Tricuspid atresia with transposition of the great arteries in adolescents and adults: current state and late complications

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SUMMARY The outcome was reviewed in 17 patients aged 15–40 years with tricuspid atresia and transposed great arteries selected by survival beyond age 14 years. Only five lead normal lives (ability index 1 or 2); the rest are dead or disabled. Arrhythmias occurred in seven. Maintenance of sinus rhythm is important because incessant atrial arrhythmias cause serious symptomatic deterioration. Pulmonary vascular disease and subaortic stenosis were important determinants of late mortality and morbidity. Because the mortality associated with the Fontan operation was high in these patients it should be performed with impeccable surgical technique and only in those who fulfil all the selection criteria for the operation. A shunt is the preferred option when any of the criteria are not met.

Palliative surgery and the Fontan operation have modified the course of tricuspid atresia and transposed great arteries, but little is known about the outcome in adults with this condition. We have reviewed 17 adolescents and adults with this anomaly and have assessed the factors which contributed to their death and disability.

Patients and methods

We reviewed all patients with angiographically proven tricuspid atresia and transposed great arteries who were seen at the National Heart Hospital, London, since 1965 and who survived beyond age 14 years. The 17 patients (aged 15–40 years) were divided into two groups—group A: 12 patients with palliative shunts or no previous surgery (six are alive to date) and group B: five patients who had a Fontan operation or modification at 15–21 years; only one is alive. The causes of death were analysed and necropsy data were examined when they were available. All living patients were examined in 1986 and had chest x-ray, electrocardiogram, exercise test by the modified Bruce protocol, and 24 hours of ambulatory electrocardiography. The left ventricular ejection fraction was measured by cross-sectional echocardiography and equilibrium electrocardiogram-gated radionuclide angiography. The ability index was determined in each patient to evaluate their quality of life (figure).1

Results

GROUP A: PATIENTS WITH PALLIATIVE SHUNTS OR NO SURGERY

Table 1 shows the data on 12 patients. Nine had pulmonary stenosis. Two (cases 2 and 5) had shunt operations before the age of one, a further five needed surgery during the first decade, and one had a Blalock-Taussig shunt at 17 years. One patient had her first shunt operation at 34 years. Seven patients had Blalock shunts; two a Glenn anastomosis (cases 5 and 9), and three patients had a direct aorto-pulmonary anastomosis—Potts (case 2), Waterston (case 12), or from ascending aorta to left pulmonary artery (case 5). Three patients without pulmonary stenosis (cases 4, 7, and 11) survived more than 14 years without surgery, but had severe pulmonary vascular disease and presented with serious symptoms (haemoptysis, severe hypoxia) in the second and third decades. In a fourth patient (case 5) pulmonary vascular disease only developed after three previous shunt procedures, almost certainly as a
result of a direct aorto-pulmonary artery shunt performed when she was eight years old.

Mortality
Six patients aged 15–22 years died—three at operation. One with the Eisenmenger reaction (case 7) had massive haemoptysis from an infarct in the right lung and died after emergency lobectomy. Another with a failing Glenn anastomosis (case 9), small pulmonary arteries, and mild subaortic stenosis died at an attempted Blalock shunt. This was her third operation. The third death occurred in a patient (case 8) with angina, mild aortic valve stenos, and severe subaortic stenosis; she died in asystole after extensive resection (>4 g) of muscle from the subaortic right ventricle; the ventricular septal defect was large.

Patient 10 died suddenly nine months after an initially successful Blalock shunt performed at the age of 17 years; increasing hypoxia suggested thrombosis of the shunt. A patient (case 12) with an ability index of 2 died from an intracerebral haemorrhage. This was probably caused by habitual glue sniffing; two previous shunts were functioning. Patient 11 died in chronic left ventricular failure with pulmonary vascular disease, mitral regurgitation, and atrial fibrillation.

Survivors: ability index and exercise capacity
Four of the six survivors lead nearly normal lives (ability index of 1 or 2) (table 1). Two patients with pulmonary vascular disease (cases 4 and 5) are more severely limited (ability indices of 3 and 4).

