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When does death occur in an acardiac twin? Ultrasound diagnostic difficulties

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1 Introduction

Fetal acardia, a rare abnormality seen only in multiple pregnancy, occurs once in every 35.000 births [5]. The malformation affects one fetus only and is thought to derive from an umbilical artery to artery anastomosis within a fused monochorial and monoamniotic placenta. Consequent haemodynamic alterations have been suggested to result in reverse arterial perfusion of the 'perfused' twin, by its sibling, the 'pump' twin [8].

The twin reverse arterial perfusion sequence (TRAP) is associated with four major types of abnormality: Acardius acephalus, with absence of fetal head and thoracic organs, myelacephalus with partially developed head and extremities, amorphus where there is no recognisable human shape, and acornus where only the fetal head develops [2]. TRAP sequence is lethal for the affected fetus, and causes death in up to 50% of normal co-twins, either from intrauterine cardiac failure or the sequelae of prematurity [8]. Antenatal recognition of the condition is imperative if prospective management is to be instituted to improve the outcome [6]. The purpose of this communication is to report difficulties in ultrasound diagnosis, and to stress the risk of sudden intra uterine death of the pump twin in absence of compromise on ultrasound, Doppler studies or fetal blood sampling.

2 Case report

A routine ultrasound scan at 17 weeks on a 22-year-old primigravid patient showed the presence of monoamniotic twins with polyhydramnios. Twin I was normal but twin II was considered to have anencephaly and ascites. Fetal heart activity was seen in each twin (figure 1). Because twin I appeared entirely normal, and twin II was considered non viable, the parents were not offered selective fetocide and the pregnancy continued. Repeat scan at 22 weeks showed that the heart of twin II had stopped beating and this fetus had become grossly oedematous. Its umbilical cord was tightly wrapped around the leg of the other twin (figure 2), in which growth, anatomy, and Doppler blood flow velocity waveforms in the umbilical artery remained normal. Fetal blood sampling, performed at 24 weeks from the cord of twin I to exclude genetic abnormalities and DIC and confirm its well being, showed normal karyotype (46 xx), blood gases, haematological and clotting studies. The procedure was completely uncomplicated but three days later the mother reported lack of fetal

Curriculum vitae

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Figure 1. Ultrasound scan taken at 17 weeks. On the left a sagittal view of the chest of the acardiac monster showing the structure which appeared to be pulsating. On the right a similar view of the normal twin.

movements and twin I was found on ultrasound to be dead.

Labour was induced with vaginal prostaglandins, resulting in an uncomplicated delivery of two female infants weighing 200 and 780 g. Post mortem revealed that twin II was anencephalic and acardiac (figure 3). There were early signs of maceration, and a double aortic arch arose from the lungs without any evidence of cardiac tissue, while the oesophagus, stomach and liver were absent. A cervical spina bifida was also present. Twin I was normal, but on histological examination fibrin clots were found in the lungs, kidneys and liver. Both artery to artery and vein to vein anastomoses were identified within the monochorionic placenta, which weighed 330 g. The cord of twin II, which was centrally inserted and contained three vessels, was tightly wrapped around the other twin's leg, producing trophic changes and constriction phenomena (figure 4).

3 Discussion

The diagnosis of acardiac monster must be made antenatally if the prognosis for the normal co-twin is to be improved in TRAP. However it is only recently that this abnormality has been identified antenatally on ultrasound [3]. The diagnosis is straightforward in the presence of an amorphus mass of tissue or of an acephalus acardiac twin associated with an anatomically normal co-twin [7], which may show signs of cardiac failure. The diagnosis can however be more difficult with malformations such as anencephaly, especially if the normal co-twin has no evidence of chronic cardiac failure.

In our case the assessment was made even more difficult by normal heart activity seen in the acardiac fetus on the initial scan. Retrospective analysis of the original ultrasound scan together with the autopsy findings led us to speculate that

