Case Report

Paediatric Surgry Section

Thoraco-abdominal Anomalies in a Dicephalic Parapagus Twin

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ABSTRACT

Conjoined twins represent a rare but fascinating congenital condition, the aetiology of which remains obscure. 'Parapagus' is a fairly new term, in which the twins lie side to side, with ventrolateral fusion but there is some confusion in the literature. Parapagus twins arise from streaks that lie adjacent to one another, potentially allowing cross-signaling to occur, with the development of laterality defects. In this article, we demonstrate the various anomalies of the thoracic and the abdominal cavity, with an embryological reason behind their development. An accurate antenatal assessment allows the parents to be counseled in depth as to the likely outcome of the pregnancy and the chances of postnatal separation and survival..

Key Words: Conjoined twin; Parapagus; Dicephalic twin; Monozygotic twins

INTRODUCTION

Conjoined twins are being increasingly accepted into our everyday lives, principally because of the increasing number of cases where attempts have been made to separate them surgically and also because of the curiosity towards the science behind their development.

'Dicephalus' is a subset of parapagus, in which the twins share a common body from the neck or upper chest downwards, having only a pair of limbs and a set of reproductive organs [1]. This anomaly represents less than 0.5 % of all the reported cases of conjoined twins [2].

Most authors report that 70-95% of the conjoined twins are females [3,4] one study cited a nearly equal male: female ratio. [4] The reason for a greater number of females in most series of conjoined twins is not known. Milham suggested that it might be due to the early loss of conjoined male embryos [5].

Conjoined twinning arises when the twinning event occurs at about the primitive streak stage of development i.e., at about 13-14 days after fertilization and it is always associated with the monoamniotic, monochorionic type of placentation [6].

The frequent anomalies which are associated with conjoined twinning are the duplication of the visceral organs, omphalocele, facial clefts, meningomyelocele, an imperforated anus and cardiac defects [7].

After reviewing a lot of literature which dealt with the subject of conjoining, we became aware that the complexity of the union in our case needed an extensive study and evaluation before the surgical separation of the conjoined twins.

CASE REPORT

A two-headed, conjoined twin was donated to anatomy department for preservation by the Paediatric Department, Chigateri General Hospital, Davangere, India. The twin was delivered as stillborn to a primigravida by caesarean section. The mother had limited antenatal care and no antenatal ultrasonogram was done earlier. There were no records of previous congenital anomalies in the siblings of the family members.

An autopsy revealed that the twin had two heads and necks and the fusion of the trunk below the neck with the right twin head was in better alignment with the spine. Externally, a normal trunk, abdomen, upper and lower limbs, a single set of ambiguous genitalia and an anus were seen.

An infantogram showed two separate vertebral columns terminated abruptly with a single sacral component and two sets of ribs, one from each vertebral column were projected ventrally to meet a single sternum.

The twin was carefully dissected and a photographic record was made of all recognizable anomalies.

