

Acardius Acephalus: A Case Report and Review of Literature

H Nachchat¹, M Chigri¹, N Sahli^{2*}, F Abdeddine¹, S Amrani¹, M Youssfi¹ and S Bargach¹

¹Department of Gynecology and Obstetrics, Maternity Souissi, University Mohammed V, Rabat, Morocco

²Department of Radiation Oncology, Regional Center of Oncology of Tangier, Morocco

***Corresponding Author:** N Sahli, Department of Gynecology and Obstetrics, Maternity Souissi, University Mohammed V, Rabat, Morocco.

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Abstract

Background: A cardiac-acephalic or commonly Twin Reversed Arterial Perfusion (TRAP) sequence, is a rare complication unique to monozygotic twin pregnancies.

We are reporting an interesting and rare case of an acardiac-acephalic twin with favorable evolution of his pump twin.

Case Presentation: This case report case involved a 31-year-old G2P3, she had 24 weeks gestation when she presented to our department. On obstetric ultrasonography, she had an intrauterine monozygotic monoamniotic twin pregnancy with the presence of a normal fetus pump and a acardiac acephalus fetus. The doppler study showed a retrograde arterial flow to the acardiac fetus.

Weekly ultrasound monitoring was without particularity and cesarean was performed at 36 weeks of gestation with a an alive, male baby without any external abnormalities and the acardiac acephalic twin.

Conclusion: We present a case report of acardius acephalic twin to highlight the importance of early diagnosis which can help to plan the monitoring and treatment for improving the survival of the pump twin.

Keywords: *Acardiac-Acephalic; Twin; Pregnancy; Complications*

Abbreviation

TRAP: Twin Reversed Arterial Perfusion

Introduction

Acardiac-acephalic or commonly Twin Reversed Arterial Perfusion (TRAP) sequence, is a rare complication unique to monozygotic twin pregnancies in which a twin with an absent or rudimentary heart ("acardiac twin" or "parasitic twin") is perfused by its co-twin ("pump twin") via arterial anastomoses on the placental surface.

The upper body and head of the acardiac twin are often poorly developed and may be absent.

The pump twin is at risk of developing heart failure and other complications that may lead to preterm birth or demise, which are common without intervention [1].

We present a case report of acardius acephalic to highlight the importance of early diagnosis which can help to plan the treatment for improving the survival of the pump twin.

Case Report

A 31-year-old female G2P3 presented to our unit at 24 weeks of gestation for a follow-up of her current pregnancy. Her medical and family history were without particularity.

Her clinical examination revealed a uterus of 26 week's size. On obstetric ultrasonography, she had an intrauterine monochorionic monoamniotic twin pregnancy with the presence of a fetus pump with normal heart activity (Figure 1) and a acardiac acephalus fetus (Figure 2), also we noted a single placenta and a thin membrane separating the two fetuses.

