Left ventricular function in double inlet left ventricle before the Fontan operation: comparison with tricuspid atresia

ANDREW N REDINGTON, BRODIE KNIGHT, PAUL J OLDERSHAW, ELLIOT A SHINEBOURNE, MICHAEL L RIGBY

From the Brompton Hospital, London

SUMMARY Fourteen patients with double inlet left ventricle and nine patients with tricuspid atresia had biplane left ventricular angiography with simultaneous measurement of left ventricular pressure by micromanometer. Age distribution, haemodynamic function, and previous palliative operation were similar in the two groups. Left ventricular volumes were calculated frame by frame throughout the cardiac cycle by Simpson's rule. The end diastolic volume index was similar in the two groups, but the ejection fraction was significantly lower in tricuspid atresia. Left ventricular peak filling and emptying rates were also lower in tricuspid atresia, although heart rates in the two groups were similar. End diastolic shape index was significantly higher in patients with tricuspid atresia, indicating a more globular shape, and changed less during systole, suggesting differences in the mechanism of ejection between the two groups. Analysis of pressure-volume loops showed normal phase relations between pressure and volume, but systolic stroke work was reduced in tricuspid atresia and correlated with stroke volume and shape change.

Left ventricular function was impaired in patients with tricuspid atresia when compared with those with double inlet left ventricle and this finding may reflect structural differences caused by the absence of one atrioventricular connection.

Various radical palliative procedures based on Fontan's original operation have been introduced to correct the circulation in patients with tricuspid atresia or double inlet left ventricle.\textsuperscript{12} It has been suggested that "normal" left ventricular function is of fundamental importance to survival after these procedures.\textsuperscript{3} More recently, however, several groups have shown depressed left ventricular ejection fraction in patients with tricuspid atresia both before and after primary palliative procedures such as systemic to pulmonary shunting and pulmonary artery banding.\textsuperscript{4-44} There is little published data on ventricular function in double inlet left ventricle but our own preliminary data have suggested that some indices of left ventricular function are normal or near normal in these patients.\textsuperscript{7} It is somewhat surprising, therefore, if ventricular function is an important determinant of outcome, that perioperative mortality is lower in patients with tricuspid atresia than in patients with double inlet left ventricle undergoing modified Fontan procedures.\textsuperscript{8}

We studied left ventricular function in a group of patients with double inlet left ventricle and compared these results with similar measurements in patients with tricuspid atresia in order to explore the possible causes of any differences between the two groups and to assess the influence of ventricular function on the outcome of corrective surgery.

Patients and methods

TERMINOLOGY

The segmental approach to the classification of univentricular hearts has been described in detail elsewhere.\textsuperscript{9} In hearts with a univentricular connection the atrial chambers connect either entirely or predominantly to one ventricle. In all patients in this study the atria connected to a main chamber of left ventricular morphology and the right ventricle was rudimentary.

Patients were described as having double inlet left
ventricle when one atrioventricular valve and more than 50% of the other valve were connected to a morphologically left ventricle. Under these circumstances one or other of the atrioventricular valves may be stenotic, imperforate, straddling, or overriding; these additional abnormalities are described separately (table 1). When there was a common atrioventricular valve the connection was said to be double inlet when > 75% of the valve was connected to the left ventricle.

The other group had classic tricuspid atresia, that is absence of the right atrioventricular connection with interposition of sulcus tissue between right atrium and the rudimentary right ventricle. No patient had the much rarer abnormality of atrioventricular concordance with imperforate right atrioventricular valve. The distinction between these two abnormalities was made by cross sectional echocardiography according to previously described echocardiographic criteria.10

The position of the rudimentary right ventricle and the ventriculoarterial connection in each patient was determined by cross sectional echocardiography and angiography.

