The Palliative Mustard Operation for Double Outlet Right Ventricle or Transposition of the Great Arteries Associated with Ventricular Septal Defect, Pulmonary Arterial Hypertension, and Pulmonary Vascular Obstructive Disease

A Report of Eight Patients

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SUMMARY Five patients with double outlet right ventricle, ventricular septal defect, pulmonary arterial hypertension and pulmonary vascular obstructive disease and three patients with complete d-transposition of the great arteries, ventricular septal defect, pulmonary arterial hypertension and pulmonary vascular obstructive disease underwent an elective Mustard baffle operation. The ventricular septal defect was not closed. A large patent ductus arteriosus was divided in three patients. Seven of the eight patients are alive five to 32 months after surgery; one patient died 11 months after surgery. Cyanosis, dyspnea on exertion, and exercise limitation improved initially in all and has persisted in the survivors. In pre and postoperative hemodynamic studies in four patients, systemic arterial oxygen saturation and effective pulmonary blood flow increased from mean values of 70% to 90% and 1.7 L/min/m² to 3.3 L/min/m², respectively. Absolute systemic and pulmonary flows, and pressures and resistances, were not significantly altered.

Criteria for selection of patients with transposition of the great arteries or double outlet right ventricle who would benefit from a palliative Mustard procedure (Mustard atrial baffle without closure of the ventricular septal defect) are: 1) severe symptoms; 2) pulmonary arterial hypertension (75% systemic) with pulmonary vascular obstructive disease; and 3) pulmonary arterial oxygen saturation greater than systemic (ascending aorta) arterial oxygen saturation by approximately 10%.

THE RESULTS OF SURGICAL MANAGEMENT OF PATIENTS WITH TRANSPOSITION OF THE GREAT ARTERIES (d-TGA) and intact ventricular septum have consistently improved since the introduction of Mustard's intra-atrial baffle procedure. However, at least one third of patients with d-transposition have an associated ventricular septal defect (VSD). This group of patients is seldom severely hypoxic in infancy, in contrast to babies with an intact ventricular septum, but is at high risk to develop congestive heart failure, pulmonary arterial hypertension (PAH), and pulmonary vascular obstructive disease early in life. Palliation with a pulmonary artery band carries a high mortality, and complete repair in infancy was not possible. In the past most of these patients who were not successfully banded died in congestive heart failure in infancy or early childhood. A small number, however, developed pulmonary vascular obstructive disease (PVO) and survived into later childhood and adolescence.

This report presents the surgical experience at the Children's Hospital Medical Center, Boston, Massachusetts, in eight consecutive patients with either d-transposition of the great arteries and ventricular septal defect, or double outlet right ventricle (DORV) with a subpulmonary ventricular septal defect and no pulmonary stenosis (Taussig-Bing malformation or partial transposition). All eight patients had PAH and PVO. Four patients had a patent ductus arteriosus (PDA), and two, coarctation of the aorta. The clinical, anatomic, and pathophysiologic features of these patients are described and the surgical management and results using the Mustard procedure are presented.

Materials and Methods

Following Lindesmith's report of successful palliative Mustard operation in patients with TGA, VSD, and PAH, 19 patients with marked cyanosis and exercise intolerance underwent detailed cardiac catheterization and angiographic studies in our institution to identify individuals suitable for the proposed surgical intervention. In some patients the cardiac anatomy was unknown or incompletely diagnosed; others were thought, on clinical grounds, to have transposition with a ventricular septal defect and pulmonary vascular obstruction. The major hemodynamic criteria sought were 1) markedly elevated pulmonary arterial pressure; 2) markedly elevated pulmonary arteriolar resistance; 3) pulmonary arterial oxygen saturation appreciably greater than ascending aortic saturation. The important anatomic prerequisites were 1) complete or partial transposition of the great arteries; 2) ventricular septal defect; and 3) two well-developed ventricular sinuses with two atrioventricular valves. Eight patients from the initial group of 19 fulfilled the criteria and have undergone surgical treatment (table 1). One 23-month-old patient (CHMC #83-71-12) with TGA, VSD, and PAH is excluded from this
report because the VSD was patched 12 hours after the Mustard operation. Although significant PAH was noted preoperatively, measurement of a significant left-to-right shunt in the operating room led to the decision to close the VSD. The child died several hours later; postmortem examination revealed Grade III vascular obstructive disease in both lungs.

