CASE REPORT

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# Management of monochorionic monoamniotic twin pregnancy: literature review and case report

## Manejo del embarazo gemelar monocoriónico monoamniótico: revisión de la literatura y reporte de caso

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**ABSTRACT**

In view of the presentation of a case of monochorionic monoamniotic twin pregnancy, the literature on its diagnosis and management was reviewed by electronic search in the Medline, OVID, Cochrane, and PubMed databases between 1966 and 2019. The key words used in the inquiry were: pregnancy, twins, monoamniotic, cord entanglement. There is a high risk of sudden fetal death in monoamniotic twins caused by entanglement of the umbilical cords, so timely diagnosis of chorionicity and amnionicity in twin pregnancy by ultrasonography may provide guidance for close fetal surveillance and thus improve obstetric outcome.

Key words: Chorion, Amnion, Pregnancy, Twins, monozygotic, Umbilical cord, Fetal death

**RESUMEN**

A propósito de la presentación de un caso de embarazo gemelar monocoriónico monoamniótico, se revisó la literatura sobre su diagnóstico y manejo mediante búsqueda electrónica en la base de datos de Medline, OVID, Cochrane y PubMed entre los años 1966 y 2019. Las palabras clave utilizadas en la indagación fueron: embarazo, gemelos, monoamniótico, enredamiento de cordones. Existe un riesgo alto de muerte fetal súbita en gemelos monoamnióticos causado por el enredamiento de los cordones umbilicales, por lo que el diagnóstico oportuno de la corionicidad y amnionicidad en el embarazo gemelar mediante ecografía puede dar la pauta para una vigilancia fetal estrecha y mejorar así el resultado obstétrico.

Palabras clave. Corion, Amnios, Embarazo gemelar, Gemelos monocigóticos, Cordón umbilical, Muerte fetal

## Introduction

The prevalence of dizygotic twins varies between populations, whereas the prevalence of monozygotic twins is relatively stable worldwide.

Monoamniotic twins account for approximately 0.01 % of spontaneously conceived pregnancies and 5 % of monochorionic twin pregnancies. Monoamniotic twins include conjoined or Siamese twins, with an incidence of 1 in 50.000 pregnancies[(1)].

Monoamniotic twin pregnancies are the least common type and have many of the same complications as biamniotic monochorionic twin pregnancies (e.g., feto-fetal transfusion syndrome). However, monoamniotic twin pregnancy is characterized by increased risks of congenital anomalies[(2,3)] and umbilical cord intertwining with potential sudden fetal death[(4)].

In twin pregnancy, the post-fertilization division of the zygote determines the type of chorionicity and amnionicity. Monoamniotic monochorionic placentation occurs when that division occurs between days 8 to 12 post-fertilization. In comparison, biamniotic twins are the result of division between day 4 and 7 for monochorionic placentation and between day 1 and 3 for dichorionic placentation.

The factors responsible for the timing of embryo splitting are unknown. The use of assisted reproductive techniques appears to play a role, as in vitro fertilization increases the frequency of monozygotic twinning.

In some studies, manipulation of the zona pellucida, which is performed with intracytoplasmic sperm injection and assisted hatching, increased the frequency of monoamniotic twins[5].

Ultrasonography is an effective prenatal diagnostic tool to determine amnionicity and chorionicity. This is of utmost importance, because monochorionic twins have a shared fetoplacental circulation, which puts them at risk for serious pregnancy-specific complications, such as feto-fetal transfusion syndrome (FFTS), twin anemia-polycythemia sequence (TAPS), and selective fetal growth restriction (selective IUGR)[6].

These complications increase the risk of neurological morbidity and perinatal mortality in monochorionic twins compared to dichorionic twins. Also, prenatal identification of monochorionic monoamniotic twins (MCMA) is critical, because monoamnionicity carries a higher risk of adverse perinatal outcomes compared to biamnionicity[7].

In expert hands, the optimal time to determine chorionicity and amnionicity is in the first trimester after seven weeks, with a sensitivity ≥ 98 %, while in the early second trimester the sensitivity is 90 %[8-10].

The identification of two separate placentas is a very reliable indicator of dichorionic twins. This indicator is generally only useful in early pregnancy, as separated placentas often appear fused later in gestation.

The intergemellar membrane is absent in a monoamniotic monochorionic twin pregnancy, so great care must be taken to confuse this condition with a biamniotic monochorionic twin pregnancy complicated with severe FFTS, where the intergemellar membrane is completely folded over the donor fetus, making it almost impossible to visualize ultrasonographically.

The observation of a triangular projection of tissue extending between the layers of the intergemellar membrane from the placenta (lambda sign) determines the diagnosis of diamniotic bicornuate twin pregnancy[11].

On the other hand, diamniotic monochorionic twin pregnancy is appreciated sonographically with the T-sign, which refers to the appearance of the thin intergemellar membrane composed of two amniotic layers when it detaches from the placenta at a 90-degree angle[12].

There are additional findings, such as the visualization of a single vitelline vesicle with two fetal poles, strongly suggesting the presence of monoamniotic twins[13,14]. However, the diagnosis should always be corroborated by 12-week ultrasonography[15,16].

