Echocardiographic assessment of conjoined twins.


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ABSTRACT

Background: To determine the accuracy of pre- and postnatal echocardiography in delineating the degree of cardiac fusion, intracardiac anatomy (ICA) and ventricular function of 23 sets of conjoined twins with thoracic-level fusion presenting to a single centre over a 20 year period.

Methods: We assessed 13 thoracopagus, 5 thoraco-omphalopagus and 5 parapagus pairs presenting to our institution between 1985 and 2004 inclusive. Echocardiographic data was analyzed together with operative intervention and outcome. Twins were classified according to the degree of cardiac fusion: separate hearts and pericardium (group A, n=5), separate hearts and common pericardium (group B, n=7), fused atria and separate ventricles (group C, n=2), and fused atria and ventricles (group D, n=9).

Results: The degree of cardiac fusion was correctly diagnosed in all but one set. ICA was correctly diagnosed in all cases, although the antenatal diagnosis was revised postnatally in 3 cases. Abnormal ICA was found in one twin only in two group A pairs, one group B pair, and both group C pairs. All group D twins had abnormal anatomy. Ventricular function was good in all twins scanned prenatally, and postnatally function correlated well with clinical condition. Thirteen sets of twins in groups A-C underwent surgical separation; 16/26 survived. There were no survivors from groups C or D.

Conclusions: Pre- and postnatal echocardiography accurately delineates cardiac fusion, ICA and ventricular function in the majority of twins with thoracic-level fusion. It is integral in assessing feasibility of separation. The outcome in twins with fused hearts remains dismal.
INTRODUCTION

The incidence of conjoined twins is reported to be in the range of 1 in 50,000 to 1 in 100,000,[1] but as 60% are stillborn or die shortly after birth, the true incidence is around 1:200,000 live births.[2] Females predominate in the ratio of 3:1. Twins are classified according to the major site of union, to which the ending “–pagus” is added, meaning “fixed” (Figure 1). Thoracopagus (joined at thoracic level) twins are the most common, accounting for 40% of cases, followed by omphalopagus (joined at the abdomen, but often including the lower thorax), accounting for 32%.[2] Other forms include pyopagus (sacral fusion), ischiopagus, and craniopagus. The term parapagus is used to describe twins in whom there is extensive side-to-side fusion (formerly known as thoraco-omphalo-ischiopagus, or dicephalus). Although there may be cardiac abnormalities in any type of fusion, thoracopagus twins have the highest incidence of abnormal cardiovascular findings with a 90% incidence of shared pericardium,[3] and major myocardial connections in some 75% of cases.[4] The extent of cardiac fusion and intracardiac anatomy (ICA) in conjoined twins not only determines the potential for surgical separation but also long-term survival. Surgical separation is rarely feasible in complex fused hearts although there may be occasional exceptions.[5][6]

We report the efficacy of both pre- and postnatal echocardiography in the evaluation of 23 sets of conjoined twins with thoracic-level fusion over a 20 year period at a single centre. To our knowledge, this represents the largest single-centre series of conjoined twins with cardiac involvement.

PATIENTS AND METHODS

Between 1985 and 2004, 23 sets of conjoined twins with thoracic-level fusion were referred to assess the feasibility of surgical separation. Two further sets without thoracic-level fusion (1 omphalopagus and 1 pyopagus) were excluded from this analysis. There were 18 sets of females and 4 sets of male twins. In the remaining set, sex was not determined at prenatal assessment. Six sets of twins were referred from overseas while the remaining 17 sets were from the United Kingdom and the Republic of Ireland. There were 13 sets of thoracopagus twins, 5 sets of thoraco-omphalopagus twins, and 5 sets of parapagus twins. All except 2 sets were diagnosed prenatally, between 14 and 38 weeks gestation, and delivery was by caesarean section. Fetal echocardiography was performed on 12 sets of twins later in the series, 10 at Great Ormond Street Hospital and 2 at other fetal medicine units. Diagnostic echocardiographic images were obtained as early as 14 weeks’ gestation. In general the left-sided twin was termed twin 1 and the right-sided twin termed twin 2. The remaining 11 sets underwent postnatal echocardiogram between one day and three years of age. A modification of standard echocardiographic views was performed as dictated by the accessibility of subcostal and precordial views. Cardiac catheterisation was undertaken in 3 sets of twins early in our experience. The twins were classified according to the degree of cardiac fusion: group A separate hearts and pericardium, group B separate hearts and common pericardium (Figure 2), group C fused atria and separate ventricles, and group D fused atria and ventricles (Figures 3 and 4).
RESULTS