All eight patients underwent resting, supine right and left heart catheterization, biventricular angiography, and angiotography (tables 1 and 2, figs. 1–3). Sedation was achieved with a mixture of Meperidine (2 mg/kg, not exceeding a total of 50 mg), Thorazine (0.5 mg/kg, not exceeding a total of 12.5 mg) and Phenergan (0.5 mg/kg, not exceeding a total of 12.5 mg) administered intramuscularly approximately 1/2 hour before the procedure. Cardiac catheterization and angiography were accomplished without difficulty. Blood oxygen saturations were measured with an oximeter (American Optical Corporation Model 10840) in all eight patients at preoperative catheterization and in four patients at postoperative catheterization. Ear Oximeter determinations (Waters Instrument Corporation, Model XE 340) were measured in the four patients who have not undergone recatheterization. Minute O₂ consumption (VO₂) was measured directly in three of eight patients at the preop study, and in three of four of the postop study; an assumed value was used in the remainder. Calculation of cardiac output and pulmonary blood flow, by the Fick principle, was possible preoperatively in four patients (1, 2, 4, 5) without PDA, and in one with a hemodynamically insignificant PDA (3). In the three patients with pulmonary-to-aorta shunting, as demonstrated by reversed differential cyanosis (ascending aortic oxygen saturation less than descending aortic oxygen saturation), neither absolute systemic nor pulmonary blood flow could be calculated. Effective pulmonary blood flow was calculated in each patient at both studies. Postoperative resting right and left catheterization and angiography have been obtained in four patients and are compared to preoperative findings (table 2). In addition, to assess the status of the pulmonary vascular bed, systemic and pulmonary blood flows and pressures were measured before and after the administration of tolazoline (Priscoline) (1 mg/kg i.v.) in one patient (4) prior to surgery and in two patients (1, 6) following surgery. Oxygen was administered at catheterization to two patients (4 and 6) preoperatively, and one patient (6) at postoperative evaluation. Pre- and postoperative bicycle exercise studies were attempted in two (2, 7) and completed in one (fig. 4). Bilateral lung biopsies were available in two patients (3, 5).

Table 1 presents the clinical data and anatomic features at operation. Seven of eight patients were males; age at operation ranged from 5-6/12 to 21-10/12 years, median age 13 years. Three patients had complete d-transposition of the great arteries (fig. 1) and two of these had left juxtaposition of the atrial appendages (JAA); one of these latter two had a small right atrial chamber. All three had a left superior vena cava (LSVC), and one of these also had a small patent ductus. Five had double outlet right ventricle with subpulmonary VSD (fig. 2) and three of these also exhibited large

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**Table 1. Clinical and Anatomic Data**

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<tr>
<th>Pt/Sex/Age (yrs)</th>
<th>Anatomic Diagnosis</th>
<th>PDA</th>
<th>NYHA Class</th>
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<td></td>
<td></td>
<td>Pre</td>
<td>Post</td>
<td></td>
</tr>
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<td>1/M/15</td>
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<td>I-II</td>
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<td>PAB, atrial septectomy</td>
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<td></td>
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<tr>
<td>4/F / 5 1/12</td>
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<tr>
<td>5/M/12 11/12</td>
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<tr>
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**Abbreviations:** Coarctation = coarctation of the aorta; DORV = Double outlet right ventricle; d-TGA = d-transposition of the great arteries; JAA = juxtaposition of atrial appendages; LSVC = left superior vena cava to coronary sinus; NYHA = New York Heart Association; PAB = patent ductus arteriosus; RA = right atrium; T-B = Taussig-Bing malformation; VSD = ventricular septal defect

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**Table 2. Hemodynamic Data**

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<tr>
<th>Pt</th>
<th>SA O₂</th>
<th>PAO₂</th>
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<th>PBF L/min/m²</th>
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<th>PAP mm Hg</th>
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<th>Rp units</th>
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<td>NC</td>
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<td>NC</td>
<td>NA</td>
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</tr>
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</table>

*Ear oximetry.
†Proximal to right peripheral pulmonary artery stenosis.

