

# Trap Sequence: Case Report

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TRAP (Twin Reversed Arterial Perfusion) sequence is a rare syndrome seen in monochorionic twin pregnancies. It occurs in 1% of monochorionic twin pregnancies. It is characterized with a recipient fetus exhibiting lethal anomalies including acardia and a pump fetus supplying blood by vascular anastomosis in the placenta. Pump twin is structurally normal but heart failure may develop with cardiomegaly, pericardial and pleural effusion, ascites and polyhydramnios also seen during pregnancy. This anomaly has high mortality for pump twin and is always lethal for recipient. Here, a TRAP sequence case diagnosed at 14 weeks of gestation in antenatal period and followed up spontaneously until labor, with the demand of the family is presented.

**Keywords:** TRAP, Acardiac twin

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## Introduction

Twin reversed arterial perfusion (TRAP) sequence known as acardiac malformation, is a rare complication unique to monozygotic multiple gestations. It occurs in approximately 1 per 35.000 pregnancies<sup>1</sup> and 1/30 in monochorionic triplets.<sup>2,3</sup> Pathophysiology is not entirely understood as yet. The retrograde transfer of blood from the pump twin to the acardiac twin occurs at the level of the shared placenta through anomalous arterial-to-arterial connections. The blood reaching the acardiac twin is poor in oxygen and preferentially perfuses the lower extremities and body, resulting in lower concentrations of oxygen reaching the superior body and head. Perfusion of the acardiac twin with poorly oxygenated blood is thought to be the cause of the predictable pattern of multiple anomalies seen in these fetuses, which are more pronounced in the superior aspect of the body and head.<sup>4</sup>

Here, we report a case of TRAP sequence diagnosed at 14 weeks of gestation and followed up spontaneously until labor,

with the demand of the family is presented. Also clinical presentation, prognostic factors and therapeutic options were discussed in this case report.

## Case Report

A 25-year-old woman, gravida 8, para 1, was seen at 14 weeks for a screening ultrasound (US) examination in our department. We diagnosed a monochorionic-monoamniotic pregnancy with a TRAP sequence. The pregnancy was spontaneous; dating was obtained by menstrual history and confirmed at 14 weeks by US measurement of crown-rump length (CRL). The acardiac twin (fetus A) was found to be substantially enlarged due to an important skin edema (hydrops fetalis) (Figure 1). The pump twin (fetus B) showed normal biometry on US with a biparietal diameter (BPD) of 28 mm, head circumference (HC) of 108 mm, abdominal circumference (AC) of 88 mm, and femur length (FL) of 14.5 mm. The CRL measurement of acardiac twin (fetus A) was 50.2 mm. The fetus A had acephaly, there was no fetal heart and the arms appeared abnormally rudimentary. The lower limbs and the lower part of the body were normal (Figure 1). Discrepancy in CRL measurements was greater than 3% between the TRAP twin and the isolated normal twin. Patient and family were informed about gestation and its prognosis. Selective cord coagulation was recommended for treatment but they denied and decided spontaneous follow up of pregnancy. Follow-up US examinations using a Sonoace 9900 (Medison; Seoul, Korea) equipped with a volumetric 6.5 MHz two-dimensional transducer showed persistent fetal blood flow for perfusion in acardiac twin.

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Figure 1: TRAP sequence in monozygotic twins (Acardiac acephalic hydropic fetus)

Amniocentesis was also performed, and the two fetuses had normal karyotype (46 XX; 46 XX). The patient was discharged 24 hours after the procedure. The following weeks were uneventful, and Doppler studies confirmed persistent umbilical blood flow in the hydropic twin and normal resistance in both uterine arteries with a mean pulsatility index (PI) of 1.2 and without notches. Neonatal respiratory distress syndrome prophylaxis was performed at 28 weeks [betamethasone (Celestone Chronodose, Schering-Eczacıbaşı, Lüleburgaz, Turkey) two doses a 12 mg intramuscular with another dose 12 h later] to accelerate fetal lung maturation. Labor started spontaneously after a few days, at 28 weeks of gestation. Tocolysis was performed but it was not successful and birth of twins occurred at 28 weeks of gestation. The first twin had a birth weight of 1120 g and an Apgar score of 9/10; the second twin was ex and had a birth weight of 550 g (Figure 2). The normal twin was hospitalized for nearly 35 days and had a normal course.

