



IJCRR

Section: Healthcare

ISI Impact Factor  
(2019-20): 1.628

IC Value (2019): 90.81

SJIF (2020) = 7.893



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# Acardiac Acephalic Fetus: A Rare Complication of Twin Pregnancy, A Case Report and Review of the Literature

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## ABSTRACT

**Introduction:** The acardiac acephalic fetus or twin reversed arterial perfusion (TRAP) sequence, is a rare complication specific to monochorionic twin pregnancy. It is characterized by the development of arterio-arterial and veno-venous anastomosis leading to one of the twins' predominance.

**Objective:** Our objectives are to describe this pathology and its etiopathogenic factors and emphasize the need for an early diagnosis and adapted therapeutic management to improve the prognosis of the pump twin.

**Clinical case study:** We report the case of an acardiac fetus from a twin pregnancy of a 32 years old-patient, low socio-economic level. She was admitted at 40 weeks of amenorrhea according to the date of her last period, in the expulsion phase, giving vaginal birth to a first twin of normal appearance and then an acardiac fetus weighing 1000g.

**Presentation:** In our study, the prevalence rate of TRAP is estimated at 1 in 44,000 births. Its diagnosis is based on Doppler ultrasound from the 1st trimester which allows visualizing the reversal of vascular flow in the acardiac recipient fetus and during the 2nd trimester, the MRI allows to confirm the diagnosis. In our case, the pregnancy was poorly followed, no ultrasound examination was performed and the diagnosis of the acardiac fetus was made at the time of delivery.

**Conclusion:** An acardiac fetus is never viable. However, several complications can affect the transfusion twin, which underlines the importance of an early prenatal diagnosis allowing adequate therapeutic management to improve the prognosis of the healthy twin.

**Key Words:** Acardiac acephalic fetus, Twin pregnancy, Antenatal diagnosis, Complication, Prognosis

## INTRODUCTION

Described for the first time by Benedetti in 1533, the acardiac acephalic fetus also named TRAP (twin-reversed arterial perfusion) sequence or acardius acarnius or acardiac mass is a rare and severe complication of monochorionic twin pregnancies.<sup>1,2</sup> Its prevalence is estimated at 1/35000 births and affects 1% of monozygous twin pregnancies, the risk of recurrence is estimated at 1/10000.<sup>3,4</sup>

The TRAP sequence is characterized by the development of arterial-arterial and venous anastomosis between the two cord insertions leading to an inverted infusion via the umbilical artery of a twin and thus the predominance of one of the twins.<sup>3</sup> Hemodynamic abnormalities are the cause on the one hand of the lack of development of cardiac structures with

morphological abnormalities in the transfused fetus, and the other hand of the increase of myocardial work of the healthy fetus or pump.

Perinatal complications of twin pregnancy with an acardiac fetus are dominated by heart failure of the healthy co-twin, polyhydramnios and prematurity. The prognosis of the healthy twin is fraught with high mortality with an estimated risk of perinatal death of 50-70%.<sup>5</sup> The management of twin pregnancies with acardiac fetuses is based on the assessment of the prognosis of the healthy twin by early antenatal ultrasound and a therapeutic attitude of which aim is to improve the heart function of the pump fetus by prescribing digitalis or selective interruption of the circulation of the acardiac fetus. Our objectives are to describe this pathology and its etiopathogenic factors and emphasize the need for an early

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ISSN: 2231-2196 (Print)

ISSN: 0975-5241 (Online)

Received: 17.10.2020

Revised: 05.12.2020

Accepted: 03.02.2021

Published: 04.06.2021

diagnosis and adapted therapeutic management to improve the prognosis of the pump twin.

## CLINICAL CASE STUDY

This is a 32-year-old pregnant woman, married, housewife, low socio-economic level, group 0+, second primitive gestation. The pregnancy was poorly followed with the realization of a single syphilitic serology that came back negative. The patient reports the notion of taking fenugreek and plants during the first trimester of pregnancy and the history of diabetes in the family, without the notion of inbreeding. Furthermore, she did not receive any iron or folic acid supplementation.

Admitted at 40 weeks of amenorrhea (WA), in the expulsion phase with a ruptured water pocket, his general and clinical examination was without particularity. The delivery took place by natural childbirth giving birth to a first twin who had no abnormalities, then a dystocic extraction of an acardiac fetus in the form of an amorphous, oedematized mass, weighing 1000g, with a single lower limb, having 4 toes, and a trunk, without heart activity and head or upper limbs, with the presence of a short umbilical cord (**Figure: 1**). The deliverance was rapid with the expulsion of a single placenta evoking a monochorionic monoamniotic twin pregnancy.

## DISCUSSION

Multiple pregnancies are associated with a high risk of mortality and morbidity due to prematurity, cardiovascular and neurological complications. This risk is 3 to 5 times higher in the case of monochorionic twin pregnancy than in bichorionic pregnancies, this is probably due to fetoplacental vascular anastomosis.

