Senning repair for transposition of the great arteries in the first week of life

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ABSTRACT Infants with d-loop transposition of the great arteries (d-TGA) may have unacceptable results after balloon septostomy while awaiting surgery. It has been our policy to repair defects in infants with d-TGA and intact ventricular septum or small ventricular septal defect at the time of diagnosis. We report our experience with the Senning operation in 18 newborns less than 1 week of age. The mean age at operation was 3 days (range 12 hr to 7 days) and the mean weight was 3.5 kg (range 2.8 to 4.8). There were two early postoperative deaths (11%) and one late death (5%). Early mortality was associated with preoperative acidosis and congestive heart failure. Late mortality was associated with severe left ventricular outflow tract obstruction (LVOTO). The 15 long-term survivors have been followed for an average of 27 months and 11 of the 16 perioperative survivors have undergone postoperative catheterizations. There was no evidence of systemic or pulmonary venous obstruction. One patient developed LVOTO that led to his death. Two patients had residual atrial shunts. Electrocardiograms revealed no major arrhythmias. All patients are clinically asymptomatic. Good hemodynamic, electrocardiographic, and clinical results can be obtained with correction of d-TGA in the first week of life.

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MANAGEMENT OF d-loop transposition of the great arteries (d-TGA) has undergone dramatic change in the past 35 years. The development of the Blalock-Hanlon,1, 2 Mustard,3 Senning,4, 5 and Jatene procedures, along with Rashkind balloon septostomy,7 has led to significant decreases in mortality and morbidity.

Recently, interest has been focused on the correct age for repair. The trend has been toward earlier correction to avoid morbidity and mortality.8–17 The long-term results of early repair are comparable to those of later repair without the risks associated with uncorrected d-TGA.11, 16, 17

While palliation is achieved in many infants with d-TGA with balloon septostomy, significant complications can occur while these infants are awaiting surgery. Gutgesell et al.18 have reported a 22% incidence of death, cerebrovascular accident, or obstructive pulmonary vascular disease in infants with palliated d-TGA awaiting surgery.18 Because of this unacceptable morbidity and mortality, it has been the policy at our institution to repair defects in all infants with d-TGA at the time of diagnosis. We report the results of 18 infants with d-TGA corrected with the Senning procedure during the first week of life.

Methods

Patients. From April 1979 through January 1984, 18 infants 1 week or less old underwent Senning repair for d-TGA. The patient population was derived from records of the Departments of Pediatric Cardiology and Thoracic Surgery. Infants with a patent ductus arteriosus (PDA) or small ventricular septal defect (VSD) without adequate mixing were included. Initial evaluation included cardiac catheterization at our institution in 13 and at a referring hospital in five.

Surgical procedure. A modified Senning operation was performed in all 18 infants. The atrial patch was augmented with Knitted Dacron in 11 patients and enlargement of the new pulmonary venous atrium with a pericardial patch was necessary in one patient. All PDAs were ligated and three patients had suture closure of a VSD. Hypothermic circulatory arrest was used in all patients (18°C in 17 and 20°C in 1). The mean period of circulatory arrest was 57 min (range 47 to 77).

Follow-up. Clinical history, physical examination, and growth parameters were reviewed along with chest roentgenograms and electrocardiograms. Postoperative cardiac catheterization was performed in 11 patients. Measurement for left ventricular outflow tract obstruction (LVOTO) was performed if clinically indicated by elevated left ventricular pressures or by the presence of a murmur suggestive of LVOTO. Pulmonary venous pressures were measured if indicated by clinical evidence of obstruction. Systemic venous obstruction was determined by measuring the gradient between the mean superior vena cava or inferior vena cava pressure and the mean syste-
ic venous atrial pressure. Intracardiac shunts were determined by oximetry data.

**Survival analysis.** Survival was analyzed by the Kaplan-Meyer method for actuarial survival.19

**Results**

**Patient profile.** Eighteen neonates with d-TGA underwent Senning repair between April 1979 and January 1984 (table 1). This group comprised all but five newborns with d-TGA seen at our institution during that period; one infant was excluded because repair was delayed until 2 months of age and four infants were excluded because they had VSDs. Six of the patients had no associated cardiac defects, five had associated PDA, four had an associated VSD, and three patients had both PDA and a VSD. LVOTO was not seen preoperatively. All of the patients with VSDs had small defects without adequate mixing. The age range at operation was 12 hr to 7 days (mean 3 days), and the weight range was 2.8 to 4.8 kg (mean 3.5). Sixteen patients had undergone Rashkind balloon septostomy before operation.