Cardiac fusion: The type of conjoin was the major determinant of degree of cardiac fusion, with the majority of thoraco-omphalopagus twins in group A (Table 1), the majority of parapagus twins in group B (Table 2), and the majority of thoracopagus twins in groups C and D (Tables 3 + 4). In the 15 sets of twins where anatomy could be verified by surgery or postmortem, the degree of cardiac fusion was correctly diagnosed in all but one set (twins C2). In these twins fetal echocardiography at 22 weeks was suggestive of cardiac fusion, with a common atrial mass and two atroventricular valves overriding a complex ventricular structure. Two great arteries arose towards the right-sided twin, but no outflow tracts could be seen arising towards the left-sided twin. However postnatal echocardiography suggested contiguous but non-fused hearts, although the pericardium was shared. Twin 2 (right) had normal ICA, but twin 1 had a severely hypoplastic heart which was only about 20% of the expected size. The atroventricular valves were severely regurgitant, and there was a large VSD. Two small great arteries arose in parallel from the ventricular mass. The twins underwent surgery when twin 1 suffered an acute hemodynamic decompensation on day one of life. At separation, a single pericardial sac was identified, and the atria were found to be connected. When this connection was divided, twin 1 succumbed. Soon after pericardial closure twin 2 developed severe pulmonary hypertension, and she died 33 hours later.

In one further set of twins (C1), a left atrial communication was correctly diagnosed on postnatal echocardiogram, but at surgery an additional aberrant artery was found arising from the descending aorta of twin 1 which inserted into the aortic arch of twin 2 between the origin of the left carotid artery and the left subclavian. It became clear that twin 1 was partially perfusing twin 2, and when this vessel was clamped twin 2 died. Twin 1 survived surgery but died unexpectedly at 6 weeks of age. Despite extensive investigation, no cause was found.
Table 1. Separate hearts, separate pericardium (Group A).

<table>
<thead>
<tr>
<th>Twins</th>
<th>Type</th>
<th>Timing of diagnosis</th>
<th>Twin 1 ICA / Twin 2 ICA</th>
<th>Surgery</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>A1</td>
<td>female</td>
<td>Thoraco-omphalopagus</td>
<td>17 weeks gestation, postnatal echo</td>
<td>normal / normal</td>
<td>elective separation at 10 weeks</td>
</tr>
<tr>
<td>A2</td>
<td>female</td>
<td>Thoraco-omphalopagus</td>
<td>17 weeks gestation, postnatal echo</td>
<td>normal / normal</td>
<td>emergency separation at 1 day</td>
</tr>
<tr>
<td>A3</td>
<td>male</td>
<td>Thoraco-omphalopagus</td>
<td>17 weeks gestation, fetal echo</td>
<td>normal / normal</td>
<td>emergency separation at 1 day</td>
</tr>
<tr>
<td>A4</td>
<td>female</td>
<td>Parapagus</td>
<td>14 weeks gestation, fetal echo</td>
<td>normal / AVSD, DORV, ant Ao, PAT</td>
<td>emergency separation at 1 day</td>
</tr>
<tr>
<td>A5</td>
<td>female</td>
<td>Thoraco-omphalopagus</td>
<td>20 weeks gestation, fetal echo</td>
<td>normal / VSD (repaired at 8 weeks)</td>
<td>elective separation at 7 weeks</td>
</tr>
</tbody>
</table>

Table 2. Separate hearts, common pericardium (Group B).

