Third-trimester ultrasound diagnosis of twin-to-twin transfusion syndrome (TTTS) - A review of two cases

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ABSTRACT
Twin-to-twin transfusion syndrome (TTTS) is an unbalanced net transfusion of blood between twin fetuses through placental anastomoses. It is a complication of monochorionic diamniotic (MCDA) twin gestation in which one of the twins is the donor while the other twin acts as the recipient. TTTS without treatment is a major cause of increased morbidity and mortality among MCDA twin pregnancies. Ultrasound diagnosis and staging of TTTS are based on Quintero staging which influences the choice of management. We present two cases of ultrasound diagnosed TTTS. A 27-year-old G2P1+0 with twin gestation who had an ultrasound scan at 34 weeks gestation, and a diagnosis of Quintero stage 1 TTTS was made. She had an emergency caesarian operation and was delivered of two live babies. A 24-year-old G1P0+0 with twin gestation who had an ultrasound scan at 35 weeks gestation, with a diagnosis of Quintero stage 5TTTS. The donor twin was alive while the recipient twin had hydrops fetalis with no cardiac activity. An emergency caesarian section was done and the donor twin survived. TTTS is a common complication among MCDA twins and it is a cause of increased morbidity and mortality among them. Early diagnosis through ultrasonography and prompt management will help reduce the morbidity and mortality associated with the syndrome.

Key words: Third trimester; twin-to-twin transfusion syndrome; ultrasound.

Introduction
Twin-to-twin transfusion syndrome (TTTS) is an unbalanced net transfusion of blood between twin fetuses through placental anastomoses occurring between 15 and 26 weeks of gestation. It affects majorly the monochorionic diamniotic (MCDA) twin gestation, although it has been described in some monochorionic monoamniotic (MCMA) twin pregnancies. About 10%–15% of monochorionic pregnancies will develop TTTS. In the 19th century, Friedrich Schatz, a German obstetrician, extensively studied the twinning process and was the first to postulate a relationship between vascular anastomoses and the development of TTTS. Monochorionic twins normally exchange blood during gestation, and two patterns of intertetal transfusion have been described based on ultrasound studies. The most frequent pattern which occurs in 90% of cases results from constant but balanced bidirectional interfetal transfusion between the twins. This means that transfusion from one is counterbalanced by transfusion from the other. In 10% of cases, a chronic imbalance in net flow develops, resulting in twin-to-twin transfusion (TTT). If this condition progresses, the recipient twin develops a generalized swelling which increases the risk of heart failure and hydrops fetalis. There is also polyhydramnios due to increased urinary output. Meanwhile, the donor twin remains small and underdeveloped with decrease in urinary output, which...

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may lead to renal failure and oligohydramnios. One cannot obviously expect the classical oligo-polyhydramnios sequence observed in MCDA twins in MCMA twins, so the diagnosis is based on the observation of polyhydramnios and discordant bladder size.[4]

A staging method for TTTS was developed by Quintero which is now widely used in assessing the severity of the syndrome and when to intervene.[5] The current treatment modalities for TTTS include serial amniodrenchure[6] and fetoscopic laser coagulation of vascular anastomoses.[7] Without treatment, TTTS is a major cause of increased morbidity and mortality among MCDA twin pregnancy.[5-7]

We present two MCDA twin gestations with TTTS diagnosed by ultrasound in the third trimester.

**Case study 1**

A 27-year-old G2P1+0 (1A) presented to the ultrasound unit of the antenatal clinic of the University College Hospital for a routine third-trimester ultrasound scan. The pregnancy was booked at 29 weeks + 1 day gestational age (GA) at the antenatal clinic of the University College Hospital, Ibadan. There were no complaints so far during the course of pregnancy. She had two previous ultrasound scans in private clinics which revealed normal development of the twin gestation.

Ultrasound findings revealed live twin gestation at a GA of 34 weeks + 1 day; they were in two different sacs; the first twin [Figure 1] had increased amniotic fluid around it with an unstable lie. The deepest vertical pool (DVP) was 11.1 cm and the placenta was seen anteriorly. The fetal heart rate was 135 bpm with regular rhythm. The urinary bladder was seen and the estimated fetal weight (EFW) was 2.25 kg.

The second twin [Figure 2] appeared stuck to the superior portion of the uterine cavity with minimal movement and minimal fluid seen around it. The DVP was 2.2 cm. Minimal urine was noticed within the urinary bladder. The fetal heart rate was 148 bpm. Doppler of both umbilical and middle cerebral arteries was done and they showed normal Doppler indices. Figure 3 shows Doppler ultrasound of the middle cerebral artery of the donor twin. EFW was 2.2 kg. The cervical os was closed. An impression of MCDA twin gestation at 34 weeks GA with stage 1 TTTS was made.

The patient was referred to the emergency unit of the antenatal clinic where she was scheduled for an emergency lower segment caesarean which was done the same day. She was delivered of two live male neonates. Twin I (recipient twin) weighed 2.25 kg with Apgar score of 6/1 9/5, while twin II (donor twin) also weighed 2.25 kg with Apgar score of 8/1 9/5. Both babies were discharged home on the fourth day of life.