RESULTS

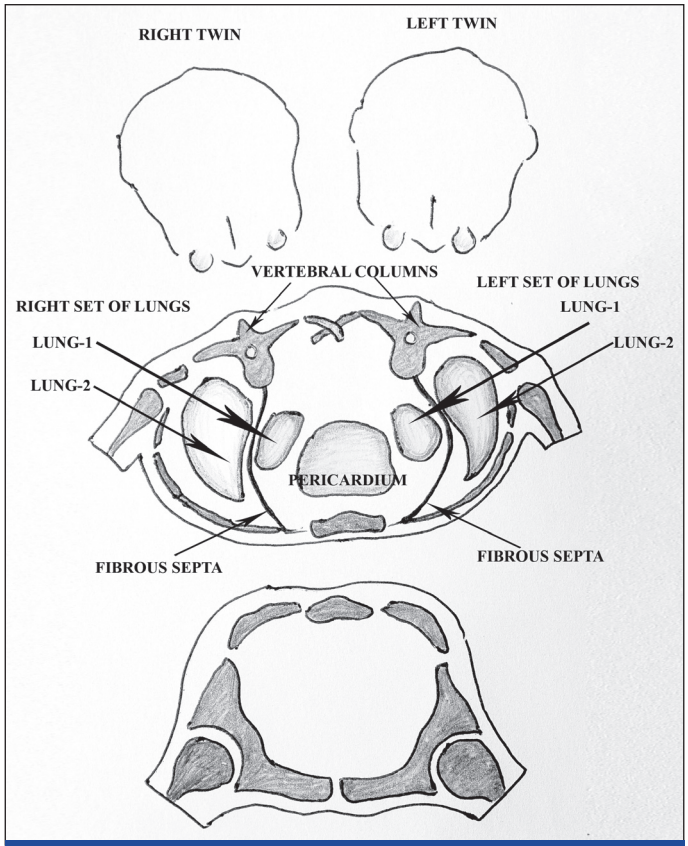
Within the thorax, [Table/Fig-1&2] two tracheae (one from each twin) led to two set of lungs, whose lobes occupied three pleural cavities. The median pleural cavity contained two lungs, one from each twin and a pericardial cavity more anteriorly.

Two lateral pleural cavities were separated from the median pleural cavity by two fibrous septae [Table/Fig-3] which extended from the posterior surface of the sternum to the anterior surface of the vertebral columns.

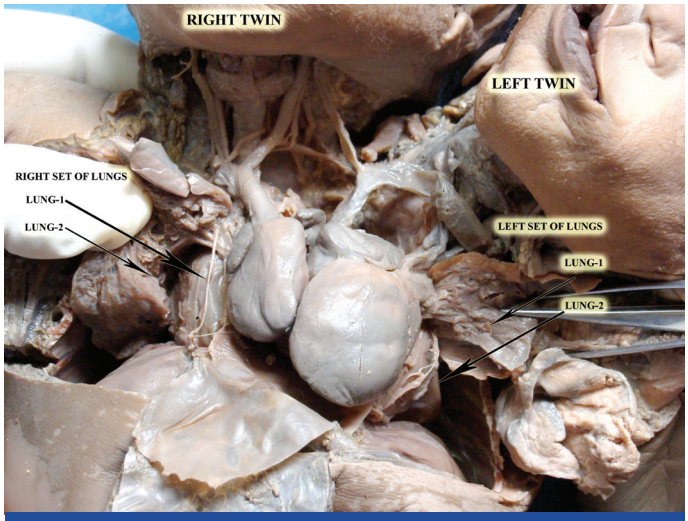
The trachea from the right twin divided into two primary bronchi which opened into the two right lungs. Of which the lateral one was well developed and the medial one was hypoplastic [Table/Fig-2]. No hilar structures were identifiable. Similarly, the left twin's trachea bifurcated into two, which entered into the two left lungs, of which the medial lung was poorly developed.

The mouth, pharynx and the oesophagus of each twin were normal till the level of the 7th cervical vertebra. Upon entering into the thorax each ran in different courses.

The oesophagus of the right twin passed to the right of the media stinum between the trachea and the vertebral bodies passed more anteriorly within the fibrous septa [Table/Fig-3] located between located between the two right lungs. Then, it crossed the midline of the thorax and pierced the diaphragm to continue as a single



[Table/Fig-1]: Cross-sectional illustration of dicephalic twin at the level of head, thorax and pelvis levels

