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Monochorionic twin pregnancies: a systematic approach to diagnosis and management

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Abstract

Background: Monochorionic Twins (MC) account for approximately 20% of all twin pregnancies and are associated with increased perinatal morbidity and mortality compared to dichorionic twins. Complications unique to these pregnancies include Twin-To-Twin Transfusion Syndrome (TTTS), Twin Polycythaemia Anaemia Sequence (TAPS), Selective Intrauterine Growth Restriction (sIUGR) and death of the co-twin. Adhering to a systematic and objective approach of management, can lead to early recognition and the commencement of appropriate interventions to prevent further morbidity.

***Objectives*:** This review highlights how complications specific to MC twins can be detected and suggests management strategies that can be instituted to improve outcomes.

Data source: The Medline, PubMed, Cochrane and TRIP databases were searched for original research and reviews on monochorionic pregnancies using the strategy; monochorionic pregnancy with/and complications OR management.

Data synthesis: Conference proceedings, case reports and personal communications were excluded though expert opinions from major bodies were considered. Only articles in English were included in this review. These were filtered for English language, human subjects and were restricted to the period 2000-2014.

Conclusion: Monochorionic twin pregnancies are associated with higher incidence of adverse perinatal outcomes compared to dichorionic pregnancies. With appropriate management and diagnostic strategies most of these complications can be recognized and prevented.

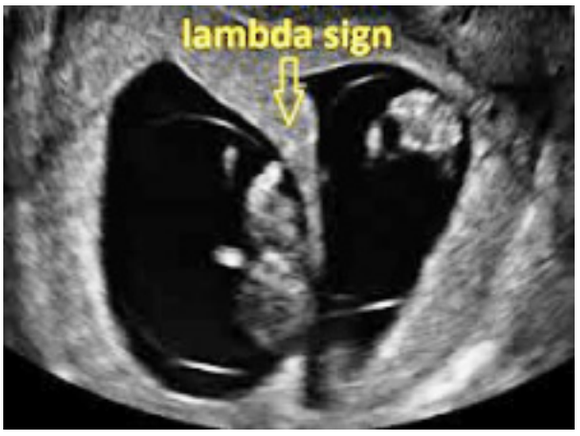
Introduction

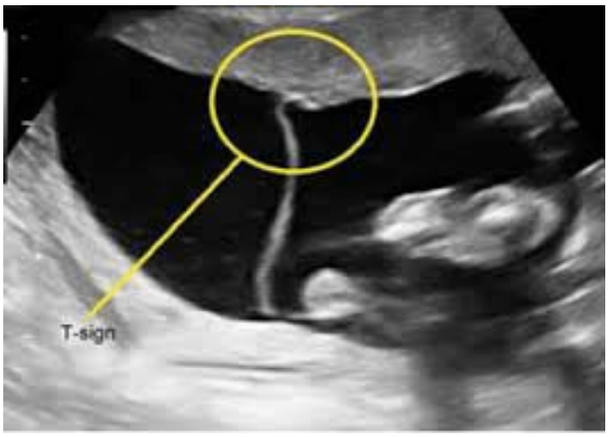
Monochorionic twins account for 1 in 250 of all pregnancies and 20% of twin gestation. One out of every twenty twin pregnancies due to assisted reproductive techniques is also monochorionic. While all monochorionic twins are monozygotic, about a third of monozygotic twins are dichorionic. This is due to early cleavage of the morula between day 1 and 3 resulting in separate placentae and amniotic sacs (1, 2). Compared to singletons, high order pregnancies are generally associated with increased perinatal mortality and morbidity but this risk tends to be much higher in monochorionic than in dichorionic pregnancies. A landmark observational study by Sebire and co-workers (3) reported that one out of every three monochorionic pregnancies will have complications. Furthermore, the perinatal mortality is twice as high in monochorionic pregnancy compared to dichorionic pregnancy. Monochorionic pregnancies are also 4 to 5 times more likely to suffer neurological morbidity compared to dichorionic pregnancy (3). There is therefore need for increased antenatal surveillance for all monochorionic pregnancies in order to identify these complications and initiate appropriate interventional strategies. This review presents a systematic approach to the diagnosis and management of monochorionic twin pregnancies.

Why determine chorionicity?

Chorionicity is key to the classification of twin pregnancy. This is best determined by ultrasound performed before 20 weeks. Beyond this gestation it is very difficult to define chorionicity with certainty. Monochorionic twins are identified by the presence of an inter-twin membrane….the T- sign...as opposed to the lambda or 'Twin-Peak' sign seen in dichorionic pregnancies (Figure 1).

