

# Corrected Transposition: Anatomic Repair Using the Hemi-Mustard Atrial Baffle and Bidirectional Superior Cavopulmonary Connection



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Anatomic repair for congenitally corrected transposition requires thoughtful surgical planning at both initial (neonatal or late) presentation, and during definitive repair. An algorithmic approach to the overall management of this lesion, with its many associated intracardiac anomalies, is presented. Modified atrial switch with bidirectional superior cavopulmonary connection is commonly utilized and demonstrates favorable results through a 20-year experience. Herein, technical considerations learned during the implementation of this strategy are described and emphasize the concept, in selected cases, of native pulmonary root preservation by translocation as an adjunct that is uniquely suited by adopting this approach.

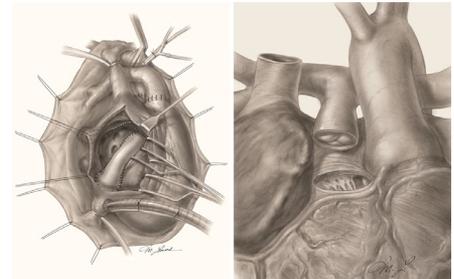
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Management for corrected transposition continues to evolve. Historically, addressing associated lesions (ie, ventricular septal defect (VSD), systemic tricuspid regurgitation, pulmonary stenosis) and preserving ventricular discordance, that is physiologic repair, was the mainstay of surgical therapy. An impetus to restore the morphologic left ventricle to the systemic circulation prompted the development of anatomic repair, through a combined atrial and arterial double switch operation. Several recent series demonstrate that there is great variability in both the approach and outcome to which individual centers ascribe and achieve [1–6].

In 2011, our group introduced a variant approach to the anatomic repair, that is, modified atrial switch via hemi-Mustard with bidirectional superior cavopulmonary connection (SCPC), and reported on our initial experience with 48 patients over a 15-year period [7,8].

Herein, we describe nuances learned during the implementation of this technique and emphasize the concept, in selected



Hemi-Mustard creation and main pulmonary artery translocation.

## Central Message

Corrected transposition requires thoughtful surgical planning at initial presentation through to definitive repair. This can be effectively accomplished through a modified atrial switch procedure, incorporating the hemi-Mustard and bidirectional superior cavopulmonary connection, and in select instances, with native pulmonary root translocation.

cases, of native pulmonary root preservation by translocation as an adjunct that is uniquely suited by adopting this approach.

## APPROACH

Corrected transposition is associated with several lesions, including pulmonary stenosis (PS) in 60%, VSD in 70%, Ebsteinoid tricuspid valve in 20%, and complete heart block with 1–2% increasing risk per year of life [4,7,8]. Our surgical approach is tailored to the combination of lesions that are associated and summarized in [Table 1](#).

## TECHNICAL CONSIDERATIONS

### Early Interventions

Patients that are duct-dependent (ie, severe PS) may require an aortopulmonary shunt to prevent severe cyanosis. This can be performed in standard fashion.

A pulmonary artery band is utilized in two distinct situations: (1) to control excess pulmonary blood flow (ie, VSD without significant PS) and (2) to train the deconditioned left ventricle (LV) (ie, no VSD without significant PS). Band placement is critical and should be secured to the distal pulmonary trunk in

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# USE OF HEMI-MUSTARD AND BIDIRECTIONAL SUPERIOR CAVOPULMONARY CONNECTION

**Table 1** Stanford Children’s Health Treatment Algorithm for Common Anatomic Variants of Corrected Transposition

Associated Lesion	Physiology	Early Intervention	Definitive Intervention
VSD only	<ul style="list-style-type: none"> <li>• LV remains pressure-loaded</li> <li>• Adequate semilunar valves</li> </ul>	PAB to control PBF	ASO, VSD closure, hemi-Mustard, SCC
VSD/PS	<ul style="list-style-type: none"> <li>• LV remains pressure-loaded</li> <li>• Inadequate neo-Ao valve for ASO</li> </ul>	PAB vs shunt to control PBF vs no intervention	LV-Ao baffle, hemi-Mustard, SCC; RV-PA conduit vs PA translocation
PS only	<ul style="list-style-type: none"> <li>• LV loses pressure-load</li> <li>• Inadequate neo-Ao valve for ASO</li> <li>• Lack of VSD for baffle</li> </ul>	Shunt to achieve adequate PBF vs no intervention	Best candidate for avoiding intervention; modified double switch in unique cases
None	<ul style="list-style-type: none"> <li>• LV loses pressure-load</li> <li>• Adequate semilunar valves</li> </ul>	PAB for LV training	ASO, hemi-Mustard, SCC

