

Reminder of important clinical lesson

Lessons from monochorionic twin delivery

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Summary

The presence of acute peripartum anaemia in a monochorionic twin pregnancy represents a clinical challenge requiring prompt recognition and management. Twin-to-twin transfusion syndrome (TTTS) is a major complication of these pregnancies and a medical emergency in its acute form. Acute intrapartum fetoplacental transfusion (AIFT) has been reported infrequently. The authors present a case of a probable acute TTTS in an uneventful monochorionic monoamniotic twin pregnancy, where typical ultrasound criteria for long-standing TTTS were absent. The first twin was born pale, hypotonic and developed hypovolemic shock due to acute anaemia. Soon after birth, she presented with seizures and a cerebral ultrasound detected a large parieto-occipital infarction. The second twin, although plethoric, was clinically well. The risk of acute TTTS and AIFT, although infrequent and unpredictable, should be kept in mind when planning delivery of monochorionic twins, because the consequences for one or both twins can be disastrous.

BACKGROUND

The twin-to-twin transfusion syndrome (TTTS) is a severe complication of monochorionic twin pregnancies. It results of an abnormal sharing of placental blood, through vascular anastomosis, which are present in most monochorionic placentae.¹ Although intertwin transfusion can occur through these anastomosis, TTTS only arises if a circulatory imbalance is present, which can result in hypovolemia, hypotension, growth-restriction and oligo-anuria in the donor, and hypervolemia, hypertension, polyuria and ultimately heart failure in the recipient. Therefore, the classic ultrasound signs of TTTS include progressive growth discordance, oligohydramnios or anhydramnios and a small or absent bladder filling in the donor, in contrast to polyhydramnios and distended bladder in the recipient.¹ Its incidence varies between 5% and 15% and accounts for approximately 12% of perinatal deaths.²⁻³ If severe or left untreated, the mortality can reach up to 90%.⁴

It usually begins in the second trimester of pregnancy, but can occur at any stage and rarely peripartum, particularly after clamping of the umbilical cords.¹ In acute TTTS, the typical ultrasound criteria for long-standing TTTS are absent. Acute TTTS is a medical emergency.²

Fetoplacental transfusion is the entry of fetal blood into maternal circulation and has a wide spectrum of clinical variation.⁴ It can present as life-threatening anaemia with devastating consequences for the fetus or newborn, such as neurologic injury, stillbirth or neonatal death. When it occurs during delivery it is designated as acute intrapartum fetoplacental transfusion (AIFT).⁴ In monochorionic monoamniotic twin pregnancies it has been described in the second twin, after delivery of the first one. Clinically relevant AIFT occurs rarely in monochorionic twin pregnancies.²⁻⁴ A probable explanation is that the time between twin deliveries is usually short enough to prevent

development of severe hypovolemia, and so, many fetoplacental transfusions may go unrecognised.⁴

CASE PRESENTATION

A 32-year-old primigravida with an uneventful monoamniotic, monochorionic twin pregnancy of 31 weeks, presented to our Hospital due to premature rupture of membranes with discharge of slightly bloody amniotic fluid. Serial ultrasounds during pregnancy showed appropriate growth of both twins, normal amniotic fluid volume, bladders of comparable size and normal nuchal translucency. After 2 h, a caesarean section was performed. The first female-twin was born pale and hypotonic but recovered well without resuscitation. Her Apgar score was 8 and 9 at 1 and 5 min, respectively. The second female-twin was born well, with a plethoric appearance. Birthweights were similar and adequate to gestational age, 2180 g for the first twin and 2185 g for the second. Both were admitted to neonatal intensive care unit, in spontaneous breathing with good oxygen saturations.

Soon after birth the first twin developed respiratory distress: tachypnoea of 115 breaths per minute, expiratory grunt, intercostal retractions and oxygen needs (FiO₂ 0.25). She was pale, with poor peripheral perfusion, heart rate of 142 per minute and hypotensive blood pressure of 32/13 mm Hg, mean of 23 mm Hg. Her haematocrit was 23.4%, haemoglobin concentration 7.7 g/dl, leucocyte and platelet count were normal and C reactive protein was negative. Her diuresis and renal function were normal. She was started on ampicilin and gentamicin. The second twin was clinically and haemodynamically stable with a venous haematocrit of 58% and haemoglobin concentration 19 g/dl.

After infusion of 10 ml/kg of normal saline and transfusion of 20 ml/kg of packed red blood cells, the first twin

recovered haemodynamic stability and, by the 5th hour, her haematocrit had risen to 35% and the haemoglobin concentration to 11.4 g/dl. By the 6th hour she experienced frequent tonic seizures, with dancing movements of the eyes and myoclonic jerks that were controlled with phenobarbital. She had no electrolyte imbalance or hypoglycaemia. Because of multiple apnoeas needing bag-valve-mask ventilation, she was intubated and required assisted ventilation for 48 h. Serial cerebral ultrasound revealed a large parieto-occipital infarction. The brain MRI (10th day) showed multiple haemorrhagic foci, cavities, vasogenic and cytotoxic oedema and thromboembolic events suggestive of ischaemia-reperfusion injury. Both mother and newborn extended coagulation panels were normal. Blood culture was negative. An abnormal neurological examination persisted throughout the entire admission.

