Mechanisms of tricuspid valve regurgitation in hypoplastic left heart syndrome: a case-matched echocardiographic–surgical comparison study

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Aims

The multifactorial mechanisms of tricuspid valve (TV) insufficiency in patients with hypoplastic left heart syndrome (HLHS) include structural anomalies of TV leaflets and ventricular dilatation. We hypothesized that 2-D echocardiography underestimates the importance of TV structural abnormalities, whereas surgical assessment underestimates the importance of motion abnormalities, and compared echocardiographic assessment with surgical description.

Methods and results

Two independent experts retrospectively reviewed echocardiograms of all patients who had staged single-ventricular palliation and TV repair during January 1998–December 2008, and compared with case-matched controls who did not require TV repair. Primary and secondary mechanisms of TV insufficiency were categorized, and surgical findings ascertained from operation records. There were 32 patients with a median age of 5.9 months (0.3–140) and 32 matched controls. On echocardiographic review, an abnormality of at least one leaflet was noted in every patient (100%) vs. in only 14 controls 14 (44%) (P, 0.001). Leaflet prolapse was described in 22 (69%), and the restriction of a leaflet in 20 (69%). Agreement between the experts was excellent (κ = 0.64–0.88). On surgical inspection, annular dilatation was found in 17 (53%), and leaflet dysplasia in 14 (44%). Agreement between echocardiographic and surgical assessment was poor (κ < 0.6).

Conclusion

Important structural abnormalities are common in patients with HLHS and TV insufficiency, some readily identified by 2-D echocardiography. However, there are significant discrepancies between echocardiographic and surgical findings. Echocardiographic assessment is sensitive to detect leaflet motion abnormalities, but not leaflet structural abnormalities. Both echocardiographers and surgeons should be aware of these limitations when planning surgical interventions.

Keywords

Single-ventricle physiology • Tricuspid valve • Valve repair

Introduction

Tricuspid valve (TV) insufficiency is associated with a poor long-term outcome in patients with hypoplastic left heart syndrome (HLHS) undergoing staged palliation.1 The presence of severe tricuspid regurgitation (TR) has traditionally been a contraindication for choosing the single-ventricle palliation pathway, or is considered an indication for surgical valve repair either during one of the palliative stages or as a separate surgical intervention.1–6 Our recent published experience shows that TR of at least mild grade is common, being present in approximately one-third of infants at their evaluation prior to stage II intervention.7 Before surgical intervention on the valve, a detailed description of the TV apparatus is required, as the mechanisms of TR in this condition are usually complex and multifactorial.8,9 Factors which contribute to TR include annular dilatation, right ventricular (RV) dysfunction, valve prolapse, and structural anomalies of the TV leaflets (bicuspid or quadricuspid valves, accessory orifices, leaflet malformation, leaflet prolapse, leaflet restriction or ‘tethering’, and wide variability in the anatomy of the supporting apparatus).10 Chronic volume...
overload leads to progressive ventricular dilatation, which contributes to TR, and further exacerbates the volume overload.

A recent 3-D echocardiographic (3DE) study demonstrated tricuspid annular anatomy in HLHS to be abnormal compared with normal controls with the loss of the normal 3-D shape of the annulus. No good data are available on how reliably pre-operative transthoracic echocardiography can describe the mechanisms of TR and how it relates to surgical findings. As the valve leaflets can be difficult to visualize, the hypothesis behind the current study was that 2-D echocardiography underestimates the importance of structural TV anomalies as the leaflet morphology can be difficult to assess using both 2-D transthoracic or transoesophageal echocardiography (TEE) and 3DE. Therefore, the echocardiographic description was compared with the surgical description of the mechanism of regurgitation.

Methods
We retrospectively reviewed all patients with HLHS who underwent staged single-ventricle palliation in our institution between January 1998 and December 2008, and underwent TV repair either at the time of one of their staged operations or as a sole procedure between stage II and stage III palliation. All patients with HLHS who underwent TV repair were included in the study. Each patient (case) undergoing TV repair surgery was matched to a control patient in the cardiac surgery database, using diagnosis, ventricular function, type of palliation, body weight, and body surface area. The control patients were identified from the echocardiographic database as having no/mild TR at the same stage in the palliative strategy as the matched case. For a detailed description of the case matching, we refer to Honjo et al.7 The purpose of the case matching in the current study was to compare the TV characteristics and the RV size and the function between a group of patients with moderate-to-severe regurgitation and a group with less-than-moderate regurgitation. As no reference data were present for this patient group, we created our own control group by case-matching in our surgical database.

