DOI: 10.7860/IJARS/2020/45098:2561

Case Report

Radiology Section

Twin-Twin Transfusion Syndrome: Antenatal Sonographic Diagnosis-A Case Report

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ABSTRACT

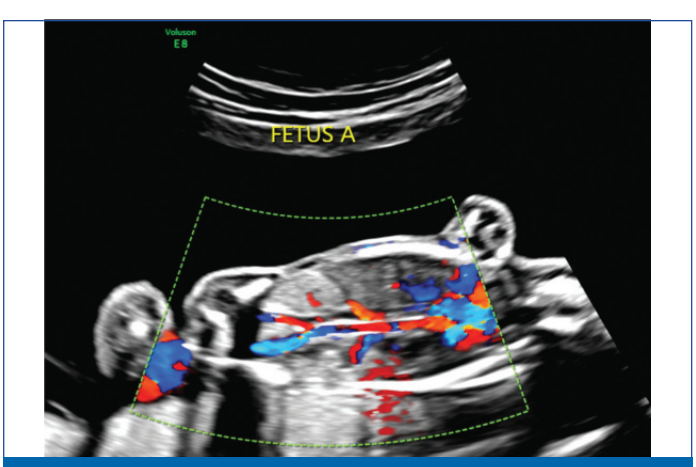
Twin-Twin Transfusion Syndrome (TTTS) is a complication seen in monochorionic pregnancies due to unbalanced vascular anastomosis in placenta. It results in high perinatal mortality, thereby, warranting an early diagnosis. It has an incidence of 1-3 per 10,000 births. A case of TTTS is presented which was diagnosed on fetal ultrasound at 20 weeks of gestation. The donor twin had oligohydramnios with collapsed urinary bladder throughout the examination. The recipient twin had polyhydramnios, mitral as well as tricuspid regurgitation, pulmonary artery stenosis, high resistance flow in the umblical artery, reversal of A wave in ductus venosus and fetal ascites. To make the diagnosis of TTTS there should be a presence of monochorionic diamniotic pregnancy with polyhydramnios in the recipient twin and oligohydramnios in the donor twin.

Keywords: Monochorionic, Polyhydraminos, Ultrasound

CASE REPORT

A 20-year-old healthy primigravida presented to the Department of Radiodiagnosis and Imaging of the Institute for second trimester anomaly scan. Her gestational age was 20 weeks 1 day according to her last menstrual periods. There was no significant or relevant past medical or surgical history. The patient was afebrile with blood pressure of 110/86 mmHg, heart rate of 78/minute, respiratory rate of 19/minute and SpO2 of 99%. The ultrasound examination revealed two live intrauterine fetuses separated by a thin membrane separating the two. The placenta was single and was along the posterior wall of uterus. The fetus towards right side of mother was labelled as twin A. It was seen in cephalic presentation and had mean gestational age according to ultrasonographic biometry of 18 weeks 4 days±10 days and estimated fetal weight of 247±36 grams.

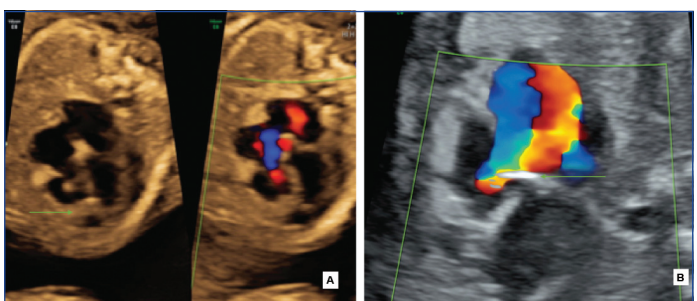
The amniotic fluid was reduced with single deepest cord free vertical pocket of 1.9 cm. The fetal heart rate on M mode was 150-160 bpm. Fetal heart situs and axis was normal. The four chamber view showed normal symmetry of the chambers. Three vessel views showed normal orientation and size of the main pulmonary artery, aorta and the superior vena cava. The Right and Left Ventricular Outflow Tracts (RVOT and LVOT) were normal. No evidence of valvular regurgitation was seen. Ductus venosus showed normal waveform on spectral Doppler. The fetal kidneys appeared normal. The fetal urinary bladder was seen in collapsed state during the one hour course of examination [Table/Fig-1]. No evidence of oedema, pleural effusion or fetal ascites was seen.



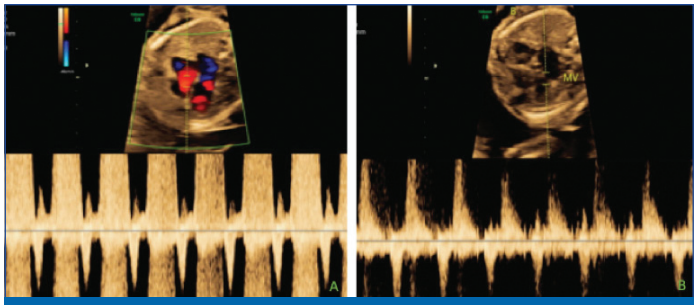
[Table/Fig-1]: Ultrasonographic image showing collapsed urinary bladder in twin A.

