CASE REPORT

A Case of Twin to Twin Transfusion Syndrome

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SUMMARY

Twin-to-twin transfusion syndrome (TTTS) is the result of an intrauterine blood transfusion from one twin (donor) to another twin (recipient). TTTS only occurs in monozygotic (identical) twins with a monochorionic placenta. The donor twin is often smaller with a birth weight 20% less than the recipient's birth weight. The donor twin is often anemic and the recipient twin is often plethoric with hemoglobin differences greater than 5 g/dL **Key words:** Twin to twin transfusion syndrome, Acardiac twinning, Monochorionic diamniotic pregnancies

INTRODUCTION

Twin-to-twin transfusion syndrome (TTTS), also called twin oligoamnios polyhydramnios sequence (TOPS) ¹

Twin to twin transfusion syndrome (TTTS) is the result of transfusion of blood from one fetal twin to another twin. The blood transfusion from the donor twin to the recipient twin occurs through placental vascular anastomoses. The most common vascular anastomosis is a deep, artery-to-vein anastomosis through a shared placental cotyledon.

Twin-to-twin transfusion syndrome (TTTS) is a serious complication in about 10% to 20% of monozygous twin gestations with an incidence of 4% to 35% in the United States. Severe TTTS is reported to occur in 5.5% to 17.5% of cases. TTTS is a progressive disease in which sudden deteriorations in clinical status can occur, leading to death of a cotwin. Up to 30% of survivors may have abnormal neurodevelopment as a result of the combination of profound antenatal insult and the complications of severe prematurity. This article presents an overview of what is known about the pathophysiology and the diagnosis of TTTS, the role of echocardiography in TTTS, treatment options available for TTTS, complications of treatment for TTTS, and short- and long-term outcomes of TTTS²

Prenatal interventions in TTTS improve the outcome. Most cases of in-utero interventions should only be performed in specialized centers as a part of a clinical trial protocol³.

Amnioreduction remains a treatment option for pregnancies with twin-to-twin transfusion syndrome (TTTS) not meeting criteria for laser surgery or those in which it is not feasible. Amnioreduction is a relatively simple treatment which does not require sophisticated technical equi`pment.4.

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Figure-1: Monozygotic twins with monochorionic, diamniotic placentation

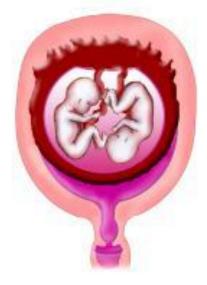


Figure-2: Monozygotic twins with monochorionic, monoamniotic placentation.

CASE REPORT

A 28-year old primigravida conceived after IUI, referred to Shaikh Zayed Hospital at 33⁺ weeks of gestation with a diagnosis of Acardiac Twin Pregnancy. She was advised elective caesarean section after steroid cover. But patient did not follow. After 2 days she came back with absent fetal movements. Her USG was done which confirmed Acardiac Twin Pregnancy with IUFD of normal twin. Her elective caesarean section was done after basic workup. Indication for elective LSCS was transverse lie of 1st twin which was IUFD.

She delivered first male fetus of 34 weeks gestation, weighing 2.6.Kg. It was pale and mildly oedematous with vesicles all over the body. No gross anomaly was seen. It was "Donor Fetus"

2nd twin was "Acardiac Acephalus Fetus" It has abdomen and lower limbs only. Thorax, head and arms were absent. Only two pits of eyes and one dimple of oropharyngeal pouch were present. The whole structure was plethoric (recipient fetus). Monochorionic placenta weighing 650g was delivered afterwards. Parents refused for autopsy, postoperative period was unremarkable and patient discharged on 5th day of operation.

DISCUSSION

Acardiac twinning, a sequalae of TRAP syndrome is a rare complication of multiple pregnancies. It affects 1% of monozygotic twins⁵. This complication is unique to "monochorionic monoamniotic pregnancies" Mostly results from "artery" to "artery" anastmosis.⁶ Donor fetus pump blood to other fetus with inadequate pressure and reduced oxygenation. This ultimately results in disruption and reduction of existing structures of "recipient fetus". Acardiac twin can be one of four types depending upon amount of blood supplied by its donor.

