Seminars in Pediatric Surgery 24 (2015) 217–220

Contents lists available at ScienceDirect

Seminars in Pediatric Surgery

journal homepage: www.elsevier.com/locate/sempedsurg

The management of conjoined twins: Cardiology assessment

Rachel E. Andrews, MA, MRCP, Robert W.M. Yates, MB, BCh, FRCP, Ian D Sullivan, FRACP*

Cardiothoracic Unit, Great Ormond Street Hospital, London WC1N 3JH, UK

ARTICLE INFO

Keywords:
Conjoined twins
Cardiac defects
Echocardiography
Prenatal diagnosis

ABSTRACT

Structural cardiac defects occur in at least 1 twin in about 75% of conjoined twins with thoracic level fusion. Outcomes after surgical separation of thoracic level conjoined twins have been favorable when the hearts have been separate. However, even in this situation, the outlook is poor for an individual twin with an important cardiac defect. Arterial anastomosis between twin circulations is an important additional consideration, with poor outcomes for perfusion recipient twins. Surgical separation is contraindicated when ventricular level cardiac fusion exists. Cardiac assessment is a key component of prenatal counseling.

Monochorionic twinning and cardiac defects

The risk of cardiac anomaly is increased in monochorionic twin pregnancy, with a risk gradient related to the timing of the division of the single fertilized embryo.[1] When there is a monochorionic, diamniotic pregnancy, the risk of primary structural cardiac abnormality in at least 1 twin is about 7%.[2] This is about 10 times the baseline population risk for an unselected singleton pregnancy. In the rare monochorionic, monoamniotic twin pregnancies, the risk of heart abnormality in at least 1 twin may be over 50%.[2] If a monochorionic twin is affected, the risk to the other for cardiac abnormality is reported to be about 25%. Concordance for cardiac abnormalities might be anticipated[3] but occurs in fewer than 20% of cases, emphasizing that epigenetic factors are important.[4,5] Interestingly, disturbance of laterality, meaning heart defects in the context of right or left atrial isomerism, are over represented.[6] Consequently, it is not surprising that cardiac defects are common in conjoined twins, affecting at least 1 twin in over 50% of conjoined twin pregnancies with thoracic level fusion,[7,8], and as many as 79% in a recent fetal series.[9]

High rates of termination of pregnancy and intrauterine death mean that the precise incidence of conjoint twinning is difficult to define. A global study described 383 sets of conjoined twins from more than 26 million births reported from 21 surveillance programs.[10] The total prevalence was 1.47 per 100,000 births. Of pregnancies delivered, 46% were live born, 27% stillborn, and 27% underwent termination of pregnancy. There was a trend for higher prevalence in Mexican and South American populations, whose pregnancy outcomes included 70–80% live births, as elective termination of pregnancy for fetal anomaly was classified as "not permitted" in these populations. The authors also estimated the prevalence of conjoined twins in spontaneous abortions as 3.03 per 1000.

The estimated prevalence would mean that about 11–12 pregnancies complicated by conjoint twinning would be expected annually in the United Kingdom, possibly increased in the present era because of the increased incidence of monozygous twinning after conception using assisted reproductive techniques. However, it has been estimated that when termination of pregnancy, intrauterine death, and early neonatal mortality are considered, the incidence of conjoined twins surviving long enough for surgical separation to be considered in the United States is about 1:320,000 births.[11]

Cardiac defects in conjoined twins

Fascination with the subject means that the literature relating to conjoined twins is extensive, but consists largely of case reports or very small series. An extensive literature review aimed to clarify cardiac involvement in as many as 1262 sets of all types of conjoined twins, among whom there were 192 pairs with some degree of cardiac fusion.[8] The analysis was inevitably hampered by the variation in description of morphology, and the historic nature of many of the numerous references.

We reviewed our institutional experience of cardiac assessment of conjoined twins with thoracic level fusion in the 20 year period 1985–2004.[7] This included thoracopagus, thoraco-omphalopagus and parapagus conjoined twins, in whom the nature of cardiac fusion, or the presence of separate hearts, is not apparent from external inspection. The diagnosis of conjoint twinning was made

* Corresponding author.
E-mail address: ian.sullivan@gosh.nhs.uk (I. Sullivan).

http://dx.doi.org/10.1053/j.sempedsurg.2015.07.003
1055-8586/

prenatally in all but 2 sets, both born overseas. Only 2 of the affected 23 pregnancies were terminated. This suggests that the decision to commit to the pregnancy, usually with the parental aim or expectation of postnatal surgical separation, had been made prior to referral to our hospital. A large Brazilian single center description of fetal diagnosis of conjoined twins over a similar time period was published recently.[9] In contrast to our experience, they described termination of pregnancy in 17 of 53 pregnancies, somewhat at variance with the registry data described above.[10]

Cardiac investigation

The widespread use of late first trimester screening for chromosome abnormality in first world populations, which utilizes fetal ultrasound findings together with maternal age and biochemistry, means that most surviving conjoined twins will now be diagnosed at this relatively early gestation in these populations. The exact details of cardiac anatomy may be difficult to define at this stage, but the presence or absence of cardiac fusion, or the presence of a major cardiac defect in 1 twin of a pair with separate hearts, should be evident. More detailed analysis of the intracardiac anatomy including the extent of cardiac fusion is often possible from early in the second trimester, but details of extracardiac circulatory anastomoses may be harder to define, and may not become apparent until after delivery. Postnatally, cardiac structure and function is still best assessed by echocardiography. However, there has been an emerging role for contrast-enhanced computed tomography giving data in 3-dimensional planes, which is especially useful for the postnatal assessment of venous and arterial anatomy. Diagnostic cardiac catheterization and angiography is invasive and probably obsolete and has not been used by our team since the 1980s. Cardiac magnetic resonance is disadvantaged by long acquisition times and the requirement for general anesthesia but has an important role in non-cardiac assessment.

