

The twin reversed arterial perfusion syndrome (TRAP sequence)

Jakub Wyroba^{1,2,A–B,D,F} , *Magdalena Maria Piróg*^{3,D–E} ,
Joanna Figula^{4,B–C} 

¹ Malopolski Institute of Fertility Diagnostics and Treatment – KrakOvi, Krakow, Poland

² Fertility Disorders Clinic, Medical College, Andrzej Frycz Modrzewski Krakow University, Poland

³ Clinic of Gynaecological Endocrinology at the Jagiellonian University Medical College in Krakow, Poland

⁴ Artvimed, Krakow, Poland

A – Research concept and design, B – Collection and/or assembly of data, C – Data analysis and interpretation, D – Writing the article, E – Critical revision of the article, F – Final approval of the article

Abstract

Background: The twin reversed arterial perfusion syndrome (TRAP sequence) is a rare congenital sequence and a serious complication of monochorionic multiple pregnancies. The frequency of this sequence is estimated at 1:35,000 pregnancies and 1% of monochorionic pregnancies. In this sequence, one fetus is described as a “semi-donor” or “twin-pumping” (autosite), and the second as a twin acardiac (acardiac twin). The cause is considered to be arteriovenous anastomoses in the placenta, which lead to blood transfusion between the twins.

Material and methods: In this article, we report a case of monochorionic monoamniotic pregnancy resulting from natural fertilization, complicated by TRAP sequence.

Results: The patient gave birth by cesarean section at 34 weeks’ gestation to a neonate weighing 2350 g, length 43 cm, with an Apgar score of 8/10.

Conclusions: The outcome of a course of maternity care and possible treatment options in the event of TRAP sequence, as reviewed in the literature, are presented in the article.

Keywords: monochorionic pregnancy complications, Twin reversed arterial perfusion (TRAP sequence)

Introduction

Reversed arterial perfusion sequence (TRAP), also known as twin reversed arterial perfusion sequence (TRAP), is a rare congenital anomaly that constitutes a serious complication of multiple monochorionic pregnancies. Its incidence is estimated at one in 35,000 pregnancies and 1% of monochorionic pregnancies [1]. In this syndrome, one foetus is referred to as the “donor twin” or “pumping twin” (autosite), while the other is

✉ Jakub Wyroba, e-mail: jwyroba@uafm.edu.pl

referred to as the acardiac twin. The cause of this sequence is believed to be arteriovenous anastomoses within the chorionic plate. This abnormal vascularization of the shared placenta leads to blood transfusion between the twins. Deoxygenated blood flows through the umbilical artery of the anaemic twin and returns through the umbilical vein, leading to underdevelopment of the upper body parts of this twin [2]. In this situation, the donor twin is forced to supply blood to the other foetus through the collateral arterial circulation of the shared placenta. This may result in the development of congestive heart failure in the pumping twin. The presence of the sequence increases the risk of preterm birth due to increasing polyhydramnios or iatrogenic factors. Given the estimated mortality rate of acardiac twins, the goal of obstetric care is to save the donor twin by isolating their circulatory systems. This is achieved by caesarean delivery [3], endoscopic vascular ligation [4], intravascular insertion of a metal coil [5], ultrasound-guided chemical embolization [6,7] (e.g., with 100% ethanol), or laser obliteration [8].

Case Report

A 30-year-old patient in her first pregnancy, 22 weeks pregnant, with RhD serological incompatibility, was scheduled for a consultation and further diagnostic evaluation at the Clinic of Gynaecological Endocrinology at the University Hospital in Krakow. Previous ultrasound examinations at another centre had reported abnormal findings. At 13+3 weeks of gestation, the patient underwent a first-trimester genetic ultrasound using a GE Voluson 730 Expert system with a 4-8 L RAB transducer. An acardiac foetus was suspected.

From the interview it emerged that the patient had conceived naturally after four or five months of trying. Four months before conception, she began taking folic acid 0.4 mg/day and took iodine for the first three months of pregnancy. The patient had a seven-day viral infection during which she did not receive pharmacotherapy.

At 23+0 weeks of gestation, a follow-up ultrasound was performed, and the diagnosis of an acardiac foetus was confirmed (twin A: CRL – 78.6 mm, BPD – 57.3 mm, HC – 211 mm, OFD – 73.3 mm, AC – 185 mm, FL – 38.7 mm; twin B: FL = 38.4 mm).

At 24+0 weeks of gestation, the patient underwent an MRI (Table 1). At 26+2 weeks of gestation, the patient was readmitted to the Clinic due to threatened preterm labor caused by a urinary tract infection. Antibiotic therapy improved her general condition and resolved her dysuria. Steroid therapy was then administered. The patient was discharged home in good

general condition, with a viable pregnancy. Her due date was scheduled for May 8, 2012, according to Naegele.