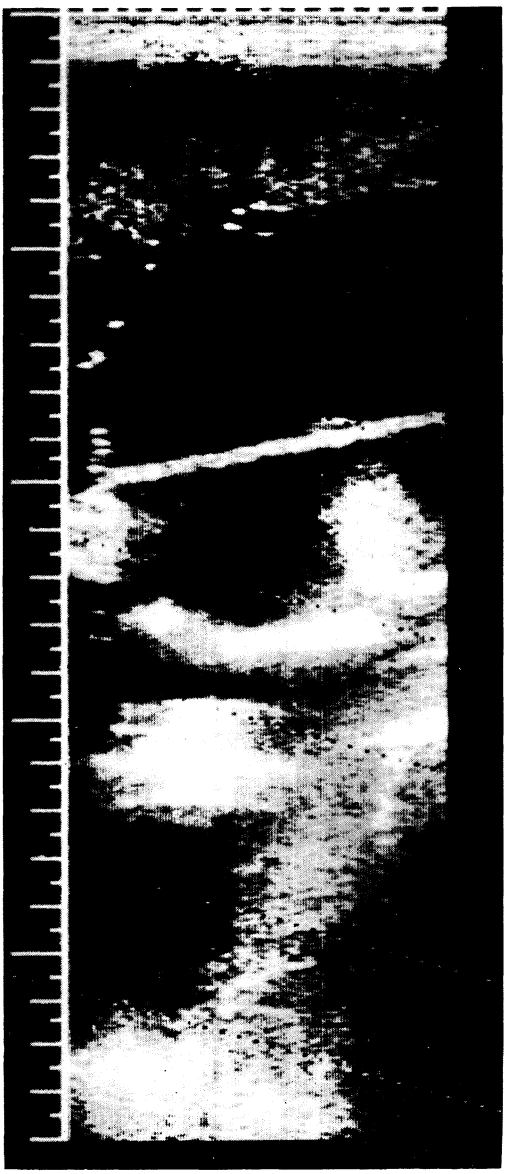

Figure 2. Ultrasound scan of the umbilical cord of the acardiac monster tightly wrapped around the leg of the pump twin.

the heart motion seen was a mass of lung tissue with dilated vessels which pulsed, as a consequence of the reverse perfusion, at the same cardiac rhythm as the other twin. The eventual disappearance of this pulsation may have occurred as a consequence of the interruption of the reverse perfusion when the cord became tightly wrapped around the leg.

Absence of fetal heart activity on ultrasound is the gold standard for diagnosis of intrauterine fetal death, yet in acardia this assessment is not possible. The demonstration of reflex movements

Figure 3. The acardiac twin.

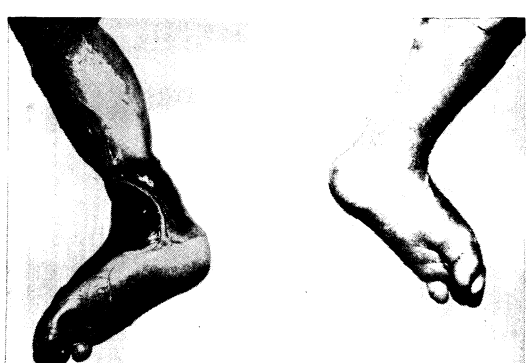

Figure 4. Twin I, the "pump" twin. Note the signs of early maceration and the oedema of the right foot, caused by the entangled cord of twin II.

in acardiac fetuses suggests that they are still 'alive', but no information exists on the accuracy of absent movements in diagnosing fetal death. Diagnostic difficulty may also arise in the interpretation of reflex movements in monoamniotic twins when, like in our case, the dead twin is 'towed' around the amniotic cavity by its co-twin. In view of the original ultrasound, we assume that both fetuses were alive at 17 weeks, and subsequent fetal death was diagnosed when no heart activity was seen in the abnormal fetus. However this sign in the acardiac fetus is dependant upon the heart activity of the other twin and unimpeded perfusion through placental anastomoses. We suggest that in the presence of cord entanglement, perfusion varied with the degree of tautness. At autopsy, only mild maceration was found in the acardiac fetus, suggesting that tissue death had occurred much more recently than suggested by the three weeks of absent cardiac activity seen on scan. Presumably intermittent loosening and tightening of the loop of cord with consequent resumption and interruption of reverse perfusion could have occurred until shortly before delivery, leading to a state of progressive avascular necrosis or 'partial' death. Under these circumstances the ultrasound detection of intermittent pulsatile activity in the 'dead' fetus may pose ethical and diagnostic difficulties.

Several possibilities exist for the cause of death in the pump twin. The normal co-twin often develops chronic cardiac failure because of the reverse perfusion, producing a mortality of 50% [8]. In our case there was no evidence of compromise in the pump twin, on the basis of both the normal haematological and gas values obtained at fetal blood sampling, and of the normal ultrasound and doppler studies. Specifically, there was no evidence of cardiac failure with only mild ventriculomegaly in the absence of oedema or hepato-splenomegaly being found at autopsy. Fetal death as a complication of blood sampling appears unlikely since the procedure was uncomplicated, and the death occurred at least 48 hours after normal fetal heart rate and movements were recorded following the procedure.