**Case study 2**

A 24-year-old primigravida with twin gestation presented
for routine ultrasound scan at 35 weeks GA at the ultrasound unit of the antenatal clinic of the University College Hospital, Ibadan. She booked the index pregnancy at 14 weeks GA and had four previous ultrasound scans at outside facilities which revealed MCDA twin gestation. No abnormality was detected.

Our ultrasound finding revealed twin female fetuses at a GA of 34 weeks + 1 day; they were in two different sacs; the first twin [Figure 4] showed no cardiac activity and it showed fluid within both pericardial and peritoneal cavities with skin edema. There was increased amniotic fluid around it with DVP of 12.3 cm [Figure 5a].

The second twin [Figure 5b] appeared stuck to the superior portion of the uterine cavity with minimal movement and minimal fluid seen around it with a DVP of 1.8 cm. Minimal urine was noticed within the urinary bladder. The fetal heart rate was 152 bpm and EFW was 2.5 kg, which was small for the GA and suggestive of some form of intrauterine growth restriction (IUGR). Doppler ultrasound of the umbilical artery, however, showed normal Doppler indices [Figure 6]. An impression of MCDA twin gestation at 34 weeks GA with TTTS Quintero stage 5 was made. An emergency caesarian section was done the same day and the donor twin survived with an Apgar score of 81 and fetal weight of 2.65 kg. She was discharged home on the fifth day of life, hale and hearty.

**Discussion**

TTTS, also known as feto-fetal transfusion syndrome and twin oligohydramnios-polyhydramnios sequence is a complication of disproportionate blood supply between monochorionic twins, which results in high morbidity and mortality.[1-4] The presence of placental vascular anastomoses is an etiological factor for the development of TTTS.[3] Injection studies of twin placentaes have shown that such anastomoses are almost invariably present in monochorionic twins and extremely rare in dichorionic twins.[3,7-9]

Three types of anastomoses have been documented in monochorionic twin pregnancies: from artery to artery (AA), from vein to vein (VV), and from artery to vein (AV). AV anastomoses are unidirectional and are referred to as “deep” anastomoses since they proceed through a shared placental cotyledon, whereas AA and VV anastomoses are bidirectional and are referred to as “superficial” since they lie on the chorionic plate. AV anastomoses are found in 90%-95% of MCDA pregnancies, AA in 85%-90%, and VV in 15%-20%. TTTS is caused by net imbalance of blood flow between the twins due to AV anastomoses.[8-10]

The classification of TTTS is currently defined by a staging method by Quintero et al.[5-11] which is based on ultrasound and Doppler studies [Table 1]. Five possible stages have been described in the progression of TTTS. This staging method is now widely used in assessing the severity of the syndrome and when to intervene. There has been a lot of debate about the Quintero staging method; it has been said that stage 1 does not necessarily have the best outcomes because some degrees of cardiac dysfunction have been noted within this stage. Another criticism is that it does not accurately
TTTS usually presents in the second trimester and it is a dynamic condition that can remain stable throughout gestation, may regress spontaneously, progresses slowly over the weeks, or rapidly within a period of days with rapid deterioration in the well-being of the twins. Both cases presented were detected in the third trimester, which may be as a result of lack of expertise by the sonographers who scanned them before their presentation at our facility.

The management options for TTTS include expectant management, amnioreduction, fetoscopic laser photocoagulation of placenta anastomoses, septostomy, and selective reduction. The management option varies depending on the GA and stage at the time of diagnosis. Recommendations about the timing of delivery vary and many factors such as the response to treatment, fetal growth, and disease progression determine the timing of delivery. However, the median GA at delivery for most studies which used laser treatment was 33–34 weeks, especially in severe cases. For stages I and II with reassuring surveillance, delivery is delayed till 34–37 weeks. The two cases presented had emergency caesarean sections as soon as the diagnosis was made. This was due to the fact that both were detected beyond fetal viability in the third trimester and also because surveillance and other less invasive therapeutic facilities are not available and routinely practiced at our center. This underscores the need for manpower development and funding in the area of feto-maternal care in the tropics.

**Conclusion**

TTTS is a common complication among MCDA twins and it is a cause of increased morbidity and mortality among them. Early sonographic diagnosis and prompt management by well-trained and equipped obstetric team would in no small measure stem the tide in the rising morbidity and mortality associated with the condition especially in low-income countries.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Table 1: Quintero staging**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Ultrasound parameter</th>
<th>Criteria</th>
</tr>
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<tbody>
<tr>
<td>I</td>
<td>DVP of amniotic fluid</td>
<td>DVP &lt; 2 cm in donor sac; DVP &gt; 8 cm in recipient sac</td>
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<tr>
<td>II</td>
<td>Fetal bladder</td>
<td>Non-visualization of fetal bladder in donor twin over 60 min of observation</td>
</tr>
<tr>
<td>III</td>
<td>Umbilical artery, ductus venosus, and umbilical vein Doppler waveforms</td>
<td>Absent or reversed umbilical artery diastolic flow, reversed ductus venosus a-wave flow, pulsatile umbilical vein flow</td>
</tr>
<tr>
<td>IV</td>
<td>Fetal hydrops</td>
<td>Hydrops in one or both twins</td>
</tr>
<tr>
<td>V</td>
<td>Absent fetal cardiac activity</td>
<td>Fetal demise in one or both twins</td>
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</tbody>
</table>

DVP: Deepest vertical pocket
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Conflicts of interest
There are no conflicts of interest.

References