Six patients performed an exercise test; three of the four with no pulmonary vascular disease reached stage III; one (case 2) stopped in stage II. The two patients with pulmonary vascular disease stopped at stage 0.

Left ventricular ejection fraction measured by radionuclide angiography ranged from 36% to 64% (mean 47). Neither of the two patients with impaired ventricular function (cases 3 and 4) have had previous shunts, but both are hypoxic and have polycythaemia that requires regular venesection. One

Table 1  Clinical data on 12 patients with tricuspid atresia and transposition of the great arteries who had palliative procedures or no surgical treatment

<table>
<thead>
<tr>
<th>Case No</th>
<th>Age</th>
<th>Sex</th>
<th>Congenital pulmonary stenosis</th>
<th>Operations performed and age (yr)</th>
<th>Ability index in 1986</th>
<th>Arrhythmias (%)</th>
<th>LVEF</th>
<th>Pulmonary vascular disease</th>
<th>Cause of death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alive:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>40</td>
<td>F</td>
<td></td>
<td>Blalock shunt (dextrocardia)</td>
<td>2</td>
<td>AF</td>
<td>64</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>2*</td>
<td>28</td>
<td>F</td>
<td></td>
<td>Potts anastomosis (1)</td>
<td>2</td>
<td>Multifocal AT</td>
<td>52</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>27</td>
<td>M</td>
<td></td>
<td>Pulmonary valvotomy (7)</td>
<td>2</td>
<td>0</td>
<td>36</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>26</td>
<td>F</td>
<td>0</td>
<td>Blalock shunt (2/12)</td>
<td>4</td>
<td>VT</td>
<td>38</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>22</td>
<td>F</td>
<td>+</td>
<td>Ascending Ao-LPA shunt (8)</td>
<td>3</td>
<td>0</td>
<td>49</td>
<td>+ Acquired late</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>20</td>
<td>M</td>
<td>+</td>
<td>Blalock shunt (6)</td>
<td>1</td>
<td>0</td>
<td>45</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Dead:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>22</td>
<td>M</td>
<td>0</td>
<td>R lower lobectomy (22)</td>
<td>0</td>
<td>Couplets, VPCs</td>
<td>0</td>
<td>+</td>
<td>Operative</td>
</tr>
<tr>
<td>8†</td>
<td>22</td>
<td>F</td>
<td>+</td>
<td>Blalock shunt (2)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>Operative</td>
</tr>
<tr>
<td>9†</td>
<td>18</td>
<td>F</td>
<td>+</td>
<td>Blalock shunt (4)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>Operative</td>
</tr>
<tr>
<td>10</td>
<td>17</td>
<td>F</td>
<td>+</td>
<td>Blalock shunt (17)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>Operative</td>
</tr>
<tr>
<td>11‡</td>
<td>17</td>
<td>M</td>
<td>0</td>
<td>Blalock shunt (3)</td>
<td>0</td>
<td>AF</td>
<td>0</td>
<td>+</td>
<td>Inoperable</td>
</tr>
<tr>
<td>12</td>
<td>15</td>
<td>M</td>
<td>+</td>
<td>Waterston anastomosis (14)</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>+</td>
<td>Intracerebral haemorrhage</td>
</tr>
</tbody>
</table>

*This patient has subsequently had a Fontan operation. †Patients 8 and 9 also had subaortic stenosis. ‡Patient 11 also had mitral regurgitation. AF, atrial fibrillation; Ao, aorta; AT, atrial tachycardia; LPA, left pulmonary artery; LVEF, left ventricular ejection fraction; R, right; VPCs, ventricular premature complexes; VT, ventricular tachycardia.
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Table 2  Clinical data on five patients with tricuspid atresia and transposition of the great arteries with a Fontan procedure