<table>
<thead>
<tr>
<th>Twins</th>
<th>Type</th>
<th>Timing of diagnosis</th>
<th>Twin 1 ICA / Twin 2 ICA</th>
<th>Surgery</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>B1</td>
<td>Female</td>
<td>Thoracopagus</td>
<td>38 weeks gestation, postnatal echo and catheter</td>
<td>normal / PAT/VSD, on prostaglandin</td>
<td>emergency separation at 3 days</td>
</tr>
<tr>
<td>B2</td>
<td>Male</td>
<td>Parapagus</td>
<td>At delivery, postnatal echo</td>
<td>normal / normal</td>
<td>elective separation at 8 months [7]</td>
</tr>
<tr>
<td>B3</td>
<td>Female</td>
<td>Parapagus</td>
<td>18 weeks gestation, postnatal echo</td>
<td>normal / normal</td>
<td>elective separation at 3 years [8]</td>
</tr>
<tr>
<td>B4</td>
<td>Male</td>
<td>Parapagus</td>
<td>22 weeks gestation, postnatal echo</td>
<td>normal / normal</td>
<td>elective separation at 10 months</td>
</tr>
<tr>
<td>B5</td>
<td>Female</td>
<td>Thoracopagus</td>
<td>15 weeks gestation, postnatal echo</td>
<td>normal / normal</td>
<td>elective separation at 3 months [9]</td>
</tr>
<tr>
<td>B6</td>
<td>Female</td>
<td>Parapagus + normal triplet</td>
<td>20 weeks gestation, fetal echo</td>
<td>normal / normal</td>
<td>separation declined</td>
</tr>
<tr>
<td>B7</td>
<td>Female</td>
<td>Thoraco-omphalopagus</td>
<td>At delivery, postnatal echo</td>
<td>normal / normal</td>
<td>elective separation at</td>
</tr>
</tbody>
</table>
Table 3. Fused atria, separate ventricles (Group C).

<table>
<thead>
<tr>
<th>Twins</th>
<th>Type</th>
<th>Timing of diagnosis</th>
<th>Twin 1 ICA / Twin 2 ICA</th>
<th>Surgery</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>C1</td>
<td>thoracopagus</td>
<td>18 weeks gestation, postnatal echo</td>
<td>Normal / AVSD, hypoplastic LV</td>
<td>emergency separation at 5 days</td>
<td>twin 2 died in surgery, other died at home</td>
</tr>
<tr>
<td>C2</td>
<td>thoracopagus</td>
<td>22 weeks gestation, fetal echo</td>
<td>hypoplastic heart, large VSD, ant Ao / normal</td>
<td>emergency surgery at 1 day</td>
<td>twin 1 died in surgery, other shortly after</td>
</tr>
</tbody>
</table>

Table 4. Atrial and ventricular fusion (Group D, all thoracopagus).

<table>
<thead>
<tr>
<th>Twins</th>
<th>Timing of diagnosis</th>
<th>No. of AV valves / ventricles</th>
<th>Twin 1 /Twin 2 great arteries</th>
<th>Timing of delivery / death</th>
</tr>
</thead>
<tbody>
<tr>
<td>D1</td>
<td>? gestation, postnatal echo and catheter</td>
<td>2 / 4</td>
<td>DOV, PAT / DOV, PAT</td>
<td>term / 3 days</td>
</tr>
<tr>
<td>D2</td>
<td>28 weeks gestation, postnatal echo and catheter</td>
<td>2 / 2</td>
<td>DOV with ant Ao, PS / DOV, NRGA, hypoplastic Ao</td>
<td>37 weeks / 4 weeks</td>
</tr>
<tr>
<td>D3</td>
<td>18 weeks gestation, fetal echo</td>
<td>1 / 3</td>
<td>DOV with ant Ao / single outlet</td>
<td>TOP at 20 weeks</td>
</tr>
<tr>
<td>D4</td>
<td>17 weeks gestation, fetal echo</td>
<td>2 / 4</td>
<td>NRGA / VA discordance, IAA</td>
<td>term / first week of life</td>
</tr>
<tr>
<td>D5</td>
<td>28 weeks gestation, fetal echo</td>
<td>1 / 4</td>
<td>VA discordance / single outlet</td>
<td>34 weeks / 1 hour</td>
</tr>
<tr>
<td>D6</td>
<td>14 weeks gestation, fetal echo</td>
<td>1 / 2</td>
<td>DOV, ant small Ao / PAT, MAPCAS</td>
<td>36 weeks / 4 weeks</td>
</tr>
<tr>
<td>D7</td>
<td>15 weeks gestation, fetal echo</td>
<td>1 / 4</td>
<td>aortic atresia / single outlet</td>
<td>28 weeks / 30 minutes</td>
</tr>
<tr>
<td>D8</td>
<td>20 weeks gestation, fetal echo</td>
<td>2 / 3</td>
<td>NRGA / DOV with ant Ao, PS</td>
<td>TOP at 20 weeks</td>
</tr>
<tr>
<td>D9</td>
<td>15 weeks gestation, fetal echo</td>
<td>2 / 3</td>
<td>DOV, ant Ao / VA discordance</td>
<td>35 weeks / 30 hours</td>
</tr>
</tbody>
</table>

