Hemodynamic Studies in Children Four to Ten Years after the Mustard Operation for Transposition of the Great Arteries


SUMMARY
Fourteen patients have been studied hemodynamically 4-10 years (mean 5.5 years) after the Mustard operation for transposition of the great arteries. Investigation was directed principally at 1) the detection of baffle obstruction by catheterization of the pulmonary veins (PV) and venae cavae and recording of synchronous right ventricular (RV) and pulmonary capillary wedge pressures (PCW); 2) the detection of intra-atrial baffle leaks by dye curves and selective angiography; 3) the assessment of RV and LV function by calculating peak VCE (dp/dt/28p) from high fidelity recordings in 11 patients. Severe baffle obstruction to the PV return was found in only one patient. The others had no or minimal gradients between RV end-diastolic and PCW pressures (mean 1.3 ± 0.69 mm Hg). Cardiac output was normal at rest (4.1 ± .22) and increased to 7.1 ± .62 L/min/m² (+73%) but the gradient between the RV end-diastolic and PCW pressures remained insignificant (2.2 ± 1.13 mm Hg). No evidence of caval obstruction was found in any patient. Baffle leaks were found in five patients with mild bidirectional shunting. All arterial oxygen saturations were above 90%. Mild tricuspid regurgitation was demonstrated in two patients by RV angiography and was absent in 12 others. The contractility index peak VCE averaged 1.87 ± .122 sec⁻¹ for the RV and was significantly lower in the LV (1.53 ± 1.35 sec⁻¹, P < 0.01). Only one patient presented significantly decreased RV contractility with a peak VCE of 1.07 sec⁻¹ and poor contraction on the RV angiogram. These data indicate that the long-term prognosis after the Mustard operation should be good in most patients and that the right ventricle is capable of functioning at the level of contractility of a normal left ventricle.

THE PROGNOSIS FOR PATIENTS with complete D-transposition of the great arteries (TGA) has improved markedly since the introduction and acceptance of balloon septostomy and the intra-atrial baffle operation. The intra-atrial baffle redirects venous return to the heart, producing functional but not anatomical correction of the defect. Clinical studies of early and late postoperative survival are well documented and have shown the modification of the natural history of TGA, which may follow a successful intra-atrial baffle operation. A previous report of the clinical course of long-term survivors followed at the Hospital for Sick Children, Toronto for up to eight-and-a-half years postoperatively, suggested that 73 of 80 long-term survivors were leading a normal life. Reported hemodynamic studies, however, have been few in number and have principally involved patients who were in the early postoperative period when investigated or who had postoperative complications requiring hemodynamic assessment.

In this paper we report the results of hemodynamic and angiographic studies in 14 children four to ten years after the intra-atrial baffle operation for TGA.

Patient Material and Methods

The 14 children represented all patients at the Sick Childrens Hospital, Toronto who had a complete right and left heart catheterization study at an interval greater than four-and-a-half years following the intra-atrial baffle repair.
operation. Their clinical course paralleled that of other long-
term survivors reported previously. The ages of the 14 children at the time of the intra-atrial baffle repair ranged from 18 months to 4⅞ years. The interval
between surgery and investigation ranged from 4.4 to 10
years with a mean of 5.5 years. Thirteen of the children had
TGA alone and one child had TGA associated with an inter-
ventricular septal defect. The preoperative clinical and
hemodynamic data are summarized in table 1. Eleven of the
14 children had had Blalock Hanlon procedures, and two
had had Sterling Edwards procedures. The pulmonary
artery pressure was measured in only two preoperatively.
The majority of these patients had been investigated pre-
operatively and treated palliatively before Rashkind balloon
septostomy was an accepted procedure and before current
techniques for entering the pulmonary artery had been
refined. Pericardium was used in all cases for construction of
the atrial baffle. The operative techniques employed have
been reported previously.

**Procedure**

All the subjects were lightly sedated with a standard
premedication containing demerol, phenergan and chlor-
promazine.