## Discussion

TRAP sequence is a rare complication of multiple pregnancies for which the pathophysiology is not entirely understood as yet. The mortality rates for pump twins have been

noted to be as high as 50% to 70%. The reported perinatal mortality of the pump twin is approximately 55%.<sup>5</sup>

Management of twin pregnancies complicated by TRAP sequence includes imaging evaluation and close monitoring to decide whether and when invasive treatment is deployed. Doppler US remains the standard method of diagnosis of prognostic factors in TRAP sequence. Estimated fetal weight is one of prognostic factors in TRAP.<sup>6</sup> TRAP is characterized with a recipient, acardiac fetus and a pump fetus supplying blood by vascular anastomosis in the placenta. After perfusion of the acardiac twin and further deoxygenation, the blood is returned to the venous circulation of the pump twin, bypassing the placenta and thus contributing to chronic hypoxemia and growth restriction of pump twin. Furthermore, the increasing size of the hydropic acardiac twin may lead to a significant increase in intrauterine volume, with the consequence of premature labor and preterm delivery.<sup>7</sup> Moore et al.<sup>5</sup> calculated the ratio of fetal weights between the acardiac twin and its pump cotwin in 49 sets of twins with TRAP sequence by US. They reported increased rates of poor perinatal outcome due to preterm delivery, polyhydramnios and congestive heart failure when the weight ratio was greater than 70%.<sup>8</sup> It was reported that if this ratio was greater than 70%, preterm labor occurred



Figure 2: The acardiac twin with acephaly, only rudimentary upper limbs, and normal twin after delivery

in 90%, polyhydramnios in 40% and congestive heart failure in 30%.<sup>5</sup> Also low pulsatility index of umbilical artery in pump twin was suggested as a sign of poor prognosis.<sup>9</sup> Since then, many US calculations have been developed using AC or fetal length to calculate estimated weight ratios as prognostic indicators for the pump twin.<sup>4,10,11</sup> In our case, weight ratio of twins was smaller than 70%, Doppler studies confirmed normal resistance in both uterine arteries with a mean pulsatility index (PI) of 1.2 and without notches and these were all good prognostic factors. Besides, chromosomal abnormalities were detected at an incidence of 9% in pump twins<sup>12</sup> while they have been reported in ratio of 30% in acardiac twins<sup>13</sup> and this can also influence prognosis and decisions about fetal intervention. In present case report, amniocentesis was done and result was reported as normal karyotype for both fetuses.

In literature, there are some controversies about conservative and interventional treatment of TRAP sequence. Conservative management includes close observation and non-surgical intervention focus on the prevention of complications, by serial amniocentesis or maternal use of indomethacin to decrease amniotic volume and maternal use of digoxin to treat congestive heart failure.<sup>14,15</sup> On the other hand, there have been many successful interventions described, associated with increased survival of the pump twin. These therapies all attempt to interrupt the vascular connections between the twins<sup>2,13</sup> include US- or fetoscopic-guided therapies aimed at obliteration of either the umbilical cord or the main intraabdominal vessels of the acardiac fetus.<sup>2,3,16-18</sup> Such procedures are endoscopic umbilical cord ligation,<sup>19</sup> sclerosis of the umbilical cord with alcohol,<sup>20</sup> US-guided thermo coagulation of the umbilical cord and aorta,<sup>21</sup> endoscopic laser coagulation of the umbilical artery<sup>22</sup> and bipolar coagulation of the umbilical cord.<sup>23</sup> Intrafetal radiofrequency ablation (RFA) of the acardiac twin, with ablation performed within the fetal abdomen at the umbilical cord insertion, has been associated with survival rates as high as 95%.<sup>10,24</sup> Quintero et al.<sup>25</sup> described umbilical cord occlusion either by ligation with a suture with or without trans-section of the cord or by laser photocoagulation of the cord or of the arterio-arterial and veno-venous anastomoses in 51 patients. Their technical success rate was 88%. In contrast, umbilical cord ligation with trans-section of the cord seemed to be technically the most complicated procedure, with a preterm premature membrane rupture (PPROM) rate of 36% (4/11) within three weeks in cases that underwent successful cord occlusion.<sup>25</sup> Tan and Sepulveda<sup>26</sup> compared the cases treated by heterogeneous cord occlusion techniques such as embolization, cord ligation, laser coagulation, bipolar and monopolar diathermy with the cases treated by various intrafetal ablation techniques such as alcohol, monopolar diathermy, interstitial laser and radiofrequency. They concluded that intrafetal ablation is associated with a higher clinical success rate than the cord occlusion technique, with an

overall pump twin survival rate of 76%. The main complication associated with these techniques was PROM. Hecher<sup>27</sup> reported that PROM occurred in only 18% of cases at a median interval of nine weeks after the laser surgery, and PROM rate before 34 weeks was reported as 24% if the bipolar coagulation was indicated.<sup>27</sup>

