

Quiz

Complex braided true umbilical cord knots (TUCK) and cord entanglement with fetal demise in monochorionic monoamniotic twin pregnancy: a catastrophic sequela-what is your diagnosis?

A 23-year-old Gravida 3, Para 1, with one living and one abortion, with a history of one previous lower segment Cesarean section and a short inter-conception period of 16 months, conceived spontaneously during the lactational period. She was followed up outside and had irregular antenatal visits, with the first visit at 20 weeks. Ultrasound confirmed twin pregnancy with no congenital anomalies. She was taking iron and calcium supplements regularly. She presented late to our institution at 34+2 weeks in latent labour and all her investigations were reviewed. Ultrasonography revealed intrauterine fetal demise of one twin, with an overall liquor volume (amniotic fluid index) of 18 cm (Table 1). On the initial physical examination, the body mass index was 22 kg/m², with no signs of tachycardia, fever, pallor, icterus, or edema. On per abdomen examination, the abdomen was over-distended, and multiple fetal parts were palpable with the presence of scar tenderness. On vaginal examination, the cervical os was 1 cm dilated, uneffaced, with intact membranes. A strip NST of the live fetus was reactive, with a baseline fetal heart rate of 150 bpm. Based on these findings, the patient was admitted, and relevant hematological and biochemical investigations were sent (Table 1). The patient underwent an emergency Cesarean section. Intraoperatively, the lower uterine segment was well formed and the previous scar was thinned out; the amniotic bag ruptured, and the liquor was clear. Placenta delivered completely with membranes intact (Figure 1). Umbilical cord abnormality was noted and is shown in Figures 2 and 3. Both female babies were extracted as vertex presentation. The first neonate cried after initial steps of resuscitation, had a one minute APGAR score of 3, and weighed 2150 g. On re-evaluation at 5 minutes, the APGAR score improved to 7. The second twin was dead with no signs of life and with a birth weight of 2200 g. An autopsy was performed on the dead fetus (Table 1). Grossly, no congenital anomalies were noted; however, skin peeling was seen (Figure 4). No significant neonatal complications occurred in the surviving twin. After counselling, the placenta was sent for histopathological examination, the umbilical cord and fetal tissue of the intrauterine fetal demise fetus was sent for genetic analysis.

What is the complication and diagnosis in this case?

Received: October 16, 2025 **Accepted:** January 19, 2026 **Epub:** March 03, 2026



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DOI: 10.4274/jtgga.galenos.2026.2025-10-8

Cite this article as: Devalla A, Pagolu A, Gopidas GS, Veldi S, Palati B. Complex braided true umbilical cord knots (TUCK) and cord entanglement with fetal demise in monochorionic monoamniotic twin pregnancy: a catastrophic sequela-what is your diagnosis? J Turk Ger Gynecol Assoc.



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Answer

This is a monochorionic monoamniotic (MCMA) twin pregnancy with true umbilical cord knot (TUCK) and cord entanglement with a single intrauterine fetal demise.

Diagnosis of twins is made by ultrasound examination between 11+2 weeks and 14+1 weeks (crown-rump length 45–84 mm) to assess fetal viability, gestational age and chorionicity. Monochorionic twin pregnancies have a single placental mass and a thin inter-twin membrane that inserts into the placenta at a perpendicular plane (T-sign).

Monozygotic twinning occurs rarely in natural conception, with an overall incidence of 0.4–0.45% (1). MCMA twins are quite uncommon, constituting around 1% of all twin pregnancies and 5% of all monochorionic pregnancies (1). Specific complications associated with monochorionic twin (2) pregnancy are shown in Table 2. Pregnancies complicated by TUCK are associated with a 4–10-fold increased risk of

stillbirth, attributed to obstruction of umbilical venous return from the placenta and up to 11% perinatal morbidity resulting in intrauterine fetal demise or growth restriction (3). It is therefore important to consider that, in contrast to many other etiologies of stillbirth, umbilical cord accidents are likely acute, and thus affected fetuses are less likely to benefit from the compensatory system of redistribution of oxygenated blood towards essential fetal organs (central nervous system, heart and adrenal glands), as may occur in association with uteroplacental insufficiency (3).

