

Case Report

Conjoined twins: report of two cases and review of literature

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Received: 22 May 2014

Accepted: 10 June 2014

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ABSTRACT

Two pairs of conjoined twins were encountered at the SCB medical college cuttack, Odisha, India during the past 3 years. In the first set the prenatal diagnosis was not established and the case was referred to us from other hospital with diagnosis of twin pregnancy and prolonged second stage of labour. Emergency caesarean section done and a pair of thoraco-omphalopagus twins delivered. Both babies died soon after birth. In the second case the conjoined twins were diagnosed prenatally at 20 weeks of gestation in routine anomaly scan as thoraco-omphalopagus twin with fetuses were positioned face-to-face and fused from sternum to umbilicus. Fetuses were found to have separate hearts, a fused liver and separate upper and lower gastrointestinal tracts. Each twin had two normal appearing kidneys and a urinary bladder. The couples were counselled about the various management options and referred to higher centre. But she was admitted at 34 weeks of gestation with preterm labour and emergency caesarean section done and a pair of male thoraco-omphalopagus twins delivered. Both babies were alive and referred to higher centre for further management. The prenatal identification of conjoined twins is of cardinal importance for the planning of delivery and possible separation.

Keywords: Conjoined twins, Thoraco-omphalopagus, Monozygotic, Prenatal diagnosis, Surgical separation

INTRODUCTION

Conjoined twins are a rare complication of monozygotic pregnancies. The incidence of conjoined twins is approximately 1:50000 in utero to 1:250000 live births.¹ Conjoined twins are generally incompatible with life i.e. 65% of cases are stillborn while of those that are born alive, 35% die within the first 24 hours.² Only 25% survive to an age where surgical separation can be considered.³ Prognosis is related to the vitality of the shared organs and gravity of the accompanying congenital anomalies, as well as to the location and extent of adhesion. The prenatal identification of conjoined twins is of cardinal importance for the planning of

delivery and possible separation.⁴ The cases were presented because of their rarity.

CASE REPORT

Case 1

Our first case a G₃P₂L₂, 28-years Hindu female referred from peripheral hospital, admitted in our labour room as a case of prolonged second stage of labour with twin pregnancy. This was unsupervised pregnancy till onset of labour. She was given trial for vaginal delivery and referred to us in second stage of labour as head failed to descend even after 4 hours. Her previous two pregnancies were uneventful vaginal delivery and at both babies were

normal. There was no family history of twin pregnancy, or ingestion of ovulation inducing agents, drug or radiation exposure in first trimester. Examination revealed features of obstruction and emergency caesarean section done. To our surprise a pair of male thoraco-omphalopagus twins delivered (Figure 1). The babies were delivered at 33 weeks of gestation with combined birth weight of 3.5 kg. The fetuses were positioned face-to-face and fused from upper thorax to umbilicus with single umbilical cord. Both the babies died soon after birth and the parents denied autopsy because of religious belief.



Figure 1: Conjoined male thoraco-omphalopagus twins with single umbilical cord.

Case 2

The second case was a G₃P₁A₁, 30 year old Hindu female came for routine antenatal check-up at 20 weeks of gestation and diagnosed prenatally as a case of thoraco-omphalopagus conjoined twin. Fetuses were found to have separate hearts, a fused liver and separate upper and lower gastrointestinal tracts. Each twin had two normal appearing kidneys and a urinary bladder. Neither there was any family history of multiple pregnancy or congenital anomaly nor had she taken any drugs or exposed to radiation during first trimester. She had one previous normal vaginal delivery and spontaneous abortion at 2 months of gestation. The couples were counselled about the various management options and referred to higher centre. But she was admitted at 34 weeks of gestation with preterm labour and emergency caesarean section done and a pair of male thoraco-omphalopagus twins delivered. The babies were positioned face-to-face and fused from sternum to umbilicus with single umbilical cord (Figure 2). The combined birth weight was 4.5 Kg. Placenta was Monochorionic & Monoamniotic and there was a succenturiate lobe (Figure 3) found around 10 cm away from main placental mass. The babies were resuscitated by neonatologist and admitted to NICU. Post-operative

period was uneventful and both babies were referred to higher centre for further management and possible separation.



Figure 2: Conjoined male omphalopagus twins with single umbilical cord.



Figure 3: Succenturiate lobe in monochorionic and monoamniotic placenta.

