Cardiovascular anomalies in thoracopagus twins and the importance of preoperative cardiac evaluation


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Four pairs of thoracopagus twins have been described. Cardiac catheterisation was performed in all the cases. Angiocardiographic and necropsy findings suggest that the most common abnormality was some form of univentricular heart. The communication between the 2 hearts was at atrial level in 2 cases. Separation was performed in 1 of these cases but only 1 of the twins survived for 14 hours after operation. It is suggested that full cardiac catheterisation with selective angiocardiogram is essential before separation is considered. Identical heart rates were observed in each pair and there was invariably a major communication between the hearts of the twins.

The incidence of conjoined twins has been estimated at about 1 in 50 000 births (Morrison, 1963; Amiel, 1967). Frequently the twins are born dead but more than 300 cases of survival, from a few hours to 63 years, have been documented (Tandon et al., 1974). Survival depends on the site of union of the twins and the resultant fusion of vital organs. Thoracopagus twins are joined from the anterior part of the thorax down to the umbilicus. Nearly 75 per cent of such twins have serious cardiovascular anomalies which influence the prospects of successful surgical separation (Leachman et al., 1967; Nichols et al., 1967). Ten attempts at surgical separation have been discovered in a review of the reports published since 1925, but there has not been a successful separation in twins sharing a heart or part of it (Mulcare et al., 1970). However, with recent advances in paediatric cardiovascular surgery, it should be possible to separate some of them, and hence it is essential to carry out full preoperative cardiovascular investigation.

The purpose of this communication is to report an attempted surgical separation and to review the cardiovascular abnormalities in 4 pairs of thoracopagus twins seen at The Hospital for Sick Children, London. In all 4, simultaneous electrocardiograms from the twins recorded an identical heart rate, suggesting the association of fused hearts.

Case 1

The twins, both female, were born after an elective lower segment caesarian section at 39 weeks' gestation. The pregnancy was uneventful and there was no history of any ingestion of drugs during pregnancy. The parents were both 29 years old and have one other child—a girl, 2 years old, who is normal. The combined weight of the twins was 4·9 kg. Respiratory difficulty was noticed soon after birth. The twins were intubated and transferred to The Hospital for Sick Children, Great Ormond Street.

On examination, they were joined across the chest and abdomen over a distance of 15 cm. Their facial appearances were normal. There was an infraumbilical hernia with a single umbilical cord containing two umbilical arteries and two umbilical veins. All the peripheral pulses were felt, at a rate of 90 per minute, and simultaneous limb lead electrocardiograms confirmed identical heart rates for the twins. Because of rapid deterioration in the condition of Twin B, cardiac catheterisation was performed and revealed the following.

TWIN A

A distinctly separate inferior vena cava led to the right atrium. The right ventricle was entered from
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the right atrium. The pulmonary artery originated from the right ventricle. Right ventriculography showed a free communication with the (morphological) right ventricular chamber of Twin B. There was a hypoplastic left ventricle giving rise to a hypoplastic aorta.

TWIN B
The right atrium was entered from the inferior vena cava. The same ventricular chamber was entered via two different atrioventricular valves. Ventriculography confirmed a primitive ventricle with a subaortic outlet chamber.

Cardiac arrest occurred soon after the angiocardiogram in Twin B. Despite resuscitative measures, the twins died.

Necropsy confirmed the angiocardiographic findings. In Twin A there was a mitral valve hypoplasia with hypoplastic left ventricle and bicuspid aortic valve. Twin B had a primitive ventricle with two atrioventricular valves and subaortic outlet chamber. There was a free communication between the two 'right' ventricular chambers of the twins.

In addition to the cardiovascular anomalies the small intestines between the duodenum to ileum were fused. There was also continuity between the liver tissue of the twins.

Case 2

The conjoined twins were admitted to The Hospital for Sick Children, Great Ormond Street, in August 1968, immediately after birth. The mother was a primigravida aged 24, and the twins were born by breech extraction at 32 weeks' gestation. They were female and their combined weight was 3.17 kg. They were joined face-to-face from about the third rib to the umbilicus.

