Case Report/Caso Clínico

Acute per-caesarean twin-to-twin transfusion syndrome or twin anemia-polycythemia sequence?

Sindrome de transfusão feto-fetal agudo per-cesariana ou sequência anemia-policitémia?

Rui Artur Coimbra*, Ana Patricia Domingues**, Clara Morais***, Etelvina Fonseca***, Paulo Moura****

Obstetrics Department, Coimbra University Hospitals

ABSTRACT

Monochorionic twin pregnancies present a significantly high rate of fetal mortality and morbidity, in part due to the twin-to-twin transfusion syndrome (TTTS) and its consequences. TTTS typically occurs in the second trimester and generally develops chronically. The risk of TTTS is unpredictable and despite its classical chronic development, it can also occur in a sub-acute or even acute manner. In the literature, few cases describe an acute or sub-acute peri-partum TTTS. The authors present one case of acute per-caesarean twin-to-twin transfusion syndrome that could also be classified as a twin anemia-polycythemia sequence (TAPS), a type of chronic twin-totwin transfusion associated with chronic anemia in the donor and polycythemia in the recipient, without twin oligo-polyhydramnios sequence (TOPS).

Keywords: Keywords: monochorionic twin pregnancy, twin-to-twin transfusion syndrome, acute per-partum syndrome, twin anemia-polycythemia sequence.

INTRODUCTION

The incidence of spontaneous multiple pregnancies is about 1%, being twin pregnancies in most of the cases; one third of these will have a monochorionic placentation. In general, the mortality of twins is 5-10 times greater than single pregnancies. Monochorionic twins are considered a high risk pregnancy due the higher incidence of fetal and perinatal morbidity and mortality. The rate of fetal loss in dichorionic twins is about 2% and in monochorionic it is about 10%. That high mortality is related to TTTS, higher prematurity and congenital malformations.

TTTS is the result of vascular connections between placentas that allows the connections of both fetoplacental cir-
culations (this connections are found in 98% of monocho-
rionic-diamniotic placentas). Therefore, unidirectional or
unbalanced blood flow is necessary to explain the changes
noted in TTTS. These anastomoses may be arterial to ve-
nous (AV), arterial to arterial (AA), and venous to venous
(VV). In fact, the majority of monochorionic-diamniotic
placentas have AV connections and its presence does not
necessarily results in TTTS. The presence of AA anasto-
moses was associated with a lower likelihood of TTTS.
Pathological studies have demonstrated that AV anasto-
moses are deep, but the vessels that feed them are invariably
superficial. In about 15-30% of the monochorionic twins,
the flow imbalance in AV communications of one fetus, the
donor, to the other, the receptor, will result in the TTTS.
The clinical case presented, at time of occurrence, was clas-
sified as a TTTS intra-partum, a very rare and unpredicta-
ble situation, even with a regular surveillance to delivery.
With the recently described as twin anemia-polycythemia
sequence, this earlier diagnose is contested and the value of
accurate Doppler evaluations, namely the middle cerebral
artery peak systolic velocity (MCA-PSV), assume another
importance.

**CASE REPORT**

AMRG, 30 years, second time pregnant, with a previous
spontaneous abortion at 6 weeks of gestation, iatrogenic
hypothyroidism (thyroidectomy in 2008, for papilar carc-
noma), had a spontaneous monochorionic diamniotic twin
pregnancy diagnose at 12 weeks. The prenatal surveillance
included a regular ultrasound (every 2 to 3 weeks) with
normal fetal anomaly scan and echocardiography in both
fetus.

The patient was hospitalized at 31 weeks, for sur-
veillance and fetal pulmonary maturation protocol, as
some Doppler abnormalities were seen: Umbilical Artery
(UA) Doppler of first fetus (F1) revealed resistance (IR)
and pulsatility index (IP) >p90 with diastolic flow pre-
sent and without evidence of centralization – middle ce-
rebral artery (MCA) Doppler was normal with a peak sys-
tolic velocity within normal range. Anatomy, growth and
amniotic fluid were normal. She had hospital discharge 7
days latter with normalization of UA Doppler, but was re-
admitted at 34 weeks again with UA Doppler abnormality
of F1 and fetal growth of both fetus on the 5th percentile.
The scan performed at 35 weeks and 1 day revealed: an
estimated fetal weights (EFW) of F1 is 1900 grams(gr) and
F2 is 1650gr, amniotic fluids (AF) were normal and AU
and MCA Doppler’s evaluations were also normal. Two
days later, F1 UA doppler revealed IR>p90 with diasto-
ic flow present and peak systolic velocity of MCA nor-
mal for both fetus (MCA-PSV of F1=45.5cm/s(p10) and
F2=65.0cm/s(p80)). Due to the gestational age it was deci-
ded to perform an elective caesarean, under epidural anal-
ge sia, with easy extraction of both fetus (with a time break
of one minute). Two males were born, the first with 1760gr,
Apgar index (AI) of 9/10/10 and pH artery/vein=7.15/7.20
and the second with 1710gr, AI of 9/10/10 and pH artery/
vein=7.17/7.22. Macroscopically the placenta was mono-
chorionic diamniotic, which was histological confirmed,
but the vascular connections have not been evaluated.

Because one of the twins has born pale and the other
plethoric, the pediatric team has picked blood from both
umbilical cords.