**Abbreviations:** EPBF = effective pulmonary blood flow; NA = not available; NC = not calculated; PAH = pulmonary arterial pressure, mean; PAO₂ = pulmonary arterial oxygen saturation; PBF = pulmonary blood flow; Post = postoperative; Rp = resistance pulmonary (clinical units); Rs = resistance, systemic (clinical units); SA O₂ = systemic arterial oxygen saturation; SBF = systemic blood flow; SAP = systemic arterial pressure, mean.
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Results

Clinical

All eight patients survived surgery and improved subjectively, with decreased cyanosis and dyspnea (New York Heart Association functional classification I–II, table 1). One patient (8) died unexpectedly at home eleven months after surgery.

Postoperative care was relatively uncomplicated. All patients were weaned easily from cardiopulmonary bypass, with nearly normal initial postoperative arterial oxygen saturations (on 100% oxygen). Over the next few days a gradual decline in arterial oxygen saturation (fig. 5) developed which, as demonstrated in several patients by a difference in the simultaneous pulmonary venous atrial and systemic arterial saturations, resulted from intracardiac right-to-left shunting, presumably on the basis of a transient increase in the pulmonary vascular resistance. This was well tolerated by our patients, who were “accustomed” to low arterial oxygen saturation, and soon resolved. The presence of significant right-to-left shunting in the early postoperative period demonstrated the patients’ dependence upon an open ventricular septum to allow decompression of the pulmonary circulation. Furthermore, the persistence of this dependence upon right-to-left shunting was demonstrated later in four patients at postoperative catheterization and in two patients at graded bicycle exercise.
dystrophy and cutaneous angiokeratoma. Renal function studies were normal at that time.

At age 46 years, he experienced the onset of anorexia, nausea, pleuritis, nocturia and swelling of his face and ankles. Subsequently, renal failure developed and the patient was referred to the University of Minnesota Hospitals for biochemical confirmation of Fabry disease (see table 1), dialysis and renal transplantation. Physical examination showed his blood pressure to be 170/60 mm Hg. A grade III/VI blowing apical systolic murmur was present and thought to represent mitral insufficiency. No diastolic murmur was heard. The electrocardiogram showed sinus tachycardia, sinus arrhythmia, left ventricular hypertrophy and strain. There were numerous unifocal ventricular beats. Thoracic roentgenogram showed slight cardiomegaly.

The patient underwent unilateral nephrectomy and splenectomy. He was maintained on chronic hemodialysis and subsequently received a cadaver renal allograft. His postoperative course was complicated by pneumonia and then by a severe rejection episode which did not respond to antirejection therapy. The allograft was removed; he was again maintained on chronic hemodialysis and subsequently he underwent another cadaver allograft transplantation. Shortly thereafter the allograft was rejected, his condition deteriorated, sepsis developed and he died.

Postmortem Examination

Postmortem examinations were performed in patients 1 and 2. The tissues of each patient were studied by both histochemical and ultrastructural techniques. In addition, the concentrations of the accumulated glycosphingolipids in Fabry disease were quantitated in portions of the cardiac valves, atria, ventricles, lung tissue, and selected arterial vessels from patient 1.

Anatomic Findings

The anatomic findings were similar in both patients. The hearts weighed 800 and 440 grams, respectively; in patient 1 each heart chamber was enlarged, while in patient 2 the left and right ventricles were enlarged. Interchordal hooding of the tricuspid valvular leaflets was present but the chordae tendineae and papillary muscles were normal. The right ventricles were dilated and in patient 1 focal scarring of the endocardium was present and the free wall of the right ventricle was hypertrophied (1.0 cm in thickness). The pulmonary valves were normal.