Ao, aorta; ASO, arterial switch operation; LV, left ventricle; PA, pulmonary artery; PAB, pulmonary artery band; PBF, pulmonary blood flow; RV, right ventricle; SCC, superior cavopulmonary connection.

situations where the pulmonary valve may be utilized in the future as the neo-aortic valve. Tightness of the pulmonary artery band is titrated in the usual fashion when balancing pulmonary blood flow (ie, monitoring the direction of intracardiac shunt flow, assessing systemic oxygenation saturation and partial pressure). For left ventricular training, intraoperative measurements of the proximal pulmonary root and right atrial pressures are coupled with real-time transesophageal echocardiography to determine ultimate band tightness. The band is tightened with a goal of achieving systemic level pulmonary artery pressure, without causing elevated LV filling pressure (ie, right atrial pressure >8 mm Hg), global or regional wall motion abnormalities, worsening mitral valve regurgitation, and/or increasing arrhythmia burden. In our experience, LV training typically requires an initial band placement that can achieve half to two-thirds systemic level pulmonary artery pressure, and a revision tightening 6 months subsequently, to achieve systemic level [9].

Patients are reassessed every six months, with cardiac magnetic resonance imaging, transthoracic echocardiography, and cardiac catheterization. A patient is considered for definitive double switch repair when the LV can tolerate >90% systemic pressure, LV mass has achieved >60 g/m<sup>2</sup>, mitral regurgitation remains mild or better, LV end-diastolic pressure <8–10 mm Hg, and LV ejection fraction remains >50%.

In select situations, patients with adequate semilunar valves may undergo neonatal definitive repair. Elevated pulmonary vascular resistance precludes use of the SCPC in these cases, and a full Senning atrial switch is performed with the arterial switch procedure.

## Definitive Repair: Double Switch Procedures

We typically delay definitive management to (1) avoid the elevated pulmonary vascular resistance of the neonatal period, thus enabling appropriate antegrade superior cavopulmonary flow, and (2) to reduce the risk of late obstruction that can occur with neonatal repair in those patients requiring an LV-aortic intracardiac baffle. The Ebsteinoid tricuspid valve is usually not addressed surgically unless regurgitation is severe,

as this reliably improves once the valve is positioned in the pulmonary circulation and unloaded via the superior cavopulmonary connection. Heart block is addressed as needed, and prophylactic epicardial pacing wires are placed when any pre-existing evidence of atrioventricular node dysfunction is documented.