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>Sex</th>
<th>Type of Fontan</th>
<th>Ability index</th>
<th>Arrhythmia (%)</th>
<th>Cause of death</th>
</tr>
</thead>
<tbody>
<tr>
<td>18</td>
<td>F</td>
<td>Direct RA appendage to PA</td>
<td>2</td>
<td>Bigeminy</td>
<td>26</td>
</tr>
<tr>
<td>19</td>
<td>M</td>
<td>Dacron tube + homograft RA to PA</td>
<td>VT, paced</td>
<td>Reoperation. Thrombus in conduit + PAs. Right PA stenotic beyond conduit</td>
<td>Bleeding at cardiac transplantation</td>
</tr>
<tr>
<td>21</td>
<td>M</td>
<td>Homografts RA to PA (x 2 = 2 outlets from RA. Inflow homograft IVC PAs remade with Dacron + homograft RA to PA)</td>
<td>15</td>
<td>18</td>
<td>26</td>
</tr>
<tr>
<td>15</td>
<td>M</td>
<td>Homografts RA to PA (x 2 = 2 outlets from RA. VSD enlarged)</td>
<td>15</td>
<td>18</td>
<td>26</td>
</tr>
</tbody>
</table>

Patient 14 also had severe subaortic stenosis and patient 17 also had moderate aortic stenosis.

Arrhythmias

Five of the 12 patients had arrhythmias. Atrial fibrillation caused symptomatic deterioration in two, which improved in one (case 1, who also had a transient stroke) after cardioversion. She has remained in sinus rhythm on quinidine for four years. In three patients asymptomatic arrhythmias were found on Holter monitoring: non-sustained multifocal atrial tachycardia (case 2), non-sustained ventricular tachycardia (case 4), and ventricular couplets (> 30/h) (case 8).

GROUP B: PATIENTS WITH A FONTAN OPERATION

Table 2 summarises the data on these five patients. All had pulmonary stenosis. Four had had a Blalock shunt in the first decade, and one (case 13) had had a pulmonary valvotomy and a Potts shunt. All patients had improvement in their clinical state after the shunt operation. A Fontan operation, with various modifications, had been performed at 15–21 years (table 2).

Mortality

There were four deaths, three in patients who were not ideal candidates for the procedure. Three who died at reoperation had not had symptomatic benefit from the Fontan operation. Patient 14, with small pulmonary arteries and severe subaortic stenosis, developed severe right-sided congestion and eventually his conduit and pulmonary arteries became thrombosed. He died at reoperation. Patient 15 had adequate preoperative left ventricular function, but heart block developed at operation and he required an epicardial pacemaker. He was well enough to return to work (ability index 2) but left ventricular function deteriorated. One year after the Fontan operation ventricular tachycardia developed. This was abolished by amiodarone. His ejection fraction fell to 10% and cardiac transplantation was performed, but he died from uncontrollable bleeding. Patient 16 had poor pulmonary artery development and despite refashioning of the pulmonary arteries with Dacron and a homograft right atrial pressure was high after the Fontan procedure and increased over the next three years. At reoperation the pulmonary arteries were again refashioned but the right atrial pressure remained high (20 mm Hg) and she died on the second postoperative day. A 15 year old boy died on the second day after a Fontan operation. Before operation he had moderate subaortic stenosis (gradient 40 mm Hg at rest) with a left ventricular end diastolic pressure of 20 mm Hg and a pulmonary artery pressure of 34/24 mm Hg.

Survivor: ability index and exercise capacity

One patient (case 13) was alive nine months after the Fontan operation. She had an ability index of 2, left...
ventricular ejection fraction was 26%, and Holter monitoring showed runs of bigeminy. She reached stage II of the exercise test and was stopped by dyspnoea.

Discussion

Tricuspid atresia and transposition of the great arteries is uncommon, and without early surgery the prognosis is usually poor because of hypoxia from low pulmonary blood flow in those with pulmonary stenosis or left ventricular failure in those with high pulmonary blood flow. In this series, however, two patients with pulmonary stenosis survived to 17 and 34 years without surgical treatment, and two patients with the Eisenmenger reaction survived to 22 and 26 years without surgical intervention. The proportion of patients with this anomaly who survive to adolescence is unknown. The patients we saw were referred to the National Heart Hospital because they had complications and because of our known interest in adult congenital heart disease.