- Acardiac ancepus has body, lower limbs and partially formed head.
- Acardiac acephalus has abdomen, lower limbs without thorax, anus or head.
- Acardiac amorphous: snapless mass with no recognizable organs but only some axial structure.
- d. Acardiac acormus: only cranial development.⁷ Donor twin is at increased risk of sudden IUD in absence of any compromise on USG, Doppler studies or fetal blood sampling. But, in the most of cases Doppler studies show increased SD ratio in umbilical artery⁸.

Acardiac twin pregnancy is at increased risk of preterm labour (79%), polyhydramnios (51%) cardiac failure of pump twin (28%) and pump liver IUD (25-

50%).

Mortality rate for acardiac twin is 100%. Perinatal mortality rate for pump twin is 35-50%⁹

Screening for TTTS: Increased nuchal tranclucency at 10-14 weeks on USG. is associated with an increased risk for subsequent development of TTTS.

Intertwine membrane folding on USG between 16 &17 weeks gestation the likelihood ratio for the subsequent development of TTTS is 4.2%.

Velementous insertion of cord increases the risk for TTTS. Cord insertion can be reliably determined at 16 weeks. Combination of an eccentric cord insertion in one twin and marginal or velementous cord insertion in other twin is more frequently seen in TTTS prevalence rate in TTTS is 32%

Management: Of these pregnancies is controversial and range from conservative to invasive. Both strategies show similar success rate¹⁰.

Among invasive techniques "laser ablation of anastmoses" is the only option with promising results 11. purpose is to arrest shunting of blood and transfer of vasoactive mediators, should be done between 16 to 26 weeks. Overall survival of both twins is 36-50% and that of one twin is 77-90% average gestational age at birth: 32.1 weeks

Outcome depends upon gestational age at birth and whether intrauterine fetal brain ischemia occurred, lower the gestational age at birth greater the risk for longstanding neourologic or pulmonary sequelae.

CONCLUSION

Early identification of the monochorionic twin pregnancies at increased risk of TTTS would assist patient counseling and planning of follow up. Timely treatment of TTTS might improve outcome by preventing PPROM and cervical shortening which are important risk factors for preterm birth. Antenatal care should be provided in tertiary care centre. After birth treatment depends on infant specific symptoms. donor twin may need a blood transfusion to treat anemia. Recipient twin may need to have the volume of body fluid reduced, this may involve exchange transfusion, medications may be given to treat heart failure.

REFERENCES

- Chalouhi GE, Deloison B, Ville Y. Management of twinto-twin transfusion syndrome. Gynecol Obstet Fertil 2012;40:174-81.
- Habli M, Lim FY, Crombleholme T. Twin-to-twin transfusion syndrome: a comprehensive update. Clin Perinatol 2009;36:391-416.
- Dabrowska K, Gadzinowski J. Are in-utero interventions justified? Perspective of neonatologists. Part III. Twin to

- twin transfusion syndrome and congenital heart defects. Ginekol Pol 2011;82:525-32
- Li X, Morokuma S, Fukushima K, Otera Y, Yumoto Y, Tsukimori K, et al. Prognosis and long-term neurodevelopmental outcome in conservatively treated twinto-twin transfusion syndrome. BMC Pregnancy Childbirth 2011;11:32.
- James WH. A note on epidemiology of acardiac monstens. Teratology 1977.
- Sen S, Esen UI, Šturgiss SN, Twin reversed arterial perfusion syndrome. J Perinat Med 2002;30:267-72.
- Mohanty C, Mishra OP, Singh CP, Das BK, Singla PN. Acardiac anomaly spectrum. Obstet Gynecol 1998;91:818-21.

- 8. Shever DM, Armstrong B, Shah Y. Prenatal sonographic diagnosis, Doppler velocimetric umbilical cord studies and subsequent management of acardiac twin pregnancy. Obstet Gynecol 1998;74: 472-5.
- Healey MG. Acardia: predictive risk factors for co-twin's survival. Teratology 194;50:205-13.
- 10. Sullivan AE, Varner MW, Ball RH, Jackson M, Silvar RM. The management of acardiac twins: a conservative approach. Ann Obstet Gynecol 2003; 189:1310-3.
- 11. Avias F, Sunderji S, Gimpelson R, Cotton E. Treatment of acardiac twinning. Clin Dysmorphol 1997;6:359.