International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Mohamedalhalabi H. Int J Reprod Contracept Obstet Gynecol. 2026 Nov;15(11):4595-4598
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20263694

Case Report

Successful management of a monochorionic monoamniotic twin pregnancy with extensive cord entanglement

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Received: 17 September 2026
Revised: 03 October 2026
Accepted: 05 October 2026

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ABSTRACT

Monochorionic monoamniotic (MCMA) twin pregnancy is rare and is associated with fetal loss, congenital anomalies, prematurity and umbilical cord entanglement. We report the management and outcome of a 29-year-old primigravida in whom a single placenta, two fetuses and no intertwin membrane were identified at 9 weeks of gestation. Surveillance included ultrasound assessment every 2 weeks from 16 weeks, umbilical artery and middle cerebral artery Doppler studies, fetal echocardiography at 20-22 weeks, and inpatient fetal surveillance from 30 weeks. A planned caesarean birth was performed at 32+1 weeks, consistent with guidance recommending planned preterm birth for uncomplicated MCMA twins. Two female neonates weighing 1.5 kg and 1.4 kg were delivered. Both received neonatal intensive care for 15 days and were discharged in stable condition. Maternal recovery was uncomplicated. Marked entanglement of the two umbilical cords was documented intraoperatively. This case illustrates that extensive cord entanglement may coexist with a favourable outcome when early diagnosis, structured surveillance, planned preterm caesarean birth and neonatal support are available.

Keywords: Monoamniotic twins, Monochorionic twins, Cord entanglement, Caesarean section, Preterm birth

INTRODUCTION

Monochorionic monoamniotic (MCMA) twins share one placenta and one amniotic cavity and account for approximately 1% of twin pregnancies.[1] The absence of an intertwin membrane permits umbilical cord entanglement, while the shared placental circulation creates risks that include twin-twin transfusion syndrome (TTTS), selective fetal growth restriction, twin anaemia-polycythaemia sequence and acute haemodynamic imbalance.[1,2]

Early determination of chorionicity and amnionicity is therefore central to risk assessment and referral to an experienced fetal medicine service.[2] We describe an MCMA pregnancy with extensive cord entanglement at delivery and favourable maternal and neonatal outcomes after planned caesarean birth at 32+1 weeks.

CASE REPORT

A 29-year-old primigravida (G1P0) with an unremarkable medical history attended routine antenatal care. She did not smoke, drink alcohol or use recreational substances, and there was no known family history of twin pregnancy or genetic disease. Ultrasonography at 9 weeks showed two fetuses with synchronous cardiac activity, a single placenta and no visible intertwin membrane, consistent with an MCMA twin pregnancy.

The patient received counselling about the diagnosis, anticipated complications, surveillance and planned preterm birth. Serial ultrasonography was undertaken every 2 weeks from 16 weeks to assess fetal growth and features of monochorionic complications. Surveillance included Doppler assessment of the umbilical and middle

cerebral arteries. Fetal echocardiography was performed at 20-22 weeks.

She was admitted at 30 weeks for intensified fetal surveillance. The documented regimen included daily cardiotocography and weekly biophysical profile assessment. Neonatal support was arranged in anticipation of preterm birth. No emergency indication for delivery was documented. Colour Doppler ultrasonography at 31+0 weeks demonstrated intertwined umbilical vessels consistent with cord entanglement (Figure 1).

A planned caesarean section was performed at 32+1 weeks as the intended timing and mode of birth for the MCMA pregnancy. Two female neonates were delivered, weighing 1.5 kg and 1.4 kg. Both neonates received care in the neonatal intensive care unit for 15 days and were discharged in stable condition. The mother's postoperative recovery was uncomplicated. Extensive entanglement of the umbilical cords was observed and photographed during the operation (Figure 2).

