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Case Report

# Respiratory Distress of Mother Secondary to Severe Acute Polyhydramnios Secondary to Spontaneous Quadruplet Pregnancy and TRAP (Twin Reversed Arterial Perfusion) Sequence in Ambo University Referral Hospital, Ambo, Ethiopia. A Case Report

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**Keywords**

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**Abstract**

**Background:** High order multifetal gestation is important because these pregnancies are higher risk of maternal, fetal, and neonatal complications than singleton pregnancies. Quadruplets can be fraternal (multizygotic), identical (monozygotic), or a combination of both. Multizygotic quadruplets occur from four unique egg/sperm combinations.

Spontaneous quadruplet pregnancy is very uncommon with an incidence rate of 1 in 512000 to 1 in 677, 000 births. TRAP sequence is a serious complication that occurs exclusively in monochorionic pregnancies. It is caused by one or more abnormal vascular connections between fetuses that occur most commonly on the surface of the placenta or, in some cases, within the umbilical cord.

**Case Summary:** A 35 years old Gravid 11 Para 7 Abortion 3 and molar pregnancy one who doesn't remember her last normal menstrual period but claimed to be ammenorhic for the last five months presented to our facility with shortness of breath and on evaluation there was features of respiratory distress and abdomen was extremely distended and on ultrasound quadruplet with severe polyhydramnios and one of the fetus with major congenital anomalies which considered as cystic hygroma later after delivery diagnosed as Acardiac quadruplets. After discussion with the patient pregnancy was terminated and three of quadruplet without major congenital anomalies whiles the other was Acardiac quadruplet.

**Conculsion:** Mono chorionic quadruplet with Acardiac is very rare and up to my knowledge not reported yet. Early evaluation of patient with ultrasound will help to identify the problem and early intervention is possible before the patient develops symptoms and complications.

## INTRODUCTION

High order multifetal gestation is important because these pregnancies are higher risk of maternal, fetal, and neonatal complications than singleton pregnancies.

Twin reversed arterial perfusion sequence (TRAP) refers to a rare, unique complication of monochorionic twin pregnancy in which a twin with an absent or a non-functioning heart ("Acardiac twin") is perfused by its co-twin ("pump twin") via placental arterial anastomosis. The Acardiac twin usually has a poorly developed heart, upper body, and head. The pump twin is at risk of heart failure and problems related to preterm birth.

TRAP sequence occurs in about 1 percent of monochorionic twin pregnancies and 1 in 35,000 deliveries [1].

The development of the Acardiac anomaly is a rare complication of monozygotic multiple pregnancies. Acardiac foetuses were first described by Benedetti in 1533 [2] (twin obstructed).

The Acardiac twin is transfused by the normal co twin by means of reversal of circulation through large vein to vein and artery to artery anastomosis and has no direct communication with placenta. The outcome is 100% fatal for Acardiac twin with a 50-75% mortality of the normal pump twin [2].

The abnormal circulatory pattern provides perfusion of deoxygenated blood from the pump twin to the lower half of the recipient twin via its iliac arteries, but poor perfusion of the upper torso and head. Unequal vascular perfusion from the pump twin may contribute to the evolution of a variety of structural abnormalities in the recipient twin [3].

Many of the bizarre defects are felt to be caused by low oxygen tension and this causes dramatic alteration in the twin fetal physiology and high prenatal mortality.

The Acardiac twin is a parasite, putting the pump fetus at risk of high output cardiac failure [4].

Four distinct morphological types have been described in different literature and standard text books [5].

**Acardiac acephalous**: It is the most common type (60-75%) with developed pelvis and lower limbs, may have arms, but thoracic organs and head absent

**Acardius anceps**: The most differentiated type of Acardiac twins with established body and extremities, but only partially formed head and face (20%).

**Acardiac acormus**: In which only cephalic structures were detectable, is a sporadic type (10%).

**Acardius amorphous**: Consists of shapeless mass of tissue containing no recognizable human structures.

TRAP should be suspected in monochorionic twin pregnancies when one fetus appears anatomically normal and the other lacks apparent cardiac structures and/or activity.

The degree of abnormality in the Acardiac twin varies widely, ranging from a fetus with well developed lower extremities, pelvis, and abdomen to a tissue mass that is not readily recognizable as fetal parts. The cranium may be absent or present with cranial defects, such as anencephaly or holoprosencephaly.

In addition, there can be limb defects, anterior abdominal wall defects, and absence of lungs, kidney, spleen, and/or liver [6]..

In addition to the absent or non-functioning heart, hallmarks of the Acardiac twin include massive edema of the head, trunk, and upper extremities. This edema exceeds that which might be attributable to autolysis and is related to the altered perfusion [6].