In patient 1, the left atrium was enlarged and showed endocardial thickening; the left ventricle was enlarged, hypertrophied, and showed moderate to severe endocardial thickening. A jet lesion was present on the posterior wall of the left atrium. The mitral valve showed thickening and hooding of the leaflets (fig. 1 left). A large round defect (1.2 cm in diameter) was present in the anterior mitral leaflet. Two tags of tissue protruded into the lumen of this defect. The chordae were normal and the papillary muscles were thickened. Marked thickening of the left ventricular wall was present; the free wall measured 3.5 cm in thickness. Focal scarring was present on the anterior free wall. A small area of calcification, 2 × 3 mm, was present in the region of the attachment of the anterior mitral leaflet close to the base of the aortic valve. The aortic valve had three cusps which showed moderate calcification (fig. 1 right). Several small excrescences of calcification were present on the inside of the cusps. A small defect, 2 mm × 1.5 mm, was present in the right aortic cusp. An area of hemorrhage (healed vegetations) was present on the posterior cusp.

In patient 2, the left atrium was normal, but the left ventricle was enlarged and moderately hypertrophied. The free wall of the left ventricle measured 1.8 cm in thickness. The leaflets of the mitral valve were slightly thickened and showed moderate interchordal hooding (fig. 2); the chordae tendineae were normal and the papillary muscles were thickened and shortened. The aortic valve was normal.

The aorta of patient 1 was thickened and moderately dilated and the coronary arteries showed grade I atherosclerotic changes. In patient 2, the coronary arteries appeared normal.

Figure 1. Left) Mitral valve from patient 1 showing thickening and hooing of the anterior leaflet. Perforation in the anterior leaflet presumably resulted from bacterial endocarditis; chordae tendineae were normal. Right) Aortic valve from patient 1 showing three cusps and moderate calcification.
following surgery, presumably of an arrhythmia. An autopsy was not obtained. Two children (4 and 6) were severely limited prior to operation, and following surgery their behavior and activity are described as normal by their parents. One nineteen-year-old (7) who was severely limited is now employed as a carpenter. One patient (2) developed the sick sinus syndrome postoperatively and required a demand pacemaker; nonetheless, his subjective exercise capacity has considerably improved. Growth among the eight patients was not altered appreciably; during the brief follow-up period of five to 32 months, seven of the eight patients have remained at their preoperative height and weight level (\( \pm 3\% \) in 6/7). A five-year-old girl (4) has increased from the third to the tenth percentile in height and weight.

The mean preoperative hematocrit was 64.3 \( \pm 6.0\% \) (range 54.33–69.7\%) and when measured in the late postoperative period had significantly \((P \leq .01)\) decreased to mean 47.0 \( \pm 4.5\% \) (range 42.4–50.9\%). The three patients with thrombocytopenia prior to surgery had normal platelets postoperatively as estimated by peripheral blood smears.

Pre- and postoperative bicycle ergometry exercise was attempted in two patients. Patient 2, although demonstrating subjective improvement as well as increased arterial oxygen saturation following surgery, was unable to complete the exercise study either before or after surgery. Patient 7 (fig. 5) successfully completed the exercise study. Prior to surgery, exercise was terminated at a level of 20\% predicted workload on account of severe hypoxemia (pO\(_2\), 23 mm Hg) and metabolic acidosis. Following surgery, the patient demonstrated a threefold increase in exercise capacity (fig. 5); hypoxemia and acidosis appeared at higher levels of effort. The patient is back at work as a carpenter and reports no major difficulty.

**Physiologic**

Figure 6 illustrates the pre- and postoperative ascending aorta (AAO) and pulmonary arterial oxygen saturations. The mean systemic arterial oxygen saturation increased markedly following surgery \((P < .001)\), rising from 71 \( \pm 7\% \) to 91 \( \pm 4\% \), and exceeded slightly the pulmonary arterial oxygen saturation measured prior to surgery. As expected, the pulmonary arterial saturation in the four patients catheterized after surgery declined, although the change was not so great as observed in the systemic arterial oxygen saturation.