The institutional Research Ethics Board approved the study and waived the requirement for patient consent.

Echocardiographic assessment
In our institution, all patients undergo a detailed transthoracic examination (TTE) prior to any surgical intervention. This pre-operative TTE is performed according to a standardized imaging protocol. Children between 3 months and 3 years are sedated using a standardized sedation protocol. For the cases, the transthoracic 2-D echocardiograms obtained in the patients prior to surgical TV repair were retrospective-ly reviewed by two observers. They were blinded to each other’s findings as well as to the surgical findings. The mechanisms of valvar regurgitation were described according to a modified Carpentier classification11–13 (see Supplementary data online, Appendix; Figure 1). This included the prolapse of one of the leaflets (defined as the free edge of any leaflet overriding the plane of the annulus during systole) due to either chordal elongation or rupture of a papillary muscle, restriction of leaflet motion due to chordal shortening or tethering of a leaflet, annular dilatation, valve leaflet dysplasia (defined as an abnormality of the leaflet, such as thickening, or nodular irregularity of the leaflet edge or presence of an extra zone of apposition/cleft in one of the leaflets), abnormality of the papillary muscle, ventricular dilatation, and the presence of endocardial fibroelastosis (EFE) on the ventricular surface or on the papillary muscles (as suggested by hyperechogenicity on the ventricular surface or papillary muscles). Dysplastic leaflets and abnormal papillary muscles could be present with or without leaflet motion abnormalities (prolapse or restriction). The echocardiograms of the case-matched controls were reviewed to describe TV anomalies in cases with no, trivial, or only mild TR. TR grade was numerically expressed as 0 (none or trivial), 1 (mild), 2 (moderate), or 3 (severe).

Ventricular function was assessed by subjective assessment (categorized as normal, mildly, moderately, or severely depressed), as well as more quantitatively by calculating RV fractional area change [(end-diastolic area – end-systolic area)/end-diastolic area]) from the apical four-chamber views. The TV annulus was measured, and the z-score based on the patient’s body surface area was calculated using our own echocardiography laboratory’s database. RV dimensions were measured from the apical four-chamber view at the level just below the tricuspid annulus and at the mid-cavity level. The sphericity index was calculated by measuring the RV width mid-cavity, and dividing it by the RV length (mid-annulus to RV apex dimension) from the apical four-chamber view.14

As different abnormalities can be present simultaneously, the two echocardiographers defined what they thought to be the primary or secondary mechanism contributing to TV regurgitation. Interobserver agreement was evaluated between the two experienced observers.

Surgical assessment
At the time of surgical repair, the TV was inspected by the surgeon. After opening the right atrium, the surgeon inspects the valve morphology and the competency of the valve is tested using a saline test. When the surgeons perform a TV repair, they write a detailed report on the operative findings including a detailed description of the mechanisms of regurgitation and techniques used to try to reduce the amount of regurgitation. For the purpose of this study, the surgical description of the valve anatomy was retrospectively reviewed based on the surgical notes. To describe the surgical anatomy, the same classification as the one used for echocardiographic description of the valve leaflet was used. The surgical description was available for the patients undergoing valve repair and not for the case-matched controls.

Surgical indication and techniques
In our centre, we routinely inspect and repair TVs in patients with equal or greater than mild-to-moderate TR, preferably at the time of the second palliative stage, the bidirectional cavo pulmonary shunt.7 The patients who developed significant TR after stage II palliation underwent TV repair as a sole procedure as the combination of both is considered a high-risk procedure. We performed TV repair and Fontan simultaneously only when TV repair could be accomplished in a reasonable amount of time and with a good result so that the TV repair did not add any additional risk to the subsequent postoperative Fontan physiology. It is extremely rare for us to intervene on the TV at stage I palliation, and a neonate with HLHS and severe TV regurgitation may be directed towards primary heart transplantation rather than staged palliation.