**Abbreviations for Tables 1-4:** ant Ao, anterior aorta; AV, atrioventricular; AVSD, atrioventricular septal defect; DO(R)V, double outlet (right) ventricle; IAA interrupted aortic arch; LV, left ventricle; MAPCAS, multiple aortopulmonary collateral arteries; NRGA, normally related great arteries; PAT, pulmonary atresia; PHT, pulmonary hypertension; PS, pulmonary stenosis; TOP, termination of pregnancy; VA, ventriculo-arterial; VSD, ventricular septal defect.
Intracardiac anatomy: As far as could be assessed by repeated echocardiography, catheterization, surgery or postmortem, the ICA was correctly diagnosed in all 18 sets seen postnatally. Of the 7 sets who underwent both pre- and postnatal echocardiography, the prenatal diagnosis had to be revised postnatally in 3: one ventricular septal defect (VSD) turned out to be an atrioventricular septal defect (AVSD) (twins A4); one perimembranous VSD was only seen on postnatal echo (twins A5); and one suspected single outlet heart had normal ventriculo-arterial connections (twins B6). Abnormal ICA was far more frequent in thoracopagus twins (23/26, 88%) than in either thoraco-omphalopagus twins (1/10) or parapagus twins (1/10). It affected one twin only in two sets from group A, one set from group B, and both sets from group C. All twins in group D had very abnormal anatomy, which was discordant in 8/9 pairs. This is described below:

Twins D1: shared a single atrial chamber into which 2 inferior vena cavae, 3 superior vena cavae and the pulmonary veins from each twin drained ipsilaterally. There were 2 separate atrioventricular valves connected to ventricles which communicated via a large muscular VSD. There was a rudimentary anterior ventricle on each side, connected to each main ventricle via a VSD, which gave rise to a patent aorta and an atretic pulmonary artery in each twin. Drawings of the postmortem findings are shown in Figure 3.

Twins D2: shared a large common atrium, into which hepatic veins drained from twin 2, and a single inferior vena cava drained from twin 1. The pulmonary veins of each twin drained ipsilaterally. There were 2 atrioventricular valves and 2 ventricles, each of which were double outlet. From the ventricle on the side of twin 1, the great arteries arose in parallel, with the aorta anterior, and a stenotic pulmonary artery posterior. From the ventricle on the side of twin 2, the great arteries were normally related, but the ascending aorta and arch were hypoplastic.

Twins D3: shared a common atrium which received systemic and pulmonary veins from each twin, and a common atrioventricular valve committed to 2 large ventricles. Two great arteries arose in parallel from the ipsilateral ventricle to one twin, and one great artery to the other twin arose from a rudimentary ventricle which communicated with the other large ventricle.

Twins D4: shared a large common atrium. Twin 1 had a common atrioventricular valve orifice overriding 2 ventricles, of which the left was smaller. The great arteries were normally related. Twin 2 had a single atrioventricular valve connected to a good-sized left ventricular cavity, a small restrictive VSD and hypoplastic right ventricle. There was ventriculo-arterial discordance with a small aorta arising from the hypoplastic right ventricle, and type B interruption of the aortic arch. The left ventricular cavities of each twin were fused, with at least 2 large defects allowing communication between them.

Twins D5: shared a complex fused heart with 2 atrial cavities. There was a common atrioventricular orifice overriding 4 ventricles which were all in communication. Two
great arteries arose in parallel in twin 1, but only a single arterial trunk arose in twin 2. Ventricular function was good, but there was severe atrioventricular valve regurgitation, accompanied by severe ascites most marked in twin 2.

Twins D6: shared a complex fused heart which lay mainly in the thorax of twin 1. There was total anomalous pulmonary venous drainage from both twins to an enlarged right atrium, and a common atrioventricular orifice overriding 2 ventricles. The left ventricle was hypoplastic. Two great arteries to twin 1 arose in parallel from the right ventricle, but the aorta was smaller than the pulmonary artery. The aorta to twin 2 arose from the hypoplastic left ventricle, but the pulmonary artery in this twin was atretic with multiple aortopulmonary collateral arteries.

Twins D7: shared a complex conjoined heart with 2 atria and 4 ventricles, of which 2 were rudimentary. There was a common atrioventricular valve, with moderate regurgitation. There were 2 great arteries arising towards twin 1, with the pulmonary artery much larger than the aorta at 15 weeks gestation, but only 1 great vessel arising towards the twin 2. By 20 weeks gestation the aortic valve in twin 1 had become atretic, and there was retrograde flow around the aortic arch from the duct.

