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Case Series

Successful clinical outcome in complicated monochorionic twins: case series

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ABSTRACT

Monochorionic twins have some unique complications like twin-to-twin transfusion syndrome (TTTS), twin anemia polycythemia sequence (TAPS), twin reverse arterial perfusion syndrome (TRAP) and conjoined twins which are otherwise not seen in dichorionic twins. In this case series, we report 3 cases of monochorionic twins who had complications. Our first case had monochorionic twin with a pump fetus and a TRAP. The second case had TTTS stage I and after spontaneous preterm premature rupture of membranes at 32 weeks, delivered both fetuses vaginally at an interval of about 3 hours. Our third case had TTTS stage II and she underwent selective fetoscopic laser photocoagulation of intertwin anastomotic vessels at about 25 weeks and subsequently multiple episodes of amnioreduction till 31 weeks when she underwent emergency caesarean delivery. Our study highlights the need for constant vigilance in patients with monochorionic twins to look for complications like TTTS and TRAP.

Keywords: Monochorionic diamniotic twins, Monochorionic monoamniotic twins, Dichorionic diamniotic twins, TRAP, TTTS, Laser photocoagulation

INTRODUCTION

Based on the number of placenta, twins are classified as dichorionic if both fetuses have different placenta and monochorionic twins if both fetuses share same placenta. Ultrasound can be employed to define chorionicity and it has been repeatedly shown that the highest accuracy is before fifteen weeks of pregnancy.1 Monochorionic twins have higher rates of complications as compared to dichorionic twins. Monochorionic twins can be divided into mainly two types i.e., monochorionic diamniotic if both fetuses are in different amniotic sacs and monochorionic monoamniotic twins if both fetuses are in the same amniotic sacs. The complications in monochorionic monoamniotic twins is much higher as compared to monochorionic diamniotic twins. The complications unique to monochorionic placentation include twin-to-twin transfusion syndrome (TTTS), twin anemia polycythemia sequence (TAPS), twin reverse arterial perfusion syndrome (TRAP) as well as the

conjoined twins.

CASE SERIES

Case 1 TRAP

A-26-years old G4P1L1A2 lady with monochorionic monoamniotic twins complicated with TRAP sequence was referred at 34-week 3 days gestation to our institute for evaluation and management. Her first pregnancy had resulted in spontaneous miscarriage at 7 weeks. Second pregnancy was dichorionic diamniotic twins which had also resulted in spontaneous miscarriage at 8 weeks. Third pregnancy was terminated at 32 weeks due to severe preeclampsia. She delivered a 1 kg female baby by Caesarean section and the child is presently alive and healthy. This was her fourth spontaneous conception. At our institute, she was found to have monochorionic monoamniotic twins with one live fetus i.e., pump twin and the other acardiac acephalous TRAP twin with a single

entwined conjoined umbilical cord of both fetuses with central insertion on the placental surface. Ultrasound showed pump twin growth corresponding to 32 weeks with estimated weight corresponding to 10th percentile and amniotic fluid index 13 cm. The cerebroplacental ratio was at 1st percentile. Absence of head, heart, thorax and upper extremity was noted in the TRAP fetus. Few pelvic structures were seen with presence of both lower limbs, marked soft tissue edema and talipes equinovarus in both feet. Umbilical cord showed single artery and single vein in a cardiac fetus which joined the umbilical cord of pump fetus to form a single entwined umbilical cord with single central insertion on placental surface placed anteriorly. Fetal echocardiography of pump twin was normal. After administration of steroids for lung maturity, the twins were delivered by LSCS at 34 weeks 5 days gestation as she had undergone preterm caesarean delivery in her previous pregnancy. Twin 1 was male with birth weight of 2.1 kg and normal APGAR score and did not show any signs of cardiac failure in the postnatal period. There were no anomalies noted in the newborn. In twin 2 male TRAP, only few abdominal structures and lower limbs were seen with no signs of life (Figure 1 and 2).