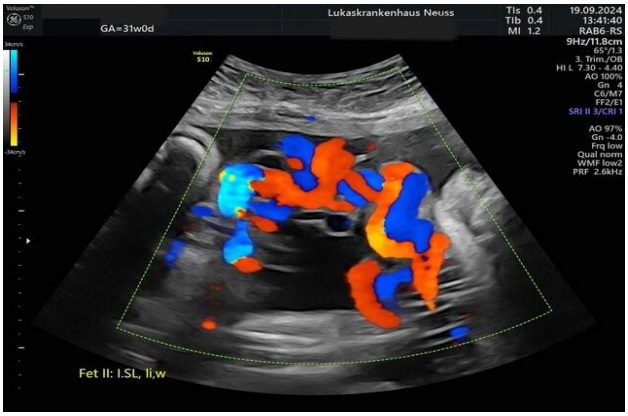


Figure 1: Colour Doppler ultrasound at 31+0 weeks showing intertwined umbilical vessels consistent with cord entanglement in the reported MCMA twin pregnancy.

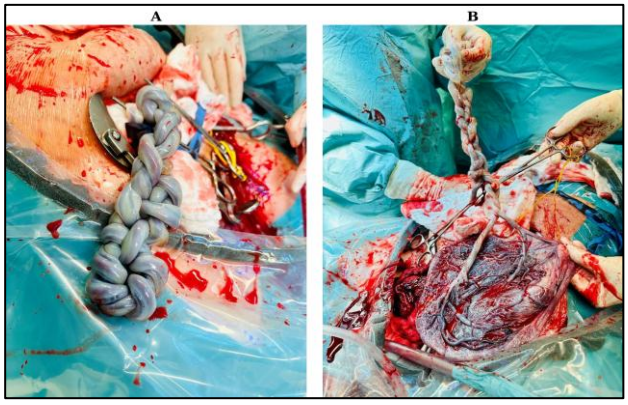


Figure 2 (A and B): Intraoperative photographs from the reported patient showing extensive umbilical cord entanglement. (A) Densely intertwined umbilical cords after delivery. (B) Shared placenta and entangled cords at caesarean birth.

DISCUSSION

This case brings together the central clinical challenges of MCMA pregnancy: reliable early diagnosis, surveillance for complications that may evolve rapidly, uncertainty about the best location and intensity of monitoring, and the need to balance the risk of unexpected fetal death against the morbidity of planned preterm birth. MCMA twins comprise approximately 1% of twin pregnancies and represent the highest-risk form of twin gestation.[1] Their risk is not explained by cord entanglement alone. A single shared placenta creates vascular connections between the fetal circulations, while the absence of an intertwin membrane permits unrestricted fetal movement and contact between both umbilical cords. The resulting clinical spectrum includes congenital anomalies, TTTS, selective fetal growth restriction, twin anaemia-polycythaemia sequence, acute intertwin haemodynamic events, spontaneous preterm birth and fetal death.[1,2]

The diagnosis at 9 weeks was therefore an important strength in this patient's management. Chorionicity and amnionicity are most accurately assigned in the first trimester, before progressive fusion of the amnion and chorion makes membrane assessment more difficult. NICE recommends using the number of placental masses, the presence and thickness of amniotic membranes, and the lambda or T sign when a twin pregnancy is first detected.[3] Updated ISUOG guidance notes that the amniotic sacs can be distinguished from approximately 8 weeks and recommends transvaginal assessment and tertiary review when uncertainty remains.[2] Early classification is clinically meaningful because an MCMA diagnosis changes counselling, surveillance, referral and delivery planning. It also avoids misclassifying a monochorionic diamniotic (MCDA) pregnancy with an inconspicuous or disrupted septum as a true monoamniotic pregnancy.

The two-weekly ultrasound strategy from 16 weeks used in this case is consistent with contemporary monochorionic surveillance principles. RCOG recommends serial fetal biometry and umbilical artery Doppler assessment at 2-week intervals from 16 weeks, with middle cerebral artery peak systolic velocity assessment from 20 weeks to screen for twin anaemia-polycythaemia sequence.[1] In MCMA twins, TTTS may be harder to recognize because both fetuses occupy the same amniotic cavity; the classic oligohydramnios-polyhydramnios sequence seen in separate sacs cannot be applied in the usual way. Evaluation must instead integrate fetal growth, bladder filling, Doppler findings, cardiac function and fetal heart-rate patterns.[2,5] Fetal echocardiography at 20-22 weeks, as documented here, was also clinically appropriate because monochorionic gestations have an increased prevalence of cardiac and other structural anomalies.

