Long-term Results After Atrial Repair of Transposition of the Great Arteries in Early Infancy

LYNN MAHONY, M.D., KEVIN TURLEY, M.D., PAUL EBERT, M.D., AND MICHAEL A. HEYMANN, M.D.

SUMMARY  Fifty-two patients younger than age 100 days who had an intact interventricular septum or a small ventricular septal defect underwent atrial repair of d-transposition of the great arteries (d-TGA). No patient died. To assess long-term results, we evaluated all 36 patients who had been followed for at least 1.5 years (mean 2.7 years) after surgery. The physical findings, chest roentgenograms, ECGs and echocardiograms were reviewed. Catheterization was done 6–60 months (mean 15 months) after surgery in 28 patients. Growth was normal in all but three patients. Neurologic development was abnormal in six patients (delayed speech in one patient, learning disability in three patients and preoperative cerebral infarction in two patients). The ECG showed sinus rhythm in 24 patients, minor abnormalities in nine and major dysrhythmias in three. Catheterization showed a normal cardiac index in all 28 patients. No intracardiac shunt was detected in 19 of 24 patients in whom complete oximetry data were available. One patient required reoperation for persistent atrial shunt and subsequent pacemaker placement. Two patients required baffle revision for symptoms related to superior vena caval obstruction. Right ventricular end-diastolic pressure was less than 15 mm Hg in all of 18 patients evaluated. We conclude that atrial repair of d-TGA in early infancy can be performed with a low mortality rate and a low incidence of late complications.

THE SURVIVAL rate of children with d-transposition of the great arteries (d-TGA) (concordant atrioventricular and discordant ventriculoarterial connections) and an intact interventricular septum or a small ventricular septal defect improved dramatically after balloon atrial septostomy and the atrial baffle operation were introduced.1,2 Balloon atrial septostomy in the first days of life provides palliation until an atrial baffle operation can be performed.

The optimal age for an atrial baffle operation is controversial. Early repair has been recommended for several reasons. First, although many infants are clinically stable after successful balloon atrial septostomy, before atrial repair they are at risk of death and complications including moderately severe hypoxemia, polycythemia, cerebrovascular accidents,11,12 growth retardation,13 and early onset of pulmonary vascular disease.14 In addition, infants who do not improve after balloon atrial septostomy require urgent further intervention. Because balloon atrial septostomy usually accomplishes an adequate tear in the interatrial septum, surgical atrial septectomy is generally not of further benefit, as failure of atrial mixing is probably related more to ventricular compliance and vascular resistance than to inadequate communication.5,6 Further, surgical atrial septectomy subjects the infant to a significant risk of death and morbidity, especially phrenic nerve palsy.8,10

To avoid these complications, we perform a balloon atrial septostomy at the initial cardiac catheterization on all these patients. Patients who are clinically stable after several days of observation and have a systemic arterial oxygen tension (Pao2) greater than 25–30 mm Hg and no acidemia are discharged. If they continue to do well, an atrial baffle procedure is done electively at age 2–3 months. However, if the Pao2 is not adequate after septostomy, the patient is given an infusion of prostaglandin E1 (PGE1)15 and, if necessary, a transfusion of packed red blood cells to keep the hematocrit at 50–55%. If the patient improves with PGE1 infusion, it is continued for several days to allow the normal postnatal decrease in pulmonary vascular resistance to occur. If the patient does not improve or continues to require PGE1, to maintain an adequate Pao2, an atrial baffle operation is done regardless of age.

The complications of atrial baffle operations in older infants and children have been well documented.16–30 It is important to determine whether performing these...
procedures at this earlier age subjects the patients to increased complication rates or to different complications. We therefore evaluated the clinical course and postoperative studies in 36 consecutive patients who underwent surgery at younger than 100 days of age and were followed for at least 18 months after surgery.

Methods

From January 1975 through July 1981, 52 patients younger than age 100 days underwent an atrial baffle procedure for repair of d-TGA. A Mustard procedure was done on 36 patients and a Senning procedure on 16 patients. No patient died.

The study group consisted of all 36 of these patients who were more than 18 months postoperative; follow-up data were available for all patients. All patients had undergone cardiac catheterization and balloon atrial septostomy in the first week of life. A surgical atrial septectomy was performed in one patient at another hospital. Thirty patients had an intact interventricular septum and six had a small ventricular septal defect that was not repaired. At surgery, the patients were 4–98 days old (median 52 days) (fig. 1), and weighed 2.3–6.6 kg (median 3.7 kg) (fig. 2). A Mustard procedure was done on 33 patients and a Senning procedure on three patients. These procedures were done as described previously. 4, 5, 31

The patients were followed for 1.5–5.6 years (median 2.7 years). Clinical history and physical findings, including height and weight, were reviewed. Growth was considered normal if measurements were within 2.5 SD of the mean for age. 32 Development was evaluated both by history and by observation. Developmental testing was performed unless clinically indicated. The latest available chest roentgenogram and standard 12-lead ECG from each patient were reviewed. Echocardiograms were performed on 27 patients. Cardiac catheterization was performed on 28 patients 6–60 months (median 15 months) after surgery. Right-heart catheterization was performed in 26 patients; two patients had complete obstruction of the superior vena cava (SVC) and both iliac veins, and thus the right heart could not be catheterized. Left-heart catheterization was performed in 18 patients. In some completely asymptomatic children, the patient’s cardiologist thought that complete catheterization or retrograde arterial catheterization was not indicated.

