Cumhuriyet Medical Journal, 44(2): 223-227, 2022
DOI: http://dx.doi.org/10.7197/cmj.1110498

Cumhuriyet Medical Journal

| cmj.cumhuriyet.edu.tr | Founded: 2004 Available online, ISSN:1305-0028 Publisher: Sivas Cumhuriyet Üniversitesi

A Case of Trap Sequence (Acardiac Twin) Treatment with Conservatively

Rabia Şerife Beğendik[1a*], İsmail Yaman[1b], Begüm Kurt[2c], Nazan Yurtçu[2d]

[1]Sivas Cumhuriyet University, Faculty of Medicine, Sivas Turkey, [2]Sivas Cumhuriyet University, Faculty of Medicine, Department of Obstetrics and Gynaecology, Sivas, Turkey

**Corresponding author*

Case Report

History

Received: 29/04/2022
Accepted: 15/07/2022

ABSTRACT

TRAP sequence (twin reversed arterial perfusion) is a complication with a poor prognosis, seen only in monochorionic twin pregnancies, known as acardiac twin formation[1]. In the very early stages of the formation periods of twin pregnancies, as a result of the single formation of the placenta and the defective formation of the cord vessels of the second baby (acardiac twin), the heart and accordingly the brain development cannot occur in the baby with placenta abnormalities; but other parts of the body can develop in different combinations. Therefore, the second baby can develop from a separate creature to the other baby as a parasite. The normal twin, is known as the pump twin and provides the blood circulation of the acardiac twin. These inter twin anastomoses lead to circulatory confusion, the reverse circulatory morphogenesis interrupted and acardiac twinning occurs[2]. This case is presented because the patient had a healthy twin who was born due to the onset of preterm labor that was not terminated at the request of the family, and the other twin with acardiac anencephaly. Ablation treatment could not be performed due to the delay in our case, and the pump twin was delivered in a healthy way.

Keywords: Acardiac, monochorionic, pregnancy, pump twin, RFA ablation, TRAP

Konservatif Yöntem ile Yönetilen Trap Sekansı (Akardiak İkizlik) Vakası

Süreç

Geliş: 29/04/2022
Kabul: 15/07/2022

ÖZ

TRAP sekansı (ters arterial kanlanma sekansı) akardiak ikiz oluşumu olarak bilinen sadece monokoryonik ikiz gebeliklerde görülen kötü prognoza sahip bir komplikasyondur [1]. İkiz gebeliklerin oluşum evrelerinin çok erken dönemlerinde plasentanın tek oluşması ve ikinci bebeğin kordon damarlarının kusurlu oluşumu neticesinde, ikiz eşi olan bebekte kalp ve buna bağlı olarak beyin gelişimi oluşamaz, ancak vücudun diğer uzuvları değişik kombinasyonlarda gelişebilir. Dolayısı ile ikinci bebek ayrı bir canlı varlıktan ziyade diğer bebeğe bir parazit eklenti olarak gelişebilir. Normal ikiz, pompa ikiz olarak bilinir ve akardiak ikizin kan dolaşımını sağlar. İkizler arası bu anastomozlar dolaşım karmaşasına yol açar, oluşan tersine dolaşım morfogenezi duraksatır ve akardiak ikizlik ortaya çıkar[2]. Bu olgu ailenin isteği doğrultusunda sonlandırılmayan erken doğum eyleminin başlaması nedeniyle doğumu gerçekleştirilen sağlıklı ikiz eşi ve akardiak anensefali olan diğer ikiz eşi olması nedeniyle sunulmaktadır. Ablasyon tedavisi olgu geç kalındığı için yapılamamış olup pompa ikiz sağlıklı bir biçimde doğurtulmuştur.

