CLINICOPATHOLOGIC CORRELATIONS

Conjoined Thoracopagus Twins

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SUMMARY A case of vaginally delivered stillborn female thoracopagus conjoined twins is presented. Each twin had a cleft lip and cleft palate. They shared a common pericardial sac and a common abdominal cavity. The liver was conjoined, and the small bowel was common to both twins. One twin was aplastic. Although the twins were monozygotic, their viscera were neither identical nor mirror-image to each other, and the visceral anomalies were unique to each twin.

The hearts were fused, and communications existed between the circulations at the atrial and ventricular levels. One twin had single atrium, solitary (right) ventricle, malposed great arteries, pulmonary valvular atresia and infundibular stenosis, right aortic arch with aberrant left subclavian artery and with left ductus arteriosus, and atresia of the common pulmonary vein. The other twin had two atrial septal defects of the fossa ovalis and sinus venosus types, partial anomalous pulmonary venous connection, persistent left superior vena cava to coronary sinus, solitary (left) ventricle, malposed great arteries, and left aortic arch with aberrant right subclavian artery and with left ductus arteriosus.

There was atrial and ventricular fusion. On hypothetical grounds, and in a comparable anatomic situation, salvage of one twin might be possible by surgical means, while the other twin was sacrificed.

IN THE PROCESS OF DEVELOPMENT of two individuals from a single fertilized ovum, a spectrum exists including parasitic monsters, conjoined twins, and two separate monozygotic twins. Conjoined twins are classified according to the area of union, the most common site being the anterior thoracic and upper abdominal midline (thoracopagus). Although most thoracopagus twins are externally symmetrical, their viscera are not necessarily either identical or mirror-images of each other. In this report, the findings in a case of thoracopagus conjoined twins are presented, with particular emphasis on the complex cardiovascular malformations which were unique to each twin. Consideration is given to potential for surgical correction by sacrifice of one of the twins.

Clinical Findings

The mother was 24 years old and had had one previous uncomplicated pregnancy. Both pregnancies were fathered by the same person. Neither the father nor the mother had family histories of twin gestations or congenital anomalies. In the current case, twins were suspected before birth, even though two separate heart beats were never documented. The prenatal course was characterized by polyhydramnios but was otherwise uncomplicated. Spontaneous labor developed during the twenty-ninth week of gestation.

At that time, fetal heart tones were absent. By abdominal roentgenography, two fetuses were identified in unusual position and attitude, with both fetal heads in vertex position. During delivery, thoracopagus conjoined twins were identified. Vaginal delivery was accomplished, utilizing Elliot forceps for the first twin and internal cephalic version of the second twin. Each twin was a stillborn female.

One large placenta was delivered, with one chorion and one amnion. A single umbilical cord was present, which contained two arteries and one vein.

Autopsy Findings

Though joined ventrally, the twins faced each other somewhat obliquely, like a partially-opened book. The fetus to the observer's left will be called Twin A, and the one on the right will be called Twin B. The terms "right" and "left" will now be used only in relation to each twin.

The combined mass of the twins was only 1500 grams. The crown-heel length, head circumference and chest circumference of Twin A were 14.5, 10.5 and 12.0 cm, respectively, while corresponding measurements for Twin B were 13.5, 10.3 and 12.0 cm. The blood type of each twin was A-negative, the same as that of the mother.

Twin A had a cleft lip and cleft palate to the left of its midline, while Twin B had the same abnormalities to the right of its midline. Although the twins appeared as mirror-images externally, their thoraco-abdominal organs were not mirror-images of one another. The twins were joined from the sternal manubrium to the level of the single umbilicus.

The twins shared a common anterior mediastinum, with fusion of the pericardial sacs, cardiac structures and thymus glands. The sternum was absent in each twin. The peritoneal cavities of the twins were continuous above the level of the umbilicus and associated with a common upper anterior abdominal wall. The liver was conjoined and contained two gallbladders. Twin A had one spleen, while no spleen was present in Twin B.

Each twin had its own esophagus, stomach and proximal duodenum. The second part of each duodenum fused to form a common distal duodenum. There was also a common jejunum and ileum for both twins. A Meckel diverticulum measuring 2.0 cm in length was present. Immediately beyond this structure, an ileum and colon continued in Twin A, and the colon arose in Twin B.

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Each twin had a normally-formed female urogenital system, pancreas, pair of adrenals, central nervous system and respiratory system. Each twin had a trilobed right lung and a bilobed left lung.

**Cardiovascular System**

The thoracopagus twins had a common pericardial sac. There was also atrial and ventricular fusion, and inter-cardiac communications existed between the atria and between the ventricles. Figure 1a and b summarize the anatomic findings.

