

Stage V Twin-To-Twin Transfusion Syndrome With Hydrops Fetalis And Intrauterine Demise: A Case Report

Dr Anila Reddy Chilukuri^{1*}, Dr Kamal Patil², mrityunjay chandrashekar metgud³, Mahadevi savanur⁴, Anusha malapure⁵, vinutha hanji⁶

^{1*}PG JR-3, MS Obstetrics and Gynecology, Jawaharlal nehru medical college, belagavi. email id - dr.anila3@gmail.com

²Professor, MD Obstetrics and Gynecology, Jawaharlal nehru medical college, belagavi. Email Id - kamalpatil1967@yahoo.co.in

³Professor, MD Obstetrics and Gynecology, Jawaharlal nehru medical college, belagavi. Email id - metm67@gmail.com

⁴Associate Professor, MS Obstetrics and gynecology, Jawaharlal nehru medical college, belagavi. Email id - drdevim30@gmail.com

⁵Assistant Professor, MS Obstetrics and gynecology, Jawaharlal nehru medical college, belagavi. Email id - anushamalapure@gmail.com

⁶Assistant Professor, MS Obstetrics and gynecology, Jawaharlal nehru medical college, belagavi. Email id - vinuhanji@gmail.com

ABSTRACT

Background: Twin-to-twin transfusion syndrome (TTTS) is a severe complication of monochorionic diamniotic (MCDA) twin pregnancies caused by unbalanced placental vascular anastomoses. Advanced stages are associated with high perinatal mortality, particularly when diagnosis is delayed.

Case Presentation: We report a case of Stage V TTTS in a 32-year-old G3P2L2 woman at 28 weeks and 3 days period of gestation. Ultrasonography revealed classical features of TTTS, including oligohydramnios in the donor twin (twin A) and marked polyhydramnios with hydrops fetalis in the recipient twin (twin B). The recipient twin had intrauterine fetal demise at presentation. The donor twin showed relatively preserved growth but evidence of compromised intrauterine conditions. Due to the advanced stage and poor prognosis, the patient underwent induction of labor. The donor twin was delivered alive with low birth weight and required admission to neonatal intensive care unit (NICU), while the recipient twin was delivered as a macerated stillbirth with features of hydrops fetalis.

Conclusion: This case highlights the rapid progression and poor prognosis of advanced TTTS, especially with late diagnosis. Early identification of chorionicity, regular antenatal surveillance, and timely referral for fetal therapy are crucial to improve outcomes and prevent such adverse complications.

Keywords: Twin-to-twin transfusion syndrome; Monochorionic diamniotic twins; Hydrops fetalis; Intrauterine fetal demise; Quintero staging; Polyhydramnios-oligohydramnios sequence; Fetoscopic laser photocoagulation; High-risk pregnancy

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INTRODUCTION

Twin-to-twin transfusion syndrome (TTTS) is a serious and potentially life-threatening complication unique to monochorionic twin pregnancies, arising due to unbalanced placental vascular anastomoses. It occurs in approximately 10–15% of monochorionic diamniotic (MCDA) twin gestations and contributes significantly to perinatal morbidity and mortality (2). The overall prevalence is estimated to be 1–3 per 10,000 births, with TTTS accounting for a considerable proportion of fetal loss and adverse neonatal outcomes in twin pregnancies (4).

The pathophysiology of TTTS involves abnormal intertwin blood flow through shared placental vessels, most commonly arteriovenous anastomoses. This results in a net transfer of blood from one twin (the donor) to the other (the recipient). The donor twin becomes hypovolemic, leading to reduced renal perfusion, oliguria, and oligohydramnios (2,12). In contrast, the recipient twin develops hypervolemia, which results in

polyuria, polyhydramnios, and progressive cardiovascular overload. Over time, this imbalance can lead to severe complications such as cardiac dysfunction, hydrops fetalis, and even intrauterine fetal demise, particularly in advanced stages of the disease (4). TTTS is typically diagnosed antenatally using ultrasonography. The hallmark diagnostic feature is the presence of discordant amniotic fluid volumes, commonly referred to as the polyhydramnios-oligohydramnios sequence (4,5). Additional sonographic findings may include fetal growth discordance, abnormal Doppler studies, absence of the donor twin's bladder, and features of hydrops in the recipient twin. The severity of TTTS is classified using the Quintero staging system, which ranges from Stage I (mild disease with preserved bladder visibility in the donor twin) to Stage V, which represents intrauterine demise of one or both fetuses (2,5).

The clinical course of TTTS is often unpredictable and can progress rapidly, particularly during the second

*Author for Correspondence: dr.anila3@gmail.com

trimester (2). Advanced stages of the disease, especially Stages III to V, are associated with poor prognosis, with reported perinatal mortality rates as high as 70–100% when untreated or diagnosed late. Even in cases where one twin survives, there is a significant risk of long-term neurological impairment due to intrauterine hypoxic injury or hemodynamic instability (6).

