

## Case Report

### Twin to Twin Transfusion Syndrome (TTTS) : A high index of suspicion is mandatory

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#### Abstract

Twin-Twin transfusion syndrome (TTTS) leads to high rates of perinatal morbidity and mortality due to its poorly understood etiology and difficulty in diagnosis and treatment. Early diagnosis during foetal ultrasound is, therefore important in reducing the morbidity and mortality rates. TTTS is a phenomenon almost exclusive to monochorionic twin pregnancies. TTTS is associated with high rates of perinatal mortality. The donor twin is characterized by oliguria, oligohydramnios or anhydramnios, growth restriction and abnormal umbilical artery by Doppler velocimetry. The recipient, on the other hand, is characterized by polyuria polyhydramnios abnormal venous dopplers cardiac enlargement /failure, and eventually hydrops. The present study reported a case of TTTS an unusual presentation.

**Key words:** : Monochorionic twin, Twin Discordance, Stuck twin

#### Introduction

Twin-to-twin transfusion syndrome (TTTS) is a rare complication of pregnancy that develops in 10-15% of twins when the babies share the same placenta (monochorionic). TTTS occurs in 5-38% of monochorionic twins<sup>(1)</sup>. It may develop any time in the second or third trimester of pregnancy. The diagnosis of twin-twin transfusion syndrome is largely based on sonographic finding of disparity in liquor volumes between the two amniotic sacs. The extreme variant of TTTS, the "stuck twin" phenomenon has been reported in 8% of twin pregnancies as seen in our case. Outcome is dependent upon gestational age at birth, the lower the gestational age at birth, the greater the risk for long standing neurologic or pulmonary sequelae.<sup>[1]</sup> Monochorionic twin pregnancies are considered high risk situations and require close monitoring. Here we present a case of gravida4 para1abortion2 with 34-36weeks of twin gestation with TTTS stage3 with stuck twin A (donor). Due to rarity of presentation of stuck twin phenomenon & its unusual presentation in late third trimester, we are reporting this case.

#### Case History

G4P1L1A2 was referred to our hospital at 8 ½ months of amenorrhoea with scan report showing

Mono Chorionic Diamniotic twin (MCDA) gestation with growth discrepancy between the twins and doppler changes. She had her ANC's outside and her scan at 8wks had showed twin gestation (chorionicity not mentioned). Subsequent scan done at 16 wks was normal without any discrepancy in growth or liquor. She had no history suggestive of TTTS like abdominal distension, respiratory distress or preterm labour. Previous obstetric history - 1st pregnancy- full term normal delivery, male baby 3 years alive & healthy, 2nd pregnancy-spontaneous abortion at 2months, 3rd pregnancy MTP at 40days of gestation. Her previous cycles were regular LMP - 20/03/13, EDD- 27/12/13. There was no history of twinning in the family. On examination she was moderately built and nourished. Pallor +, No pedal oedema, PR-72/min, BP-120/70mmHg, RS-normal, CVS-S1S2 heard, P/A- uterus was over distended, relaxed, SFH 38cms, AG 102cms, multiple fetal parts felt, FHS of one fetus heard distinctly which was 132bpm. P/V -cervix posterior, uneffaced, os closed. On impression, to rule out whether it is G4P1L1A2 at 36 weeks with twin pregnancy or TTTS or Single fetal demise. On investigations, Blood group - O Positive, Hb-10g/dl, Serology was negative, Urine routine is normal.

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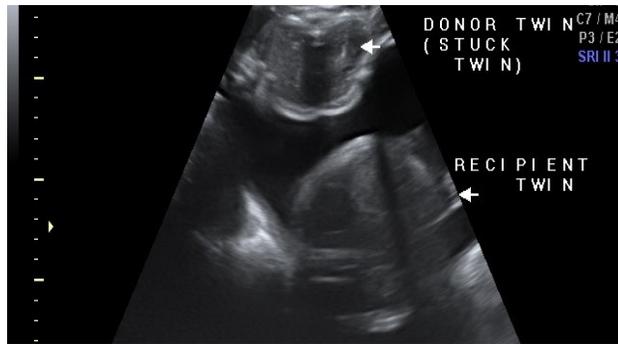


Fig:1 – Showing Twin A (Donor twin, Stuck twin) & Twin B (recipient twin)

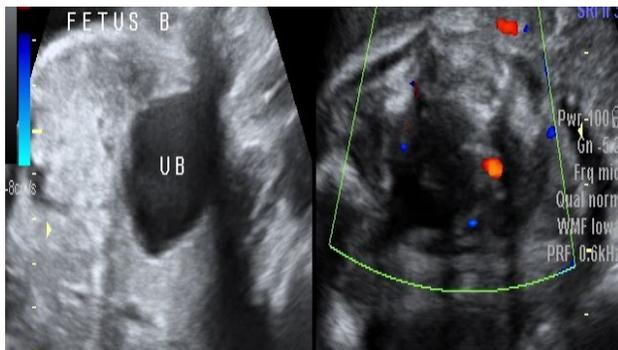


Fig:2 – showing Fetus “B” recipient twin with full bladder, fetus “A” showing twin with empty bladder