**Atrial Complex and Pulmonary Veins**

In Twin B, two atria were present (fig. 2a), while in Twin A a single atrium existed. The latter chamber was in free communication with the right atrium of Twin B. Four atrial appendages were present, two for each twin. In the single atrium of Twin A, a cord-like muscle bundle represented the only tissue resembling a septum. The superior and inferior venae cavae of Twin A joined the right side of the single atrium, but neither a coronary sinus nor pulmonary veins joining the atrium were present for this twin (fig. 2b). The pulmonary veins joined a common venous sinus from which no gross vessel emanated (so-called atresia of common pulmonary vein).

The atrial portion of Twin B was partially divided into right and left chambers. This partition was comprised of a delicate fibrous membrane (septum primum?) with total absence of tissue resembling septum secundum. Two atrial septal defects were present, one being of the fossa ovalis type and the second being present near the junction of the superior vena cava with the right atrium of this twin.

In Twin B, bilateral superior venae cavae and a single inferior vena cava were present. The left superior vena cava joined the coronary sinus. The right atrium also

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**Figure 1.** Schematic diagram of the observed cardiac structures. a) The atria and ventricles. Twin A has a single atrium, while Twin B has right and left atria (RA(B) and LA(B)) with two atrial septal defects. In Twin A, superior and inferior venae cavae (SVC (A) and IVC (A)) join the single atrium of that twin, while similar structures (SVC (B) and IVC (B)) join the right atrium of Twin B. The persistent left superior vena cava (LSVC) of Twin B joins its coronary sinus (CS). In Twin A, the pulmonary veins (insert) join to form a blind sac (PV (A)). In Twin B, the right upper pulmonary vein (PV-RU) joins the right atrium of that twin near a sinus venosus type of atrial septal defect, while the other pulmonary veins (PV-LU; PV-LL; PV-RL) of Twin B join its left atrium. Only two atrioventricular valves are present, one resembling a tricuspid valve (TV) on the side of Twin A and a mitral valve (MV) on the side of Twin B. The ventricular mass comprises two ventricles, that of Twin A resembling a right ventricle (RV) and that of Twin B resembling a left ventricle (LV). Two ventricular septal defects (VSD (I) and (II)) are present in the septal wall between the ventricles of Twin A and that of Twin B. b) The ventricles and great vessels. From the ventricle of Twin A (RV), which resembles a right ventricle, arise the great vessels of that twin. These are malposed and associated with atresia at the pulmonary valve. The aortic arch is right-sided and gives rise to an aberrant left subclavian artery (LS), while the ductus arteriosus (LDA) arises at the origin of the left common carotid artery (LC) and terminates in the left pulmonary artery. From the ventricle of Twin B (LV), which resembles a left ventricle, arise the great vessels of that twin. These are also malposed, but without pulmonary stenosis. The aortic arch is left-sided and gives rise to an aberrant right subclavian artery (RS). The ductus arteriosus (LDA) is left-sided, arising proximal to the level of the left subclavian artery and terminating in the left pulmonary artery. Ao = ascending aorta, PT = pulmonary trunk, RPA = right pulmonary artery, LPA = left pulmonary artery, RC = right common carotid artery.
anomalously received the right upper lobe pulmonary vein of this twin, while its left atrium received the veins of the right lower lobe and entire left lung.

**Ventricular Complex**

Externally, the ventricular structure was represented by one mass common to the two twins. Internally, half of this received blood from the common atrium of Twin A and blood from the right atrium of Twin B. This half of the ventricular mass will be called the "ventricle of Twin A." The other half of the ventricular mass received blood from the left atrium of Twin B and will be called the "ventricle of Twin B." From the exterior, the apex of the ventricle of Twin A was directed superiorly toward the great vessels of Twin B, while the apex of the ventricle of Twin B was directed inferiorly and toward Twin A (fig. 3a). Internally, the ventricle of Twin A was a solitary chamber having the morphologic features of a right ventricle, while the ventricle of Twin B was a solitary chamber having the morphologic features of a left ventricle.

The two ventricular chambers shared a common wall or septum in which there were two defects. The first was small and lay behind the tricuspid septal leaflet (to be described) of Twin A. The second defect was larger, and was represented by an oblique channel in the muscle of the septum (fig. 3b).

**Atrioventricular Valves**

Two distinct atrioventricular valves were present (fig. 4). One had the structure of a tricuspid valve and arose partially from the single atrium of Twin A and partially from the right atrium of Twin B. It led entirely into the ventricle of Twin A. The other atrioventricular valve resembled a mitral valve and arose entirely from the left atrium of Twin B. It led into the ventricle of Twin B. The tricuspid valve was wider than the mitral valve.