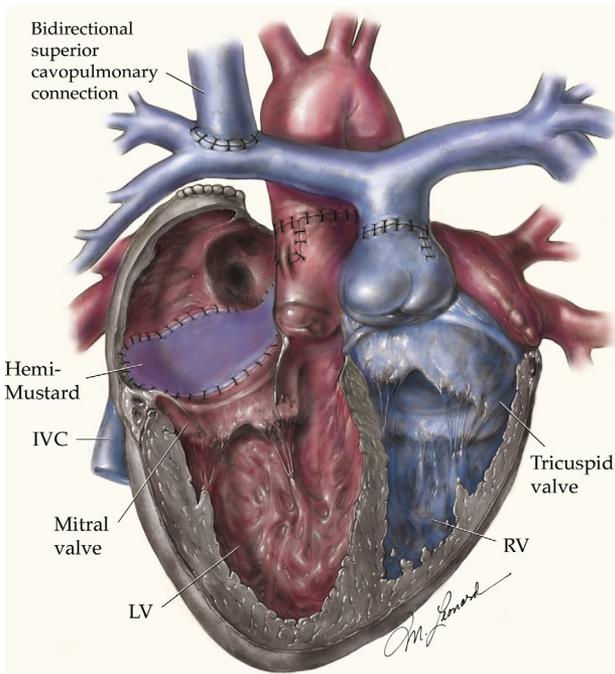
## Cannulation and CPB

Definitive repair is approached via primary or redo sternotomy. Aortic cannulation is performed at the junction of the ascending aorta and innominate artery in the event a LeCompte maneuver is to be performed. The superior vena cava is cannulated at the junction of the innominate vein in preparation for SCPC. The inferior vena cava (IVC) is cannulated at the junction of the hepatic veins in preparation for hemi-Mustard. An LV vent is positioned through the right upper pulmonary vein. Any prior aortopulmonary shunt or pulmonary artery band is dissected and controlled during the initiation of cardiopulmonary bypass. Core cooling to 25–28°C is commenced. Ventriculoarterial reconstruction and hemi-Mustard are performed under aortic cross clamp and cardioplegic arrest. Right ventricular to pulmonary artery (RV-PA) conduit and bidirectional SCPC are performed after the aortic cross clamp is removed during rewarming on cardiopulmonary bypass.

## Creating Ventriculoarterial Concordance

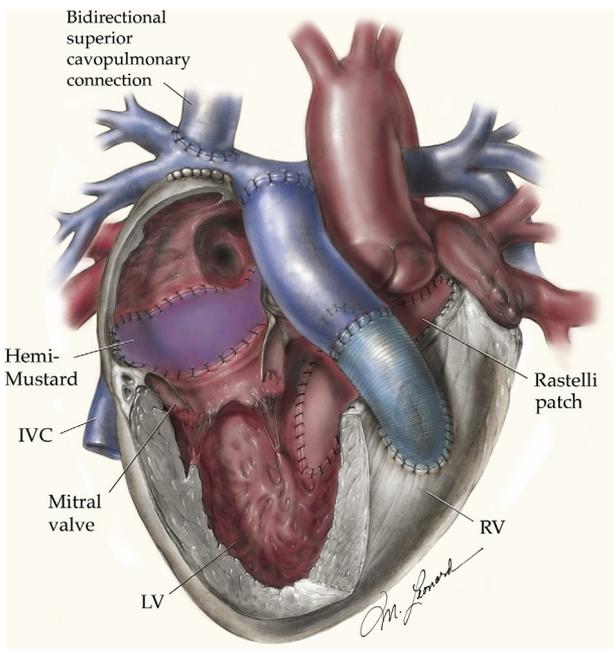
If arterial switch is to be performed (Fig. 1), this is commenced and completed in the standard fashion used for simple transposition. A LeCompte maneuver is not always performed and depends on the relative orientations of the aortic and pulmonary roots.

If an intracardiac LV-aortic baffle is to be performed (Fig. 2), a site for the right ventriculotomy is carefully assessed. Crossing coronary arteries are avoided. An appropriate path for eventual conduit placement is visualized. If pulmonary root translocation is to be considered, greater scrutiny is required, as the native pulmonary artery is limited both in length and in its ability to be repositioned. Generally, the ventriculotomy



**Figure 1** Definitive double switch repair for patients with two adequate semilunar valves. Arterial switch, hemi-Mustard, superior cavopulmonary connection.

should be positioned within the conus of the right ventricle. The VSD is then visualized, and an intracardiac LV-aortic baffle is created with a circular-shaped patch of autologous or bovine



**Figure 2** Definitive double switch repair for patients with an inadequate pulmonary valve (ie, valvar pulmonary stenosis). Left ventricle – aorta baffle, hemi-Mustard, right ventricle – pulmonary artery conduit, superior cavopulmonary connection.

pericardium, bringing the left ventricle in continuity with the transposed native aortic valve.

