The morphologically tricuspid valve in hypoplastic left heart syndrome

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Abstract

Objective: Competence of the tricuspid valve is crucial for survival of children with hypoplastic left heart syndrome. We studied the morphology and topology of the valvar and subvalvar structures, trying to identify abnormalities which could impair valvar function. Methods: A total of 82 specimens with hypoplastic left heart syndrome were examined pathologically. Measurements of valvar dimensions were taken, significant dysplasia of the valvar leaflets was noted and the muscular and tendinous supporting structures determined. The findings were correlated to the subgroups of hypoplastic left heart syndrome. Results: Of the hearts, 10 (12%) showed a bileaflet right atrioventricular valve, 27 (33%) a moderately and 2 (2%) a severely dysplastic tricuspid valve. The majority of the abnormalities was found in hearts with a patent mitral valve. In 79% of the hearts with mitral atresia, the septal surface was concave instead of convex to the right ventricular lumen and the direct tendinous attachments of the septal leaflet replaced by a multitude of freestanding papillary muscles. The number of direct septal attachments was significantly higher in hearts with a patent mitral valve. Conclusions: The tricuspid valve in hypoplastic left heart syndrome can differ from the valve seen in normal patients. The subvalvar apparatus is different in hearts with mitral atresia, whereas dysplasia of the leaflets occurs more often together with mitral stenosis. These features should be considered in reconstructive operations as well as during diagnostic procedures. © 1997 Elsevier Science B.V.

Keywords: Aortic atresia; Aortic stenosis; Hypoplastic left heart syndrome; Mitral atresia; Mitral stenosis

1. Introduction

Hypoplastic left heart syndrome represents a spectrum of congenital malformations, which, without surgical intervention, is uniformly fatal within the first weeks of life. Besides the possibility of cardiac transplantation in the newborn or infant, reconstructive surgical techniques have now been in use for over a decade. Norwood and his colleagues established a three-staged concept for treatment, resulting in a modified Fontan-circulation after initial augmentation of the hypoplastic ascending aorta [1,2]. Since mortality of these complex surgical procedures is steadily decreasing, a growing number of children live with the morphologically right ventricle, and the corresponding right atrioventricular and ventriculoarterial valves, supporting the systemic circulation. Significant tricuspid regurgitation occurs only rarely, but, if present, can cause severe clinical problems [3–6]. Mild regurgitation, frequently observed, does not represent a risk factor for operative treatment, but its long-term effects are, as yet, not known [7,8].

Although the morphology of the overall hypoplastic left heart syndrome has been extensively reviewed, relatively little attention has been paid to the potentially crucial tricuspid valve [9–13]. Structural abnormalities of the leaflets are rare, but, to the best of our knowledge, the supporting subvalvar structures have been studied only by Bharati and Lev [14]. To add further information, therefore, we studied the morphology and topology of the valvar and subvalvar apparatus of the tricuspid valve in 82 hearts falling in the spectrum of hypoplastic left heart syndrome, focusing on abnormal-
of the ventricular septum, as seen between adjacent leaflets. Special attention was paid to the arrangement of the zones of apposition (commissures) of each leaflet. The nature, number, and location of the papillary muscles were noted, together with the arrangement of the zones of apposition (commissures) between adjacent leaflets. Special attention was paid to the supporting structures of the septal leaflet. We also assessed the shape of the ventricular septum, as seen from the right ventricle, as being convex (bulging into the right ventricular cavity as in normal hearts), straight, or concave (giving the right ventricular cavity an almost circular cross-sectional appearance).

Of the 82 patients, 42 were male and 40 were female. The age at the time of death ranged between 1 day and 1 year, with a mean of 22 ± 61 days. Of 8 patients who had undergone surgery: 3 died after a first stage Norwood procedure, 2 after implantation of a systemic-to-pulmonary arterial shunt; 2 after aortic valvar commisurotomy; and 1 after resection of an aortic coarctation. Associated cardiac malformations, other than those belonging to the category of hypoplastic left heart syndrome or concerning the tricuspid valve, were rare. A large defect within the oval fossa was found in one, partially anomalous pulmonary venous connection in another, multiple but diminutive ventricular septal defects in two; and bilateral arterial ducts in a final patient.

### 3. Results

Of the hearts, 38 (46%) showed mitral atresia and aortic atresia, 27 (33%) showed mitral stenosis and aortic atresia, 16 (20%) showed combined mitral and aortic stenosis and 1 heart (1%) showed mitral atresia with aortic stenosis. The dimensions of the right ventricle, the tricuspid valvar orifice, and the width of each leaflet are shown in Table 1. There is no statistically significant difference in those dimensions between the three groups of specimens.

