Results of the Arterial Switch Operation in Patients With Transposition of the Great Arteries and Abnormalities of the Mitral Valve or Left Ventricular Outflow Tract

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Between January 1983 and October 1989, 290 patients underwent an arterial switch operation for transposition of the great arteries; 30 (10.3%) of the patients had abnormalities of the left ventricular outflow tract or mitral valve, or both. These abnormalities included isolated pulmonary valve stenosis (n = 9), septal (dynamic) subpulmonary stenosis (n = 5), anatomic (fixed) subpulmonary stenosis (n = 7), abnormal mitral chordae attachments (n = 2) or a combination of abnormalities (n = 7). There were two early deaths, one of which was due to previously unrecognized mitral stenosis and a subpulmonary (neo-aortic) membrane and one late death due to presumed coronary obstruction.

Of the nine patients with pulmonary valve abnormalities due to either a bicommissural (n = 5) or a thickened tricuspid valve, only one underwent valvotomy. Peak systolic ejection gradients in these nine patients measured preoperatively ranged from 0 to 50 mm Hg. At follow-up study 5 to 30 months postoperatively, the neo-aortic valve gradient was ≤15 mm Hg in all patients; three patients had mild neo-aortic regurgitation. Preoperative gradients may overestimate the degree of obstruction because of the increased pulmonary blood flow present in transposition.

No patient with "dynamic" subpulmonary obstruction before the arterial switch operation had a surgical procedure performed on the left ventricular outflow tract; none had evidence of subaortic obstruction after the arterial switch. Likewise, no patient with anatomic subpulmonary obstruction due to accessory valve tissue (n = 5) or a subpulmonary membrane (n = 2) had a residual left ventricular outflow gradient or abnormal antroventricular valve function after the arterial switch operation with tissue or membrane resection. Isolated mitral valve abnormalities were rare, being limited to two children with abnormal chordal attachments. The only patients with significant residual left ventricular outflow obstruction were three children with a combination of abnormalities that included obstruction due to posterior deviation of the infundibular septum.

These data indicate that mild pulmonary valve abnormalities, dynamic or surgically remediable subpulmonary stenosis or abnormal mitral valve attachments do not preclude a successful arterial switch operation; however, left ventricular outflow tract obstruction associated with posterior deviation of the infundibular septum may result in residual obstruction.

The arterial switch operation is the preferred surgical treatment for transposition of the great arteries. The theoretic advantages of the arterial switch operation include having the left ventricle and mitral valve in the systemic circulation and a reduction in the incidence of postoperative atrial arrhythmias. These benefits have been confirmed in several follow-up studies (1-6).

In transposition of the great arteries several anatomic and physiologic variations occur that may potentially affect the outcome of an arterial switch operation. Guidelines for selecting candidates for this surgical procedure have evolved and include 1) a left ventricle capable of supporting the systemic circulation after surgery, 2) a coronary artery pattern that is amenable to transfer to the neo-aorta without distortion or kinking, and 3) left ventricular inflow and...
outflow tracts free of significant structural abnormality. Our experience and that of others (6-11) suggest that the left ventricle is prepared to assume the systemic circulatory load if the arterial switch operation is performed shortly after birth or if the left ventricular pressure is near systemic levels, and most coronary artery patterns in transposition of the great arteries are amenable to an arterial switch operation. However, the impact of anatomic abnormalities of the mitral valve and left ventricular outflow tract has not been adequately explored.

Structural abnormalities of the left ventricular outflow tract may occur in up to 33% of patients with transposition of the great arteries (12,13) and mitral valve abnormalities (mostly minor) have been detected in up to 71% of autopsy specimens with transposition (14,15). The types of abnormalities that might contraindicate an arterial switch operation remain to be defined. Therefore, we reviewed our results of the arterial switch operation in patients with transposition of the great arteries and abnormalities of the mitral valve or left ventricular outflow tract, or both, with particular attention to surgical management and follow-up.

