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A Rare Case Report on Twin Reversal Arterial Perfusion

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Abstract:*Twin-Reversed Arterial Perfusion (TRAP sequence)is a rare complication of monochorionic twins. TRAP sequence is known as acardius or chorioangiopagusparasiticus. It occurs in 1% ofmonochorionic twin pregnancies and in 1 in 35,000 pregnancies1. TRAP sequence is characterized by a structurally normal pumptwin perfusing an anomalous twin. In TRAP sequence, one twinis usually developmentally normal (pump twin) and the other twin has a serious condition like missingaheart (acardiac) or a head (acephalic) or both, that prevents it from surviving on its own.The term "reversed perfusion" is used to describe this scenario because bloodenters the acardiac/acephalic twin through reversed flow through its umbilical artery and exits through theumbilical vein, which is opposite to the normal blood supply of the fetus. The mortality of the acardiac twin is100%, and the perinatal mortality of the pump twin is reported to be around 50%.TRAP sequence was diagnosed by ultrasoundat SAH@ RC ultrasound centre and referred for management2.*

Keywords: *Monochorionic twins; Pump twin; Reverse flow; Acardiac / Anceps.*

I. Introduction

Acardiac Twin or TRAP Sequence is a very rarecondition with an incidence of 1 in 35,000pregnancies**,** occurring in approximately 1% ofmonochorionic twins. One twin is structurallynormal and is referred to as pump twin becauseit pumps blood to other twin which is abnormal. Abnormal in the form of consisting only legs and lower half of the body but no upper half of the body, head or heart. In TRAP sequence the deoxygenated blood from the normal twin enters theabdomen of the acardiac fetus allowing forsome development of lower body.Once blood reaches the upper half of the body oxygen saturation is extremely low halting development of this area. The risk is directly dependent on the size of the acardiac twin and the mortality rate for the normal (pump) twin is approximately 50%.

II. Case Report

A 26 years old Gravida 2 para 1 living 1 was 19 weeks of gestation with twin pregnancy of monochorionicdiamniotic diagnosed at 16th week scan was been followed in our hospital and diagnosed as TRAP when sent for anomaly scan at SAH &RC.

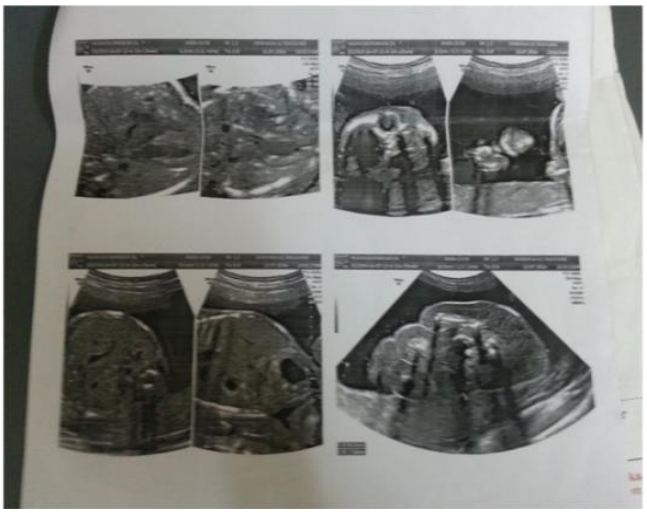

Figure – 1

Twin A with polyhydramnios of AFI 8 cm. Twin B is grossly edematous with bilateral Talipesequinovarus. Fetal spine is present, rudimentary calvarial bones present. Upper limb is not seen and anterior abdominal wall defect is present. Color Doppler study shows twin reverse arterial perfusionof umbilical artery ratio 2.49. .

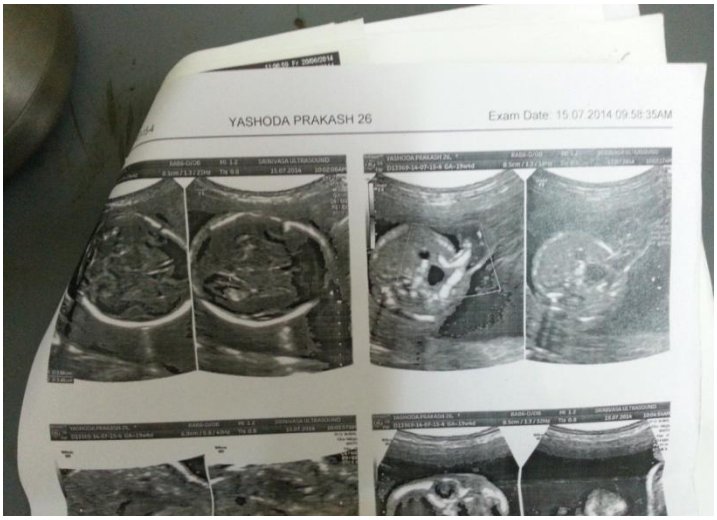

Figure – 2

On examination uterus was 32 weeks size relaxed.Patient was been explained regarding the treatment options and complications associated with it.Patient wanted to go for termination so consent was taken and induced delivered a live male baby 500gm and acardiacacephalus baby 500 gm.Acardiac twin showed anterior abdominal wall defect, no cranium,no left upper limb and baby is fully edematous and well developed lower limbs.