Results

General Clinical

All patients survived the immediate postoperative period. There have been no late deaths. Serious complications immediately after surgery included transient seizure activity after a cardiac arrest associated with induction of anesthesia in one patient and chylothorax that required extended hospital treatment in two other patients. No other serious perioperative problems were noted.

Growth, as measured by height and weight, was normal in all but three patients; one dysmorphic child was short and two others were slightly underweight for age (fig. 3). Five children had head circumferences greater than 2.5 SD above the mean for age. Three of these children were asymptomatic, and computed axial tomography (CAT) scans of the head done in two of these children were normal. The other two children were subjectively thought to have behavior abnormalities, and CAT scans showed mild cortical atrophy and possible early hydrocephalus. These two children had complete obstruction of the superior vena cava and their symptoms resolved after baffle revision. A repeat CAT scan in one patient was normal.

Development is apparently normal in all but six children. One patient has an unexplained delay in speech development, two patients have learning disabilities and one child who is dysmorphic has marked psychomotor developmental delay. The other two had preoperative cerebral infarctions that were diagnosed on CAT scans; these patients have residual hemiplegia. Two other patients in whom preoperative cerebral infarctions were diagnosed by CAT scan appear to be developing normally.

All patients have apparently normal exercise tolerance when compared with siblings and peers. No for-
Normal exercise testing was done because of the young age of the patients. No patient had evidence of congestive heart failure. However, one patient was treated with digoxin because of tricuspid incompetence. Occasional episodes of cyanosis were noted in only one patient, who had a large right-to-left shunt across the baffle; this shunt was subsequently repaired.

The chest roentgenograms showed normal or minimally increased heart size and right ventricular prominence in all patients. No patient had pulmonary edema. Pulmonary vascular markings were moderately increased in three patients who had left-to-right shunts.

Electrocardiography

Sinus rhythm (normal P-wave axis and contour) was present in 33 of the 36 patients (92%) at discharge from the hospital; two patients had ectopic atrial pacemakers, and one patient had junctional rhythm. At the time of this report, sinus rhythm was present in 24 patients (67%) (table 1). Serious dysrhythmias were noted in only three (8%) patients: One patient was in sinus rhythm until she underwent repair of a baffle leak, after which she developed third-degree atrioventricular block and required permanent pacemaker placement; the second patient had supraventricular tachycardia and is treated with digoxin; and the third patient has atrial flutter, which is treated with multiple medications. Two other patients had transient, asymptomatic dysrhythmias (sick sinus syndrome and supraventricular tachycardia) documented on ECG, but are now in sinus rhythm and taking no medication.

Echocardiography

M-mode and two-dimensional echocardiograms of 27 patients showed that the cardiac chamber dimensions and wall thicknesses were within the expected profile for patients after atrial repair of d-TGA.34 Typical findings included increased right ventricular wall thickness and chamber size. The atrial baffle was in the usual position in the apical four-chamber and subcostal views.35 Contrast echocardiography,36 performed on 17 patients, showed complete SVC obstruction in four, partial SVC obstruction in three and no evidence of obstruction in the remaining 10.

Cardiac Catheterization

Oximetric data showed no intracardiac shunts in 19 of the 24 patients (79%) in whom complete data were available. The mean cardiac index was 3.3 ± 1.2 l/min/m² (± SD, n = 24). A small (< 2.5 l/min/m²) left-to-right shunt was noted in three patients and a small right-to-left shunt in one patient. One patient required revision of the atrial baffle because of a large right-to-left shunt.

A mean pressure difference of more than 10 mm Hg between the SVC and systemic venous atrium was observed in seven of the 28 patients (25%) (table 2). Two of these patients have undergone baffle revision because of mild cerebral atrophy and early hydrocephalus diagnosed by CAT scan. The other patients are asymptomatic. One of the two patients with postoperative chylothorax had complete SVC obstruction but was asymptomatic; the other had no SVC obstruction. No patient had obstruction at the inferior limb of the baffle (mean pressure difference of < 5 mm Hg between the inferior vena cava and systemic venous atrium). Only two patients had left ventricular outflow tract obstruction (pressure difference of < 40 mm Hg, table 3); in one it was present before surgery. Both were asymptomatic and neither has required surgery. The pulmonary arterial pressure and pulmonary vascular resistance were normal in all 25 patients evaluated. One patient had pulmonary venous obstruction (mean pressure difference of 10 mm Hg between pulmonary vein and pulmonary venous atrium), but she is asymptomatic.