Methods

Since 1983, the arterial switch operation has been the surgical procedure of choice for patients with transposition of the great arteries and forms of double outlet right ventricle with transposition-like physiology at The Children's Hospital in Boston. The preoperative evaluation in all patients has included cardiac catheterization with angiography (either at the referring hospital or at The Children's Hospital) as well as two-dimensional echocardiography with Doppler interrogation in our institution. Echocardiographic, cardiac catheterization and operative reports of all patients who underwent an arterial switch operation from January 1983 through October 1999 were reviewed to identify abnormalities of the mitral valve, pulmonary valve or left ventricular outflow tract.

The surgical management of patients with transposition of the great arteries and either an intact ventricular septum (16,17) or a ventricular septal defect (18), our postoperative follow-up protocol (1,2) and the methods used in our noninvasive laboratory for quantifying valvular regurgitation and stenosis by Doppler echocardiography (18,19) have been reported previously.

Study patients. During the study period, 290 patients underwent an arterial switch operation; 33 had abnormalities of the mitral valve or left ventricular outflow tract detected by echocardiography or at catheterization, or both. Among these, two patients originally thought to have minor accessory mitral valve tissue but no obstruction by echocardiography were found to have no gradient at cardiac catheterization and a normal pulmonary valve at surgery. Another patient thought to have accessory mitral valve tissue but no obstruction by echocardiography was found to have a normal valve at surgery. These three patients were excluded from further analysis. The remaining 257 patients (88.3%) of the total cohort of 290 form the basis of this report.

During the same time period, 52 children with transposition of the great arteries and ventricular septal defect with severe left ventricular outflow tract obstruction (9 of whom had pulmonary atresia) were considered poor candidates for an arterial switch operation and underwent a Rastelli-type procedure. These patients are not included in this report.

Results

Pulmonary valve abnormalities (Table I). Nine patients were identified with isolated abnormalities of the native
Table 2. Dynamic Subpulmonary Obstruction in Five Patients

<table>
<thead>
<tr>
<th>Pt. No.</th>
<th>IVS/VSD</th>
<th>Age at Op (mo.)</th>
<th>PreOp LV-AO (mm Hg)</th>
<th>Discharge Echo LV-AO (mm Hg)</th>
<th>Age at Last Cath (mo.)</th>
<th>LV-AO (mm Hg)</th>
<th>AR</th>
<th>Last Echo Age (mo.)</th>
<th>LV-AO (mm Hg)</th>
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<th>Last Follow-Up Age (mo.)</th>
<th>Diastolic Murmur</th>
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<td>None</td>
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<td>48 (C)</td>
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<tr>
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<td>None</td>
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</tr>
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<td>12</td>
<td>None</td>
<td>76</td>
<td>None</td>
<td>None</td>
<td>76</td>
<td>None</td>
</tr>
</tbody>
</table>

*Patient died 26 h after surgery (see text). Abbreviations as in Table 1.

pulmonary valve. Two of the nine patients had an intact ventricular septum and the remaining seven had a ventricu-
lar septal defect. The peak systolic ejection gradient before
the arterial switch operation ranged from 0 to 50 mm Hg.
Patients with a ventricular septal defect tended to have a
larger gradient. Eight patients (aged 6 days to 11 months)
underwent a primary arterial switch operation, whereas one
patient (Case 5) underwent a rapid two stage arterial switch
operation as described by Jonas et al. (20). At operation, one
patient had a trileaflet valve with a severely hypoplastic left
anterior cusp, three had a nodular, thickened trileaflet valve
and five had a bicommissural pulmonary valve. Only one
patient (Case 8) underwent valvotomy. All nine patients
survived operation. Echocardiography with Doppler study
before hospital discharge revealed one patient with a trivial
(10 mm Hg) maximal instantaneous gradient across the
neo-aortic valve; the other eight had no measurable gradient.
Two patients had mild neo-aortic regurgitation; one of these
was the patient who had pulmonary artery banding as part of
the two stage arterial switch operation.

Mid-term evaluation by cardiac catheterization (n = 5) or
Doppler echocardiography (n = 7), or both, 4 to 27 months
after surgery has shown no patient with significant (>15 mm
Hg) neo-aortic valve stenosis. Three patients (all with a
bicuspid valve) have an audible diastolic murmur of neo-
aortic regurgitation; in two of the three this finding was
confirmed by echocardiography. All nine patients are alive
and asymptomatic at a mean follow-up interval of 19 months
(range 4 to 20) after operation.

