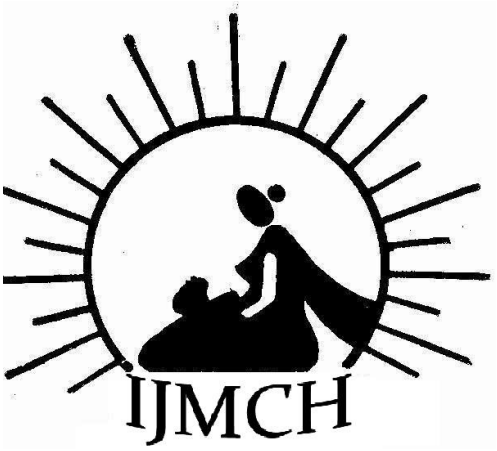
Volume 14 (2), 2012

A Case of Acardius Anceps with PPROM and Breech Presentation – A Successful Conservative Approach

Kaur Manpreet
Patil Neelamma
BI Surekha

www.ijmch.org



INDIAN JOURNAL OF MATERNAL AND CHILD HEALTH

Acardiac twinning occurs once in 35,000 births & 1 in 100 monozygotic twins. Though it is so rare, it presents with different types of complications which put the managing obstetrician in dilemma.

CASE REPORT

A Case of Acardius Anceps with PPROM and Breech Presentation – A Successful Conservative Approach

Kaur Manpreet,*# Patil Neelamma,# BI Surekha***#**

***Professor, **Associate Professor, ***Consultant radiologist, #Department of Obstetrics & Gynecology, BLDE University's Shri B.M. Patil Medical College Hospital and Research Centre, Bijapur. #Sai Scanning Center, Bijapur.**

Corresponding Author: Dr. Manpreet Kaur,

E mail: dr_mjtehalia@yahoo.co.in

ABSTRACT

Acardiac twinning occurs once in 35,000 births & 1 in 100 monozygotic twins. Though it is so rare, it presents with different types of complications which put the managing obstetrician in dilemma. We had a case of acardiac twin pregnancy with PPROM with breech presentation of first normal twin which was managed conservatively with minimal risks to the normal twin. This case is being described in a setting where cesarean sections for very preterm babies are many times deferred because of limitations of neonatal care facilities and financial constraints.

Key words: ***Multiple gestations, Acardiac twin, PPROM***.

INTRODUCTION

Acardiac twinning is an obstetric challenge. It is a rare development, occurring once in 35,000 births, or 1 in 100 monozygotic twins[(1)] and constitutes a group of the most bizarre malformations in human conception. That apart, the management dilemma that an acardiac twin pregnancy poses makes it an interesting case report. This case is being reported not only for its rarity, but also for the additional obstetric complications that it presented with, that is, PPROM (32 weeks) with breech presentation of first, normal twin. This case is being described in a setting where cesarean sections for very preterm babies are many times deferred because of limitations of neonatal care facilities and financial constraints.

CASE REPORT

A 22 yr primigravida with spontaneous conception and seven months pregnancy, presented with mild pain abdomen & PV leak since 4 hrs. On examination uterus was over distended (34wks size) & relaxed. Multiple fetal parts were palpable & one fetal pole had entered the pelvis. Only one FHS could be localized. Speculum examination showed leakage of clear liquor. P/v examination revealed a closed, partially effaced cervix with breech at o station. Her previous scan had given a diagnosis of an MCDA (Monochorionic, Diamniotic) pregnancy, with an anencephalic, hydropic, nonviable fetus, along with a second alive, normal fetus of 11 weeks 6 days. A second scan had diagnosed a fetus papyraceous with a normal second twin of 20 weeks gestation. A rescan now showed twin fetuses, single fundal placenta, a normal alive twin in breech of 32 weeks gestation and a second, acardiac twin with mild polyhydramnios.

After detailed discussion with the patient regarding perinatal outcome the patient was admitted for conservative management. Complete bed rest with foot end elevation, i/v antibiotics (Ampicillin, Metronidazole, Gentamycin), Injection Hydroxyprogesterone Caproate 500 mg i.m. weekly and prophylactic steroids were instituted. Routine blood and urine investigations were within normal limits. CRP was negative.USG after one week showed severe oligohydramnios, no organomegaly or ascites in the normal twin. It was decided to continue pregnancy upto 34 weeks of gestation, if possible. Daily clinical monitoring for chorioamnionitis and fetal compromise was done.CRP was repeated twice & was negative. P/v examination on day four revealed Cervix one finger tight, 80% effaced, with breech at +1 station. The stay of the patient was uneventful except for continued leakage. At 34 weeks completed, that is, ten days of conservative management, the patient was induced with 25 mcg. Misoprostol vaginally, followed by Oxytocin. Patient delivered after seven hours of induction. An alive, preterm, female baby of weight 1.8 kg delivered as assisted breech & an acardiac twin weighing 1.3 kg by breech extraction, five minutes later. The first twin appeared normal in all aspects, but because cardiac failure is possible even post delivery, was admitted to NICU for observation. Congestive cardiac failure developed on the third day & was managed conservatively. Baby was discharged on day 10 in a satisfactory condition. Baby had normal development on follow up examination upto 1 yr.

