

Leucomalacia In A Twin Complicating Twin-To-Twin Transfusion Syndrome TTTS With IUFD Of The Second Twin: A Case Report And Literature Review

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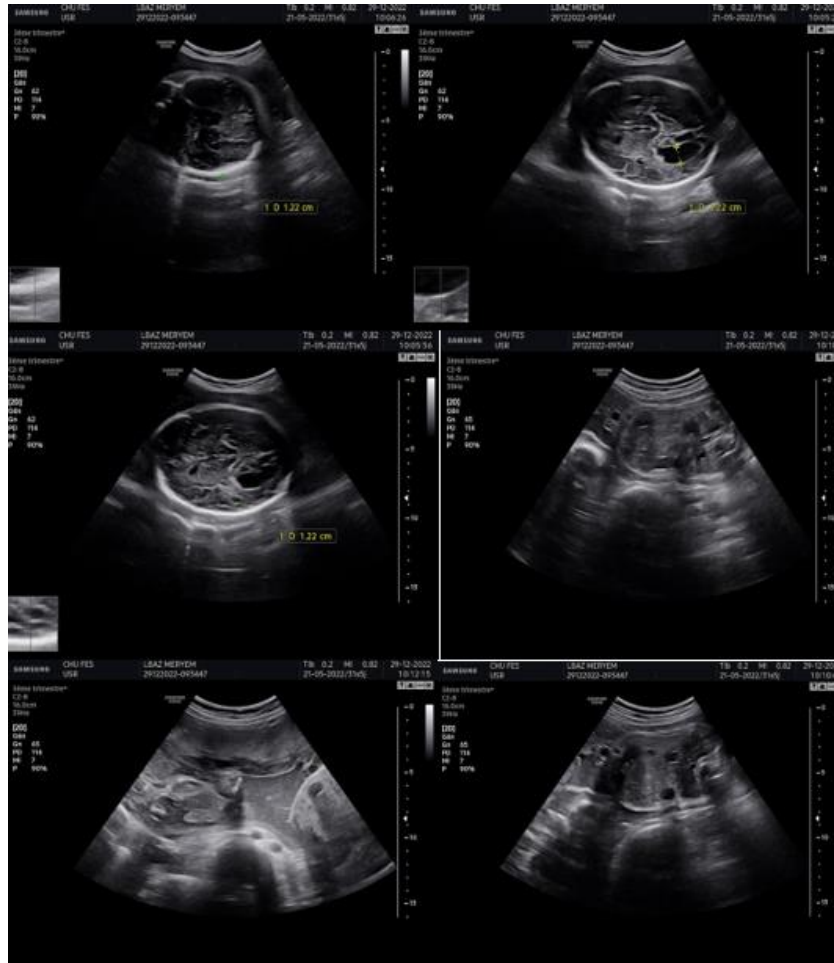
I. Introduction:

- Twin-to-Twin Transfusion Syndrome (TTTS) is a rare but serious complication of monochorionic twin pregnancies, characterized by an imbalanced blood flow between twins due to placental vascular anastomoses. This condition can lead to significant morbidity and mortality for both fetuses.
- Periventricular leukomalacia (PVL) is a severe ischemic brain injury that primarily affects preterm neonates but can also complicate certain pathological twin pregnancies. Twin-to-Twin Transfusion Syndrome (TTTS), a complication unique to monochorionic pregnancies, is characterized by a hemodynamic imbalance between the two fetuses due to placental vascular anastomoses.
- In this context, the in utero demise of one twin (IUFD) further exacerbates complications for the surviving twin. The resulting ischemia, inflammatory processes, and metabolic disturbances significantly increase the risk of brain injuries, particularly periventricular leukomalacia.
- Here, we report a rare case of leukomalacia in a surviving twin, complicating TTTS with IUFD of the co-twin. Through this case report and a review of the literature, we aim to highlight the pathophysiological mechanisms, diagnostic challenges, and therapeutic considerations of this complex situation.

II. Case Presentation

- A 30-year-old patient, G3P2 (2 live births via vaginal delivery), with no notable pathological history, presented to our facility for follow-up of a monochorionic diamniotic twin pregnancy estimated at 33 weeks of gestation.
- The clinical examination revealed a patient who was conscious and hemodynamically and respiratory stable, with normocolored conjunctiva, a normal heart rate, normal blood pressure, and afebrile. Obstetric examination found a longitudinally distended uterus, the perception of two fetal poles, and a uterine height consistent with the gestational age.
- The obstetric ultrasound concluded a twin pregnancy with a single Grade II placenta (Granum), showing a vacuolated appearance on one side.
- Twin 1 (J1): Positive cardiac activity, cephalic presentation, estimated fetal weight of 1441 g, normal amniotic fluid quantity, with mild ventriculomegaly measuring 12 mm. There were multiple anechoic images in the cerebral parenchyma suggestive of porencephaly secondary to leukomalacia, along with several cerebral and intestinal calcifications. Umbilical Doppler was normal (0.51), while the ductus venosus showed a negative A-wave.