On examination there was a single umbilical cord in the midline. They were both cyanosed and acidemic. There was no evidence of cardiac failure and the peripheral pulses were considered to be normal. Electrocardiogram showed synchronous electrical activity.

They underwent cardiac catheterisation and angiography on the day of admission. Because of the poor condition of the twins full data were not obtained, though it was clear that there was free communication between the two hearts at atrial level, and in both there was total anomalous pulmonary venous drainage. Operation was considered impossible and the twins died soon after cardiac catheterisation was completed.

Necropsy showed the following.

TWIN A
There was tricuspid and pulmonary atresia as well as total anomalous venous drainage to the coronary sinus. The right atrium communicated with the left atrium via a patent foramen ovale. There was a small partially closed ductus arteriosus which communicated directly with small pulmonary arteries.

TWIN B
Again there was total anomalous pulmonary venous drainage to the coronary sinus. In addition there was aortic atresia and a very small left ventricle (hypoplastic left heart syndrome). The mitral and tricuspid valves were normal. There was a communication between the left and right atria via a patent foramen ovale. The pulmonary arteries appeared normal and there was a huge ductus arteriosus.

There was free communication between the two hearts, from the right atria of Twin A to the coronary sinus of Twin B.

There was also a large fused liver. The gastrointestinal system was otherwise normal.

Case 3

The conjoined twins, both female, were admitted to The Hospital for Sick Children, in May 1969, aged 24 hours. They were born at 39 weeks' gestation by ceasarian section. Their combined weight was 5.2 kg. Their mother was 30 years of age and had had two previous miscarriages. The twins were joined from about 2 cm below the suprasternal notch downwards to the upper edge of the common omphalocele.

Fig. 1 Bipolar lead II electrocardiogram in case 3. Upper tracing (Twin A), lower tracing (Twin B) show identical ventricular rate. Ventricular extrasystole (beat 2) in both tracings. Sinus capture (last beat).
On admission their general condition was good and they were acyanotic. The peripheral pulses were all easily felt and there was no clinical evidence of congestive heart failure. The electrocardiogram showed nodal rhythm, and there was evidence of two electrically independent hearts (Fig. 1).

Cardiac catheterisation on the day of admission showed the following.

**TWIN A**
There was a single atrium and the two ventricles were connected by a ventricular septal defect. An inferior ventricle gave rise to an anterior aorta and an anterosuperior ventricle gave rise to a posterior pulmonary artery. The aorta and pulmonary artery appeared normal.

**TWIN B**
There was a single atrium and a single ventricle. The pulmonary artery and aorta appeared normal and were normally related. There was a ductus arteriosus with a shunt from pulmonary artery to aorta on angiography.

The two hearts communicated at atrial and ventricular levels.

These findings rendered surgical intervention impossible, but oral feeding was started. On the fourth day of life the twins became cyanosed with a gradual increase in respiratory rate and eventually died later that day.

Necropsy showed distortion of the thoracic cage with a cleft-like space on the right which had considerably reduced the space available for pulmonary development so that both lungs had a pronounced degree of hypoplasia. There was a single large fused heart shared by the twins, and the cardiac lesions were those described at catheterisation. There were thus three ventricles with one common chamber.

The bowel of both twins was normal. The liver of each twin was fused in the mid-line to form one very large organ weighing 150 g; separate gall-bladders and bile ducts were present and the pancreas in both cases was normal. No other abnormalities were detected.

**Case 4**

The twins, both female, were born after an elective lower segment caesarian section. The pregnancy had been uneventful, and the parents already had one normal child. Simultaneous electrocardiograms (Fig. 2) showed identical heart rates, and the twins were transferred to The Hospital for Sick Children, in May 1976. The combined weight of the twins was 4·8 kg. The twins were joined from the lower one-third of the sternum down to the umbilicus. There was a small omphalocele at the lower part of the junction. The external features of the upper and lower parts were normal. All peripheral pulses were felt and both twins were tachypnoeic. Twin B was moderately cyanosed.

