

# Cardiovascular anomalies in a dicephalus dipus dibrachius twin

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## Abstract

Conjoined twins of dicephalus type are extremely rare. We present a report of stillborn di-cephalus dipus dibrachius twin. There were two complete heads on two necks, one thorax, one abdomen, and externally normal two arms and two legs. From a thorough understanding of the anatomy of these twins we can gain insight into their developmental process and devise appropriate surgical strategies for separation.

Key words: Conjoined twins; Dicephalus; Parapagus; Teratology

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## Introduction

Dicephalus dipus dibrachius is an extremely rare form of conjoined twinning in which the infant has two arms, two legs, one trunk, but two heads. [1] Conjoined twins represent one of the rarest forms of congenital anomalies with an incidence of 0.2 : 10,000 pregnancies, but only 0.05 : 10,000 live births.[2]

The definitive cause of dicephalus twinning is unknown, and there are no known risk factors that are specific to this anomaly. In a study conducted by Tippi et al., 28% of conjoined twins died during intrauterine life, 54% during immediate postnatal life and 18% survived [3]. The present case is reported with an emphasis on the complex cardiovascular malformations which were noted in each twin since the architecture of the heart becomes the most vital organ during separation and survival of the twins.

## Case report

A 24 year old woman, primigravida, at 32 weeks of gestation delivered a set of conjoined twin(Fig.1) by caesarean section. The pregnancy was conceived spontaneously. The patient denied any contact with potential hazards like any teratogenic agents. There were no congenital anomalies in the obstetric history of the other siblings of the family. The twin had two heads and necks and fusion of the trunk below the neck with the right head is in better alignment with the spine(Fig.1&4). Both babies showed normal features with mild degree of cleft upper lip in right twin. Single pair of upper and lower limbs with no obvious congenital malformations were noted. The abdomen was soft with a single umbilical cord containing two arteries and one vein. Examination of the genitalia showed an ambiguous genitalia and a patent anus between genitalia and coccyx bone. Posterior aspect of the twin presented meningomyelocele slightly to the left of the median plane.

Babygram (Fig.4) showed two skulls and two separate vertebral columns which have an articulation with a single bony pelvis. There were two separate sets of ribs with four clavicles, of them lateral two were taking part in shoulder girdle formation and medial two overlapping each other posteriorly without giving any attachments.

## Autopsy findings

When viewed from the posterior aspect twin 1 was on the observer's right and twin 2 on the observer's left. The terms "right" and "left" will hereafter be used in relation to each twin (Fig.2). Both the twins shared a common thoracic cavity with a single sternum. When the thoracic cavity was opened a common anterior mediastinum was found. On opening the pericardium, two separate, closely apposed hearts were exposed. The two cardia are placed side by side. The left heart was slightly larger than the right and were joined at the atrial levels. Four atrial appendages were present, two for each twin. The atrial complex was a common chamber posterior to the ventricles. A single large tributary which could be taken as inferior venacava drains into the common atrial chamber. In this case, the heart could not be assigned to either twin and therefore, it is referred to as the shared heart within a common peri-cardial sac(Fig.3).

In right twin(Fig.2&3), a small tributary joins the right atrial chamber and is formed by the joining of internal jugular vein and subclavian vein of the right side. This could be taken as superior vena cava of the right twin. The two atria have communicated with each other through a wide communication, which could be taken as an en-larged foramen ovale. Left atrium on receiving the pul-monary veins, opened into a common atrial chamber. Two ventricles were separated by an incomplete septa with no outflow tract from the right ventricle. While the aorta arose from the left ventricle arching towards right side and continued as right descending thoracic aorta. A small branch from the concavity of aortic arch, close to heart bifurcates into two to supply both the lungs on the right side thorax cavity.

In left twin(Fig.2&3), a large tributary to the left of the former tributary which is formed by the joining of both internal jugular veins, inferior thyroid vein and subclavian vein of the left twin with the left internal jugular vein of the right twin. This could be taken as superior vena cava of the left twin. The right and left ventricles are separated by a distinct interventricular septum. The pulmonary trunk takes its origin from the right ventricle and is giv-ing branches to both the lungs which are on the left side. Ductus arteriosus is a wide vessel and has the same cali-ber as of pulmonary trunk and is connecting the right ven-tricle with the aortic arch close to the origin of the left subclavian artery. Ascending aorta has its origin from the left ventricle and is continued as the arch of aorta which has a normal branching pattern and is then continued as left descending thoracic aorta. The right and left descend-ing thoracic aorta have joined with each other just above the diaphragm to form a common descending thoracic aorta which has an opening in the diaphragm to be con-tinued as common abdominal aorta which has a normal branching pattern. Upon an interior dissection it is observed that the two atria have a wide communication which could be a wide patent foramen ovale and this common atrial cavity has a communication with a remnant of a possible common sinus venosus (Fig.3). Upon dissection of the interior of the ventricles of both the twins, it is observed that the atrio-ventricular communication is present in both cases with the cusps of the wall which is hypoplastic. The chor-dae tendinae of the papillary muscles are reaching the cusps. The interior of the both ventricles of either twins present only two papillary muscles, chordae tendinae of each reaching the cusps. Ventricular wall musculature shows that trabaculae cornae are not well formed.