<table>
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<th>TABLE 1. Results of Most Recent Standard 12-lead ECGs.</th>
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<td>ECG diagnosis</td>
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<td>Sinus rhythm</td>
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<td>Sinus rhythm with occasional junctional escape beats</td>
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<td>Passive dysrhythmias</td>
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<td>Ectopic atrial pacemaker</td>
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<td>Junctional rhythm</td>
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<td>Major dysrhythmias</td>
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<td>Third-degree atrioventricular block</td>
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<td>Supraventricular tachycardia</td>
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<td>Atrial flutter</td>
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<th>TABLE 2. Superior Vena Cava Obstruction Measured by Mean Pressure Difference Between Superior Vena Cava and Systemic Venous Atrium</th>
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<td>Pressure difference (mm Hg)</td>
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<td>&gt; 10</td>
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<td>Complete obstruction</td>
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tomatic. The right ventricular end-diastolic pressure was less than 10 mm Hg in 14 of the 18 patients (78%) evaluated (table 4). No patient had a right ventricular end-diastolic pressure greater than 15 mm Hg.

Angiography confirmed the SVC obstruction in seven patients. A large right-to-left shunt across the baffle was seen in one patient. One patient had tricuspid insufficiency, but he is taking digoxin and is asymptomatic. The right ventricle was enlarged in all patients, but contractility was normal.

**Discussion**

Infants with d-TGA are at risk of complications, including cerebrovascular accidents, growth retardation and pulmonary vascular disease. In addition, prolonged exposure of tissues such as brain and myocardium to low oxygen tensions may damage these tissues and impair long-term function. Although early surgical correction might be expected to prevent most of these complications, we considered it important to assess whether operation at an early age was associated with increased mortality or morbidity or a change in the type of morbidity when compared with a more traditional approach.

Our results show that atrial repair of uncomplicated d-TGA in early infancy is safe and has an excellent outcome. The mortality rate is lower than that reported in older children and is similar to that reported more recently in small groups of younger children.

Most of our patients showed no symptoms of cardiorespiratory dysfunction. Exercise tolerance was evaluated subjectively because the patients were too young to perform exercise studies. Previous reports have documented abnormal cardiorespiratory responses to exercise in asymptomatic children after atrial repair of d-TGA, but the patients in those reports were older at surgery than our patients.

Motor and psychological development were not formally evaluated unless clinically indicated. Although development was grossly normal in most of these patients, more problems may become apparent as these children enter school.

The reported incidence of dysrhythmias after atrial baffle operations has been 10–80%.

Among our patients, 67% had sinus rhythm (normal P-wave axis and contour) by standard 12-lead ECG and only 8% had active dysrhythmias. Although the true incidence of dysrhythmias, especially those considered "passive," is probably higher and could be better detected by 24-hour monitoring, we agree with Southall et al. that it is important not to ascribe all dysrhythmias to surgical intervention. These investigators reported intermittent junctional escape rhythms, atrial premature complexes (fewer than 12/hour), complete sinoatrial block and 2:1 sinoatrial block in normal neonates and young children. The lower incidence of dysrhythmias in our report and in more recent reports, compared with that in earlier series, appears to be related to surgical technique, including the use of deep hypothermia and total circulatory arrest to improve exposure and the placement of a single atrial cannula and baffle sutures to minimize damage to the sinus and atroventricular nodes and to their blood supplies. Since the follow-up period is relatively short in some of our patients, continued observation is important.

Echocardiography is useful in evaluating patients after atrial baffle procedures. The ability to assess ventricular chamber size and wall thickness, left ventricular outflow tract obstruction, intracardiac shunts, and SVC obstruction is helpful for deciding appropriate times for recatheterization. The echocardiograms correctly predicted all instances of severe SVC obstruction. We have found two-dimensional contrast echocardiography particularly useful in assessing SVC obstruction in asymptomatic patients and those with increased head circumference and chylothorax.