In conclusion, there are significant risks involved with these procedures, so prognostic information about the pump twin is important for decisions regarding deployment and timing of therapeutic intervention.<sup>28</sup> Several experts support the current approach, stating that expectant management is only appropriate in the absence of poor prognostic features.<sup>29</sup> Perinatal mortality of cases when treated conservatively is between 10-50%.<sup>30</sup> Sullivan<sup>17</sup> reported one intrauterine fetal death out of 10 cases, and cautioned against aggressive intervention and recommended conservative approach. In our case, preterm labor started and birth occurred at 28 weeks of gestation. By advanced neonatal care, prognosis of pump twin was good but of course, it is impossible to predict handicaps about developmental stages of childhood. However, regarding the neurodevelopmental outcome of the surviving pump twin, a recent series of cord coagulations in complicated monochorionic multiple pregnancies suggested that interventions at an advanced gestational age, and when signs of cardiac decompensation were already present, correlated with mental delay and unfavorable neurological outcome.<sup>6</sup> To overcome these controversies about management of TRAP sequence, advanced researches including higher number of cases are needed.

## TRAP Sekansı: Olgu Sunumu

TRAP (Twin Reversed Arterial Perfusion) sekansı monokoryonik ikiz gebeliklerde görülen, çok ender bir sendromdur. Monokoryonik ikiz gebeliklerde %1 oranında görülmektedir. TRAP sekansında multipl anomalilere sahip, kalbi gelişmemiş bir fetus ile bu fetusu plasentadaki vasküler anastomozlar yoluyla besleyen pompa fetusun varlığı söz konusudur. Pompa fetus yapısal olarak normaldir ancak bu fetusta, gebelik esnasında kardiomegali, perikardial efüzyon, plevral efüzyon, asit ve polihidramnios ile birlikte kalp yetmezliği, gelişebilir. TRAP sekansı pompa fetus için yüksek mortalite oranına sahip iken, alıcı fetus için hemen her zaman ölümcüldür. Burada ondördüncü gebelik haftasında tanı konulan, ailenin istemi doğrultusunda konservatif gözlem ile takip edilen, akardiyak asefalik monokoryonik ikiz olguyu sunduk.