We have previously described the surgical techniques we used.7 In brief, local annuloplasty and commissuroplasty were the two major techniques used in patients with annular dilatation and/or leaflet prolapse. Complete obliteration of the posterior leaflet was occasionally performed. ‘Clefts’ or pseudo-commisures were primarily closed with interrupted polypropylene sutures. Dysplastic leaflet edges were approximated with interrupted fine polypropylene sutures to...
improve the linearity of the coaptation area. Less commonly used tech-
niques were chordal shortening for chordal elongation, or an Alfieri
stitch. Intra-operative saline testing of valve competence is routinely
carried out in our institution, with additional repair attempted if
good results are not demonstrated.

Statistical analysis
Continuous variables are stated as means with standard deviations if
normally distributed, and medians with interquartile ranges if not. A
P-value of <0.05 was considered significant throughout. Analysis of
categorical variables was done using Fisher’s exact test. Continuous
variables were analysed with Student’s t-test in the case of normally
distributed variables, and Mann–Whitney U test for non-normally dis-
tributed variables. Variables identified as being significant in the univari-
ate analysis were further evaluated by stepwise conditional logistic
regression to obtain a model to predict which features were more
likely to be seen in patients who had required surgery. The level of
agreement between observers was evaluated using the κ coefficient.
A κ-value of >0.6 was considered to be indicative of good interobser-
ver agreement.

Results
Thirty-two patients with HLHS underwent TV repair (‘cases’) (Table 1). The median age at repair was 5.9 months (range 0.3–
140.0), and the median age of controls at the time of their
matched operation was 5.9 months (range 0.2–76.7) (not signifi-
cantly different). The median weight at TV repair was 5.8 kg
(3.0–38.6), and 5.4 kg (range 2.5–12) in controls (not significantly
different).

Echocardiographic assessment
Every patient undergoing valve repair was noted to have an abnor-
mality of at least one leaflet compared with 14 (44%) of controls
(P < 0.001). The anterior leaflet was assessed as abnormal in all
but one case (97%), the septal leaflet was abnormal in 29 (91%),
and the posterior leaflet in 19 (66%). The abnormality most com-
monly noted to be causing TV insufficiency was the prolapse of at
least one leaflet in 22 (69%) cases, followed by restriction of a
leaflet in 20 (63%) cases (Table 2). The prolapsing leaflet was anter-
ior in 20 (59%), posterior in 4 (12%) and septal in 10 (29%)
patients undergoing TV repair. Although 19 (59%) patients were
noted to have dysplastic leaflets on echocardiographic review, no
‘clefts’ (additional zones of apposition) were identified in any of
these valve leaflets.

The categorization of primary and secondary mechanisms of re-
gurgitation is shown in Table 3. Interobserver agreement was ex-
cellent for the assessment of TV abnormalities and mechanisms
of valvar regurgitation, with κ-values ranging from 0.64 to 0.88.

By univariate analysis, leaflet prolapse, leaflet restriction, annular
dilatation, leaflet dysplasia, or ventricular dilatation was present in a
significant proportion of patients who had required surgery. Multi-
variate analysis identified annular dilatation (P = 0.05, OR 4.1, CI
1.0–16.7) and leaflet restriction (P = 0.032, OR 5.8, CI 1.2–to 28.9)