Fig:3 – Showing gross difference between the weights of the babies

On performing scanning in our hospital we found that MCDA twin – Twin A (Donor twin) (stuck twin) – cephalic presentation, corresponding to 29 weeks, small for gestational age with anhydramnios (Deepest Vertical Pocket-0), reversal of flow in the umbilical artery, empty urinary bladder. Twin B (Recipient twin) – breech presentation, corresponding to 34 weeks, polyhydramnios (Deepest Vertical Pocket -12cms), over distended urinary bladder and pulsatile umbilical vein waveforms. TTTS grade 3 with stuck donor twin. With adequate blood patient was taken up for emergency LSCS – 1st twin delivered by breech extraction with excess liquor, 2nd twin with membrane tightly wrapped around the

baby was cephalic presentation. Baby details- Recipient twin (1<sup>st</sup> twin) - female, 2.01kg good APGAR. Donor twin (2<sup>nd</sup> twin, stuck twin) - female, 1.3kg good APGAR. Placenta - Single with thin intervening membrane suggestive of MCDA placenta. Recipient twin – Echo showed VSD, USG abdomen showed persistent full bladder. There was difference of > 5g Hb between the twins, weight difference of 0.7kg(40 %). Blood group of the babies - O Positive.

## Discussion

Approximately 75% of monozygotic twins are monochorionic. Twin to twin transfusion syndrome (TTTS) is a specific condition complicating 10-15% of monochorionic multiple pregnancy<sup>(1)</sup>. Additional findings may include a stuck donor with non-visualization of the bladder, arterial and/or venous Doppler abnormalities, hydrops, and an absence of arterioarterial anastomoses. The natural history of severe TTTS is well established. Mortality rates approach 80 to 100 percent if left untreated, especially when it presents at less than 20 weeks gestation<sup>(2)</sup>. Vascular anastomoses are present in virtually 100% of monochorionic twin pregnancies, whereas twin-twin transfusion occurs (with rare exception) in 5-10% of monochorionic pregnancies<sup>(3, 4)</sup>. The reason for the occurrence of twin-twin transfusion syndrome in only a small proportion of the monochorionic twin pregnancies with vascular anastomoses is unknown. The intraplacental anastomosis is usually situated in a single, shared cotyledon of the common placenta, and is usually arteriovenous but may be arterial-arterial anastomoses. A distinction between the acute and chronic forms of the syndrome can be made on the basis of weight discrepancy and haemodynamic changes. Infants with the chronic form have discrepancies in birth weight exceeding 15% and the peripheral blood film of the donor twin may show hypochromic microcytic anaemia and erythroblastosis<sup>(1)</sup>. TTTS has been classified into 5 stages based on sonographic findings by Quintero et al<sup>(5)</sup>.

The best treatment of cases presenting before 26 weeks of gestation is fetoscopic laser ablation of the intertwin anastomoses on the chorionic plate<sup>(7)</sup>. There are no formal contraindications of laser therapy, although a short cervical length (<15 mm) may indicate a higher-risk of preterm delivery. Preliminary evidence suggests that cervical cerclage might reduce this risk. Moreover, perinatal survival rates after laser therapy were higher when the TTTS was Stage 1 or 2 than when it was Stage 3 or 4. Increasingly, therefore, attention is being paid to the timely diagnosis and treatment of TTTS<sup>(6)</sup>. In the absence of complications after laser treatment, planned delivery is recommended from 34 weeks and no later than 37 weeks. There is

controversy about the best way to manage TTTS diagnosed early in the 3rd trimester. Apart from expectant management or amniodrainage, elective delivery is an option.

However TTTS is a relatively rare complication occurring in third trimester<sup>(8)</sup>. This is particularly troublesome given that two structurally normal fetuses are involved. Hence it poses a management challenge. Clinicians caring for women with monochorionic pregnancies should have a strong clinical suspicion for TTTS. Sonographic signs of monochorionic diamniotic twins include a single placenta, a thin dividing membrane, a "T"-sign, and gender concordance. Before ruling out monochorionic monoamniotic twins in cases where no dividing membrane is seen, a diligent search for a thin membrane tightly wrapped around one twin should be performed.

Other sonographic findings include

1. Abnormal amniotic fluid volume
  - a) One sac with oligohydramnios, deepest vertical pocket < 2.0cm
  - b) One sac with polyhydramnios, deepest vertical pocket > 8.0cm
2. Persistent urinary bladder findings
  - a) Small or no bladder visualized in twin with oligohydramnios
  - b) Large bladder visualized in twin with polyhydramnios
3. Estimated fetal weight discordance (>20% of larger twin's estimated weight)
4. Appearance of a "stucktwin"
5. Hydropsfetalis (presence of one or more of the following in either twin)
  - a) Skin edema (> 5mm thickness of scalp skin)
  - b) Pericardial effusion
  - c) Pleural effusion
  - d) Ascites.

Other sonographic findings that may prove to be of prognostic significance include - The presence of a hypertrophied, dilated heart, with absence or reversal of flow in the ductus venosus during atrial contraction. In the donor, the heart may be dilated, the bowel is hyperechogenic, and there is absent end-diastolic flow in the umbilical artery. Outcome is dependent upon gestational age at birth, the lower the gestational age at birth, the greater the risk for long standing neurologic or pulmonary sequelae. Monochorionic twin pregnancies are considered high risk situations and require close monitoring.

## Conclusion

Appropriately timed diagnosis of twin-twin transfusion syndrome is crucial because a delay in diagnosis may result in a delay in treatment and increased

perinatal mortality and morbidity. To accomplish this we have to do 2weekly monitoring by ultrasound complimented by Doppler where necessary once monochorionic diamniotic twin is diagnosed.

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