Table 1  Sequential diagnosis and previous operation in 14 patients with double inlet left ventricle and nine with tricuspid atresia

<table>
<thead>
<tr>
<th>Patient</th>
<th>AV connection*</th>
<th>RV to LV relation</th>
<th>VA connection</th>
<th>Associated lesions</th>
<th>Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Double inlet left ventricle</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>2 valves</td>
<td>Right</td>
<td>Concordant</td>
<td>Absent left PA</td>
<td>Waterston shunt</td>
</tr>
<tr>
<td>2</td>
<td>2 valves</td>
<td>Left</td>
<td>Concordant</td>
<td>Pulmonary atresia</td>
<td>Waterston and Blalock shunt</td>
</tr>
<tr>
<td>3</td>
<td>Imperforate</td>
<td>Right</td>
<td>Single outlet</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>2 valves</td>
<td>Right</td>
<td>Discordant</td>
<td>Subpulmonary stenosis and valvar PS</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>2 valves</td>
<td>Left</td>
<td>Discordant</td>
<td>Valvar PS</td>
<td>Blalock shunt</td>
</tr>
<tr>
<td>6</td>
<td>2 valves</td>
<td>Right</td>
<td>Discordant</td>
<td>Ao interruption</td>
<td>Repaired Ao interruption, PA band</td>
</tr>
<tr>
<td>7</td>
<td>2 valves</td>
<td>Left</td>
<td>Discordant</td>
<td></td>
<td>PA band</td>
</tr>
<tr>
<td>8</td>
<td>Common</td>
<td>Right</td>
<td>Discordant</td>
<td>Ao coarctation</td>
<td>Repaired coarctation, PA band (tight + +)</td>
</tr>
<tr>
<td>9</td>
<td>2 valves</td>
<td>Right</td>
<td>Concordant</td>
<td>Pulmonary vascular disease</td>
<td>—</td>
</tr>
<tr>
<td>10</td>
<td>2 valves</td>
<td>Right</td>
<td>Discordant</td>
<td>Ao coarctation</td>
<td>Repaired coarctation</td>
</tr>
<tr>
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<td>Discordant</td>
<td></td>
<td>PA band</td>
</tr>
<tr>
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<td>2 valves</td>
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<td>Discordant</td>
<td>Ao interruption, restrictive VSD</td>
<td>Repaired Ao interruption, enlarged VSD, PA band</td>
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<tr>
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<td>Right</td>
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<td></td>
<td>PA band</td>
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<tr>
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<td>2 valves</td>
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<td>Discordant</td>
<td>Double outlet LV PS</td>
<td>Shunt</td>
</tr>
<tr>
<td>15</td>
<td>Tricuspid atresia</td>
<td>Discordan</td>
<td>Double outlet RV</td>
<td>Restrictive VSD, Subpulmonary stenosis, pulmonary valve stenosis</td>
<td>PA band</td>
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<td>16</td>
<td>Tricuspid atresia</td>
<td>Discordant</td>
<td>Double outlet RV</td>
<td>Subpulmonary stenosis, pulmonary valve stenosis</td>
<td>Shunt</td>
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<tr>
<td>17</td>
<td>Tricuspid atresia</td>
<td>Concordant</td>
<td>Double outlet RV</td>
<td>Subpulmonary stenosis</td>
<td>Shunt</td>
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<tr>
<td>18</td>
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<td>Concordant</td>
<td>Double outlet RV</td>
<td>Subpulmonary stenosis</td>
<td>Shunt</td>
</tr>
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<td>Discordant</td>
<td></td>
<td></td>
<td>Band</td>
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<td>Discordant</td>
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<td></td>
<td>Band</td>
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<td>Concordant</td>
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<td></td>
<td>PA band</td>
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<tr>
<td>22</td>
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<td>Concordant</td>
<td></td>
<td></td>
<td>Shunt</td>
</tr>
<tr>
<td>23</td>
<td>Tricuspid atresia</td>
<td>Single outlet</td>
<td></td>
<td>Pulmonary atresia</td>
<td>Shunt</td>
</tr>
</tbody>
</table>

*All patients have situs solitus.

PA, pulmonary artery; Ao, aortic; PS, pulmonary stenosis; VSD, ventricular septal defect; AVV, atrioventricular valve; RV, right ventricle; LV, left ventricle.
Table 2  Haemodynamic data (mean (SD)) recorded before angiography

<table>
<thead>
<tr>
<th></th>
<th>Tricuspid atresia</th>
<th>Double inlet left ventricle</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peak LV systolic pressure (mm Hg)</td>
<td>100 (24)</td>
<td>108 (21)</td>
<td>NS</td>
</tr>
<tr>
<td>LV end diastolic pressure (mm Hg)</td>
<td>9 (4)</td>
<td>8 (4)</td>
<td>NS</td>
</tr>
<tr>
<td>Aortic oxygen saturation (%)</td>
<td>86 (3)</td>
<td>84 (8)</td>
<td>NS</td>
</tr>
</tbody>
</table>

