

# A CASE OF THORACOPAGUS TETRABRACHIUS TETRAPUS

## Anatomical and Obstetrical Aspects

by

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### *Introduction*

The great rarity of conjoint twins demands a detailed study of all such freaks. The problem has been of interest to embryologists and obstetricians alike. But recently, the subject has assumed a greater importance, since the surgical separation of Siamese twins of Kano by Ian Aird (1954). This problem of surgical separation demands that the gross anatomy of these cases should be very well known. Therefore, the obstetrical and anatomical aspects of a case of Thoracopagus are being presented below.

### *Case Report*

T. R., aged 28 years, was admitted for her fourth delivery on 14th April 1960 at 10 P.M. She was in labour since 6 P.M. membranes had ruptured at 7 P.M., and a foot had prolapsed at 8 P.M. Of her 3 previous pregnancies, the second had ended in a twin delivery with survival of the second baby.

The uterus was tense, presentation and

position were difficult to make out. Foetal heart sounds were not heard. Uterus was contracting every 3 minutes. When examined internally, both legs with breech of a foetus were found at the vulva, cord was palpable, but there were no pulsations. On deeper examination a bridge of soft tissue was felt connecting the two bodies, possibly at a level above the umbilicus.

As contractions of the uterus were strong, it was decided to terminate labour by the vaginal route under anaesthesia with a provisional diagnosis of a xiphopagus conjoined twin. An attempt to turn the joint area, so as to deliver the babies without destruction, had to be given up for lack of uterine relaxation and space.

The fourth leg was pulled out, and with traction on legs it was noticed that there was a common cord to both foetuses. Deeper examination revealed a thoracopagus union. A Jardine's hook was passed and, in the attempt to separate the two foetuses, it was found necessary to cut viscera which included a liver bridge and a heart. When arms were delivered, it was found impossible to extract one head at a time. Decapitation of the larger foetus completed the delivery. The third stage and puerperium were uneventful.

### Anatomical Findings

The conjoint twins were full-term females, weighing 6 lbs. 14 oz. Externally they presented a gap in thorax and upper abdomen, through which the viscera of the two were in continuity. Both upper and lower extremities of both the foetuses were fully developed and did not show any evidence of malformations.

The line of fusion extended from the third ribs of both the foetuses and continued down to the hypochondria and epigastrium. As a result of this, the sternae were represented by two cartilaginous manubria. In this area, the line of fusion involved all the layers from skin down to the bones. The union of the twins was of face to face type.

**Right Foetus.** On opening the thoracic cavity, a very large thymus was seen, completely masking the left lung. The two pleural cavities were well formed and fully separated from other cavities. Right lung was divided into four lobes, the accessory lobe being the infracardiac one. Left lung was made up of three lobes, with a very well marked lingula.

The two hearts were united along their adjacent surfaces. The heart of the right foetus was a four chambered one, showing an interatrial septal defect. The right atrium received the two vena cavae. The left atrium received three pulmonary veins at its right end and was fused with the sinus venosus of the heart of the left foetus. The mitral opening was very wide. Tricuspid opening was normal. The auricular appendices were well formed. The left ventricle was fused with the right ventricle of the left foetus on its external aspect only. The cavities of the two did not communicate. The papillary muscles of the mitral valve were rudimentary. Aortic valve was normal. Right ventricle, tricuspid and pulmonary valves were normal. The relative positions of aorta and pulmonary trunks were normal. Ductus arteriosus was normal. No interventricular septal defect was found. The arch of the aorta gave origin to right common carotid and right subclavian arteries instead of the brachiocephalic.

In the heart of the left foetus, the atria were represented by a common chamber

which received the entire venous drainage of the body, namely, the right and left superior vena cavae, inferior vena cava and two right and one left pulmonary vein. From the right and left aspects of this chamber the respective auricular appendices were projecting. This chamber communicated with left atrium of the heart of the right foetus, and can be called a Sinuatrial chamber of the heart of the left foetus. Right ventricle was massive and occupied the anterior aspect. The left ventricle occupied a posterior position. The two ventricles freely communicated with each other through a wide interventricular septal defect. There was a dextroposition of aorta and pulmonary trunk with both the vessels arising from both the ventricles. The valves of the heart, ductus arteriosus and branching of arch of the aorta were normal.

The right lung was divided into four lobes including an infracardiac lobe, and the left lung was divided into two lobes only.

The twins had a common diaphragm, lying at the normal level. The diaphragm had no congenital defects.

The two livers were united in such a way that the fusion took place on the right surfaces of both the livers. It can be surmised that this took place by a rotation of the right liver through 180°, as evidenced by the dextroposition of the gall bladder and entry of inferior vena cava into the liver. This rotation was permitted by persistence of a duodenal mesentery. Consequently, the right common bile duct opened on the right side of the duodenum. A single umbilical vein entered the midline portion of the liver and divided into right and left branches. A tongue-like process of liver tissue overlay that spot.

Rest of the gastrointestinal tracts of both the foetuses were separate. Stomach, pancreas and spleen were normal in both foetuses. Except for the presence of Meckel's diverticulum in both the foetuses, gastrointestinal tracts did not show any abnormality.

