

Transposition of the Great Arteries and Intact Ventricular Septum: Anatomical Repair in the Neonate

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ABSTRACT Fourteen neonates 18 hours to 32 days old with transposition of the great arteries (TGA) and virtually intact ventricular septum (IVS) underwent arterial switch operations under deep hypothermic circulatory arrest. Preoperative left ventricular to right ventricular peak systolic pressure ratios ranged from 0.7 to 1.0 (mean, 0.92), and the echocardiogram showed a centrally positioned ventricular septum in 10 patients and a rightward displaced ventricular septum in 4. One patient died twelve hours after operation. Postoperative complications included bleeding from the left coronary artery-pulmonary artery anastomosis (1 patient), stenosis of the pulmonary artery-aorta anastomosis requiring reoperation (2 patients), transient ST segment and T wave abnormalities consistent with ischemia (3), and development of pathological Q waves suggestive of clinically silent infarction (2).

The capacity of the left ventricle in a neonate to effectively take over the systemic circulation was clearly demonstrated. A longer follow-up period is needed to assess late ventricular function, coronary ostial growth, growth of the aorta-pulmonary artery anastomosis, late aortic valve (anatomical pulmonary valve) function before definitive recommendations about the superiority of the arterial switch operation in neonates with TGA plus IVS can be formulated.

Intraatrial repair by the Senning or Mustard technique has dramatically enhanced the prospects for survival of children with transposition of the great arteries (TGA). Over the years, the operative mortality and the incidence of many late complications of these operations have decreased significantly. Supported by the observation that right ventricular (RV) failure or tricuspid regurgitation develops late in some patients after atrial inversion repair, an impression exists that the right ventricle is somehow inherently different from the left and may not be well suited to support the systemic circulation indefinitely. Indeed, there is suggestive evidence that the right ventricle in the systemic circulation responds

differently from the left when challenged with increased afterload by the infusion of methoxamine hydrochloride [1]. Also, atrial dysrhythmias related to dysfunction of the sinus and atrioventricular nodes are associated with serious morbidity and mortality in a significant number of patients who have undergone atrial inversion repair of TGA.

Jatene's successful arterial switch operation in 1975 resulted in a renewed and increasing interest in anatomical repair of TGA [2]. Jatene recognized early in his experience that a left ventricle that has developed for some time in the low-resistance pulmonary circuit may fail acutely following an arterial switch operation. Consequently, he recommended the operation principally for patients with TGA and a ventricular septal defect (VSD) or a large patent ductus arteriosus (PDA). Since TGA most often is not associated with either of these anomalies, for anatomical repair to have wide application in the management of TGA, a more comprehensive selection process is necessary. Yacoub and associates [3] have investigated the concept of preparing the left ventricle for systemic pressure work in patients with TGA and intact ventricular septum (IVS) by preliminary banding of the pulmonary artery followed later by debanding and an arterial switch operation. Alternatively, it can be reasoned that the left ventricle of the newborn should be well suited for systemic function, as it has supported systemic pressure during gestation.

This report constitutes our initial experience with the arterial switch operation in neonates with TGA and IVS.

Material and Methods

From May, 1983, through December 1, 1983, 14 consecutive neonates with TGA {S,D,D} and IVS underwent an arterial switch operation. There were 9 boys and 5 girls. They ranged in age from 18 hours to 32 days (mean age, 6.6 days) and in weight from 1.8 to 4.4 kg (mean weight, 3.4 kg). Ten of these neonates (71.4%) underwent the arterial switch operation during the first week of life. Ratios of RV to left ventricular (LV) pressure ranged from 0.7 to 1.0 (mean, 0.92) (Table). Associated cardiovascular lesions included PDA (10 patients), a small VSD (2), coarctation of the aorta (1), and left juxtaposition of the atrial appendages and dextrocardia (1).

Before the arterial switch operation, 8 patients had a balloon atrial septostomy, 9 received prostaglandin E₁, and 1 had resection of coarctation of the aorta. The preoperative electrocardiogram demonstrated right-axis deviation and pure R wave in V₁ in 12 and an R wave/S wave ratio of less than 1 in V₆ in 9 neonates.

