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Case Report

A thoracophagus conjoined twins with myelomeningocele: an unusual case

Saunitra A. Inamdar, Shruti S. Goel*, Vaidehi S. Subhedar

Department of Obstetrics & Gynecology, JNMC, Sawangi (M), Wardha, Maharashtra, India

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***Correspondence:**

Dr. Shruti S. Goel,

Email: shruti_7589@yahoo.co.in

ABSTRACT

Conjoined twins represent one of the rarest forms of twin gestation. Conjoined twinning is rare, occurring in about 1% of monochorionic twins with an estimated incidence ranging from 1:30,000 to 1:2,00,000 live births and 1 in 650 to 900 twin deliveries. A 21 year old gravida 2, with an abortion, a resident of Gijnakheda, Deoli Wardha came to our hospital, AVBRH Sawangi and presented with history of amenorrhoea of six months. She had her first ultrasonography done which demonstrated thoracophagus conjoined twins. Patient was admitted in view of management of conjoined twins. She was posted for elective caesarean section on 5/11/11. A conjoined twin of combined weight around 2 kgs was extracted out. Both the foetuses were female with multiple congenital anomalies. In conclusion, conjoined twins are associated with a high perinatal mortality; therefore making an early diagnosis by means of ultrasonography, gives parents a chance to elect pregnancy termination.

Keywords: Conjoined twins, Thoracophagus, Prenatal diagnosis

INTRODUCTION

Diaphragmatic hernias complicating pregnancy is rare. Conjoined twins represent one of the rarest forms of twin gestation.¹ Conjoined twins are a subset of monozygotic twins in which incomplete embryonic division occurs on 13 to 15th days following conception resulting in varying degrees of fusion between the two foetuses.²

Conjoined twinning is rare, occurring in about 1% of monochorionic twins with an estimated incidence ranging from 1:30,000 to 1:2,00,000 live births and 1 in 650 to 900 twin deliveries. An increased incidence of 1:14,000 to 1:25,000 is described in various parts of South East Asia and Africa. Some 40% to 60 % of conjoined twins are reported to be still born and approximately 35% of live births do not survive beyond first 24 hours. Prognosis is related to the vitality of shared organs and gravity of the accompanying congenital anomaly as well as location and extent of adhesions. There is a reported female preponderance of 3:1.²

CASE REPORT

A 21 year old gravida 2, with an abortion, a resident of Gijnakheda, Deoli Wardha came to our hospital, AVBRH Sawangi and presented with history of amenorrhoea of six months. She had no previous antenatal visits. She had her first ultrasonography done which demonstrated thoracophagus conjoined twins. Patient was admitted in view of management of conjoined twins. She is gravida 2 with an abortion which was a spontaneous abortion at two months for which she underwent check curettage at civil hospital, Wardha. She had her last menstrual period on 11/4/11. According to Naegele's formula her expected date of delivery was 18/1/12. Her previous menstrual cycles were regular. Her previous medical, surgical and obstetrical history was unremarkable. There was no family history of twinning on either maternal or paternal sides and no history of intake of medication or exposure to radiation. She was diagnosed as a sickling trait. On general examination her vitals were stable. On per abdominal examination fundal height was corresponding to 28 weeks, foetal heart rate was 140 beats per minute. Relatives were informed about the malformation and poor chance of survival of the twins. They decided to terminate the pregnancy. A written informed consent was taken from the relatives regarding the termination.



Figure 1: Conjoined twins delivered by LSCS.

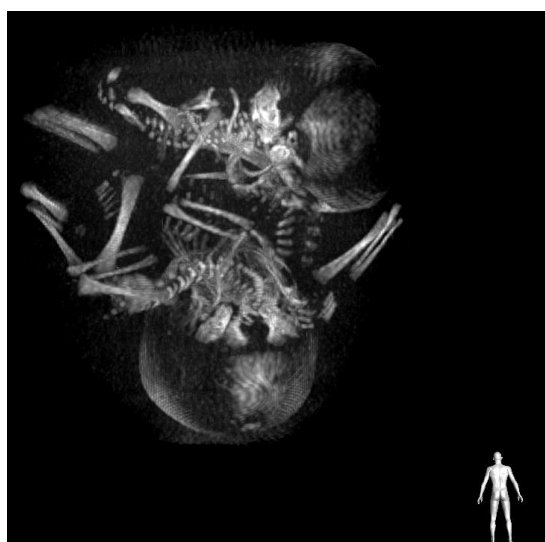


Figure 2: X-ray of conjoined twins.

Ultrasonography demonstrated thoracoabdominal conjoined twins of gestational age 24.1 weeks with two heads seen separately with single heart, stomach and bladder with probable fusion of spines of foetuses with closed myelomeningocele. Placentation was monoamniotic and monochorionic type. Cardiac activity was noted with probable congenital structural defects.

She was posted for elective caesarean section on 5/11/11. A conjoined twin of combined weight around 2 kgs was extracted out. Both the foetuses were female with multiple congenital anomalies. First baby was extracted by vertex presentation followed by the trunk with malformed trunk of second twin followed by the head. Baby showed few limb movements after birth. Immediately the neonates were resuscitated and shifted to neonatal intensive care unit. On examination the fetal biometry of both twins was consistent with the menstrual age. The upper limbs of both the twins were normal. Foetuses were positioned face to face and fused from abdomen to lower thorax. Foetuses were found to share the heart, bladder and liver. One of the foetus had

myelomeningocele. The neonates were shifted to NICU where they were shifted to ventilator support. Though prompt resuscitative measures were taken, the twins died within 2 hours of birth from cardiopulmonary failure.

DISCUSSION

Conjoined twins are rarely encountered and very few obstetricians are confronted with conjoined twins in their professional life.² Conjoined twins are classified according to the most prominent site of conjunction and the aspect of embryonic disc.¹

Two contraindicating theories exist to explain the origins of conjoined twins. The traditional theory is fission, in which the fertilised egg splits partially and conjoined twins represent delayed separation of the embryonic mass after day 12 of fertilisation. The second theory is fusion, in which a fertilised egg completely separates, but stem cells find like -stem cells on the other twin and fuse the twins together.³⁻⁵

Antenatal diagnosis of conjoined twins is usually uncommon, the fact that they are conjoined is usually not determined until late in gestation or during parturition.^{6,7} The diagnostic modalities available for antenatal diagnosis of conjoined twins includes radiography, ultrasonography and magnetic resonance imaging. Gray and associates established a list of criterion for antepartum diagnosis of conjoined twins: the heads are at same level and body plane; the spines are in unusual proximity; the spines are usually extended; the foetuses do not change position relative to each other after movement or manipulation.⁸ A detailed survey of the vasculature of fused vital organs is very important in determining the prognosis of surgical separation and also owing to the high frequency of associated anomalies related to fusion, which includes neural tube defects, orofacial clefts and diaphragmatic hernia.²

In conclusion, conjoined twins are associated with a high perinatal mortality; therefore making an early diagnosis by means of ultrasonography, gives parents a chance to elect pregnancy termination. Due to improved operative techniques outcome in conjoined twins has improved significantly.

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