Surgical separation

We classified twins with thoracic level fusion (thoracopagus, thoraco-omphalopagus, or parapagus) according to the degree of cardiac fusion.[7] Group A had separate hearts and pericardial sacs, Group B had separate hearts contained within a common pericardial sac, Group C had atrial level fusion but separate ventricles, and Group D had fused atria and ventricles. The group from Sao Paulo used the same classification.[9] In their large series of conjoined twins diagnosed prenatally, surgical separation was attempted in only 5 of 33 live born sets. Only 6 of 10 operated twins were reported to be alive after separation. In all of these survivors, the hearts had separate pericardial sacs, and were described as “normal.”

Our experience is biased by referral patterns, often influenced by the prospect of surgical separation. This was performed in all 5 group A twin pairs, with 7 of 10 long-term survivors. A fatality occurred in a twin just prior to arrival at our hospital, and the other twin was kept alive by manual compression of the conjoined area followed by emergency surgery on arrival. The co-twin survived. Both of a parapagus twin pair, one of whom had a complex heart defect with features typical of right atrial isomerism, died after emergency surgery on the first day of life, which was performed because of severe pulmonary hypertension in the twin with a structurally normal heart. In Group B (separate hearts and common pericardium), surgical separation was performed in 6 of 7 twin pairs. Separation was not attempted in a parapagus pair. There have been 9 of 12 long-term survivors. There were 2 in-hospital post-operative deaths. One was a twin born in 1985, who had pulmonary atresia with a ventricular septal defect. Following separation the arterial duct dependent pulmonary blood flow was maintained with prostaglandin. Subsequently the baby developed obstructive jaundice necessitating a choledochoenterostomy, followed by a laparotomy 5 weeks later. She suffered a cardiopulmonary arrest and died 2 days after this, possibly due to closure of the arterial duct despite the continued prostaglandin infusion. Another separated twin had impaired biventricular function post-operatively, not apparent pre-operatively, which may have been the main factor in her demise. This was early in our experience, and has informed a low threshold for inotropic support in the perioperative period since. The remaining death in this group occurred in an infant at home, several months after separation, probably secondary to aspiration.

In contrast to this favorable experience, attempted separation of conjoined twins was unsuccessful in both pairs with atrial level fusion (group C). Both of these were emergency operations at 5 and 1 days of age respectively, required because of hemodynamic instability. Apart from the atrial level fusion, each of these twin pairs had 1 abnormal and 1 essentially “normal” heart. In the first of these pairs, an arterial collateral vessel arising from the aortic arch in 1 twin was contributing to the perfusion of the other. The recipient twin, who had an unbalanced atrioventricular septal defect with a small left ventricle, died intraoperatively when this vessel was clamped. The “donor” twin survived surgery, but died unexpectedly at home 6 weeks later. In the other Group C pair, 1 twin had an extremely small heart, occupying only about 20% of the expected volume, in addition to a ventricular septal defect, transposed great arteries and severely regurgitant atrioventricular valves. This twin died immediately after division of the atrial communication. Her co-twin died on the second post-operative day in association with severe pulmonary hypertension.

Surgical separation has not been attempted in any group D twin pairs (atrial and ventricular fusion). None have survived beyond the neonatal period.

It is not difficult to understand why the outlook is so bleak when there is ventricular cardiac fusion. Ventricular level fusion is associated with complex structural abnormalities at veno-atrial, atrioventricular and ventriculo-arterial levels. The defects of laterality evident in monochorionic, monoamniotic separate twins indicate that such disturbances of right–left cardiac development are almost certain to be more profound in conjoined twins. Additionally, the preponderance of so called “right-sided” heart abnormalities [9] can most simply be explained by severe disturbance of looping of the embryonic primitive heart tubes. Ventricular level fusion consistently involves fusion of left ventricular myocardium.[11] “Anchoring” of left ventricular myocardium with separate, but abnormal, looping of that part of each primitive heart tube, which would have formed the right ventricle and arterial outlets in normal cardiac development, results in variable right ventricular hypoplasia, and abnormalities of both the atrioventricular and ventriculo-arterial connections. This can provide at least a superficial explanation for the morphology seen in most complex fused hearts (Figures 1 and 2).

Other than cardiac fusion, the 2 main cardiac concerns are structural defects of one or both separate hearts, and contribution to the perfusion of 1 twin by cardiac output from the other. Combined data from Great Ormond Street Hospital and Sao Paolo [7,9] report 24 pairs of conjoined twins with separate hearts. Of these, important structural cardiac defects were described in 10 of 48 (21%) hearts. There were no survivors among the twins with cardiac abnormalities, whether or not separation was attempted, with the exception of an infant from our series who underwent successful closure of a large ventricular septal defect a few days after separation, and is now a healthy 11-year-old girl.