Table 1 Patient characteristics

<table>
<thead>
<tr>
<th>Patient characteristics</th>
<th>Cases</th>
<th>Controls</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, median (range)</td>
<td>5.9 months (0.3–140.0)</td>
<td>5.9 months (0.2–76.7)</td>
</tr>
<tr>
<td>Weight, median (range)</td>
<td>5.8 kg (3.0–38.6)</td>
<td>5.4 kg (range 2.5–12)</td>
</tr>
<tr>
<td>TV z-score, mean ± SD</td>
<td>3.2 ± 1.4</td>
<td>1.6 ± 1.9</td>
</tr>
<tr>
<td>RV FAC, mean ± SD</td>
<td>34 ± 14%</td>
<td>35 ± 8%</td>
</tr>
<tr>
<td>Sphericity index, mean ± SD</td>
<td>0.34 ± 0.14%</td>
<td>0.35 ± 0.08%</td>
</tr>
</tbody>
</table>
to be the two factors that were most likely to be identified in patients who required TV repair. RV function as assessed either subjectively or by RV fractional area change was not significantly different between groups; 0.34 ± 0.14% in cases, compared with 0.35 ± 0.08% in controls, P = 0.715. The RV size, measured by the RV diastolic area indexed to the body surface area, was not significantly different between groups; 31.9 ± 8.8 cm²/m² in cases, compared with 27.8 ± 8.1 cm²/m² in controls, P = 0.06. RV geometry as assessed by the sphericity index was not significantly different between cases and controls; 0.99 ± 0.14 compared with 1.01 ± 0.13, P = 0.574. The TV annulus z-score was significantly larger in cases than in controls (1.60 ± 1.86 vs. 0.8 ± 0.23, P = 0.0249), suggesting more annular dilatation in the cases requiring valve surgery.

### Echocardiographic–surgical correlation

There was considerable variation between the echocardiographic assessment of the TV and the surgical assessment based on direct visualization of the valve. The presence of annular dilatation was the only feature on which there was good agreement between echocardiographers’ and surgeons’ assessments (κ = 0.72). Eighteen (53%) patients were judged to have no abnormality of any of the three leaflets, compared with all patients on echocardiographic review. In all patients judged at surgery to have no leaflet abnormality, annular dilatation was present, and chosen as the primary mechanism of regurgitation. Additional zones of apposition or ‘clefts’ were seen in 13 (38%) patients at surgical assessment, although none was detected on echocardiographic review. In only 8 (24%) patients, the prolapse of a leaflet was seen intra-operatively, compared with 23 (68%) cases detected on echocardiographic review. Chordal elongation, which was thought by the echocardiographers to be the most common cause of leaflet prolapse, present in 19 (59%) patients, was thought to be present in only 3 (9%) of the patients during inspection by the surgeons. Restriction of leaflet motion was judged by the surgeons to be present in only 1 (3%) patient, compared with 20 (63%) patients on echocardiographic review. The surgeons did not comment on ventricular dilatation, nor on EFE. Table 4 shows the TV abnormalities described by the surgeons, with the echocardiographic assessment for comparison. When the assessment of abnormalities of valve leaflet or papillary muscles was compared between echocardiographic and surgical assessment, there was poor agreement, with all k-values <0.6. The surgeons labelled the primary mechanism of TR to be annular dilatation in the majority (17, 53%) of cases, followed by dysplasia of TV leaflets in 14 (44%) of cases. There was poor agreement between surgical and echocardiographic assessments of primary mechanism, with a k-value of <0.6.

We divided the patients into two groups according to the type of surgery they underwent: an annuloplasty only, or an

### Table 2 Echocardiographic assessment of TV abnormalities in cases and controls

<table>
<thead>
<tr>
<th>Variable</th>
<th>Echocardiographic review</th>
<th>Surgical assessment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>TV repair cases, n (%)</td>
<td>Controls, n (%)</td>
</tr>
<tr>
<td>Leaflet prolapse</td>
<td>22 (69)</td>
<td>14 (44)</td>
</tr>
<tr>
<td>Leaflet restriction</td>
<td>20 (63)</td>
<td>10 (31)</td>
</tr>
<tr>
<td>Annular dilatation</td>
<td>29 (91)</td>
<td>13 (41)</td>
</tr>
<tr>
<td>Leaflet dysplasia</td>
<td>19 (59)</td>
<td>5 (16)</td>
</tr>
<tr>
<td>Abnormal papillary muscle</td>
<td>3 (9)</td>
<td>1 (3)</td>
</tr>
<tr>
<td>Ventricular dilatation</td>
<td>32 (100)</td>
<td>21 (66)</td>
</tr>
<tr>
<td>EFE</td>
<td>6 (19)</td>
<td>12 (38)</td>
</tr>
</tbody>
</table>

NS, not significant; N/A, not applicable.