The acardiac twin was 24 cm in length with Twin Weight Ratio(TWR) of 68%. Lower limbs were well formed upto mid-calf and absent below. External genitalia appeared normal (female). Anal opening was absent; a hair covered area on top of a stunted trunk was present. Rudimentary buds, probably representing facial structures were present. A thin, friable, two vessel cord was found attached in the area of apparent chest. The attachment

of the acardiac twin's cord, either to a vessel in the placenta or directly to the pump twin's cord (the acardiac twin has no villus circulation of its own), could not be ascertained, as the entire length of 9 cm came out with the fetus. Autopsy showed absent heart, lungs and liver while hypoplastic intestines, kidneys, pancreas and spleen were present. Microscopy revealed normal layers of intestine with congestion of spleen. Unfortunately, the placenta could not be sent for pathological examination.

DISCUSSION

Monozygotic twinning (MZ) occurs in 3.5/1000 deliveries,(2) with evidence showing its increasing incidence with ovulation induction(3) and assisted reproduction.(4) MZ twinning may result in symmetric division forming either normal, "identical" twins, or conjoint twins. Asymmetric twinning may lead to the formation of Endoparasites or the fetus in fetu, Ectoparasites, which need to be differentiated from fetiform teratomas, and finally, the Acardiac twins. Various terminologies are in use for the anomaly-CAPP or Chorioangiopagus parasiticus twin, which implies that there is conjunction in the chorionic circulation in these twins. This term has been suggested to be the most accurate and inclusive term for this abnormality. It is also known as Acardiac twin, but evidence of a rudimentary heart has been found in some cases.(5) Lastly, the TRAP sequence, which describes the entry of deoxygenated blood from the pump twin into the abnormal (parasitic) twin through its umbilical artery, that is, a reversal of flow in the artery, and exit through the umbilical vein.

The various theories put forward for the CAPP twins are the Primary Developmental theory and the Secondary vascular destructive theory.[5] According to the first, it is the failure of development of the heart, the absence of an umbilical artery or the presence of chromosomal abnormalities which may be responsible for this anomaly; but rudimentary heart, normal three vessel cords and normal chromosomal patterns have been reported in some cases. The second postulates that because the abnormal twin gets perfused with deoxygenated blood from the pump twin, the resulting tissue hypoxia causes the resorption of the already formed tissues. As the lower parts of the body receive the blood earlier, the caudal end of the fetus is slightly better developed than the cephalic tissues.

The physical abnormalities present in the acardiac twin form the basis for the classification of such twinning.[6] The simplest classification includes Acardiac anceps (some cranial structures present), Acardius acephalus, Acardius acormus and Acardius amorphous. The acardiac twin is often hydropic; lower limbs are relatively well developed; variable perineal structures are present and a trunk with a normally situated cord maybe present. The upper body is replaced by a dome like structure, and rudimentary arms and buds representing facial structures may be present. Brain development is absent or minimal, but recently, an acardiac twin with an almost fully developed brain has been described.[7] X-ray may show corresponding structures. On autopsy, thoracic and visceral structures are varyingly absent or rudimentary. The outcome is uniformly fatal for this twin. The brunt of the abnormality is borne by the pump twin. This twin is pumping blood to maintain two circulations. Hence, there is first the development of a high cardiac output and increased renal perfusion, leading to the development of polyhydramnios. Eventually, RVH develops, leading to cardiac failure, pericardial effusion, hepatosplenomegaly and ascites. Subsequently, hydrops due to cardiac overload and hypoproteinemia occurs. Perinatal mortality in the pump twin ranges from 50 – 75 %.[8]

Moore *et al*[9] reported factors for a favorable outcome in a study of 49 cases of acardiac twins and concluded that more the TWR (ratio of the wet weight of the acardiac twin to weight of pump twin), worse is the prognosis. With TWR > 70% , the incidence of preterm delivery is 90%, hydramnios 40% and pump twin CCF 30%;with TWR < 70%, it falls to 75%, 30% and 10% respectively. A rough estimate of the weight of the acardiac twin can be made by comparing the ratio of the perimeter of the acardiac and pump twin or applying the formula of the prolated ellipsoid, that is, length of acardiac twin times abdominal diameters, divided by 2.[10] Dashe *et al*[11] described the utility of Doppler velocimetry in predicting the outcome in TRAP sequence in six cases. They concluded that pump twins with small resistive indices difference with the acardiac twin, had poorer outcomes

