



## Case report

# Successful intrauterine treatment with radiofrequency ablation in twin reversed arterial perfusion (TRAP) sequence: a case report and review of literature

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### ABSTRACT

Twin reversed arterial perfusion (TRAP) sequence is a rare complication of monochorionic multifetal pregnancies, and affects approximately 1% of monochorionic twins with a prevalence of 1/35000 pregnancies. This condition belongs to the broad spectrum of twin-to-twin transfusion syndrome and is presumed to result from the fused placentation of monochorionic twins, in which vascular anastomoses arise between the 'pump' twin and that of the 'perfused' acardiac twin. The severity of this syndrome depends upon the type of anastomoses and the timing of their establishment. Here we present a case of acardiac twin pregnancy successfully treated by obliteration of blood supply to the acardiac twin using radiofrequency ablation and review the current concepts in the pathogenesis, diagnosis and treatment of TRAP sequence.

**Key words:** Twin reversed arterial perfusion syndrome, acardiac twin, acardiac malformation, monochorionic pregnancy, treatment, radiofrequency.

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## Introduction

Twin reversed arterial perfusion (TRAP), also known as acardiac malformation or acardiac twin, sequence is a rare complication of monochorionic multifetal pregnancies that affects approximately 1% of monochorionic twins with an incidence of 1/35000 pregnancies [1-3].

This condition belongs to the broad spectrum of twin-to-twin transfusion syndrome and is presumed to result from the abnormal placentation of monochorionic twins in which vascular anastomoses arise between the arterial circulation of the hemodynamically larger 'pump' twin and that of the recipient 'perfused' twin. The severity of this syndrome depends upon the type of anastomoses and the timing of their establishment [4].

Circulation is accomplished by the heart of the pump twin who is at risk of heart failure. Since growth of the acardiac twin depends on blood supply by the pump twin, growth of the acardiac twin cause hyperdynamic circulation causing heart failure, fetal hydrops, and polyhydramnios in the pump twin [5]. The perfused twin may display severe and sometimes lethal anomalies, such as acardia and acephalus. The mortality risk of the pump twin has been

reported to be 50- 75% [6-7]. Early diagnosis is essential for proper management to save pump twin [4].

The natural history of TRAP sequence may vary from occasional patient reaching term to severe cardiac failure that causes intrauterine death of pump twin. In severe cases salvage invasive procedures were used to disconnect twins [8].

Here we present a case of acardiac twin pregnancy in which obliteration of blood supply to the acardiac twin using radiofrequency ablation (RFA) successfully salvaged the pump twin.

## Case

A 27-year-old woman, gravida 2 para 0 abortus 1 was referred to our tertiary referral center at 24 weeks' of gestation because of a twin pregnancy complicated with signs of skin edema, ascites and extremity abnormalities at the acardiac twin. The diagnosis of twin pregnancy was established at 8 weeks' gestation that one of the twins with absent cardiac activity.

The patient was counseled regarding the high risk of perinatal morbidity and mortality associated with the con-



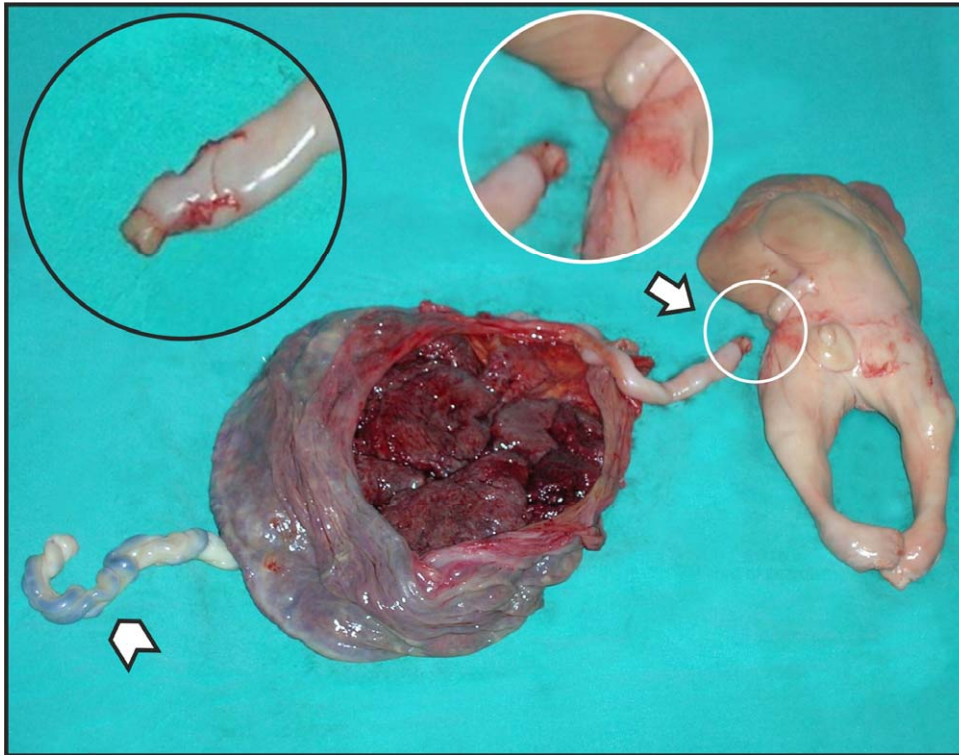


Figure 1. Radiofrequency ablated umbilical cord supplying male acardiac twin (arrow, insets) and umbilical cord supplying pump twin (arrowhead).

dition and was followed for correct diagnosis of the abnormality. Subsequent ultrasonographic examinations revealed a monochorionic-diamniotic twin pregnancy. Also absence of head and upper extremities, pes equinovarus in lower extremities, and ascites with large cystic spaces without cardiac activity were noted. Pulsed Doppler imaging showed reversed flow through the single umbilical artery of the acardiac fetus, establishing the diagnosis of TRAP sequence. The pump twin's umbilical arterial S/D ratio was 2.8, resistance index was 0.61, and pulsatility index was 0.94. Intrauterine ablation of umbilical cord of acardiac twin was decided on development of cardiomegaly in the pump fetus at 27 weeks of gestation.