LV, left ventricular.

mark, coinciding with the R wave of the patient's electrocardiogram, was recorded on the cinefilm. After the position of the rudimentary ventricle had been displayed by echocardiography, the angio-
graphic projections were selected to show the interventricular septum. The projections selected were direct anteroposterior and lateral when the rudimentary chamber was to the left of the main chamber and 30° right anterior oblique-60° left anterior oblique when it was to the right. In 12 of the patients with double inlet left ventricle and in five with tricuspid atresia left ventricular pressure was measured with a micromanometer tipped catheter (Millar Instruments) during ventriculography. The output of this catheter was recorded at a paper speed of 200 mm/s along with an electrocardiogram and an event mark that coincided with the appearance of contrast medium in the ventricle. Thus it was possible to synchronise the angiogram and pressure recordings. At the end of the study the patient was moved 10 cm with a catheter in the left ventricle for later calibration of the cinefilm.

**Analysis of angiograms**

Only normally conducted beats were analysed. Extrasystoles and post-extrasystoles were excluded. Our method of analysis has been described in detail previously. Briefly, successive cineframes were projected on to a Summagraphics digitising plate interfaced with a Prime 750 computer. The left ventricular outline, excluding the rudimentary right ventricle, was traced frame by frame, starting and finishing with frames corresponding to the R wave of the electrocardiogram. Approximately 100 x,y coordinates were recorded for each outline and stored on magnetic tape for later analysis. This procedure was repeated for the same cycle in the other angiographic view and finally the corresponding left ventricular pressure trace was digitised on the same equipment.

From this stored data the following were derived:

**Left ventricular volume**

Because the shape and position of these ventricles are abnormal it is inappropriate to measure their volume from single plane angiograms according to the geometric assumptions derived from the normal left ventricle. In this study we calculated ventricular volume from the orthogonal biplane angiograms using Simpson's rule. This is a multiple slice method which overcomes some of the geometric assumptions inherent in other methods of calculating left ventricular volume. Although no angiographic method is perfect, our method, previously described in detail, was accurate when compared with left ventricular cast volumes measured by fluid displacement. Briefly, the left ventricle was divided into 40 equal slices along its long axis. The cross sectional area of each segment was assumed to be an ellipse and its major and minor dimensions were calculated from each of the angiographic views. The volume of the ventricle was derived from the sum of the volume of each slice by applying Simpson's rule (fig 1).

Ventricular volume was measured frame by frame throughout the cardiac cycle and displayed along with its first derivative, dVol/dT (fig 2). End diastolic and end systolic volumes were later corrected for body surface area and expressed in ml/m². The ejection fraction was calculated in the usual way. Peak rates of ventricular filling and emptying were taken as the maximum positive and negative deflections on the first derivative of the volume-time curve and were normalised by dividing by end diastolic volume (units = s⁻¹).

**Ventricular shape index**

This is a measure of the roundness of the outline of the ventricular cavity (fig 2) that is independent of volume. Calculated as 4π (cavity area)/(perimeter)² × 100 it has a maximum value of 100 when the cavity outline is circular and 0 when the cavity is obliterated. The normal left ventricle shows a progressive reduction in shape index throughout systole, as it becomes a less circular shape. In this study end diastolic, end systolic, and percentage systolic change were taken as an average of the two angiographic views.

**Pressure-volume relations**

The frame by frame measurement of ventricular volume has been described above. The derived volume-time curve and the simultaneous digitised pressure trace were combined to give an instantaneous pressure-volume loop. The left ventricular systolic stroke work performed on the circulation is equal to the area enclosed by the loop and this was measured by automatic planimetry and expressed in J/beat/m² body surface area (1 J = 1.36 × 10⁻⁴ mm Hg.ml). The cycle efficiency of the loops was expressed as a ratio of the area of the loop to that of a square or rectangle that just encloses it (that is the ratio of actual work done to the maximum possible for...
ventricle with the same stroke volume and pressure change).

**STATISTICAL ANALYSIS**

Group data were compared by standard t tests and the null hypothesis was rejected when \( p < 0.05 \). We used linear regression analysis by the method of least squares to examine correlations between variables.

