

Twin-Reversed Arterial Perfusion (TRAP) Sequence: Case Report and Literature Review

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ABSTRACT

Introduction and Importance: The TRAP sequence or cardiac mass is a serious pathology that occurs in monochorionic twin pregnancies, characterised by the absence of development of cardiac structures with a series of malformations in the transfused fetus, which is never viable, and numerous complications affecting the transfused twin.

Case Presentation: We report a case of TRAP sequence in a presumed pregnancy at 7 months.

Discussion: The TRAP sequence is an exceptional and specific complication of monochorionic twin pregnancies, affecting on average one birth in 35,000 and 1% of monozygotic pregnancies. The differential diagnosis includes in utero fetal death of a twin without anomalies, anencephaly associated with cystic hygroma and placental tumour. Management of the TRAP sequence in complicated twin pregnancies is aimed at the survival and compensation of the transfused fetus.

Conclusion: Analysis of this observation highlights the importance of prenatal diagnosis in the prevention of its consequences.

Keywords

TRAP sequence, Acardiac mass, Monochorionic twin pregnancies.

Introduction

The TRAP sequence or cardiac mass is a serious pathology that occurs in monochorionic twin pregnancies and multiplies the risk of this developmental anomaly by four. It has an incidence of 1 in 35,000 pregnancies and is characterised by the presence of a malformed fetus with a rudimentary, non-functioning or absent heart and without direct placental perfusion by arterio-arterial communication from the transfused twin, if this is followed by a double deoxygenated perfusion after perfusion of the transfused twin, communication which then returns to the transfused twin by venovenous communication. The natural history of the disease is

classic, with the transfused twin dying of heart failure in 35-55% of cases. However, there is no evidence regarding the short and long term morbidity and mortality of the transfused twin who is chronically exposed to a lower oxygen saturation.

This poor prognosis is at the origin of the outline of several techniques of invasive intrauterine therapy which have allowed the interruption of the flow towards the acardiac fetus, improving the prognosis of the fetus transfused, without defining the gestational age appropriate for this procedure. The aim of this study is to examine the basic aspects and the current status of this pathology, with priority given to early diagnosis, and the alternatives to prenatal invasive treatment. The impact on quality of life is significant and can be severe.

Case Report

This is a 23-year-old patient, primigravida, with a history of first-degree consanguinity. Admitted to our structure for delivery of a presumed unmonitored pregnancy at 7 months. On admission, the patient was found to be in labour with 9 cm dilated cervix, 8 hours of ruptured membranes and cephalic presentation. Uncollected BCFs. Antenatal ultrasound revealed a twin pregnancy. The biometrics of the first twin were an estimated abdominal circumference of 254.3 mm, corresponding to 32 WA + 3J, and an estimated femur length of 58.9 mm, corresponding to 31SA + 6J, with an estimated fetal weight of 1920 ± 325 g, without cardiac

activity. An oval formation with an axis of 17 cm was highlighted, with tissue consistency, without visualisation of the cardiac region. A single posterior corporeal-fundal placental mass was also noted. Biological assessment showed no abnormality. At the end of the delivery, a cephalic extraction was performed and a female baby weighing 1905g was delivered. The general examination showed a fetus with neonatal cyanosis and no cardiac activity (Figure 1). The cardiac mass was delivered second, presenting with a trunk, malformed limb buds and an umbilical cord (Figure 2). Fetopathological examination was suggested but refused by the family.



Figure 1: Transfuser twin.



Figure 2: Acardiac twin.

Discussion

The TRAP sequence is an exceptional and specific complication of monochorionic twin pregnancies, affecting on average one birth in 35,000 and 1% of monozygotic pregnancies [1,2]. It is secondary to arterio-arterial and venovenous anastomoses at the level of the placenta, which are frequent and connect the circulations of the transfusion twin to the transfused twin. The pathogenesis of TRAP remains controversial [3]. The structure of the acardiac twin can vary from a well-differentiated fetus to one with no recognisable anatomy. Most commonly, the acardiac twin is anencephalic with no identifiable upper extremities, with a central trunk and the presence of a vertebral column. An umbilical cord with two vessels is found in more than two thirds of cases. Without a circulatory or lymphatic system, this twin often presents with severe subcutaneous oedema and cystic hygroma, which may be the cause of its severe deformity [3].

With the introduction of first trimester ultrasound, the diagnosis of the TRAP sequence is becoming earlier and earlier. The initial diagnosis is made in the presence of a fetus with severe malformation in a monochorionic twin pregnancy. The diagnosis is suggested by the disparity in size between the two fetuses, incomplete development of the head, trunk and upper limbs, deformity of the lower limbs, severe subcutaneous oedema and cystic hygroma, and the absence of cardiac activity in the child. In one of the two fetuses, even if the presence of cardiac activity does not exclude the diagnosis, the reverse flow in the umbilical artery towards the acardiac fetus on colour doppler and the aortic flow in the cephalic direction allow the diagnosis to be made. The demonstration of blood flow within a mass without cardiac activity is inherent to this anomaly [4].

The differential diagnosis includes in utero fetal death of a twin without anomalies, anencephaly associated with cystic hygroma and placental tumour [5]. The main prognostic factors to determine the condition of these twins and guide the action to be taken are the size of the acardiac fetus and the cardiovascular condition of the transfused fetus, namely tricuspid insufficiency or outright heart failure, hence the benefit of ultrasound monitoring by an experienced operator [6]. Treatment of the TRAP sequence in complicated twin pregnancies aims to achieve survival and compensation of the transfused fetus. Most fetal medicine centres delay treatment until 16-18 WA, according to Quintero et al. reaching this deadline could be fatal for the transfusion twin and suggests early treatment. If the diagnosis is not made in the first trimester, an expectant attitude may be most appropriate.

Various types of prenatal therapy are now available, and the occlusion of the vessels of the acardiac fetus, so well described

by Nicolaides et al. [7]. Coagulation of the umbilical cord by the energy of a laser beam has good results when the gestational age of the patient is less than 20-24 weeks. Intra-fetal therapies consist of ablation of fetal acardiac vessels in the abdominal aorta or pelvic vessels, which are identifiable regardless of the location of the placenta, umbilical cord, or amount of amniotic fluid [8,9].

Conclusion

The TRAP sequence in monochorionic pregnancy is characterised by the presence of a malformed fetus with a rudimentary or absent heart structure, perfused by A-A and V-V communication with the transfusion twin. The diagnosis is often made early in the first trimester and separation of the placental communication with the transfusing fetus is a safe alternative when the umbilical cord of the acardiac fetus is not identifiable. It is therefore important to determine the gestational age of the patient at the time of diagnosis to determine the benefits of expectant management versus invasive management.

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