- The patient delivered prematurely at 35 weeks of gestation via vaginal delivery:
 1. First twin: Alive, Apgar score 10/10, birth weight: 1544 g.
 2. Second twin: Intrauterine fetal demise (IUFD), estimated fetal weight: 300 g.
- Two hours after delivery, the first twin developed respiratory distress associated with cyanosis, which unfortunately resulted in the neonate's death.

III. Discussion

- TTTS is a complex condition, occurring in 15% of monochorionic twin pregnancies, equivalent to 1 case per 2000 pregnancies [8]
- The excess mortality and morbidity in monochorionic pregnancies are primarily due to vascular anastomoses on the chorionic plate, which connect the circulations of the two fetuses. The death of one monochorionic fetus leads to the exsanguination of its co-twin, resulting in death in 20–30% of cases. When the co-twin survives, ischemic complications, particularly cerebral, occur in a similar proportion. The most acute complication of these pregnancies is twin-to-twin transfusion syndrome (TTTS), characterized by polyuric hydramnios in the recipient twin and oliguric oligohydramnios in the donor twin.
- TTTS is associated with a perinatal mortality rate of 90% and severe neurological sequelae in survivors

T1	T2
Crown-to-rump length discordance > 6 mm	Discordance persistante de taille des vessies
Amniotic fluid discordance	Membrane not visible or against a fetus
Nuchal translucency of 1 embryo Discordance of the two nuchal thicknesses greater than 20%	Sign of the membrane folding
	Hydramnios and oligohydramnios

Tableau I. Signes échographiques à rechercher en faveur du. STT

Stade I	Bladder visible in the donor
Stade II	Bladder not visible in the donor
Stade III	Abnormality in at least one Doppler (absent or reversed diastolic flow in the umbilical artery of the donor and/or absent or reversed flow in the ductus venosus of the recipient)
Stade IV	Hydrops in the recipient
Stade V	Fetal death.

Table II. Classification for Assessing TTTS Severity [6].

- **A Second-Trimester Sign to Identify: The "Membrane Folding" Sign** (Table I). A second-trimester sign that should be better recognized is the "folding sign" of the membrane (Figure 1) [7]. When there is an increase in amniotic fluid in one sac and a decrease in the other, the membrane folds. This sign occurs in about 14% of twin pregnancies evaluated in the second trimester. When this sign is present, the risk of TTTS development is close to 50% [6]. In such cases, it is crucial to reassess the patient within 8 days.
- **Case Study:** In our case, TTTS was diagnosed at stage V. TTTS is associated with a perinatal mortality rate of 90%, with severe neurological sequelae observed in 20–40% of survivors, who are typically born at an average gestational age of 25 weeks.
- **Delivery Timing and Recommendations:** According to CNGOF recommendations, it is reasonable to consider delivery between 34 and 37 weeks of gestation [9]. There is ongoing debate regarding whether delivery should occur closer to 37 weeks or remain near 34 weeks in these cases. The mode of delivery depends on obstetric conditions, with cesarean section often performed.
- Iterative amniotic fluid drainage was historically the only prenatal therapy for TTTS, allowing the survival of at least one twin in approximately 50% of cases, with delivery occurring around 28 weeks and a risk of sequelae in about 20% of survivors.
- Surgical fetoscopic treatment of TTTS, specifically laser coagulation of vascular anastomoses on the chorionic plate, has significantly improved outcomes. This procedure ensures the survival of at least one twin in approximately 80% of cases, with an average gestational age at delivery of 33 weeks and a risk of sequelae in survivors reduced to less than 10%, primarily neurological.
- **Twin Anemia-Polycythemia Sequence** TAPS must be recognized by obstetricians. This condition, characterized by anemia in one fetus and polycythemia in the other, occurs in 10–15% of cases after laser treatment and in 5% of uncomplicated monochorionic pregnancies. Its diagnosis, facilitated by systematic measurement of the middle cerebral artery velocity in both fetuses, requires specific management.

IV. Conclusion

- There is a clear benefit to the early diagnosis of TTTS, which can be facilitated by biweekly ultrasound scans, as recommended for monochorionic twin pregnancies.
- TTTS is a severe pathology that has been the subject of significant research in recent years, leading to a better definition of its management.

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