Figure 1: Single placenta with acardiac twin.



Figure 2: Acardiac acephalus twin.

Case 2 TTTS

One 30 years old G5P1L1A3 lady with monochorionic diamniotic twin pregnancy with stage 1 TTTS was referred to our hospital at 30 weeks. The same was confirmed at our institute (Figure 3). Her first pregnancy was uneventful and she had term vaginal delivery of a live healthy baby. The 2nd, 3rd and 4th pregnancies had resulted in spontaneous first trimester miscarriages. This was her 5th pregnancy by spontaneous conception. At 32 weeks gestation, on ultrasound the largest vertical pocket of liquor in Twin A was 16 cm and in Twin B it was 1.8 cm. Urine was seen in bladder of both twins. She had spontaneous rupture of membranes 2 days later. Injection Dexamethasone for fetal lung maturity and Injection Magnesium sulphate for fetal neuroprophylaxis was administered. Soon, it was followed by spontaneous onset of labour. The first twin had spontaneous vertex delivery vaginally. It was a male baby weighing 1.525 kg. Patient ceased to have uterine contractions after birth of the first baby. Mother and 2nd twin was kept under constant monitoring and expected management was planned. The second twin had vaginal assisted breech delivery after 3 hours of the birth of the first twin. It was a male baby weighing 2.03 kg with normal APGAR scores. Postnatal recovery of both twins was good. There was no postpartum hemorrhage, sepsis or any other complications in the mother.



Figure 3: Monochorionic diamniotic twin.

Case 3 TTTS treated with laser photocoagulation

Another 26 years old G3P1L1A1 lady with monochorionic twins presented at 23 weeks 5 days gestation to our institute with exaggerated abdominal distension. On evaluation, she was found to have polyhydramnios in one sac and oligohydramnios in the other sac. The urinary bladder could not be visualized in the donor fetus. Doppler studies of both twins were within normal limits. There were no features of hydrops in any of the fetuses. The couple was counseled about selective fetoscopic laser photocoagulation of intertwin anastomosis. Risks of procedure like intrauterine fetal demise, preterm premature rupture of membranes and preterm delivery were explained. The couple was also briefed beforehand

about inability to do procedure in some patients because of fetal position and other technical difficulties. Also benefits of the procedure in the form of normalization of amniotic fluid in both sacs, normal expected growth for twins and prolongation of pregnancy duration were explained. The couple after counseling opted for the procedure in the patient. She was given a course of antenatal steroids and she underwent selective fetoscopic then photocoagulation of inter twin anastomosis. About 2 weeks later, she went into preterm labour which was successfully managed with tocolytics. Hydramnios in recipient sac reappeared. In view of recurring severe polyhydramnios in recipient sac with maternal discomfort, amnioreduction was done multiple times (five times to be exact) at an interval of about 7 to 10 days under ultrasound guidance with 20 G size 15 cm long amniocentesis needle. The minimum amniotic fluid volume drained was 1300 ml and maximum were 3000 ml. This patient developed gestational diabetes and intrahepatic cholestasis of pregnancy later on. Also, Selective growth restriction in donor fetus increased and growth discordance reached 40% at about 31 weeks. There was absent end diastolic flow in umbilical artery of recipient fetus (Figure 4). At 31 weeks 2 days gestation, emergency LSCS was done in view of Stage III TTTS, growth discordance, IHCP and severe polyhydramnios in recipient sac. The first twin was male weighing 1.46 kg. The second twin was also male weighing 960 grams. Both newborns were admitted in NICU and managed by neonatologist. There was uneventful recovery of both twins and mother.

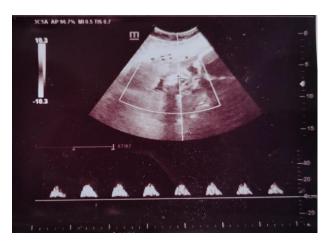


Figure 4: Absent end diastolic flow in recipient fetus.

