

Case Report

Conjoined Twins (Thoraco-Omphalopagus)

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ABSTRACT

Conjoined twins are a rarely seen congenital anomaly with severe mortality. Among the different variety of conjoined twins, Thoraco-omphalopagus is the most common type, wherein the two foetuses are joined at thorax and upper abdomen region. In this type of twins usually there is single heart, but lungs will be separate. In GIT, the foregut will be separate, but midgut may be shared by both twins. Hindgut will be separate. The exact cause is unknown, but it is mostly considered to be an irregular division of the zygote. One such case was observed during routine foetal autopsy performed in Dept of Anatomy, GMCH-32, and Chandigarh. The mother was 21yrs old prime gravida and the condition was diagnosed at the time of USG examination at 13+6 weeks of gestational age. Autopsy was performed after taking full consent. The foetuses had single umbilical cord and sex of both the foetuses was male. After autopsy it was found that both foetuses shared single heart, stomach, small intestine, large intestine, liver and spleen. However there was development of separate lungs and organs of Genito-urinary system. There are two theory proposed for the formation of the conjoined twins. A fusion theory which is more accepted and other one is fission theory. The exact mechanism of formation of twins, obstetrical and surgical importance and other details will be discussed in detail with the available literature.

KEY WORDS: Conjoined Twins, Thoraco-omphalopagus, Congenital Anomaly.

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Online Access and Article Informtaion

Quick Response code



DOI: 10.16965/ijims.2015.130

International Journal of Integrative Medical Sciences

www.imedsciences.com

Received: 12-10-2015

Accepted: 02-11-2015

Reviewed: 12-10-2015

Published: 10-11-2015

Source of Funding: Self

Conflicts of interest: None

INTRODUCTION

Babies come into the world heralding the good news that the human species with all its diversities and complexities is still going good. Though a new-born brings in its wake untold happiness to those around, there are some unfortunate babies whose birth is clouded with sadness and worry for the parents because of the birth defects [1] in them that are manifest either immediately defects like cleft lip, cleft palate, Down syndrome, autism, muscular dystrophy, neural tube defects, congenital heart disorders to rare birth defects such as cleft foot and hand, club foot, aglossia and albinism, to name a few

after birth or after a while, depending on the nature of the congenital abnormality. Birth defects include abnormalities in the new born baby's (structure, function or body metabolism), which usually lead to physical and mental disabilities and can even be fatal sometimes. There are many causes for birth defects involving a wide range of factors some due to hereditary abnormality, some caused by chromosomal disorders or genetic disorder and some others caused by environmental agents.

All over the world as per the 2015 studies Congenital anomalies affect approximately 1 in 33 infants leading to 6.6% deaths in infants and

causing significant morbidity in children. Birth defects are present in about 3% of new-borns [2]. Congenital anomalies resulted in about 632,000 deaths per year in 2013 down from 751,000 in 1990 [3-6]. The type with the greatest death are congenital heart disease (323,000), followed by neural tube defects (69,000) [7]. Many studies have found that the frequency of occurrence of certain congenital malformations depends on the sex of the child [4-8]. Now various techniques are available to detect congenital anomalies in fetus before birth. About 3% of new-borns have a "major physical anomaly", meaning a physical anomaly that has cosmetic or functional significance [9]. Physical congenital abnormalities are the leading cause of infant mortality in the United States, accounting for more than 20% of all infant deaths. Seven to ten percent of all children will require extensive medical care to diagnose or treat a birth defect [10]. Among new-borns is between 2-3% that is similar to that found in the industrialized world (ICBDMS, 1991) Congenital anomalies account for 8-15% of perinatal deaths and 13-16% of neonatal deaths in India.

Conjoined twins are rarely seen congenital anomaly with severe mortality occurring in 1:50,000 - 1:60,000 live births. Among the different variety of conjoined twins, Thoraco-omphalopagus is the most common type, wherein the two fetuses are joined at thorax and upper abdomen region. Teratologists have proved that congenital abnormalities in animals can be induced but no one has been able to prevent the occasional occurrence of conjoined twins. Until this is achieved; surgeons will be concerned with repair procedures, primarily depending upon the morphology of the abnormality. The opportunity to dissect ischiopagus, whose incidence among conjoined twins is very low (6%) is worth reporting [11].