The fetus towards left side of mother was labelled as twin B. It was seen in transverse lie with mean gestational age according to ultrasonographic biometry of 19 weeks±10 days and estimated fetal weight of 347±51 grams. The fetal heart rate on M mode was 137-147 bpm. The amniotic fluid was on higher side with single deepest cord free vertical pocket of 8.2 cm. Fetal heart size was enlarged with cardio-thoracic ratio of 0.65. It was normal in situs, position and axis. On four chamber view, the right ventricle of fetal heart showed thickened echogenic walls- suggestive of hypertrophy. The left ventricle showed mild thickening of its walls. There was thickening with raised echogenicity of the tricuspid valve leaflets- suggestive of dysplastic valve with evidence of tricuspid regurgitation seen on Color Flow Imaging (CFI) [Table/Fig-2]. Mitral valve also appeared echogenic with evidence of mild mitral regurgitation [Table/Fig-3].

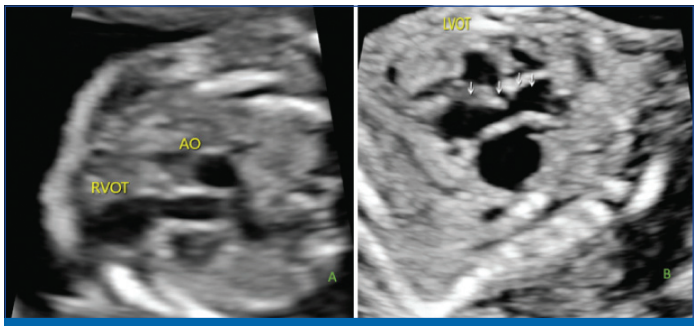
RVOT appeared narrowed with smaller diameter of the pulmonary artery in comparison to the aorta on three vessel view- suggestive of pulmonary artery stenosis [Table/Fig-4]. Spectral Doppler showed reversal of flow on CFI. LVOT appeared grossly normal. Colour doppler revealed high resistance flow in the umblical artery with Resistive Index (RI) -0.88, Pulsatility Index (PI)- 2.09 and systolic/diastolic (S/D)- 8.53. However, no reversal of end diastolic flow was seen [Table/Fig-5]. Ductus venosus showed reversal of A wave on spectral Doppler [Table/Fig-6]. Umblical vein showed pulsatile flow. Urinary bladder of fetus was normally visualised. There was free fluid seen in fetal peritoneal cavity [Table/Fig-7].



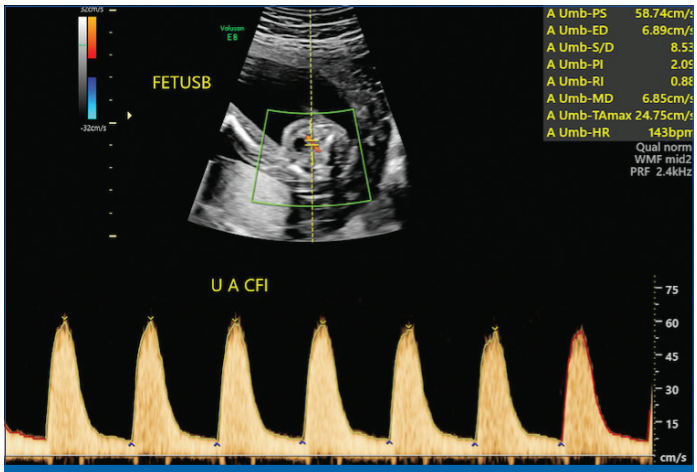
[Table/Fig-2]: Ultrasonographic images showing: a) right ventricular hypertrophy with aliasing of blood flow at tricuspid valve; b) Dysplastic tricuspid valve with tricuspid regurgitation.



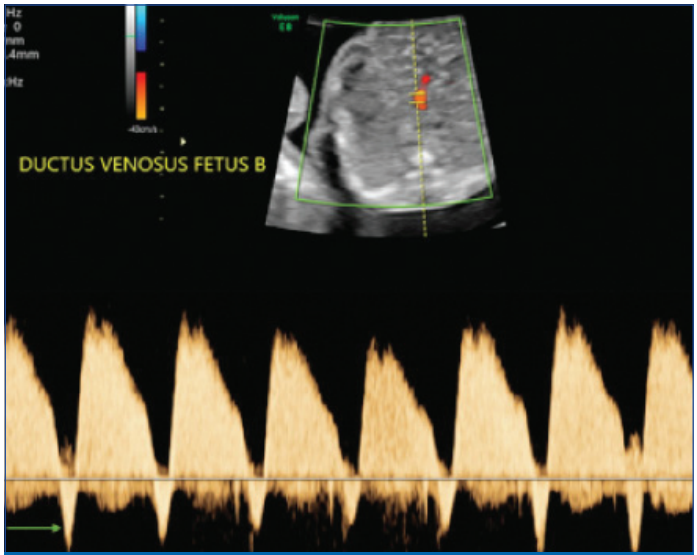
[Table/Fig-3]: Doppler ultrasound images showing: a) tricuspid regurgitation; and b) mild mitral regurgitation.



