International Journal of Academic Health and Medical Research (IJAHMR)
ISSN: 2643-9824
Vol. 7 Issue 1, January - 2023, Pages: 81-84

TRAP Sequence: A Case Report.

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Abstract: *Acardiac mass or TRAP sequence is a rare and serious pathology of monochorionic pregnancy. Its frequency is 1/35 000. During this pathology one of the two twins has no heart, and its vascularization is ensured against the current by the second one via very wide arterial and venous anastomoses, responsible for a hemodynamic imbalance. The diagnosis must be made in the first trimester and the evolution can easily be made towards the high flow heart failure of the "Pump" twins, justifying a follow-up every 15 weeks, which will be even closer if signs of heart failure set in. Diagnosis and follow-up are based on ultrasound with fetal Doppler studies. We report the case of a twin pregnancy with TRAP sequence discovered on ultrasound at 23 weeks of amenorrhea, with delivery at 27 weeks.*

Keywords: TRAP sequence, twin pregnancy, prenatal diagnosis.

1. Introduction:

Acardiac mass or TRAP sequence is a rare and serious pathology of monochorionic pregnancy.
During this pathology one of the two twins has no heart, and its vascularization is ensured against the current by the second one via very wide arterial and venous anastomoses, responsible for a hemodynamic imbalance.
The diagnosis can be made in the 1st trimester and the evolution can easily be made towards the high flow heart failure of the "Pump" twins, justifying a follow-up every 15 weeks, and which will be even closer if signs of heart failure set in.

This pregnancy is generally overhung by a great morbidity requiring a rigorous monitoring, with sometimes the necessity of an "exclusion" of the acardiac by funicular obliteration or coagulation by interstitial laser. The need to anticipate this situation and the specificity of this management should lead the patient to be referred to a specialized center as soon as the diagnosis is made.

2. Clinical case:

We report the case of a twin pregnancy with TRAP sequence discovered on ultrasound at 23 weeks of amenorrhea, in a patient who consulted for a diagnosis of amenorrhea. This ultrasound showed the existence of a twin pregnancy most probably monochorionic and mono-amniotic, with a twin without any particularity, next to a very heterogeneous tissue mass with liquid and bone images roughly reminding the two lower limbs and a trunk with a spinal axis at the level of which a very short umbilical cord takes birth.
Initially the pumped fetus did not show any signs of fetal distress or anemia. The option of feticide by electrocoagulation was excluded since the free portion of the cord was very short and very difficult to visualize on ultrasound. Weekly ultrasound monitoring to detect early signs of heart failure with pulmonary maturation by corticosteroid therapy at 28 SA if no abnormality.

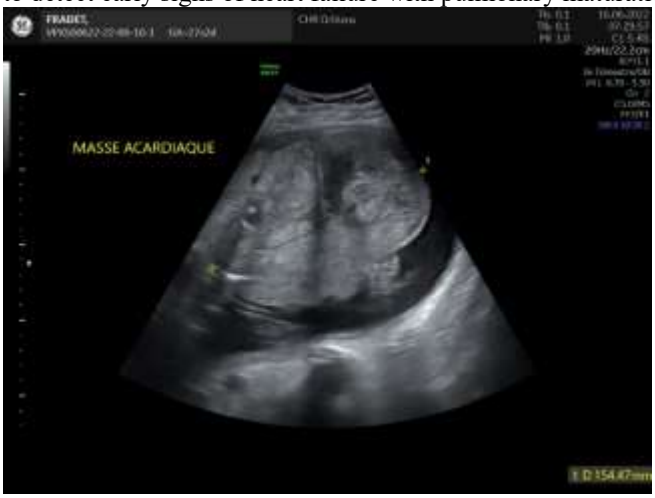

Figure 1 : Acardiac mass : heterogeneous tissue mass with liquid and bone images roughly reminding the two lower limbs and a trunk.

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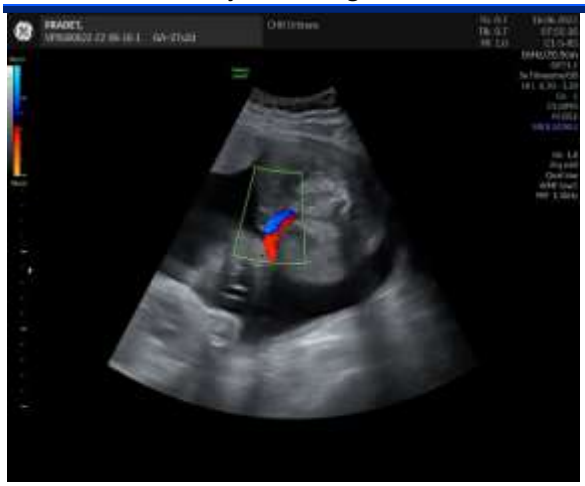

Figure 2 : the free portion of the ombilical cord.

At 27 weeks + 4 days, anomalies appeared on the fetal Doppler: increase in the velocity of the middle cerebral artery, tricuspid leakage, and increase in the resistance index of the ductus veinosus, with hydramnios, hence the decision to perform a fetal extraction by the high route, for fetal rescue.