Anahtar sözcükler: Akardiyak, gebelik, ikiz, monokoryonik, RFA ablasyon, TRAP

[a] *tempus.ticking@gmail.com* *https://orcid.org/ 0000-0002-8967-5017*
[b] *md.ismailyaman@gmail.com* *https://orcid.org/ 0000-0001-8949-9052*
[c] *dr.begumkurt@yahoo.com.tr* *https://orcid.org/ 0000-0002-7166-3130*
[d] *nazanyurtcu@cumhuriyet.edu.tr* *https://orcid.org/ 0000-0003-4725-043X*

How to Cite: Beğendik RŞ, Yaman İ, Kurt B, Yurtçu N (2022) A Case of Trap Sequence (Acardiac Twin) Treatment with Conservatively, Cumhuriyet Medical Journal, June 2022, 44 (2): 223-227

Introduction

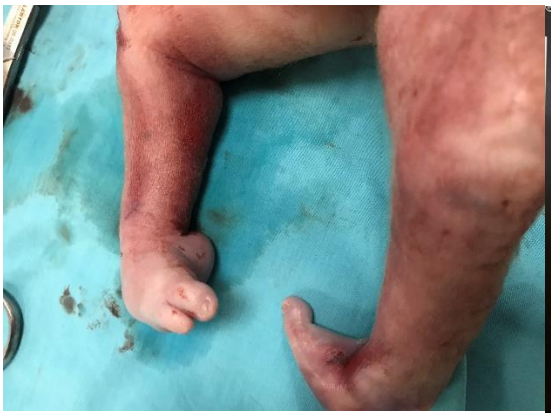
The incidence of Trap sequence in the world is 2,6% of monozygotic twins and it is a malformation seen in 1 in 35.000 pregnancies, and this rate is increasing gradually[3]. Today, the reason for the increase in the incidence of TRAP sequence; it may be diagnosed by ultrasonography, which is widely used in the first trimester, and in vitro fertilization, which causes an increase in the incidence of multiple pregnancies. Acardiac Twins or TRAP Sequence (Twin reversed arterial perfusion) is a rare complication of monokaryonic twin pregnancy. While acardiac twin is a severely abnormal twin with rudimentary or no heart (acardiac twin); there is a twin perfusing their common twin through abnormal arterio arterial anastomoses (pump twin). Acardiac twin is completely dependent on the circulatory support of the pump twin, and the lower extremities and cranium, which in some cases do not develop at all, are often underdeveloped. Therefore, the acardiac twin cannot survive in the postnatal period. As a result, if untreated, the circulatory load supports the acardiac twin, while the pump twin is at risk for preterm birth and death caused by heart failure and other complications. A multiloculated dorsal cystic hygroma is usually present. The acardiac twin is usually oligohydramniotic[4]. TRAP Sequence can also be seen in monochorionic triplet and high level multiple pregnancies. Despite the presence of trunk and extremity movements in the twin with multiple anomalies, the diagnosis can be made in the antenatal period when the heart cannot be demonstrated by ultrasonography and vascular anastomoses in the placenta are revealed by doppler ultrasonography. While the acardiac twin progresses with a mortal course, the normal twin may be exposed to polyhydramnios, which may cause heart failure and premature birth after developing circulatory load. This case is presented because of the acardiac twin who was delivered by cesarean section at the age of 29 weeks and 6 days without any intervention due to the family's request for follow up of the pregnancy.

The exact pathogenesis of the TRAP sequence is not fully known. There are two theories of its pathogenesis. In the first one; abnormal arterio arterial anastomoses in early embryogenesis, supply deoxygenated blood to an acardiac twin with no direct placental perfusion. This is the reason for acardiac twin liveIn the second theory; an acardiac twin with defective early cardiac embryogenesis has low systemic arterial pressure and has backward blood flow from its twin, and normal tissues in the acardiac twin are partially resorbed. Regardless of the exact cause, the net result is that the acardiac twin is dependent on the pump twin for circulatory support from the beginning of the first trimester[5]. In the TRAP sequence, the pump twin maintains a normal fetal circulation pattern. In addition, part of the cardiac output passes through one or more arterio arterial anastomoses and then continues retrograde to one or both umbilical arteries and the systemic circulation of the acardiac twin, thus creating "reverse" circulation perfusion. Reverse circulation with relatively deoxygenated blood from the pump twin contributes to the development of a wide variety of structural abnormalities in the acardiac twin. It usually shows varying degrees of necrosis, poor development and absence. The lower half of the acardiac twin is thought to receive a share of the pump twin's circulatory support, which may be why the fetal abdomen, pelvis, and lower extremities are relatively better developed than the trunk, upper extremities, and head[6,7].