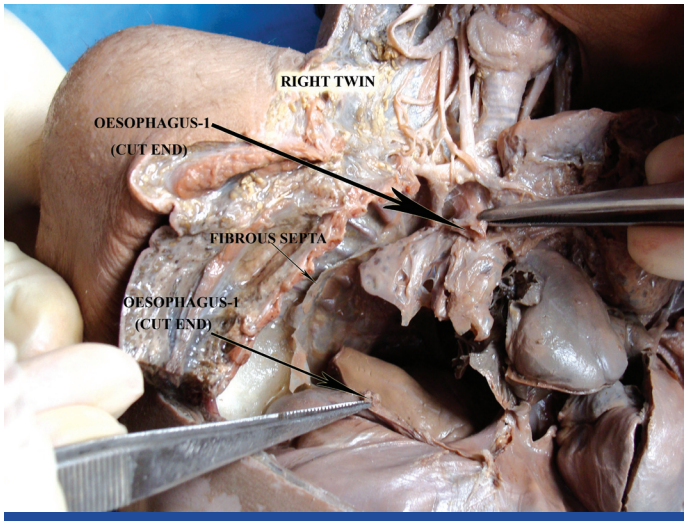


[Table/Fig-2]: Photograph of dissected twin showing two sets of lungs and a shared heart (pericardium removed) in a thoracic cavity

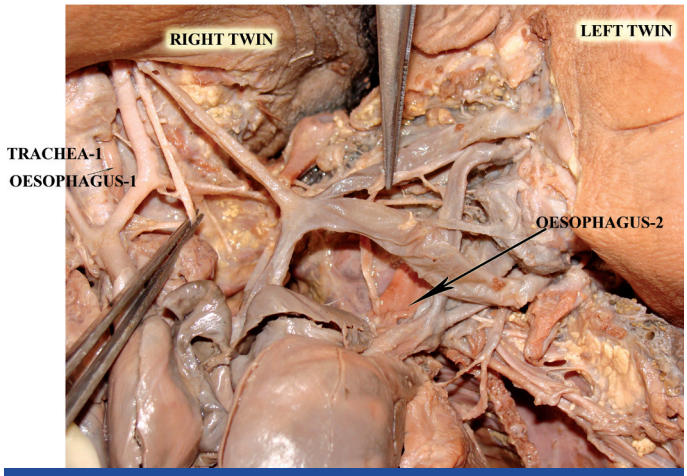
stomach in the abdominal cavity. The oesophagus of the left twin [Table/Fig-4] became obliterated at the level of the root of the neck as a thin sheath and disappeared.

The stomach and the intestine were single and normal in shape as per the size of the foetus. The lobes of the liver were enlarged and the gall bladder was normal. A single spleen which was common to both the twins was located below the left side of the diaphragm.

The urogenital system revealed the absence of the kidneys at their normal locations. Both the kidneys were fused and were located above the level of the diaphragm in the thoracic cavity, with a larger defect which was formed in between the two vertebral columns. There were two ureters which opened into a single bladder normally. The suprarenal glands could not be identified.



[Table/Fig-3]: Photograph of dissected twin showing the root of the neck with esophagus of left twin (esophagus-2) being obliterated



[Table/Fig-4]: Photograph of dissected twin showing fibrous septa between each set of lungs and esophagus of right twin (esophagus-1) continues as a single stomach in the abdominal cavity

Ambiguous genitalia were noted externally with no internal genital organs of either sex.

DISCUSSION

There are two theories of how conjoined twins are formed. The more widely accepted one is the "fission theory", which states that conjoined twins occur when a fertilized ovum begins to split into identical twins, but is somehow interrupted during the process and develops into two partially formed individuals who are stuck together [1].

Spencer argued that conjoined twinning cannot possibly result from a "fission event", and can result from the fusion of monoamniotic twins. He proposed that two monovular embryonic discs may lie adjacent to one another at various angles, and may become secondarily united rostrally, caudally, laterally or dorsally and symmetrically or asymmetrically but always homologously [8].

It might seem logical to assume that dicephalus twins arise from two separate, nearly parallel notochords on one embryonic disc, very close together caudally, but with varying degrees of separation rostrally.

By the end of the fourth week, except for the area of the umbilicus, the entire embryo becomes covered by intact ectoderm and all the possible sites of conjoined twins may have either closed or moved to locations which are inaccessible for possible secondary fusion [8].

As it is postulated that intact skin will not fuse with intact skin, including the ectoderm of the embryo, there are some specific locations in which the ectoderm is normally absent or normally destined to fuse or to breakdown and to determine whether these areas are analogous to the sites where conjoined twins are to be united. Or, the union of conjoined twins can occur only where the surface ectoderm is absent or normally programmed to become disrupted or united. This thesis not only speaks fusion but also argues against fission [8].