### Table 3 Primary and secondary mechanisms of regurgitation determined by surgical assessment and echocardiographic review

<table>
<thead>
<tr>
<th>Variable</th>
<th>Echocardiographic review</th>
<th>Surgical assessment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Primary mechanism of regurgitation, n (%)</td>
<td>Secondary mechanism of regurgitation, n (%)</td>
</tr>
<tr>
<td>Prolapse of leaflet</td>
<td>19 (59)</td>
<td>5 (16)</td>
</tr>
<tr>
<td>Restriction of leaflet</td>
<td>2 (6)</td>
<td>12 (38)</td>
</tr>
<tr>
<td>Annular dilatation</td>
<td>8 (25)</td>
<td>5 (16)</td>
</tr>
<tr>
<td>Leaflet dysplasia</td>
<td>3 (9)</td>
<td>5 (16)</td>
</tr>
</tbody>
</table>
annuloplasty plus additional surgery to the valve leaflets or chords. When the echocardiographic findings between these two groups were compared, the only statistically different finding was the presence of restriction of a leaflet; 2 (6%) patients had restricted leaflet motion in the annuloplasty-only group compared with 19 (59%) in the annuloplasty plus additional surgery group, \( P = 0.01 \).

**Surgical outcome**

Our surgical outcomes have been previously reported\(^7,15\) for a cohort that includes this group of patients. Severity of TR was significantly reduced from the mean grade of 2.2 \( \pm \) 0.5 pre-operatively to 0.8 \( \pm \) 0.6 post-operatively (\( P = 0.03 \)), and although many patients re-developed significant TR within 1 year of surgery, few patients required re-repair of the TV if ventricular function was preserved.

**Discussion**

This study looks at the differences between echocardiographic and surgical assessment of TR in patients with HLHS requiring surgical intervention on the TV. Based on the echocardiographic assessment, annular dilatation and valve prolapse were the most common primary mechanisms, while during surgery, annular dilatation and valve dysplasia were identified as being the most common mechanisms. Despite good inter-observer agreement between echocardiographers, the agreement between the echocardiographic and surgical assessment is poor. This suggests that echocardiographic assessment and surgical visual assessment are focused on different aspects of TV abnormalities.

**Valve leaflet motion abnormalities**

Some of the findings are not really surprising. The prolapse and restriction of the leaflet motion were often not detected by the surgeon, as leaflet movement abnormalities are difficult to detect intra-operatively, and echocardiography is a much more sensitive method of detecting this.\(^1\)\(^6\) Adult echocardiographic studies examining the mitral valve also demonstrate echocardiography to be a sensitive tool for diagnosing abnormalities of leaflet motion, but to be somewhat limited in the diagnosis of anatomical abnormalities.\(^1\)\(^7\) As valve motion abnormalities were also commonly detected in patients who did not require TV valve surgery, the contribution of some degree of valve prolapse as well as restricted motion to TR is debatable, and based on our data, echocardiography seems to over-diagnose the importance of prolapse and restricted valve motion. This is probably due to the associated annular and ventricular dilatation present in almost every patient requiring surgical intervention. The septal tricuspid leaflet chordal attachments and papillary muscle are laterally displaced in the presence of RV dilatation, causing the echocardiographic appearance of restricted motion,\(^1\)\(^8\),\(^1\)\(^9\) and the lack of coaptation between the septal and anterior and posterior leaflets can give the appearance of leaflet prolapse in the plane of the TV annulus. Takahashi et al. demonstrated this well using 3DE,\(^7\) showing that leaflet tethering and prolapse are associated with significant TR in patients with HLHS. They showed that in these patients, the papillary muscle is displaced laterally, and the shape of the tricuspid annulus becomes more planar.