The second twin was well, without signs of cardiac failure. She was jaundiced from day 3, without criteria for phototherapy. Her cerebral ultrasound was normal.

At day 18, both twins were breastfed and exhibited good weight gain, and were discharged home with scheduled follow-up.

The placenta's pathological examination revealed insertion of the umbilical cords close to each other with severe subchorial congestion without thrombosis. Flow cytometry to identify fetal haemoglobin in maternal blood for detection of fetomaternal haemorrhage was not performed.

At present, (18 months) both twins have a normal growth, but while the second twin is a healthy baby, the first twin has spastic cerebral palsy.

DISCUSSION

The presence of acute peripartum anaemia in one twin and plethora in the other, led to the exclusion of infection and, on the other hand, evoked a possible acute TTTS.

In our case, typical ultrasound criteria for long-standing TTTS were absent. There was no birthweight discrepancy. The first twin was born pale and hypotonic and soon after birth presented with hypovolemic shock due to severe anaemia, possibly secondary to acute transfusion to the recipient twin or the placenta. A possible mechanism is the existence of bidirectional flow in 'superficial' arterioarterial or venovenous anastomosis, instead of unidirectional flow in 'deep' arteriovenous anastomosis.⁵

Intertwin haemoglobin difference was superior to 10 g/dl (TTTS is usually considered when >5 g/dl) nevertheless haemoglobin and haematocrit values of the second twin were borderline normal, in the superior range.²

Moreover, the second twin was always clinically stable in contrast to the severe presentation of the first twin. So, we can argue that although acute TTTS is the most probable explanation, the occurrence of AIFT during delivery cannot be excluded with certainty.

In the literature, AIFT is described after successful delivery of first twins, with rapid clinical deterioration of the second twins. One hypothesis is that substantial amounts of blood are transfused through anastomosing vessels, due to decreased resistance in the first twin's part of the placental vasculature after umbilical cord clamping.⁴ In our case,

the donor twin was the first to be delivered. The physiological mechanism for an overwhelming intrapartum transfusion is still unclear, but is known to occur without any recognised eliciting factor.⁴

A positive maternal Kleihauer screen or flow cytometry would support the diagnosis of AIFT, but neither was performed as this hypothesis was not immediately evoked.⁴

Maternal perception of decreased fetal movement is the most common antenatal presentation of fetomaternal haemorrhage, and was not present in our case, which gives further support to an acute peripartum event.⁴

A less likely diagnosis that could also explain acute anaemia in one twin and premature rupture of membranes is an intra-amniotic haemorrhage, but the amniotic fluid was only slightly bloody.

Rapid initiation of treatment is essential for an optimal neurodevelopment outcome. In our case, despite the uncertainties of the diagnosis, haemodynamic and haematological disturbances were promptly recognised and managed. Unfortunately, the first twin sustained important neurological sequelae.

In conclusion, although infrequent and unpredictable, the risk of acute TTTS and AIFT should be kept in mind when planning delivery of monochorionic twins, because consequences for one or both twins can be disastrous.

Learning points

- ▶ Acute peripartum anaemia in a monochorionic twin pregnancy represents a clinical challenge requiring prompt recognition and management.
- ▶ A cause to consider is acute twin-to-twin transfusion syndrome, a rare medical emergency associated with important morbidity and mortality.
- ▶ Another entity to have in mind is AIFT. Clinically relevant AIFT occurs rarely in monochorionic twin pregnancies, but can present as life-threatening anaemia with devastating consequences for the fetus or newborn.
- ▶ Regardless of final diagnosis, timely correction of haemodynamic and haematological disturbances is essential for survival and for an optimal neurodevelopment outcome.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **El Kateb A**, Ville Y. Update on twin-to-twin transfusion syndrome. *Best Pract Res Clin Obstet Gynaecol* 2008;**22**:63–75.
2. **Seng YC**, Rajadurai VS. Twin-twin transfusion syndrome: a five year review. *Arch Dis Child Fetal Neonatal Ed* 2000;**83**:F168–70.
3. **Clouqueur E**, Boukerrou M, Nayama M, et al. [Acute per-partum fetofetal transfusion. A case study on two sets of twins]. *J Gynecol Obstet Biol Reprod (Paris)* 2006;**35**:283–7.
4. **Uotila J**, Tammela O. Acute intrapartum fetoplacental transfusion in monochorionic twin pregnancy. *Obstet Gynecol* 1999;**94**(5 Pt 2):819–21.
5. **Bermúdez C**, Becerra CH, Bornick PW, et al. Placental types and twin-twin transfusion syndrome. *Am J Obstet Gynecol* 2002;**187**:489–94.

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