

**Case Report**

**CORD ENTANGLEMENT: A COMPLICATION OF MONOCHORIONIC MONOAMNIOTIC PREGNANCY LEADING TO SECOND TRIMESTER PREGNANCY LOSS**

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**ABSTRACT**

The incidence of MAMC twins is 1 in 10,000 pregnancies and 1% of all twin pregnancies and it has been associated with a high incidence of perinatal mortality of 28-47%. Cord entanglement, a condition unique and specific to MAMC pregnancies, occurs in 23 % to 80% of the cases and it has been related to high perinatal mortality. We report here a case of cord entanglement at 24 weeks pregnancy leading to demise of both fetuses. Cord entanglement can be a major cause of fetal loss and therefore parental counseling is important. Early diagnosis, aggressive antenatal monitoring and termination of pregnancy after 32 weeks can improve perinatal losses.

**Keywords:** *Twin Pregnancy, Monochorionic Monoamniotic Twin Pregnancy, Cord Entanglement*

**INTRODUCTION**

The incidence of MAMC twins is 1 in 10,000 pregnancies (Cordero *et al.*, 2006) and 1% of all twin pregnancies. MAMC twins have been associated with a high incidence of perinatal mortality of 28-47% (Eserdag *et al.*, 2010). Perinatal mortality and perinatal morbidity is mainly due to cord accidents, fetal anomalies, prematurity, discordant anomaly, TRAP, TTS, twin locking, spontaneous miscarriages, and conjoint twins (Roque *et al.*, 2003; Hack *et al.*, 2009).

Cord entanglement, a condition unique and and specific to MAMC pregnancies, occurs in 23 % to 80% of the cases (Roque *et al.*, 2003; Pasquini *et al.*, 2006; Ezra *et al.*, 2005) and it has been related to high perinatal mortality (Eserdag *et al.*, 2010). There is still no consensus in literature for the management and the mode of delivery of these rarely encountered cases. We report here a case of cord entanglement at 24 weeks pregnancy leading to demise of both fetuses.

**CASES**

A primigravida presented in emergency with pregnancy of 24 weeks gestation and labor pains. She was an unbooked case. On general examination mild pallor was seen. Per abdomen examination uterus was 20-22 weeks size. She was having good uterine contractions.

Per vaginum examination revealed cervix 3-4 cms. dilated, fully effaced with bulging bag of membranes. No investigations or USG report was available with her. Her routine investigations done were normal. She expelled spontaneously after an hour, twin dead fetuses with entanglement of cord about 5-6 times. Peeling of skin was visible at places.

The cord was also seen around the neck of fetus (Figure 1). Placenta was single with no dividing membranes in between. The fetuses weighed around 750 gms each.

**DISCUSSION**

Incidence of twins is only 2.5% of all pregnancies (Cordero *et al.*, 2006). Only one-third of all twins are monozygotic and 75% of the monozygotic twins are monochorionic. Among monochorionic twins, about 2% are monochorionic-monoamniotic (MAMC) (Cordero *et al.*, 2006).

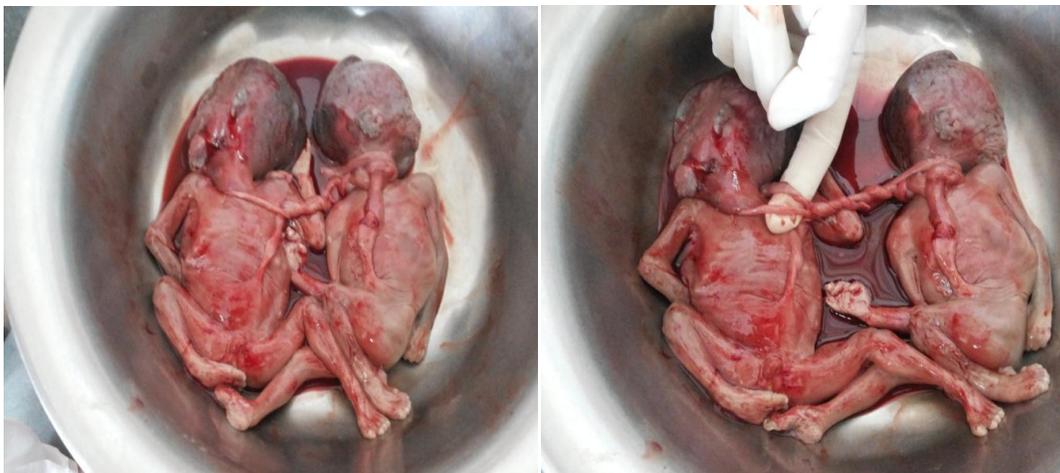
MAMC arise from a single blastocyst where the zygotic division occurred between 8-12 days. Monoamniotic twins are connected by two umbilical cords to a single placenta inside a single amniotic sac, hence, cord entanglement is expected resulting in cord compression, true knots, cord occlusion and associated with a high perinatal mortality.

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Mono-amniotic twins are at increased risk of unexpected fetal demise. In a series of mono-amniotic twins reported by Hack *et al.*, (2009) there were 22 intrauterine deaths, of which eight double fetal deaths and six single fetal deaths after 20 weeks. The cause was unknown or attributed to cord accidents in 7, while twin-to-twin transfusion syndrome and selective reduction for discordant anomalies accounted for the remaining. Ezra *et al.*, (2005) has reported cord entanglement as cause of intra-uterine death in 80%. In our case also fetal demise of both twins occurred at 24 weeks gestation.

