



## Discordant Twin Pregnancy with One Malformed Fetus – Color Doppler Follow Up for Expectant Management

## KEYWORDS

Discordant, acardia, twin

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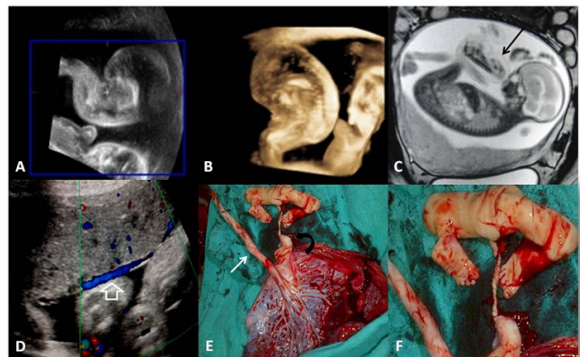
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**ABSTRACT** Monochorionic twin discordant with congenital anomalies introduce additional burdens to obstetricians in their endeavor to save the normal co-twin. The wide array of treatment options that have been described all incur significant morbidity and none of them are unequivocally optimal. Whether to embrace the antepartum use of aggressive intervention techniques or take the conservative anticipative approach remains a topic of debate and clinical dilemma. In our view, a serial evaluation of the color Doppler parameters in such patients might help us to triage the appropriate candidates for expectant management and lead to reduced rate of overtreated patients. In this study, we describe our experience with monochorionic twinning discordant with fetal acardia and anencephaly managed conservatively through color Doppler surveillance

## INTRODUCTION

Monochorionic twinning presents unique sets of challenges to obstetricians. Of the numerous complications that attend multiple gestation premature delivery, preeclampsia, polyhydramnios, placental abnormalities and postpartum hemorrhage are the most common [1]. Monochorionicity of twins prompt other complications as well, many of which are related to complex placental vascularity and development of intertwin blood vessel anastomoses. Three types of anastomoses have been recognized – the arterio arterial (AA), veno venous (VV) and the deeper arterio venous (AV) communications. Flow in the arteriovenous anastomoses is unidirectional and their presence along with the absence of compensatory AA or VV anastomoses can lead to Twin Twin Transfusion syndrome (TTTS) [1]. The association of monochorionic twin gestation with congenital anomalies in any one of the fetus, can be highlighted by the fact that observations on monochorionic twins have led to better understanding of many congenital anomalies [1]. Presence of structural anomalies puts the apparently normal co-twin to additional potential risks. Anencephaly, for instance, is associated with development of polyhydramnios, premature rupture of membranes and intrauterine death of the normal twin [2]. Another concern is discordant growth leading to TTTS. Fetal acardia, another rare anomaly, is seen only with multiple pregnancy and has an incidence of 1/34,600 births [3]. A consistent feature of this condition is presence of artery to artery anastomoses within fused placenta which leads to reversed arterial perfusion of the acardiac “perfused twin” by its sibling, the “pump twin”. The reported mortality of the normal pump twin is extremely high (50 -70%) and has been attributed to increase cardiac overload on pump twin in an effort to perfuse its acardiac sibling [4,5]. Presence of such anomalies raises a clinical dilemma, as to which is the best management strategy to minimize the potential risk to normal twin. The reports of increased mortality of the normal co-twin have prompted the use of invasive techniques intended to selectively occlude umbilical flow to acardiac twin or selective feticide of anencephalic twin. However selective feticide after 16 weeks of gestation has been associated with increased risk of miscarriage

[2]. Procedures described to selectively occlude the umbilical vessels to acardiac twin all incur significant morbidity [6]. Thus expectant management of such cases by color Doppler monitoring should be discussed and emphasized widely. In this article, we describe our experience with conservative management in cases of monochorionic twin, discordant with anencephaly and fetal acardia, which followed favorable outcome on serial color Doppler surveillance.

