Twin-to-twin transfusion syndrome: a case report. Antepartum prediction of underlying placental vascular pattern in monochorionic twin pregnancies may be possible

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A case of twin-to-twin transfusion syndrome is described. Comparing data of serial antepartum ultrasonography with a haemodynamic model suggests the possibility of predicting the underlying placental vascular anatomy. It is suggested that serial ultrasonography, including full biometry, pulsatility indices of the umbilical arteries, foetal echocardiography, assessment of amniotic fluid indices and foetal bladder filling could serve as ultrasound parameters for pattern recognition of the underlying placental vascular anomaly. Biometry should be plotted serially in a difference/average plot. Future application of such intensive ultrasound monitoring in monochorionic twins, as soon as monochorionicity is established, may distinguish those monochorionic twins who may benefit from treatment from those whom it would be better only to observe.

Keywords: twin pregnancy, twin to twin transfusion syndrome, vascular anastomoses, monochorionicity, placenta, laser

Introduction

The twin-to-twin transfusion syndrome (TTTS) is a serious complication of monochorionic twins with considerable perinatal morbidity and mortality.1,2 Although the syndrome has received much attention since Schatz's publications3 more than a century ago, its underlying pathophysiology is poorly understood and diagnostic criteria are not unequivocally agreed.4 Neonatal criteria as formulated by Blickstein5 may not apply in utero. As a consequence, antenatal treatment is surrounded by controversy.6,7

The basis of the syndrome is a placental vascular anomaly.8 Placental anastomoses linking the two foeto-placental circulations produce an uncompensated net transfusion of blood from the donor to the recipient twin. Remarkably, however, the placenta does not seem to play a major role in therapeutic management decisions, and is rarely adequately investigated following delivery. Insufficient data are available relating diagnosis, foetal growth and therapy with placental anastomotic patterns. Our long term objectives are first to develop methods which allow antenatal assessment of placental anastomotic patterns, secondly to select subsequent rational therapy. We present a case of a monochorionic twin pregnancy showing serial ultrasonography obtained biometry, clinical and placental follow-up, and evaluation of the data by a haemodynamic model.

Case

A 34-year-old woman, gravida2, para1, was seen in her local hospital at 17 weeks and 6 days amenorrhea, because her pregnant uterus was too large for gestational age. Ultrasound examination revealed twins, and no intertwin membrane was seen.
Already a slight difference in foetal size was observed. Repeat ultrasonography one week later showed a very thin intertwin membrane, one placental mass, no lamda sign. Detailed scanning showed an increased echogenicity of the intestines, raising suspicion of a bowel occlusion in the smaller twin, and an enlarged heart with a dubious lining of pericardial effusion, raising suspicion of a congenital heart defect in the larger twin. The obvious difference in biometry and a considerable difference in amniotic fluid compartments of the twins raised suspicion of TTTS. Referral to a tertiary centre followed. At 19 weeks and 2 days amenorrhoea biometry indicated estimated foetal weights of 342 and 227 g (Hadlock), oligohydramnios and polyhydramnios (amniotic fluid index: 23 cm). A distinct difference in heart/thoracic ratios was present. The larger twin had a slightly enlarged heart, with a thin lining of pericardial effusion, and some tricuspid regurgitation was observed. No structural anomaly of the heart was seen. The larger twin had an enlarged, fully filled bladder during the complete examination. The smaller twin had increased echogenicity of the bowel compartment, but no abnormal dilatations were seen. Some bladder filling was observed. When one week later the discrepancy in fluid compartments further increased (AFI:26 cm), further deterioration to a full blown oligohydramnios/polyhydramnios sequence with one twin stuck was feared. The patient was transferred to a centre for laser ablation of the connecting placental vessels (Dr J Deprest, KU Leuven, Belgium). As some amniotic fluid and bladder filling in the smaller twin were still present, no immediate action was advised. Close follow-up was scheduled. If necessary, laser intervention could be performed at any time. At follow-up during the ensuing weeks stabilisation of the situation was observed. No further increase in amniotic fluid volume was seen. No preterm contractions and no discomfort arose from the polyhydramnios. No interventions, such as puncture of the intertwin membrane, amniocentesis or laser therapy were performed. Figure 1 shows the estimated foetal weights during the entire observation period. At 27 weeks hospitalisation was ordered for bed rest and foetal monitoring by daily cardiotocography, because the pulsatility index of the umbilical artery of the smaller twin was high (2.6) with absent end-diastolic flow (Figure 2). Three courses of corticosteroids were administered for foetal lung maturation. At 33 weeks a slight decrease in heart rate variability of the smaller twin was observed. A Caesarean section under spinal anaesthesia was performed. Two girls were delivered of 1585 g and 2135 g and in good condition (Apgar scores 8 and 9 after 1 and 5 min in both girls). The monochorial diamniotic placenta weighed 740 g. The cord of the recipient was centrally inserted and that of the donor velamentously. By inspecting the cotyledonar masses supplied by each chorial vasculature, it was estimated, that there was a 3:1 unequal sharing of the placental mass (recipient:donor). By dye-injection of the placental vasculature a large artery-to-artery (A–A) anastomosis of 4 mm in diameter and a large artery-to-vein (A–V) anastomosis of 6 mm in diameter, and a smaller A–V of less than 1 mm in diameter were visible.

