Hemodynamic Data and Angiographic Findings after Mustard Repair for Complete Transposition of the Great Arteries

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SUMMARY
Eleven children with uncomplicated transposition of the great arteries repaired by the Mustard procedure were studied by atrial pacing, cardiac catheterization, and cineangiography. Nine had persistent postoperative ectopic cardiac rhythms. Atrial pacing in five suggested sinus node damage in four and A-V junctional conduction tissue damage in one.

In seven children dye-dilution curves indicated small right-to-left shunts. In all seven superior vena cava cineangiograms showed leaks in the baffle at the superior atrial junction. In the other 10, sequential pressures in the superior vena cava, physiologic right atrium, and inferior vena cava showed no evidence of obstruction to systemic venous return. The three complications related to placement of the atrial baffle seemed to be due in large part to technical difficulties in suturing the baffle in the vulnerable sinus node region.

There were no differences in the postoperative hemodynamics between the five patients repaired at a younger age because of severe cyanosis and the five larger children who had been repaired electively. Our results support the aim for early surgical correction in uncomplicated transposition of the great arteries.

Additional Indexing Words:
Postoperative cardiac arrhythmias Atrial pacing Dye-dilution curves
Swan-Ganz catheter Superior vena cava obstruction Intraatrial baffle leaks

Since 1964 the Mustard procedure has become the preferred method for surgical correction of complete transposition of the great arteries.¹ ² Several centers have reported their results with this technic, but the long-term prognosis after total repair is not yet established.³⁻¹⁰ Walldhausen and associates reported some of the clinical findings in their patients followed up to 9 months after repair.⁸ Tynan and associates recently reported the cardiac catheterization findings in 18 patients with postoperative complications, including tricuspid valve insufficiency, pulmonary venous obstruction, and superior vena cava

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Completed while Dr. Rodriguez-Fernandez was a Research Fellow of the Maryland Heart Association.

Supported in part by U. S. Public Health Service Grant 5R01 HE107718, the Maternal and Child Health Service Project Grant 12 H201, and Research Career Development Award 5KO-HE-09761-05.

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Received March 7, 1972; revision accepted for publication May 10, 1972.
obstruction.\textsuperscript{9, 10}

Between 1965 and 1971, 12 children with uncomplicated complete transposition of the great arteries survived open-heart repair with the Mustard procedure at The Johns Hopkins Hospital. The early surgical results in eight of these patients were previously reported.\textsuperscript{5} Our study is concerned with the postoperative hemodynamic data and the electrocardiographic and angiographic findings in 11 of the 12 survivors. The parents of the 12th one refused permission for postoperative cardiac catheterization.

\textbf{Material and Methods}

\textbf{Material}

The nine boys and two girls ranged in age at the time of postoperative evaluation from 2.7 to 10.8 years (mean 6.3 years). At preoperative cardiac catheterization the 11 children were cyanotic with hematocrits ranging from 54 to 76\% (mean 67\%) and systemic oxygen saturations ranging from 52 to 80\% (mean 68\%) (table 1). All were on maintenance digoxin. The chest X-rays showed increased pulmonary vascularity and cardiomegaly, and the electrocardiograms consistently showed sinus rhythm, right-axis deviation, and right ventricular hypotrophy.

Six patients (cases 1–6) were repaired electively between 3 and 7 years of age when they weighed 12.0 kg or over. Five (cases 7–11) were repaired at a younger age because of severe cyanosis and rising hematocrit. The second group was between 2 and 3 years old and weighed 9.6–12.0 kg at the time of surgical repair.

In six of the 11 patients pericardium was used to fashion the intraatrial baffle. In the five more recent operations the baffle was constructed of knitted Dacron fabric as previously reported by Danielson and associates\textsuperscript{7} (fig. 1). This material prevents the possibility of postoperative pericardial shrinkage producing venous obstruction,\textsuperscript{10} and avoids the problem of using pericardium in patients with post-Blalock-Hanlon pericardial adhesions.\textsuperscript{6}

In six operations a U-shaped atriotomy incision was performed which turned an atrial flap, based on the A-V groove. In the five more recent procedures the atriotomy was modified to an oblique incision extending from the tip of the right atrial appendage to the right atrium-inferior vena cava junction. This incision may reduce the likelihood of damage to the sinus node, and facilitates the use of a patch to enlarge the atrium in small infants.