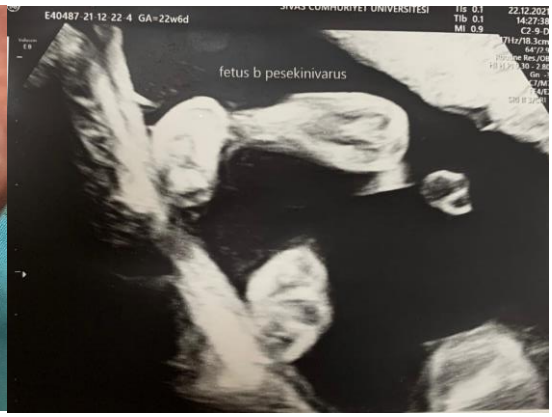
Case Report

A 21 years old patient, who had her first pregnancy (spontaneously pregnant without assisted reproductive pregnancy)applied to our perinatology outpatient clinic with monoamniotic monokaryonic twin pregnancies. According to the last menstrual period, the thoracic cavity of the acardiac twin fetus was narrow in the ultrasonography of the patient who was 20 weeks and 6 days pregnant, and the lungs and heart could not be visualized. In addition, neck and cranial structures above the thorax level are not seen. Although fetal intra abdominal structures could not be seen clearly, diffuse ascites and clustered intestinal loops with increased echogenicity were observed. It was found to be compatible with diffuse circumferential cystic hygroma and hydrops was extending in both thighs, being more common in the fetal body part. Club foot deformity draws attention in both feet (Picture 1). Umbilical cord calibration decreased and single artery and single vein were observed (Picture 2,3 and 4). In the obstetric examination, 22 weeks and 6 days of monochorionic monoamniotic twin pregnancy and TRAP sequence findings were detected in ultrasonographyIn ultrasonography examinations are; as of the week of fetal plans, it was followed as normal as possible. No cardiomegaly or tricuspid regurgitation was observed. Anasarca style widespread edema and cystic areas were observed on the skin. Ascites in the abdomen and pes equinovarus deformity in the lower extremities were observed. Retrograde arterial flow to the acardiac twin was observed in doppler ultrasonography. The patient and her husband were informed about the trap sequence and adverse obstetric outcomes. Amniocentesis and microarray examination were recommended because the risk of genetic anomaly increased in cases with trap sequence. Pregnancy termination was offered as an option. The patient and her husband decided to continue the pregnancy. It was explained that they could be referred to an advanced center for fetal cord occlusion (RFA ablation, bipolar cord ablation). The patient and her husband accepted the evaluation with periodic ultrasonographic examination and preferred the conservative method. Routine controls of the patient who was referred from another center at 20 weeks and 6 days pregnant were followed up 11 times until delivery, once or more than once a week. The patient was admitted to our emergency department at 29 weeks of gestation due to pain. The patient had uterine contractions. She was admitted to the obstetrics service due to premature

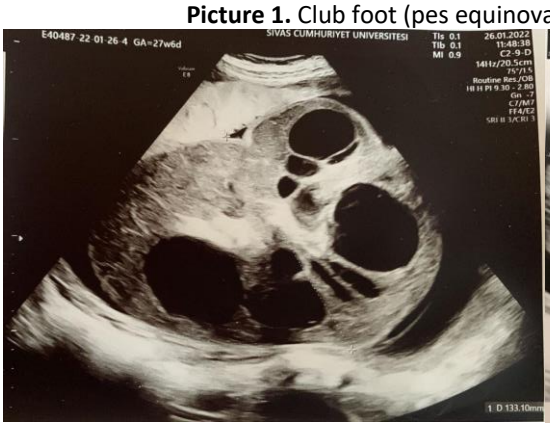
birth. Clinical observation was made. The patient, whose cervical length was measured as 25 mm, was started on antenatal steroid treatment and 2 g maintenance treatment after 4 g magnesium sulfate loading. After the completion of the treatment periods, the contractions continued, and the patient was started on nifedipine as tocolytic therapy, and delivery was planned because the uterine contractions could not be controlled and the cervical length was shortened. A male baby with a cesarean section of 1781 g, 43 cm, 1 min/ 5 min Apgar score of 4/8 was delivered in a healthy condition (Picture 5). Then later, acardiac/anencephaly twin and placenta were delivered (Pictures 6,7,8 and 9). Acardiac/anencephaly twin and placenta were sent to the pathology laboratory. In the pathological examination report, mature immature intermediate and terminal chorionic villi are observed in the placenta material; surrounded by membranes, with a 17 cm length and 1,5 cm diameter umbilical cord with a central entrance. Umbilical cord containing 2 arteries,1 vein,chorionic and amniotic membranes section were observed The pump twin was hospitalized in the neonatal intensive care with the diagnosis of prematurity. Examination and follow up were performed in terms of anomalies that may accompany the pump twin (Cardiology examination and abdominal ultrasonography were performed to check for heart failure and cardiological anomalies.). There is no abnormality in the results.