Figure 1: The lambda/Twin Peak sign for dichorionic diamniotic (DCDA) twins and T sign depicting the monochorionic diamniotic (MCDA) twins





Misdiagnosis of chorionicity has been associated with a delay in recognition of complications such as Twin To Twin Transfusion Syndrome (TTTS). Consequently, perinatal mortality is much higher in pregnancies that had an initial wrong determination of chorionicity (4). Indeed, the Southwest Thames Obstetric Research Collaborative (STORK) study group concluded in one of their series that in the absence of structural abnormalities and aneuploidies, chorionicity was the single best determinant of adverse perinatal outcomes in twin pregnancies (5). It is therefore important that all twin pregnancies are classified as either monochorionic or dichorionic. This forms the basis for triage and subsequent management strategies. A diagnosis of twin pregnancy should therefore not be entertained without specification of chorionicity. In the event this is not possible due to late presentation (a common occurrence locally...), the author advises that the clinician should assume the pregnancy to be monochorionic to avoid unexpected outcomes.

Data source and synthesis

The Medline, PubMed, Cochrane and TRIP databases were searched for original research and reviews on monochorionic pregnancies using the following strategy; monochorionic pregnancy WITH/AND complications OR management.

Conference proceedings, case reports and personal communications were excluded. Consensus opinion and expert opinions by major obstetric societies were, however, considered. The articles were filtered for English language, human subjects and were restricted to the period ranging from 2000-2014 except for one landmark study conducted in 1997. Select articles on the pathogenesis of monochorionic twin gestation were included. Articles addressing controversies in the management of monochorionic pregnancies were excluded.

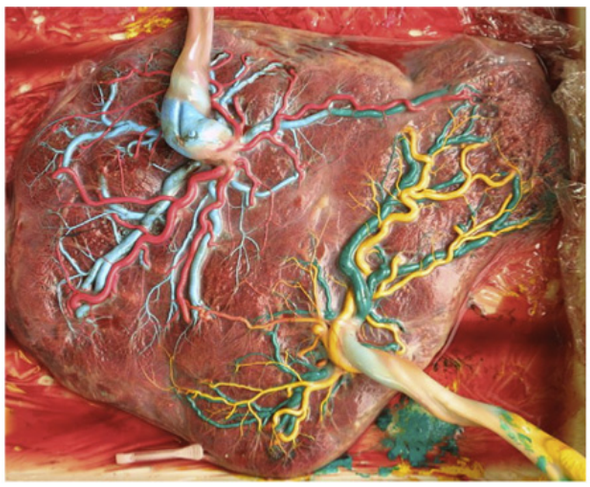
Prenatal follow-up for twin pregnancy

The prenatal follow-up for monochorionic twins should be more frequent. Initially this is done every 2 weeks until 26 weeks for early identification of TTTS. At each of these visits an ultrasound should be performed specifically for detection of differences in amniotic fluid, bladder size and abdominal circumference. From the third trimester, Middle Cerebral Artery (MCA) doppler should also be included to rule out Twin-Anaemia Polycythemia Sequence (TAPS). Follow-up scans should be performed ideally by personnel with experience in MC twins. The women should also have advanced scans done at 12, 20, 28 and 32 weeks. These scans should be performed by high level fetal medicine specialist or obstetricians/ radiologist with interest in fetal medicine and should include anatomy, fetal biometry and doppler. Fetal echocardiography should be considered, taking in mind the higher prevalence of cardiac defects in this population (6, 7).

Complications of monochorionic pregnancies and its management

Complications of monochorionic twins are either due to arterio-venous anastomosis or unequal placental sharing (Figure 2). Both of these phenomena result in unequal distribution of blood and hence nutrients in one twin at the expense of the other. The severity depends on the number and size of the anastomoses and the level of disparity in placental sharing (8, 9). These complications can however be recognized and appropriate management strategies instituted that could improve the prenatal outcomes of either one or both babies.

Figure 2: A monochorionic twin placenta following vascular injection studies, demonstrating multiple vascular anastomoses, including both clear arterio-arterial and arterio-venous communications (Reproduced with permission from Black M *et al 2010*)



1. Twin-to-Twin Transfusion Syndrome (TTTS)

TTTS occurs when one twin (the donor) pumps its blood into the recipient twin. This occurs via arterio-venous anastomosis and the outcome is the donor

twin becoming plethoric and anuric with severe oligohydramnios while the recipient is overloaded becoming hydropic with polyhydramnios and diuresis. This is associated with a more than 90% mortality of both twins and risk of severe neurological impairment in the survivors. The mother is also at an increased risk of developing mirror syndrome (4,10-13). TTTS commonly occurs between 16 and 26 weeks gestation. It is therefore safe to assume TTTS will not occur after this period.