**Preoperative and postoperative profile.** Preoperative complications consisted mainly of hypoxemia and congestive heart failure. Seven patients underwent a trial of prostaglandin E, for persistent hypoxemia, and three of the patients received digoxin for congestive heart failure. Three patients underwent repeat Rashkind septostomy because of persistent hypoxia. Other preoperative complications were transient supraventricular tachycardia during catheterization in one patient and a blood culture positive for *Staphylococcus aureus* in one patient.

After surgical repair, all of the patients were weaned from cardiopulmonary bypass. Seven patients had transient postoperative complications that resolved without sequelae, while two patients had complications that led to their deaths. Transient complications included junctional rhythm in four, supraventricular tachycardia in one, ventricular fibrillation in one, cardiac arrest in one, and seizures in one patient. Two patients with transient junctional rhythms developed passive arrhythmias on follow-up; one of these patients had first-degree atrioventricular block and the other had ectopic atrial pacemaker and occasional junctional beats. Fatal complications included ventricular failure in one patient and severe ischemic central nervous system damage in one patient. No patient demonstrated evidence of systemic or pulmonary venous obstruction postoperatively. However, one patient who died had evidence of systemic venous obstruction and required augmentation of the systemic atrium during weaning from bypass. No patient underwent reoperation for systemic or pulmonary venous obstruction.

Among the 16 perioperative survivors the duration of ventilator support ranged from 1/2 to 7 days (mean 1.5). The duration of hospitalization was from 7 to 21 days (mean 11).

**Mortality.** The overall mortality was 17% (3/18; table 2). Two patients died during the perioperative period (perioperative mortality 11%) and one patient died at 33 months of age.

The two patients that died during the perioperative period had very complicated preoperative courses. Both patients developed severe acidosis and hypoxia that required urgent surgical correction. One patient whose pH was less than 7.03 for 5 hr before surgery developed severe neurologic dysfunction shortly after surgery. He died on the tenth postoperative day without neurologic recovery. The second patient was responsive only to deep pain with nonreactive pupils upon arrival at our institution. The emergency surgical procedure was complicated by obstruction of the superior and inferior venae cavae when the patient was first weaned from cardiopulmonary bypass, which required augmentation of the systemic atrium. The patient was weaned from bypass the second time, but died a short time later from continued hypotension and poor ventricular function. Autopsy in both infants revealed good surgical repair without evidence of systemic or pulmonary venous obstruction.

The patient who died late after surgery was found to have LVOTO and a residual VSD on postoperative catheterization at 12 months of age. At the initial catheterization he was thought to have a small 3 to 4 mm supracristal VSD, but LVOTO was not found. At 27 months of age, he underwent patch closure of the VSD, pulmonary commissurotomy, and infundibular resection. Postoperative catheterization revealed residual severe LVOTO. At 33 months of age, he under-

**TABLE 1**

<table>
<thead>
<tr>
<th>Patient characteristics</th>
<th>18</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of patients</td>
<td>18</td>
</tr>
<tr>
<td>Ratio of male/female patients</td>
<td>15/3</td>
</tr>
<tr>
<td>Age at operation (days)</td>
<td>3.0 (0.5)</td>
</tr>
<tr>
<td>Mean (SEM)</td>
<td>0.5-7.3</td>
</tr>
<tr>
<td>Range</td>
<td>3.5 (0.1)</td>
</tr>
<tr>
<td>Weight at operation (kg)</td>
<td>2.8-4.2</td>
</tr>
<tr>
<td>Associated defects</td>
<td>5</td>
</tr>
<tr>
<td>PDA</td>
<td>4</td>
</tr>
<tr>
<td>VSD</td>
<td>3</td>
</tr>
<tr>
<td>PDA VSD</td>
<td>16</td>
</tr>
<tr>
<td>Rashkinds</td>
<td>11</td>
</tr>
</tbody>
</table>

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TABLE 2
Findings at follow-up