All patients underwent preoperative echocardi-

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Preoperative Left Ventricular and Right Ventricular Peak Systolic Pressures and Pressure Ratios

Age at Operation (day)	LV Pressure (mm Hg)	RV Pressure (mm Hg)	LV/RV Pressure Ratio
1	50	52	0.9
2	52	55	0.9
2	48	48	1.0
2	70	70	1.0
2	65	68	0.9
2	70	70	1.0
2	50	52	0.9
4	75	75	1.0
5	60	62	0.9
7	70	95	0.7
10	60	78	0.7
10	62	62	1.0
14	58	67	0.8
32	82	92	0.8
6.6	62.2	67.5	0.92

LV = left ventricular; RV = right ventricular.

graphic evaluation and heart catheterization; LV and RV pressures were measured in all patients, and the LV/RV pressure ratios were calculated (see Table). The echocardiogram showed a rightward displacement of the ventricular septum (type I) in 4 patients (28.6%) and a centrally positioned ventricular septum (type II) in 10 patients (71.4%). None had a type III or leftward displaced septum.

All infants were intubated and artificially ventilated for a minimum of twenty-four hours after operation. Postoperative assessment included continuous monitoring of systemic arterial blood pressure, right and left atrial pressures, and the ECG.

Surgical Technique

Deep hypothermic circulatory arrest was used for all neonates. Following a median sternotomy, cardiopulmonary bypass was instituted by cannulation of the distal ascending aorta and placement of a single venous cannula through the right atrial appendage. By means of a heat exchanger interposed on cardiopulmonary bypass, core cooling was carried out with cold (15°C) perfusion until the nasopharyngeal and rectal temperatures reached 20°C. During core cooling, the ductus arteriosus was divided and both the left and right pulmonary arteries were dissected beyond their point of branching. At 20°C the aorta was clamped, cardioplegic solution was infused, and cardiopulmonary bypass was stopped. The duration of circulatory arrest ranged from 47 to 64 minutes. The right atrial cannula was then removed. In those patients who had a balloon atrial septostomy, the atrial septal defect was closed.

While the patient was on bypass, the coronary arteries

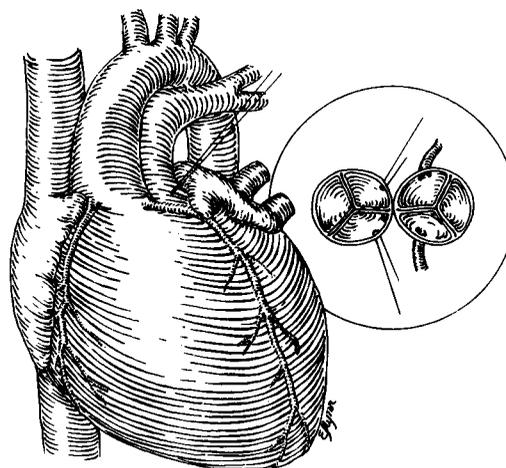


Fig 1. Sites of implantation of the left and right coronary arteries on the pulmonary artery are marked by reference stitches.

were inspected and the sites where the left and right coronary arteries were to be implanted on the pulmonary artery were marked by reference stitches (Fig 1).

During circulatory arrest, the aorta was transected approximately 1 cm distal to the coronary arteries and the main pulmonary artery was divided in its midportion. In the neonates, we encountered little discrepancy between the sizes of the great arteries. The left and right coronary ostia were excised using either a button of aortic wall (Fig 2A) or a segment of aortic wall extending from the rim of the aortotomy incision to the base of the coronary sinus (Fig 2B). We made certain to include the largest possible segment of aortic wall around the coronary ostium. An equivalent oval segment of pulmonary arterial wall (previously oval) (Fig 3) was then excised, and the coronary arteries were sutured to the pulmonary artery using a continuous 7-0 polydioxanone (PDS) suture material. Next, the distal pulmonary artery was brought anterior to the ascending aorta (Lecompte maneuver) [4], and the proximal pulmonary artery was anastomosed to the distal aorta using a continuous 6-0 PDS suture (Fig 4). At this point in the operation, the right atrial cannula was replaced and cardiopulmonary bypass was restarted. During rewarming, the sites of explantation of the coronary arteries were repaired using either a patch of circular Gore-Tex (Fig 5A) or a segment of pericardium (Fig 5B). Finally, the proximal aorta was sutured directly to the distal pulmonary artery with a continuous 6-0 PDS suture (see Fig 5). At a rectal temperature of 36°C, cardiopulmonary bypass was discontinued.