The diagnostic criteria for TTTS

a. Confirmed monochorionic pregnancy at 11-14 weeks.
b. Polyhydramnios in the recipient twin with a maximum vertical pocket of >8cm (a cut off of 10cm after 20 weeks has been used by the Eurofetus group).
c. There is oligohydramnios in the donor with a maximum vertical pocket <2cm.
d. There is discordant fetal bladders with markedly enlarged bladder in the recipient while that of the donor is very small or non-visible.

The severity of TTTS was described by Quintero *et al* (14) as shown in Table 1.

Table 1: The Quintero classification of twin to twin transfusion syndrome

Stage	Classification
I	There is discrepancy in amniotic fluid volume with oligohydramnios of a maximum vertical pocket (MVP) < 2cm in one sac and polyhydramnios in the other sac (MVP >8cm) The bladder of the donor twin is visible and Dopppler studies are normal
II	The bladder of the donor twin is not visible (during length of examination, usually around 1 hour) but Doppler studies are not critically abnormal
III	Doppler studies are critically abnormal in either twin and are characterized as abnormal or reversed end-diastolic velocities in the umbilical artery, reverse flow in the ductus venous or pulsatile umbilical venous flow
IV	Ascites, pericardial or pleural effusion, scalp edema or overt hydrops present
V	One or both babies are dead

Management of TTTS

Fetoscopic laser coagulation of the placental anastomosis has been shown to be associated with up to 90% chances of survival of at least one fetus with minimal risk of neurological damage compared to amnio-drainage (15-17). The procedure involves fetoscopic visualization of the anastomoses and their coagulation. Previously this was done selectively i.e. only the anastomotic vessels were coagulated. This increased the risk of persistent deeper anastomoses that could eventually result either in recurrence of TTTS or later development of TAPS. To address this drawback, the Solomon's technique which involves coagulation along the entire placental equator has been described and current evidence reports it to be superior to selective coagulation (18). However, fetoscopy and laser coagulation are available in select centers even in the developed countries.

In the absence of fetoscopy it may still be logical to offer amnio-drainage. It is not clearly known how this intervention works, but it has been thought that draining the amniotic fluid from the polyhydramnios twin may relieve some pressure on the blood vessels especially the arterio-arterio anastomoses hence improving the blood flow to the other twin. This procedure is associated with upto 50% survival rate of at least one twin, however the neurological damage associated with the surviving twin is almost twice as high compared to fetoscopic laser coagulation (19). Other techniques such as septostomy have been abandoned.

2. Twin Anaemia Polycythemia Sequence (TAPS)

This condition occurs in 3-5% of monochorionic twins and is thought to be due to persisting anastomoses that lead to a state of chronic perfusion of one twin to the other (20). The result is one fetus being anaemic and the other being polycythemic. Unlike TTTS, this manifests during the late second and third trimester. As such the related mortality is less than that associated with TTTS. TAPS could also follow laser treatment for TTTS, when there are some persistent anastomoses after selective coagulation. The prognosis of the latter tends to be worse than the spontaneous type (21). The diagnostic criteria for TAPS includes: (22)

Prenatal: Middle Cerebral Artery Peak Systolic Velocity (MCA-PSV) >1.50 MoM in the donor and MCA-PSV <0.80 MoM in the recipient

Postnatal: Inter-twin haemoglobin difference >8.0g/dl and inter-twin reticulocyte count ratio (donor/recipient) >1.7

Management of TAPS

Most cases of TAPS, especially the spontaneous type, can be managed expectantly with favorable outcomes. Serial measurement of MCA-PSV could be used to determine any worsening condition and the Umbilical Artery (UA) and Ductus Venous (DV) waveforms should be used for the timing of delivery (23).

Intra-peritoneal transfusion of the anaemic fetus has been attempted with good results but it has been argued by some that such an intervention may not be beneficial as all it does is to provide the anaemic twin with more reservoirs to donate (24).

3. Selective intra-uterine growth restriction- sIUGR

Selective IUGR, complicates about 10% of monochorionic twin gestation and could result in the death of one twin if interventions are not timely. The pathophysiology behind sIUGR is unequal sharing of the placental territory. The diagnosis is made when there is >25% discordance in weight, although early recognition could be made by a discrepancy in abdominal circumference (25). A classification system (Table 2) to determine the severity and prognosis of sIUGR has been developed based on the characteristic of the umbilical artery wave forms into type I, II, III (26). The perinatal outcome is dependent on the severity.