Figure 1. Photograph of Dicephalus dipus dibrachius twin

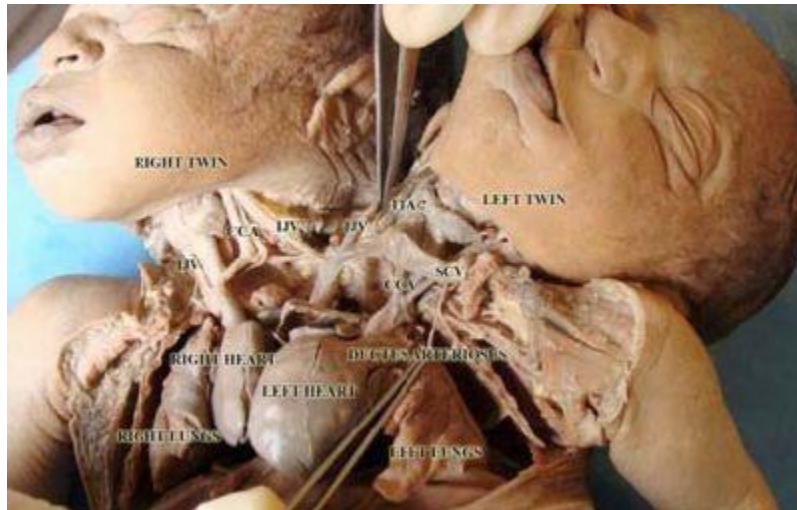


Figure 2. Anterosuperior view of Dicephalus twin showing conjoined heart and its great vessels.

Abbreviations [Figure 3]

