 hours. Although the intertwin membrane was no longer visible, the UCE had become unclear. The NST remained variable and reactive, without decelerations. By 27 weeks, fetus A's EFW dropped to 9,2nd percentile, with normal UA doppler and abnormal MCA doppler; while fetus B's EFW was 0,4th percentile, with intermittently absent UA diastolic flow and abnormal MCA dopplers. The following day, the patient had regular contractions and 4 cm dilation, progressing to active labor within 3 hours, leading to a cesarean section. Newborn A weighed 930 g (APGAR 8/9/9), and B weighed 685 g (APGAR 6/8/9). The umbilical cords were entangled in a complex knot (Figure 2). Both twins were discharged from the neonatal intensive care unit after 77 days. Histopathological exams confirmed amniocity and chorionicity.

We report a case of an intrauterine environment change from diamniotic to monoamniotic due to



FIGURE 1. Ultrasound image of umbilical cord entanglement between the fetuses.



FIGURE 2. Complex umbilical cord entanglement between the fetuses with multiple loops and knots.

intertwin membrane rupture. This may occur spontaneously (infection, excessive fetal movements, polyhydramnios, maternal trauma) or iatrogenically (invasive procedures)^{2,3,4}. Antenatal diagnosis is rarely described, suggestive ultrasound findings are an absent/disrupted membrane, twins being close to each-other and UCE⁴. A case series reported UCE incidence at 72% in MCMA twins and 43% in MCDA with intertwin membrane rupture³. Various groups reported different approaches, caesarean delivery is often scheduled between 32 and 35 weeks⁵. Concluding, our case and others confirm that, though rare, spontaneous septostomy in an MCDA pregnancy can cause complications like UCE, typically seen in monoamniotic twins. Careful monitoring of the membrane and cords is essential, and difficulty visualizing the membrane should raise suspicion of rupture, guiding prenatal care and delivery planning.

REFERENCES

1. Liberty G, Reyzer SM, Shenhav S, Anteby EY, Cohen SM, Yagel S. Ultrasound diagnosis of first trimester umbilical cord entanglement in monochorionic monoamniotic twins – case report and review of the literature. *BMC Pregnancy Childbirth*. 2024 Nov 28;24(1):801. doi: 10.1186/s12884-024-06962-6. PMID: 39604909; PMCID: PMC11603871.
2. Razavi B, Kasraeian M, Hashemi A, Moradi Alamdarloo S, Najib FS. Complex twisted knots of umbilical cord in a monochorionic-diamniotic twin gestation: a case report. *Galen Med J*. 2020 Sep 16;9:e1878. doi: 10.31661/gmj.v9i0.1878. PMID: 34466603; PMCID: PMC8343711.
3. Suzuki S. Case series of monoamniotic and pseudomonoamniotic twin gestations. *ISRN Obstet Gynecol*. 2013;2013:369419. doi: 10.1155/2013/369419. Epub 2013 Feb 21. PMID: 23509635; PMCID: PMC3595717.
4. Boccherini C, D'Ambrosio V, Marcocchia E, Vena F, Di Mascio D, Corno S, Bartolone M, Giancotti A. Monochorionic-Diamniotic Twin Pregnancy Complicated by Spontaneous Septostomy and Cord Entanglement: A Systematic Review, Evaluation of Complication Rates and Presentation of an Additional Case. *Clin Exp Obstet Gynecol*. 2022;49(6):123. doi: 10.31083/j.ceog4906123.
5. Lee KJ, Kim MK, Lee SY, Lee WS, Lee YH. Spontaneous rupture of the dividing membrane in a monochorionic pregnancy resulting in a pseudo-monoamniotic pregnancy with cord entanglement. *J Obstet Gynaecol Res*. 2012 May;38(5):863-6. doi: 10.1111/j.1447-0756.2011.01795.x. Epub 2012 Mar 26. PMID: 22449420.

AUTHORS' CONTRIBUTIONS

MFC prepared the clinical case, conducted the scientific research and wrote the manuscript. VT obtained the ultrasound images and reviewed the manuscript. CM collected the images and reviewed the manuscript.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest. No funding.

PATIENT CONSENT

Informed consent was obtained

ACKNOWLEDGMENT

Fetal Medicine and Prenatal Diagnosis Unit team, Maternal-Fetal Pathology team.

CORRESPONDENCE TO

Maria Flores-Casteleiro
E-mail: mariafcasteleiro@gmail.com
<https://orcid.org/0009-0003-5077-0871>

RECEIVED: 14/01/2025

ACCEPTED: 03/05/2025