**Conotruncus**

From the ventricle of Twin A (resembling a right ventricle) arose both great arteries of that twin (fig. 5a). These were malposed, the aorta arising anteriorly and to the right of the pulmonary trunk. Although the aortic valve was normal, the pulmonary valve was atretic and beneath it was a stenotic infundibular channel. There was discontinuity between the tricuspid valve, on one hand, and the aortic and pulmonary valves, on the other, yielding a picture of bilateral coni.

From the ventricle of Twin B (resembling a left ventricle) arose both great arteries of that twin (fig. 5b). These vessels were malposed, the aorta arising anteriorly and to the right of the pulmonary trunk. The aortic and pulmonary valves were normal. Although there was a mitral-pulmonary valvular continuity, the mitral and aortic valves were separated by a muscular subaortic conus.

**Aortic Arch**

Twin A had a right aortic arch with aberrant origin of the left subclavian artery as the fourth branch of the arch (fig. 5a). There was no right ductus arteriosus, but a left ductus arteriosus was present and ran from the aorta at the level of the origin of the left common carotid artery, above, to the left pulmonary artery, below.

Twin B had a left aortic arch with aberrant right subclavian artery (fig. 5b). Only a left ductus arteriosus was present; this joined the aortic arch proximal to the level of origin.
of the left subclavian artery and inserted into the left pulmonary artery.

Coronary Arteries

In each twin, the aorta arose in front of the pulmonary artery. Each aorta had two coronary ostia, and the anterior aortic cusp was the noncoronary cusp in each twin (fig. 6). In each twin, its left coronary artery gave rise to the anterior descending and circumflex branches. The right coronary artery gave off a conus branch in each twin. At the base of the ventricular mass, there was direct anastomosis of the right coronary artery of Twin B to the circumflex branch of Twin A. The posterior descending artery arose in Twin A from its right coronary artery and in Twin B from its left circumflex artery.

Comment

Conjoined twins develop from a single fertilized ovum and result from incomplete fission of the embryo before the third week of gestation. Although monozygotic twins occur in 0.39-0.41% of births, conjoined twins are much rarer and account for only 0.002% of births. Conjoined twins have a striking sex predilection, with 70-95% being females. Most are premature, and many are stillborn. Conjoined twins are classified according to the site of union, about 75% having anterior thoraco-abdominal fusion (thoracopagus). The present case is that of stillborn female premature thoracopagus conjoined twins.

In conjoined twins, the severity of visceral malformation depends upon the extent of the area of union. The pleural, pericardial and peritoneal cavities may be separate or may be common to both twins. In all described cases, as in ours, the livers are fused, and the hearts and portions of the gastrointestinal tract are usually fused in cases with large areas of union.

In reported cases, the sternum is found to be partially or totally absent in all, the pericardial sac common in 90% and the hearts fused in 75%. The severity of cardiac fusion varies and includes (a) separate hearts, (b) atrial fusion with separate ventricles, and (c) atrial and ventricular fusion. To our knowledge, no case exhibiting fusion of the great vessels has been reported. In atrial fusion with separate ventricles, at least one of the hearts is usually severely malformed and often both are malformed. No cases of ventricular fusion without atrial fusion have yet been reported.

In the present case, the sternum was absent, the pericardial sac was common, and complex atrial and ventricular fusion was present.

The most common atrial malformations in thoracopagus twins include single atrium and large atrial septal defect. Often, at least one of the thoracopagus twins has a single atrioventricular valve or an atrioventricular canal. The most common ventricular malformations in thoracopagus twins include single ventricle and large ventricular septal defect.

Classically, although the great arteries are not fused, they are often transposed (or malposed in the case of single ventricle or double outlet ventricle) or exhibit persistent truncus arteriosus. Pulmonary atresia is relatively common.

The first essential for successful surgical separation of thoracopagus twins is to obtain live births. This is best accomplished by identification of the condition before labor sets in, in order that delivery by cesarean section may be done. Attempted vaginal delivery results in a high incidence of fetal death.

The antenatal diagnosis of thoracopagus twins may be suspected by roentgenographic study. The diagnosis of
Figure 4. a) The atrioventricular valve lying between the single atrium (SA (A)) of Twin A and the right atrium (RA (B)) of Twin B, on one hand, and the ventricle of Twin A ("RV"), on the other, has a tricuspid structure. The cord of muscle which attempts to form a septum between the single atrium of Twin A and the right atrium of Twin B is represented by the letter C. b) The left atrium (LA (B)), the mitral-like valve and the ventricle ("LV") of Twin B. So-called telangiectasias of the mitral valve are represented by the dark spots, a feature common in newborns.

