

Case Report

Acardiac Acephalus Twin diagnosed at delivery: A case report

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ABSTRACT

Abnormal placental vascular anastomoses in monochorionic twins can give rise to a rare congenital anomaly known as Acardiac twin. With the advancement of antenatal care and ultrasonography many such cases are diagnosed early. However, in underdeveloped country like Nepal where early antenatal care coverage rate is low such cases may be diagnosed late. I report one such case of Acardiac acephalus twin diagnosed during delivery.

Key Words: Acardiac acephalus twins, Monochorionicity, Twin Reversed Arterial Perfusion

INTRODUCTION

Twin-Reversed Arterial Perfusion (TRAP) Sequence or *Acardiac twin* is a rare condition complicating about 1% of monochorionic twins pregnancies and 1 out of 35000 deliveries [1]. Multifetal pregnancy is an obstetric condition that present high risks both for the mother and the fetuses. Likely complication for the unborn children range from spontaneous abortion, congenital malformations to lifelong disabilities. As the number of fetuses increases, chance of favourable outcome decreases. Twin fetuses can result from fertilization of two separate ova - Dizygotic or Fraternal twin and less commonly by the division of a single fertilised ovum-Monozygotic or Identical twin [2]. The twinning mechanism may be aberrant giving rise to many types of malformations. Even though usually incomplete splitting of the embryo is considered as the main cause of

aberration, there is a possibility of early secondary fusion of two separate embryos. A peculiar feature that may develop in a monozygotic twin is *monochorionicity*. In this condition the twins develop in two separate amniotic sacs but have a common surrounding chorion. A single chorion leads to the anatomical sharing of the two fetal circulation in the form of artery-to-artery and vein-to-vein anastomoses. Monochorionic twins present 20% of all twins and have higher chances of complications due to this sharing of circulation. In monochorionic twins pregnancy there is 6 times higher chance of fetal demise between first trimester and 24 weeks in comparison to a dichorionic twin pregnancy [3]. Due to the circulation anastomoses there develops a rare condition known as Twin-Reversed Arterial Perfusion (TRAP) Sequence. In the TRAP sequence there is usually a normally developed fetus along with another fetus that lacks heart and

other structures. The normally developed donor companion of recipient acardiac twin usually shows features of heart failure. In TRAP sequence, deoxygenated blood flows from the umbilical artery of the pump twin in a reversed direction into the umbilical artery of the perfused twin, via an artery-to-artery anastomosis and usually returns via a vein-to-vein anastomosis back to the pump twin. The perfused twin is thus a true parasite and does not have any functional cardiac activity, hence also the name "Acardiac twin" [3]. The deoxygenated blood goes preferentially to the iliac vessels of the recipient twin. This preferential blood supply leads to the perfusion of the lower body and disrupted growth and development of the upper body structures. Failure of the development of head results in the "Acardiac acephalus" twin. The condition is associated with a high risk of perinatal death caused by a combination of high-output cardiac failure in the donor twin and polyhydramnios - related preterm birth [4].

CASE REPORT:

Mrs AK, 19 years old Primigravida presented to the Emergency room of the hospital with the chief complaint of amenorrhoea for 8 months, pain abdomen and watery discharge per vaginum for 4 hours. She was not sure of her Last Menstrual Period and had a single antenatal checkup 3 weeks back before coming to our hospital. The scan done by the ultrasonologist during antenatal checkup showed a single live fetus of 35 weeks in cephalic presentation with adequate liquor weighing 2530 grams.

On examination the lady had a Blood Pressure of 120/80 mm of Hg. Her uterus was Term size and head was engaged with a regular FHR of 139/mt. Uterine contractions were

mild. In P/V examination the os was 4 cm dilated with 50% effacement. Head was at-1 station. The membrane was absent and liquor was clear. A scan was performed by the ultrasonologist and it showed a single foetus with Amniotic Fluid Index of 9.3. Augmentation of labor was decided with oxytocin. After 4 hours of augmentation no progress in labor was noted and a decision to perform emergency cesarean section was made. A male baby of 3 kg was delivered with an Apgar score of 8/10 and 9/10. While removing the placenta a fleshy mass was noted along with the placenta which resembled lower half of a fetus. This foetus has no head, well developed lower limbs, underdeveloped male genitalia and a short umbilical cord (Image 1).



Image: 1

X-ray revealed well developed bones of the left leg and femur, metatarsals and phalanges in the right leg. X-ray also showed unfused pelvic bones (Image 2). Placenta weighed 550 gms and had one normal and another short cord (Image 3). The healthy twin had no signs of heart failure (Image 4). Both mother and baby had an uneventful post op. period. BP of the mother rose to 140/80 mm of Hg on two occasions in first 24 hours of surgery but

remained in the range of 110-120 upon 70-80 mm of Hg in the remaining hospitalized days. Mother and baby were discharged on 7th post op day. A follow up visit after one week showed no maternal or fetal complication.



Image: 2



Image: 3



Image: 4

DISCUSSION

Monochorionicity is best diagnosed in first trimester. Early diagnosis of monochorionic twins provides enough time and opportunity to an obstetrician for identification and management of monochorionic twins related complications.

Various authors recommend early diagnosis of TRAP and applications of interventions to arrest the circulation of the acardiac twin and thereby improving the outcome of donor cotwin [5, 6]. Late diagnosis with compromised twins may be an indication for termination of pregnancy in view of unfavourable outcome for unborn children and mother [7, 8].

CONCLUSION

Finally, In underdeveloped countries there is a need to encourage mothers and their families for early antenatal checkup. Radiologists / Ultrasonologists need to be vigilant to pick any abnormal finding. Such efforts will provide both the mother and the unborn children a better prognosis.

REFERENCES

1. James WH. A note on the epidemiology of acardiac monsters. *Teratology* 1977; 16: 211-216.
2. Cunningham FG, Levono KJ, Bloom SL, Spong CY, Dashe JS, Hoffman BL, Casey BM, Sheffield JS. *William's Obstetrics*. 24th ed. McGraw-Hill Education; 2014.
3. Lewi L, Deprest J, Hecher K. The vascular anastomoses in monochorionic twin pregnancies and their clinical consequences. *Am J Obstet Gynecol* 2013; 208(1):19-30.
4. Moore TR, Gale S, Benirschke K. Perinatal outcome of forty-nine pregnancies complicated by acardiac twinning. *Am J Obstet Gynecol* 1990; 163: 907-12.

5. Hecher K, Lewi L, Gratacos E, Huber A, Ville Y, Deprest J. Twin reversed arterial perfusion: fetoscopic laser coagulation of placental anastomoses or the umbilical cord. *Ultrasound Obstet Gynecol* 2006; 28: 688-91.

6. Lee H, Wagner AJ, Sy E, et al. Efficacy of radiofrequency ablation for twin-reversed arterial perfusion sequence. *Am J Obstet Gynecol* 2007 ;196:4591.e1-4.

7. Rohilla M, Chopra S, Suri V, Aggarwal N, Vermani N. Acardiac- Acephalus Twins: A Report of 2 Cases and Review of Literature. *Medscape J Med* 2008; 10(8): 200.

8. Sogaard K, Skibsted L, Brocks V. Acardiac twins: pathophysiology, diagnosis, outcome and treatment. Six cases and review of the literature. *Fetal Diagn Therapy* 1999;14: 53-59.

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