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 chorioangiopagus parasiticus. It occurs in 1% of monochorionic twin pregnancies and in 1 in 35,000 pregnancies. TRAP sequence is characterized by a structurally normal pump twin perfusing an anomalous twin. In TRAP sequence, one twin is usually developmentally normal (pump twin) and the other twin has a serious condition like missing heart (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 blood enters the acardiac/acephalic twin through reversed flow through its umbilical artery and exits through the umbilical vein, which is opposite to the normal blood supply of the fetus. The mortality of the acardiac twin is 100%, and the perinatal mortality of the pump twin is reported to be around 50%. TRAP sequence was diagnosed by ultrasound at SAH & RC ultrasound centre and referred for management.

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

I. Introduction

Acardiac Twin or TRAP Sequence is a very rare condition with an incidence of 1 in 35,000 pregnancies, occurring in approximately 1% of monochorionic twin pregnancies. One twin is structurally normal and is referred to as pump twin because it 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 the abdomen of the acardiac fetus allowing some 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 monochorionic diamniotic diagnosed at 16th week scan was been followed in our hospital and diagnosed as TRAP when sent for anomaly scan at SAH & RC.
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 perfusion of umbilical artery ratio 2.49.

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 500 gm 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.
III. Discussion

Twin reverse arterial perfusion syndrome (TRAP) was first defined by Grunewald in 1942. TRAP syndrome occurs in monochorionic gestation form. In which case one twin has Acardia (the recipient) with no heart and the other twin is structurally normal (pump or donor twin). Due to the absence of heart in the acardiac twin the pump twin supplies deoxygenated blood via vascular anastomoses to the acardiac twin. Acardia twin does not send blood to placenta and all its blood comes from and goes back to the circulation of the pump twin, through the vascular connections on the surface of the shared placenta. It is therefore structurally normal twin perfusing an anomalous recipient twin via an artery-to-artery anastomosis in a reverse direction. Thereverses flow is through its umbilical artery and exits through the umbilical vein which isopposite to the normal blood supply of the fetus. Deoxygenated low-pressure blood from the pump twin which would normally return to the placenta, instead flows directly to the acardiac twin, resulting in a wide array of structural abnormalities, caused by arterioarterial and veno-venous placental anastomoses. The acardiac twin is usually grossly abnormal with severe reduction anomalies of the upper part of the body as seen in our present case. Mortality of the pump twin is 50-75% usually due to the result of heart failure and ofthe acardiac is 100%.

Schatz (1898) classified acardia into two main groups: Hemiacardius (imperfectly formed heart) and Holoaacardius (absence of heart). The first type is Acardiusacephalus, where no cephalic structures present. (head & upper extremities are lacking. It is most common variety. The second is acardius anacorpus where some cranial structure and neural tissue or brain tissue is present. The body and extremities are also developed. The third is acardius acorvulus with cephalic structure but no truncal structures are present. i.e., Head without a body. The umbilicalcord is attached to the head. It is rarest form of Acardia. The fourth type is Acardius amorphus without distinguishable cephalic or truncal structure. It is least developed and not recognizable as human form with minimal development. This differs from Teratomas only by its attachment to an umbilical cord. (3 and 4). Therapeutic options targeted at interrupting the vascular anastomosis between the twins under ultrasound guidance using fetoscope. Several different techniques have been used to treat TRAP sequence by interrupting the connection between the acardiac twin and the pump twin to increase the chances that the pump twin will survive. These techniques include cord occlusion by embolization, ligation, laser photocoagulation, monopolar and bipolar diathermy. Intrafetal ablation has also been performed by alcohol injection, monopolar diathermy, interstitial laser, and radiofrequency ablation (RFA). It is also important to exclude a chromosomal abnormality prior to offering a fetoscopic procedure in TRAP sequence since the incidence of chromosomal abnormality in the pump twin may be as high as 9 percent.

IV. Conclusion

Accurate antenatal diagnosis is essential to improve the prognosis of this rare entity of TRAP sequence. Improved imaging techniques like 2D ultrasoundography, 3D ultrasonography and transvaginal Doppler ultrasonography have made the diagnosis of Acardia possible even in the first trimester of pregnancy by detecting in vivo flow in the recipient acardiac fetus. Early diagnosis may help to reduce the risk of such complications. There could be 95% survival in the pump twin with an average age at delivery between 36 and 37 weeks. The prenatal diagnosis of TRAP sequence enables both invasive and conservative treatment to be offered to the mother to reduce the amount of compromise of the pump twin.

References

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