Though, Rossi and Prefumo, (2013) in his systemic review, has concluded that cord entanglement does not contribute to prenatal morbidity and mortality in monoamniotic twin pregnancies. Prematurity, congenital abnormalities and twin-to-twin transfusion syndrome are the more common causes of perinatal mortality in MCMA twins after excluding lethal anomalies and pregnancies that had reached 20 weeks (Hack *et al.*, 2009). The low friction surfaces of the cords, the cushioning effect of Wharton's jelly and the resistance of the vessels to compression ameliorate compromise in umbilical circulation (Vijayan and Saraswathi, 2015).

Perinatal mortality in MAMC twins rises as gestation advances. It has been estimated that perinatal mortality rises at a rate of 2-4% per 2 week interval from 15-32 weeks, 11% from 33-35 weeks and 21.9% from 36-38 weeks therefore, premature delivery with steroid cover at 32 weeks has been advocated by Roque *et al.*, (2003). Case reports have indicated fetal demise at 30-31 weeks gestation (Aggarwal and Terhase, 2015).



**Figure 1: Monochorionic Monoamniotic Dead Twin Fetuses with Entanglement of Cord 5-6 Times, also around the Neck of both Fetus; Peeling of Skin Visible at Places**

### **Management**

No treatment can reverse MAMC twin condition. Hence, early diagnosis, regular and aggressive fetal surveillance and early elective cesarean is recommended to improve the outcome.

Early USG signs for diagnosis of MAMC include single gestation sac with single yolk sac. Presence of intertwin membrane excludes diagnosis of MAMC. A high fetal loss is anticipated and therefore, parental counselling should be done.

A detailed USG nearing 20 weeks including fetal heart chamber should be done. Aggressive monitoring starting at 25-26 weeks of gestation, when fetal viability is reached, can help to lower the risk of fetal death considerably (Jo *et al.*, 2011). Although, there are arguments to support a policy of increased surveillance, parents should be warned that demise can occur in between evaluations. The presumed mechanism is cord compression with or without acute intertwin transfusion, which may be detectable on fetal heart rate tracings and Doppler examination. Cord compression is expected to cause spontaneous decelerations. Doppler flow velocimetry reflect hemodynamic alteration in fetoplacental circulation secondary to narrowing of umbilical vessels involved in cord entanglement such as notch in umbilical

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artery waveform (David *et al.*, 2002). Dual gate Doppler seems to be more accurate than conventional single gate Doppler for diagnosis of cord entanglement, because of confirmation of 2 different heart rate at the same time (Ito *et al.*, 2017).

Reported time of delivery varies between 32-35 weeks and should be weighed against the risks of neonatal respiratory disorder. Death of a co-twin, doppler ultrasound reports may be taken into consideration for individualizing time of delivery.

According to the best available evidence, about 1 in 25 mono-amniotic pregnancies is complicated by fetal demise after 32 weeks. In comparison, the risk of neonatal death associated with preterm birth at 32 weeks is about 1 in 100. At this gestational age, therefore, delivery carries a smaller risk of death than remaining in utero. A similar analysis can be made for diamniotic twins: after 32 weeks, the risk of demise is about 1 in 80 in utero, so an elective preterm birth would generally not be beneficial in diamniotic twins (Lewi, 2010).

Perinatal mortality in MAMC twins is mainly a consequence of conjoined twins. TRAP, discordant anomaly and spontaneous miscarriages before 20 weeks gestation (Hack *et al.*, 2009; Dias *et al.*, 2010). Expectantly managed mono-amniotic twins after 20 weeks have a good outcome irrespective of the finding of cord entanglement. The practice of elective very preterm delivery or other interventions to prevent cord accidents in mono-amniotic twins therefore, should be re-evaluated (Dias *et al.*, 2010).

Though, there have been reports of successful vaginal deliveries with entangled cords and with true umbilical cord knots (Demaria *et al.*, 2004). RCOG has recommended delivery by cesarean section at 32 weeks. Caesarean section avoids the risk of cord prolapse and inadvertent clamping of entangled cord of 2<sup>nd</sup> twin.

An experimental drug Sulindac has been used for medical amnioreduction. This drug lowers the amount of fluid in the amniotic sac thereby reducing the amount of fetal movement. This is thought to lower the chances of cord entanglement or compression. However, this drug has not been studied in a large number of pregnancies and its potential side effects are unknown. Pasquini *et al.*, (2006) has reported 100% perinatal survival rate (after exclusion of all losses before 20 weeks' gestation) with use of sulindac therapy. If a severe anomaly is diagnosed in one twin, selective feticide can be performed by fetoscopic or ultrasound-guided coagulation and subsequent transection of the cord (Dias *et al.*, 2010; Demaria *et al.*, 2004).

### **Conclusion**

MAMC is an uncommon condition where high perinatal loss is expected. Cord entanglement can be a major cause of fetal loss and therefore, parental counseling is important. Early diagnosis, aggressive antenatal monitoring and termination of pregnancy after 32 weeks can improve perinatal losses. Newer technologies like fetal reduction and medical amnio-reduction might have saved such complication as in our case.

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