

# POOR LONG-TERM OUTCOME IN A SURVIVOR PRESENTING WITH THE TWIN REVERSED ARTERIAL PERFUSION SEQUENCE *IN UTERO*: A CASE REPORT

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The twin reversed arterial perfusion (TRAP) sequence is a very rare complication of multiple gestations and is associated with a high mortality rate, reaching more than 50% in pump twins. The four common complications are preterm labor, polyhydramnios, fetal congestive heart failure, and fetal death of the pump twin *in utero*. Prenatal diagnosis during early pregnancy is possible using detailed ultrasonographic examination. Therapies, including conservative treatment and invasive procedures, are directed toward achieving optimal maintenance of pump twins based on clinical presentation. Risk factors for pump twin mortality include a high twin-to-twin weight ratio, acardiacus anceps, low umbilical artery pulsatility index, and a rapid growth rate in the acardiac twin. Herein, we present a case of TRAP sequence in a patient who underwent conservative treatment and had a poor neurologic outcome during long-term follow-up. Although the experience is still limited, early diagnosis of TRAP sequence and more aggressive treatment, instead of an expectant approach, might be a better option.

**Key Words:** acardiac twin, monochorionic twin, twin reversed arterial perfusion sequence  
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Anomalous reversal of umbilical arterial flow, in which umbilical arterial blood flows from the pump twin to the acardiac twin via a placental arterio-arterial anastomosis, is called twin reversed arterial perfusion (TRAP) sequence [1]. It is a very rare complication of multiple gestations, with an incidence of one in every 35,000 pregnancies, and is observed in 1% of monochorionic twin pairs [2,3]. Most acardiac fetuses are found in twin pregnancies, with only 8% in triplets [3]. Depending on the variable stages of developmental disruption, acardiac anomalies are divided into four types, including acardiacus anceps, acardiacus acephalus, acardiacus acormus, and acardiacus amorphus [4]. The four common complications are preterm labor,

polyhydramnios, fetal congestive heart failure, and fetal death of the pump twin *in utero* [2,3]. However, information regarding the pump twin, including long-term outcome and prognostic factors, is relatively limited. Herein, we present a case of a survivor pump twin with TRAP sequence who had a poor neurologic outcome on long-term follow-up.

## CASE PRESENTATION

A 28-year-old woman pregnant with twins had one fetal demise, diagnosed on ultrasonography by a local practitioner at 13 weeks of gestation. She was referred to our hospital at 30 weeks of gestation with pregnancy-induced hypertension. Sonography showed one twin with normal anatomy but symmetric growth restriction, and the other with acardia and hydrops fetalis. The patient underwent regular weekly follow-up at our clinic, and

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during that period, the pump twin showed a pulsatility index in the umbilical arteries of 1.0 and no evidence of fetal heart failure on ultrasonography. The fetal monitor revealed a reactive pattern in the fetal heart rate.

The patient underwent Caesarean section at 35 weeks of gestation due to malpresentation of the twins, severe polyhydramnios, and pre-eclampsia. A male baby, with a birth weight of 1,450 g and Apgar scores of 6 (1 min) and 7 (5 min), was admitted to the neonatal intensive care unit (NICU) due to prematurity, very low birth weight, and respiratory distress syndrome. His body weight and length, head circumference, and abdominal circumference were below the 10<sup>th</sup> percentile of the normal growth curve. The acardiac twin weighed 4,850 g and had an edematous appearance. It had a partially developed head with no separation of the eyelids, anophthalmia, and no ears. It also had a deformed upper trunk, hypoplastic upper limbs, and an abdominal wall defect, but had a relatively well developed lower trunk with an ambiguous genital organ (Figures 1 and 2). There was a high weight ratio of the acardiac twin to the pump twin (3.34). The monochorionic, monoamniotic placenta weighed 950 g.

The pump twin had a normal cranial ultrasound at 2, 9, and 36 days of age. Ventriculomegaly and a cyst on the right germinal matrix were detected by cerebral ultrasonography at 3 months of age, and computerized tomography (CT) scan showed mild atrophy of the brain. Follow-up ultrasonography disclosed ventriculomegaly, a subependymal cyst, increased subdural space, cortical atrophy, and hydrocephalus during the first year of life. A repeat CT scan at 18 months of age confirmed the cortical atrophy. Neurologic hearing loss was diagnosed during detailed neurologic examinations and cerebral palsy was confirmed using the standard criteria, including persistent abnormality of movement and posture resulting in impaired function, caused by a non-progressive lesion on the immature brain. In addition, there was severe mental and motor developmental delay, as assessed by the Bayley Scales of Infant Development (Table) at 3 years of age. The survivor pump twin was followed up in the Pediatric Department of our hospital until he reached 3 years of age.