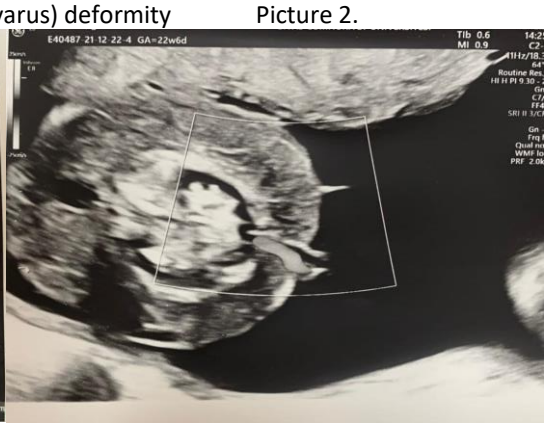
Picture 1. Club foot (pes equinovarus) deformity



Picture 2.

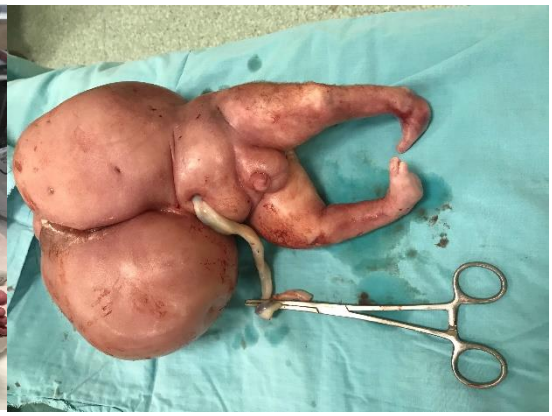


Picture 3.

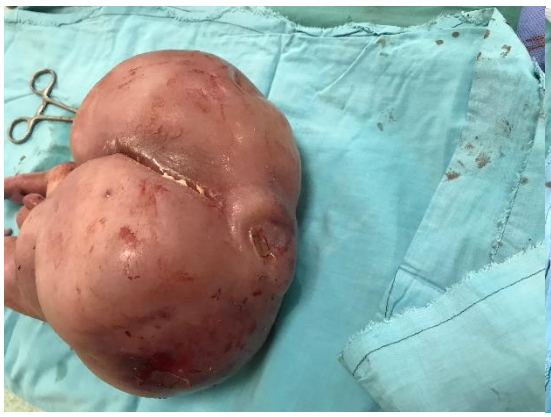


Picture 4.