Twins D8: shared a common atrium, into which the systemic and pulmonary veins drained ipsilaterally. Two atrioventricular valves were committed to one large central ventricle, which was connected to a rudimentary ventricle on each side via a VSD. Twin 1 had normally related great arteries, but in twin 2 the arteries arose in parallel from the ipsilateral rudimentary ventricle, with the aorta anterior. The pulmonary artery was hypoplastic. The echocardiographic images are shown in Figure 4.

Twins D9: shared a heart which lay mainly in the thorax of twin 1, and consisted of a common atrium and 2 atrioventricular valves opening into 2 main ventricles which were connected by an inlet VSD. There was an additional rudimentary ventricle on the left, also connected via a VSD. This gave rise to 2 great arteries to twin 1, which arose in parallel. The 2 main ventricles each gave rise to a great artery to twin 2, but there was ventriculo-arterial discordance.

**Ventricular function**: this was good in all sets scanned prenatally, and postnatally correlated well with clinical condition. There was concern over poor preoperative ventricular function with high postoperative inotrope requirements in some twins early in the series, most notably twins B3.[8] These parapagus twin girls were referred at 3 years of age for separation, and twin 2 had moderately reduced left ventricular function preoperatively. Following separation her ventricular function deteriorated further despite aggressive fluid/blood replacement and inotropic support, and she died on the third postoperative day. At postmortem, the myocardium appeared pale and poorly developed, although myocardial histology was normal. In view of this, subsequent sets were given prophylactic preoperative inotropes if there was any concern over cardiac function, with good effect.
Separation: Thirteen sets of twins underwent surgical separation, 5 from group A, 6 from group B, and 2 from group C. Surgery was not offered to twins in group D. Six were undertaken as emergency procedures within the first week of life, and 7 were performed electively between 3 months and 3 years of age. In one emergency separation, one twin had died on the journey to our unit and the conjoined area had to be compressed digitally until the surviving twin could be separated (Twins A3). There were 16 out of a possible 26 survivors, 7 from group A (70%), and 9 from group B (75%); 4 following emergency separation (33%), and 12 following elective separation (86%). There were no survivors from group C or D. The surgical figures include 3 deaths which occurred relatively late following separation; in twins B1, B4 and C1 (see above). After separation of the B1 twins, the duct-dependent circulation of twin 2 was maintained with prostaglandin. She subsequently developed obstructive jaundice which necessitated a choledochoenterostomy, and then a further laparotomy 5 weeks later. Two days after this she suffered a cardiopulmonary arrest and died. Postmortem examination confirmed the cardiac anatomy and revealed the arterial duct to have closed. The B4 twins had a very prolonged postoperative course because of difficulties with wound closure, and were discharged after 11 months. One twin subsequently died following an episode of aspiration.
DISCUSSION

Although the literature relating to conjoined twins is extensive, there are few large series reported, and comparison is difficult because of continued discrepancies in nomenclature, despite standardised systems being proposed.[10] Particular difficulty occurs in the classification of thoracopagus, thoraco-omphalopagus and omphalopagus twins. In this series we have described twins with ventral thoracic-level fusion as thoracopagus, and ventral abdominal fusion extending to the lower part of the thorax as thoraco-omphalopagus. We have excluded omphalopagus cases with ventral lower abdominal fusion only. Of the 23 cases, thoracopagus connections were the most common, and were associated with a greater degree of cardiac fusion and a higher risk of abnormal ICA than either thoraco-omphalopagus or parapagus connections. This finding agrees with that of both postmortem [11] and surgical series.[6][12][13] In Group D, the number of ventricles varied between 2 and 4, although 1 or 2 were often rudimentary. The anatomy of the atrioventricular junction was very variable, but even in hearts with more than 2 ventricles, there were never more than 2 atrioventricular valves. There was a common valve in 4/9 sets. There were either 3 or 4 great arteries in each pair, but one or more was hypoplastic or atretic in 8/9. The concordance rate for abnormal intracardiac anatomy was low, consistent with non-conjoined monochorionic twins.[14]

Ventricular fusion has been reported in parapagus twins[11][12][13][15] but this was not seen in our series, although 4/5 had a common pericardium. Defects of lateralization, including right and left atrial isomerism and mirror imagery are particularly common in parapagus twins,[15] although they are also seen in other forms.[16] Usually it is the right-sided twin that is affected. In one pair of parapagus twins from this series, the right-sided twin was isomeric (twins A4). Unfortunately there was no postmortem to confirm this. Although postmortems were always requested, the majority of parents declined (11 of 16 pairs in which one or both babies died).