Figure 7 portrays the systemic blood flow and pulmonary blood flow in the five preoperative patients without a significant PDA (patients 1–5) and in the four patients postoperatively (two of whom, 6 and 7, had a PDA divided at operation). While the absolute pulmonary and systemic flows did not change appreciably in three of the four patients with follow-up catheterizations, the low preoperative effective pulmonary blood flow of 1.1 to 1.9 L/min/m\(^2\) rose to normal levels (2.7–3.4 L/min/m\(^2\), \( P < 0.001 \)) in all four.

Tolazoline failed to change calculated flows and resistances both in the one preoperative patient and in the two postoperative patients. Oxygen administration, on the other hand, increased the pulmonary arterial saturation in both patients studied, indicating an increase in pulmonary arterial flow, and a reduction in the pulmonary resistance.

Mean pulmonary arterial pressure was equal to or greater than 75% of systemic pressure in all patients before the operation, including one patient (2) with an ineffective pulmonary artery band (fig. 8, table 2). There was little change in the four patients evaluated after surgery. In three (1, 2, 4) of the five patients (1–5) in whom both pulmonary
blood flow and mean pulmonary arterial pressure could be measured preoperatively, pulmonary vascular resistance equaled or exceeded 50% systemic. In one (3), pulmonary vascular resistance could not be calculated because of right pulmonary arterial stenosis; lung biopsies taken at surgery disclosed significant Grade III and IV pulmonary vascular obstructive disease in the right and left lungs, respectively. In the fifth patient (5), pulmonary vascular resistance was one-third of systemic; however, bilateral lung biopsies obtained three years previously (following an atrial septectomy) disclosed Grade III changes in both pulmonary vascular beds.

**Discussion**

Medical therapy for patients with the complex physiology of transposition and pulmonary vascular obstructive disease fails to alleviate symptoms and prevent development of complications. Total repair, in the presence of severe pulmonary vascular obstructive disease, is associated with an unacceptably high mortality. A recent report from this institution described symptomatic improvement as well as increased arterial oxygen saturation in a similar group of patients after surgical atrial septectomy. This procedure is particularly useful in patients with atresia or stenosis of an...
atrioventricular valve who have an inadequate interatrial communication. In contrast, patients similar to those under discussion should be more improved by a palliative Mustard procedure.

Information derived from these patients, along with the experience of others,14, 19-28 clearly demonstrates an increase in the systemic arterial oxygen saturation following a palliative Mustard procedure. All four patients evaluated postoperatively increased their ascending aortic saturations in excess of what would be expected from a simple exchange of pulmonary arterial and systemic arterial oxygen saturation (table 2, fig. 6). Pulmonary arterial pressure and pulmonary blood flow are not altered by the operation, whereas systemic blood flow tends to decrease somewhat (fig. 7). As expected, pulmonary vascular resistance, although not calculated in four of the eight postoperative patients, remains is unchanged at systemic levels in three of the four postoperative patients, and is unchanged in the other one.

The major hemodynamic alteration that occurs following the placement of the intra-atrial baffle is the net gain (greater than simple reversal of systemic and pulmonary saturations) in the systemic arterial oxygen saturation. This finding may be attributed to the twofold increase in the effective pulmonary blood flow (fig. 7) and, in part, to the reduction in the amount of systemic venous blood recirculated directly to the aorta, related to an increase in systemic vascular resistance (illustrated by patient 2 in our series).26 These two factors produce a near reversal in the relative proportions of fully saturated pulmonary venous blood and desaturated systemic mixed venous blood reaching the aorta.26 Thus, these patients are converted from individuals with parallel circulations to individuals with circulations in series, associated with a small bidirectional shunt, pulmonary arterial hypertension and pulmonary vascular obstructive disease, in effect, the Eisenmenger syndrome.15, 26