Dynamic subpulmonary stenosis (Table 2). Five patients
had dynamic subpulmonary stenosis due to posterior bowing
of the ventricular septum in systole (Fig. 1 and 2). Three had
an intact ventricular septum and two had a small, restrictive
ventricular septal defect. Before the arterial switch opera-
tion, the peak systolic ejection gradient ranged from 25 to 55
mm Hg. At the time of arterial switch operation, the patients
ranged in age from 2.5 to 7.3 months. No fixed abnormalities
of the left ventricular outflow tract were reported at surgery.

One patient (Case 10) with an intact ventricular septum
and a preoperative left ventricular/right ventricular pressure
ratio of 0.6, died 26 h after the arterial switch operation. The
postoperative course was marked by elevated left atrial
pressure, pulmonary hemorrhage and a left pneumothorax.
Autopsy revealed a nonobstructive small linear fibrous eleva-
tion on the left ventricular septal surface at the point of
apposition with the anterior leaflet of the mitral valve.

The remaining four patients had no evidence of subaortic
obstruction by echocardiography or Doppler study at dis-
charge; one patient had mild aortic regurgitation. Mid-term
evaluation by cardiac catheterization (n = 3) and Doppler
echocardiography (n = 3) 7 to 72 months after operation has
revealed no subaortic obstruction. These four patients are
alive and asymptomatic at a mean follow-up period of 54
months (range 21 to 75) after operation.

Figure 1. Patient 12. Left ventricular angiogram (long axial oblique projection) before operation. The peak systolic ejection gradient across the left ventricular outflow tract was 40 mm Hg as a result of posterior deviation of the ventricular septum causing dynamic subpulmonary obstruction.
Figure 2. Patient 11. Echocardiographic images (long-axis parasternal view) before (Pre-ASO, top panel) and after (Post-ASO, bottom panel) the arterial switch operation. A 48 mm Hg peak systolic ejection gradient was measured across the left ventricular (LV) outflow tract before surgery. The interventricular septum protruded into the left ventricular outflow tract, with coexisting systolic anterior motion of the mitral valve, causing dynamic subaortic obstruction. After operation, the left ventricular outflow tract was unobstructed. A = anterior; Ao = aorta, L/I = leftward and inferior; LA = left atrium; PA = pulmonary artery.

Anatomic subpulmonary stenosis (Table 3). Seven patients had isolated fixed subpulmonary obstruction due to accessory atrioventricular (AV) valve tissue in five patients (Fig. 3 and 4) and to a subpulmonary membrane in two patients (Fig. 5). One patient (Case 19) had previously undergone coarctation repair and pulmonary artery banding. Before the arterial switch operation, the peak systolic ejection gradient across the left ventricular outflow tract ranged from 24 to 70 mm Hg. One patient (Case 15) with low left ventricular pressure underwent a rapid two stage arterial switch operation (20). During the arterial switch operation, the accessory AV valve tissue was resected in three of the five patients with this anomaly and retracted through the ventricular septal defect in the other two; in the two patients with a subpulmonary membrane the membrane was resected through the native pulmonary (neo-aortic) valve. All seven patients survived surgery. Echocardiography with Doppler study at discharge revealed no subaortic obstruction with mild neo-aortic regurgitation in three patients, including both patients whose subpulmonary membrane was resected through the neo-aortic valve and the one patient whose ventricular septal defect was closed through this valve.

The child who underwent a two stage arterial switch operation died of unexplained causes 7 months after operation. Although no autopsy was performed, this child had a single left coronary artery and presented with acute congestive heart failure and arrhythmias. Coronary obstruction was the presumed cause of death. Of the six survivors, mid-term evaluation by catheterization (n = 4) or echocardiography with Doppler study (n = 5), or both, 6 to 36 months after operation revealed no residual left ventricular outflow obstruction. Three patients have mild neo-aortic regurgitation. The six survivors are all asymptomatic at a mean follow-up interval of 26 months (range 6 to 36) after operation.

