

# Diagnosis, evaluation, and delivery of primipara with twin reversed arterial perfusion (TRAP) syndrome

V. Paunović<sup>1</sup>, M. Zamurović<sup>1</sup>, T. Paunović<sup>1</sup>, Z. Protrka<sup>2</sup>

<sup>1</sup>University Clinic for Obstetrics and Gynecology "Narodni front", Belgrade; <sup>2</sup>University Clinical Centar, Kragujevac (Serbia)

## Summary

Twin reversed arterial perfusion (TRAP) sequence is a rare complication in monochorionic twin pregnancy. It is the most serious type of transfusion syndrome (TTTS). One twin has serious malformations, and due to the fact that the heart is almost always missing, it is often called acardiacus. TRAP sequence is present in less than 1% of monochorionic pregnancies, while the incidence compared to normal pregnancies is one in 35,000 pregnancies. This paper presents the diagnostics, evaluation, and completion of pregnancy in a primipara with twin pregnancy complicated by the TRAP syndrome.

**Keywords:** TRAP syndrome; Twins; Acardiacus.

## Introduction

Twin reversed arterial perfusion sequence, also called TRAP sequence, TRAPS, or acardiac twinning, is a rare complication of monochorionic twin pregnancies. It is a severe variant of twin-to-twin transfusion syndrome (TTTS). TRAP sequence occurs in 1% of monochorionic twin pregnancies and in one in 35,000 pregnancies overall [1].

This anomaly is characterized by the twins' blood systems being connected instead of independent. One twin, called the acardiac twin or TRAP fetus, is severely malformed. The heart is missing or deformed, hence the name acardiac, as are the upper structures of the body. The legs may be partially present or missing, and internal structures of the torso are often poorly formed. The other twin is usually normal in appearance. The normal twin, called the pump twin, drives blood through both fetuses. It is called reversed arterial perfusion because in the acardiac twin the blood flows in a reversed direction.

TRAP sequence can be diagnosed using obstetric ultrasound. Doppler examination will confirm that blood flow in the acardiac twin is in the reverse direction, entering via the umbilical cord artery and exiting through the vein [2, 3].

After diagnosis, ultrasound and amniocentesis are used to rule out genetic abnormalities in the pump twin. A procedure may then be performed which will stop the abnormal blood flow. The acardiac twin may be selectively removed. The umbilical cord of the acardiac twin may be surgically cut, separating it from the pump twin, during fetoscopic cord occlusion. Also a radiofrequency ablation needle may be used to coagulate the blood in the acardiac twin's um-

bilical cord; this latter procedure is the least invasive. These procedures greatly increase the survival chances of the pump twin, to about 80% [4]. If left untreated, the pump twin will die in 50-75% of cases.

## Case Report

An expectant mother, 38-years-old, first reported to the clinic in week 24 of a twin pregnancy for evaluation. Immediately after admission, obstetric ultrasound was performed. Color Doppler exam verified the presence of abnormal circulation. After detailed examinations, it was ascertained to be a case of twin pregnancy complicated by TRAP syndrome. Protocol called for amniocentesis. It showed a normal karyotype in the first twin - donor. Two weeks later, in gestational week 26, spontaneous delivery began. A live new born girl weighing 900 grams was delivered vaginally. She was immediately placed under pediatric care and then transported to Institute for Neonatology. Upon the birth of the first twin - donor, the umbilical cord was clamped and an ultrasound examination repeated, confirming the presence of a mixed tumefact consisting of solid and bone tissue in close contact with the placenta, corresponding to the second twin - the acardiacus. Slightly after an hour after from the birth of the first twin, a profuse bleeding occurred *ex utero*. Because of profuse bleeding and suspected placental abruption, not being able to extract the second twin - acardiacus, vaginally, the decision was made to finish the delivery of the acardiacus surgically, by cesarean section in mother's best interest. The cesarean section, performed in the usual way, gave birth to an anomalous fetus weighing 1,500 grams (Figure 1).

## Discussion

The prognosis in TRAP syndromes is lethal for acardiac twin. The fetal mortality of the pump twin is extremely high



Figure 1. — Acardiac twin.

Histopathological examination confirms a case of monochorionic diamniotic placenta with marginal insertions of both cords and anomalous acardiacus. After three months under intensive care, the first twin (donor) was discharged from the Institute for Neonatology in satisfactory health condition.

[50-75%] because of high output cardiac failure. References mention only rare individual cases of twin pregnancy complicated by TRAP syndrome left untreated.