DISCUSSION

Conjoined twinning is a rare aberration of monozygotic monoamniotic twinning and results in fusion of the twins at any part of their body. Conjoined twinning is usually thought to occur with late division (post day-13) of the embryonic disc and “conjoined” might actually be a misnomer since it is a failure of fission.³ The incidence of monozygotic twinning is independent of race, maternal age and geography. There is a predominance of females in conjoined twins on the order of 3:1 and no increase in risk is known with parity, race, maternal age or heredity.⁵ But incidentally both set of twins were male in our cases. Conjoined twins are classified according to the adhesion

regions: thoracopagus (thorax), omphalopagus (abdomen), pygopagus (sacrum), ischiopagus (pelvis), craniopagus (cranium), cephalopagus (face), and rachipagus (dorsal). Thoracopagus is the most frequent type. 75 % of conjoined twins are seen as thoraco-omphalopagus.⁶ If they have separate sets of organs, chances for surgery and survival are greater than if they share the same organs.

Prenatal diagnosis is most important in order to have the opportunity to choose any option and Ultrasonography and MRI are the tools of first choice in this aspect.⁷⁻⁹ Unfortunately this was not done in our first case and she landed in obstructed labour with undiagnosed conjoined twins. An evaluation consisting of a level II ultrasound, fetal echocardiogram and fetal MRI is particularly important for conjoined twins, as the location and extent of where the twins are joined and what organs are shared plays a crucial role in deciding whether the twins will be separable. Fetal ultrasound is a safe, non-invasive procedure that uses high frequency sound waves to provide detailed high-resolution images, including 3-D and 4-D views. Fetal echocardiogram is a non-invasive ultrasound procedure that assesses the structure and function of the fetal heart. Fetal echocardiogram is the most important test for the evaluation of conjoined twins, as it determines if the twins share a heart and if so, where that connection occurs. In fetal MRI mainly the recently used Ultrafast fetal MRI which does not require sedation of mother or muscle relaxants for the fetus to obtain images, the indicators of conjoined twins include lack of a separating membrane between the twins, inability to separate the fetal bodies, and constant position of the fetal heads.¹⁰

Our first case was thoraco-omphalopagus, the most common type (75%) of conjoined twin where the two babies lie face to face and share a common sternum, diaphragm and upper abdominal wall. In cases of conjoined hearts at ventricular (pumping chamber) level, there are no known survivors in literature.¹⁰ Our second case was also a thoraco-omphalopagus twin where the babies face one another and are joined at the anterior abdominal wall from lower sternum to umbilicus. The hearts are separate, the peritoneal cavity of one tends to be joined with the other, but upper intestinal tracts are usually separate. In the majority of cases, a bridge of liver connects the infants which was also present in our case. The prognosis and possible separation is better in this case.

Pygopagus represents about 20% of cases, joined at the buttocks and perineum, facing away from each other. A significant length of the sacrum may be fused, and as a result, the twins often share the sacral spinal canal. A single lower rectum and anus is common, and often the lower genital tract and external genitalia are fused. Ischiopagus represents less than 5% of cases, the connection occurs at a single bony pelvis. Four normal legs may be attached, but often two of the four are fused

into one malformed limb. The intestinal tracts often join and empty into a single colon. Craniopagus which is the least common type of conjoined twins, accounting for 2% of cases, is represented by fusion of the skull. The twins often share large Dural sinuses and vascular structures. Rarely, the brains are separated by bone and each brain has separate leptomeninges. In others, the brains are connected, or separated only by arachnoid, making separation extremely difficult and dependent upon a superior sagittal sinus for each brain.¹⁰

Treatment approach includes mostly termination of pregnancy. If a decision for continuation of pregnancy is taken, then congenital cardiac anomalies, degree of organ sharing and congenital anomalies should be determined by fetal echo with fetal MRI.⁶ Delivery should be planned in a tertiary centre with facilities of possible separation. Mode of delivery is caesarean section and a vertical uterine incision is often required.⁶ Prognosis depends on the location and extent of the conjoinment, presence of additional anomalies in the fetuses, whether the vital organs are shared or not, the extent of the shared organs which determines decisions for surgical separation or termination of pregnancy.

CONCLUSION

This case highlights the features of an extremely rare fetal malformation. Once the diagnosis of conjoined twins is established, the family should be informed in detail. Besides the choice of termination, if continuation of pregnancy and a postnatal separation procedure is desired they should be provided with detailed information on prognosis and results.

ACKNOWLEDGEMENTS

We would like to thank the dept. of Obstetrics & Gynecology, SCB medical college, Cuttack, Odisha for their valuable support and co-operation of patients and their families admitted to these hospitals.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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DOI: 10.5455/2320-6012.ijrms20140851

Cite this article as: Rathod S, Samal SK. Conjoined twins: report of two cases and review of literature. *Int J Res Med Sci* 2014;2:1203-6.