In 11 of the 14 children venous catheterization was
performed by the percutaneous technique through the femoral
vein. In the remaining three children a catheter was passed from a right sided antecubital vein in two cases and a left
sided antecubital vein in one. The venous catheter was passed
to the systemic venous atrium and then to the left
ventricle. Systemic venous obstruction was assessed by
pressure recordings as the catheter was withdrawn from the
right atrium to the venae cavae or vice versa. In all 14
patients the main pulmonary artery (MPA) was entered
from the left ventricle with either an end hole catheter and
guide wire or a flow guided Swan-Ganz balloon catheter and
pulmonary artery wedge pressures were obtained.

In all 14 cases a femoral artery was catheterized per-
cutaneously and an end hole or angio catheter passed retro-
gradely to the ascending aorta, right ventricle and across the
tricuspid valve to the new pulmonary venous atrium. In all 14
cases the right and left pulmonary veins were probed and
pressure recordings obtained as the catheter was withdrawn
from the pulmonary veins to the right ventricle. Simultaneous pressure recordings were made of 1) the
pulmonary venous atrial and systemic venous atrial pressures,
and 2) the pulmonary capillary wedge and right
ventricular end-diastolic pressures.

All pressures were measured by Statham strain gauge
transducers placed at a level one-third of the chest thickness
below the sternal angle.

To detect any residual shunt, measurements of O₂ content
were made in all chambers by a Waters cuvette oximeter and
in 12 patients indocyanine green dye was injected into the
superior and inferior venae cavae and dye curves recorded
from the femoral artery.

Cardiac output measurements were made by indicator
dilution technique, using indocyanine green dye, a Harvard
constant withdrawal pump and a Waters 250 densitometer.
The dye was injected into the MPA and sampled from the
aorta.

Observations of pressures and cardiac output were
repeated following intravenous infusion of isoproterenol.
The rate of infusion of isoproterenol was adjusted to give an
increase in heart rate of at least 50%. Cardiac output deter-
minations were repeated at what was judged to be the
sustained peak effect of the drug and simultaneous
pulmonary venous capillary and RV end-diastolic pressures
were recorded.

Right ventricular and left ventricular function was
assessed by calculating the peak velocity of the contractile
element (peak VCE); this was obtained from high fidelity
pressure tracings using a transducer-tipped catheter
(Statham p86 No. 5F). The tracings were analyzed on an
X-Y digitizing table and the value dp/dt/28p (from total
pressure) calculated every 5 msec during the isovolumic
phase for three consecutive contractions. An average peak
value for VCE was then obtained from the plot dp/dt/28p
against pressure.

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**Table 1. Preoperative Hemodynamics and Clinical Data**

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<th>RA (mean)</th>
<th>LA (mean)</th>
<th>LV</th>
<th>RV</th>
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<th>SaO₂ (%)</th>
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Abbreviations: LA = left atrium; RA = right atrium; LV = left ventricle; RV = right ventricle; MPA = main pulmonary artery;
SaO₂ = systemic arterial oxygen saturation; BH = Blalock Hanlon; SE = Sterling Edwards; NE = not entered.
Table 2. Postoperative Hemodynamic Data

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Abbreviations: SVC = superior vena cava; IVC = inferior vena cava; PVA = pulmonary venous atrium; SVC = systemic venous atrium; RV = right ventricle; SR = sinus rhythm; LV = left ventricle; MPA = main pulmonary artery; FV = pulmonary vein; SaO₂ = systemic arterial oxygen saturation; T1 = tricuspid incompetence.
Results

The hemodynamic findings in the 14 children at the time of postoperative study are shown in table 2.

Intra-atrial Baffle Function

There was no evidence of systemic venous obstruction in any of the 14 patients. The pressure difference between the venae cavae and the new systemic venous atrium did not exceed 2 mm Hg in any child. In two patients superior vena caval angiography suggested slight narrowing at the superior vena cava-atrial junction but no pressure difference was found in either case (fig. 1).

In seven patients the mean pulmonary venous atrial and systemic venous atrial pressures were identical; five of the seven had small intra-atrial baffle defects. In two of the seven patients the pulmonary and systemic venous atrial a and v waves were identical; in the remaining five the a wave or a cv wave was greater in the pulmonary venous atrium than in the systemic venous atrium.