**Twin 1** presented a whiteness cutaneous-mucosa and
a respiratory distress syndrome always without oxygen
need. It had also an asymptomatic anemia, with arterial
pressure and cardiac frequency normal: day 0 (D0) with
erthrocytes-2.40x10^6/microL, haemoglobin (Hg)-9.1g/
dl; hematocrit level (Hct) -27%, platelets - 302000 U/L
and in D3 with Hg-10.0g/dL; Hct-29.1%, 320000 platelets.
Other morbidities included ictericia/hyperbilirubinaemia
without criteria for phototherapy. This neonate got medical
discharge on D8.

**Twin 2** had a plethoric aspect with a polycythemia and
thrombocytopenia: D0 with erythrocytes-6.49x10^6/mi-
croL, Hg-25.3g/dL; Hct-71.4%, 121000 U/L platelets and
D3 with Hg-26.7 g/dL; Hct-61.4%, 68000 U/L platelets being
held sanguine exchange (blood by fisiologic serum) wi-
without intercurrences. On D4 Hg was 21.7g/dL, Hct-61.4%,
68000 U/L platelets and on D7 the Hg value was 23.2 g/dL;
Hct-67.9%, 111000 platelets. Ictericia/hyperbilirubinaemia
with criteria for phototherapy also occurred, from D1 to
D5. Clinical discharge took place on D8.

In Pediatric evaluation, neurodevelopment outcome at
6 and 12 months was normal for both children

**DISCUSSION**

The TTTS is defined by the presence of polihydramnios
(maximum vertical pocket of amniotic fluid of &gt;8cm) in
one fetus and oligohydramnios (maximum vertical pocket
of fluid of&lt;2 cm) in another. The staging system propo-
sed by Quintero defined 5 stages: stage I, with a oligo/
polihydramnios sequence, the bladder of the donor twin
was still visible, whereas in stage II, the bladder was not
visualized but the doppler studies were normal; by stage
III, there are doppler abnormalities; in stage IV, hydrops
is present; and in stage V, there is a demise of one or both twins. Untreated TTTS that develops earlier than 26 weeks has a perinatal mortality rate of 90%.10

It has been suggested that some features in first trimester scan will be able to predict TTTS in monochorionic twin pregnancies. These features included crown-rump-length (CRL) and nuchal translucency (NT) discrepancy (the prevalence of increased NT thickness in at least one of the fetuses that subsequently develop TTTS was described to be about 30%, compared to 10% of those that do not develop TTTS); and abnormal Doppler flow velocity waveform in the ductus venoso (DV).11

In this particular case reported and in all performed ultrasounds there were no suggestive signs of TTTS. Even the last one, performed in the same day of the delivery, also didn’t reveal any suspicion. The short time between the last ultrasound evaluation and the caesarean (one hour) excludes any chronic or sub-acute TTTS. The caesarean held without incidents and fetal extractions were simple and fast. Immediately, the newborns were delivered into the care of the Pediatric team, who suspected of TTTS.

A twin anemia-polycythemia sequence (TAPS) is a newly described form of chronic twin-to-twin transfusion associated with chronic anemia in the donor and polycythemia in the recipient, without twin oligo-polyhydramnios sequence (TOPS).12

In present case we admit the possibility of a spontaneous TAPS (although it is more often described after laser surgery). Both forms are characterized by the presence of a large inter-twin hemoglobin difference at birth without signs of TOPS as seen injuries the typical form of chronic TTTS. Whereas iatrogenic TAPS occurs in up to 13% of cases after laser therapy, the spontaneous form seems to complicate 5–6% of monochorionic twin pregnancies.

TAPS can be diagnosed both pre and postnatally. The prenatal diagnostic criteria is based on the inter-twin discordance in Doppler ultrasound measurement of middle cerebral artery peak systolic velocity (MCA-PSV). As described by Mari et al., MCA-PSV measurement >1.5MoM in one twin suggests severe anemia and a simultaneous decrease of the MCA-PSV in the co-twin suggests polycythemia. The postnatal diagnosis is based on three criteria: marked intertwin hemoglobin differences at birth, reticulocytosis in the donor and placental injection examination showing very small superficial AV anastomoses. The case reported occurred in 2009 and, although MCA was evaluated in all ultrasounds performed (regarding de IP, IR and S/D index), the PSV wasn’t described in all the reports, so it’s not possible to classify this case as a TAPS, remaining the doubt based on the post-natal observation.

The understanding of the pathogenesis of TAPS is still incomplete but it appears to be mediated by a few small AV anastomoses. Chronic inter-twin transfusion through these few minuscule AV anastomoses may occur so slowly that compensatory hemodynamic mechanisms maintain normal blood volume in both fetuses. The placental vascular pattern may also be dynamic as spontaneous thrombosis or infarction may occur as well as revascularization.

The prenatal monitoring of MCDA twins should imperatively include the measurement of the MCA-PSV of both fetuses during each follow-up visit, even in all uncomplicated pregnancies with absence of inter-twin discordance in amniotic fluid volumes. Although, the sensibility and the specificity of this prenatal marker (MCA-PSV) haven’t yet been demonstrated.

The special interest this clinical case lies on the need of MCA-PSV Doppler evaluation for a differential diagnostic between the rare form of acute TTTS (that may occur in any gestational age and in a sudden form) and the recently described TAPS.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

REFERENCES