Kalyani and Bindra described a case of TRAP syndrome in which the acardiacus lived longer than the pump twin who had neurological problems and survived for 24 hours while acardiac twin was stillborn [5].

References describe cases of expecting mothers who did not accept the risks of interventions, such as the electrocoagulation of the cord during fetoscopy. The problem lies in high risk for the donor twin in the remaining part of the pregnancy. The outcome of the pregnancy is uncertain for this twin, considering that the other twin – acardiacus, already suffers from intrauterine malformations and for him/her the outcome is surely lethal, usually already in the uterus, and most definitely after the delivery [6].

The outcome of pregnancies complicated by TRAP syndrome depends on conditions of the mother and of the donor twin. The donor twin is supervised by cardiotocography and color Doppler ultrasound. Problems may appear due to the fact that such pregnancies end significantly before the term, so the twin expected to survive is often in danger because of early gestational age and consequent circulation problems, causing most authors to decide to end the pregnancy surgically, i.e. by cesarean section. On the other hand, vaginal birth with constant cardiotocographic monitoring of the condition of the donor involves the vaginal delivery of the acardiacus as well, which can be expected to be longer and requires extreme patience. Delivery of the placenta has its own risks, due to the position of the

placenta, its morphology, pathological lesions expected, and connected with the TRAP syndrome, that can lead to bleeding during third and fourth stages of labour, sometimes requiring surgical intervention.

## Conclusion

Diagnosis is necessary for proper prenatal management and can be established by Doppler examination of the umbilical artery of the abnormal twin. Optimal management of acardiac twin pregnancy is controversial as it is seen rarely and with different variations. The goal of therapy is the salvage of the pump twin. Fetal and neonatal mortality rates of the pump twin is high. Therefore, invasive methods toward eliminating the acardiac twin are selected. Discontinuation of the blood flow to the acardiac twin is the method that is applied by many authors. These methods include endoscopic umbilical cord ligation, sclerosis of the umbilical cord with alcohol, thermocoagulation of the umbilical cord and aorta under ultrasonography guidance, and coagulation of the umbilical cord [7]. Cases such as the present are rare where pregnancy ends in spontaneous childbirth at the end of the second trimester and where the donor twin survives after intensive supervision, while the acardiacus requires cesarean section delivery in the interest of the mother because of the complications during delivery.

## References

- [1] Chandramouly M., Namitha.: "Case series: TRAP sequence". *Indian J. Radiol. Imaging*, 2009, 19, 81.
- [2] Okai T., Ichizuka K., Hasegawa J., Matsuoka R., Nakamura M., Shimodaira K., *et al.*: "First successful case of non-invasive in-utero treatment of twin reversed arterial perfusion sequence by high-intensity focused ultrasound". *Ultrasound Obstet. Gynecol.*, 2013, 42, 112.
- [3] Ichizuka K.I., Hasegawa J., Nakamura M., Matsuoka R., Sekizawa A., Okai T., Umemura S.: "High-intensity focused ultrasound treatment for twin reversed arterial perfusion sequence". *Ultrasound Obstet. Gynecol.*, 2012, 40, 476.
- [4] Tsao K., Feldstein V.A., Albanese C.T., Sandberg P.L., Lee H., Harrison M.R., Farmer D.L.: "Selective reduction of acardiac twin by radiofrequency ablation". *Am. J. Obstet. Gynecol.*, 2002, 187, 635.
- [5] Kalyani R., Bindra S.M.: "Twin Reversed Arterial Perfusion Syndrome (TRAP or Acardiac Twin)-A Case Report". *J. Clin. Diagn. Res.*, 2014, 8, 166.
- [6] Pepe F., Teodoro M.C., Luca C., Privitera F.: "Conservative management in a case of uncomplicated trap sequence: a case report and brief literature review". *J. Prenat. Med.*, 2015, 9, 29.
- [7] Buyukkaya A., Tekbas G., Buyukkaya R.: "Twin Reversed Arterial Perfusion (TRAP) Sequence; Characteristic Gray-Scale and Doppler Ultrasonography Findings". *Iran J. Radiol.*, 2015, 12, e14979.

Corresponding Author:

M. ZAMUROVIC, M.D.

University Clinic for Obstetrics and Gynecology  
"Narodni front", 62, Kraljice Natalije St.

11000 Belgrade (Serbia)

e-mail: mzamurovic@gmail.com