

Conservative Management of Twin Reversed Arterial Perfusion Sequence (TRAP): Case Report

“Twin Reversed Arterial Perfusion (TRAP)” Sekansında Konservatif Yaklaşım

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ABSTRACT Twin reversed arterial perfusion (TRAP) sequence is a rare but severe complication of monochorionic multiple pregnancies. Perinatal mortality for the pump twin is around 35-50%. A woman aged 32, gravida 2, para 1 was referred for twin pregnancy and in utero fetal demise of co-twin at 28 weeks of gestation. Sonography demonstrated TRAP sequence. Cord occlusion was not performed since it had a higher mortality rate than an appropriately grown fetus delivered at 28 weeks of gestation. She delivered a healthy 2000 g female infant at 31+2 gestational week by cesarean section for footling presentation during preterm labor. The acardiac acephalus twin was 1400 g. Pump twin had an uneventful postnatal course. In the absence of poor prognostic factors, pump twins can be managed conservatively if mortality rate due to procedure exceeds perinatal mortality for that particular gestational age.

Key Words: Pregnancy; pregnancy complications; pregnancy outcome

ÖZET İkiz ters dönmüş arteriyel perfüzyon (TRAP) sekansı monokoryonik çoğul gebeliklerin nadir ama ağır bir komplikasyonudur. Perinatal mortalite %35-50 arasında verilmektedir. Otuziki yaşında gravid 2 para 1 olan ve 28 haftalık ikiz gebeliği bulunan bir kadın, ikiz eşinin intrauterin ölümü nedeniyle kliniğimizde değerlendirilerek sonografide TRAP sekansı saptandı. Kord oklüzyonu nedeniyle perinatal mortalite oranı 28 haftada doğan normal bir fetusunkinden fazla olacağından, bu işlem yapılmayan hasta yakın takibe alındı. 31+2 gestasyonel haftada ayak gelişi ile preterm doğum nedeniyle sezaryen yapılan hastaya 2000 g sağlıklı bir kız bebek doğurtuldu. Akardiyak ikiz 1400 gramdı ve pompa olan fetusta postnatal dönemde bir özellik saptanmadı. Kötü prognostik faktörlerin yokluğunda, eğer prosedüre bağlı perinatal mortalite o gestasyonel hafta için olandan fazla ise tedavide konservatif kalınabilir.

Anahtar Kelimeler: Gebelik; gebelik komplikasyonları; gebelik sonucu

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Twin reversed arterial perfusion (TRAP) sequence is a rare, but severe complication of monochorionic multiple pregnancies. It affects about one in 35.000 pregnancies, one in 100 monozygotic twins, and one in 30 monozygotic triplets. It is characterized by a structurally abnormal, acardiac twin perfuse through a single artery-to-artery anastomosis by the normal, ‘pump’ co-twin. Although the pathogenesis is still under debate, the primary defect is believed to be the single artery-to-artery and vein-to-vein anastomoses between the cords or on the surface of the placenta. Retrograde perfusion of deoxygenated and nutrient-poor low pressure blo-

od from the 'pump' twin is believed to result in disruptions and incomplete morphogenesis of the recipient twin, the acardiac anomaly spectrum.¹

The natural course of TRAP sequence may differ from patient to patient and the reported perinatal mortality for the 'pump' twin is around 35-50%.² Pregnancies reaching term uneventfully and spontaneous cessation of reversed umbilical arterial flow have been reported.³ However, continuous growth of the acardiac fetus may increase the cardiovascular load of the 'pump' twin leading to congestive heart failure, polyhydramnios and in utero fetal demise. In addition, the mass effect of acardiac twin along with polyhydramnios increases the risk of preterm delivery.⁴ Several prognostic factors such as the acardiac-to-pump twin weight ratio, cardiac failure in the pump twin, polyhydramnios, and certain morphologic characteristics of the acardiac fetus and Doppler indices of the umbilical arteries have been suggested and individualization of management has been advocated. Here we present a case of TRAP sequence managed conservatively in our institution late at second trimester and propose a gestational age based management. Informed consent of the patient was obtained.

CASE REPORT

A 32-year-old, gravida 2, para 1 woman was referred to our institution with the diagnosis of twin pregnancy and in utero fetal demise of the co-twin at 28 weeks of gestation. Sonographic examination revealed monochorionic-diamniotic twin pregnancy. Twin A was appropriately grown, consistent with the last menstrual period. Amniotic fluid volume was normal and no structural anomalies were detected. Twin B appeared grossly abnormal (Figure 1). Cranium and upper extremities were not visualized. There was no cardiac activity. Generalized subcutaneous edema and large cystic spaces, resembling a giant cystic hygroma, were seen. The spine and lower extremities were rudimentary. Twin B appeared almost the same size as Twin A. Color Doppler ultrasonography demonstrated reversed umbilical arterial blood flow towards the acardiac fetus, confirming the diagnosis of TRAP sequence. Umbilical venous and ductus venosus



FIGURE 1: A view of the acardiac twin during ultrasound.