The site and the extent of the union are reflected in the varying degrees of involvement of the viscera, particularly the heart, diaphragm, the gastrointestinal tract and the urogenital system.

The respiratory system arises as a diverticulum on the ventral aspect of the primitive foregut and in the dicephalus, usually develops into individually owned trachea and lungs for each twin. As the duplication of the oesophagus extends further caudally in the dicephalus, a second trachea and a pair of lungs will develop [9].

As ventrally conjoined twins are postulated to be united over one yolk sac, some portion of the gastrointestinal tract should remain single (undivided), the location and the length of the shared viscera varying with the site and the extent of the primary union of the embryonic discs. Dicephalus twins generally have two oesophagi which may join in the middle of the thorax or both may empty into a single stomach [9].

As the duplication of the vertebral columns increases toward the caudal aspect in the dicephalus, the oesophagus doubles and the diaphragms are defective in only one-third. Most of the defects are located between the two vertebral columns, with an occasional notation that each of the two conjoined diaphragms is attached to a different spine. The hernia which is associated with these posterior diaphragmatic defects protrudes into the posterior aspect of the conjoined thorax [9].

CONCLUSION

Conjoined twins should be suspected in all monochorionic, monoamniotic twin pregnancies and careful sonographical assessment should be undertaken to exclude any of the classical signs which are suggestive of conjoined twins and to identify the severity of the shared foetal organs for perinatal management.

When serious malformations that are incompatible with post natal life are diagnosed early, as in our case the selective termination of a single baby is impossible. We therefore agree with Owolobi et al [10]. that the termination of the pregnancy should be advised in cases where dicephalic twins are detected early in utero.

REFERENCES

[1] Bondeson J. Dicephalus conjoined twins: A historical review with emphasis on viability. *J Pediatr Surg* 2001; 36: 1435-1444.
[2] Singhal AK, Agarwal GS, Sharma S, Gupta AK, Gupta DK. Parapagus conjoined twins: complicated anatomy precludes separation. *J Indian Assoc Pediatr Surg* 2006; 11(3):145-147.
[3] Harma M. Harma M, Mil Z, Oksuzler C. Vaginal delivery of dicephalic parapagus conjoined twins: case report and literature review. *Tohoku J. Exp Med* 2005; 205: 179-185.
[4] Barth RA, Filly RA, Goldberg JD, Moore P, Silverman NH. Conjoined twins: prenatal diagnosis and assessment of associated malformations. *Radiology* 1990; 177: 201-207.
[5] Mabogunje OA, Lawrie JH. Conjoined twins in west Africa. *Archives of Disease in Childhood* 1980; 55: 626-630.
[6] Kaufman MH. The embryology of conjoined twins. *Childs Nerv Syst* 2004; 20: 508-525.
[7] Romero R, Pilu G, Jeanty P, Ghidini A, Hobbins JC. Prenatal diagnosis of congenital anomalies. Ed. Norwalk: Appleton and Lange; 1988. pp 405-409.
[8] Spencer R. Theoretical and analytical embryology of conjoined twins: part I: Embryogenesis. *Clin Anat* 2000; 13: 36-53.
[9] Spencer R. Theoretical and analytical embryology of conjoined twins: part II: Adjustments to union. *Clin Anat* 2000; 13: 97-120.
[10] Owolobi AT. Oseni SB, Sowande OA, Adejuyigbe O, Komolafe EO, Adetiloye VA, Komolafe A, et al. Dicephalus dibrachius dipus conjoined twins in a triplet pregnancy. *Trop J Obstet Gynaecol* 2005; 22: 87-88.

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DECLARATION ON COMPETING INTERESTS:

No competing Interests.

Date of Submission: **Apr 18, 2011**
Date of Peer Review: **Jun 03, 2011**
Date of Acceptance: **Jul 01, 2011**
Online First: **Jul 25, 2011**
Date of Publishing: **Aug 08, 2011**