Pregnancy was monitored weekly until 33 weeks' without any signs of heart failure and polyhydramnios. Patient was presented with preterm rupture of membranes in the same week. After 10 hours of admission caesarean section was performed with progress of labor. Two male fetuses were delivered weighing 1760 g (pump twin) and 370 g (acardiac twin).

## Discussion

The acardius fetal malformation, occurring once in every 35000 births, was first described in 1533 [9]. The phenotype of the acardiac twin is classified according to the mode of development and growth. The TRAP sequence leads mainly to the '*acardius acephalus*' phenotype (no cranial or thoracic structures and usually malformed upper extremities). However, three other phenotypes '*acardius anceps*' (some cranial structures), '*acardius amorphous*'

(the most malformed structure with only a shapeless mass of tissue without a recognizable human part) and '*acardius acormus*' phenotypes have also been described. The rarest phenotype is '*acardius acormus*' that exhibits cranial elements but no body structure [8].

Although exact cause is not known, there are two theories about development of acardiac fetus; First, fetal fetal anomaly develop secondary to cardiac malformation [10-11]. According to second theory TRAP sequence is a part of the spectrum of twin-to-twin transfusion syndrome (TTTS). Artery-to-artery or vein-to-vein anastomoses of monochorial placenta are thought to cause reverse flow from pump twin to acardiac twin. Since blood reaches the lower part of trunk, secondary atrophy of the upper parts including heart develops [12-13]. Benirschke, proposed the cause of placental abnormality is the result of segregation of uneven number of cells during monozygotic twinning [14]. Irrespective of the primary cause, observation of fetal cardiac activity in TRAP sequence cases in early pregnancy supports second theory. Early *in utero* death of a twin might result in reverse perfusion through artery-to-artery or vein-to-vein anastomoses [7]. In accordance with this theory, TRAP sequence is rarely associated with chromosomal abnormalities [9].

Diagnosis of TRAP sequence continues to be problematic. Frequently, the acardiac fetus is mistaken for a twin who has died or an anencephalic fetus. The ultrasonographic features of absent head and trunk regions and increased soft tissue in the body aid in correct diagnosis. In rare cases cardiac activity might be seen due to rudimen-

tary cardiac structure [15]. Recently, pulsed Doppler examination has been used to show reversed flow through the umbilical artery at the acardiac twin [15-16]. Typically, perfusion through single umbilical artery is observed in Doppler examination [15].

The recognition of this abnormality in the antenatal period becomes important for the proper management [15]. The goal of perinatal management of acardiac twin pregnancy is to deliver a mature pump twin without heart failure or fetal hydrops. The clinical dilemma regarding the decision to intervene and the exact time of this intervention is still unsolved, exact criteria, such a stage-related therapy have not been defined [8]. Since there are limitations of conservative management, several methods of intrauterine treatment of an acardiac twin pregnancy have been proposed [17-18], medical or invasive procedures, with the common purpose to relieve hyperdynamic circulation of the pump twin by cessation of blood perfusion to the acardiac twin.

Medical treatment with digoxin and indomethacin which are associated with limited success that may be appropriate for mild cases.

Although all proposed methods are effective, there are marked differences in technical difficulty and invasive potential. The methods of vasooclusion are varied and reports have included the use of alcohol [17], RFA [18], umbilical cord ligation [19-20], bipolar [21] and monopolar [22] ultrasound-guided thermo coagulation, coils [23], and fetoscopic [24] and ultrasound-guided [25-26] laser coagulation, due to the limited number of cases, superiority of one method over the other is difficult to assess.

Although, there is a classification system for the management of acardiac malformation [27], evidence from other mono chorionic twins with TTTS showed that interventions at an advanced gestational age and when signs of cardiac decompensation were already present correlated with mental delay and unfavorable neurological outcome [15]. Deoxygenated blood from acardiac twin passes directly to the pump twin causes chronic hypoxia irrespective of the presence of cardiac decompensation [21]. Recently it was reported that selective reduction of the acardiac twin with RFA was found to be minimally invasive and effective for intrauterine treatment of acardiac twin pregnancy [18].

The prognostic factors for the pump twin is based on the retrospective study of Moore et al [28], who summarized 49 cases of acardiac twins. In this study perinatal mortality rate was %55. Review of the literature mortality rates of the pump twin with fetal surgery occurred is 13.6% in comparison with 50% mortality with expectant management and weight ratio of 0.7 and above is associated with poor prognosis [29]. Prognosis of triplet pregnancies complicated with TRAP sequence is very poor irrespective of treatment [30].

Some physicians believe that conservative treatment should be considered unless there are signs of rapid growth of the recipient twin or a dimension ratio >0.5 were noticed. In mono chorionic pregnancies complicated with TTTS endoscopic ablation of anastomoses has become the only invasive fetal therapy proven by randomized controlled trial to improve perinatal survival [31]. According

to this evidence most physicians believe prophylactic cord occlusion should be offered to all cases of TRAP sequence.

Endoscopic laser coagulation and sonography-guided umbilical cord ligation are the preferred modalities of treatment before and after 24 weeks of gestation, respectively.

In the present case, obliteration of blood perfusion by the acardiac twin with RFA was successful without technical difficulty. We recommend that therapeutic intervention must be introduced before the onset of intrauterine fetal complications as long as there is clear data about the long term neurological sequela of the pump fetuses.

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