A right atrioventricular valve with only two leaflets was found in none of the hearts with combined left-sided atresia, but was seen in 4 (15%) of the hearts with mitral stenosis and aortic atresia, and 6 (38%) of the hearts with combined stenosis, with an overall incidence of 12.2%. We found 2 hearts with a quadricuspid right atrioventricular valve. Accessory orifices were present in 2 hearts, both with mitral stenosis and aortic atresia (Fig. 1 and Fig. 2).

Dysplasia of the tricuspid valvar leaflets was distributed with similar frequency throughout the groups (Table 2), but a markedly higher incidence of structural or architectural abnormalities was found in presence of a patent mitral valve (Fig. 1a). Ebstein’s malformation
was discovered involving both the tricuspid and mitral valves in one case (Fig. 1b). Interestingly, all 3 hearts from patients who died after the first stage of a Norwood procedure showed abnormalities of the tricuspid valve; in one case the heart had three delicate leaflets but prolapse of the antero-superior leaflet, another heart showed a bileaflet right atrioventricular valve with moderately dysplastic leaflets and the third heart had severely dysplastic leaflets as part of a parachute malformation.

Our observations concerning the supporting apparatus of the tricuspid valve reflected the known variability of right ventricular structures. Nevertheless, we noted some features obviously related to the hypoplastic left heart syndrome. In all but two of the hearts with combined mitral and aortic atresia, three distinct groups of papillary muscles were identified. They were the prominent anterior papillary muscle, a more variable group of inferior papillary muscles, and a medial

![Fig. 1.](a) A heart with mitral atresia and aortic stenosis displayed to show the right side. The leaflets of the tricuspid valve are thickened with rolled edges (arrowheads). The medial papillary muscle is not evident. A small ventricular septal defect (VSD) is present. (b) This heart with mitral stenosis and aortic atresia had Ebstein’s malformation affecting both the tricuspid and mitral valves. The tricuspid valve has been opened through its major orifice. Note the apically displaced hinge of the septal leaflet (small arrows). The regular atrioventricular junction is marked by the broken line. The medial papillary muscle is absent. In its place is an anomalous muscle band (cut through between the stars) attached to the supraventricular crest and the parietal wall. The open arrowheads indicate two accessory orifices.

![Fig. 2.](a) The right side of this heart shows a severely malformed tricuspid valve with a bizarre arrangement of valvar apparatus. The septal leaflet has multiple insertions to papillary muscles located postero-inferiorly, and in the apex instead of direct attachments to the septum. A bridge of valvar tissue between the septal and antero-superior leaflets results in the formation of an accessory orifice (arrowhead). (b) The left side shows a tiny accessory orifice (arrowhead) in the mitral valve, a short antero-lateral papillary muscle and direct attachments of the mural leaflet to the ventricular wall. The stars indicate the coronary sinus.

### Table 2

<table>
<thead>
<tr>
<th>Subtype of HLHS</th>
<th>TV-dysplasia</th>
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<tbody>
<tr>
<td></td>
<td>Moderate</td>
</tr>
<tr>
<td>MA/AA (n = 38 + 1)</td>
<td>6 (15%)</td>
</tr>
<tr>
<td>MS/AA (n = 27)</td>
<td>13 (48%)</td>
</tr>
<tr>
<td>MS/AS (n = 16)</td>
<td>8 (50%)</td>
</tr>
<tr>
<td>Total (n = 82)</td>
<td>27 (33%)</td>
</tr>
</tbody>
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AA, aortic atresia; AS, aortic stenosis; HLHS, hypoplastic left heart syndrome; MA, mitral atresia; MS, mitral stenosis; TV, tricuspid valve.
papillary muscle arising from the supraventricular crest and supporting the zone of apposition between the septal and the antero-superior leaflets. In hearts with a patent mitral valve, the supraventricular crest was markedly shallow, so that in 12 (44.4%) of the hearts with mitral stenosis and aortic atresia, and in 9 (56.3%) of the hearts with combined stenosis, no distinct medial papillary muscle could be identified. The zone of apposition between the septal and antero-superior leaflets was supported mostly by tendinous cords attached to the supraventricular crest.