**Results**

No patient had important atrioventricular valve regurgitation. Table 3 summarises the data derived from the angiograms. Mean (SD) end diastolic volume was 136 (32) and 129 (40) ml/m² for the groups with double inlet left ventricle and tricuspid atresia respectively (\( p = \text{NS} \)). End systolic volume, however, was significantly lower in patients with double inlet left ventricle (45 (18) vs 64 (26) ml/m² (\( p = 0.05 \)). Thus ejection fraction was higher in the double inlet group (74 (9) vs 51 (6.5)%, \( p < 0.001 \)). Furthermore, nine patients in the double inlet group had an ejection fraction of \( \geq 75\% \) and only one patient had an ejection fraction of \( < 60\% \).

In contrast none of the patients with tricuspid atresia had an ejection fraction of \( > 60\% \) (fig 3). There were no differences in ventricular volumes or ejection fraction when patients in the various operative subgroups were compared and there was no significant relation between ejection fraction and age, age at or time from operation, or aortic oxygen saturation for either group.

The ventricular shape index at end diastole (averaged from the two angiographic views) was higher in tricuspid atresia (88 (6) vs 81 (6) \( p < 0.05 \)), indicating a more globular shape. Systolic shape index change in patients with tricuspid atresia was reduced (5.8 (4.3) vs 18 (7)%, \( p < 0.001 \)), suggesting differences in the mechanism of ejection between the two groups. There was a significant correlation between systolic shape index change and ejection fraction (all patients, \( r = 0.63, p < 0.02 \)).

Peak rates of ventricular filling corrected for end diastolic volume were 5.9 (2.5) and 4.1 (1.2) s⁻¹ for the groups with double inlet or tricuspid atresia respectively (\( p < 0.05 \)). There was a strong negative correlation between peak filling rate and RR interval in the patients with double inlet left ventricle (\( r = -0.72, p < 0.001 \)) but not in those with tricuspid atresia (\( r = -0.47, p = \text{NS} \)). Time to peak filling rate was less in tricuspid atresia (66 (22) vs 82 (40) ms) but this difference did not reach statistical significance. The peak rate of ventricular emptying was also reduced in patients with tricuspid atresia.
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catheterisation (oxygen saturation = 62%), the pressure-volume loop showed an abnormal diastolic limb with a much reduced cycle efficiency (45%).

Systolic stroke work, measured by planimetry of the loops, was lower in tricuspid atresia and correlated (p < 0.05) with stroke volume and shape change (all patients, r = 0.73 and 0.7 respectively).

Discussion

Infants with univentricular atrioventricular connection account for up to 10% of symptomatic patients presenting with congenital heart disease. In many of these patients the condition can be palliated in early life and later such patients may be suitable for a corrective procedure. The accurate assessment of ventricular function in these patients, both before and after such procedures, is therefore of considerable importance.

Unlike the normal left ventricle, which approximates to a prolate ellipsoid, the left ventricle in these patients may vary considerably in size, shape, and position. Whatever method of measurement is used these factors are likely to influence the results when assessing ventricular function from angiograms. In particular, the application of algorithms derived for the measurement of normal left ventricular volume from single plane angiograms is not valid. In this study we used a computer assisted method to measure left ventricular volume from biplane angiograms using a modification of Simpson's rule.
This has previously been shown to be an accurate method of volume measurement that overcomes some of the geometric assumptions inherent in single plane derivations. The ventricle is divided into 40 elliptical slices and the major and minor dimensions of these are taken from each of the angiographic views. Several groups have used similar methods and have reported raised ventricular volumes and depressed ejection fraction in patients with tricuspid atresia, but there have been few attempts to measure more sensitive indices of function or to compare function in these patients with those who have other forms of univentricular atrioventricular connection. Left ventricular end diastolic volume, corrected for body surface area, was similar in our patients with tricuspid atresia and those with double inlet left ventricle. As might be expected, however, the absolute values for end diastolic volume were considerably higher than normal in these ventricles receiving both systemic and pulmonary venous return. The ejection fraction in our patients with tricuspid atresia was similar to that reported with both angiographic and radionuclide techniques. Unlike other studies, however, we were unable to confirm any significant relation in our small study between ejection fraction and age, previous systemic-to-pulmonary shunt procedures, or systemic oxygen saturation. Indeed the finding of normal, and often supranormal, ejection fraction in the patients with double inlet left ventricle, of similar age range, haemodynamic function, and distribution of previous palliative procedures suggests that these factors may not be primary determinants of the reduced ejection fraction seen in patients with tricuspid atresia.