Results

Deaths

There have been 1 hospital death and no late deaths. The 7-day-old patient died twelve hours after operation. This patient had an accessory right coronary orifice supplying a rather large right conal branch. In the attempt to

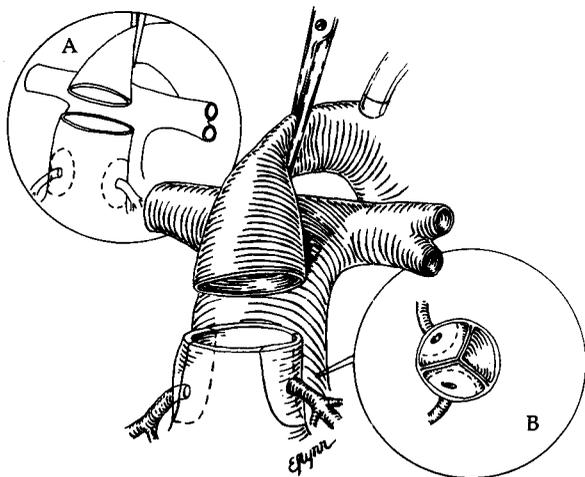


Fig 2. The aorta is transected and the left and right coronary arteries are excised using (A) either a button of aortic wall or (B) a segment of aortic wall extending from the rim of the aorta.

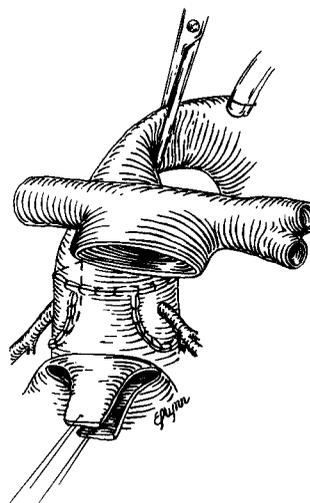


Fig 4. The distal pulmonary artery is brought anterior to the ascending aorta, and the proximal pulmonary artery is anastomosed to the distal aorta.

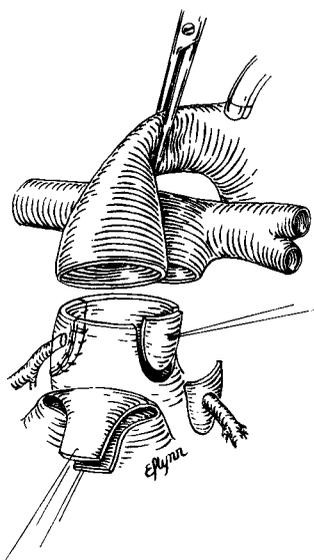


Fig 3. An equivalent segment of pulmonary arterial wall (previously marked) is excised, and the coronary arteries are sutured to the pulmonary artery.

include this conal branch with the main right coronary artery, undue tension on and some distortion of the right coronary artery-pulmonary artery anastomosis occurred. Right coronary flow was compromised; the child remained in a low-output state. At the time of cardiac arrest, the right ventricle was ischemic. The postmortem examination confirmed the distortion and partial occlusion of the right coronary artery.

Complications

One patient required reoperation after four hours because of bleeding from the left coronary artery-pulmonary artery anastomosis. Among the first 5 patients seen, severe stenosis developed at the pulmonary

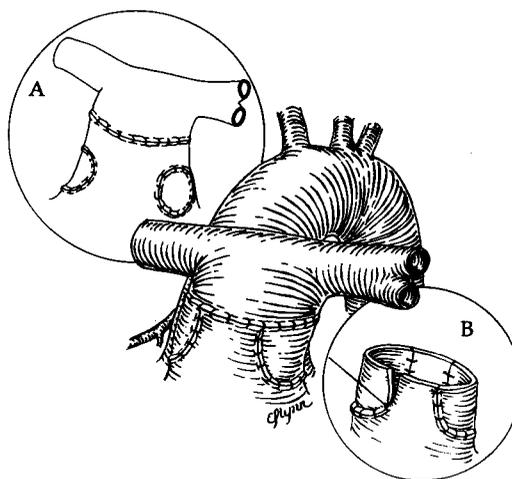


Fig 5. The sites of coronary artery explantation are repaired using either (A) a patch of Gore-Tex or (B) a segment of pericardium. Finally, the proximal aorta is sutured to the distal pulmonary artery.

artery-aorta anastomosis in 2. In both, cardiac catheterization revealed suprasystemic RV pressures. Angiograms showed a localized obstruction at the anastomotic site. A two-dimensional echocardiogram demonstrated leftward displacement (type III) of the ventricular septum. Both patients underwent successful reoperation four and five months after the original operation. The supravalvular pulmonary stenosis was relieved by placement of a Gore-Tex patch.