Finally, ultrasonographic detection of umbilical cord entanglement is pathognomonic for monoamniotic twins and can be seen as early as the late first trimester[17,18].

Regarding complications, monochorionic monoamniotic twin pregnancy, like diamniotic monochorionic twin pregnancy, may present with FFTS, selective IUGR, preterm delivery and, particularly, the unique complication of monoamniotic pregnancies, such as umbilical cord entanglement.

The high mortality rate of monoamniotic twins has been related to sudden death because of such umbilical crosslinking. Intermittent occlusion of the umbilical blood vessels may be associated with neurological morbidity, whereas prolonged occlusion may be lethal[19].

Monoamniotic twins have a high mortality rate, which has been reported in up to 50% of cases. Monoamniotic twins have a high risk of congenital malformations, reaching a frequency of 38-50 %, usually affecting only one twin[20,21].

Monoamniotic twin pregnancy should be managed in a specialized center, as it requires close surveillance.

Given the high frequency of congenital malformations in this group of twins, it is essential to perform a specialized ultrasound between 12 and 14 weeks of gestation and a detailed morphological ultrasound between 16 and 18 weeks[22].

The antenatal management plan should include monitoring of fetal well-being, so ultrasound surveillance is recommended every 2 weeks after week 24[23].

Umbilical artery Doppler evaluation should be performed at least weekly. Abnormal Doppler ultrasound findings vary in severity from the presence of a diastolic notch to absent or reverse diastole[24]. The presence of such a notch may reflect hemodynamic alterations in the fetal-placental circulation secondary to narrowing of the umbilical vessels involved in umbilical entanglement[25].

Regarding in-hospital versus home or outpatient surveillance, the MONOMONO study evaluated the difference in obstetric outcome between the two types of surveillance. 195 pregnant women with an uncomplicated monoamniotic twin gestation (390 fetuses) were included. Of these, 75 (38.5 %) were managed as inpatients and 120 (61.5 %) as outpatients. The overall perinatal loss rate was 10.8 % (42/390), with a peak fetal mortality rate of 4.3 % (15/348) occurring at 29 weeks' gestation. On that basis, there was no statistically significant difference in fetal mortality rates between inpatient management initiated at 26 weeks compared with outpatient surveillance[26].

Elective cesarean termination of monoamniotic gestations at 32-33 weeks is recommended to avoid potential sudden death due to cord entanglement and extreme prematurity[27]. Neonatal outcomes are reasonably good at 32 weeks for infants cared for in well-equipped neonatal intensive care units[28]. If the fetus has not had pulmonary maturation before 28 weeks because of threatened preterm delivery, administration of a new course of antenatal corticosteroids is recommended at the time of hospital admission.

## Case report

We present the case of a 37-year-old female patient with type 2 diabetes mellitus in glycemic control with diet and hypothyroidism under treatment with levothyroxine, managed in a maternal-fetal medicine unit of a private hospital.

Her first trimester ultrasound determined a monochorionic monoamniotic twin gestation and her noninvasive prenatal genetic testing was reported as normal, with both female fetuses.

On second trimester morphologic ultrasound, the anatomy of both fetuses was normal, as was their growth. Also, sonographic signs of umbilical cord entanglement were already identified.

At 28 weeks, during ultrasound surveillance, the diagnosis of intrauterine growth restriction was established in both fetuses (Figure 1) and umbilical cord entanglement was evident (Figures 2 and 3). The Doppler spectral representation of the umbilical artery showed a protodiastolic notch (Figure 4), so the diagnosis of 28-week monochorionic monoamniotic twin pregnancy complicated with IUGR in both fetuses, umbilical cord entanglement, history of previous cesarean section, hypothyroidism and pregestational diabetes was established.

In-hospital management for antenatal surveillance was decided. During hospitalization at the maternity center, strict electronic fetal monitoring was performed 3 times a day and Doppler ultrasound, evaluating blood flow in the umbilical artery, middle cerebral artery, and venous ductus twice a week.

It was decided to terminate the pregnancy at 32 weeks and 4 days after administering 2 cycles of pulmonary maturity inducers and neuroprotection with magnesium sulfate the night before surgery.

The cesarean section was performed without complications, obtaining two female newborns with immediate crying and weights of 1,750 and

Figure 1. Graph of the growth pattern of both fetuses. A downward growth curve is shown with fetus A at percentile 4 and fetus B at percentile 2.

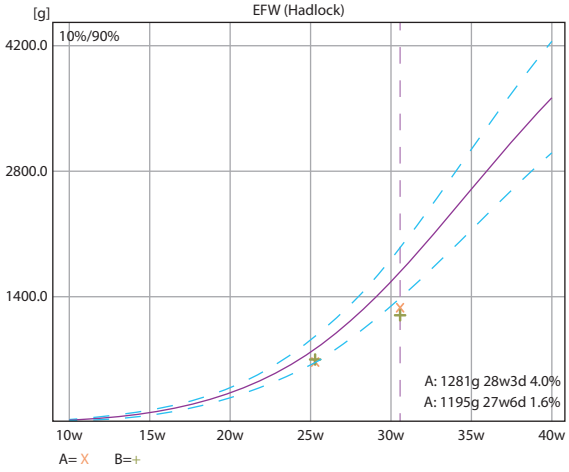


Figure 2. Grayscale image showing umbilical cord entanglement.