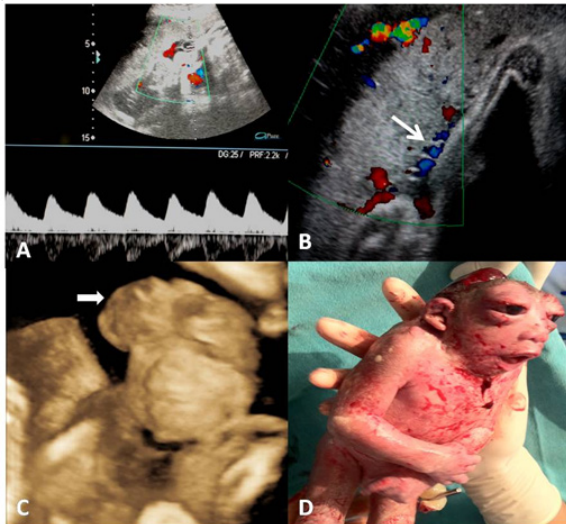
CASE REPORT  
PATIENTS

**Patient 1 (Figure 1)**

A 26 years female, G2 P1+0L1 with a previous history of lower segment caesarean section presented to department of obstetrics at 21 weeks of gestation for routine antenatal check up. The present pregnancy was a spontaneous conception and patient had no history of oral contraceptive pill intake or family history of congenital disorders. Ultrasound revealed monochorionic monoamniotic gestation with acardiac twinning. The normal twin showed sonographic gestational age corresponding to clinical gestational age with fetal heart rate of 163 beats per minute. On color Doppler interrogation, fetal umbilical and middle cerebral arteries showed normal flow velocity waveforms and Doppler indices. USG of acardiac twin showed absence of head and thorax with

deformed bones. Extensive subcutaneous edema was noted with talipes equinovarus deformity. Estimated fetal weight by abdominal girth was approximately 1/10<sup>th</sup> of the pump twin. The findings were confirmed by T2 weighted MRI sequences. The twins had a fused anteriorly placed placenta, and the amniotic fluid was adequate. Power Doppler revealed chorionic plate vessels crossing the placenta which showed bidirectional spectral waveforms on pulse wave interrogation suggestive of arterioarterial anastomoses. No arteriovenous anastomoses could be identified. In view of late presentation, normal color Doppler indices expectant management was planned with color Doppler every three weeks and cardiocotography (CTG) every two weeks. Follow up scan did not show any evidence of cardiac overload, intrauterine growth retardation or polyhydramnios in the normal twin.

Various Doppler parameters were monitored at 3 week intervals. Table 1 summarizes the umbilical artery pulsatility index of cases at first, penultimate and the last Doppler examination which shows significant difference in Pulsatility index between pump and acardiac twin. Also the twin weight ratio remained at approximately 1:10. The pulse wave analysis of arterioarterial anastomoses did not show significant change in peak systolic velocities. These factors instigated us towards expectant management. Patient had scar tenderness at 37 weeks gestation, and caesarean section was performed. She delivered the 1st twin by breech. The surviving male baby weighed 2980 mg at birth with no obvious congenital anomaly and cried immediately after birth. Second twin was delivered along with delivery of placenta. Postnatal period was uneventful. Twin 1 was active; accepting feeds well, alive and well at time of discharge. The acardiac twin weighed 280 mg and a plain radiograph of acardiac twin was taken which showed both femurs and incompletely formed pelvic bones.



**Patient 2 (Figure 2)**

A-27-year old woman presented to obstetric department for regular antenatal check up at 25 week gestation. Sonography revealed mono chorionic monoamniotic twin gestation discordant with anencephaly. The biometric development of the normal twin was concordant to clinical age (25 weeks) with no obvious fetal structural abnormality. The co-twin was anencephalic and of 24 weeks by biometry. Polyhydramnios was present. Fetal heart rates of both fetuses were within normal limit (140 and 134 beats /min respectively). Color Doppler was performed which showed normal umbilical artery and ductus venosus spectral waveform in both twins. Color Doppler also revealed several chorionic plate vessels with aliasing artifacts suggesting bidirectional flow. Pulse wave imaging showed bidirectional waveforms suggesting arterioarterial anastomoses. No deeper vascular anastomoses could be identified. MRI was also performed which showed no obvious structural abnormality in the normal twin. The couple was

informed, in view of late presentation, normal color Doppler indices and absence of vascular anastomoses, expectant management was planned with color Doppler every three weeks and CTG every other week. Despite the development of polyhydramnios on follow up scans, this patient was managed conservatively. The amniotic fluid index during successive examinations remained between 25-28. The decision in favor of expectant management, rather than amniocentesis was based on the non progression of the polyhydramnios, both in terms of the discomfort to the patient and based on amniotic fluid index calculation. Also no significant discordance between pulsatility indexes was noted between the twins. At 38 weeks of gestation, caesarean section was performed because of institutional protocol to operate all the cases of mono chorionic pregnancy. The anencephalic infant died soon after birth (birth weight 1300 gram). The surviving infant cried immediately after birth and had normal apgar score. The infant was accepting feeds normally and weighed 2510 gr. The infant was discharged on 7<sup>th</sup> day and was followed up monthly for initial 3 months and showed normal growth.