Different management strategies for reducing the morbidity and mortality of the pump twin, both interventional and expectant, have been reported. The indications for intervention are when the acardiac twin continues to grow, the gestation is previable and the pump twin either develops polyhydramnios or cardiac failure. The various methods tried include ligation, laser[12] and radiofrequency ablation[13] of the acardiac umbilical cord under fetoscopic guidance, embolization of circulation with fibrin or glue,[14] placement of steel coils in the umbilical artery, close to the abdominal wall of the acardiac twin,[15] thermo coagulation and "sectio parva".[16] Expectant management of the acardiac twin pregnancy includes close clinical, ultrasonographic and Doppler velocimetric monitoring, maternal administration of digoxin and indomethacin[17] and serial amniocentesis, to improve prognosis. Sullivan *et al*[8] described a case series of 10 acardiac twins and highlighted the success of expectant management in these with close antepartum surveillance. Our case, too, highlights the potential for the conservative management of acardiac twinning in resource limited settings.

CONCLUSION

Previously, interventional management was considered as the appropriate approach to improve the prognosis of the pump twin. Lately, the focus is shifting to expectant management. Close monitoring of such a pregnancy can improve prognosis without procedures for which expertise and specialized equipment is neither cheap, nor easily available and where limitation of perinatal and neonatal care make very early termination of pregnancy, especially by the cesarean route, an undesirable option.

REFERENCES

1. Thelmo MLC, Fok RY, Shertukde SV. Acardiac twin fetus with severe hydrops fetalis and bilateral talipes varus deformity. Fetal Pediatr Pathol 2007;26(5-6):235-42.
2. Gall SA. Epidemiology. In: Patterson AS, ed. Multiple Pregnancy and Delivery, Chapter 1.Missouri: Mosby-Year Book, Inc., 1996.
3. Derom C, Vlientinck R, Derom R, Van Den Berghe H, Thiery M. Increased monozygotic twinning rate after ovulation induction. Lancet 1987;1(8544): 1236-8.
4. Wenstrom KD, Syrop CH, Hammit DG, Van Voorhis BJ. Increased risk of monochorionic twinning associated with assisted reproduction. Fertil Steril 1993;60(3):510-4.

5. Baldwin VJ. Anomalies of Monozygotic Duplication. In: Pathology of Multiple Pregnancy, Chapter 10.Springer-Verlag New York Inc., 1994.
6. Smith J, Gilson G, Del Valle G, Joffe G, Chatterjee M *et al*. Twin, acardiac, acephalus.TheFetus.net.1991;1:3-19.
7. Peterson BL, Broholm H, Skibsted L, Graem N. Acardiac Twin with Preserved Brain. Fetal Diagn Ther.2001; 16:231-3.
8. Sullivan AE, Varner MW, Ball RH, Jackson M, Silver RM. The management of acardiac twins: A conservative approach. Am J Obstet Gynecol 2003; 189:1310-3.
9. Moore TR, Gale S, Benirschke K. Perinatal outcome of forty-nine pregnancies complicated by acardiac twinning. Am J Obstet Gynecol 1990; 163:907-12.
10. Grab D, Schneider V, Keckstein J, Terinde R. Twin, acardiac, outcome. Fetus 1992; 2(1):11-3.
11. Dashe JS, Fernandez CO, Twickler DM. Utility of Doppler Velocimetry in predicting outcome in twin reversed-arterial perfusion sequence. Am J Obstet Gynecol 2001; 185:1-7.
12. Sepulveda W, Hasbun J, Dezerega V, Devoto JC, Alcalde JL. Successful sonographically Guided Laser Ablation of a Large Acardiac Twin at 26 weeks Gestation. J Ultrasound Med 2004; 23: 1663-1666.
13. Tsao K, Feldstein VA, Albanese CT, Sandberg PL, Lee H et al. Selective reduction of acardiac twin by radiofrequency ablation. Am J Obstet Gynecol 2002; 187:635- 40.
14. Holzgreve W, Tercanli S, Krings W, Schuierer G. A simpler technique for Umbilical Cord Blockade of an Acardiac Twin. New Engl J Med.1994; 331:56-7.
15. Hamada H, Okane M, Koresama M, Kubo T, Iwasaki H. Fetal Therapy in utero by blockage of the umbilical blood flow of acardiac monster in twin pregnancy. Nippon Sanka Fujinka Gakkai Zasshi.1989; 41(11):1803-9.
16. Robie GF, Payne Jr GR, Morgan MA. Selective Delivery of an Acardiac Acephalic Twin. N Engl J Med 1989; 320(8):512-3.
17. Simpson PC, Trudinger BJ, Walker A, Baird PJ. The intrauterine treatment of fetal cardiac failure in a twin pregnancy with an acardiac, acephalic monster. Am J Obstet Gynecol 1983; 147(7):842-4.

Photograph: Acardiac baby with normal twin