Only one patient had evidence of pulmonary venous obstruction (C.B.). This boy was first studied seven years postoperatively. He had been suspected on clinical grounds of having pulmonary hypertension and this diagnosis was confirmed at cardiac catheterization. However, the diagnosis of pulmonary venous obstruction had not been made because neither a pulmonary venous nor reliable pulmonary artery wedge pressure was obtained. At restudy, nine years postoperatively, the pulmonary artery wedge pressure was elevated (table 2). An end-hole catheter was passed retrogradely across the tricuspid valve into the pulmonary venous atrium, which appeared to be divided into two chambers. The catheter passed into the pulmonary veins and a withdrawal recording demonstrated a high pressure chamber proximally communicating via a small orifice with a low pressure chamber distally (fig. 2). An angiogram was performed with the catheter positioned at the orifice of the left upper pulmonary vein and demonstrated a diaphragm-like structure to be the site of obstruction within the pulmonary venous atrium (fig. 3). In spite of the severe baffle obstruction demonstrated at catheterization, this boy only became dyspneic on severe exertion and clinically was judged to have only mild impairment of his effort tolerance.

Another patient (M.S.) had pulmonary hypertension when restudied five years postoperatively but no evidence of pulmonary venous obstruction. The left ventricular pressure preoperatively was 32/2 mmHg and there was no evidence angiographically of a ventricular septal defect. The interval between his preoperative study and operation was 18 months.

Seven of the 14 children had intra-atrial baffle defects

FIGURE 1. Superior vena caval angiogram demonstrating narrowing at the superior vena caval-atrial junction in the absence of a pressure difference. The uppermost arrow indicates an intra-atrial baffle defect.

FIGURE 2. Pressure record obtained within the new pulmonary venous atrium as the catheter is withdrawn through the narrow orifice produced by intra-atrial baffle obstruction.

FIGURE 3. Angiogram with a catheter positioned just below the orifice of the left upper pulmonary vein demonstrating that a diaphragm-like structure appears to be the site of obstruction within the pulmonary venous atrium.
demonstrated by angiography (fig. 1) or dye curves. None was cyanotic and none had an arterial saturation below 90.

The most common site of deficiency in the atrial baffle was at the superior vena cava-atrial junction and this was so in four of the seven cases. One child with angiographic evidence of a baffle leak had no evidence of intra-cardiac shunting, as determined by oximetry or dye curves; mild bi-directional shunting was present in the other six children (table 2).

Tricuspid Regurgitation

Two patients had a trivial degree of tricuspid regurgitation. Both had an intact ventricular septum when investigated preoperatively and tricuspid regurgitation was not noted at that time. Neither was in sinus rhythm postoperatively.

Main Pulmonary Artery — Left Ventricular Pressure Differences

Three patients had systolic pressure differences of greater than 20 mm Hg between the main pulmonary artery and the left ventricle. In one patient (G.T.) studied in the early postoperative period, the left ventricular pressure was 49/0 mm Hg; this increased to 80/6 mm Hg at restudy eight years postoperatively and was associated with a pressure difference of 60 mm Hg between the left ventricle and the main pulmonary artery.

Cardiac Output

The cardiac index at rest was measured in ten of the 14 patients and ranged from 3.2 to 5.1 L/min/m². The cardiac index was measured in the same subjects following administration of isoproterenol. The mean cardiac index for the group rose from 4.1 L/min/m² to 7.9 L/min/m². The increase in cardiac output usually resulted in a small diastolic pressure difference around the baffle but this did not exceed 5 mm Hg in any patient (mean 2.2 ± 1.13).

Contractility

For the right ventricle peak VCE averaged 1.87 ± .122 sec⁻¹. Only one child had a significant decrease in the right ventricular peak VCE with a value of 1.07 sec⁻¹; poor performance was also observed on the right ventricular angiogram in this child. Right ventricular angiography in the other ten children suggested a good performance in seven and a slight decrease in performance in two cases; in one case multiple premature ventricular contractions made assessment of performance impossible. For the left ventricle there was a somewhat larger scatter but the mean value for peak VCE was 1.53 ± .135 sec⁻¹ and was significantly lower than for the right ventricle (P < 0.01) (fig. 4).