Historically conjoined twins have been classified on the basis of cardiac fusion into groups with separate hearts with common pericardium, fused atria with normal ventricles, and fused atria and ventricles.[4] However this does not take into account twins with separate hearts and pericardium, which occurred in both thoraco-omphalopagus and parapagus twins in this series. Our classification includes this group of patients. While surgical survival rates were not significantly different in pairs with or without pericardial fusion, detailed knowledge of the anatomy allows meticulous planning of the operative strategy, which is a key factor in successful surgery.[2]

An accurate understanding of the degree of cardiac fusion and the ICA is essential to determine potential for surgical separation in twins with thoracic–level fusion. It has been our policy to offer surgery to twins in Groups A–C. All parents of twins with type D anatomy were counselled that separation would not be feasible. In addition, accurate assessment of ventricular function is also essential for successful surgery. The unexpected death in twin B3-2 secondary to ventricular dysfunction caused us to institute prophylactic inotropy in twins B5, with good effect.

The experience reported in this series was accumulated over 20 years, and the quality and availability of imaging modalities has changed considerably over this time. Echocardiography has provided accurate assessment of cardiac fusion, ICA and
ventricular function throughout, so that cardiac catheterization and angiography were performed in only 3 cases early in the experience. Angiography may occasionally still be required if the anatomy severely restricts the echocardiographic windows,[17][18] or if there is persisting uncertainty about the degree of cardiac fusion.[19]

As the resolution of ultrasound has improved, it has become possible to assess the heart prenatally at earlier gestations. Diagnostic images have been reported as early as 9 weeks’ gestation.[12] In our series, although all twins from the UK and Ireland were diagnosed antenatally, not all were referred for prenatal echocardiography, even as late as 1998. Referral rates were higher for thoracopagus twins, but as abnormal ICA may be seen in all 3 types of thoracic-level fusion, we would recommend that all types should be referred. Prenatal diagnosis is technically easier for the operator, as it avoids the anatomical constraints of scanning fused chests postnatally, and fetal fluid-filled lungs may allow better images. Recently it has become possible to assess conjoined fetuses using 3-dimensional echocardiography, and this may be helpful for the parents as the images are easier to understand.[20]

MRI has an increasing role in multi-system evaluation of conjoined twins prior to separation,[21][22] and has the advantage of being able to produce 3-dimensional reconstructed images in any orientation[23] as well as providing information about intracardiac and great vessel blood flow. Multislice CT scanning with its rapid acquisition times may also be useful, particularly in assessing details of arterial and venous anatomy. However, these sophisticated imaging techniques will continue to be used as adjuncts to echocardiography, which is likely to remain the bedrock of cardiac assessment for the foreseeable future.

Results from surgical separation have improved over time,[1] helped by advances in imaging allowing for increasingly meticulous preoperative planning, and in intensive care.[24] Echocardiography performed by an experienced operator is a key part of both pre- and postoperative management, and provides an accurate, safe, non-invasive, and easily portable method of assessing the cardiovascular system in these complex infants.

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CONFLICTS OF INTEREST: none

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**FIGURE LEGENDS**

**Figure 1.** Commoner forms of conjoined twins: a craniopagus, b thoracopagus, c omphalopagus, d parapagus, e ischiopagus, f pyopagus.

**Figure 2.** Contiguous but non-fused hearts with no dividing pericardial wall between them: a) twins B1, 1985, with subaortic VSD in the twin on the right; b) twins B5, 1994, with improved resolution compared to a). RV, right ventricle, LV, left ventricle.

**Figure 3.** Drawings from the postmortem of twins D1: a) anatomical drawing of the systemic and pulmonary venous connections. IVC, inferior vena cava, (L)SVC, (left) superior vena cava. b) schematic drawing of the arrangement of the ventricles and great arteries. V, ventricle, PA, pulmonary artery.

**Figure 4.** Fetal echocardiographic images at 20 weeks gestation (Twins D8, 2004): a) fused atria and ventricles; b) great arteries arising in parallel from a rudimentary ventricle. The pulmonary artery is hypoplastic.
REFERENCES

rudimentary ventricle
main ventricle
aorta
hypoplastic pulmonary arteries
atrium
rudimentary ventricle