Figure 3. Color Doppler study denotes cross-linking and entanglement of the umbilical cords.

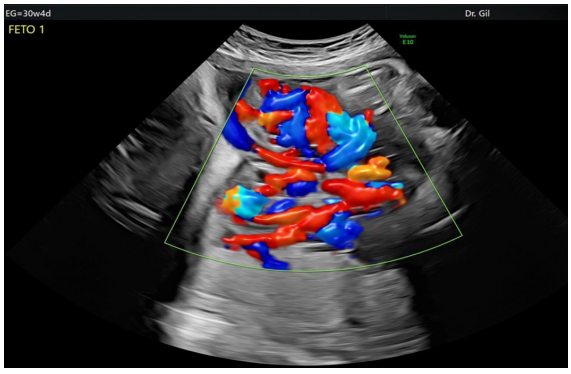
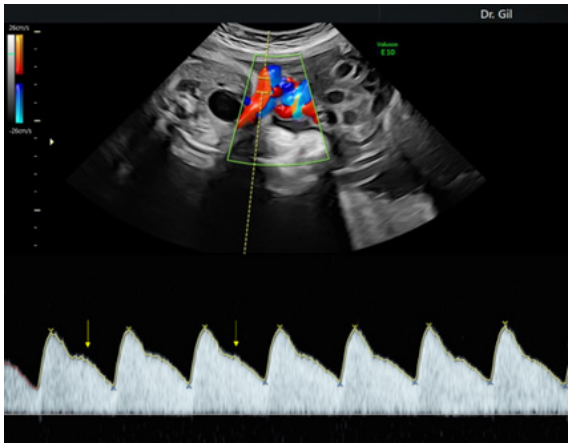



Figure 4. Doppler study of the umbilical artery showing protodiastolic notch (yellow arrow) due to umbilical compression by cord entanglement.




1,560 grams and Apgar of both babies of 8 at one minute and 9 at 5 minutes, having as the most important finding the presence of intertwined umbilical cords and with two true knots between them (Figures 5 and 6).

The patient was discharged on the third post-cesarean day with favorable evolution. The twins remained hospitalized in the neonatal ICU for 27 days in an incubator without major respiratory distress. During their hospitalization they gained weight and achieved spontaneous suction, being discharged with weights of 2,100 and 2,040 grams.

Figure 5. Postnatal evaluation showing umbilical cord intertwining and entanglement.

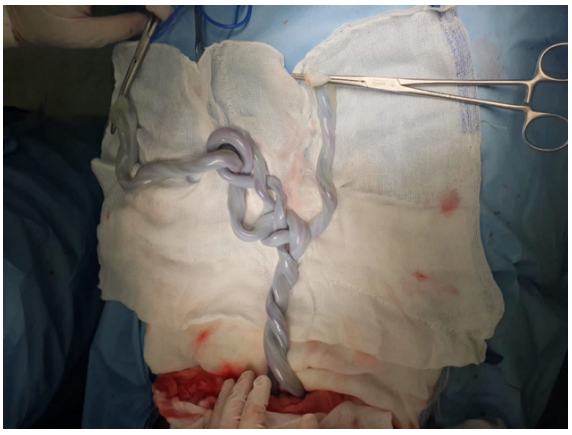


Figure 6. Postnatal evaluation showing the proximity between the placental insertions of the umbilical cords.

## Discussion

Perinatal mortality is the most important complication of monoamniotic placentation, either due to sudden fetal death secondary to umbilical cord entanglement or due to fetal growth disturbances and prematurity.

Umbilical cord entanglement is one of the main causes of fetal death in these pregnancies, which occurs due to the proximity of the placental insertions of the umbilical cords.

The key point in the management of twin pregnancies is the timely ultrasound diagnosis of chorionicity and amnionicity, since once these are determined, an antenatal surveillance plan can be implemented, thus reducing perinatal morbidity and mortality.

The present case is important because it shows how a monochorionic monoamniotic twin gestation complicated with IUGR and umbilical cord entanglement can be successfully managed in a tertiary level center, both prenatally and postnatally.

It is important to highlight the permanent fetal surveillance in complicated monoamniotic twin pregnancies and the resolution of the pregnancy by elective cesarean section at 32-33 weeks after pulmonary maturation and fetal neuroprotection.

In the present communication, in-hospital surveillance was decided due to the diagnosis of IUGR in both fetuses and the presence of notch in the spectral Doppler wave of the umbilical artery, which is an indicator of funicular compression.

Delivery in a well-equipped neonatal intensive care unit is associated with a low risk of neonatal mortality or severe morbidity.

Finally, the available evidence is insufficient to allow a strong recommendation on the optimal gestational age for planned delivery of these pregnancies, but this case clearly demonstrates that individual maternal and fetal management should be individualized for each pregnant woman.

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