The well-documented and difficult to manage abnormalities in the hemostatic mechanisms17-20 of these patients and the associated risk of cerebrovascular accidents21 are both favorably altered by a reduction in hematocrit and return to normal coagulation.24

The hallmark of suitability for operation appears to be systemic arterial (AAO) to pulmonary arterial (PA) oxygen saturation difference of approximately 10% in the absence of mixing through atrial or great vessel communications. Two patients (5 and 8) had an AO-PA difference of 6% and 8% respectively. One had had a previous atrial septectomy, allowing considerable interatrial mixing. Following the palliative Mustard he was symptomatically improved with a 15% increase in arterial saturation (5, table 2). The second (8) had a large PDA with significant PA to AO shunt; he also had severe pulmonary regurgitation, which probably increased interventricular mixing during the diastolic regurgitant flow. This patient, who improved the least in terms of symptoms and saturation increase (table 2, 8) died suddenly during the night at home eleven months following surgery.

Selection for either palliative or corrective (Mustard and VSD closure) operations is of critical importance in borderline cases (e.g., those with high pressure in the pulmonary artery, but less than 50% systemic resistance in the pulmonary vascular bed, similar to 5), since patients with TGA and even mild pulmonary vascular obstructive disease experience a 40% mortality following total correction.9 Pharmacological maneuvers such as tolazoline and oxygen administration are of limited value and may yield divergent results; tolazoline administration suggested in our patients a fixed resistance in the pulmonary vascular bed, whereas oxygen administration, given to two of the three tolazoline-treated patients, resulted in an apparent increase in the pulmonary blood flow suggesting a labile pulmonary vascular resistance. At the time of surgery, multiple lung biopsies may afford additional information on the status of the pulmonary vascular bed.

Atrial septectomy has been a useful palliative procedure for patients with similar hemodynamic and anatomic features22; the resulting increase in interatrial bidirectional shunting produces a moderate increase in the systemic arterial oxygen saturation. In contrast, the palliative Mustard procedure reverses and separates the systemic and pulmonary venous returns, resulting in a far greater increase in the effective pulmonary flow, often to normal levels (fig. 7). In comparing the data from this study with those patients who have undergone atrial septectomy at this institution, postoperative arterial oxygen saturations in the palliative Mustard group are higher (mean = 91%) than in the surgical septectomy group (mean = 75%). Surgical mortality and morbidity did not differ appreciably, and three patients who had previously undergone a surgical septectomy underwent a palliative Mustard operation later. Thus, our experience indicates that selected patients with DORV or d-TGA, VSD, PAH, and PVO can be safely and effectively palliated with the Mustard operation, enjoying the benefits of the correction of the transposition physiology (parallel circulations) without being subjected to the risks of concomitant VSD closure. By the same token, one should remember that this is not a corrective procedure, and that the long-term prognosis of these converted Eisenmenger patients is unknown.

Finally, it is interesting to note that although the intracardiac anatomy of d-TGA and DORV may usually be distinguished from one another by angiography (figs. 1, 2), the clinical, physiologic and surgical results do not appear to be related to these anatomic differences. Nonetheless, associated lesions such as PDA, JAA, LSVC, and others are of great importance in the surgical management and should be sought for by thorough preoperative study.

Summary

Eight patients with double outlet right ventricle or complete transposition, ventricular septal defect, severe pulmonary arterial hypertension, and pulmonary vascular obstructive disease are described. A palliative Mustard operation was performed in each one. We believe that this procedure is the treatment of choice in those severely symptomatic patients with 1) severe pulmonary vascular obstruction and severe pulmonary arterial hypertension, and 2) PA saturation greater than SA saturation by approximately 10% (absence of significant interventricular mixing). It should be emphasized, however, that this operation is palliative in both intent and result, is not without risk, and does not alter the pulmonary pathophysiology nor totally correct the intracar-
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diac defects. Nevertheless, it appears that this procedure can be performed in selected patients with reasonable safety and appreciable relief of symptoms.

References

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