Electrocardiographic Findings

In 11 of the 13 survivors, the ECG showed a marked increase in the R wave/S wave ratio in lead V_6 during the immediate postoperative period. It occurred in 3 pa-

tients by day 2, in 2 by day 3, and in the remainder by day 7.

The initial QRS vector was altered in 8 patients; 5 showed new Q waves in leads II, III, and aV_r , and 3 with preexisting Q waves in those leads had larger Q waves postoperatively. In none of these 8 patients were other signs of ischemia or infarction present.

In 3 infants, transient ischemic ST segment and T wave changes were noted on days 1 through 3 postoperatively. These changes were accompanied by the appearance of new Q waves suggestive of infarction in 2 of them.

Comment

In normally related great arteries, there is a linear correlation between the thickness of the LV free wall and the age of the patient [5]. No such correlation has been demonstrated between RV free wall thickness and age. The peak systolic wall stress may be a major stimulus for increasing ventricular wall thickness. As LV dimensions and systemic blood pressure increase with growth, the LV wall thickness correspondingly increases to maintain a constant peak systolic wall stress. Since the pulmonary artery pressure decreases after birth, there is no such stimulus for increasing RV wall thickness.

In patients with TGA, the right ventricle is the systemic ventricle and the left ventricle is the pulmonary ventricle; the two circulations are in parallel. At birth, LV free wall thickness is similar to that in normal hearts.

After birth, as pulmonary vascular resistance and pressure decrease, the "normal" increase in LV wall thickness is curtailed. The curtailment is more marked in simple TGA plus IVS than in TGA with VSD and LV hypertension [6]. Left ventricular to RV pressure ratios in neonates with TGA plus IVS also decrease appreciably (from a mean of 0.80 to a mean of 0.45) by 6 months of age. The progressive decline in LV peak systolic pressure and the subsequent reduction in ratio of LV free wall thickness to chamber dimension may limit LV pressure work capabilities.

Furthermore, in normal hearts the ventricular septum is functionally committed to the left ventricle; thus, the prolate ellipsoid configuration of the left ventricle is ideally suited to minimize wall tension. Reversal of septal curvature, demonstrated echocardiographically in patients with either RV volume or pressure overload, has also been noted in infants with TGA plus IVS [7]. End-diastolic and end-systolic minor-axis ratios and the radius of curvature of the ventricular septum determined by two-dimensional echocardiography show a significant correlation with LV/RV peak systolic pressure ratios. Equilibration of LV and RV pressures in the fetus maintains LV geometry until after birth when the increase in systemic and the decrease in pulmonary artery pressure eventually cause a rightward displacement of the ventricular septum (type I). Type II, or intermediate position, and type III, leftward deviation of the ventricular septum, indicate either a volume or pressure over-

load of the right ventricle. Among the neonates in this series, type II predominated, as anticipated; none had type III septal configuration. It is of interest that in a 3-month-old infant with TGA and IVS (not included in this series of neonates) who underwent an arterial switch operation, the left ventricle failed to maintain an effective cardiac output after operation. The arterial switch operation had to be undone and converted (successfully) to a Senning repair. The preoperative echocardiogram in this patient showed a type III septal configuration. At present we know of no reliable test capable of predicting at what pressure the left ventricle becomes inadequate to sustain the systemic circulation. After the establishment of more clinical correlations and further refinements in technique, including mechanical or pharmacological afterload challenge of the left ventricle, echocardiographic definition of septal position may prove a valuable noninvasive marker to evaluate the capability of the left ventricle to perform systemic pressure work.