Table 1. MRI report from 18.01.2012
<p>The examination was performed using SSET2, FSPGRT1fs, DWI, and 2DFIESTA sequences in the coronal, transverse, and sagittal planes, with slice thicknesses of 4 mm, 5 mm, and 8 mm.</p> <p>Pregnancy I, 24 weeks.</p>
<p>Twin pregnancy. The placenta was located on the posterior wall, with discreetly higher signal levels visible in the central part – lobular oedema? No cystic lesions or haemorrhages were detected. The amount of amniotic fluid was normal.</p> <p>Fetus A: alive, male, in a breech presentation. Eyeballs and facial structures were properly developed. The skull was normal and symmetrically arched. Brain structures were developed appropriately for the foetus’s age – the neural tissue layer thickness and signal were normal, the ventricular system was not dilated and without displacement. The cerebellum and pons were normal. The spinal canal was of normal width, symmetrical, and without signs of clefting. A single spinal cord was visible within the canal. The nuchal cistern was of normal width.</p> <p>The heart and lungs were normal. The liver, spleen, stomach, small intestines, large intestine, kidneys, and urinary bladder were normally developed. The foetal limbs were normally developed. The feet and hands were normal.</p> <p>Fetus B: most likely male, with a narrow, single-vessel umbilical pedicle, lower limbs, and pelvis (dystrophic bony structures) visible, with significant subcutaneous tissue oedema (16.7 mm thick). The remaining organs of the second foetus were missing.</p>
<p>No septum/membrane was detected separating foetuses A and B, suggesting a monoamniotic pregnancy.</p>
<p>The image is consistent with reverse arterial perfusion syndrome of the acardius amorphous type, and no neural tissue was detected, ruling out acardius myelacephalus.</p>

On March 28, 2012, at 34+0 weeks’ gestation, the patient was readmitted to the Clinic due to amniotic fluid leakage. An ultrasound was performed to assess the pregnancy (Figs. 1 and 2), and a CTG was obtained (Fig. 3).

The pregnancy was terminated by caesarean section using the Misgav-Ladach procedure. She delivered a live son weighing 2350 g and measuring 43 cm in length, with an Apgar score of 8/10 cm, and a stillborn foetus weighing 98 g and measuring 12 cm in length (Figs. 4 and 5).

An extensive general and neurological examination of the newborn revealed no abnormalities. The newborn was septic, and given the significant obstetric history, empirical antibiotic therapy was prescribed and continued for nine days.

Blood and urine cultures were negative. During hospitalization, the newborn had good circulation, and basic vital signs were normal. From birth, the newborn was enterally fed his mother's milk with the addition of a pre-term formula. The newborn gained weight normally. On the second day, he was vaccinated against hepatitis B and BCG. Due to RhD incompatibility (mother: blood group B, RhD negative; father: RhD positive), a qualifying test for administration of anti-D immunoglobulin was performed. No anti-D antibodies were detected in the test; therefore, the patient was qualified for administration of 300 mg of anti-D immunoglobulin the day after caesarean section. Due to an uncomplicated postpartum period, the patient was discharged home in good general and local health on the fourth day after delivery.