Case Report

The different variety of conjoined twins Thoraco-omphalopagus is the most common type, wherein the two fetuses are joined in the thorax and upper abdomen region. In this type of twins usually they will share a single heart, but lungs will be separate. The GIT system the foregut part

will be separate, but midgut may be shared by both twins. Hindgut will be separate. The exact cause is unknown, but it is mostly considered to be irregular division of the zygote. Here we are reporting a case of Thoraco-omphalopagus. During a routine foetal autopsy, a foetus was found to have a thoracophagus in the Dept. of anatomy, GMCH-32, Chandigarh. The mother was 21yrs old primi-gravida and the condition was diagnosed at the time of USG examination at 13+6 weeks of gestational age. A primigravid was diagnosed thru USG as Thoracophagus and pregnancy was terminated and abortus was sent to our department for autopsy. Antenatal history was normal with routine check-ups and intake of vit-B12 and folic acid tablets. Medical and past history was not significant. No significant family history. Autopsy was performed after taking full consent.

OBSERVATIONS

External examination:

- Both were male foetus.
- Foetuses were fused at thorax and upper abdomen region.
- Both foetuses had separate urogenital openings.
- Both foetuses had imperforate anus.
- Limbs were separate.
- Left side foetus showed amniotic bands at midhigh level on left side.
- They had single umbilical cord.

Internal Examination:

- Incision was given to open the fused region.
- The thoracic and abdominal cavities were opened.
- Following observations were seen.
- Single heart.
- Trachea fused in the middle region. In upper and lower part they were separate to both foetuses. They both had separate lungs.
- In GIT: both foetuses had separate oesophagus which opened into common stomach. Small intestine and large intestine were shared inside both foetuses. Sigmoid colon was seen on right side only.
- Liver was present on right foetus and spleen in left foetus.

In Genito-Urinary system: Both foetuses had their own genito-urinary system.

Fig. 1: Showing the Internal Examination of the Conjoined Twins (Thoraco-Omphalopagus).



Fig. 2: Showing the External Examination of the Conjoined Twins (Thoraco-Omphalopagus).

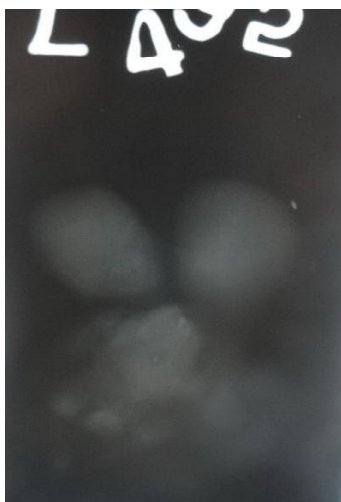


Fig. 3: X-Ray showing the External Examination of the Conjoined Twins (Thoraco-Omphalopagus).

DISCUSSION

Conjoined twinning was first described in 1100AD. The first case diagnosed by USG was reported in 1977. It is rare abnormality estimated to occur in about 1% of monozygotic twins. Depending upon the site of fusion it is divided into different types of conjoined twins. Robertson

in 1953 studied conjoined twins and determined the incidence of various types as being 73% thoracopagus, 19% Pygopagus, 6% ischiopagus and 2% craniopagus. The female conjoined twins are two to three times more common than male twins. Higher incidence of anomalies in still births [12] and the incidence of congenital anomalies were significantly higher in preterm babies [13], low birth weight infants [14], mother's age above 35 years [15], increased maternal age [16], and increased birth order. The factors that significantly increase the risk of congenital malformations to be presence of hydramnios, maternal febrile illness in the first trimester, past history of abortions, diabetic mother, eclampsia, previous abortion and history of congenital heart disease in previous child or malformed babies. Mother having diabetes mellitus, arterial hypertension, and hypothyroidism shows a positive association [17] with congenital malformations. The annual report of Indian Council of Medical Research [18] says that the commonest congenital malformations are cardiac in nature. Cardiovascular, musculoskeletal and genito-urinary were the most commonly affected systems in a descending order of frequency. With special reference to the neural tube defect (NTD) [19], the incidence of NTD has markedly reduced in the developed countries following mass promotion and mandatory prescription of folic acid for pregnant mothers. The incidence of congenital heart disease was the leading congenital malformation followed by musculoskeletal system.