Umbilical proximal cord insertion is common in MCMA placentas (53%) and is a causal factor underlying cord entanglement (4). The ultrasound diagnosis is challenging, and cord accidents are often an incidental finding during delivery (5). Enhanced three-dimensional sonographic resolution, along with the extensive use of colour Doppler, has significantly improved the accuracy of prenatal diagnosis of TUCK. The reliability of detection largely depends on whether

Table 1. Hematological, radiological, biochemical investigations

Date	Investigation	Impression
	Blood group	A positive
7/8/25	Hb (g/dL) WBC (10³/μl) Platelet count (10³/μl)	12.1 9.92 236
7/8/25	Fibrinogen	470 ng/mL
7/8/25	KFT (urea/creatinine)	8/0.4 mg/dL
7/8/25	LFT	Within normal limits
Fetal investigations		
4/3/25	Anomaly scan outside	Monochorionic monoamniotic twins (MCMA)~20+2 weeks Placenta anterior, liquor adequate, no gross anomalies detailed
3/7/25	Growth scan outside	Monochorionic monoamniotic twins ~29+5 weeks, AFI-15cm, placenta anterior, cephalic presentation, normal Dopplers Twin A: EFW 1407 gm Twin B: EFW-1421gm
7/8/25	Growth scan at institute	Monochorionic monoamniotic twins, AFI-18 cm, placenta anterior, normal Dopplers Twin A: Live intrauterine fetus ~ 34+3 weeks, cephalic, EFW 2368gm Twin B: Intra uterine fetus ~ 33+3 weeks, with no cardiac activity, EFW-2093gm
7/8/25	Fetal autopsy	Umbilical cord: Length-42 cm, spiralling normal, frilling in the umbilical cord, cord hematoma, reddish discoloration caused by haemolysis. Report: Female fetus of approximately 34 weeks' gestational age, appropriate for age with no external congenital anomalies. Ascending colon with subhepatic caecum → suggestive of intestinal malrotation developmental anomaly. Comma-shaped left kidney → minor renal anomaly
7/8/25	Fetal radiological imaging X-ray	Normal
7/8/25	Chromosomal micro array	Normal karyotype
7/8/25	Histopathological examination	Third-trimester MCMA placenta with mature vascularized villi with intervillous fibrin deposition with dystrophic calcification, with central cord insertion. Both umbilical cords show three vessels. Membranes are unremarkable
CBC: Complete blood count, LFT: Liver function test, RFT: Renal function test, AFI: Amniotic fluid index, EFW: Estimated fetal weight, KFT: Kidney function test, Hb: Haemoglobin, WBC: White blood cell		



Figure 1. Monochorionic placenta with central insertion of two umbilical cords

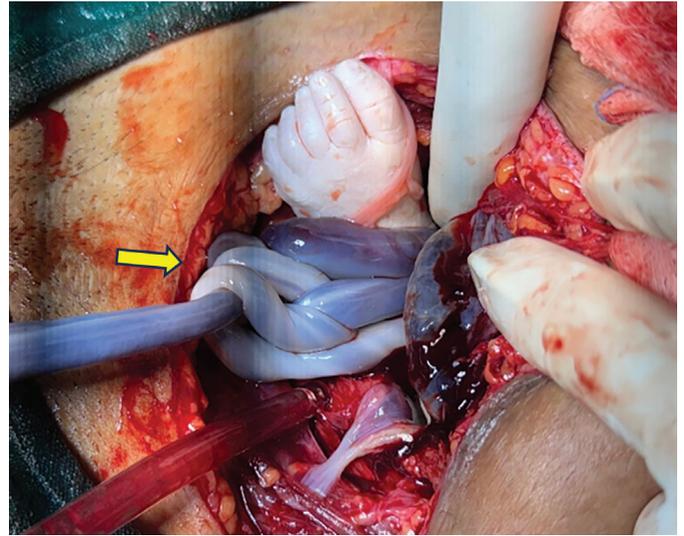


Figure 3. Cord entanglement around the deceased fetal foot (yellow arrow)