The anemic and hypotrophic baby was hospitalized in neonatology and presented respiratory distress, dying at 2 days of life.

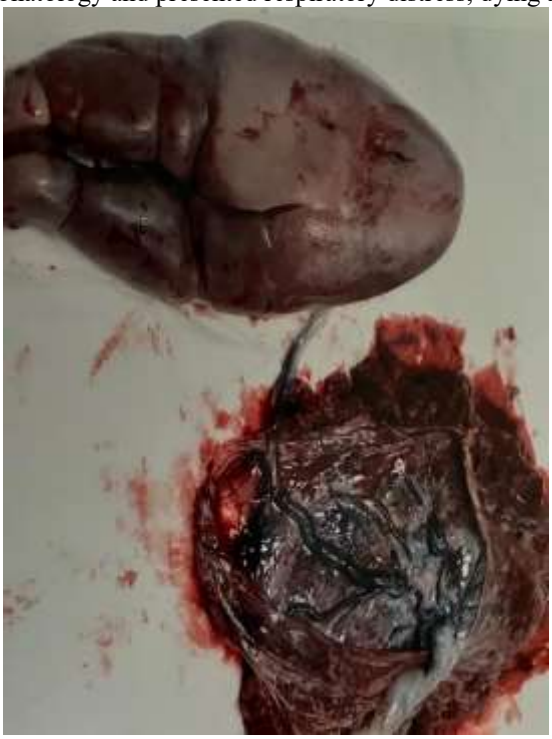

Figure 3,4: Acardiac mass after delivery

3. Discussion:

The acardiac fetus or acardiac monster is a serious and rare malformation, its incidence is estimated at 1 / 35 000 pregnancies, it concerns monochorionic twin pregnancies whether they are mono or bi-amniotic. It is currently referred to as reverse arterial perfusion syndrome.

The acardiac mass presents as a lower half of the body usually without upper limbs, cephalic extremity and heart. Abnormalities of abdominal wall closure are often present with lower limbs present in whole or in part, and sometimes abdominal organs in place.

There is usually significant edematous infiltration and in all cases the heart is absent or rudimentary. Less complete forms leave part of the cephalic extremity (base of the skull) and the upper limbs more or less complete, creating the acardiac-acephalic or acardiac-anencephalic monster.
From a pathophysiological point of view, there is not a well established etiology, especially with the description in the literature of TRAP sequence cases with different sexes.
Generally it is the association of several pathological processes during fertilization, leading to a very early defect of blood perfusion. During this pathology, the blood flow becomes unidirectional, coming from the normal twin called Pump, towards the 2nd acardiac. This perfusion is then reversed for the second, with insufficient perfusion of the upper half, hence its regression. For the pump twin who loses part of his blood, an increase of the cardiac flow is noted which can evolve towards cardiac insufficiency with hydramnios.

The diagnosis can be made in the first trimester during the prenatal ultrasound where the color Doppler plays a crucial role.
In the first trimester, in a monochorionic pregnancy, the discovery of an abnormal embryonic structure, poorly defined, with no visible cardiac activity, should not lead to a hasty conclusion of embryonic evanescence with an embryo in the process of lysis, but requires a control of the ultrasound within 2 weeks in order to judge its evolution;
Later, an edematous mass will be visualized, with disorganized anatomical elements, very hypoplastic for the upper part (often acephalic), associating liquid areas (occluded digestive segments) and echogenic (disorganized bone structures). The lower hemisphere is often better preserved in its structure with individualizable lower limbs. A progressive oligohydramnios is often observed.

The vitality of the acardiac is objectified by its growth and the presence of an umbilical flow: reversed pulsatility if the implantation of the cord can be observed in Doppler. Because of the vascular flight that it causes, the pregnancy can be complicated by hydramnios and rapidly evolving cardiac insufficiency in the "pump" twin (30% of cases) that can lead to its death in utero. It should therefore benefit from rigorous surveillance, sometimes requiring "exclusion" of the acardiac by funicular obliteration or interstitial laser coagulation. The need to anticipate this situation and the specificity of this management should lead the patient to be referred to a specialized center as soon as the diagnosis is made.
The prognosis of this pregnancy depends essentially on the weight of the acardiac mass and the degree of its development. Thus a weight lower than ¼ of the pump twin is generally a good prognosis. Also the more the acardiac twin has more organ the more the risk of complications is real. The appearance of hydramnios, cardiomegaly, pericardial effusion, tricuspid insufficiency, negative A wave on the ductus venosus, pulsatile umbilical vein, as well as signs of anemia are all also considered factors of cardiac decompensation of the healthy twin, and therefore of poor prognosis

The management of this pathology consists of occluding the suppletive vascularization of the acardiac twin, which allows the hemodynamic situation of the pump twin to be restored. It is done by laser coagulation of the cord of the acardiac twin, generally before 24 hours.
Thereafter, it is advisable to monitor the pump twin by performing Doppler of the ACM in search of anemia by bleeding from the pump twin into the acardiac twin.

4. Conclusion:

TRAP sequence is a serious pathology fortunately rare which concerns monochorionic twin pregnancies. The diagnosis is ultrasonographic. The management by vascular occlusion of the acardiac mass, allows to improve the prognosis.

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