In the literature, the acardiac acephalic fetus is a rare complication, affecting one in 35,000 pregnancies, or 1% of all monozygous pregnancies.<sup>3,4</sup> In our study the prevalence rate is estimated at 1 in 44,000 births. TRAP is more common in nulliparous women as in our case, and it is three times higher in monozygous triplets than in twins, with an incidence of chromosomal abnormalities of 9%.<sup>5</sup>

The etiopathogenia of this entity is explained by several theories. Some say that it is the presence of primary cardiac dysmorphogenesis that causes the placental vascular anastomosis necessary for the development of the acardiac fetus, others think that it is the opposite; it is the presence of inverted vascular flow that will be the cause of heart atrophy. Nowadays, the association of the two theories with circulatory insufficiency of early-onset (8-12WA) and the occurrence of venous and arterial-arterial placental anastomosis is accepted. As a result, the acardiac fetus is a true parasite that receives blood from the pumping twin via the

umbilical arteries in retrograde flow via anastomosis of the arterial-arterial and venous-venous plate.<sup>6,7</sup> This oxygen-poor blood is responsible for the lack of development of the head, heart, and upper limbs of the acardiac twin, hence the twin-reversed arterial perfusion (TRAP) sequence name.<sup>8</sup>

This phenomenon of twin reversed arterial perfusion was observed by U. Gembruch et al. on doppler ultrasound after the death of the transfuser twin at 25WA occurring as part of a transfuser-transfused syndrome of a biamniotic monochorionic twin pregnancy, without any congenital defects or heart defects detected in the two twins. This observation lasted 12 hours and then disappeared without repercussion on the living twin.<sup>9</sup>

The role of prenatal ultrasound performed between 6 and 20 weeks old in the diagnosis of congenital malformations, in particular cardiac malformations, has been well evaluated and demonstrated by Lekshmi et al<sup>10</sup>. The diagnosis of the acardiac acephalic fetus is based on Doppler ultrasound from the 1st trimester which allows visualizing the reversal of vascular flow in the acardiac recipient fetus and during the 2nd trimester, the MRI allows to confirm the diagnosis. In our case, the pregnancy was poorly followed, no ultrasound examination was performed and the diagnosis of the acardiac fetus was made at the time of delivery. It was an acardius acephalus according to the classification of Napolitani and Schreiber (1960) which distinguishes 4 groups.<sup>11</sup>

- Acardius anceps: The head is poorly formed
- Acardius acephalus: The head is absent
- Acardius acormus: The presence of the single head
- Acardius amorphous: Unrecognizable anatomy

Another classification differentiates :

- Cases where the heart is not completely formed: Hemiocardius
- cases where the heart is absent: Holoacardius.

This is the second complication of vascular anastomosis of monochorionic placentas after transfuser-transfused syndrome,<sup>12</sup> with a mortality rate for the healthy twin “pump” estimated at 50-70%.<sup>5</sup> The risk of perinatal mortality of the pump twin may be due to congestive heart failure, polyhydramnios, and premature delivery. The rate of complication is proportional to the ratio of the weight of the parasitic twin to that of the pump twin.<sup>7,13</sup> Thus, the mortality rate is 64% when the size of the acardiac fetus is greater than 50% of the size of the pump. When the acardiac twin exceeds 70% of the size of the pump twin, premature delivery reaches 90%,<sup>5</sup> polyhydramnios 40%, and heart failure 30%.<sup>8</sup>

The therapeutic management in cases of acardiac fetuses is highly discussed and controversial. However, several authors opt for expectation from the beginning of pregnancy; with weekly monitoring by performing regular ultrasounds to monitor the state of growth, and look for signs

of heart failure in the twin pump.<sup>13</sup> Evolution can be normal without any complication. In case of hydramnios or heart failure in the pump twin, medical treatment is considered with indometacin, amniocentesis to reduce the volume of amniotic fluid and Digoxin to treat congestive heart failure in the pump fetus.

Invasive therapeutic protocols are also proposed such as, stopping infusion of the acardiac twin by percutaneous radio frequency; reliable and effective technique despite the risk of premature membrane rupture according to the study of P. Cabassa et al.<sup>14</sup> Other techniques are used, such as injecting thrombogenic products or sclerosing agents such as alcohol, which results in embolization of the umbilical cord of the parasitic twin.

The ligation of the umbilical cord by thermo coagulation or laser coagulation under endoscopic or ultrasound guidance performed at the beginning of the 2nd quarter (from 18WA), was tested by K. Hecher et al on 60 cases with a success rate of 82%, and a survival rate of 80%, with 67% of cases survived beyond 36WA.<sup>15</sup> Ultrasound-guided thrombosis of an umbilical cord artery and neonatal MRI surgery can be performed if the healthy twin shows signs of failure.<sup>5,8</sup>

## CONCLUSION

In a monochorionic twin pregnancy, the diagnosis of acardiac twins must be suspected and detected early by antenatal Doppler ultrasound in the absence of cardiac structure or activity of one of the twins, and MRI, with regular follow-up for adverse prognosis factors in the healthy fetus so that an intervention can be planned early enough to avoid the onset of serious complications from the pump twin.

## ACKNOWLEDGEMENTS

We would like to thank everyone who has contributed to this study.

Ethical approval and consent to participate

The Ethical Committee of Biological Research, Faculty of Medicine and Pharmacy – Rabat, approved the study, n°: 20/16. The oral consent to participate was obtained from the parents.

## Funding

Not applicable

## Competing interests

The authors declare that they have no competing interests.

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**Figure 1:** Acardiac acephalic fetus with only one lower limb, four toes, one trunk, no cardiac activity, no head or upper limbs.