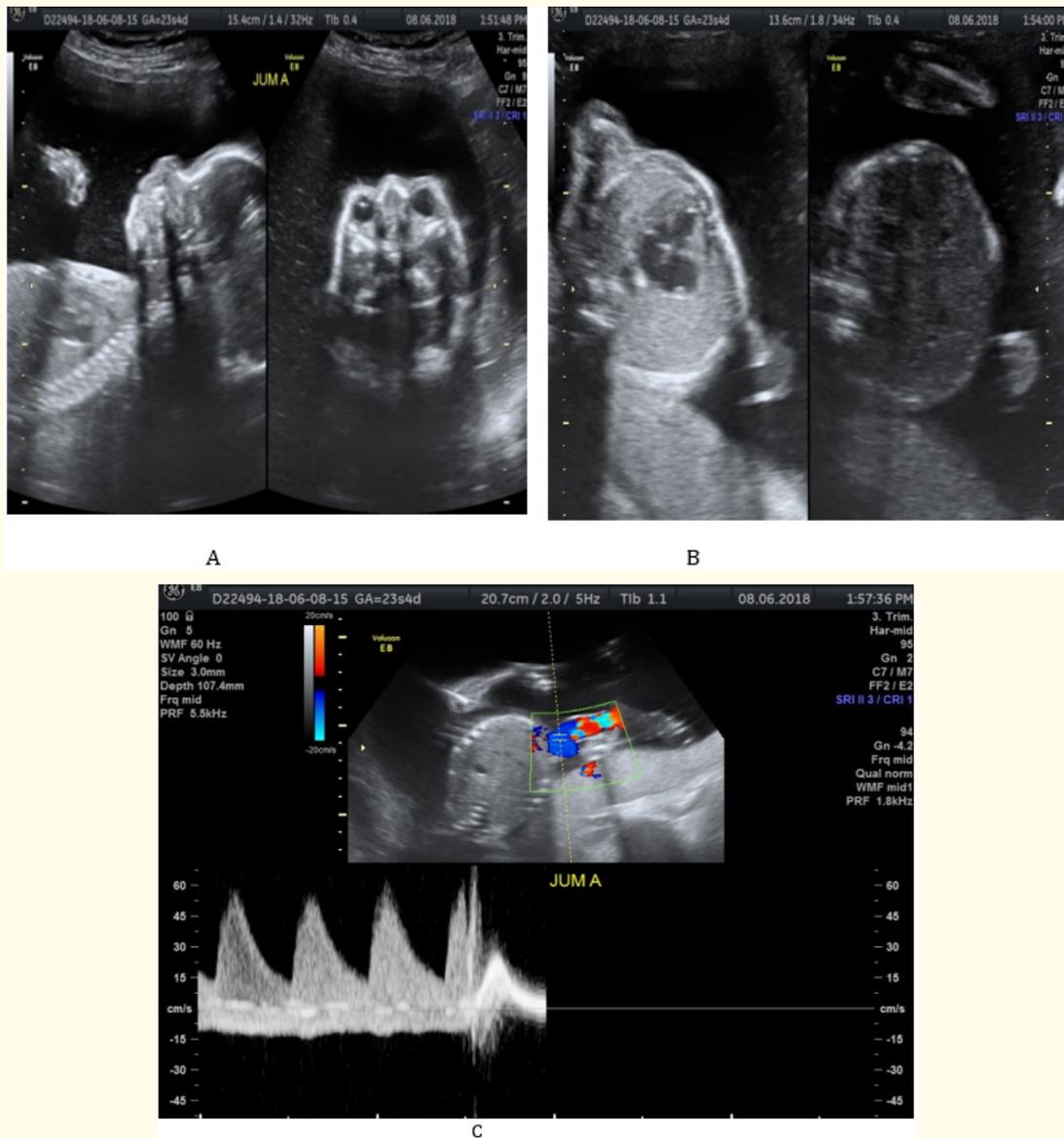


Figure 1: Pump Fetus A. Normal cephalic pole B. Normal cardiac cavities C. Doppler of the umbilical cord.

