

Detection of twin reversed arterial perfusion sequence at the time of first ultrasound screening and its spontaneous resolution: a case report

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SUMMARY: Detection of twin reversed arterial perfusion sequence at the time of first ultrasound screening and its spontaneous resolution: a case report.

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Since monochorionic twins share the same placenta, there is the possibility of the development of placental anastomoses and communication between the fetuses. The extreme form of

this anastomosis is the twin reversed arterial perfusion sequence (TRAPs). Some authors believe that prophylactic cord occlusion should be offered to all pregnancies of TRAPs. However exact criteria to deciding whether and when intervene have not been defined and it is not a procedure without complications. Anyway a spontaneous resolution is possible, even if rare. Here we present one of these cases of TRAPs with a good outcome without treatment. Weekly ultrasound is useful for the monitoring of TRAPs pregnancy to avoid intervention because spontaneous resolution is possible, as demonstrated in our experience.

KEY WORDS: Twin reversed arterial perfusion sequence - Acardiac twin - Monochorionic twin pregnancy - Ultrasound diagnosis.

Introduction

In monochorionic twin pregnancy fetuses result from a single zygote and share the same placenta. This occurs spontaneously in 0.3-0.4% of all pregnancies (1-4). Since the placenta is unique, there is the possibility of the development of anastomoses and communication between the two fetuses. The extreme form is the twin reversed arterial perfusion se-

quence (TRAPs). It is a rare condition that occurs in 1:9500 to 1:11000 pregnancies (5) (0.3-1% of monochorionic ones) (6-8). This is characterized by the presence of an acardiac twin (or with a severely malformed heart) that is hemodynamically dependent on a normal co-twin (pump twin). The malformation in the acardiac twin is incompatible with survival, while the normal twin has a variable mortality rate between 50-70% caused especially by congestive heart failure (9) and one third of them die before 16 weeks (10). Early sonographic diagnosis, ultrasound follow-up, and intervention are the tools of prenatal care associated with improvement in survival. Here we present the case of a pregnancy complicated by TRAPs and its outcome.

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Case report

A 41-year-old patient came to our attention at a postmenstrual age of 10 weeks+2 days after an in-vitro-fertilized twin gestation. She had performed elsewhere an ultrasound at 5 weeks of pregnancy that described the presence of two gestation bags and two embryos with heartbeat. At our first visit a 2D ultrasound defined it as monochorionic-diamniotic pregnancy. Twin A appeared grossly abnormal, acephalic and acardiac, without upper limbs, while sketched lower limbs were detected (Figure 1). Additionally, cystic hygroma and a single umbilical artery were seen. Twin B appeared grossly normal with a Crown-rump length (CRL) of 34,6 mm according to the amenorrhea. Suspecting a TRAPs, an extensive sonographic evaluation was instituted with a Doppler 3D study. This technique showed the artery-

to-artery anastomosis in the placental bed with a reversed flow in the acardiac twin's umbilical artery (Figure 2). Three-dimensional surface at 11 weeks gestation enhanced the extent of the malformations of the acardiac twin. A consultancy at a specialized center proposed an invasive treatment by laser coagulation within the 12th weeks of pregnancy. Informed about risks and benefits, the patient preferred the classic treatment at the 16th week with meanwhile periodic ultrasound checks. At the 12 weeks+1 day of gestation the ultrasound documented the absence of heartbeat and active movements of the malformed fetus that measured 30 mm with widespread edema. There was flow in the umbilical cord and normal amount of amniotic fluid. Morphology and size of fetus B were normal (CRL 61 mm). Nasal bone was visualized; no abnormalities were found in cardiac efflux. The venous duct wave and the amount of amniotic fluid appeared normal.

At the 13 weeks+3 days of gestation, the patient underwent villocentesis: the results confirmed the normal 46, XX karyotype of the healthy fetus. At the 13 weeks+5 days in the fetus B a mild polyhydramnios followed by slight dilatation of the left ventricle occurred. After seven days wave A of the venous duct inverted.

Consequently, we planned to laser coagulation of placental anastomoses for the 15th week. Meanwhile, the ultrasound monitoring continued to document the haemodynamic decompensation of the fetus B and the volumetric growth of the fetus A (57x40 mm at the 14th week).

Unexpectedly, at the last check the day before the surgery, ultrasound examination showed no flow in the umbilical cord. In the following days, there was an improvement of the polyhydramnios, a reduction of dilatation of the left ventricular chamber and the positivization of the wave A in the venous duct of the healthy fetus, showing a spontaneous resolution of the TRAPs. The patient was followed with weekly controls and four echocardiography every month, until 32th week. The delivery took place at the 39th week by caesarean section (self-determination). The newborn weighed 2850 gr and the Apgar Score was 9/10 at the first minute and 10/10 at the fifth one. Adjacent to the placenta the residue of the acardic fetus could be seen (Figure 3). Today the baby is ten months old and her development continues regularly.



Figure 1 - Two dimensional cross section of the malformed twin at 15th week: grossly abnormal, acephalic and acardiac twin, with and without misurations.



Figure 2 - Color pulsed Doppler waveform of umbilical artery of the acardiac twin.