IJV : Internal Jugular vein

CCA : Common carotid artery

SCV : Subclavian vein

SCA : Subclavian artery

ITA : Inferior thyroid artery

ITV : Inferior thyroid vein

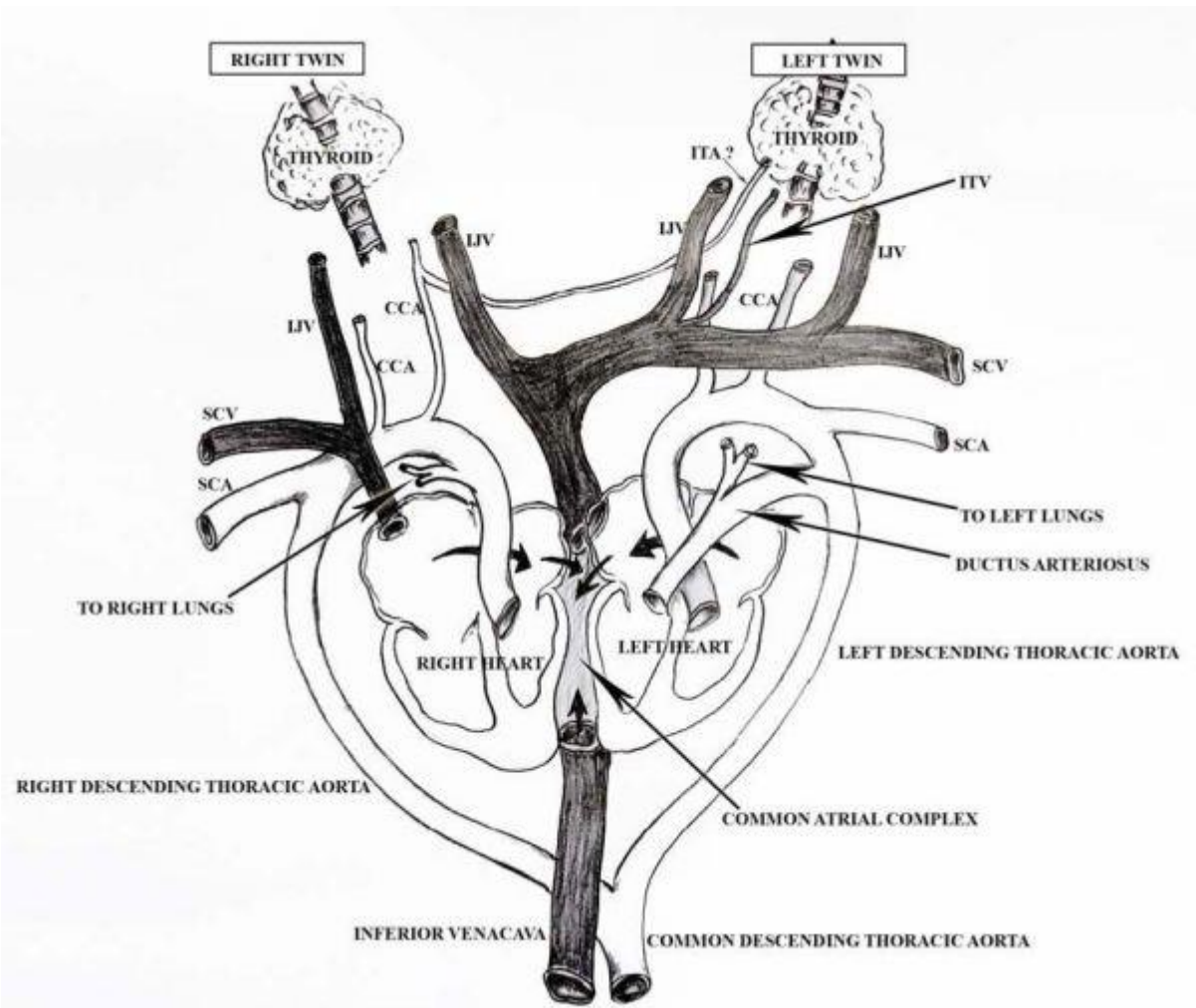


Figure 3. Schematic illustrations of the conjoined hearts and its great vessels

## Discussion

Cranial duplication are found in 75% of all double mon-sters, but duplications of the entire head and neck are uncommon.[4]. Two theories have been proposed to explain conjoined twinning. More widely accepted is the “fission theory” which states that conjoined twins occur when a fertilized ovum begin to split into identical twins, but somehow interrupted during the process and develops into partially formed individuals who are stuck together [5]. The second theory was proposed by Spencer, which states that a fertilized ovum divides completely into two embryonic discs whose unusual proximity results in sec-ondary fusion into conjoined twins as the embryo en-larges.[6]

Dicephalic twins with duplicated axial structures seems to be readily induced by fission accidents. The extreme examples of these fission – produced twins would be the production of identical twins by complete separation of the embryo into equal halves at any time after the first cleavage to the blastocyst stages.[7] Newman noted : The existence of mirror image symmetry in the components of double monsters argues against the theory that they have arisen through the fusion of separate embryonic axes.[8] Complex anomalies of the heart are seen more often in dicephalus than other types of conjoined twins, and might result from disturbed cross – signaling between tissues in adjacent primitive streaks, as suggested in animal mod-els.[9,10]