The pump twin may have signs of high-output cardiac failure: hydramnios, cardiomegaly, pericardial and pleural effusions, ascites, and tricuspid regurgitation. The development of cardiac failure in TRAP sequence is related to the ratio of the size of the Acardiac twin to that of the pump twin. When this ratio exceeds 0.70 (i.e., the calculated weight of the Acardiac twin is 70 percent or greater compared to the weight of the pump twin), the risk of congestive heart failure in the pump twin is approximately 30 percent, compared to a risk of 10 percent when the ratio is less than 0.70 [7].

## CASE REPORT

**A** 35 years old Gravid XI Para VII Abortion II (spontaneous) and one molar pregnancy for which suction curettage done three years back, didn't remember her last normal menstrual period but complained ammenorhic for the last five months presented to our hospital after she referred from Jaldu primary hospital with the impression of respiratory distress secondary to poly hydramnios plus triplet pregnancy for further investigation and management.

Upon on arrival to our hospital she was complaining shortness of breath and upon on physical examination she was in respiratory distress with flaring of ala nasi ,RR=32per minute deep breathing ,other vital sign within normal range ,abdomen was extremely distended ,difficult to appreciate fetal part and there was pedal pitting edema bilaterally

Iv line was opened, the patient was put on intranasal oxygen and taken to ultrasound room for further evaluation.

On Ultrasound evaluation there was excessive amniotic fluid which single deepest pocket measured 16.68 cm and there were four fetuses seen. Three of them their cardiac motion were seen, no gross anomalies seen and their gestational age were in the average of 19-21 weeks and one thick dividing membrane was also seen

The fourth fetus had excessive cystic like appearance on the whole of his neck, that extends down ward to back and the only small skull bone was seen. No cardiac motion was seen and Gestational age from bowed femoral length was 14 weeks (Figure 1 and Figure 2).

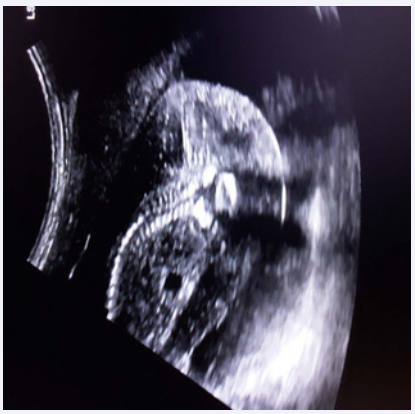

**Figure 1** Ultrasound picture of Acardiac quadruplet at Ambo University Referral Hospital, Ambo, Western Ethiopia, December 16, 2019.

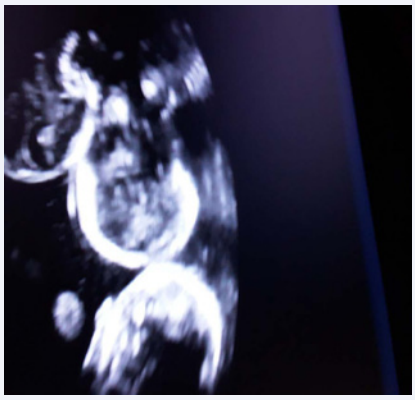

**Figure 2** Ultrasound picture of head of triplets at Ambo University Referral Hospital, Ambo, Western Ethiopia, December 16, 2019.

After ultrasound evaluation the assessment of respiratory distress secondary to severe poly hydramnios plus quadruplet with severe cystic hygroma of fourth fetus was made and after discussion with the patient and patient family termination of pregnancy was decided to relieve patient from her current problem.

After termination of pregnancy decided pelvic examination was done and cervix was 2 cm dilated ,station was high, intact membrane ,soft and anterior cervix ,60% effaced and with favourable cervical score induction with 5IU oxytocin was started and the dose escalated every 30 min and on the second phase of oxytocin drip cervix was dilated to 5cm membrane bulged and controlled artificial rupture of membrane done and after 4 hours she gave explusions of Q1----male,200 gram, Q2------male,200 gram,Q3---- male,200 gram and Q4---400 gram male with whole oedematous body, short extremities, no head and eye seen but umbilical cord with single vein and two arteries seen which had appearance of Acardiac features of Q4 (Figure 3 and Figure 4).

After explusions fetuses, 10IU oxytocin given and placenta was removed by controlled cord traction.

The patient discharged on second day of expulsion with stable vital sign and advised on family planning and she decided to take at her nearby hospital

## CASE DISCUSSION

A multiple pregnancy may be the result of the fertilization of a single egg that then splits to create identical fetuses, or it may be the result of the fertilization of multiple eggs that create fraternal fetuses, or it may be a combination of these factors. A multiple pregnancy from a single zygote is called *monozygotic*, from two zygotes is called *dizygotic*, or from three or more zygotes is called *polyzygotic*. Similarly, the siblings themselves from a multiple birth may be referred to as monozygotic if they are identical or as polyzygotic if they are fraternal.