The newborn infants were admitted to our neonatal intensive care ward. First haemoglobin levels were 11.7 mmol/l (recipient) and 11.0 mmol/l (donor). First postpartum blood pressure readings were 78/42 mmHg (mean arterial pressure (MAP): 59 mmHg) for the recipient and 48/25 mmHg (MAP: 38 mmHg) for the donor. The recipient twin had
tricuspid regurgitation and signs of decompensation cords. She was treated with furosemide. In the neonatal course she developed an intra-ventricular haemorrhage grade II with evidence of venous infarction. The donor twin’s neonatal course was uneventful. After one week both twins were transferred to the neonatal ward of their local hospital. Follow-up of both children at the child neurology department of the Free University Amsterdam (Dr Niokiktien) shows no developmental problems in either of the children to date (3 months old).

Comparison with the haemodynamic model

A haemodynamic computer model simulating the consequences of different types of placental vascular architecture in monochorionic twins has been proposed previously. This model shows in cases with unequal placental sharing and an A–V anastomosis compensated by an A–A anastomosis a decreasing effect of the difference between the two foetal weights normalised to the average of the two foetal weights. This plot of the ‘difference over the average ratio’ flattens out longer into the pregnancy. This is in contrast to an unopposed A–V anastomosis, where the curve steeply increases further (Figure 3). The ultrasound evidence of tricuspid regurgitation and increased heart/thoracic ratio in the larger twin is indicative of functional A–V anastomosis. The difference in the pulsatility indices of the arteriae umbilicales (from 19 weeks onwards: unchanged) reflects the unequal placental sharing. Flattening out of the difference/average plot and stabilising of the increase in amniotic fluid index of the recipient indicates compensation to the A–V shunt. An A–A anastomosis must be functioning. Comparing our ultrasound data with the model predictions (Figure 4), we feel that the model predicted to some extent the underlying placental vascular anatomy.

Discussion

We propose that it should be possible to predict the underlying placental anatomy by frequent detailed ultrasound scanning including full biometry, as soon as monochorionicity is established. Collecting consecutive cases of monochorionic twins using serial ultrasound monitoring and with definite pathological studies of the placentae is desperately needed for antepartum classification of monochorionic twins according to their placental problem. Early serial ultrasound scanning including full biometry, constructing growth curves with ‘difference:average ratio plots’ could make early recognition possible. This serial ultrasonography should also include assessments of the foetal hearts. Differences in heart/thoracic ratios or appearance of tricuspid regurgitation can be considered evidence for the haemodynamic effects of foeto-foetal transfusion. Also differences in foetal bladder filling and in amniotic fluid compartments should be looked for. The ratio of the pulsatility indices of the umbilical arteries may serve as an indicative measure of placental sharing, as the umbilical artery pulsatility index reflects placental resistance, which is proportional to placental mass. From these sonographic data, the various patterns predicting the underlying placental problem may ensure better diagnostic antepartum criteria. Hopefully this type of pattern recognition

Figure 3 Difference:average ratio of estimated foetal weight for the single arterio-venous component and unequal placental sharing (A–V), and for the arterio-venous plus arterio-arterial anastomotic pattern and unequal placental sharing (A–V–A–A)

Figure 4 Difference:average ratio of the estimated foetal weights, defined as the difference between the two measured weights divided by the average value of the two weights, as a function of gestational age. The solid line is from the model results shown in Figure 1. The offset of 0.45 at early gestational age corresponds to the 1:2.6 unequal placental sharing. The dashed line is a least squares fit of the clinical data.
will lead to early identification of cases requiring treatment.

We propose that an expert team comprised of an experienced ultrasonographer, a dedicated obstetrician, a neonatal intensive care specialist, a pathologist experienced in developmental pathology, and a developmental neurologist should manage mono-chorionic twin pregnancies.

References