### Creating Atrioventricular Concordance

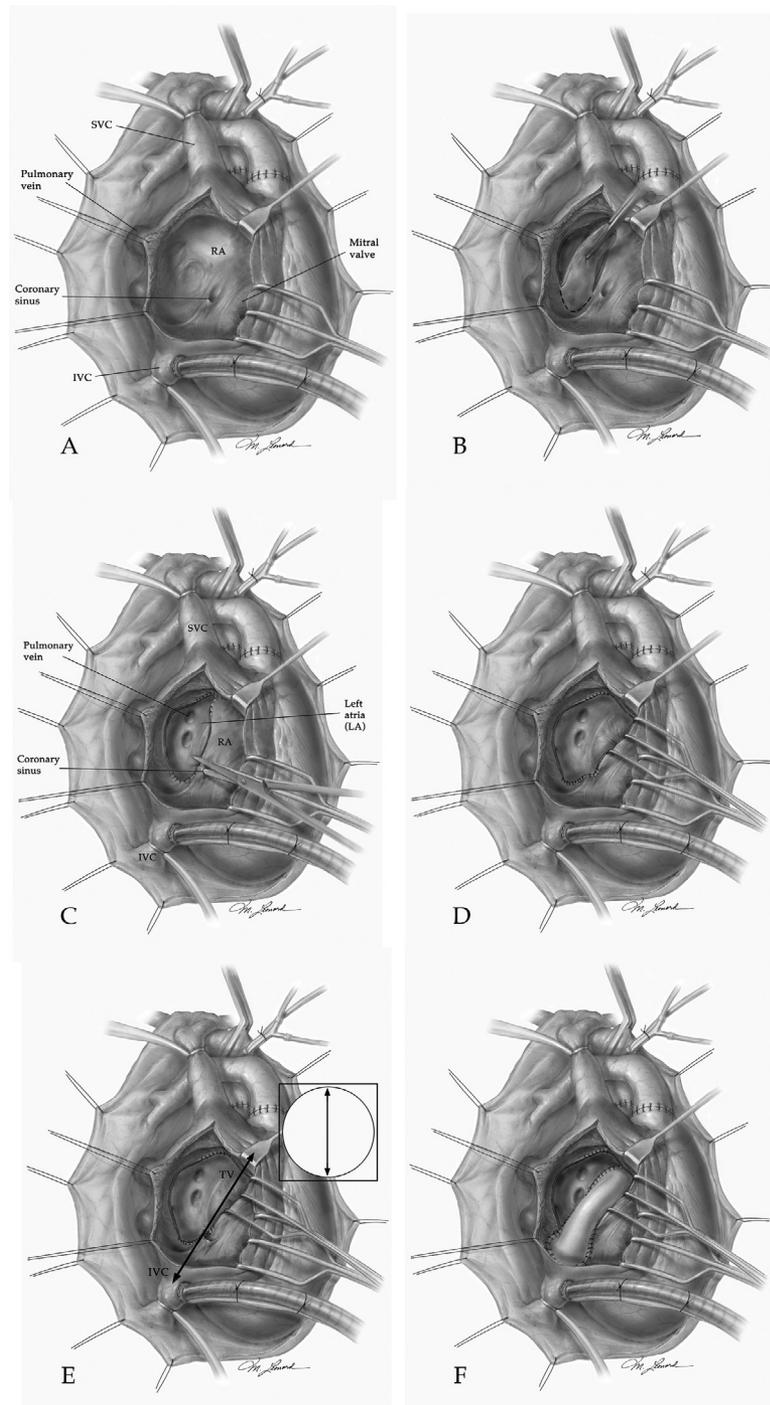
The atrial switch is performed after the ventriculoarterial reconstruction (Fig. 3A–F). A right atriotomy that traverses from the right atrial appendage to the lateral IVC-RA junction enables appropriate visualization of the systemic veins and mitral valve. This incision terminates about 1 cm from the IVC insertion into the right atrium, facilitating placement of the hemi-Mustard patch. The atrial septum primum is then generously excised; this is carried aggressively cephalad and caudad until epicardial fat is entered in both directions. Areas of weakness are reinforced and re-endothelialized with interrupted suture. The creation of an unrestricted common atrium is imperative to prevent obstruction of flow from pulmonary veins to the mitral valve, and IVC to tricuspid valve. A GoreTex patch is then sewn to baffle IVC flow to the more posterior tricuspid valve. This baffle is most vulnerable to obstruction as its path crosses the atrial septal plane. As such, the coronary sinus is unroofed, and the baffle is shaped in a circular fashion to prevent narrowing at its waist. The atriotomy is closed.