Types of Conjoined Twins:

Inferior Conjunction	Lower body is single or twins joined by some lower portion of body
Diprosopus	Two faces with one head and body
Dicephalus	Two heads with one body
Ischiopagus	Joined by inferior sacrum and coccyx
Pygopagus	Joined by posterolateral sacrum and coccyx
Superior Conjunction	Upper body is single or twins joined by some upper body portion
Dipygus	Single head, thorax, abdomen with two pelvis and four legs
Syncephalus	Facial fusion with or without thoracic fusion
Craniopagus	Joined at the head
Middle Conjunction	Fusion of the midportion of the body, separate above and below
Thoracopagus	Thoracic fusion
Omphalopagus (Xiphopagus)	Joined from umbilicus to xiphoid cartilage
Thoraco-omphalopagus	Thoracic and abdominal fusion
Rachipagus	Vertebral fusion above the sacrum

The exact aetiology for the occurrence of twins is unclear. But two theories have been proposed to explain this phenomenon. The traditional theory is fission theory, in which the fertilized egg splits partially and conjoined twins represent delayed separation of the embryonic mass after day 12 of fertilization. The second theory is fusion, in which a fertilized egg completely separates, but stem cells (which search for similar cells) find like stem cells on the other twin and fuse the twins together. Generally this type of cases normal delivery is not possible if the fetuses survive up to term.

Prenatal ultrasonography leads to early detection of malformations and facilitates early surgical intervention. Ultrasound at around 20 - 21 weeks has long been considered for screening pregnancies for structural malformations. Use of second trimester ultrasound for detection of chromosomal anomalies was first suggested in 1985 [20]. Chromosomal defects were progressively found to be associated with certain sonographic features, including biometric parameters (e.g., short length of femur and humerus, pyelectasis, large nuchal fold, ventriculomegaly, early fetal growth restriction) and morphologic signs (e.g., choroids plexus cysts, echogenic bowel, echogenic intracardiac focus). Data on the validity of those markers as predictors of chromosomal anomalies (mostly related to Down syndrome) are at variance depending upon the author [22]. There may be multi factorial reasons for these namely lack of awareness among people to get level 3 ultrasound during antenatal period, poverty, lack of proper health services in periphery. The main purpose of the study is we need to involve and make aware all the health care workers who are providing maternal and child health care working in government or private sector so as to quantify exact prevalence rate of congenital malformations involving any particular system. Mortality and morbidity can be because of sepsis, infections, prematurity or delay in treatment due to late admissions after diagnosis due lack of money, poor background and lack of awareness. This study definitely helps to know the pattern of congenital anomalies and their outcome in this area so that strategies for prevention, early detection and timely manage-

ment can be sort out.

Management: Prenatal diagnosis by USG should be done. Surgery to separate conjoined twins may vary from relatively simple to extremely complex, depending on point of attachment and the internal organs that are shared, but it is life threatening.

CONCLUSION

The prognosis of this type of twins is generally poor. Once conjoined twins have been diagnosed, characterization of the type and severity of the abnormality should be assessed using three-dimensional USG or MRI. Termination of pregnancy should be suggested to the family.

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How to cite this article:

Prasad Arun, Kapoor Kanchan, Sharma Anshu, Abraham Joseph. Conjoined Twins (Thoraco-Omphalopagus): A Case Report. *Int J Intg Med Sci* 2015;2(10):170-174. **DOI:** 10.16965/ijims.2015.130