\textbf{Methods}

Postoperative cardiac catheterization was performed in the 11 children by the percutaneous technic through a femoral vein.\textsuperscript{11} A femoral artery was also entered percutaneously with a no. 18 Longdwell Teflon needle. The venous catheter was advanced from the inferior vena cava into the physiologic right atrium, and from the atrium into the superior vena cava and left ventricle, in all patients except one (case 6). The pulmonary artery was entered from the left ventricle in the 10 children with a flow-guided Swan-Ganz balloon catheter by means of a previously described technic.\textsuperscript{12}

In 11 patients 2.5 mg of indocyanine green dye was injected in the superior and inferior vena cavae, and dye-dilution curves were obtained by collecting samples from the femoral artery through a Waters X-250 densitometer. In 10 children dye was injected in the pulmonary artery, and samples were collected from the femoral artery with the same technic.

Superior and inferior vena cava cineangiograms in the posteroanterior position were done in 10 patients, and oblique left ventricular cineangiograms in three. Intracardiac electrograms were recorded in nine, and atrial pacing was performed in five instances.

\textbf{Results}

Atrial pacing in four patients (cases 2–5) with varied postoperative ectopic cardiac rhythms, including two with His junctional rhythm\textsuperscript{13} and retrograde conduction (cases 2 and 5), one with intermittent A-V dissociation

\textbf{Figure 1}

\textit{Placement of the Dacron baffle at operation. Arrows indicate the hemodynamic pathways.}

\textit{Circulation, Volume XLVI, October 1972}
<table>
<thead>
<tr>
<th>No.</th>
<th>Septostomy Type</th>
<th>Age (yr)</th>
<th>Wt (kg)</th>
<th>%</th>
<th>RA mean</th>
<th>LA mean</th>
<th>LV</th>
<th>PA</th>
<th>Syst vent</th>
<th>Syst O₂ sat (%)</th>
<th>Angiograms</th>
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<td>2 mo</td>
<td>3.5</td>
<td>3</td>
<td>6</td>
<td>6</td>
<td>58</td>
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<td>RV: ductus not patent</td>
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</tbody>
</table>

Abbreviations: BH = Blalock-Hanlon; BAS = balloon atrial septostomy; RA = right atrial; LA = left atrial; PA = pulmonary artery; NE = not entered; RV = right ventricle; LV = left ventricle; AR = aortic root; VSD = ventricular septal defect.
Electrographic, Hemodynamic, and Angiographic Findings after Mustard Repair

Table 2

<table>
<thead>
<tr>
<th>Time since repair (yr)</th>
<th>Postop cath</th>
<th>Chest X-ray</th>
<th>Cardiac rhythm</th>
<th>Atrial pacing</th>
<th>Mean pressures (mm Hg)</th>
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<tbody>
<tr>
<td>Age (yr)</td>
<td>Wt (kg)</td>
<td>%</td>
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<td></td>
<td>IVC        RA    SVC    PCW</td>
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<tr>
<td>104-04-95</td>
<td>5.4</td>
<td>8.9</td>
<td>25.7</td>
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<td>0.48       N     Int AVD</td>
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<td>8.9</td>
<td>26.4</td>
<td>25</td>
<td>0.48       N     Int AVD</td>
</tr>
<tr>
<td>100-03-26</td>
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<td>9.8</td>
<td>33.1</td>
<td>75</td>
<td>0.49       N     Int AVD</td>
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<tr>
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<td>3.0</td>
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<td>21.2</td>
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<td>0.57       N     HJR  1:1 A-V cond</td>
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<td>102-06-35</td>
<td>0.9</td>
<td>4.9</td>
<td>13.7</td>
<td>&lt;3</td>
<td>0.52       Incr CSR</td>
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<tr>
<td>21-01-55</td>
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<td>5.8</td>
<td>17.5</td>
<td>10</td>
<td>0.64       N     Int HJR</td>
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<td>120-05-15</td>
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<td>15.8</td>
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<td>0.55       Incr NSR</td>
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<td>0.7</td>
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<td>0.66       Incr CHB</td>
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<td>126-07-95</td>
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<td>0.63       Incr CSR</td>
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<td>137-08-94</td>
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<td>2.7</td>
<td>13.0</td>
<td>10</td>
<td>0.63       Incr AF</td>
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<td>130-03-01</td>
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</table>

Abbreviations: CTR = cardiothoracic ratio; PV = pulmonary vascularity; N = normal; Incr = increased; Int = intermittent; AVD = atrioventricular dissociation; HJR = His junctional rhythm; CSR = coronary sinus rhythm; NSR = normal sinus rhythm; CHB = complete heart block; AF = atrial flutter; SVC = superior vena cava; RA = physiologic right atrium; IVC = inferior vena cava; PCW = pulmonary capillary wedge; NE = not entered; PA = pulmonary artery; SA = systemic artery; + = present; 0 = absent; — = not measured; LV = left ventricle; A-V cond = atrioventricular conduction.