Mitral valve abnormalities (Table 4). Two patients with transposition of the great arteries and a ventricular septal defect had abnormalities of the mitral valve attachments. One had a cleft mitral valve with trivial regurgitation and abnormal chordal attachments to the ventricular septum with a 25 mm Hg peak systolic ejection gradient across the left ventricular outflow tract; the other had straddling of the mitral chordae to a papillary muscle group located at the junction of the right ventricular septal surface with the infundibular free wall. In this patient, a previously placed pulmonary artery band precluded accurate assessment of the degree of left ventricular outflow obstruction. In neither patient were the mitral valve attachments altered during the arterial switch operation. In the child with the straddling valve, the ventricular septal defect patch was placed so that all chordal attachments were kept on the left ventricular side.

At discharge, neither child had a significant gradient across the left ventricular outflow tract by Doppler echocardiography. One had mild mitral regurgitation (through the mitral valve cleft) and both had mild aortic regurgitation. Later catheterization and Doppler echocardiography in the patient with the cleft mitral valve revealed persistent mild mitral regurgitation.

Mixed abnormalities (Table 5). This heterogeneous group includes seven patients with various combinations of abnormalities of left ventricular inflow or outflow; all but one had a ventricular septal defect. Abnormalities of the infundibular septum contributed to the left ventricular outflow obstruction in 3 of the 7 (Cases 25 to 27). One patient (Case 27) had a subpulmonary infundibulum with close approximation of the overriding pulmonary anulus to the crest of muscular septum, whereas two patients (Cases 25 and 26) had mild posterior deviation of the infundibular septum with tunnel-
like obstruction. Various other abnormalities are described in Table 5.

One child died 3 days after the arterial switch operation. At autopsy there was mitral stenosis with thickened chordae and decreased interchordal spaces as well as a subaortic membrane that had not been recognized before operation. Echocardiographic examination at discharge in five of the six survivors revealed a 20 mm Hg gradient across the left ventricular outflow in the patient with bilateral conus and no measurable gradients in the other four patients; one patient had mild aortic regurgitation.

All three patients with left ventricular outflow obstruction associated with posterior deviation of the infundibular septum have residual outflow obstruction (Fig. 5) at mid-term follow-up study 18 to 55 months after surgery. The subaortic area appears narrow in all three patients and the gradient at catheterization ranges from 10 to 45 mm Hg. Later Doppler echocardiography in two of the three patients suggests progressive obstruction, and all three have audible murmurs of aortic regurgitation. Limited follow-up information is available on the remaining three survivors in this subgroup. Doppler echocardiography in two of the three (6 and 20 months, respectively, after operation) has shown a mild (15 mm Hg) outflow gradient and mild mitral regurgitation in one patient.

Figure 3. Patient 17. Preoperative left ventricular (A) and right ventricular (B) angiograms (long axial oblique projection). The peak systolic ejection gradient across the left ventricular outflow tract was 66 mm Hg, due to prolapsing tricuspid valve tissue (dotted lines) through a ventricular septal defect.
Figure 4. Patient 18. Preoperative echocardiographic image (subxiphoid long-axis view). The peak systolic ejection gradient across the left ventricular (LV) outflow tract was 24 mm Hg, due to prolapsing tricuspid valve tissue (arrowhead) through a ventricular septal defect. P/S = posterior and superior; R = rightward; RA = right atrium; RV = right ventricle; other abbreviations as in Figure 2.

Figure 5. Patient 21. Preoperative left ventricular angiogram (right anterior oblique projection). The peak systolic ejection gradient across the left ventricular outflow tract was 45 mm Hg, due to a subpulmonary membrane (arrows). The superior aspect of the membrane was adherent to the anterior leaflet of the mitral valve.

**Discussion**

The arterial switch operation has gained widespread acceptance as the surgical treatment of choice for infants with transposition of the great arteries. The long-term success of this approach depends, in part, on a normally functioning mitral valve and an unobstructed left ventricular outflow tract. Our results suggest that certain subgroups of children with abnormalities of left ventricular inflow or outflow may still benefit from an arterial switch operation.