It is perhaps more likely in the light of our findings that the cause of the reduced ejection fraction in tricuspid atresia is structural. The normal left ventricle ejects a considerable proportion of its stroke volume by an alteration of its shape during systole. In a previous study this normal change in shape seemed to be much reduced in patients with univentricular hearts although in four patients, all with two atrioventricular valves, this change in shape seemed to follow more closely the pattern of the normal left ventricle. In our patients with double inlet left ventricle both the end diastolic shape index and the change in shape index during systole were similar to normal values. In contrast in tricuspid atresia the left ventricle was more globular at end diastole and changed little during systole; thus virtually all the change in cavity volume in these
ventricles resulted from myocardial fibre shortening. It is possible that disruption of the normal myocardial fibre architecture, because of the absence of an atrioventricular connection, reduces the ability of the ventricle to change its cavity volume by changing shape. Indeed the ejection fraction and change in shape index were normal in a single patient studied with atrioventricular concordance and an imperforate right atrioventricular valve but otherwise similar haemodynamic function to classic tricuspid atresia (unpublished). The significant linear relation between change of shape and ejection fraction in our patients further supports this concept but cannot exclude a common, unrecognised, aetiological factor.

Frame by frame analysis of ventricular volume shows further differences in both systolic and diastolic indices of function. The peak rate of ventricular filling was significantly lower in patients with tricuspid atresia than in those with double inlet left ventricle. It is clear, however, that peak filling rate is higher in those with a double inlet than previously published normal values. It is possible that peak filling rate is higher than normal in these patients because the ventricle is filling through two atrioventricular valves. There was a significant relation between the RR interval and peak filling rate in the double inlet group ($r = 0.73$), however, and it is likely that the increase in peak filling rate was related to the relatively high heart rate in these patients. There was no haemodynamic evidence of inflow obstruction in our patients with tricuspid atresia, either across the atrial septum or across the left atrioventricular valve, and so filling was probably impaired when compared with normal. The time to peak filling rate was also reduced in these patients and this combination of an early yet reduced peak rate of ventricular filling is similar to that seen in adults with a raised end systolic volume and reduced ejection fraction secondary to ischaemic cardiomyopathy.

Despite the abnormalities described above the left ventricular pressure-volume loops showed a coordinate phase-relation between pressure and volume in most patients, and, apart from a single patient with reduced pulmonary blood flow and severe systemic desaturation, the cycle efficiency of the loops was normal. Systolic stroke work, equal to the area enclosed by the pressure-volume loop, was higher in the group with double inlet left ventricle simply because of the higher stroke volume in these patients.

The implications of these findings for corrective or radical palliative surgery are unclear. It has been suggested that normal preoperative ventricular function is an essential prerequisite for the Fontan operation or its modifications. Yet the survival of patients after these procedures appears to be higher in patients with tricuspid atresia than in those with double inlet left ventricle. It seems that in most young patients undergoing Fontan-like procedures preoperative ventricular function is not a primary determinant of survival. It is possible that the influence of preoperative ventricular function will vary depending on the type of atrioventricular connection, however. Furthermore, it is not known how preoperative ventricular function will affect performance in the survivors of these operations. We have increasing evidence that cardiac output and exercise tolerance are reduced after the Fontan procedure but there are little data relating these findings to measurements of preoperative ventricular function. A large scale longitudinal study of these patients would help to answer some of these questions.

Our results show that ventricular function in patients with tricuspid atresia is often worse than in those with double inlet left ventricle. These abnormalities are more likely to be related to structural differences rather than to differences in haemodynamic function or the effects of previous palliative operation. The higher survival after the Fontan operation of patients with tricuspid atresia suggests that ventricular function is not a primary determinant of outcome after this procedure.

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References

8 Pacifico AD, Kirklin JK, Kirklin JW. Surgical
Left ventricular function in double inlet left ventricle before the Fontan operation