**Anahtar Kelimeler:** TRAP sekansı, Akardiyak fetus

## References

1. Nakata M, Sumie M, Murata S, Miwa I, Matsubara M, Sugino N. Fetoscopic laser photocoagulation of placental

- communicating vessels for twin-reversed arterial perfusion sequence. *J Obstet Gynaecol Res* 2008;34:649-52.
2. Napolitani FD, Schreiber I. The acardiac monster: a review of the world literature and presentation of 2 cases. *Am J Obstet Gynecol* 1960;80:582-9.
  3. James WH. A note on the epidemiology of acardiac monsters. *Teratology* 1977;16:211-6
  4. Steffensen TS, Gilbert-Barness E, Spellacy W. Placental pathology in TRAP sequence: clinical and pathogenetic implications. *Fetal and Pediatr Pathol* 2008;27:13-29
  5. Moore TR, Gale S, Benirschke K. Perinatal outcome of forty-nine pregnancies complicated by acardiac twinning. *Am J Obstet Gynecol* 1990;163:907-12
  6. Lewi L, Gratacos E, Ortibus E. Pregnancy and infant outcome of 80 consecutive cord coagulations in complicated monochorionic multiple pregnancies. *Am J Obstet Gynecol* 2006;194:782-9.
  7. Chang DY, Chang RY, Chen RJ, Chen CK, Chen WF, Huang SC. Triplet pregnancy complicated by intrauterine fetal death of conjoined twins from an umbilical cord accident of an acardius. A case report. *J Reprod Med* 1996;41:459-62
  8. Bornstein E, Monteagudo A, Dong R, Schwartz N, E. Timor-Tritsch. Detection of Twin Reversed Arterial Perfusion Sequence at the Time of First-Trimester Screening, The Added Value of 3-Dimensional Volume and Color Doppler Sonography. *J Ultrasound Med* 2008;27:1105-9
  9. Dashe JS, Fernandez CO, Twickler DM. Utility of doppler velocimetry in predicting outcome in twin reversed-arterial perfusion sequence. *Am J Obstet Gynecol* 2001;185:135-9.
  10. Livingston JC, Lim J-Y, Polzin W. Intrafetal radiofrequency ablation for twin reversed arterial perfusion (TRAP): a single-center experience. *Am J Obstet Gynecol* 2007;197(399):1-3
  11. Hadlock FP, Harrist RB, Sharman RS. Estimation of fetal weight with the use of head, body, and femur measurements-a prospective study. *Am J Obstet Gynecol* 1985;151:333-7
  12. Peterson BL, Broholm H, Skibsted L, et al: Acardiac twin with preserved brain. *Fetal Diagn Ther* 2001;16:231-3
  13. Van Allen MI, Smith DW, Shepard TH. Twin reversed arterial perfusion (TRAP) sequence: a study of 14 twin pregnancies with acardius. *Semin Perinatol* 1983;7:285-93
  14. Mohanty C, Mishra OP, Singh CP, Das BK, Singla PN. Acardiac anomaly spectrum. *Teratology* 2000;62:356-9.
  15. Aggarwal N, Suri V, Saxena S, Malhotra S, Vasishta K, Saxena AK. Acardiac acephalus twins: A case report and review of literature. *Acta Obstet Gynecol Scand* 2002;81:983-4
  16. Healey MG. Acardia: predictive risk factors for the co-twin's survival. *Teratology* 1994;50:205-13.
  17. Sullivan AE, Varner MV, Ball RH, Jackson M, Silver RM. The management of acardiac twins: a conservative approach. *Am J Obstet Gynecol* 2003;189:1310-13.
  18. Sepulveda W, Sebire NJ. Acardiac twin: too many invasive treatment options e the problem and not the solution. *Ultrasound Obstet Gynecol* 2004;24:387-9.
  19. McCurdy CM Jr, Childers JM, Seeds JW. Ligation of the umbilical cord of an acardiac-acaphalus twin with an endoscopic intrauterine technique. *Obstet Gynecol* 1993;82:708-11.
  20. Sepulveda W, Bower S, Hassan J, Fisk NM. Ablation of acardiac twin by alcohol injection into the intra-abdominal umbilical artery. *Ostet Gynecol* 1995;86:680-1.
  21. Rodeck C, Deans A, Jauniaux E. Thermocoagulation for the early treatment of pregnancy with an acardiac twin. *N Engl J Med* 1998;339:1293-5.
  22. Ville Y, Hyett JA, Vandenbussche FP, Nicolaidis KH. Endoscopic laser coagulation of umbilical cord vessels in twin reversed arterial perfusion sequence. *Ultrasound Obstet Gynecol* 1994;4:396-8.
  23. Deprest JA, Audibert F, Schoubroeck DV, Hecher K, Mahieu-Caputo D. Bipolar coagulation of the umbilical cord in complicated monochorionic twin pregnancy. *Am J Obstet Gynecol* 2000;182:340-5.
  24. Tso K, Feldstein VA, Albanese CT. Selective reduction of acardiac twin by radiofrequency ablation. *Am J Obstet Gynecol* 2002;187:635-40
  25. Quintero RA, Chmait RH, Murakoshi T. Surgical management of twin reversed arterial perfusion sequence. *Am J Obstet Gynecol* 2006;194:982-91.
  26. Tan TY, Sepulveda W. Acardiac twin: A systematic review of minimally invasive treatment modalities. *Ultrasound Obstet Gynecol* 2003;22:409-19.
  27. Hecher K, Lewi L, Gratacos E, Huber A, Ville Y, Deprest J. Twin reversed arterial perfusion: Fetoscopic laser coagulation of placental anastomoses or the umbilical cord. *Ultrasound Obstet Gynecol* 2006;28:688-91.
  28. Carolina V. A, Guimaraes M, Beth M, et al. MRI findings in multifetal pregnancies complicated by twin reversed arterial perfusion sequence (TRAP). *Pediatr Radiol* 2011;41:694-701
  29. Wong AE, Sepulveda W. Acardiac anomaly: current issues in prenatal assessment and treatment. *Prenat Diagn* 2005;25:796-806
  30. Akercan F, Demirtas GS, Demirtas Ö, Kazandı M, Karadadas N. Trap (twin reversed arterial perfusion) sequence. *Ege J of Med* 2009;48(2):123-6