**Pulmonary valve abnormalities.** Nine of the 30 patients in this series had an abnormal native pulmonary valve and underwent successful repair with good mid-term results. Gradients measured preoperatively across the pulmonary valve and left ventricular outflow tract may overestimate the degree of anatomic obstruction because of the greatly increased pulmonary blood flow in children with transposition of the great arteries, especially in those with an associated ventricular septal defect. The importance of this finding is supported by the observation that patients with a peak systolic ejection gradient as high as 45 mm Hg (with a bicommissural pulmonary valve and an associated ventricular septal defect) before the arterial switch operation had no measurable gradient at the time of hospital discharge, with no surgical intervention to the pulmonary valve.

**Subpulmonary abnormalities.** Children with dynamic left ventricular outflow tract obstruction (produced by the reversed relation of right and left ventricular pressure before operation [21]), or with surgically remediable causes of obstruction (such as accessory AV valve tissue or a subpulmonary membrane) have had good mid-term results from an arterial switch operation. As originally pointed out by Yacoub et al. (22) in 14 patients, dynamic obstruction of the left ventricular outflow tract will disappear after anatomic correction once the usual interrelation of left and right ventricular pressure is restored. At hospital discharge our four survivors with isolated dynamic obstruction showed resolution of the left ventricular outflow gradients that has persisted up to 75 months after operation. Because of the potential for developing a “contact” lesion on the interventricular septal surface adjacent to the anterior leaflet of the mitral valve (13,22) (as was seen at autopsy of the child in this subgroup who died), early repair appears warranted.

Surgically remediable causes of subpulmonary obstruction in transposition of the great arteries include tricuspid valve pouch (12,13,23), aneurysm of the membranous septum (24), subpulmonary membrane (12,13) and accessory mitral valve tissue (13-15). In the five children with isolated accessory AV valve tissue in the left ventricular outflow tract, no residual obstruction or interference with normal AV valve function was detected at later follow-up study. If the subpulmonary membrane in transposition of the great arteries is similar to a subvalvular aortic membrane in normally related great arteries, resection may yield similar long-term results (25). The two patients in this series have had good short-term results, although membrane resection was performed through the native pulmonary valve, which may be responsible for the mild neo-aortic insufficiency present in both children.
Table 4. Mitral Valve Abnormalities in Two Patients

<table>
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<tr>
<th>Pt.</th>
<th>IVS/VSD</th>
<th>Age at Op</th>
<th>Cause</th>
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<th>PreOp MR</th>
<th>Surgical Therapy</th>
<th>Discharge Echo</th>
<th>Last Cath</th>
<th>Last Echo</th>
<th>Last Follow-Up</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td>(mo.)</td>
<td></td>
<td>(mm Hg)</td>
<td>AR, MR</td>
<td></td>
<td>LV-AO (mm Hg)</td>
<td>AR, MR</td>
<td>LV-AO (mm Hg)</td>
<td>AR, MR</td>
</tr>
<tr>
<td>22</td>
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<td>Clef, septal attachments</td>
<td>27 (C)</td>
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<td>None to Valve, VSD Closure†</td>
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<td>23</td>
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<td>Straddling chordae</td>
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*Could not be assessed because of coexisting pulmonary artery band; †With all mitral apparatus on left ventricular side of patch (see text). MR = mitral regurgitation; other abbreviations as in Table 1.

Table 5. Mixed Abnormalities in Seven Patients

<table>
<thead>
<tr>
<th>Pt.</th>
<th>IVS/VSD</th>
<th>Age at Op</th>
<th>Cause</th>
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<th>PreOp MR</th>
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<th>Discharge Echo</th>
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<td>(mm Hg)</td>
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*Patient fed on 3rd postoperative day (see text); †not recognized preoperatively; ‡through the neo-aortic valve; †§with all mitral apparatus on left ventricular side of patch. DMOV = double outlet mitral valve; PV = pulmonary valve; other abbreviations as in Tables 1 and 4.
Figure 6. Patient 26. Serial left ventricular angiograms (long axial oblique projection). Preoperatively (A), peak systolic ejection gradient across the left ventricular outflow tract was 60 mm Hg, due to prolapsing tricuspid valve through a ventricular septal defect, posterior bowing of the interventricular septum and mild posterior deviation of the infundibular septum. During surgery an 8 mm Hegar dilator was easily passed through the left ventricular outflow tract. Angiograms at 6 months (B) and 16 months (C) after operation reveal residual left ventricular outflow tract obstruction (arrows) due to a subaortic membrane as well as posterior deviation of the infundibular septum. A 45 mm Hg peak systolic ejection gradient was measured at the last catheterization.