## DISCUSSION

The TRAP sequence is associated with a high mortality rate, reaching more than 50% in pump fetuses [2,5]. The high mortality rate mostly results from fetal cardiac failure,

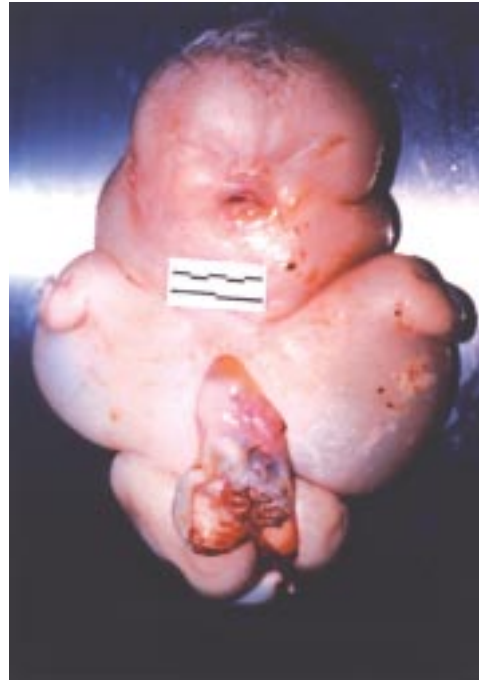


Figure 1. Frontal view of the acardiac fetus.



Figure 2. Posterior view of the acardiac fetus.

hydrops fetalis, hydramnios, and prematurity [2,3]. Ultrasonography and fetal echocardiography are valuable in the prenatal diagnosis of acardiac twins in the early second trimester [6–8]. The risk factors for mortality of the

**Table.** Bayley Scales of Infant Development in the pump twin at 3 years old

Scale	Raw score	Index	DA	Classification
Mental	*	*	14 mo	Significant delay
Motor	*	*	10 mo	Significant delay
BRS	Raw score	Percentile		Classification
Attention/Arousal	—	—		—
Orientation/Engagement	30	15		Questionable
Emotional regulation	34	16		Questionable
Motor quality	16	1		Non-optimal
Total score	81	2		Non-optimal

DA = developmental age; BRS = behavior rating scale. \*Below the lowest limit of the normal curve.

pump twin include a high twin-to-twin weight ratio (> 70%) [2], acardiacus anceps [3], a low pulsatility index in the umbilical arteries, and a rapid growth rate in the acardiac fetus [5].

Conservative treatment, such as close observation and the symptomatic treatment of hydramnios and fetal heart failure with amniocentesis, indomethacin, and digitalis, incurs a minor risk and seldom improves perinatal outcome [8]. On the other hand, invasive treatments, including hysterotomy with selective delivery of the acardiac fetus [9], interrupting vascular communications by endoscopic laser coagulation or ligation of the acardiac twin's umbilical cord [6,8], ultrasound-guided embolization of the acardiac twin's umbilical artery with absolute alcohol, or the use of thrombogenic coils, have been reported [10]. However, these procedures may lead to many complications, such as fetal death, preterm labor, premature rupture of membranes, placental abruption, intrauterine infection, hemorrhage, and maternal pulmonary edema [6,8]. The overall mortality rate of the pump twin after fetal surgery has been reported as 13.6% [6].

We chose to use careful conservative treatment, including a series of ultrasonography examinations and fetal monitors in the prenatal period. Only intrauterine growth restriction, polyhydramnios, and a huge acardiac twin were observed in the series of ultrasonography images, and no ominous fetal heart rate patterns were shown on the fetal monitor. However, severe growth restriction and poor long-term neurologic outcome, including cerebral palsy, mental and motor developmental delay, and neurologic hearing loss, were discovered later. Ahn et al reported that fetuses with central nervous system injuries in early pregnancy can

show normal intrapartum fetal heart rate patterns during labor [11]. These observations suggest that a series of ultrasonography images and fetal heart rate monitors are not the optimal tools to aid in making decisions to prevent poor long-term neurologic outcomes in these cases. It is difficult to reach firm conclusions based on such limited experience. However, an aggressive approach, with the intent of interrupting vascular communication between the twins, or using elective termination, might be a better option, based on the high mortality and morbidity in the TRAP sequence.

In summary, there are no reports of the long-term follow-up of survivor pump twins in the TRAP sequence, who may suffer a poor neurologic outcome. Although experience is still limited, early diagnosis of TRAP sequence and more aggressive treatment or elective termination might be a better option than an expectant approach. Detailed ultrasound study, which may provide a chance for early diagnosis and early management, is important in prenatal diagnosis during early pregnancy. A further investigation into the long-term follow-up of survivor pump twins with TRAP is required to confirm our observation and to standardize treatment protocol.

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