Management of TTTS depends on the stage at diagnosis, gestational age, and available expertise. Treatment options include expectant management, serial amnioreduction, fetoscopic laser photocoagulation of placental anastomoses, and selective fetal reduction in severe cases (7,8). Among these, fetoscopic laser photocoagulation is considered the gold standard therapy, as it directly targets the underlying placental vascular connections. However, access to such advanced interventions may be limited in many settings, and late presentation continues to pose a major challenge in improving outcomes.

Early identification of chorionicity and regular ultrasound surveillance are essential components of antenatal care in twin pregnancies. Current guidelines recommend close monitoring of MCDA pregnancies with serial ultrasound examinations to enable early detection of complications such as TTTS (5,10). Timely diagnosis and referral to specialized fetal medicine centers can significantly improve perinatal survival and reduce morbidity (7,11).

In this report, we present a rare case of Stage V TTTS in a monochorionic diamniotic twin pregnancy complicated by hydrops fetalis and intrauterine demise of the recipient twin. This case highlights the rapid progression of advanced TTTS and underscores the critical importance of early diagnosis, vigilant monitoring, and timely intervention in improving maternal and fetal outcomes.

CASE PRESENTATION

Patient Information and Obstetric History: A 32-year-old woman, G3P2L2, presented to our tertiary care center at 28 weeks and 3 days period of gestation with a known twin pregnancy. The pregnancy had been previously identified as monochorionic diamniotic (MCDA) on second-trimester ultrasonography. Her previous obstetric history was uneventful, with two prior full-term normal deliveries and no history of complications.

The patient reported progressive abdominal distension over the preceding few weeks, which appeared disproportionate to gestational age. There was no history

suggestive of vaginal bleeding, leaking per vaginam, fever, or decreased fetal movements. She had no known comorbidities such as hypertension, diabetes mellitus, or thyroid disorders, and her antenatal laboratory investigations had been within normal limits.

Clinical Examination: On admission, General physical examination was unremarkable. Abdominal examination showed a markedly distended uterus, with fundal height corresponding to approximately 36 weeks of gestation, significantly exceeding the period of amenorrhea. The uterus appeared tense on palpation, and fetal parts were difficult to delineate, raising suspicion of polyhydramnios. Auscultation of fetal heart sounds was inconclusive for both twins, prompting urgent imaging evaluation.

Previous Antenatal Evaluation: A prior ultrasound scan performed at 18 weeks of gestation had confirmed a MCDA twin pregnancy with a single posterior placenta. At that time, both fetuses were appropriate for gestational age, with no significant discordance in fetal weight, amniotic fluid volume, or structural anomalies. Chorionicity had been established, but there was no evidence of complications during earlier antenatal visits.

Diagnostic Assessment: A detailed obstetric ultrasonography was performed at presentation. The findings were consistent with advanced twin-to-twin transfusion syndrome (TTTS), corresponding to Quintero Stage V.

Twin A, identified as the donor twin, demonstrated oligohydramnios, with a single deepest pocket (SDP) measuring 1.69 cm (Figure 1). The fetus showed relatively preserved growth parameters for gestational age. The urinary bladder was visualized, suggesting partial preservation of renal perfusion. No gross congenital anomalies were detected.

Twin B, identified as the recipient twin, exhibited classical features of severe TTTS. There was marked polyhydramnios with a single deepest pocket (SDP) measuring 14.98 cm (Figure 2). The fetus showed diffuse subcutaneous edema involving the scalp and body, consistent with hydrops fetalis. Additionally, the stomach bubble was not visualized, suggesting compromised gastrointestinal perfusion or function. The most significant finding was intrauterine fetal demise of the recipient twin.

The placenta was noted to be single and posteriorly located, confirming monochorionicity, with evidence of a fused placental mass. These findings strongly supported the diagnosis of severe, advanced TTTS with rapid progression.

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Fig 1: Obstetric ultrasound image at 28 weeks gestation demonstrating oligohydramnios in the donor twin, with a reduced single deepest pocket(SDP) measuring approximately 1.69 cm, consistent with twin-to-twin transfusion syndrome.



Fig 2: Ultrasound image demonstrating severe polyhydramnios in the recipient twin (single deepest pocket SDP ~14.98 cm) in advanced Twin-to-Twin Transfusion Syndrome (TTTS)

Diagnosis: Based on the ultrasonographic findings and clinical presentation, a diagnosis of Quintero Stage V twin-to-twin transfusion syndrome was established, characterized by oligohydramnios in the donor twin, polyhydramnios and hydrops fetalis in the recipient twin, and intrauterine fetal demise.

Therapeutic Intervention: Given the advanced stage of disease and intrauterine demise of one twin, immediate management was indicated to prevent maternal complications and optimize the outcome of the surviving fetus. The patient and her family were counseled extensively regarding the prognosis, risks, and available management options.

A decision was made to proceed with induction of labor. The management plan was aimed at safe delivery while minimizing maternal morbidity and providing supportive neonatal care for the surviving twin.

Outcome and Neonatal Course: The patient underwent induction of labor and delivered vaginally. Twin A, the donor twin, was delivered with a birth weight of 960 grams. The neonate had Apgar scores of 6 at 1 minute and 7 at 10 minutes, indicating moderate perinatal compromise. Due to respiratory distress and prematurity, the baby was immediately shifted to the neonatal intensive care unit (NICU) for further management and supportive care.