Figure 3 - Residue of acardiac twin after birth: length 50 mm, weight 146 gr.

Discussion

The pathophysiologic mechanisms of TRAPs are not well understood. It is proved that there is artery-to-artery and vein-to-vein anastomosis through whom the twin donor (pump) sends blood to the acardiac fetus with reversal of flow in the umbilical cord vessels of the recipient twin. This has led to the

term TRAPs (11). Fully de-saturated blood flows to the upper body, leading to faulty development of the upper torso. The nourishment from the donor fetus suppresses the cardiac development in the recipient one causing acardiac or a severely malformed heart (12). Other researchers tried to propose different pathogenesis (13), but, regardless, the result is that the heart of the pumping twin has to support also the

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blood flow of the acardiac twin. This overload exposes the pump heart to the risk of heart failure and the high cardiac output increases perfusion of the kidneys, resulting in polyhydramnios. Diagnosis is made with the identification of the malformation of one fetus and of placental vascular anastomosis by Doppler. Moreover, Doppler studies show pathognomonic features of reverse flow on artery-artery anastomosis (12). The recipient twin can have different presentations based on the cephalic and trunical maldevelopment (14). The one in our case was the most common form, the acardius-acephalus one with rudimentary extremities. Nowadays, diagnosis of TRAPs can be mostly done at the end of the first trimester (15) as a consequence of the more widespread screening of chromosomal abnormalities (17). This early evaluation allows prompt intervention. The high risk of death for the pump twin has brought to the development of a wide range of intrauterine interventions: from the use of coils to the ultrasound-guided laser coagulation (17-21). Anyway it was showed that unexpected death of the pump twin may occur also after the treatment. That potential mechanisms include exsanguination into the TRAP fetus (22). In 60 consecutive pregnancies with TRAPs there were 10 (17%) spontaneous intrauterine fetal death of which 7 within 24 h mostly due to bleeding, and 3 belatedly, for cord entanglement in monoamniotic in 2 cases (23). Another complication associated is the preterm premature rupture of membrane (pPROM). This drawback occurred in 11 cases (18%) at a median of 9 weeks after the procedure according to Hecher (24). Other author found pPROM in 24% of complicated monochorionic pregnancies treated with ultrasound-guided bipolar cord coagulation (25). Also maternal complications have been reported such as bleeding, need for laparotomy, thermal injury, disseminated intravascular coagulation, chorioamnionitis and subsequent sepsis (26). Anyway, in the past, total post-procedure pump-twin mortality was found to be 13.6% (27), one quarter of the overall mortality reported for TRAPs without treatment (28). Based on this evidence, some authors believe that intervention should be offered to all TRAPs. However exact criteria to deciding whether and when intervene have not been defined. Treatment is currently reserved for after 16 weeks because early intramniotic procedures (as am-

niocentesis) showed an increased risk of miscarriage, membrane rupture and severe talipes (29-34). However prophylactic treatment at 12-16 weeks is advocated by some other authors (35). Finally, several experts support that expectant management is appropriate in the absence of poor prognostic features (36). Data of 10 cases of TRAPs treated after 16th week are available (37). Among them, there was 1 miscarriage after pPROM at 19 weeks and 9 (90%) pregnancies ended with delivery at a median gestation of 37 (28-40) weeks (37). The size of the acardiac fetus is the most important prognostic feature. Moore had already suggested in 1990 that acardiac-healty twin-weight ratio greater than 0.7 was associated with major incidence of preterm delivery (90%), polyhydramnios (40%) and heart failure (30%) (31). Most recently, Lewi demonstrated that there was no significant difference in size of the two twins in all 8 pregnancies with spontaneous demise of the pump twin (37). For these reasons, TRAPs are currently monitored every week by ultrasonography including size estimation, Doppler ultrasound and echocardiography looking for the signs of cardiac failure in the pumping twin: polyhydramnios, cardiomegaly, ascitis, and tricuspid regurgitation. In our case, the pump twin developed some of them leading to opt for an interventional behaviour. Anyway in some cases, as in ours, pregnancies ended in spontaneous resolution and term delivery. In fact, the interruption of the blood flow to the acardiac twin suppresses the cardiac stress and prevent further hemodynamic disorders. Lewi (37) reported the prognosis of 26 TRAPs. In 21% there was spontaneous cessation of blood flow to the acardiac twin and the birth of two pump twins at 37 and 40 weeks. There were two miscarriages at 19 and 23 weeks respectively. The last case pregnancy was terminated for a severe ventriculomegaly in the pump twin. Even Weisz (38) described other 6 cases. Two pregnancies were treated conservatively and ended with spontaneous cessation of blood flow in the umbilical artery. Both delivered at term. In conclusion, weekly ultrasound, including size estimation, Doppler ultrasound and echocardiography are useful for the monitoring of TRAPs pregnancy to avoid intervention because spontaneous resolution is possible, even if rare. However, long-term follow-up is necessary to rule out neurological sequelae of the pump fetuses.

Conflict of interest

The Authors report no conflict of interest.

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Disclosure statement

The Authors do not have any financial disclosures or disclosures of any other kind.

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