**Valve structural abnormalities**

Echocardiography was extremely poor at detecting structural leaflet abnormalities which were seen by the surgeons relatively frequently. Dysplasia of the TV, with irregularities of leaflet edges, and clefts in the leaflets are common abnormalities in the TV in HLHS patients,\(^1\)\(^9\) and in our patients, it was striking that a large proportion of the control group had structural abnormalities of at least one leaflet. However, these were under-diagnosed by the echocardiographic review; in fact, the echocardiographers could not detect clefts, which were seen commonly by the surgeons. In contrast to the mitral valve, it is more difficult to visualize the TV leaflets in a short axis or en face view. For the detection of additional zones of apposition in the mitral valve, this is the most commonly used echocardiographic view. In the retrospective review of the clinical echocardiograms, the short-axis views of the TV were usually not available. Therefore, the echocardiographic assessment was based on 2-D cuts through the leaflets from the four-chamber or long-axis planes, which limits the visualization of leaflet anatomy. It is possible that 3DE assessment of TV anatomy can provide better insight into the prevalence of structural TV abnormalities as this technique allows the operator to obtain en face views of the TV leaflets.\(^2\)\(^0\) The disadvantage of 3DE is its limited spatial resolution. Further study is required to look at the sensitivity and specificity of current 3DE techniques to accurately describe TV leaflet anatomy. 3DE is helpful in ascertaining the presence of prolapse and tethering of leaflets, as well as in elucidating the annular shape,\(^9\) but its use for describing more subtle structural leaflet abnormalities in the commonly dysplastic TV in these patients is less well-described.

Although surgery seems to underestimate the importance of dynamic motion of the TV leaflets, echocardiography is not very sensitive for detecting structural abnormalities of the TV leaflets. In a significant number of patients with only trivial or mild TR, abnormalities in TV motion (prolapse and restriction) are noted, without resulting in significant valve regurgitation. Although direct inspection was not available in this patient group, this seems to suggest that the combination of annular dilatation with a structural anomaly of the TV is particularly likely to cause

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**Table 4  Surgical assessment of TV abnormalities in cases undergoing TV repair, compared with echocardiographic assessment**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Echo review, ( n (%) )</th>
<th>Surgical review, ( n (%) )</th>
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<td>Annular dilatation</td>
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<td>27 (84)</td>
</tr>
<tr>
<td>Leaflet dysplasia</td>
<td>13 (41)</td>
<td>26 (81)</td>
</tr>
<tr>
<td>Abnormal papillary muscle</td>
<td>3 (9)</td>
<td>2 (6)</td>
</tr>
<tr>
<td>Ventricular dilatation</td>
<td>32 (100)</td>
<td>0</td>
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<tr>
<td>EFE</td>
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</tbody>
</table>
valve repair. This explains why a substantial proportion of patients require the use of additional surgical techniques in addition to an annuloplasty. In our centre, we do not routinely perform pre-bypass TEE of the TV. Based on these data, we revisited this practice as TEE can provide better imaging of the leaflets and the TV. Possibly, the addition of epicardial 3DE could be considered but this requires further clinical evaluation and the added diagnostic value is still uncertain.

**Endocardial fibroelastosis**

The surgeons did not rate EFE as an important cause of TR, or note its presence as being relevant to TV function. Echocardiographic review often suggests the presence of EFE in the left ventricle having an impact on septal geometry and therefore on TV function, with the potential addition of EFE on the TV apparatus. During intra-operative inspection, the TV is viewed from the right atrium, and therefore these effects of EFE are difficult to appreciate. This is an intrinsic limitation to surgical inspection.

**Tailored valve repair**

Careful inspection of the valve, by both echocardiography and direct visualization by the surgeon, allows the technique employed by the surgeon to be tailored towards the specific anatomical abnormalities and deficiencies. In our centre, partial annuloplasty and functional or anatomical commissuroplasty are the most common techniques used to repair the TV with annular dilatation and/or local prolapse. Various types of edge-to-edge repair can be employed for significant prolapse with elongated or deficient chordal support. Since the data suggest that surgical assessment often underestimates the presence or degree of preoperative leaflet prolapse or restriction, it is very important to combine echocardiographic and surgical findings to decide the subsequent repair techniques. Repairs should be tailored to the individual patient, with careful attention paid to structural abnormalities detected by echocardiography, but also having a high level of suspicion for abnormalities that echocardiography detects poorly.