Mitral valve abnormalities. Although mitral valve abnormalities in transposition of the great arteries have been reported to be as high as 71% in pathologic series (15), we have found clinically significant mitral valve abnormalities to be uncommon. Of our 290 patients who have had an arterial switch operation, only 8 (2.8%) had identifiable mitral valve abnormalities, either in isolation or in combination with abnormalities of the left ventricular outflow tract. Tissue resection was performed in one child; no surgical therapy was performed on the abnormal valve attachments or to the left ventricular outflow tract in the other patients and follow-up study nearly 3 years postoperatively has revealed only mild mitral regurgitation in one child. Straddling of the mitral valve apparatus into the right ventricular infundibulum may be associated with various degrees of left ventricular hypoplasia. If left ventricular size is adequate and the mitral valve apparatus can be kept on the left ventricular side of the ventricular septal defect patch, an arterial switch operation can be successful.

Mixed abnormalities. The group of patients with mixed abnormalities of left ventricular inflow or outflow have had the least favorable results in our series. One patient in this group died of unrecognized mitral stenosis and a subpulmonary (neo-aortic) membrane. It was difficult to make this diagnosis from the preoperative studies, even in retrospect. It may be difficult preoperatively to assess with accuracy the relative contributions of "surgically remediable" outflow obstruction (that is, AV valve tissue), the dynamic component (produced by posterior bowing of the interventricular septum) and the fixed component (produced by conal septum). In cases of more severe fixed left ventricular outflow obstruction associated with a ventricular septal defect, a Rastelli-type repair may be preferable. However, not all ventricular septal defects may be suitable for this approach, and enlargement of the ventricular septal defect may result in complete heart block or residual left ventricular outflow obstruction, or both.

The decision to perform an arterial switch operation in the setting of multiple abnormalities of the left ventricular outflow tract or mitral valve must be individualized on a case by case basis. The relative contraindications to an arterial switch operation (such as difficult coronary transfer or possible residual left ventricular outflow obstruction) must be weighed against the relative contraindications for an atrial level repair (such as right ventricular dysfunction, tricuspid regurgitation or arrhythmia) or for a Rastelli repair (ventricular septal defect size and location and the potential need for conduit revision).

Aortic regurgitation. Mild neo-aortic regurgitation has been identified in 11 (41%) of 27 late survivors, representing a higher incidence than that seen in our overall experience (1,6). In this subgroup of patients, neo-aortic valvular dysfunction may be due to the turbulent subpulmonary flow present preoperatively, prior pulmonary artery banding in some patients or the need to perform tissue/membrane resection or ventricular septal defect closure through the neo-aortic valve. At the present time, neo-aortic regurgitation has not progressed in any patient (nor has it caused left ventricular enlargement or dysfunction), but long-term serial follow-up study will be necessary.

Conclusions. Abnormalities of the mitral valve or left ventricular outflow tract do not contraindicate an arterial switch operation if they are minor in nature or surgically remediable. Gradients measured across the left ventricular outflow tract before surgery must be interpreted cautiously because resolution may occur after an arterial switch operation with the reduction of flow across the area to normal levels and the reinstitution of a normal interrelation of right and left ventricular pressure. The preoperative evaluation by echocardiography and cardiac catheterization must be enre-
fully confirmed intraoperatively by direct inspection of the left ventricular outflow tract and mitral valve apparatus. Because of the known long-term problems of atrial level repair of transposition of the great arteries, especially with an associated ventricular septal defect, we have elected to carry out the arterial switch procedure for children with left ventricular inflow or outflow abnormalities if, during operation, the left ventricular outflow tract and mitral valve appear adequate or surgically correctable and other surgical alternatives are less desirable. Systematic follow-up study appears adequate or surgically correctable and other surgical alternatives are less desirable. Systematic follow-up study remains essential to fully evaluate this approach over the long term.

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References