Left ventricular forces on the ECG increased dramatically in the majority of patients by the seventh postoperative day, corresponding to the change in relative work loads of the two ventricles. The changes in relative amplitudes of the R and S waves in the lateral precordial leads are comparable with those reported in normal infants during the first week of life [8].

In 8 patients, narrow, nonpathological Q waves developed or increased in size in leads II, III, and aV_r and were associated with a leftward shift in axis. None of these patients exhibited other evidence of ischemia or infarction. This may be an additional manifestation of the LV work load. Wide pathological Q waves developed in 2 patients, suggesting infarction. However, the hospital course of these patients did not differ from that of the other 11 patients.

Of the several potential complications after arterial switch operations in neonates, the most worrisome is that the coronary artery-pulmonary artery anastomosis may limit subsequent coronary ostial growth. Experimental evidence indicates that the inclusion of a large segment of aortic wall around the coronary orifice [9] and perhaps also the use of absorbable suture material (PDS) will prevent this potential complication. Also, the aorta-pulmonary artery anastomosis is a potential site for late obstructions. The early supravalvular pulmonary stenosis that occurred in 2 of our patients was related to undue tension and secondary flattening of the pulmonary artery because of inadequate mobilization of the distal pulmonary arteries. In both instances the stenosis was repaired easily by interposing a small patch across the narrowed anastomosis. In this age group, the use of prosthetic material to bridge the gap between the proximal aorta and the distal pulmonary artery must be discouraged. To date, we have been able to accomplish a direct anastomosis in all neonates. The Lecompte maneuver [4], anteposing the pulmonary artery to the ascending aorta, has greatly facilitated primary reconstruction of the pulmonary artery. An additional advantage in the neonatal group in contrast to older infants is the

minimal discrepancy in size between the pulmonary artery and the ascending aorta.

The single operative death in this series was due to an error in judgment. In retrospect, the accessory conal branch should have been sacrificed to avoid undue stress on the right coronary artery.

The capacity of the left ventricle in the neonate to effectively take over the systemic circulation was demonstrated by the low operative mortality and by the fact that almost none of the patients required inotropic support after operation. The stay in the intensive care unit was short and hospitalization averaged a week. We are encouraged by these initial results of arterial switch operations in such very young patients. However, a much longer follow-up period is necessary to assess late ventricular function, growth of the coronary ostium, growth of the aorta-pulmonary artery anastomosis, and late aortic valve (anatomical pulmonary valve) function before definitive recommendations about the arterial switch operation in neonates with TGA and IVS can be formulated.

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Discussion

DR. HILLEL LAKS (Los Angeles, CA): It is a privilege to discuss this important report presented by my surgical teacher, Dr. Castaneda. This paper represents a technical breakthrough, and we must ask the question of whether there is a better alternative to the hitherto standard intraatrial baffle repair for simple TGA.

Since it was proposed by Jatene in 1975, the arterial switch operation has found its greatest application in infants with TGA and large VSD. This is understandable, as the combined Senning operation and VSD closure has an early mortality of about 15%, as recently reported by the Boston group, and is associated with late morbidity and mortality. The switch operation for this indication was reported recently to have a mortality as low as 11%.

In 1980, Paul Ebert and the group in San Francisco embarked on an innovative program of performing the arterial switch procedure in the first days of life while the left ventricle, as Dr. Castaneda has explained, is still equipped to tolerate the systemic pressures. Shortly before this meeting, Dr. Ebert was kind enough to share his experience with me and agreed to have it presented in this discussion.

Two of the 6 patients died early, both deaths probably due to coronary artery distortion. One infant required reoperation for stenosis of the pulmonary anastomosis. One year after operation, previously undiagnosed pulmonary valve (that is, the new aortic valve) regurgitation developed in 2 other patients. Such regurgitation has been reported from Toronto by Drs. Williams and Trusler in a series of older children. Dr. Ebert has now discontinued this program.

The standard treatment for simple transposition is currently intraatrial baffle repair. Both the Mustard and Senning procedures have long track records and serve as a basis of comparison for newer procedures. The early mortality is now fairly standard. In several series it is 5% or less. In addition, late survival has been excellent. In his more recent experience, Trusler reported only 2 early deaths among 100 patients and in his earlier experience, there was very little falloff in the late follow-up. In the Boston experience, there are similar results with both the Senning and the Mustard operations. The intraatrial baffle repair, however, does have late sequelae, including pulmonary venous obstruction, possible RV failure and tricuspid regurgitation, and arrhythmias, as outlined in the excellent series of Bodnar and associates comparing the Mustard and Senning procedures.