Doppler findings were normal. Fetal echocardiography of twin A was normal; signs of cardiac failure including tricuspid regurgitation were not evident. The parents were informed in detail regarding the prognostic factors and possible risks, and they opted for conservative management. Fetal blood sampling was performed and normal karyotype was confirmed for Twin A. Weekly biophysical profile and follow-up scans for evaluation of signs of cardiac insufficiency, hydrops fetalis, intrauterine growth restriction, polyhydramnios and Doppler examination were scheduled.

At 29 weeks of gestation, the patient was admitted with threatened preterm labor. Sonography and Doppler findings were normal, and labor was arrested successfully with oral nifedipine. Betametasone (12 mg) was also administered intramuscularly twice in 24 hours to enhance fetal lung maturity. At 31 weeks and two days of gestation, the patient was readmitted with preterm labor. Amniotic fluid volume was at the upper limits of normal, but there were no other signs of cardiac failure. No tricuspid regurgitation and normal Doppler findings in ductus venosus was detected. However, tocolysis was unsuccessful and cesarean section due to footling presentation of the 'pump' twin was performed. A female baby, weighing 2000 g and with Apgar scores of 7 and 9, at one and five minutes, re-

spectively, was delivered. The acardiac acephalus twin weighed 1400 g (Figure 2). The placenta was monochorionic-diamniotic, and single artery-to-artery and vein-to-vein anastomoses between the cord insertions were noted on the surface (Figure 3). The pump twin had an uneventful postnatal course and was discharged in good condition.

CONCLUSION

The twin reversed-arterial-perfusion (TRAP) sequence found in monozygotic twins is a consequence of primary or secondary cardiac development disruption and direct arterio-arterial and veno-venous placental anastomoses. Many prognostic factors cited in papers and survival rate given for the pump twins are based on a retrospective study written by Moore before widespread use of sonography.² Sin-



FIGURE 2: The acardiac twin.



FIGURE 3: Appearance of the monochorionic-diamniotic placenta.

ce Moore's study, no further prognostic factors to predict the outcome of pump twin are commonly accepted and proven to contribute to survival. Prognosis of pump twin varies from case to case; some pump twins reach term without any intervention while others show rapid progress to heart failure and hydrops. Thus it is very difficult to compare the treatment alternatives without a pretreatment staging. It is a very rare condition and the scattered cases are managed in different centers with different treatment strategies. It seems that primary key point in management of the condition is timing: timing of intervention and delivery. The method of intervention may also be a secondary key point and mainly depends on the capability and experience of the centers.

Poor prognostic factors include polyhydramnios, cardiac insufficiency, large acardiac twin and rapid increase in blood flow to acardiac twin.³ The mortality rate of pump twin is mainly affected by two important factors: in utero cardiac failure and preterm birth due to polyhydramnios.⁴ Polyhydramnios should be followed up by serial sonographic evaluation. Hydrops may be a late sign of heart failure. Possible fetal cardiac insufficiency can be safely predicted by several methods such as tricuspid regurgitation and ductus venosus Doppler. In our case, twin A had no poor prognostic factors at the time of diagnosis. During serial ultrasonographic evaluation, no signs of fetal cardiac failure such as hydrops, venous Doppler abnormalities or tricuspid regurgitation occurred. However, a moderate increase in amniotic fluid volume took place. The pregnancy was allowed to continue until 32 weeks of gestation when preterm delivery ensued.

Overall perinatal mortality rate for pump twin is reported as high as 55% by Moore.² Following various therapeutic interventions such as fetoscopic or ultrasound-guided cord occlusion, the post-procedure pump twin mortality has been reported to be between 13% and 38%.⁴ Considering that the perinatal mortality rate of a correctly dated, appropriately grown singleton fetus delivered at 28-29 weeks is around 10-20%, beyond this gestational age it seems that it is not appropriate to perform

an invasive procedure which has a higher mortality rate.⁵ After 28 weeks of gestation, instead of complicated invasive procedures such as cord coagulation or intrafetal ablation, delivery of the fetus may be considered to treat ensuing fetal heart failure.⁶ Between 24 and 28 weeks of gestation pump twin must be evaluated with serial ultrasound examinations to detect the development of polyhydramnios and any signs of cardiac decompensation. In the absence of any signs of cardiac failure, polyhydramnios can be managed by serial amnioreduction at any gestational age before term. Amnioreduction is not a treatment modality that corrects the underlying mechanism; however, it may ameliorate the problems related to polyhydramnios. In case of cardiac failure, an invasive

intervention may be required to cease the blood flow to acardiac twin between 24 to 28 weeks of pregnancy. Before 24 weeks of gestation, the option of termination of pregnancy may be offered to parents. If the parents prefer to continue pregnancy, the pump twin should be managed similar to the cases managed before 28 weeks of the pregnancy.

As in our case, in the absence of poor prognostic factors, pump twin can be managed conservatively. Procedure related mortality and long-term and short-term morbidity rates of several invasive treatment choices are not available yet. In order to compare the benefits of therapeutic interventions, cases must be classified according to severity of the condition prior to the procedure.

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