Radio-diagnosis.

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INTRODUCTION:
Twin-to-twin transfusion syndrome (TTTS) occurs in either monochorionicmonoamniotic or monochorionicdiamniotic twins and is due to arteriovenous anastomosis in the common placenta. Abnormal blood vessel connections form in the placenta and allow blood to flow unevenly between the babies. One twin – called the donor – becomes dehydrated; and the other – called the recipient – develops high blood pressure and produces too much urine and over fills the amniotic sac. The donor twin appears small and the recipient twin is large, and at risk for high-output cardiac failure. The mortality rate is 40-90%, with both twins at risk.

Ultrasongraphy remains the main modality for diagnosis of twin-to-twin transfusion syndrome (TTTS).

Ultrasound diagnosis:
There is imbalance in the net flow of blood across the placental vascular communications from one fetus, the donor, to the other, the recipient.

11-14 weeks: early TTTS is suspected if there is discordance in size of the amniotic fluid sacs, ≥20% discordance in fetal nuchal translucency (NT) thickness, or absent / reversed end diastolic flow (EDF) in the ductus venosus usually in the fetus with higher NT.

≥15 weeks: oligohydramnios (deepest vertical pool of ≤2 cm) in the sac of the oliguric or anuric donor fetus and polyhydramnios (≥26 cm at 15-17 weeks, ≥28 cm at 18-20 weeks and >10 cm at ≥20 weeks) in the sac of the polyuric recipient.

The condition is subdivided into 5 stages, by Quintero et al (4), according to the clinical and Doppler finding of EDF in the umbilical artery and ductus venosus of both fetuses:

Stage 1: donor bladder visible, EDF positive in both vessels in both fetuses.

Stage 2: donor bladder not visible, EDF positive in both vessels in both fetuses.

Stage 3: EDF absent or reversed in either vessel in either fetus.

Stage 4: presence of ascites or hydrops in either fetus; usually the recipient.

Stage V - The demise of 1 or both twins has occurred.

This staging has a role in the detection and diagnosis, leading to prompt treatment and improved prognosis.

The middle cerebral artery, umbilical artery, ductus venosus in both fetuses are assessed with Doppler sonography, abnormal Doppler studies are defined as absent or reversed end-diastolic velocity in the umbilical artery, reversed flow in the ductus venosus, or pulsatile flow in the umbilical vein.

Associated abnormalities:
- The incidence of chromosomal abnormalities or genetic syndromes is not increased.

Investigations:
- Detailed ultrasound examination.
- Ultrasound scans every 1 week to monitor growth, amniotic fluid volume and pulsatility index in the umbilical artery, middle cerebral artery and ductus venosus of both fetuses.

Management:
Therapeutic amnio-reduction can stop twin-to-twin transfusion and is therefore usually an effective treatment. Although amnioreduction is the most commonly used procedure for this condition, it is still unclear whether amnioreduction can change intrauterine circulation in TTTS. In cases requiring repeated amnio reductions and showing a persistence of the clinical syndrome, changes suggesting fetal hemodynamic improvement after laser coagulation are unlikely to be observed. (2)

Most fetal interventions are currently ultrasonographically guided. Monochorionic (MC) twin gestations are high-risk pregnancies, largely due to the vascular anatomy of the shared plaentaand to the presence of vascular connections.

Discordance in amniotic fluid (but not sufficient to fulfill the oligohydramnios / polyhydramnios sequence) with normal fetal Doppler:
- Overall survival: 95%.
- Progression to TTTS: 15%.
- Ultrasound scans every 1-2 weeks to monitor evolution.

Stage 1:
- Survival: overall 85%, at least one twin 90%.
- Progression to stages 2 to 4: 20%.
- Ultrasound scans every 1 week to monitor evolution.
- Endoscopic laser ablation of communicating placental vessels if progression to stages 2-4 or increasing polyhydramnios and shortening of cervical length.

Stages 2-4:
- <28 weeks: the best management is endoscopic laser ablation of communicating placental vessels; all communicating vessels should be ablated and the area between them should also be coagulated to achieve dichorionization of the placenta.
- ≥28 weeks: the best option is to deliver by cesarean section and the timing would depend on the Doppler findings in the umbilical artery and ductus venosus of both fetuses.
  - Stage 2: survival overall 75%, at least one twin 85%.
  - Stages 3 and 4: survival overall 60-70%, at least one twin 75-85%.
  - Neurodevelopmental impairment in survivors: 5-10%.
- Follow-up after laser therapy: ultrasound scans and Doppler every 1 week until resolution of the signs of TTTS and

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normalization of Doppler findings and every 2 weeks thereafter
with special attention for signs of brain damage, recurrence of
TTTS and development of TAPS.
• Normalization of amniotic fluid volume occurs after 1 week.
Resolution of cardiac dysfunction in the recipient and of hydrops
in stage 4 TTTS usually occurs after 3-4 weeks.
• In about 1,000 cases there may be limb amputation due to
thrombotic events or amniotic bands.

Delivery:
• Vaginally at 37 weeks if there is normal growth and Dopplers in
both babies.

Recurrence: No increased risk of recurrence

Differential diagnosis:
IUGR of one twin. To distinguish this from the twin-twin transfusion
syndrome the recipient twin does not usually have polyhydramnios or
congestive cardiac failure. IUGR may occur in dichorionic
pregnancies whereas the twin-twin transfusion syndrome only occurs
in monochorionic pregnancies.

Outcome:
Perinatal mortality of 71% (when diagnosed prior to 26 weeks).

FIGURES:

REFERENCES:
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