DISCUSSION

TRAP sequence also known as acardius twin is a rare but serious complication of monochorionic multifetal gestation with an incidence of 1 in 35,000 births. TRAP is caused by a large artery to artery placental shunt often associated with a vein-to-vein shunt. With single placenta, arterial perfusion pressure of donor twin exceeds that of recipient twin who receives reverse blood flow containing deoxygenated arterial blood from its cotwin. The deoxygenated arterial blood reaches the recipient through its umbilical arteries and preferentially goes to its iliac

vessels. In our first case, the same happened and thus only the lower body was perfused and thereby resulting in disrupted growth and development of upper body.

The most commonly used classification of TRAP sequence is by Phelan and hall. It may be classified as acephalus, anceps, acormus and amorphous.² They are described as mentioned in Table 1.

Our case 1 had acephalus type of TRAP sequence. The pump twin was assessed thoroughly because it can have about 10% risk of major malformations including anencephaly, renal and skeletal anomalies.³

Table 1: Classification of TRAP sequence.

Definition	Description
Acephalus	Absent or rudimentary cephalic structures. Limbs and trunk are more or less developed. Most frequent form (>70%)
Anceps	Some cranial structures and/or neural tissue are present. It is sometimes included in the acephalus type.
Acormus	Head and cephalic structures are present but with limited or no truncal development. The head is usually attached directly to the placenta via a cord. Very uncommon.
Amorphus	Little evidence of cephalic and truncal differentiation, with very few recognizable structures such as limbs. Can resemble a teratoma, but unlike teratoma, skeletal structures and an umbilical cord is present. May represent 20% of acardia.

TTTS occurs in about 10 to 15% of monochorionic twins.⁴ The most commonly used classification system for TTTS is Quintero staging as mentioned in Table 2.⁸

Table 2: Classification of TTTS.

Stage	Classification
I	Polyhydramnios oligohydramnios sequence:
	DVP >8 cm in recipient twin and DVP <2 cm in donor twin
II	Bladder in donor twin not visible on
	ultrasound
Ш	Absent or reversed umbilical artery diastolic
	flow, reversed ductus venosus a-wave flow,
	pulsatile umbilical venous flow in either twin
IV	Hydrops in one or both twins
V	Death of one or both twins

Our second patient had stage 1 TTTS. The onset of TTTS is unpredictable, and this is why most guidelines advise biweekly monitoring with ultrasound and doppler for monochorionic diamniotic pregnancies, at least between 16 and 34 weeks of gestation.⁵ After spontaneous rupture

of membranes followed by labor and subsequently delivery of first twin, the second twin delivered 3 hours after birth of first baby. This case highlights the importance of patience of the doctor, trust of the patient with doctor and institute as well as need for constant vigilance so that if there is any need for prompt remedial measures like caesarean delivery, it can be done immediately.

Stage 1 TTTS is generally managed expectantly. Untreated TTTS is associated with more than 80% perinatal mortality and a significant rate of severe brain damage, especially in the case of survival of one twin only. In our case 3, the patient had stage 2 TTTS as urinary bladder could not be visualized on ultrasound in addition to the oligohydramnios polyhydramnios sequence. At about 25 weeks of gestation, selective fetoscopic laser photocoagulation of inter twin anastomosis was carried out with an aim to dichorionize the placenta. However, in the post procedure period also, the oligohydramnios in donor sac and polyhydramnios in recipient sac with absence of visualization of urinary bladder in donor fetus continued. Amnioreduction is an acceptable alternative in pregnancies diagnosed after 26 weeks of gestation.

CONCLUSION

The purpose of this case series is to highlight the rare complications like TRAP encountered in monochorionic twins and management of TTTS based on Quintero staging. With advancing technology in ultrasound imaging and increasing expertise in the field of fetal medicine with laser photocoagulation these is always a ray of hope for these babies.

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