Table 2: The Gratacos classification of selective intrauterine growth restriction-sIUGR in MC twins

Type	Description
I	Positive end-diastolic flow in the umbilical artery
II	Absent or reversed end diastolic flow constantly observed during all the examination
III	Intermittent absent or reversed end diastolic flow alternating over short periods of positive diastolic flow in the absence of fetal and maternal breathing

Changes UA Doppler waveforms in MC pregnancies cannot be interpreted in the same way as in other pregnancies as they represent a combination of the effects of placental insufficiency and those of inter-twin vascular connections. This fact has to be taken into consideration when interpreting these indices. However, Type I sIUGR seems to be associated with good outcomes and may be managed conservatively since the presence of a positive diastolic flow is reassuring. Type II and III have a worse prognosis and will almost always progressively deteriorate. This therefore calls for aggressive therapy before fetal demise ensues. However, the latency period between diagnosis of AEDV and delivery in these fetuses has been found to be much longer compared to singletons or dichorionic twins, probably due to the effect of 'rescue transfusions' form the larger twin thought the inter-twin blood flow interchange (7, 26).

Management of type II and III sIUGR

This could pose a difficulty ethical and medical dilemma. The treatment will depend on the gestational age, the severity of the IUGR and the wishes of the parents, taking into consideration that the demise of one fetus could adversely affect the other. The treatment of choice is to coagulate the cord of one twin. The aim is to prevent a situation where the surviving twin exsanguinates into the dead twin with a resultant ischemia of the latter's brain. Coagulation could be achieved by bipolar diathermy, laser or radio-frequency ablation. The main challenge is usually the affected twin is the one who has oligohydramnios and it is therefore difficult to access it. Amnioinfusion may improve accessibility to the amniotic sac.

Local legislation on selective termination of pregnancy has to be considered. Currently the laws in Kenya and indeed in whole of East and Central Africa are silent on the legality of such an intervention. This may change in time as we develop more capacity to recognize these complications. There are also limited neonatal intensive care units in the region and this poses a major challenge in the event elective preterm delivery is considered for the growth restricted fetuses, which is often the case.

4. Death of one twin

The risk of co-twin death is 12% in monochorionic pregnancy compared to 4% in dichorionic pregnancy (27). Death of the co-twin could either be related to sIUGR or other underlying maternal/fetal conditions. The sequela of one twin dying is that the surviving twin usually exsanguinates into the dead twin as the latter forms a low pressure zone. The result is ischemia of the vital organs especially the brain resulting in severe neurological damage in up to 20% of the survivors (28). The timing of the exsanguination is unknown but it is thought to be within few minutes of death. The problem is the extent of neurological damage does not manifest acutely and therefore one has to wait for up to 4 weeks before scanning the fetus to determine presence of any lesion in the brain. Where available fetal MRI could be used to determine the extent of this damage as early as one week after the insult. Any decision to terminate the pregnancy is best undertaken after confirming the presence of periventricular leukomalacia or other cerebral lesions.

5. Aneuploides

The diagnosis and management of aneuploidies in monochorionic twins is beyond the scope of this review, however it is important to note that this could occur. This has questioned the previous thoughts that monozygotic twins share the same genetic material. Such a discrepancy also poses an ethical dilemma of whether to end the life of one of the twins at the expense of putting the normal one at risk of preterm birth and miscarriage, or letting both of them live but still endangering the normal twin with risk of neurological damage in the event the other affected twin dies *in utero*.

6. Preterm labour

Preterm labour, whether spontaneous or as a result of obstetric intervention remains a common complication of twin gestation, the details of which are beyond the scope of this review. Most studies on prevention of preterm labour have mainly focused on singleton pregnancies, though secondary analysis of data has found some strategies such as cervical cerclage and progesterone not to confer any benefit to high order pregnancies. The use of antenatal corticosteroids in twin gestation for lung maturity is also extrapolated from studies involving singletons with very little research addressing the effect of steroids in multiple pregnancies (29).

Conclusion

Monochorionic twin pregnancies are associated with higher incidence of adverse perinatal outcomes compared to dichorionic pregnancies. With advances in prenatal care, most of these complications can now be managed with favorable perinatal outcomes. However management will depend on chorionicity and therefore its determination is key in the identifications of these types of pregnancies with timely referral to specialized care in fetal medicine centers for early identification and management of any complications that may arise.

Disclosure of interest

None to declare

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