Loosening and tightening of the cord of the perfused twin would on the other hand have produced intermittent circulatory shunts in the pump twin, with the release of de-oxygenated blood into the circulation when the cord relaxed. The resulting series of hypo- and hypertensive episodes in the normal twin, could then lead to death from vagal cardiac inhibition, acute circulatory collapse or haemorrhage. Intrauterine death can occur in 73% of monochorionic twins [1] and is often attributed to cord entanglement, absent however, in our normal twin.

The finding of intravascular fibrin deposis in the liver, lungs and kidneys of the normal twin is puzzling since the fetal clotting screen was normal 48 hours before death. Fetal disseminated intravascular coagulation has been suggested to occur after intrauterine death of one twin [4], but has not previously been reported in the pump twin in TRAP. Recurrent hypotensive and hypertensive episodes could, on the other hand, have caused multiple haemorrhages with fibrin deposition via a hydromechanical mechanism.

In conclusion, care should be taken in the ultrasound diagnosis of single intrauterine death in multiple pregnancy to exclude the presence of an acardiac fetus. The co-twin in TRAP is always at increased risk of sudden death, even without ultrasound or biochemical evidence of compromise. This case illustrates a diagnostic pitfall and rises the question of when death occurs if no heart or brain are present.

Abstract

Fetal acardia is a rare abnormality of multiple pregnancies, which is lethal for the affected fetus and can cause death in 50% of normal co-twins. Antenatal recognition with early ultrasound is essential to institute a prospective management to improve the outcome. Our communication outline the difficulties which may be encountered in ultrasound diagnosis. In particular the problem of distinguishing a fetal heart from large pulsating mediastinal vessels, which can be present in these fetuses, and the difficulty of diagnosing death in an acardiac fetus. Our report confirms that the co-twin remains at increased risk of sudden death, even without ultrasound evidence of cardiac failure or biochemical compromise. The finding in this fetus of intravascular fibrin deposits suggests the possibility of acute disseminated intravascular coagulation, not previously reported in association with an acardiac twin.

Keywords: Acardia, twins, ultrasound.

Zusammenfassung

Wann tritt beim Zwillings-Akardiakus der Tod ein? Diagnostische Probleme beim Ultraschall

Wir berichten über einen fetalen Akardiakus, bei dem im zweiten Trimenon sonographisch Herzaktionen gesehen wurden, die jedoch bei nachfolgenden Ultraschalluntersuchungen nicht mehr nachweisbar waren. Die Nabelschnur des Zwillings-Akardiakus war straff um das Bein gewickelt. Unsere Hypothese lautet, daß diese „Herzaktionen" durch Pulsationen erweiterter Thoraxgefäße hervorgerufen wurden. Der Nachweis dieser Pulsationen war abhängig von der Straffheit der verwickelten Nabelschnur – ein partielles Absterben (progressive Nekrose) war die Folge.

Großflächige Fibrinablagerungen, die sich bei der Autopsie des gesunden Zwillingspartners nachweisen ließen, lassen an eine disseminierte intravaskuläre Koagulation denken. Darüber wurde bisher im Zusammenhang mit einem fetalen Akardiakus noch nicht berichtet.

Schlüsselwörter: Akardiakus, Ultraschall, Zwillinge.

Résumé

Quand la mort survient-elle chez un jumeau acardiaque? Difficultés diagnostiques à l'échographie

Nous présentons un cas de fœtus acardiaque chez lequel une échographie du second trimestre suggérait une activité cardiaque bien que celle ci n'ait pas été observée sur les coupes ultérieures. Le cordon du jumeau acardiaque se trouvait être étroitement enroulé autour de la jambe de l'autre jumeau. Nous émettons l'hypothèse que cette activité «cardiaque» étant secondaire aux pulsations des vaisseaux dilatés dans le thorax. La présence ou l'absence de cette pulsation dépendaient du caractère plus ou moins serré de l'enroulement du cordon conduisant à un état de mort partielle (nécrose progressive). La présence de dépots de fibrine étendus trouvée à l'autopsie du jumeau normal suggère la possibilité d'une coagulation intravasculaire disséminée, ce qui na pas été antérieurement rapporté en association avec un fœtus acardiaque.

Mots-clés: Acardiaque, échographie, jumeaux.

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