The aortic cross clamp is then removed. The superior cavopulmonary connection is performed in the usual fashion. For patients who required an intracardiac baffle repair, a conduit is then placed in the usual fashion. Given the variability in heart position associated with this lesion, the RV-PA conduit may traverse to the left or right of the neo-aortic root.

### Pulmonary Root Translocation

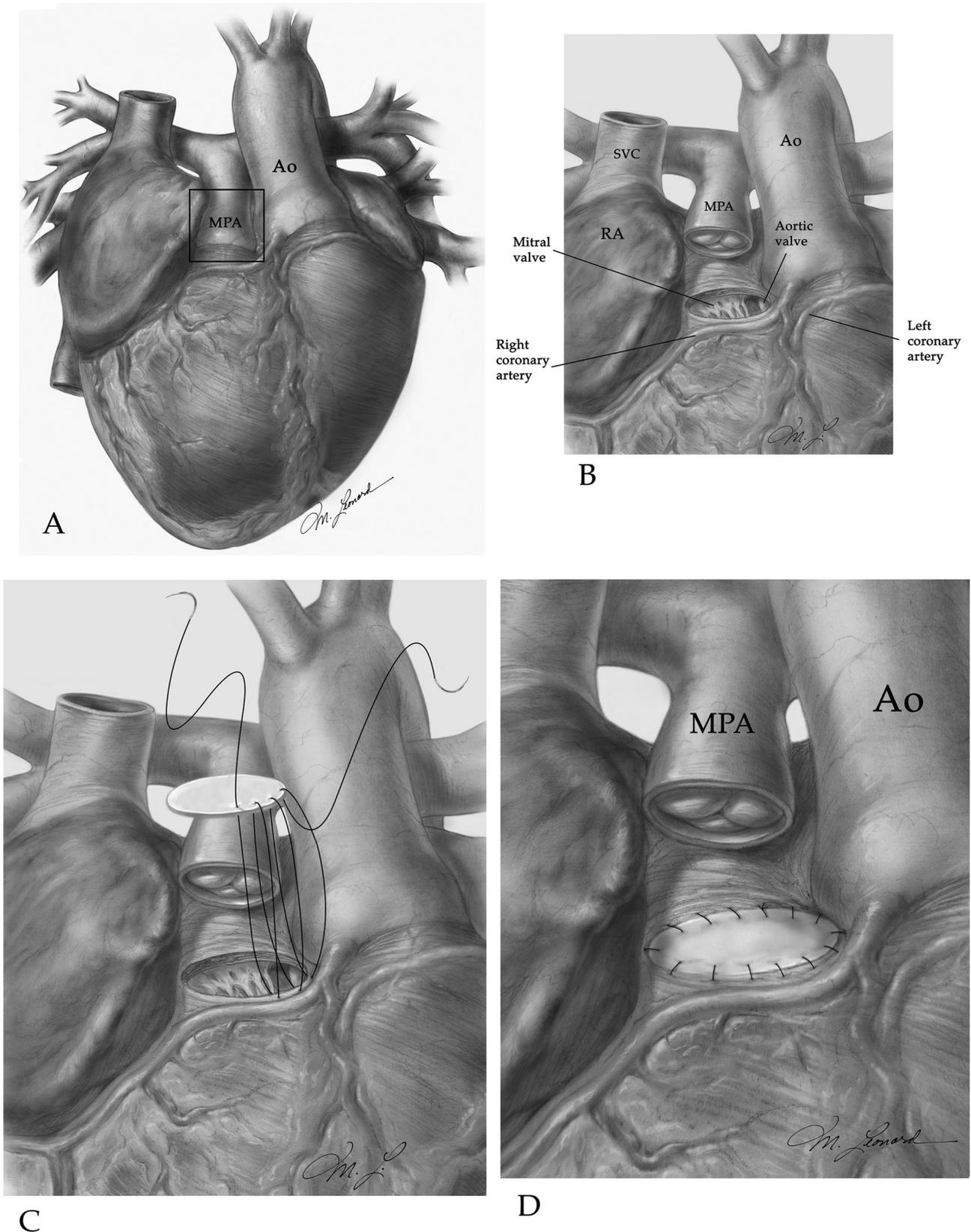
As aforementioned, ongoing experience with this technique has enabled the possibility of preserving a small but functional pulmonary trunk. Patients with VSD/PS may not have a pulmonary valve suitable for the systemic circulation, but can be considered for salvage in the pulmonary position, at a reduced total output with superior cavopulmonary offloading. This autologous valve can be durable, and grow with the patient, to reduce the likelihood of reintervention.