Other investigators have suggested that the incidence of complications resulting from the atrial baffle was increased in patients operated on at younger ages. In our patients, the incidence of shunts across the baffle detected at catheterization was lower than in other studies. Similarly, significant pulmonary venous obstruction and inferior vena cava obstruction were not present in our patients. In contrast, SVC obstruction was found in a significant number of patients. Trusler et al. reported SVC obstruction in six of 100 (mean age at operation 16 months) patients operated on after 1973; all were asymptomatic. In contrast, 10 of the 81 patients reported by Egloff et al. (mean age 9 months) had SVC obstruction and seven of these required baffle revision. Similarly, Arciniegas et al. found significant SVC obstruction in 12 of their 61 patients (mean age 28 months); three of these were symptomatic and two required reoperation. Although most of our patients were asymptomatic (probably because of decompression by the azygos vein), our concern about the relatively high incidence of SVC obstruction has led to a recent increase in the number of Senning procedures. Indeed, although a Senning procedure was performed in only three of the 35 infants in

### Table 4. Right Ventricular End-Diastolic Pressure

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<th>Pressure (mm Hg)</th>
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<td>&lt; 5</td>
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<td>5–10</td>
<td>11</td>
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<td>10–15</td>
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<td>&gt; 15</td>
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<td>Not evaluated</td>
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### Table 3. Left Ventricular Outflow Tract Obstruction Measured by Peak Systolic Pressure Difference Between the Left Ventricle and the Pulmonary Artery

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<th>Pressure difference (mm Hg)</th>
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<td>&gt; 40</td>
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the study group, this procedure was used in 13 of the 17 children operated on most recently at less than 100 days of age. Other groups have noted a low incidence of caval or pulmonary venous obstruction in patients after the Senning procedure and, although the period of follow-up is too short to reach any conclusions in our patients, we hope that the incidence of SVC obstruction will decrease. It is important to monitor head size in all children after an atrial baffle procedure and to investigate thoroughly any patient who has a head circumference greater than 2.5 SD above the mean.

The results of the postoperative hemodynamic studies were normal in most patients. The very low incidence of significant left ventricular outflow tract obstruction and uniformly normal pulmonary arterial pressures are similar to results in other reports. Tricuspid valve competence and right ventricular function continue to be a cause of concern. Tyan et al. reported tricuspid insufficiency in 17 of 21 patients, but nine of these had ventricular septal defects that were repaired. The low incidence of tricuspid insufficiency in our patients is comparable to that in other series. Although right ventricular end-diastolic pressure was normal or only slightly increased in all patients in whom it was measured, the long-term outcome for right ventricular function is uncertain. No patient had a decreased resting cardiac index and all appeared to have normal right ventricular contractility as subjectively assessed from the angiograms. Other investigators have attempted to quantify right ventricular function using ejection fraction and various indexes of contractility, but in our experience these measurements have not proved reliable in the hypertrophied, heavily trabeculated right ventricle.

We conclude that atrial repair of d-TGA can be done in early infancy with low mortality and decreased or similar complications as compared with older children. For infants not clinically stable after a balloon atrial septostomy, an atrial baffle operation is the current procedure of choice. We recommend early elective repair for other infants because of the excellent results and decreased risk of complications associated with hypoxemia.

Acknowledgment
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Blade Atrial Septostomy: Collaborative Study

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SUMMARY During the past 4 years, five institutions have collaborated in evaluating the efficacy of blade atrial septostomy. The procedure was performed in 52 patients, including 31 with transposition of the great arteries, 10 with mitral atresia, five with tricuspid atresia and six with miscellaneous anomalies. The patient’s ages ranged from 1 day to 12 years (mean 13 months). Improvement occurred in 41 of 52 patients (79%). Four patients had an intact interatrial septum, and blade atrial septostomy was successfully performed by a transseptal technique. One patient died from a lacerated left atrial wall; other complications occurred in four patients. Blade atrial septostomy is an effective palliative procedure, even when the interatrial septum is thickened or intact.

IN PATIENTS with certain varieties of congenital heart malformations, an adequate interatrial opening (IAO) is essential for survival. An IAO is important in transposition of the great arteries (TGA), as well as for obligatory interatrial shunt lesions such as mitral atresia, tricuspid atresia and total anomalous pulmonary venous return. An adequate IAO is hemodynamically beneficial to patients with double-outlet right ventricle and a restrictive ventricular septal defect (VSD) and those with a univentricular heart and a unilaterial restrictive atrioventricular valve. Balloon atrial septostomy, introduced by Rashkind and Miller in 1966,1 was the first innovative nonsurgical therapeutic modality in pediatric cardiac catheterization. Palliation has been achieved in the majority of patients who have undergone this procedure. However, in some patients, particularly older infants and children with a thickened interatrial septum, this procedure is usually not successful. In patients with an inadequate IAO after balloon atrial septostomy in infancy, the interatrial septum is often thickened, and a repeat balloon atrial septostomy is often ineffective.2

In 1973, a catheter with a built-in surgical blade was developed to enlarge the IAO in such patients. Experiments in animals substantiated its efficacy and safety,3 and clinical use of this technique has been successful in a limited number of patients.4 To extend the clinical trial of the technique, a collaborative study was organized in 1977.

In this report, we review the overall result of a collaborative study of blade atrial septostomy. The procedure is described in detail because the catheter
Long-term results after atrial repair of transposition of the great arteries in early infancy.
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