[Table/Fig-4]: Ultrasonographic images showing: a) narrowing of right ventricular outflow tract (RVOT) – suggestive of pulmonary stenosis and b) normal left ventricular outflow tract (LVOT).

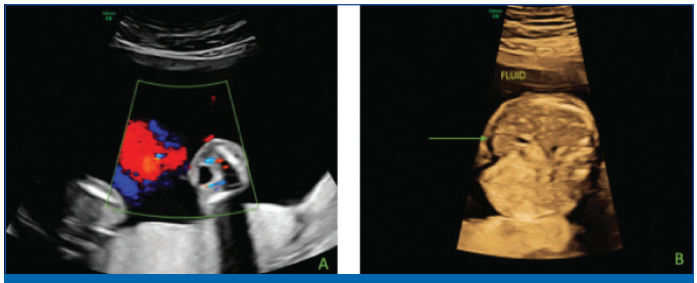


[Table/Fig-5]: Doppler ultrasound image showing high resistance flow in the umblical artery.



[Table/Fig-6]: Doppler ultrasonographic image showing reversal of A wave in ductus venosus.

On the basis of the above findings, a diagnosis of TTTS was made with twin A as donor and twin B as recipient. The mother got the pregnancy terminated at 20 weeks after consultation with the treating obstetrician.



[Table/Fig-7]: Ultrasonographic images showing: a) polyhydramnios with normal urinary bladder; b) fetal ascites.

DISCUSSION

TTTS is the result of intrauterine blood transfusion from one twin (donor) to another twin (recipient) [1]. It occurs in multiple gestation and involves chronic flow of blood from one twin to its co-twin, the prevalence of TTTS is approximately 1 to 3 per 10,000 births [2]. The aetiology is suggested to be an imbalance in the formation of arterial and venous connections of both fetuses with a single placenta. It is associated with a high perinatal mortality and neonatal morbidity and high incidence of long term neurodevelopmental morbidity in survivors [3].

The diagnosis of TTTS requires the presence of monochorionic diamniotic pregnancy, weight discrepancy between the fetuses (difference more than 20%) and oligohydramnios in one sac and polyhydramnios in the other. Oligohydramnios is defined as a maximal vertical pocket of <2 cm and polyhydramnios is defined as a maximal vertical pocket of >8 cm. Timely detection of TTTS is needed before severe complications occur, such as preterm fetal hydrops, prelabour rupture of membranes, preterm delivery or intrauterine fetal death [4].

Quintero staging system is a useful tool for describing the severity of TTTS. Stage I includes oligohydramnios in one sac and polyhydramnios in the other sac. In stage II, fetal bladder in the donor twin is not visualised which is suggestive of anuria. Abnormal doppler wave forms are seen in stage III. Stage IV includes fetal hydrops. Intrauterine death of a single fetus occurs in stage V [5].

In this case, the donor twin was in Quintero stage II and the recipient twin was in Quintero stage III as there was oligohydramnios and collapsed urinary bladder in the donor twin and polyhydramnios with abnormal doppler findings and cardiac changes were detected in the recipient twin.

In about half the cases, the heart is enlarged due to an increased myocardial thickness [6]. Tricuspid regurgitation occurs in about 30%-50% of recipients but is severe in only half of these. Mitral regurgitation on the other hand is much less frequent (6%-14% of cases) yet usually severe. In this case, the recipient twin shows ventricular hypertrophy with associated tricuspid and mitral valve regurgitation. Once a TTTS occurs, the findings tend to progress over time. There occurs worsening of ventricular hypertrophy and systolic dysfunction, which ultimately lead to fetal hydrops and intrauterine fetal demise [7]. Options for treatment of TTTS include Serial Amnioreduction (SA) fetoscopic laser coagulation of communicating vessels, septostomy, and termination [8]. Fetoscopic laser coagulation of communicating vessels is the standard therapy of treatment [9]. The treatment differs and it depends on the gestational age and stage at the time of diagnosis. Many factors like the response to treatment, growth of fetus and progression of disease determines the timing of delivery. Still, the median gestational age at delivery for the majority of the studies which used laser treatment was 33-34 weeks, particularly in severe cases [10]. In a meta-analysis and the Euro fetus study, 432 pregnancies were treated by laser treatment in comparison with SA done in 179 pregnancies. The meta-analysis of overall survival rate established that the proportion of survivor fetuses treated with

Laser Treatment was twice as great as fetuses treated with SA (66% and 47%, respectively) [11]. Neonatal deaths were in those treated by laser and 14-39% in those treated by amnioreduction [12]. Untreated TTTS generally carries a poor prognosis, with up to 90% perinatal mortality [13].

CONCLUSION(S)

TTTS is a rare and serious complication. It is important to diagnose this syndrome early and intervene in time so that severe complications including perinatal morbidity and mortality associated with this syndrome can be avoided.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: May 19, 2020
- Manual Googling: Jun 14, 2020
- iThenticate Software: Jul 13, 2020 (20%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **May 18, 2020**
Date of Peer Review: **May 29, 2020**
Date of Acceptance: **Jun 15, 2020**
Date of Publishing: **Oct 01, 2020**