The arterial switch, in contrast, promised the possibility of a cure. It is a procedure without late sequelae.

I have several questions for Dr. Castaneda. First, what will be the achievable operative mortality for this difficult procedure in the first days of life? In this series, there was 1 death out of a possible 14, which gives a mortality of 7% but 0 to 21% within the 70% confidence limit. Combining this series with that of Dr. Ebert, there were 3 deaths among 20 patients, a mortality of 15%.

Second, what will be the incidence of anastomotic stenosis? It occurred in 2 of Dr. Castaneda's 13 survivors and in 1 of Dr. Ebert's 4 survivors, an incidence of 18 percent. What will be the incidence of pulmonary valve regurgitation and of late coronary stenosis?

Third, what about the development of late LV dysfunction? Three patients in this series had ST-T wave changes, and 2 of them had pathological Q waves.

Dr. Castaneda, would you comment on the technical question of whether Dr. Pacifico's suggestion of scalloping the distal pulmonary artery instead of closing the explantation site with a patch is applicable in the neonate.

All innovative surgical procedures raise serious bioethical questions, particularly when a relatively safe alternative is available. Certainly, this excellent series makes it reasonable to continue with this experience. I again congratulate Dr. Castaneda on this outstanding surgical achievement.

DR. WILLIAM G. WILLIAMS (Toronto, Ont, Canada): Dr. Ceithaml and Dr. Castaneda have presented two very different techniques of arterial repair, one for complete transposition and one for the Taussig-Bing type of double-outlet right ventricle.

Survival after the Damus-Kaye-Stansel procedure in the Mayo series is disappointing, but I am sure we all recognize their pioneering effort in developing this new technique. My associates and I use this operation as the procedure of choice for repair of the Taussig-Bing anomaly, particularly when the arteries are side by side.

Our experience in Toronto with arterial repair of the Taussig-Bing anomaly consists of 8 infants and children. In the first 4 we used Jatene's technique; only one outcome was successful. Subsequently, 4 others underwent repair by the Damus-Kaye-Stansel procedure; 3 survived. Obviously this is not a valid statistical comparison, but we plan to continue using the latter approach in managing this particularly difficult form of double-outlet right ventricle.

Our experience with arterial repair consists of 33 patients. Overall mortality is substantial—27%—but undoubtedly can be improved with increasing experience. Our primary indication is transposition with a large VSD and without pulmonary stenosis, and was seen in 19 of the 33 patients. Three of the 4 deaths in these 19 patients were attributed to variation in coronary artery branching, that is, a single coronary artery in 1 and a circumflex branch arising from the right in 2. These anomalies have subsequently been managed successfully with Yacoub's technique of patching one side of the coronary artery button. Reverting to the Damus-Kaye-Stansel operation in this situation may not be necessary.

We have not utilized arterial repair for TGA with IVS except in the unusual circumstance in which there is a high LV pressure. Previously we thought the magnitude of this operation precluded arterial repair for the newborn. We are indebted to Dr. Castaneda's group for demonstrating the feasibility of this approach, at least in his exceptionally capable hands.

I have two practical questions for Dr. Castaneda. Why was balloon septostomy not done in infants who had a partial pressure of oxygen of less than 25 mm Hg? Do you have any special techniques of myocardial protection other than a very short cross-clamp time?

Thank you for the privilege of discussing these brilliant papers.

DR. CASTANEDA: Dr. Laks and Dr. Williams, thank you for your encouraging comments. Concerning the question of late complications, our postoperative protocol provides for one-year and five-year postoperative studies, including cardiac catheterization and cineangiography. To date, aortic regurgitation has not been identified clinically in any of the patients.

We prefer to fill the explanted areas within the proximal aorta with patches of pericardium to facilitate tailoring of the anastomosis and also to prevent undue tension.

During the initial catheterization, atrial septostomy is added only for neonates who are in metabolic acidosis or are severely hypoxic (partial pressure of oxygen of less than 25 mm Hg). Otherwise, atrial septostomy is avoided to eliminate the need for closure of the atrial septum during the switch operation.