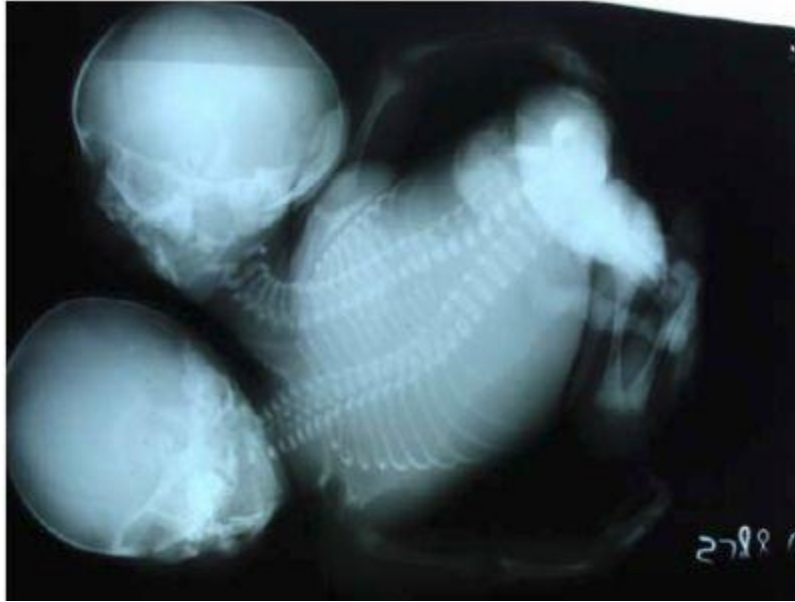


Figure 4. Radiographic image of the dicephalus dipus dibrachius conjoined twin

Partlow and his collaborators suggested that a transverse tension in the anterior part of the disc had caused a split in the chordamesoderm that divided the notochord and led to an anteriorly branched neural structures. If a blastocyst were to adhere to the uterine surface by that part of the blastocyst surface where the inner cell mass was positioned, resistance to expansion of the developing amniotic roof might result. This should compress the disc as it attempting to expand. Since the primitive node is very nearly central in the blastodisc at its first appearance, it is at the focus of forces applied peripherally on the disc. If adhesion were firm and persistent, compression of the disc might be sufficient to cause flattening of the anterior aspect of the primitive node, with bifurcation of axial structures as a consequence.[11]

Spencer concluded that fission of the developing embryo is unlikely result in conjoined twins, but that secondary fusion of two originally separate monovular embryonic discs could explain the conjoined twins. Also, he postulated that two early monovular embryonic discs may lie adjacent to one another at various angles and planes, as though floating on the surface of one sphere (the yolk sac) or on the inside of another, and may become secondarily united rostrally, caudally, laterally or dorsally, symmetrically or asymmetrically, but always homologously [12]

the embryo into equal halves at any time after the first cleavage to the blastocyst stages.[7] Newman noted : The existence of mirror image symmetry in the components of double monsters argues against the theory that they have arisen through the fusion of separate embryonic axes.[8] Complex anomalies of the heart are seen more often in dicephalus than other types of conjoined twins, and might result from disturbed cross – signaling between tissues in adjacent primitive streaks, as suggested in animal models.[9,10] Embryonic adjustments to conjunction

The fusion of two embryonic discs often requires one or both of two adaptations of the anatomic features destined to be formed at the site of union : 1) Division and diversion of midsagittal structures (i.e., oropharyngeal or cloacal membranes) resulting in separation of the primordial into equal halves that are displaced laterally and then united, one half from each twin, to form two relatively normal structures at right angles to the normal location, and 2) Aplasia of contiguous midsagittal and lateral primordial (i.e., faces, genitalia, limbs and body walls) that fail to develop, in whole or in part, as though obliterated by overlapping of developmental fields.[12]

The first mechanism, division and diversion is necessary because of the impossibility of simply burying external structures at the site of union, which would involve fusing intact ectoderm. Instead, either the oropharyngeal membrane or the cloacal membrane is divided sagittally, the two halves diverted laterally, then each half united to that of the other twin.

The embryonic aberrations found only in conjoined twins and only at the site of union are – Structures at the rostral aspect of the early embryonic disc may be involved in the union of ventrally conjoined twins, the temporal-spatial

relationship determining the extent of the fusion. Minimal rostral union will involve only the septum transversarium, but as the fusion becomes more extensive, the cardiac primordium will be included and finally, the oro-pharyngeal membrane and all the structures derived from and adjacent to it. The primitive cardiogenic areas unite after they come to reside in the (future) thorax, one from each twin on each side of the shared foregut, thus forming two jointly owned hearts. The thoracic and abdominal aortas usually follow the vertebral column of one individual fetus and provide the major blood supply for the structures of that twin.[12]

The presence of two hearts suggests that anterior ends of the early embryonic axes were separate and developed independently since the heart initially develops from cardiogenic mesoderm located cranial to the notochord. The fused "auricles" and sinus venosus suggest juxtaposition of the hearts at the stage when the atria are formed but before the definitive pleural and pericardial cavities were formed.

In the process of separation of tubular heart, separation of the common ventricular cavity has happened with a well formed interventricular septum and in continuity the conducting vessels have taken separate origin from the corresponding ventricles – aorta from left ventricle and pulmonary trunk from the right ventricle. In this case, instead of being connected with the left pulmonary artery, the ductus arteriosus is directly connected to the right ventricle and is of bigger caliber. The separation of common atria has happened but the communication between them is very wide and sinus venosus has not been partitioned.

With the presentation of both fission and fusion theories, it is observed by the present author that this particular case is more in favour of fission theory with a word of caution that the separation has happened only in the cranial part but has not extended into caudal part.

## **Conclusion**

The presence and extent of cardiac conjunction is a major determinant of the separability of conjoined twins. Cardiovascular evaluation must be directed to establish whether there is only a pericardial union, whether there is atrial connection but ventricular separation, and whether there is a ventricular connection.

No operative procedure is offered where there is complex cardiac union without the possibility of reconstructing even a single functioning heart. In our case, the anatomic structure often is such that it is unlikely that both twins will survive an attempt at separation.

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