(case 3), and one with coronary sinus rhythm (case 4), succeeded in capturing the ventricle with 1:1 conduction (table 2). In the patient with complete heart block (case 9), atrial pacing failed to capture the ventricle. Sequential mean pressures from superior vena cava to atrium, to inferior vena cava, in the 10 patients in whom the physiologic right atrium was entered, did not show a significant difference, indicating that there was no systemic venous obstruction. In addition, all 10 had normal bilateral pulmonary capillary wedge pressures suggesting that none had pulmonary venous obstruction or tricuspid valve regurgitation. Obstructive pulmonary vascular disease was not present since they all had normal pulmonary artery pressures.

In seven patients there was no evidence of obstruction across the pulmonary outflow tract. In the other three (cases 5, 7, and 10) the systolic pressure difference across the outflow tract ranged from 20 to 38 mm Hg. Left ventricular cineangiograms either in the right anterior oblique or in the left anterior oblique position showed mild thickening of the pulmonary valve in two (cases 5 and 10) and narrowing of the subvalvar outflow tract in one (case 7).

Dye-dilution curves injecting in the superior vena cava and sampling in the femoral artery in 11 patients indicated small right-to-left shunts in seven. Two of the 10 children (cases 4 and 11) in whom dye-dilution curves were done, injecting in the pulmonary artery and sampling in the femoral artery, showed small left-to-right shunts not detected by oximetry.
Superior and inferior vena cavae cineangiograms in the posteroanterior position provided a good view of the contrast flowing inside the baffle and defined its anatomic relationship to the vena cavae and mitral valve (figs. 2, 3). In the seven patients with small right-to-left shunts by dye curve, contrast material was seen to leak through the baffle into the physiologic left atrium at the superior vena cava-atrial junction (fig. 2, bottom).

**Discussion**

Postoperative evaluation carried out 7 months to 5 years (mean 2.4 years) after the Mustard procedure showed that all 11 patients had improved clinically. They were asymptomatic, acyanotic, and only two (cases 5 and 6) had not increased their weight percentiles. All had a grade II/VI systolic ejection murmur at the upper left sternal border, and one (case 4) had in addition a grade II/VI holosystolic murmur at the lower left sternal border. He was shown to have a small ventricular septal defect at postoperative catheterization, by passage of a retrograde catheter from the right systemic ventricle into the left pulmonic ventricle.

Residual cardiomegaly was present in the chest X-rays of nine of the 11 patients, and it did not seem to be related to the length of time after surgical repair. Increased pulmonary vascularity was more obvious in the five patients who had been repaired more recently.

Waldhausen and associates reported persistent postoperative arrhythmias in seven of their 18 children with uncomplicated transposition of the great arteries.9 Nine of our 11 patients had persistent postoperative ectopic cardiac rhythms. Apart from one child with complete heart block and another with atrial flutter and variable A-V block, the other seven patients with ectopic rhythms had different types of escape junctional rhythms, with predominance of His junctional pacemakers. In four of the seven (cases 2–5) 1:1 ventricular conduction was easily obtained by atrial pacing at different rates and from several
Cineangiograms after injection of contrast material in the superior vena cava (SVC) in the posteroanterior projection. Arrows indicate contrast material streaming through the baffle into the left ventricle (LV). (Top) Dye-dilution curve (inset bottom left) from SVC to femoral artery shows no evidence of right-to-left shunt. Angiogram shows no leak of contrast material across the baffle. (Bottom) Dye-dilution curve (inset bottom left) has an early appearance hump indicating a right-to-left shunt. Angiogram confirms the presence of a leak (L) across the baffle at the SVC-atrial junction.
FINDINGS AFTER MUSTARD REPAIR