The extensive operative cord entanglement is visually striking, but its interpretation requires care. Entanglement

is characteristic rather than exceptional in MCMA pregnancy: ISUOG describes it as almost universal when systematically assessed by colour and pulsed-wave Doppler, and RCOG states that it is almost always visualized.[1,2] In a systematic review of nine studies involving 114 monoamniotic twin sets and 228 fetuses with documented cord entanglement, overall survival was 88.6% (202/228) and perinatal mortality was 11.4% (26/228).[4] Of the 26 deaths, 17 occurred in utero and nine occurred at birth; only two neonatal deaths were attributed directly to cord entanglement, whereas five were related to prematurity and two to structural anomalies. The pooled comparison did not demonstrate significantly higher mortality in fetuses with entanglement than in controls.[4] Thus, the operative photographs in this report document the anatomical severity of entanglement, but they should not be used to claim that entanglement was the emergency indication for birth.

The risk of fetal loss persists despite apparently reassuring surveillance because some adverse events are sudden and may not be preceded by abnormal growth or Doppler findings. A systematic review and meta-analysis of 25 studies included 1,628 structurally non-anomalous MCMA twins alive at 24 weeks.[5] Single and double intrauterine deaths occurred in 2.5% (95% CI 1.8-3.3%) and 3.8% (95% CI 2.5-5.3%), respectively. The gestational-age-specific incidence of intrauterine death was 4.3% (95% CI 2.8-6.2%) at 24-30 weeks, 1.0% (95% CI 0.6-1.7%) at 31-32 weeks and 2.2% (95% CI 0.9-3.9%) at 33-34 weeks.[5] These non-linear estimates underline why a normal assessment at one visit cannot provide complete reassurance for the remainder of the pregnancy. They also explain the use of repeated ultrasound, cardiotocography and multidisciplinary review rather than reliance on a single test.

Whether intensified surveillance should routinely occur in hospital remains controversial. In the same meta-analysis, the pooled incidence of intrauterine death was 3.0% (95% CI 1.4-5.2%) in pregnancies managed mainly as inpatients and 7.4% (95% CI 4.4-11.1%) in those managed mainly as outpatients.[5] However, observational comparisons are vulnerable to differences in patient selection, admission thresholds, monitoring protocols and gestational age at entry. The multinational MONOMONO cohort included 195 uncomplicated pregnancies and 390 fetuses; 75 pregnancies were managed as inpatients and 120 as outpatients.[6] Overall perinatal loss was 10.8% (42/390), with the highest weekly fetal-death rate of 4.3% at 29 weeks. There was no statistically significant difference in fetal death between the strategies in the adjusted clustered analysis (3.3% vs 10.8%; adjusted odds ratio 0.21, 95% CI 0.04-1.17).[6] Inpatient care was associated with a much longer mean maternal hospital stay (42.1 vs 7.4 days), illustrating the psychological, family and resource implications of prolonged admission.

The decision to admit this patient at 30 weeks for daily cardiotocography and weekly biophysical profile assessment was therefore a defensible local strategy, but the favourable outcome cannot be attributed to hospitalisation alone. Continuous monitoring is also difficult to achieve in practice because simultaneous, interpretable fetal heart-rate traces may not be obtainable at all times in two mobile fetuses. More monitoring can identify recurrent decelerations or evolving compromise and facilitate rapid delivery, but it cannot reliably predict every acute cord or placental event. RCOG accordingly concludes that fetal mortality is similar with increased inpatient and outpatient surveillance and recommends individualised management in a fetal medicine centre with appropriate neonatal and operative resources.[1]

Timing of birth is the pivotal management decision because prolonging pregnancy may reduce prematurity-related morbidity while exposing both fetuses to continuing intrauterine risk. In the mortality meta-analysis, 37.8% of MCMA pregnancies delivered before the scheduled date, mainly because of spontaneous preterm labour or abnormal cardiotocography.[5] An English multicentre cohort reported an overall perinatal mortality of 14.7% (17/116), but the rate varied markedly with gestational age, from 69.2% among births before 30 weeks to 4.5% at 33 weeks or later.[7] Among pregnancies surviving beyond 24 weeks, the median gestational age at delivery was 33 weeks (interquartile range 32-34), usually by caesarean section.[7] These findings demonstrate that much of the observed mortality burden is concentrated in pregnancies complicated by very early fetal loss or extreme prematurity rather than among all patients reaching the planned delivery window.