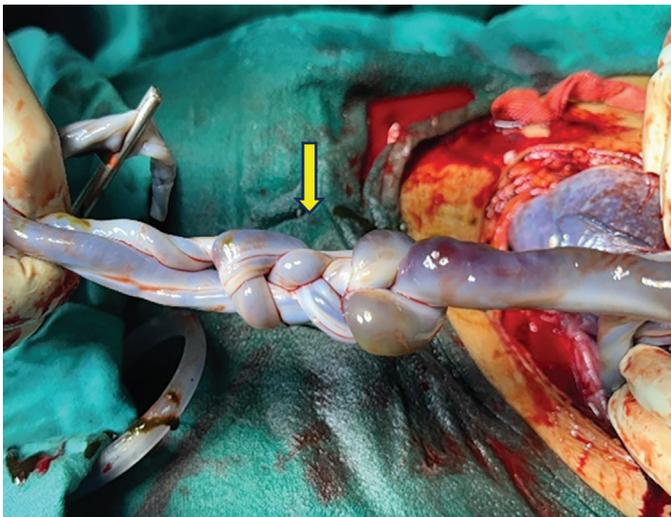


Figure 2. Twisted true umbilical cord knots (yellow arrow)

the knot lies within a visible area during sonographic scanning, the sonographer's expertise and vigilance, and the specific reporting practices of the ultrasound facility (3). However, a study by Rossi and Prefumo (6) reported that ultrasound detection of cord entanglement did not improve perinatal outcomes.

In the presented case, suboptimal monitoring, late referral and presence of undiagnosed complex braided knots contributed to the sudden intrauterine fetal demise. The incidence of single intrauterine demise occurs in 2.6-5% of all twin pregnancies (7). Intrauterine fetal demise in MCMA pregnancy has potentially significant morbid consequences for both the mother and for the co-twin, in the form of death of the surviving twin and neurological abnormality, at rates of up to 15% and 26%, respectively (2). Maternal complications



Figure 4. Live born female baby (yellow arrow), IUFD fetus (red arrow)

IUFD: Intrauterine fetal death

include coagulopathy, disseminated intravascular coagulation, iatrogenic preterm labour, and infection. Damage to the surviving co-twin after intrauterine fetal demise is explained by two mechanisms. These are thromboembolization through

Table 2. Other specific complications of MCMA twin pregnancy

Complications of MCMA twin pregnancies	Incidence
Selective growth restriction (sGR)	~20%
Twin-to-twin transfusion syndrome (TTTS)	~15–20%
Twin anaemia–polycythaemia sequence (TAPS)	~13%
Twin reversed arterial perfusion (TRAP) sequence	~1%
Cord accidents (entanglement/true knot)	~28–47%
MCMA: Monochorionic monoamniotic	

placental anastomoses causing coagulation disturbances, or acute hemodynamic imbalance due to shunt reversal into the demised twin, resulting in hypovolemic ischemic injury, predominantly neurological, in the survivor twin (8). Studies showed that the prognosis was better in dichorionic twin pregnancies compared with monochorionic twin pregnancies (9). Hillman et al. (7) reported that the frequency of mortality in the surviving fetus was 15% in monochorionic and 3% in dichorionic twin pregnancies.

Management guidelines recommend intensive antenatal surveillance by an ultrasound scan to measure fetal biometry and umbilical artery Doppler velocimetry at 2-weekly intervals from 16 weeks throughout the pregnancy, and the need to refer women to specialist centres if complications persist. Early elective Cesarean delivery planned between 32–34 weeks, may help to prevent unpredictable cord complications (2).

In a study conducted by Pasztor et al. (10), autopsy and placental examination identified the exact cause of stillbirth in 57.9% of cases. The genetic evaluation and autopsy reports mentioned no structural or chromosomal anomalies, supporting cord accident as the most likely cause of fetal demise.

In conclusion, better fetal outcomes can be achieved with greater ultrasound surveillance in a specialist fetal medicine centre. Moreover, there is a need for clinicians to counsel families about the unpredictability of cord accidents in MCMA pregnancies. Guidelines suggest that early delivery of MCMA twins between 32 and 34 weeks of gestation may reduce the risks and complications associated with this type of pregnancy. This is because umbilical cord entanglement is common in these uncommon pregnancies and is associated with an increased risk of stillbirth and intrauterine fetal demise.

Ethics

Informed Consent: Formal consent was obtained from the patient for publication purposes.

Footnotes

Conflict of Interest: No conflict of interest is declared by the authors.

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