At cardiac catheterisation, the following abnormalities were found.

**TWIN A**
A distinctly separate inferior vena cava led to the right atrium. A ventricle (morphological left ventricle) was entered from the right atrium. The ventricle lay horizontally across the thorax of Twin B (Fig. 3). The ventriculogram showed a primitive ventricle with an outlet chamber. Both the great arteries seemed to arise from the main chamber (Fig. 3a and 3b), the aorta being superiorly placed. Right atrial angiogram also opacified the inferior vena cava of Twin B.

**TWIN B**
There was a large communication between the two atria. A ventricle was entered from the left atrium. A ventriculogram confirmed primitive ventricle with obvious subpulmonary stenosis.

Left atrial injection showed the presence of a common atrium (Fig. 4). Oxygen saturations in Twin A and B suggested that the respective ventricles were separate.

It was decided to separate the twins, though the exact site of atrial communication was not known. At operation there was a common pericardial sac. There was communication between the right atrium...
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Twin A
Cephalad
Subaortic outlet chamber
Pulmonary artery
Catheter
Primitive ventricle
Caudal

Twin B
Cephalad
Aorta
Rt. atrial appendage
Rudimentary chamber
Pulmonary artery
Catheter in rt. atrium
Primitive ventricle
Caudal

Fig. 3 (a) Angiocardiogram in ventricle of Twin A and (b) line drawing of (a) show primitive ventricle with rudimentary outlet chamber (superior). Aorta is superior to pulmonary artery.

of twin B and the coronary sinus of twin A. Separation of the cardiovascular communication was achieved. The liver was divided and in twin A duodeno-ileal anastomosis was performed. Twin B died towards the end of the procedure and twin A died 14 hours later.

Necropsy showed a primitive ventricle with transposition of the great arteries in twin A. There were two atrioventricular valves leading to the common ventricle. Twin B had tricuspid atresia and pulmonary stenosis with a rudimentary right ventricle. The large left ventricle gave rise to the aorta and the pulmonary arteries were supplied by a ductus arteriosus. There was also a common atrium. The mitral valve was normal.

Discussion

The nature of the cardiovascular anomalies in our series of thoracopagus twins has been summarised

Fig. 4 Angiocardiogram of left atrium (Twin B) shows opacification of inferior vena cava. There is a primitive ventricle, a superior rudimentary outlet chamber, and pulmonary stenosis.
in the Table. The most common abnormality was some form of univentricular heart. The communication between the two hearts was at ventricular level in cases 1 and 3. Hence only cases 2 and 4 were potentially suitable for separation. However, the severe cardiovascular anomalies in the individual hearts would have required palliative procedures as well.

Cardiovascular anomalies were described in 45 sets of twins by Nichols et al. (1967) who found that 75 per cent of thoracopagus twins had common hearts and in 90 per cent there was a common pericardial sac. The cardiac malformations in their series were as follows: 2 separate ventricles (10 cases); 3 ventricles (12 cases); 2 ventricles (1 for each twin) (19 cases); and 1 ventricle for both (4 cases).

Specific cardiac malformations included pulmonary atresia and absence of ventricular septum. The cardiovascular abnormalities in our own cases are in accord with those described in this series.

The importance of preoperative cardiac evaluation has been emphasised by Simpson et al. (1970). Electrocardiographic examination in the twins seems to be a reliable guide as to whether the hearts are fused; identical heart rates were recorded in all our 4 cases and there was invariably a major communication between the hearts of the twins (Fig. 1 and 2).

Full cardiac catheterisation with selective angiocardiograms is essential before separation is considered. The exact timing of the separation is often difficult, but in our series all the twins were extremely ill and required urgent investigations. Even after successful separation, the presence of severe cardiovascular lesions may make survival unlikely without further palliative surgical procedures for each twin.

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References


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