Radionuclide measurement of left ventricular ejection fraction in tricuspid atresia

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SUMMARY In 15 patients with tricuspid atresia and one with tricuspid stenosis the left ventricular ejection fraction was measured by equilibrium gated radionuclide angiography and the results compared with those from a control group of 16 patients. The patients with tricuspid atresia had a significantly depressed ejection fraction. Those who had a surgical shunt or who had had pulmonary artery banding had significantly lower ejection fractions than the remainder. There was no significant correlation between the ejection fraction and age, the arterial oxygen saturation, or the haemoglobin concentration. Five patients were also studied during isometric exercise; three had an abnormal response. Volume overload of the ventricle is identified as one cause of the dysfunction; but other factors may be important. Radionuclide angiography offers a non-invasive method of studying ventricular function in this condition.

In recent years corrective surgery for tricuspid atresia—defined, in situs solitus, as absence of the right atrioventricular connexion—has become feasible.1 Adequate left ventricular function is an essential prerequisite to successful correction.2-3 Measurements from left ventricular cineangiograms have suggested that in many patients left ventricular function is depressed.4-7 Radionuclide angiography offers a non-invasive method for the sequential measurement of left ventricular function, but there are no reports of its use in this condition. We report the use of equilibrium gated radionuclide angiography to measure the left ventricular ejection fraction in 15 patients with tricuspid atresia and one with severe tricuspid stenosis.

Patients and methods

STUDY POPULATION
Fifteen patients with tricuspid atresia were studied; the diagnosis was confirmed in all at diagnostic cardiac catheterisation. One further patient had severe tricuspid stenosis with a hypoplastic right ventricle and a ventricular septal defect. As her diagnosis was indistinguishable clinically from tricuspid atresia she was included in the study. The patients ranged in age from 6 weeks to 34 years (mean 9-6 years). Previous corrective surgery had not been performed, but six patients had surgical shunts (4 Blalock-Taussig anastomoses or similar prosthetic shunts, one a Waterston shunt, and one a Glenn operation followed four years later by a Blalock shunt). Three patients had pulmonary artery banding in infancy; seven had not undergone any surgery.

Sixteen patients were selected as a control group as they did not have significant left ventricular pressure or volume overload and none of them was cyanosed. They were aged 1 month to 14 years (mean 7-1 years). Their diagnoses were pulmonary stenosis (10), atrial septal defect (two), and ventricular septal defect with a pulmonary to systemic flow ratio of less than 1:5:1 (four).

ANGIOGRAPHY
Equilbrium electrocardiogram gated radionuclide angiograms were acquired at rest in all the patients, and also during isometric handgrip exercise in five. The method of acquisition has previously been described and validated.8 The patients’ own red blood cells were labelled with technetium-99m in vitro by the Brookhaven method. The dose was 430 MBq/m². The projection that best separated the left ventricle from surrounding structures was chosen. Usually this was a left anterior oblique projection, but in one patient with dextrocardia a right anterior oblique projection was used. A minimum of three million counts was collected in every study.
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For the exercise study the patient compressed a dynamometer with their right hand maintaining approximately one third of their maximum compression for the period of the acquisition. Data were acquired after they had been exercising for one minute.

STATISTICAL ANALYSIS

Results are given as mean (standard deviation). Individual population means were compared by the t test and relations by linear regression analysis. The null hypothesis was rejected if p>0.05.

Results

The Table gives the patient data together with the resting and exercise left ventricular ejection fractions. The mean ejection fraction for all the study patients was 50.8(9.5)% compared with the control group, whose mean was 67.8(6.2)% (p<0.001). The mean ejection fraction for all those patients who had not had surgery was 55.7(8.9)% (n=7) and for those who had had either a surgical shunt or a pulmonary artery band 46.9(8.4)% (n=9) (p<0.05).

Even though the patients who had not undergone surgery had a higher resting ejection fraction than the remainder it was still significantly lower than that of the control group (p<0.01). There was no significant difference in values between those patients with shunts and those with banded pulmonary arteries. There was no significant correlation between the resting ejection fraction and age, haemoglobin concentration, or arterial oxygen saturation. In those patients with surgical shunts there was no significant correlation between the resting ejection fraction and the length of time since the creation of the shunt.