A prominent anterior papillary muscle was present in every heart examined, supporting the antero-superior leaflet itself as well as the zone of apposition between the antero-superior and the inferior leaflets. The inferior papillary muscle showed many variations of size and number of subunits, as is well known for normal hearts. This was the case for all the abnormal hearts examined, irrespective of the patency of the mitral valve. The support of the free edge of the septal leaflet was closely related to the morphology of the ventricular septum. In 30 (79%) of the hearts with combined left-sided atresia, the septal surface was clearly concave to the right ventricular lumen. Very few of these hearts showed the typical direct septal attachments of the septal leaflet. Instead, the leaflet was supported by a multitude of additional muscles arising from the septal part of the ventricular wall. Only in 19 and 25%, respectively, of the hearts with mitral stenosis and aortic atresia, or combined stenosis, was the septal surface concave. In the majority of these hearts, the septum was either straight or convex, as seen in normal hearts (Fig. 3). The number of direct tendinous cords arising from the septum was significantly higher in these hearts ($P = 0.0002$).

4. Discussion

The capacity of the morphologically right ventricle to support the systemic circulation has long been debated. In the very particular conditions of the Fontan circulation, as occurs after completion of the Norwood Procedure, this feature becomes even more crucial. Even if the presence of mild tricuspid regurgitation does not necessarily have a negative impact on the operative results of palliative surgery for hypoplastic left heart syndrome [7,8], there can be no question that significant atrioventricular valvar regurgitation reduces long-term hemodynamic performance after the classic or modified Fontan Operation [4,5,15]. After the early morphologic studies on hypoplastic left heart syndrome focused on the deficient structures of the left heart [9,10], the success of the Norwood Procedure drew attention to the now essential right ventricle [12–14]. The overall incidence of moderately or severely dysplastic tricuspid leaflets in our material is similar to that reported by other groups [13,14]. What has not been shown before is the markedly higher incidence of dysplastic changes in hearts with a patent mitral valve. We are unable to offer an explanation for this fact. It is surprising to find a higher degree of abnormal tricuspid leaflets in those hearts with a patent mitral valve which might be considered to be ‘closer to normal’.
As in the normal heart [15,16], the tricuspid valve was not perfectly tricuspid in some cases. The bi- or quadricuspid constitution of the valve is unlikely to affect valvar competence. More importantly, accessory orifices, though rare, was observed in two of our cases but none in the 230 specimens examined by Bharati and Lev [14]. In addition, there was I heart in our series in which Ebstein’s malformation distorted both the tricuspid and mitral valves. These malformations can certainly influence surgical outcome if missed at initial diagnosis.

Concerning the subvalvar apparatus, our findings are somewhat easier to explain. Bharati and Lev [14] described in great detail the attachment of the septal leaflet to the septum by accessory papillary muscles in their large series, and emphasised also the abnormal architecture of the other groups of papillary muscles. We were able to correlate these marked changes in intraventricular architecture to the morphology of the ventricular septum. In hearts with a minute left ventricle, the septum becomes functionally part of the free wall of the right ventricle, giving its cavity an almost circular shape with a concave septal surface. Since the tricuspid valve is much more closely related to the septum than is the mitral valve [17,18], this causes fundamental changes in the architecture of the subvalvar apparatus. The typical direct attachments of the septal leaflet are replaced by additional free-standing papillary muscles. We take these changes as an adaptive process; nature might try to prevent incompetence of the valve by giving the septal leaflet a greater degree of mobility. In hearts with a more pronounced ventricular septum, the usual architecture of the septal part of the tricuspid valve is more readily preserved.

Overall, therefore, we found markedly dysplastic tricuspid valves almost exclusively in hearts with a patent mitral valve. Since the prognosis of these patients may potentially be limited, we must try to identify them as soon as possible. We also demonstrated that the right atrioventricular valve in patients with hypoplastic left heart syndrome can be very different from the morphologically tricuspid valve in normal patients. The echocardiographer must be aware of this feature while assessing these crucial structures, as must the surgeon when it comes to reconstructive operations.

4.1. Limitations of the study

The difficulties of assessing sizes and shapes in formalin-fixed hearts are well known. Because of this, we tried to concentrate on clear morphologic features of the examined structures. The shape of the ventricular septum was so well defined in all specimens we believe that it almost certainly reflects the conditions in the living patient. Since the majority of the examined hearts came from patients only a few days old at the time of death, we could not examine the group of greatest interest, namely patients surviving all the stages of the Norwood Procedure. With a growing morphologically right ventricle, the septal structures are likely to become even more deficient, thus making the right atrioventricular valve in those patients even less a ‘normal’ tricuspid valve. The long-term anatomic and physiologic changes imposed by a circulation supported by a functionally single right ventricle remain unclear, and surely need further investigation.

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References