Both the kidneys of the right foetus and the right kidney of the left foetus were polycystic. All the four ureters were normal.

In both the foetuses, the urachus was patent near the bladder end.

Central nervous system was not examined. No evidence of abnormalities of skull or spine was seen.

The twins had a single umbilical cord which, on section, was found to be made up of one umbilical vein and four umbilical arteries.

#### *Comment*

The findings of multiple pairs of one set of extremities during an internal examination immediately arouses in the mind of the obstetrician the possibility of monoamniotic twins either separate or conjoint. A thorough search proved that no heart sounds were heard. Though pulsations were not felt in the prolapsed cord, auscultation of foetal heart sounds of good rate would require an immediate caesarean section with the possible salvage of a separate monoamniotic twin. Subsequent to the paper of Quigley (1935), several others have reported the need for extra care in the salvation of foetus in this condition, chiefly because of risks of complications of the cord.

The obstetrician is also faced with a further dilemma on the method of delivery even after a diagnosis of conjoined twins has been made, if foetal heart sounds were heard. Is he justified in performing caesarean section to deliver Siamese Twins with a prospect of their subsequent existence and possible surgical separation? Or, is he justified in performing a destructive operation with the view that as in such cases, gross anatomical abnormalities and deficiencies in development are noticed? However, from a humanitarian point of view, even with the minimum possibility of survival, it should be

felt today that a caesarean section is justifiable.

It is generally believed that monozygotic twins form as a result of cleavage of fertilised ovum into two separate embryos. Rarely, this separation is incomplete, giving rise to conjoint twins. This splitting can occur any time, between the blastula and primitive streak stage. If it is interfered with, in the period between 13th to 15th day of postfertilisation period, Siamese twins will result.

After a careful scrutiny of the anatomical findings of this case, the authors are led to believe that conjoint twins can also develop, as a result of fusion between the monozygotic twins in a common chorioamniotic sac. This surmise is substantiated by the juxtaposition of the right lateral surfaces of two livers, complete development of the extrahepatic biliary system and the presence of four umbilical arteries and a single umbilical vein in the umbilical cord. The site of fusion of the two embryos can be made out at the gap in the anterior abdominal wall in the region of the future umbilicus and its upward continuation between the two sternal plates. The probable time relationship can be located to a period just before the herniation of midgut loop. At this stage, the two sternal plates are also separate. The findings in the hearts are also suggestive of a transient interference in the morphogenetic process at this stage.

#### *Summary*

A fourth gravida, aged 28, delivered conjoint twins. The delivery was obstructed due to compound

presentation. A provisional diagnosis of Xiphipagus conjoint twins was made. Deeper examination revealed thoracopagus union. Operative interference was unavoidable. Cutting the bridge which included liver and heart along with decapitation completed the delivery.

The twins presented a face to face union, in lower part of thorax and epigastrium. The hearts were united externally over the ventricles only. In the heart of the left foetus, persistent sinu-atrial chamber and left superior vena cava, interventricular septal defect and dextroposition of vessels were noticed. Lungs showed presence of accessory lobes. The two livers were united along their right lateral surfaces. Extrahepatic biliary apparatuses were fully developed in both the foetuses. Polycystic kidneys and persistent Meckel's diverticula were observed in both the cases. A single umbilical

cord, with four umbilical arteries and one umbilical vein, was present. On the basis of the anatomical findings, it has been suggested that in this case, fusion, rather than incomplete separation of monozygotic monoamniotic twins, is responsible for the genesis of this anomaly.

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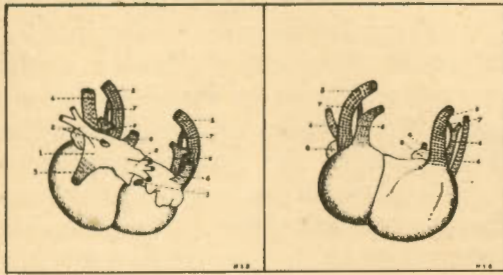


Fig. 1

Posterior and anterior aspects of the heart of conjoint twins.

1. Sinu-atrial chamber. 2. Pulmonary veins.
3. Inferior vena cava. 4. Superior vena cava.
5. Aorta. 6. Pulmonary trunk.
7. Ductus arteriosus. 8. Auricular appendix.

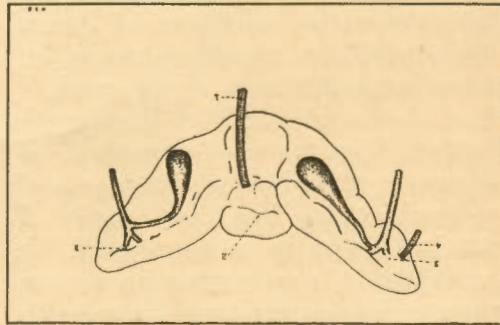


Fig. 2

Antero-inferior aspect of the livers of conjoint twins.

1. Umbilical vein. 2. Tongue like process.
3. Porta Hepatis. 4. Inferior vena cava.