## METHODS

Fetuses were first assessed on gray scale ultrasound using a 3.5 MHz curved electronic array transducer (IU-22, Philips medical systems, The Netherlands and Xario, Toshiba corp inc., Japan) for biparietal diameter, head circumference, abdominal circumference and femur lengths. In case of anencephalic twin, any discordance in abdominal circumference and femur lengths between the twins were taken note of. Amniotic fluid indices were followed on serial examinations. Fetal bladder was examined for over distension in first (polyuric recipient twin) and collapse in the other (donor twin) to rule out twin twin transfusion. Fetal heart was carefully examined for signs of failure i.e. presence of dilated and hypertrophied chambers.

Next, Doppler examination of placenta was done in each cases for identification of communicating placental vessels and use of spectral Doppler was done for characterization of the nature of such communications (arterioarterial, veno venous or arterio venous anastomoses). Careful assessment of umbilical artery flow was done regarding direction of flow to exclude reversed flow towards acardiac twin from placenta. The level of intertwin discordance in pulsatility index or umbilical artery impedance was recorded. Waveforms of fetal middle cerebral artery and ductus venosus were recorded to look for reversal of flow. MRI was performed on a 1.5 T superconducting system (Magnetom Avanto, Siemens Medical System, Erlangen, Germany) equipped with an actively-shielded whole body magnetic field gradient set using a large size receive and transmit radio frequency body coil.

**Table 1: Pulsatility Indices of Umbilical Arteries of the Normal Fetus and abnormal fetus (acardiac fetus in case 1 and anencephalic fetus in case 2)**

Case		PI (N)	PI (A)	PI(A/N)
1	1st Doppler examination	1.42	1.80	1.26
	Penultimate Doppler examination	0.80	0.99	1.23
	Last Doppler examination	0.77	1.06	1.37
2	1st Doppler examination	1.64	1.52	0.92
	Penultimate Doppler examination	0.90	0.83	0.92
	Last Doppler examination	0.96	0.93	0.96

N = normal fetus; A = abnormal fetus; PI = pulsatility index.

## DISCUSSION

Twins are frequently discordant in development but discordance with fetal congenital anomalies is higher in monozygotic pregnancy. Mono chorionic placentas virtually always come from MZ (identical) twins. Most of the discordance can be attributed to unique placental vascular relations between the twins in mono chorionic gestation. Various intertwin vascular

anastomoses of monochorionic twin placentas are perhaps the most important determinant to predict perinatal outcome in any twin gestation. This holds true for twinning discordant with congenital anomalies as well [1]. Modern Doppler Sonographic studies have come closer to understand the intra-placental vascular distribution patterns and changes inflicted in umbilical cords. Hecher et al studied 126 cases of severe TTTS through Doppler sonography and surgical findings and were able to demonstrate AV anastomoses in all of them [7]. Smith et al. (1997) provided Doppler study evidence of "improved hemodynamics" after many amnioreductions which highlights the role of Doppler as a modality to monitor the fetoplacental distribution changes [8]. Although, it would be an oversimplification to assume that the changes observed in Doppler studies will truly and always predict the intertwin vascular hemodynamics, still it is safe to state that the Doppler observations can alert the obstetricians for impending adverse perinatal outcomes. In our study, we were able to demonstrate arterio arterial (AA) anastomoses in both the cases, identified by their characteristic bidirectional spectral waveforms on pulsed wave Doppler imaging. Arterio venous connections could not be identified in either of the cases. Presence of arterio arterial anastomoses in the twin discordant with anencephaly without arteriovenous (AV) connections went some way in reassuring us that favorable outcome could be anticipated and that risks of twin twin transfusion did not apply to our case. Hence, findings in initial Doppler studies prompted us towards conservative management. In acardiac twin, the flow in arterioarterial anastomoses was monitored which did not revealed significant increase or decrease in systolic velocities.