<table>
<thead>
<tr>
<th>Perioperative survivors</th>
<th>16/18 (89%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Long-term survivors</td>
<td>15/18 (83%)</td>
</tr>
<tr>
<td>Length of follow-up (months)</td>
<td>27.0 (4.2)</td>
</tr>
<tr>
<td>Age at 1st postop. cath. (months)</td>
<td>14.6 (1.8)</td>
</tr>
<tr>
<td>Pressures at postop. catheterization (mm Hg)</td>
<td>6.7 (0.8)</td>
</tr>
<tr>
<td>Electrocardiography (n)</td>
<td>11</td>
</tr>
<tr>
<td>Sinus rhythm</td>
<td>1</td>
</tr>
<tr>
<td>SR primary AV block</td>
<td>1</td>
</tr>
<tr>
<td>Junctional rhythm</td>
<td>1</td>
</tr>
<tr>
<td>Ectopic pacemaker</td>
<td>1</td>
</tr>
<tr>
<td>Sinus bradycardia</td>
<td>1</td>
</tr>
<tr>
<td>Chest roentgenography (n)</td>
<td>12</td>
</tr>
<tr>
<td>Normal</td>
<td>1</td>
</tr>
<tr>
<td>Increased pulmonary vasculature</td>
<td>1</td>
</tr>
<tr>
<td>Increased pulmonary vasculature and enlarged heart</td>
<td>2</td>
</tr>
</tbody>
</table>

SVC = superior vena caval; SA = sinoatrial; LV = left ventricular.

went repeat infundibular resection, but some LVOTO persisted and he died after surgery from ventricular failure. Autopsy revealed evidence of the previous surgical procedures along with a stenotic pulmonary valve and biventricular hypertrophy. There was no evidence of systemic or pulmonary venous obstruction.

General clinical follow-up. The 15 long-term survivors have been followed from 9 to 64 months (mean 27; table 2). All patients were asymptomatic and doing well without limitations of activity and with grossly normal development. Two patients have weights lower than the 5th percentile for their ages, but the remaining patients are growing normally.

Chest roentgenography. Chest roentgenograms showed no evidence of pulmonary venous congestion in any patient. Twelve patients had normal cardiac silhouettes and normal pulmonary vascular markings. Two patients with a residual atrial shunt had evidence of increased pulmonary vascularity; one of these had mild cardiomegaly. One other patient had mild cardiomegaly with prominent pulmonary vascular markings.

Electrocardiography. No major arrhythmias were detected in any patient. Sinus rhythm was present in 11 patients, as determined from the follow-up electrocardiogram. Five patients had passive arrhythmias, including first-degree atroventricular block, junctional rhythm, ectopic atrial pacemaker with occasional junctional beats, and sinus bradycardia with occasional junctional beats.

The patient with the sinus bradycardia had a possible episode of supraventricular tachycardia at another hospital, but the episode was not documented and has not recurred.

Cardiac catheterization. Postoperative cardiac catheterization was performed in 69% (11/16) of the patients who survived the perioperative period. The mean age at catheterization was 14.5 months (range 4 to 26). No patient demonstrated evidence of pulmonary or systemic venous obstruction. The superior vena caval-to-systemic venous atrial gradient ranged from 0 to 2.5 mm Hg. We were unable to enter the inferior vena cava from the femoral vein in two patients, but neither showed angiographic or hemodynamic evidence of systemic venous obstruction. All patients had evidence of good systemic and pulmonary ventricular function. Two patients had evidence of residual atrial shunts. One of the patients with a residual atrial shunt had evidence of mild LVOTO (23 mm Hg gradient) that was thought to be secondary to flow. One patient had severe LVOTO, with an 85 mm Hg gradient. One patient showed evidence of mitral incompetence that was thought to be catheter induced.

Survival analysis. Survival was analyzed with the Kaplan-Meyer method for actuarial survival for small samples.\(^\text{10}\) The resulting estimated cumulative patient survival curve for the 18 patients is shown in figure 1. The estimated 2 year survival rate is 89%, with nine patients so far having survived 2 years.

Discussion

The infant with d-TGA after septostomy is at risk for many complications, including recurrent hypoxia,\(^\text{20}\) cerebrovascular accidents,\(^\text{18, 21, 22}\) pulmonary
hypertension, growth retardation, and death. Mortality for infants after septostomy and before correction varies from 12% to 28%. The high mortality for infants awaiting repair has led to earlier repair, but the optimal age has remained controversial.

Our data show that the Senning procedure can be performed in the first week of life with excellent results. The perioperative mortality of 11% (2/18) is comparable to that in other series in infants and in older patients. The two patients who died in the perioperative period were both moribund at the time of surgery because of complicated preoperative courses. The patient who died late after surgery developed intractable LVOTO and expired after the second surgical procedure to relieve the obstruction. It is encouraging that no deaths were associated with complications related to the Senning procedure; all were associated with preoperative condition or additional cardiac anomalies.