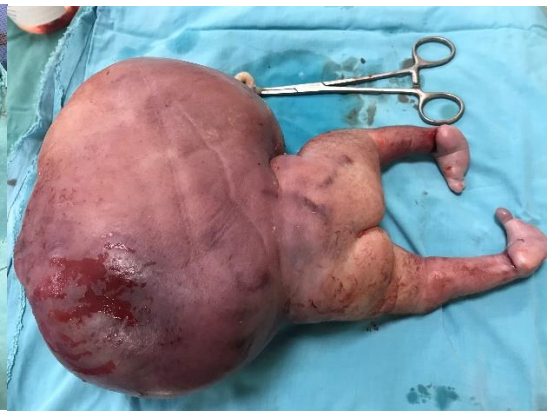
Picture 5. The Pump Twin



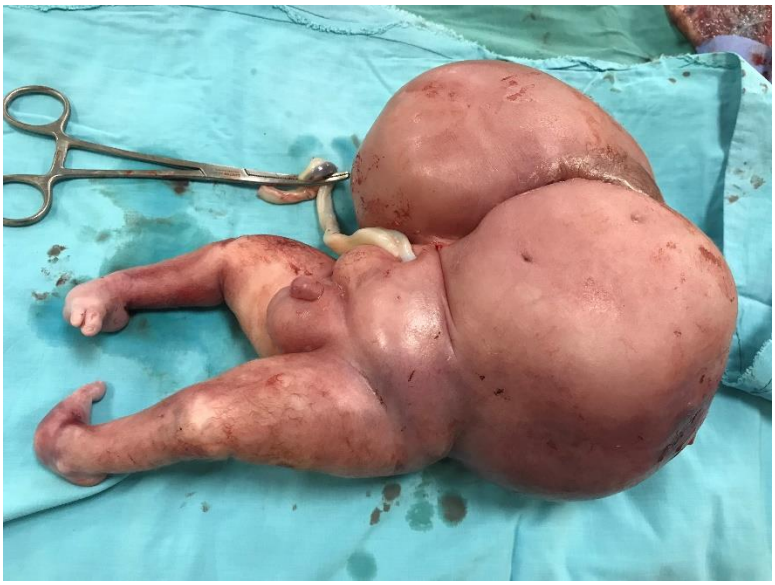
Picture 6. The Acardiac Twin



Picture 7. The Acardiac Twin



Picture 8. The Acardiac Twin



Picture 9. The Acardiac Twin

Discussion

There are vascular connections between monozygous twins. In this case, there is a blood transfusion between the twins. Mortality is very high without treatment. Severely malformed acardiac twin has no heart (holocardia) or only rudimentary cardiac tissue (pseudocardia) and has many developmental anomalies[8]. The pump twin provides blood flow to the acardiac twin with an opposite flow in umbilical artery. The mortality of the acardiac twin is 100%. In monochorionic twins, which results in the late separation of the mesothelial cell mass in the embryonic period, the anastomotic vessels provide flow in two directions. With reversible perfusion, oxygenated blood passes in the opposite direction to the acardiac twin through anastomic channels. Thus, the acardiac twin becomes dependent on the pump twin, which provides blood flow with perfusion. R C Prameela et al. in one of his studies[9] he explained:

1. In early embryogenesis, deep placental anastomoses cause malformations in the acardiac twin. As a result of early pressure decrease in a twin; it is fed from the umbilical artery by drawing the reverse flow in the other twin.
2. Cardiac development is interrupted in a twin as a result of a primary defect in embryogenesis. Then the pump twin then perfuses the acardiac twin through the arterio arterial anastomoses. These anastomoses are not the cause of cardiac anomalies; a result of cardiac anomalies.

Trap sequence is a malformation that can be diagnosed by the absence of the heart of the acardiac twin on doppler ultrasonography. In the advanced stage of the pump twin, 30% heart failure and 40% polyhydramnios may occur. With early diagnosis and treatment methods, the TRAP sequence can be determined and the acardiac twin can be saved by medical termination. Minimally invasive procedures, such as percutaneous insertion of a helical metal coil, can be performed to induce thrombogenesis in the acquis umbilical artery[10]. In 1994, Quintero et al. described the first successful fetoscopic ligation of the umbilical cord for TRAP at 19 weeks of gestation[11].

Early diagnosis and treatment of cases are very important in terms of prognosis. We have compiled 2 cases that were lost due to premature birth and delivered healthy if treatment could not be performed because it was too late.

In a study conducted by M. Chandramouly Namitha in 2009; in a 29 years old case who was found to be

carrying monochorionic monoamniotic twins in the 2nd trimester (22-23 weeks), the patient was diagnosed late and delivered prematurely when it was 27 weeks old, and the normal pump twin was also lost[12].

In our case, the pump twin was delivered in a healthy way, despite the premature birth and delayed diagnosis. Under conditions where early diagnosis is not possible and in delayed cases, conservative treatment may be successful instead of pregnancy termination.