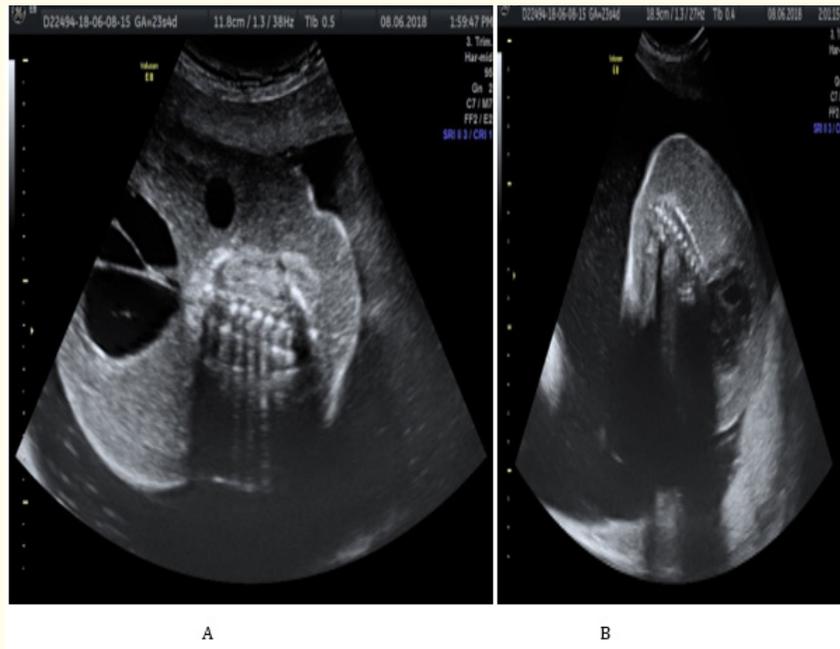
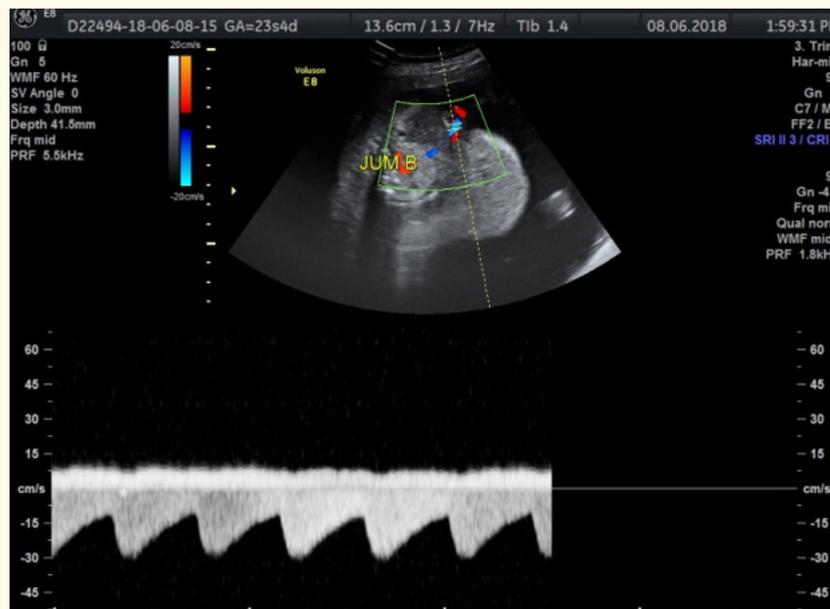


Figure 2: Acardiac Fetus. A. Absence of cephalic pole and upper limbs. B. downer limbs.

The doppler study showed a retrograde arterial flow to the acardiac fetus (Figure 3).



Figures 3: Retrograde arterial flow to the acardiac fetus.

We decided a close monitoring with weekly obstetric ultrasound scans.

At 36 week of gestation, patient came with active labor, a cesarean was realized.

The first twin - (Figure 3) an alive, male baby of 2100g without any external abnormalities and the acardiac twin (Figure 4) of 3200g who had a well-developed lower limbs and the lower trunk which was normal. It had absent development of cephalic pole, heart and upper limbs.



Figures 4: Pump twin and acardiac acephalic twin.

The pump twin was kept under supervision in the pediatric intensive care unit.

Currently, he has 8 months old and the last pediatric examination was without any particularity.

Discussion

TRAP sequence is a rare complication that occur in 2.6 percent of monochorionic twin pregnancies and 1 in 9500 to 11,000 pregnancies [2].

The exact pathogenesis of TRAP sequence is unknown [3], In one theory, abnormal arterio-arterial anastomoses in early embryogenesis give a deoxygenated blood to an acardiac fetus with no direct placental perfusion. In the other theory, an acardiac twin with defective early cardiac embryogenesis produces low systemic arterial pressure, permitting retrograde flow of blood from its pump co-twin. Regardless of the exact cause, the net result is acardiac twin dependence on a pump twin for circulatory support from early in the first trimester. The preferential vascular distribution at the lower part of the fetal body would explain the lack of thoracic development, upper limbs and/or cephalic pole.