Three of the five patients studied during isometric exercise had an abnormal response. Overall there was a mean fall in ejection fraction of 3.2(4.8)% (Figure). The fall was greatest in those patients with the lowest resting ejection fraction. In none of the patients was there a significant rise in the ejection fraction with exercise.

Discussion

These results broadly agree with those of previous invasive studies in tricuspid atresia. It is clear that left ventricular performance is depressed in the majority

Table Patient data and left ventricular ejection fraction at rest and during exercise

<table>
<thead>
<tr>
<th>Case No</th>
<th>Age (yr)</th>
<th>Resting LVEF (%) (with exercise)</th>
<th>Oxygen saturation (%)</th>
<th>Haemoglobin concentration (g/dl)</th>
<th>Surgery</th>
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<tbody>
<tr>
<td>1</td>
<td>16</td>
<td>51</td>
<td>82</td>
<td>17.7</td>
<td>Waterson shunt</td>
</tr>
<tr>
<td>2</td>
<td>22</td>
<td>34 (26)</td>
<td>85</td>
<td>18.3</td>
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<tr>
<td>3</td>
<td>11</td>
<td>60 (52)</td>
<td>85</td>
<td>13.4</td>
<td>—</td>
</tr>
<tr>
<td>4</td>
<td>20</td>
<td>58 (60)</td>
<td>84</td>
<td>19.0</td>
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<tr>
<td>5</td>
<td>5</td>
<td>67</td>
<td>85</td>
<td>15.5</td>
<td>—</td>
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<td>1</td>
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<tr>
<td>7</td>
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<td>58</td>
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<tr>
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<td>34</td>
<td>58 (59)</td>
<td>78</td>
<td>15.4</td>
<td>Blalock shunt</td>
</tr>
<tr>
<td>10*</td>
<td>3</td>
<td>50</td>
<td>81</td>
<td>19.5</td>
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<tr>
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<tr>
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<td>3</td>
<td>39</td>
<td>74</td>
<td>19.0</td>
<td>Blalock shunt</td>
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</tbody>
</table>

LVEF, left ventricular ejection fraction (measured by equilibrium gated radionuclide angiography).

*Tricuspid stenosis.
of these patients. There is evidence that prolonged volume overload of the left ventricle alone may cause moderate dysfunction. The degree of dysfunction seen here is, however, more than can be explained by left ventricular volume overload alone.

There is no straightforward relation between left ventricular function and the length of time that volume overload has been present or the degree of hypoxia. Most previous cineangiographic studies have similarly failed to correlate these variables. But in one study there was a significantly lower ejection fraction in those patients who had a surgical shunt for longer than 10 years. In all these studies left ventricular volume overload is identified as being the most significant factor associated with left ventricular dysfunction in these patients.

Normally, with isometric exercise there is no significant change in the left ventricular ejection fraction, whereas adults with coronary artery disease usually have a significant fall. In two of the patients with tricuspid atresia there was a normal response to isometric exercise; both were adults and one had a longstanding surgical shunt and the other had not undergone any surgery. Of the three patients with an abnormal exercise response, one had undergone no surgery and the other two had surgical shunts. The patient with the lowest resting ejection fraction and the greatest decrease with exercise had a Waterson shunt created 12 years previously.

Evidently, the degree of left ventricular dysfunction seen in patients with tricuspid atresia cannot be ascribed to any single factor. Although left ventricular volume overload appears to be the most important factor it must be operating in conjunction with others. Although no correlation was seen between the arterial oxygen saturation or the haemoglobin concentration and left ventricular function, the most likely associated factor would appear to be hypoxia. Alternatively, we may be observing the effects of volume overload on an intrinsically abnormal left ventricle. Some support for this hypothesis is given by the low ejection fractions found in even the youngest patients and those who had not undergone surgery. Furthermore, subtle differences from normal left ventricular function—in terms of ventricular asymmetry and shape change—have been reported in patients with univentricular-tri-ventricular connexion with a normal ejection fraction.

From the recent analysis of the risk factors for the definitive surgery of tricuspid atresia by Fontan and colleagues age at surgery is obviously important. It is not clear what role abnormalities of left ventricular performance play in the risk of such surgery, and the relation between age and left ventricular function in individual patients is not known. Having identified the left ventricular dysfunction in most patients with tricuspid atresia equilibrium gated radionuclide angiography is a valuable tool for investigating these questions.

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References