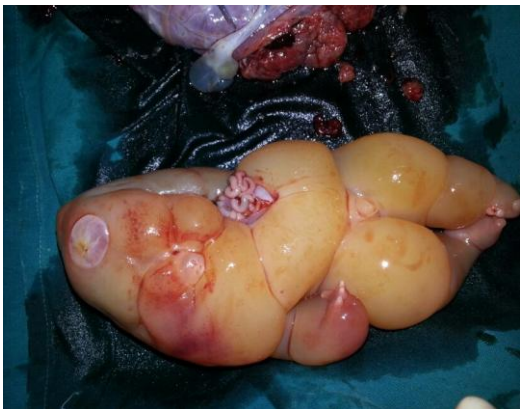

Figure – 3

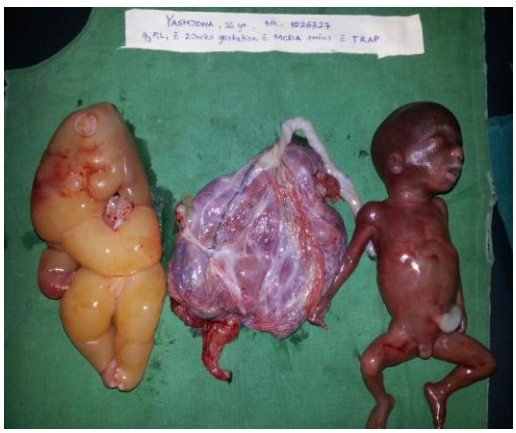

Figure - 4

III. Discussion

Twin reverse arterial perfusionsyndrome (TRAP) was first defined by Grunewaldin 1942. TRAP syndrome occurs in monochorionicgestation form. In which case one twin hasAcardia (the recipient) with no heart and theother twin is structurally normal (pump or donortwin). Due to the absence of heart in the acardiatwin the pump twin supplies deoxygenatedblood via vascular anastomoses to the acardiactwin. Acardia twin does not send blood toplacenta and all its blood comes from and goesback to the circulation of the pump twin, throughthe vascular connections on the surface of the shared placenta**.** It is therefore structurally normal twin perfusingan anomalous recipient twin via an artery-to artery anastomosis in a reverse direction**.** Thereversed flow is through its umbilical artery andexits through the umbilical vein which isopposite to the normal blood supply of thefetus. Deoxygenated low-pressure blood fromthe pump twin which would normally return tothe placenta, instead flows directly to theacardiac twin, resulting in a wide array ofstructural abnormalities, caused by arterioarterialand veno-venous placentalanastomoses. The acardiac twin is usually grosslyabnormal with severe reduction anomalies of the upper part of the body **as seen in our presentcase**. Mortality of the pump twin is 50-75%usually due to the result of heart failure and oftheacardiac is 100%.

Schatz (1898) classifiedacardia into two main groups: Hemiacardius(imperfectly formed heart) and Holoacardius(absence of heart).The first type is Acardiusacephalus, where nocephalic structures present.(head &upperextremities are lacking. It is most commonvariety. The second is acardiusanceps where somecranial structure and neural tissue or brain tissue is present. The body and extremities are alsodeveloped.The third is acardiusacormus with cephalicstructure but no truncal structures arepresent.i.e. Head without a body. The umbilicalcord is attached to the head. It is rarest form ofAcardiaThe fourth type is Acardius amorphous withno distinguishable cephalic or truncalstructure.It is least developed and not recognizable ashuman form with minimal development. Thisdiffers from Teratomas only by its attachmentto an umbilical cord(3 and 4).Therapeutic options targeted at interrupting thevascular anastomosis between the twins underultrasound guidance using fetoscope. Severaldifferent techniques have been used to treatTRAP sequence by interrupting the connectionbetween the acardiac twin and the pump twinto increase the chances that the pump twin willsurvive. These techniques include cord occlusionby embolization, ligation, laser photocoagulation,monopolar and bipolar diathermy.Intrafetal ablation has also been performed byalcohol injection, monopolar diathermy,

interstitial laser, and radiofrequency ablation(RFA)5.It is also important to exclude a chromosomalabnormality prior to offering a fetoscopic procedure in TRAP sequence since the incidenceof chromosomal abnormality in the pump twinmay be as high as 9 percent.

IV. Conclusion

Accurate antenatal diagnosis is essential toimprove the prognosis of this rare entity of TRAPsequence**.** Improved imaging techniques like 2Dultrasonography, 3D ultrasonography andtransvaginal Doppler ultrasonography havemade the diagnosis of Acardia possible even inthe first trimester of pregnancy by detectinginversion of vascular flow in the recipient

acardiac fetus. Early diagnosis may help toreduce the risk of such complications .There could be 95% survival in the pump twinwith an average age at delivery between 36 and37 weeks6. The prenatal diagnosis of TRAP sequence enables both invasive andconservative treatment to be offered to the mother toreduce the amount of compromise of the pump twin7.

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