Figure 1. TA ultrasound result - pregnancy 34 +0 weeks. - pregnancy 34 +0 weeks



Figure 2. Ultrasound result - pregnancy 34 +0 weeks

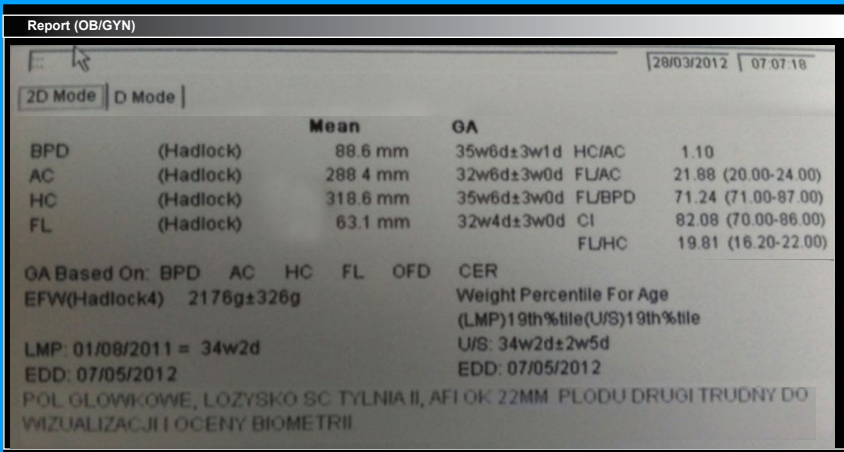


Figure 3. Recording CTG - pregnancy 34+0 weeks

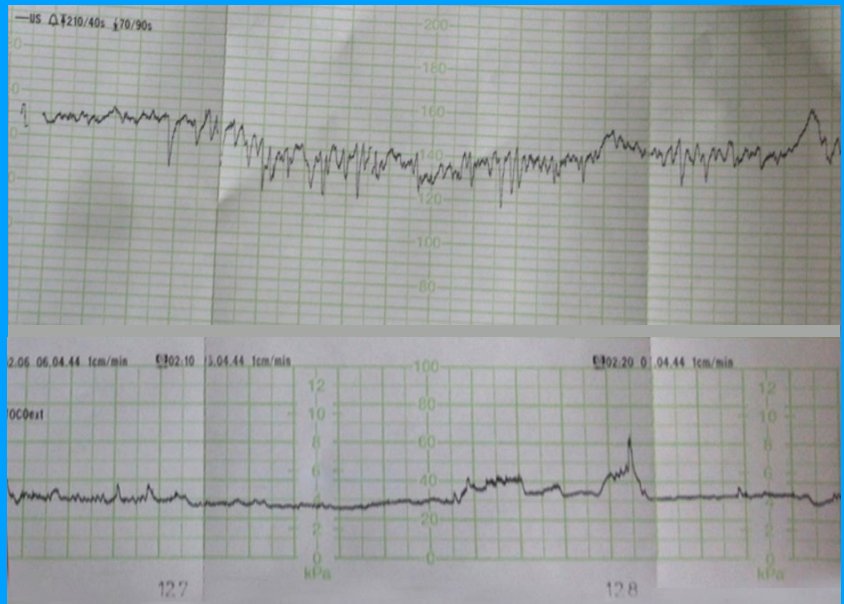


Figure 4,5. Twin B after birth

Discussion

The goal of obstetric care in pregnancies complicated by TRAP sequence is to save the donor twin by separating the circulatory systems of both twins – stopping blood flow in the abnormal twin. Of the many available methods, such as endoscopic vascular ligation, laser obliteration, and less commonly used techniques such as intravascular metal coil insertion or ultrasound-guided chemical embolization, we chose caesarean delivery. The patient was under the Clinic's constant care throughout the pregnancy. The choice of treatment in this case was based primarily on the good general condition of twin A and the absence of a heartbeat in the twin considered acardiac. In our case, the absence of any signs of foetal damage in twin A was considered unusual. The literature describes abnormalities in the donor twin, such as generalized hydrops fetalis, intrauterine growth restriction (IUGR), soft tissue oedema, pleural effusion, hepatosplenomegaly, and myocardial damage leading to foetal death, severe heart failure, pericardial effusion, or tricuspid valve regurgitation [9,10].

References

1. Chandramouly M, Namitha. Case series: TRAP sequence. *Indian J Radiol Imaging*. 2009;19(1):81–83.
2. Giménez-Scherer JA, Davies BR. Malformations in acardiac twins are consistent with reversed blood flow: liver as a clue to their pathogenesis. *Pediatr Dev Pathol*. 2003;6(6):520–530.
3. Ginsberg NA, Applebaum M, Rabin SA, Caffarelli MA, Kuuspalu M, et al.. Term birth after midtrimester hysterotomy and selective delivery of an acardiac twin. *Am J Obstet Gynecol*. 1992;167(1):33–37.

4. McCurdy CM Jr, Childers JM, Seeds JW. Ligation of the umbilical cord of an acardiac-acephalus twin with an endoscopic intrauterine technique. *Obstet Gynecol.* 1993;82(4 Pt 2 Suppl):708–711.
5. Diehl W, Hecher K. Selective cord coagulation in acardiac twins. *Semin Fetal Neonatal Med.* 2007;12(6):458–463.
6. Sepulveda W, Sfeir D, Reyes M, Martinez J. Severe polyhydramnios in twin reversed arterial perfusion sequence: successful management with intrafetal alcohol ablation of acardiac twin and amniodrainage. *Ultrasound Obstet Gynecol.* 2000;16(3):260–263.
7. Sepulveda W, Corral E, Aiello H, Otaño L, Paredes R, et al. Intrafetal alcohol chemosclerosis of acardiac twins: a multicenter experience. *Fetal Diagn Ther.* 2004;19(5):448–452.
8. O'Donoghue K, Barigye O, Pasquini L, Chappell L, Wimalasundera RC, et al. Interstitial laser therapy for fetal reduction in monochorionic multiple pregnancy: loss rate and association with aplasia cutis congenita. *Prenat Diagn.* 2008;28(6):535–543.
9. Moore TR, Gale S, Benirschke K. Perinatal outcome of forty-nine pregnancies complicated by acardiac twinning. *Am J Obstet Gynecol.* 1990;163(3):907–912.
10. Dąbrowska K, Gadzinowski J. Czy operacje wewnątrzmaciczne są uzasadnione? – perspektywa neonatologa. – Część III. Zespół Przetoczenia Między Bliźniętami (Twin to Twin Transfusion Syndrome – TTTS) i Wrodzone Wady Serca (CHD). *Ginekolog Pol.* 2011;82(7):525–532.

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