**Assessment of ventricular function and shape**

In our study, we did not find a significant difference in RV function between patients with and without TR. The problem with most echocardiographic techniques used for functional assessment is that most qualitative as well as quantitative measurements are influenced by the presence of a significant degree of TR. In the presence of severe TR, a normal ejection fraction or fractional area change can indicate underlying ventricular dysfunction, which becomes apparent only when the regurgitation is reduced. As reported by our group, the outcome of patients who underwent TV repair is less good compared with those not requiring valve repair. This seems to suggest that the underlying ventricular dysfunction contributes to the severity of TR and also possibly that the additional volume load caused by TR is not well tolerated by the systemic RV. This finding is consistent with the observation of the systemic RV in a biventricular heart. Since TV structural abnormalities are common even in those patients not requiring valve repair, it is important to assess not only the valvar anatomy, but also its degree of dysfunction. Timely TV repair will remove the additional volume load caused by the valvar regurgitation, but, if performed late, it may not improve the ventricular function substantially, as ventricular dysfunction which has been secondary to chronic volume-loading may not return to normal following TV repair. Obviously, this study does not prove the benefit of performing TV repair in this patient group as it is our institutional approach to systematically repair the valve in patients with at least moderate regurgitation, and so we do not have data on the outcome of unrepaired valves. This would require a multicentre observational study, or ideally a randomized study. This would be extremely difficult to initiate as a lot of centres use a similar treatment approach and would be reluctant not to try to repair the valve at the time of surgery.

Surprisingly, the RV size was not significantly different between patients and case controls. This suggests that important TR does not cause significantly more RV dilatation than the volume loading already inherent in the single-ventricle physiology. This could be due to the hypertrophied, and possibly relatively stiff, RV cavity acting to protect the RV from progressive RV dilatation. RV diastolic function is extremely difficult to assess in the functionally univentricular heart, especially in the presence of significant TR, so it is difficult to prove this statement. There may be inaccuracies of measurement due to the fact that we used RV areas and not RV volumes, since there is currently no reliable method in routine use for measuring RV volume in HLHS patients. Also, there was no difference in sphericity index between the cases and controls. This is not surprising, as equivalent findings have been found in the left ventricle with functional mitral regurgitation; the left ventricular size and the sphericity index have been shown to have either no or only minimal association with the degree of mitral regurgitation.

**Limitations**

In this study, we did not specifically assess the change in echocardiographic appearances post-operatively, and therefore are not able to comment on alterations in abnormalities noted according to the specific surgical techniques employed. There was a significant improvement in the TR grade post-operatively, which we have previously more extensively reported along with the clinical outcome of a group of patients which includes this cohort. The improvement in the TR grade suggests that the combined echocardiographic and surgical assessment of the TV enabled the appropriate surgical techniques to be employed.

The echocardiograms were reviewed retrospectively, and the analysis was necessarily based on the available images. The original echocardiogram was not performed with the analysis classification in mind, and therefore the imaging in some cases did not show extensive detail of the TV. Nonetheless, in all cases, the echocardiogram was sufficient to analyse the TV according to our classification. RV function remains difficult to assess by echocardiography in the context of different loading conditions. Methods used may include measurement of tricuspid annular displacement, systolic tissue Doppler velocities, and deformation imaging, but these methods are not robust, particularly in the abnormal right ventricle. The RV fractional area change measurements are difficult to perform when retrospectively reviewing a study, as images may
not have been taken specifically for this purpose, and therefore may be suboptimal, for example, the apex may be omitted.

A comparison with the control group of patients was not possible for the surgical assessment of TV abnormalities, as this group did not systematically undergo detailed inspection of the TV at the time of surgery.

Conclusions

Important structural abnormalities of the TV are common in patients with HLHS and TV insufficiency, and some of these are readily identified by two-dimensional echocardiography. There are significant discrepancies between echocardiographic and surgical findings regarding the type of structural abnormalities and mechanism of TR; echocardiographic assessment is more sensitive to detect leaflet motion abnormalities, but insensitive to detect leaflet structural abnormalities. The best repair strategy will come from a comprehensive understanding of the mechanism of the regurgitation based on functional and anatomical echocardiographic assessment combined with direct surgical inspection. A prospective study is needed to better describe the accuracy of echocardiographic diagnosis of TV abnormalities in patients with HLHS and the possible effect on long-term outcomes and surgical strategies.

Supplementary material

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

References