Neonatal morbidity provides the counterweight to early delivery. A systematic review of 14 studies comprising 685 non-anomalous MCMA pregnancies found that composite neonatal morbidity declined from 75.4% at 24-30 weeks to 65.5% at 31-32 weeks, 37.6% at 33-34 weeks and 18.5% at 35-36 weeks.[9] Respiratory morbidity decreased across the same intervals from 74.2% to 59.1%, 35.5% and 12.2%, while neurological morbidity declined from 15.3% to 10.2%, 4.3% and 0%, respectively.[9] These figures make the clinical trade-off tangible: even a short prolongation may reduce neonatal respiratory and neurological complications, but waiting beyond the recommended window exposes the fetuses to a risk of sudden death that surveillance cannot completely remove.

Current guidance resolves this balance in favour of planned preterm caesarean birth. NICE recommends planned birth between 32+0 and 33+6 weeks for uncomplicated MCMA twins, after antenatal corticosteroids have been considered, and recommends caesarean section at the planned birth.[3] NICE also advises counselling that neonatal-unit admission and respiratory problems are likely, while continuation beyond 33+6 weeks increases fetal-death risk.[3] RCOG recommends planned caesarean birth between 32+0 and 34+0 weeks.[1] The caesarean at 32+1 weeks in this case was therefore within both recommended windows. It should be described as a planned strategy for an MCMA pregnancy, not

retrospectively reclassified as emergency delivery because extensive entanglement was discovered at operation.

Contemporary outcomes are substantially better than the historically quoted mortality of 30-70%. In a Swedish single-centre cohort of 22 MCMA pregnancies and 44 fetuses, 35 fetuses (80%) were liveborn overall; among the 36 fetuses reaching 24 weeks, 35 (97%) were liveborn and there were no neonatal deaths.[8] Mean gestational age at birth was 32.5 weeks, mean birth weight was 1,910 g, and neonatal care lasted a mean of 19 days.[8] The two infants in the present report were smaller, at 1.5 kg and 1.4 kg, but both were discharged in stable condition after 15 days of neonatal intensive care. This comparison is encouraging, although the absence of detailed respiratory, infectious, feeding and neurological data prevents a more granular assessment of neonatal morbidity.

The case is clinically valuable because it links a favourable short-term outcome with direct operative documentation of severe cord entanglement. It reinforces three practical messages. First, early diagnosis creates time for informed counselling, specialist surveillance and coordinated neonatal planning. Second, cord entanglement should be expected and monitored but should not be treated as a stand-alone quantitative predictor of death. Third, the objective of management is not to eliminate all fetal or neonatal risk-an impossible goal with current tests-but to select a delivery window in which competing risks are acceptably balanced and emergency delivery remains feasible.

This report has important limitations. It describes a single pregnancy and cannot establish that any particular surveillance component caused the favourable outcome. The available record does not provide detailed longitudinal growth percentiles, Doppler values, cardiotocographic findings, antenatal corticosteroid or magnesium sulfate use, cord-blood measurements, placental histopathology, specific neonatal diagnoses or long-term neurodevelopmental follow-up. These details should not be inferred. Furthermore, published evidence is dominated by retrospective cohorts from specialist centres, with variable entry criteria and surveillance protocols; apparent differences in outcome may therefore reflect selection and referral effects. Even with these limitations, the case provides a clear illustration of evidence-aligned timing of birth and demonstrates that dramatic operative entanglement can coexist with favourable early maternal and neonatal outcomes.

CONCLUSION

Early diagnosis, specialist surveillance, planned preterm caesarean birth and coordinated neonatal care were associated with favourable short-term outcomes in this MCMA twin pregnancy. Delivery at 32+1 weeks was consistent with current guidance for planned birth and should not be presented as an emergency intervention for cord entanglement. Operative entanglement can be extensive despite reassuring short-term maternal and neonatal outcomes.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Mohamedalhalabi H. Successful management of a monochorionic monoamniotic twin pregnancy with extensive cord entanglement. Int J Reprod Contracept Obstet Gynecol 2026;15:4595-8.