Discussion

Although the intra-atrial baffle operation may result in a striking clinical improvement of the patient’s condition, concern has existed about the long-term fate and function of the pericardial baffle and the ability of the right ventricle and tricuspid valve to operate at a systemic workload. Complications relating to the placement of the pericardial baffle are now well recognized. The incidence of isolated superior vena caval obstruction is probably low and in our experience this complication does not usually require treatment. Stark et al. have emphasized the importance of preserving the aygous system at palliative surgery since this probably provides an adequate collateral circulation. When superior vena caval obstruction is sufficiently severe to require surgery it usually presents as an early problem within the first year after operation. None of the 14 patients investigated in the present study had any evidence of superior vena caval obstruction up to ten years postoperatively. Pulmonary venous obstruction due to the atrial baffle is a much more serious complication and is almost invariably associated with severe symptoms. Like superior vena caval obstruction it has usually been a complication recognized fairly early after surgery and in most reported experience has occurred within six months of operation. One of the 14 patients in this study had evidence of severe pulmonary venous obstruction when studied nine years postoperatively. An earlier study, performed seven years postoperatively, documented the presence of pulmonary artery hypertension but a reliable pulmonary artery wedge pressure was not obtained nor were the pulmonary veins entered. This case emphasizes the importance of attempting to selectively catheterize the pulmonary veins in any patient with unexplained pulmonary hypertension postoperatively. In view of the boy’s relative...
well-being throughout the postoperative period, it is difficult to speculate as to when the pulmonary venous obstruction developed.

The mechanisms responsible for baffle obstruction have not yet been well delineated. Mohri et al. experimentally confirmed graft shrinkage and thought it likely that some degree of obstruction of the patch occurred in all patients after the intra-atrial baffle operation.13 They also demonstrated, however, the growth potential of adjacent atrial tissue without any deleterious effects on the growth and function of the heart. The evidence of the present study suggests that graft shrinkage was not a problem and perhaps supports the concept of the growth potential of adjacent atrial tissue. Graft shrinkage has been the most commonly postulated mechanism for the production of baffle obstruction, but we believe that the stenosis could also result from adhesions between the pericardial baffle and adjacent right lateral atrial wall, producing a posterior and anterior pulmonary venous chamber as seen in fig. 3. The experience of several reports that baffle obstruction is more likely to occur in infancy may well reflect the greater importance in this group of patients who have smaller atria of tailoring the baffle precisely and paying careful attention to its geometry. Our experience with obstructive problems from the intra-atrial baffle has been reported previously and suggests that this should be an uncommon clinical problem.6 The present study reinforces our clinical impression that problems associated with shrinkage of the baffle are unlikely beyond one year postoperatively.

Like Rodriguez-Fernandez et al. we found that baffle leaks were common and these occurred in seven of our 14 patients.10 These leaks were not associated with any significant degree of intracardiac shunting in any patient and in particular no patient was cyanotic because of a baffle leak. The most common site of baffle detachment was at the superior vena cava and atrial junction, but in no patient was there any evidence of associated superior vena caval obstruction.

Waldhausen et al. reported tricuspid regurgitation after the intra-atrial baffle operation6 and Tynan et al. found tricuspid regurgitation in 17 of 21 survivors studied up to six years postoperatively.9 In our experience this has been a rare complication6 and in this study only two cases of mild tricuspid regurgitation were noted. In contrast to the patients reported by Tynan, 13 of the 14 patients studied by us were transpositions with an intact ventricular septum. Nine of the patients studied by Tynan had a ventricular septal defect preoperatively. One may therefore suspect a link between closure of a ventricular septal defect and tricuspid regurgitation. Tynan, however, found that there was no significant difference in the frequency of tricuspid regurgitation in his patients with an intact ventricular septum; the two patients with mild tricuspid regurgitation in this report also had an intact ventricular septum. Tynan suggested that pre-existing tricuspid regurgitation and postoperative arrhythmias were the main determinants of the postoperative tricuspid regurgitation; neither of the two patients in our study with tricuspid regurgitation was in sinus rhythm when investigated postoperatively. The mean age of Tynan's patients, both at operation and restudy, was considerably greater than the mean age of the patients in this study, and it may be that deterioration in right ventricular function and tricuspid valve competence with age prior to operation accounts for some of the difference.