Most patients (16/17 (94%)) had sufficient pulmonary blood flow to survive the first year without surgery, but 71% (12/17) needed a shunt or pulmonary valvotomy in the first decade. Perhaps those who require surgical treatment in infancy rarely survive to adolescence because they have severe pulmonary stenosis or atresia or a small ventricular septal defect.

Pulmonary vascular disease was common (4/17 patients) and was an important determinant of late mortality and morbidity. In three patients with no pulmonary stenosis (cases 4, 7, and 11) the Eisenmenger state developed early; two died before 22 years of age, and the survivor is severely disabled. In one patient (case 5) pulmonary hypertension developed over a decade after an ascending aorta to left pulmonary artery anastomosis had been constructed when she was 8; she now has an ability index of 3. When left ventricular failure developed in adults with tricuspid atresia, pulmonary congestion rapidly accelerated their deterioration, and they are unlikely to survive to the fourth decade.

The option of cardiopulmonary transplantation is more hazardous in those with previous operations because they are liable to have torrential bleeding from the chest wall during further operation. Many would argue that pulmonary artery banding should be performed in patients with heart failure in infancy to keep the pulmonary vascular resistance low, so that the Fontan procedure, considered by many to be the ultimate operation of choice for this condition, can be performed later. We are not convinced, however, that such patients with a Fontan operation performed at an early age would survive to their early twenties. Also early surgery in such patients virtually rules out subsequent successful cardiopulmonary transplantation. Furthermore, there is no doubt that in infants with heart failure, pulmonary artery banding accelerates the development of subaortic stenosis. This is why we perform pulmonary artery banding only when failure is uncontrollable or if the chest has to be opened for repair of a coarctation.

Subaortic stenosis is a serious acquired lesion, which when it is severe may cause angina (case 8). It may develop under the aorta in the subaortic "cus" or at the ventricular septal defect. Those who have subaortic stenosis at birth, or soon after, are unlikely to survive to the second decade. Four of the patients we describe had subaortic stenosis of varying degrees and two died after attempted relief of the obstruction. We believe that the outlook in patients with subaortic stenosis is poor despite the introduction of new operations. It should be assumed that sympathetic stimuli will increase the gradient. The effect of isoprenaline should be measured at the time of cardiac catheterisation to confirm this. If left ventricular function is sufficiently good we believe that β blockade may offer less hazardous short term symptomatic relief of subaortic stenosis than surgery.

Arrhythmias were common (7/17) and occurred irrespective of the type of operation and even when no operation had been performed. Thus arrhythmias appears to be part of the normal course, as they are in tricuspid atresia with normally related great arteries. Atrial arrhythmias occurred in three patients and ventricular arrhythmias in four. Neither arrhythmia was more common in those with impaired left ventricular function. Incessant atrial fibrillation, however, causes serious symptomatic deterioration and it is important to attempt early cardioversion. Medical treatment has maintained sinus rhythm in all survivors, but left ventricular function must be watched if negative inotropes are used. In one patient complete heart block developed after a Fontan operation at 21 years of age and he needed a pacemaker; the epicardial route was chosen.

Although impairment of left ventricular ejection fraction might be expected in patients with longstanding shunts, all survivors with shunts had normal ejection fractions, but their left ventricular cavities were large. Left ventricular ejection fraction did not correlate with exercise capacity, which was impaired in all patients, but despite this all patients without pulmonary vascular disease led nearly normal lives as judged by the ability index.

Should adolescents and adults with shunts or pulmonary valvotomy be referred for a Fontan oper-
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These results are depressing. We recognise that these patients were treated at a time of pioneering surgical efforts and that mistakes were made. This report illustrates the diversity of difficulties encountered in such patients. Even having learnt from our mistakes, we suspect that because of the problems inherent in the anomaly we will still not be able to prevent disabling illness in many adult survivors with tricuspid atresia and transposition of the great arteries.

Carole Warnes was supported by a grant from the British Heart Foundation.

References

Tricuspid atresia with transposition of the great arteries in adolescents and adults: current state and late complications.
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Br Heart J 1987 57: 543-547
doi: 10.1136/hrt.57.6.543

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