Few studies have taken into account preoperative morbidity and mortality along with perioperative mortality in the analysis of final outcome of treatment of d-TGA. Among infants with uncomplicated d-TGA who were treated with septostomy, Gutgesell et al. reported 8% mortality during the first month of life. Among the 1 month survivors they reported an additional 14% incidence of death or major complication in infants awaiting surgical correction. Their estimated 2 year survival for patients with uncomplicated and complicated (significant VSD and LVOTO) d-TGA was 70%. Our survival rate of 90% at 2 years compares favorably. Although the study by Gutgesell et al. included patients with complicated d-TGA and included patients who had undergone the Mustard procedure, it highlights the significant morbidity and mortality that occur in infants with d-TGA after septostomy and before surgical correction.

Postoperative complications in survivors were transient without sequelae. No patient with postoperative rhythm disturbances showed evidence of significant arrhythmias on long-term follow-up. There were no long-term neurologic sequelae in the patients who experienced cardiopulmonary arrest or seizures. While the incidence of postoperative complications was disquieting, the long-term morbidity and mortality was not affected.

The long-term complications of the Senning procedure include pulmonary and systemic venous obstruction, arrhythmias, and residual atrial shunts. The incidence of these complications with the Senning procedure is lower than with the Mustard operation, but they remain a significant problem. Some authors have suggested that complications occur more frequently with the Mustard operation in young patients, but this has not been found with the Senning procedure. Systemic venous obstruction was not found in the patients in our series. This compares favorably with the reported incidence of up to 5% in infants and up to 10% in older patients. Although the length of follow-up is limited (mean 27 months), our results are encouraging. Pulmonary venous obstruction was also not found in our patients. Others have reported incidences of up to 16% in infants and 10% in older patients.

The incidence of residual atrial shunts was 11% (2/18) in this series. Neither of the patients with a shunt is symptomatic, although one weighs less than the 5th percentile weight for his age. It is planned that both patients will have elective repair in the future. Development of atrial shunts has been reported after the Senning procedure, with an incidence up to 15% in older patients and an incidence of up to 12.5% in infants. Whether this complication is related to age and size or some other factor is not known, but it has not been a significant cause of morbidity and mortality.

The incidence of LVOTO was 11% (2/18). In one patient it was severe and led to death and in the other patient it was mild and believed to be secondary to flow from an atrial shunt. In other series, LVOTO has been reported to occur in up to 59% of the patients, with severe LVOTO in 8%. The incidence of passive arrhythmias was 25% in our series and no active arrhythmias were noted. No morbidity or mortality was related to arrhythmia. Other series have reported transient junctional rhythm postoperatively and on follow-up. The presence of active arrhythmias that require pacemaker implantation have also been reported after the Senning procedure. Whether or not passive arrhythmia will change to active ones is not known, but these patients will require close follow-up.

No patient in this series showed evidence of cardiorespiratory compromise. They were all reported to be in general good health with grossly normal development. Two patients did have weights lower than the 5th percentile for their ages. One patient had an atrial septal defect on postoperative catheterization that may have been the cause of growth failure. The other patient had a normal postoperative study and there was no identifiable cause for his low weight.

This report and others have demonstrated the ability to surgically correct the heart defects of children.
with uncomplicated d-TGA during the first week of life and the operative mortality in this and the other studies was similar. The decision of whether to use the Jatene arterial switch procedure or to perform a Senning procedure should be based on the child’s anatomy and the surgeons experience. The Senning procedure is not as technically demanding as the Jatene and may not result in as many late operative complications. The incidence of late anastomotic stricture at the coronary artery anatomosis or the arterial, pulmonary, or aortic anatomosis is not known, particularly in children who undergo the Jatene procedure as neonates. The incidence of late operative complications in this series of patients is gratifyingly low, but the incidence of late sequelae due to the right ventricle and the tricuspid valve being subjected to systemic pressures indicate the necessity for continued long-term follow-up.

Correcting heart defects in infants with d-TGA during the first week of life offers several advantages over later repair. With early repair the significant morbidity and mortality associated with palliation in infants with d-TGA can be avoided, thereby improving the overall mortality and morbidity of infants with d-TGA. Early repair also has the advantage of decreasing the duration of cyanosis, which has been associated with poor somatic growth and decreased cognitive and gross motor development and has been postulated to be associated with late right ventricular dysfunction. While the period of follow-up in this study was short (mean 27 months) and the number of patients was small (n = 18), the results presented are convincing evidence in favor of early elective repair of infants with d-TGA.

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