Only three patients in this study had a systolic pressure difference greater than 20 mm Hg between the left ventricle and the main pulmonary artery. In one of these patients it appears possible that the degree of subvalvar obstruction had increased postoperatively since the left ventricular pressure in the early postoperative period was 49 mm Hg compared with 80 mm Hg at the time of restudy. Rodriguez-Fernandez et al. reported a case of left ventricular subvalvar stenosis which appeared progressive in the postoperative period. This complication, however, seems to be very rare.16

We believe that the present study also provides some objective evidence that the long-term functional capacity of the heart, following the intra-atrial baffle operation, is well maintained, despite the presence of arrhythmias and small residual defects in the atrial baffle. The pulmonary venous atrial pressures and right ventricular end-diastolic pressures were normal at rest, and were not significantly altered after isoproterenol infusion. Despite a substantial rise in cardiac index following isoproterenol from a mean of 4.1 ± .22 to 7.1 ± .62 L/min/m², the gradient between the right ventricular end-diastolic and pulmonary capillary wedge pressures remained insignificant (2.2 ± 1.13 mm Hg).

Right ventricular function assessed by the contractility index peak VCE was within normal limits for all but one patient. This seems to be at variance with the findings of Jar- makani and Canent who, using angiographic techniques to measure the ejection fraction (EF), found depressed RV function in all their patients with TGA before as well as after operation;17 similar results have been reported by Graham et al.19 Although a good correlation exists between peak VCE and EF, these two parameters measure different characteristics of ventricular contraction.16, 17 EF is concerned with the “pump” function of the heart and is measured during the ejection period while peak VCE measures the level of contractile state from pressure recordings made during the isovolumic period. Krayenbuehl et al. have shown that EF depends not only upon myocardial contractility, but also upon physical determinants of ventricular contraction. Changes in the ejection fraction and alterations of the contractile state can occur independently of each other and in opposite directions.19 Thus concordance of results is not necessarily to be expected.

We are aware that no entirely satisfactory measurement of ventricular function exists at present, and that theoretical objections have been expressed about peak VCE and Vmax as measurements of the contractile state.19, 20 It is still believed, however, that these parameters are among the most sensitive in assessing ventricular function and they have proven useful in assessing the ventricular function in various types of congenital heart disease.16, 17, 21 We did not try to calculate ejection fraction in our cases as we found it difficult to evaluate right ventricular volumes in the heavily trabeculated RV typically seen in TGA. The values for EF found by others in TGA are very low indeed, comparable only to those seen in diffuse myocardial disease such as myocarditis or cardiomyopathy and other signs of ventricular failure might have been expected.14-16 Most of our postoperative patients had
normal physical activities, their cardiac output and RV end-
diastolic pressures were normal and none required digitalis. We therefore feel that in our patients the clinical status as well as the hemodynamic findings are in agreement with the good RV function measured by peak VCE.

Although the right ventricle in transposition patients pre and postoperatively appears to function at the same peak VCE, this does not seem to be the case for the left ventricle. The contractility index for the left ventricle in a group of preoperative transposition patients reported previously from this laboratory was significantly higher than the peak VCE for the left ventricle in the patients reported in this study. One may speculate that the left ventricle in transposition of the great arteries goes through a slow process of involution, characterized not only by changes in wall thickness, as shown by Pasternac et al., but also by change in its contractile properties. Indeed, in the patients in this study the peak VCE for the left ventricle was close to the index found for the right ventricle in a group of children of a similar age without transposition. Thus both ventricles seem to adapt to their reversed function, the RV from an early age, the LV with the passage of time.

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_Circulation_. 1976;53:532-538
doi: 10.1161/01.CIR.53.3.532

_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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