Figure 5. a) The base of the heart, the great vessels, lungs and trachea of Twin A. The aorta (Ao) is malposed. The pulmonary trunk (PT) is narrow (the pulmonary valve was atretic). There is a right aortic arch, the first branch of which is the left common carotid artery (LC) arising at the same level as the ductus arteriosus (D). The latter inserts into the left pulmonary artery (LPA). Beyond the right common carotid artery (RC) and right subclavian artery (RS), in that order, is the aberrant left subclavian artery (LS) which passed behind the esophagus. b) The heart, great vessels and lungs of Twin B. The aorta (Ao) is malposed and arises anteriorly to the pulmonary trunk (PT). The aortic arch is left-sided. The ductus arteriosus (D) runs between the aorta and the left pulmonary artery. Its aortic insertion lies proximal to the level of origin of the left subclavian artery (LS). An aberrant right subclavian artery (RS) arises distal to the origin of the left subclavian artery and passes behind the esophagus. RC = right common carotid artery; LC = left common carotid artery.
thoracopagus twins should be suspected when the following conditions are observed: 1) twins facing each other; 2) fetal heads at the same level; 3) proximity of the thoracic cages; 4) backward flexion of the cervical spines; 5) absence of the flexed fetal position; and 6) no change in fetus-fetus relationship with respect to time, manipulation or fetal movement.1,7

The experience with attempted surgical separation of thoracopagus twins shows that greater degrees of success have been obtained when separate hearts are present than when there is cardiac fusion.8

After birth the diagnosis of separate hearts is best made by showing that one twin differs from the other with respect to 1) pulse characteristics, 2) other specific hemodynamic features,9,10 or 3) separate and independent QRS complexes by simultaneous electrocardiography.6,8 Angiography is the most reliable method of determining the presence of two separate hearts or a fused heart.

If the hearts are fused, and operation is not done, death usually occurs by the age of three months. Of seven thoracopagus twins with separate hearts in whom surgical separation has been attempted, six of the 14 individuals survived, an operative mortality of 57%.5 Of three cases of thoracopagus twins with conjoined hearts in which attempts at surgical separation have been reported, the surgical plan was salvage of both twins in one case and sacrifice of one member in each of the other two cases. None of the individuals survived the first postoperative day.5,10-12 In both of the solitary salvage cases, death was attributed to respiratory insufficiency rather than the cardiac repair.6 Respiratory insufficiency was contributed to by thoracic constriction from a tight skin closure, flail chest from absence of the sternum, and concomitant pneumothorax, atelectasis or pneumonia.5,13 In the case in which salvage of both twins was attempted, death of each member occurred during the operation. The moral and ethical considerations of planned sacrifice of one member are discussed at length by Pepper.14

While in our case the twins were stillborn, it is of interest to consider the potential for salvage in the face of the anatomic states observed. On hypothetical grounds, consideration might be given to saving Twin B while sacrificing Twin A (fig. 7a and b). Leaving aside the matter of fused livers and partial fusion of the intestinal tract, one may consider surgical approaches that might preserve the fused heart for Twin B. Surgically, the atrial and ventricular septal defects would be repaired, and the superior and inferior venae cavae of Twin A would be ligated and divided. The newly constructed right atrium of Twin B would be comprised of the single atrium of Twin A and the right atrium of Twin B. The morphologic right ventricle would serve as the functional right ventricle. The atretic pulmonary trunk arising from this chamber would be severed, and the aortic valve and aorta of Twin A would be utilized as the functional pulmonary valve and pulmonary trunk for Twin B. The ascending portion of this aorta would be sectioned and connected with a tubular graft to the distal pulmonary trunk of Twin B. The left atrium and left ventricle of Twin B would

![Figure 6. Semi-schematic diagram of coronary arterial supply. a and b) Viewed from in front and from above, respectively. The aorta (Ao) arises in front of the pulmonary trunk (PT) in each twin and in each the anterior aortic cusp is the noncoronary one. The left coronary artery passes in front of the pulmonary trunk in each twin and gives rise to anterior descending (LAD) and circumflex (Cir) branches. The right coronary artery (RC) has large conal (CA) branches in each case. The posterior descending branch (PD) arises from the right coronary artery in Twin A and from the left circumflex artery in Twin B. The right coronary artery of Twin B anastomoses with the circumflex branch of Twin A. Following the surgical procedures diagrammed in figure 7, the new pulmonary trunk would supply coronary arterial blood flow to the lateral and posterior walls of the right ventricle and possibly the anterior portion of this chamber also.](https://circ.ahajournals.org/doi/fig/10.1161/01.CIR.56.3.496)
function as such. The pulmonary trunk arising from the morphologic left ventricle would be ligated proximally. The aorta of Twin B would still function as such. The major postoperative problem would be whether or not the myocardium could function adequately with part of its coronary arterial supply coming from the newly-formed pulmonary trunk.

References

Conjoined thoracopagus twins.
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