Quadruplets are much rarer than twins or triplets. As of 2007, there were approximately 3500 sets recorded worldwide. Quadruplet births are becoming increasingly common due to fertility treatments. There are around 70 sets of all-identical quadruplets worldwide. Many sets of quadruplets contain a mixture of identical and fraternal siblings, such as three identical and one fraternal, two identical and two fraternal, or two pairs of identical

Quadruplets can be fraternal (multizygotic), identical (monozygotic), or a combination of both. Multizygotic quadruplets occur from four unique egg/sperm combinations. Monozygotic multiples are the result of a fertilized egg that splits into two or more embryos. It is possible for a split to occur more than once, producing monozygotic triplets or even a rare set of monozygotic quadruplets. Fully monozygotic quadruplets are rare, representing only one in about 13 million pregnancies [8].

Quadruplets can be all male, all female, or a combination of both. Monozygotic quadruplets will always be of the same gender [9].

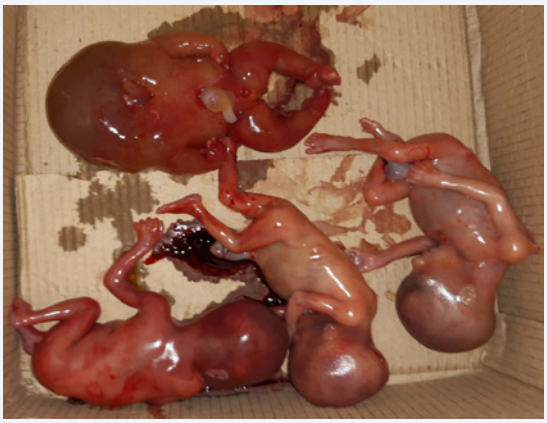

**Figure 3** Photographs of quadruplets with TRAP sequence after expulsion at Ambo University Referral Hospital, Ambo, Western Ethiopia, December 16, 2019.

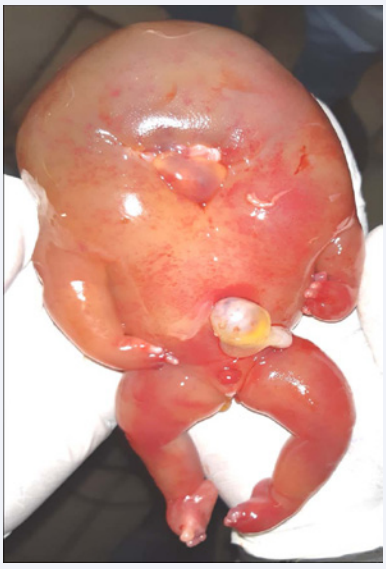

**Figure 4** Acardiac quadruplet(TRAP sequence) after expulsion at Ambo University Referral Hospital, Ambo, Western Ethiopia, December 16, 2019.

In 2017, 193 quadruplets were born. Spontaneous conception of quadruplets is rare [10]. Spontaneous quadruplet pregnancy is very uncommon with an incidence rate of 1 in 512000 to 1 in 677, 000 births [11].

The MOST (Mothers of Super Twins) organization estimates the odds at 1 in 571,787 pregnancies [12]. Most recent quadruplet births are the result of assisted reproductive techniques such as fertility-enhancing drugs or in-vitro fertilization.

TRAP sequence is a serious complication that occurs exclusively in monochorionic pregnancies. It is caused by one or more abnormal vascular connections between fetuses that occur most commonly on the surface of the placenta or, in some cases, within the umbilical cord. As a result, instead of diving down into the placenta, the blood returning from one fetus (the donor or "pump fetus") goes directly to the second fetus (the recipient) and then back to the placenta. As a result, the recipient twin receives blood that is low in oxygen and nutrient content, and that is travelling in the wrong direction through its arterial circulation; hence the term, TRAP sequence.

For pregnancies between 18 and 27 weeks of gestation, current treatment modalities target occlusion of the umbilical cord of the Acardiac twin and include laser ablation, bipolar cord coagulation, and radiofrequency ablation (RFA), which are performed with local anaesthesia and conscious sedation [13,14].

Even though different modality for the treatment of TRAP available worldwide, in our set up lack of materials, human power and experience and also considering the patent condition as well the lack of availability of neonatal intensive care unit we decided for the termination of pregnancy.

## CONCLUSION AND RECOMMENDATIONS

Twin reversed arterial perfusion sequence (TRAP), refers to a rare, unique complication of monochorionic twin pregnancy in which a twin with an absent or a non-functioning heart ("Acardiac twin") is perfused by its co-twin ("pump twin") via placental arterial anastomosis

TRAP should be suspected in monochorionic twin pregnancies when one fetus appears anatomically normal and the other lacks apparent cardiac structures and/or activity.

The Acardiac phenotype ranges from well developed lower extremities, pelvis, and abdomen to a tissue mass that is not readily recognizable as fetal parts.

Early ultrasound evaluation of pregnancies is important in order to identify the number of multiple pregnancies and early intervention.

Even though a treatment for such cases is very infant in this country some centres started feto maternal medicine speciality and such cases should referred to them if early identified

Pathology result is mandatory for such cases but not available in our facility and we recommend for the hospital management to avail for such cases.

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