In a study conducted by Enesia Mudenha Ziki et al. in 2019, in a 27 years old patient who was consulted after the death of his 26 weeks old intrauterine twin; ultrasound scan showed monochorionic diamniotic twins with normal fluid volume in the viable twin, but the nonviable twin had large edema of the fetal skin and no skull bone. The patient, who was followed up with two week ultrasound scanning and doppler velocimetry, was delivered by cesarean section when no diastolic flow in the umbilical artery was observed in the ultrasound scan performed at 34 weeks. The pump twin was delivered healthy and has no congenital anomalies[13].

After this case report, we showed that there is a possibility of giving birth to a healthy baby without termination of pregnancy in the TRAP sequence.

Results

It constitutes a scientific resource for future acardiac twin pregnancies due to the fact that this rare malformation is more mortal by adding this risk and the increasing number of Trap sequence cases [14] today. A healthy twin without any complications at birth was delivered by cesarean section.

References

1. Akercan et al. (2009). ''Trap (twin reversed arterial perfusion) sekansı''. Ege Tıp Dergisi, 48 (2): 123-126 http://egetipdergisi.com.tr/tr/download/article-file/350384

2. Etit et al. (2004). '' TRAP (Twin Reversed Arterial Perfusion) Sendromu''. Perinatoloji Dergisi Cilt:12, Sayı:1 https://www.perinataldergi.com/Files/Archive/tr-TR/Articles/PD-2004012109.pdf

3. Buyukkaya A, Guven T, Buyukkaya R. Twin Reversed Arterial Perfusion (TRAP) Sequence; Characteristic Gray-Scale and Doppler Ultrasonography Findings. Iranian Journal of Radiology https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4632132/

4. Guven F, Kantarcı F. Çoğul gebeliklerde radyoloji. Türk radyoloji seminerleri Trd Sem 2017; 5: 332-45. https://turkradyolojiseminerleri.org/content/files/sayilar/15/buyuk/332-45.pdf

5. Hecher K, Ville Y, Nicolaides KH. Color Doppler ultrasonography in the identification of communicating vessels in twintwin transfusion syndrome and acardiac twins. J Ultrasound Med 1995; 14:37-40

6. Weisz B, Peltz R, Chayen B, Oren M, Zalel Y, Achiron R, et al. Tailored management of twin reversed arterial perfusion (TRAP) sequence. Ultrasound Obstet Gynecol 2004; 23:451-5. https://www.uptodate.com/contents/diagnosis-and-management-of-twin-reversed-arterial-perfusion-trap-sequence?search=trap%20sequence&source=search_result&selectedTitle=1~8&usage_type=default&display_rank=1

7. Das K. Acardiacus anceps. J Obstet Gynaecol Br Emp 1902; 2: 34 1-35.5.

8. Gabbe ST (2012) Obstetrics: normal and problem pregnancies. (6th edn), Sanders an imprint of Elsevier, USA

9. Wong AE, Sepulveda W. Acardiac anomaly: current issues in prenatal assessment and treatment. Prenat Diagn 2005;25: 796e806.

10. Tan TY, Sepulveda W. Acardiac twin: A systematic review of minimally invasive treatment modalities. Ultrasound Obstet Gynecol 2003; 22:409-19.

11. R. C. Prameela et al. (2014). A Rare Case of Acardiac Twin: A Case Report. Int J Sci Stud 2014;2(8):254-257 https://www.ijss-sn.com/uploads/2/0/1/5/20153321/ijss_nov_cr19.pdf

12. Indian Journal of Radiology Imaging 2009, Case series: TRAP sequence https://www.thieme-connect.com/products/ejournals/abstract/10.4103/0971-3026.45352

13. Enesia Mudenha Ziki, Zvavahera Mike Chirenje, Mugove Gerald Madziyire. A case of twin reversed arterial perfusion (TRAP) sequence managed conservatively. Department of Obstetrics and Gynaecology, University of Zimbabwe, College of Health Sciences, Harare, Zimbabwe.

14. Martin J. C. van Gemert et al. (2015). Clinical and Molecular Teratology, Cilt:103, Sayı:7, Sayfa 641-642 [14 Haziran 2015] https://onlinelibrary.wiley.com/doi/10.1002/bdra.23405