Figure 3

Cineangiogram after injection of contrast material in the inferior vena cava (IVC) in the posteroanterior projection, showing the inferior contour of the baffle and the residual interatrial septum (S). Contrast material flows freely into the left ventricle (LV), and there is no leak across the baffle.

atrial sites, including the sinus node region. These results suggested that in the patients with escape junctional rhythms without heart block the most likely site of surgical damage was the sinus node itself, rather than the A-V junctional tissue or the internodal pathways. The child with complete heart block (case 9), on the other hand, suffered significant damage to the A-V junctional tissue, as demonstrated by the failure of atrial pacing to capture the ventricle. The two patients in sinus rhythm (cases 6 and 8) were among the more recent repairs, in whom an oblique atriotomy incision from the tip of the right appendage to the inferior vena cava-atrial junction was used, to avoid damage to the sinus node.

Persistent ectopic rhythms have been reported as common after the Blalock-Hanlon operation. Seven of our 11 patients were subjected to Blalock-Hanlon septostomies in infancy, but all were in sinus rhythm prior to the Mustard repair.

The three patients with mild postoperative left ventricular outflow obstruction (cases 5, 7, and 10) had elevated left ventricular pressures at preoperative catheterization. One (case 7) had a normal left ventricular angiogram preoperatively, and subvalvar stenosis in the postoperative study, suggesting that the obstruction had progressed since the preoperative catheterization. The mild thickening of the pulmonic valve in the other two children (cases 5 and 10) looked similar in the preoperative and postoperative left ventricular angiograms, suggesting that the valvar obstruction was not progressive.

The seven children with small right-to-left shunts and leaks at the superior vena cava-atrial junction had systemic oxygen saturations that ranged from 90 to 94%. The slight systemic desaturation noted by Waldhausen and associates in some of their patients may be due to similar leaks, rather than thebesian venous drainage, as suggested by Rashkind.
Figure 4

Cineangiogram in the posteroanterior projection in a patient (case 6) with postoperative complete obstruction of the superior vena cava (SVC). (Top) Injection of contrast material in the SVC shows the obstruction at the SVC-atrial junction. (Bottom) From the SVC contrast material enters the azygos vein (AV) and finally drains into the right atrium via the inferior vena cava (IVC).
One of the two patients (case 4) with small left-to-right shunts had a small ventricular septal defect. The other one (case 11) did not have a murmur suggestive of a ventricular septal defect, and preoperative left ventricular angiography and aortography had not detected a ventricular septal defect or a patent ductus. The cause of the left-to-right shunt in this case remains undetermined, but it is possible that a pulmonary vein was sutured to drain inside the baffle producing a partial anomalous venous return.6

One patient (case 6) had partial thrombotic occlusion of the inferior vena cava, and complete obstruction of the superior vena cava at the vena cava-atrial junction, that prevented entry of a catheter within the heart. A cineangiogram in the superior vena cava showed it to drain into the upper inferior vena cava via theazygous vein (fig. 4). Stark and associates reported superior vena cava obstruction in four of their 113 Mustard repair survivors and attributed it to pericardial shrinkage of the baffle.10 Our patient had a Dacron baffle, so we could not consider pericardial shrinkage as the cause of the obstruction. The inferior vena cava thrombotic occlusion probably resulted from trauma to the vessel wall at the preoperative catheterization or at the surgical repair itself.17

The three complications related to the placing of the intraatrial baffle in the 11 patients included cardiac arrhythmias in nine, leaks through the baffle at the superior vena cava-atrial junction in seven, and complete superior vena cava obstruction in one. All three seemed to be related in large part to the technical difficulties of suturing the baffle in the vulnerable sinus node region.

Five children (cases 7–11) were repaired at a younger age (mean age 2.3 years) and smaller size (mean weight 10.6 kg) because of severe cyanosis and a rising hematocrit. Preoperatively, all five had an adequate intratral communication and no evidence of significant left ventricular outflow obstruction or obstructive pulmonary vascular disease. The explanation for their early deterioration remains speculative, but our experience supports the current concept that early corrective surgery is necessary to avoid the complications accompanying severe cyanosis and polycythemia.8 Earlier correction does not seem to reduce the effectiveness of surgical repair, since there were no differences in the postoperative hemodynamics between the earlier and later repair groups.

References
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Circulation. 1972;46:799-808
doi: 10.1161/01.CIR.46.4.799

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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