Twin B, the recipient twin, weighed 1300 grams and was delivered as a macerated stillbirth. The twin B fetus showed gross features of hydrops fetalis, including generalized edema, confirming severe intrauterine compromise prior to delivery.

The surviving neonate required intensive monitoring for complications related to prematurity, low birth weight, and hemodynamic instability associated with TTTS. Further follow-up was planned to assess growth parameters, organ function, and long-term neurodevelopmental outcomes.

DISCUSSION

Twin-to-twin transfusion syndrome (TTTS) remains one of the most severe complications of monochorionic twin pregnancies, with a highly variable yet often aggressive clinical course. The present case of Stage V TTTS with hydrops fetalis and intrauterine demise highlights the adverse end of the disease spectrum, consistent with observations in existing literature.

The pathophysiological basis of TTTS involves chronic net blood transfusion from the donor to the recipient twin, primarily through arteriovenous anastomoses (Merriweather et al., 2021). Lack of compensating A-A (artery to artery anastomosis) may contribute to severe intertwin transfusion imbalance (12). This results in hypovolemia and oligohydramnios in the donor twin and hypervolemia with polyhydramnios in the recipient twin (2,12). In our case, the donor twin exhibited oligohydramnios with a measurable single deepest pocket (SDP) of 1.69 cm, while the recipient twin showed severe polyhydramnios and hydrops fetalis. Similar cardiovascular complications in the recipient twin, including fluid overload and cardiac dysfunction, have been well documented by Rotar et al. (2020), who described progressive cardiac compromise as a major determinant of poor outcomes (3).

The diagnosis of TTTS is primarily ultrasound-based, with the polyhydramnios– oligohydramnios sequence serving as a hallmark feature (Wagner et al., 2013)(4). Additional findings such as absent or reduced bladder visibility in the donor twin and Doppler abnormalities further aid in staging (5). In this case, ultrasonography confirmed advanced disease with hydrops and fetal demise, consistent with Quintero Stage V classification. The Quintero staging system remains the standard for assessing severity and guiding management (Ponce et al., 2024) (5).

Advanced stages of TTTS, particularly Stages III to V, are associated with significantly higher perinatal

mortality. Our case reflects this, with intrauterine demise of the recipient twin and compromised survival of the donor twin. Forrester et al. (2013) reported that even when one twin survives, there is a considerable risk of neurological impairment, especially following co-twin demise due to shared placental circulation and acute hemodynamic shifts (6). This underscores the importance of long-term follow-up in surviving neonates.

Management strategies for TTTS vary depending on the stage and gestational age at diagnosis. Emerging evidence supports expectant management with close ultrasound surveillance in asymptomatic patients with Stage I TTTS, while Stage II TTTS diagnosed at very early gestational ages may benefit from careful monitoring until 16–17 weeks of gestation before definitive intervention. In contrast, fetoscopic laser photocoagulation should not be delayed in advanced disease such as Stage III and IV TTTS (7). Fetoscopic laser photocoagulation is considered the gold standard, as it directly ablates the placental anastomoses responsible for the disease (Sago et al., 2018) (8). Bouchghoul et al. (2025) further highlighted that early intervention significantly improves survival rates and reduces morbidity (7). However, in our case, late presentation at 28 weeks with Stage V disease limited the available therapeutic options, and management was primarily directed toward optimizing the outcome of the surviving donor twin.

Early diagnosis and surveillance are critical in improving outcomes. Townsend and Khalil (2018) recommended serial ultrasound monitoring in monochorionic pregnancies for early detection of TTTS (10). Similarly, Hartanto et al. (2025) emphasized the challenges in early diagnosis, particularly in resource-limited settings, which may delay intervention (9). Wilpers et al. (2024) also noted that social and healthcare access factors can influence the timing of diagnosis, contributing to adverse outcomes (11). Delayed presentation at an advanced stage significantly limited the scope for definitive fetal therapeutic interventions and contributed to poor perinatal outcome. This case exemplifies the severe and rapidly progressive nature of TTTS when diagnosed late. It reinforces the importance of early determination of chorionicity, regular antenatal surveillance, and timely referral to specialized fetal medicine centers. Prompt diagnosis and intervention remain the cornerstone in reducing perinatal mortality and improving neonatal outcomes in TTTS.

CONCLUSION

Twin-to-twin transfusion syndrome is a serious and potentially fatal complication of monochorionic diamniotic twin pregnancies, particularly when diagnosed at an advanced stage. This case of Stage V TTTS with hydrops fetalis and intrauterine demise underscores the rapidly progressive nature of the condition. Late presentation significantly limits therapeutic options and is associated with poor perinatal outcomes, as reflected in the loss of one twin and the

compromised status of the surviving neonate. The findings highlight the critical importance of early determination of chorionicity, regular antenatal surveillance, and timely diagnosis using ultrasonography. Early referral to specialized fetal medicine centers and access to definitive interventions such as fetoscopic laser therapy are essential to improve survival and reduce morbidity. Increased awareness, improved prenatal care, and structured monitoring protocols are vital in preventing such adverse outcomes in twin pregnancies.

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