The prevalence of anencephaly in twins is higher than in singletons (10.4/10,000 in twins compared with 2.8/ 10,000 in singletons) and the prevalence of discordance for anencephaly is higher in monochorionic than in dichorionic twins [11]. The primary prognostic factor associated with twin pregnancies discordant for anencephaly is the development of preterm delivery due to the associated polyhydramnios which is thought to be caused by impaired swallowing reflex of the anencephalic fetus. Abrupt, uncoordinated movement of anencephalic fetus can lead to stretching and compression of the umbilical cord of the normal fetus. Other important risk factors are twin twin transfusion syndrome and intrauterine death of the anencephalic twin which leads to acute hemodynamic imbalances in the anastomotic vessels influencing prognosis of the surviving twin. Dichorionic twins with discordant anencephaly can be treated by selective fetocide however monochorionic twin pregnancies show the presence of communicating intertwin vessels. Consequently, selective fetocide can lead to transplacental passage of the injected potassium chloride or acute exsanguinations through the vascular anastomoses into the placenta of the dead anencephalic fetus [2,9,10]. Conservative management and selective fetocide by ligation of the umbilical cord or endoscopic laser coagulation of the umbilical cord vessel are the present treatment options. In our case, although patient developed polyhydramnios, the amniotic fluid index did not show significant increase during serial examinations. Also, the Doppler examination showed normal flow patterns in both cords with concordant peak systolic velocity and pulsatility index. Further, on 3D Sonographic monitoring of the cords, no evidence of any abnormal stretching of the cords was recorded. Managed expectantly, both the twins survived up to the terms when caesarean section was carried out with favorable outcome for normal twin.

Fetal acardia characterized typically by absence of fetal heart or occasionally presence of a severely malformed heart, has been described only with multiple gestation. The fact that an acardiac can develop at all is due to the presence of AA and VV anastomoses in the monochorionic placenta which brings and returns blood from pump to recipient and back to the pump twin respectively. The reversal of blood flow in recipient umbilical arteries from placenta has been proved to

exist with the use of Doppler sonography. The management of such complex anomalous condition is still a clinical dilemma for which no definite criteria have been established. Traditionally, the reported mortality of the normal twin in such cases has been 50-70% which has convinced many authorities towards active intervention to save the pump twin. Wide range of treatment options have been described ranging from selective removal of acardiac twin to various methods to interrupt umbilical artery flow (occluding metal coil, thermo coagulation, endoscopic ligation, bipolar cauterization, percutaneous injection of alcohol under Sonographic guidance, monopolar diathermy, and interstitial laser and radiofrequency ablations) to acardiac twins a conservative approach to the treatment. However, presence of such wide array of treatment option implies that none of these methods have been unequivocally described as optimal [1,11,12]. Sullivan et al (2003) studied 10 patients managed with conservative approach and inferred that mortality rate of antenatally diagnosed pregnancies complicated by TRAP sequence may be less than previously reported (10% in their study) and use of intervention techniques is therefore not justified [6]. Various methods to identify the impending risk to the normal twin have been proposed. One such method is using the ratio of the estimated weight of the acardiac twin to the pump twin (the twin-weight ratio) to guide management. A twin weight ratio greater than 50% has been associated with an increase in the risk of fetal hydrops prematurity [4]. Another method is the use of Doppler velocimetry. Greater differences in the (resistive index and pulsatility index) values between the acardiac twin and the pump twin have been associated with a more favorable pregnancy outcome [14]. In our case the twin weight ratio was approximately 1/10 in successive Sonographic examinations. The normal fetus did not show signs of hydrops or dilatation of cardiac chambers. Further, serial Doppler scans showed significant difference in pulsatility index between pump and acardiac twin which complied with our decision for expectant management.

## CONCLUSION

To conclude, with this series, we emphasize on the encouraging role of conservative management as the best approach to follow in such complex anomalies. Our experiences suggest that expectant obstetrical management should be supplemented with Doppler sonographic follow up in all such cases and previous pessimistic notion about the expectant management should be amended.

## LEGENDS

Figure 1. (A, B) 3-D USG images showing the hydropic acardiac twin (C) T2W MRI image showing the normal twin in sagittal section with parts of the acardiac hydropic twin (black arrow) (D) Color Doppler image of the placenta showing the shared chorionic plate vessel. The flow is notably homogenous representing unidirectionality (E,F) Placenta after delivery with the normal (curved arrow) and the abnormal (white arrow) cords attached to it. The acardiac twin has been left attached to the cord while the normal twin has been removed

Figure 2. (A) Spectral Doppler showing normal umbilical artery waveform in the normal twin confirms absence of twin associated complication (B) Color Doppler image showing unidirectional flow in the shared chorionic plate vessel (white arrow) (C) 3-D USG of the anencephalic fetus showing the area cerebrovasculosa (solid arrow) (D) Anencephalic fetus after delivery

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