Acardiac twin is classified according to the degree of cephalic and truncal maldevelopment [3].

1. Acardius-acephalus with no cephalic structures present. It is most common variety. This is the type seen in the present case.
2. Acardius-anceps where some cranial structure and neural tissue or brain tissue is present. The body and extremities are also developed. It is highly developed form.
3. Acardius-acormus with cephalic structure, but no truncal structures are present. The umbilical cord is attached to the head. It is rarest form of the acardia.
4. Acardius amorphous with no distinguishable cephalic or truncal structure.

Prenatal diagnosis can be made as early as the first trimester [4]. The three ultrasound findings are:

- Monochorionic multiple gestation
- One fetus with either absent cardiac activity or a rudimentary pump structure
- Doppler study demonstrating retrograde arterial flow to the acardiac fetus which is a pathognomonic finding.

The differential diagnosis of TRAP sequence includes a severely anomalous co-twin and co-twin demise. An anomalous twin unrelated to TRAP can be excluded by doppler assessment of the arterial flow pattern to that twin: it will be reversed in TRAP but normal in a purely anomalous twin. A demised twin will not have limb movement, will not have retrograde arterial flow observed on color Doppler imaging and will not continue to grow on serial ultrasound examinations, whereas an acardiac twin may demonstrate some or all of these findings.

Pregnancies with acardiac twins should be carefully assessed based on the prognostic factors. One or more poor prognostic factors raise the risk of pump twin mortality [5-8]. The principal prognostic factors are pump twin hydrops or heart failure, polyhydramnios and the ratio of the weight of the acardiac twin to that of the pump twin, when exceeded 0.70, the risks of cardiac failure, preterm delivery and polyhydramnios were 30, 90 and 40 percent, respectively, versus 10, 75 and 30 percent, respectively, when this ratio was below 0.70 [8].

Following diagnosis of TRAP sequence, patients should be referred to an experienced fetal therapy center; strategies include conservative management or acardiac twin cord occlusion.

Conservative management [9] is advised in patients if the acardiac twin is small and with no signs of cardiovascular involvement of pump twin or in cases with spontaneous closure of shunt due to tight umbilical cord wrapping. The goal is to monitor the pump twin for developing heart failure, polyhydramnios, premature birth and cord entanglement. Regular follow-up with serial sonographic examinations are necessary, but there are no robust data to support any specific evidence-based surveillance strategy for TRAP monitoring.

In our case, the ultrasound surveillance was performed every week.

Invasive treatment [10] is required when pump twin is having cardiac failure to improve the perinatal outcome, treatment for TRAP sequence is entirely focused on optimizing pump twin outcomes.

In a meta-analysis including 26 retrospective studies of TRAP sequence, intervention by cord occlusion or ablation was associated with a greater chance of live birth than conservative management (odds of miscarriage/fetal death with conservative management versus intervention: OR 2.22, 95% CI 1.23 - 4.01), with the greatest benefit in pregnancies with poor prognostic factors (odds of miscarriage/fetal death with conservative management versus intervention: OR 8.58, 95% CI 1.47 - 49.96) [10].

The three contemporary modalities for intrafetal cord occlusion therapy for management of TRAP sequence are RFA, bipolar cord coagulation and intrafetal (or interstitial) laser coagulation [11-14].

We report this case to highlight the importance of early diagnostic of such pregnancy and close monitoring by ultrasound.

Conclusion

Acardiac-acephalic twin is a rare complication of monochorionic twin gestation. The present case reported an acardiac-acephalic twin. This is the most common form of all acardiac twins. Early diagnosis and ultrasound monitoring may help to plan the treatment for improving the survival of the pump twin.

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Competing Interests

The authors declare that they have no competing interests. Consent for publication Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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