Appropriate technique is critical in both the harvesting and positioning of this autograft, and we have found it best to perform the pulmonary root harvest as the first step of the operation, if it is to be considered. A small exploratory incision in the pulmonary trunk is performed to allow direct visualization of the pulmonary valve to ensure that the valve appears suitable. Particular attention (Fig. 4) to (1) the pulmonary-mitral fibrous continuity, (2) nearby coronary arteries, and (3) the aortic annulus, is imperative, as injury to any of these crucial structures is difficult to repair. Once the autograft is harvested, it is assessed for length and orientation relative to where the planned ventriculotomy will be. As is important for the LeCompte, pulmonary root translocation requires aggressive mobilization of the branch vessels through the lobar and segmental levels to avoid unnecessary tension that can result in stenosis. The harvest site is also carefully closed with a circular patch of autologous or bovine pericardium. Primary closure is



**Figure 3** Hemi-Mustard. (A) A right atriotomy that traverses from the right atrial appendage to the lateral IVC-RA junction enables appropriate visualization of the systemic veins and mitral valve. This incision terminates about 1 cm from the IVC insertion into the right atrium, facilitating placement of the Hemi-Mustard patch. (B) The atrial septum primum is then generously excised; this is carried aggressively cephalad and caudad until epicardial fat is entered in both directions. (C, D) Areas of weakness are reinforced and re-endothelialized with interrupted suture. The coronary sinus is unroofed. The creation of an unrestricted common atrium is imperative to prevent obstruction of flow from pulmonary veins to the mitral valve, and IVC to tricuspid valve. (E, F) A GoreTex patch is then sewn to baffle IVC flow to the more posterior tricuspid valve. This baffle is most vulnerable to obstruction as its path crosses the atrial septal plane. As such, the baffle is shaped in a circular fashion to prevent narrowing at its waist.

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**Figure 4** Pulmonary artery root harvest. (A) Importantly, the pulmonary root is nestled between the native aortic root and right coronary artery, and in fibrous continuity with the anterior leaflet of the mitral valve. (B) Precise excision enables harvest without damage to the autograft or the surrounding structures, and (C, D) patch closure of the native left ventricular outflow tract prevents distortion.

ill-advised, as distortion of the aforementioned neighboring structures (ie, mitral valve, aortic valve, coronary artery) would ensue.

Positioning the translocated pulmonary root is performed after cross clamp removal, but prior to SCPC. Much like conduit placement, the native annulus is anastomosed posteriorly in running fashion to the ventriculotomy. Anteriorly, this is usually not possible due to foreshortening, and hood augmentation with a piece of homograft or pericardium is completed. The SCPC is anastomosed after the translocated pulmonary arterial system is in its final position to ensure appropriate orientation.

## DISCUSSION

Our longitudinal experience with this population encompasses 98 patients over 20 years, of which 49 have undergone arterial switch, and 49 intracardiac baffle repair. Seventy-seven underwent modified atrial switch by hemi-Mustard with SCPC offloading. Three patients have undergone native pulmonary root translocation. We have had three operative mortalities.

Of the 77 patients having undergone modified atrial switch, none have required reintervention for sinus node dysfunction or baffle obstruction. One patient developed atrial tachyarrhythmia requiring catheter-based ablation, and one infant required SCPC takedown for intractable chylothorax. Importantly, the use of the hemi-Mustard and SCPC has resulted in significantly longer freedom from intervention for RV-PA conduit failure [7,8].

Of the three patients who underwent pulmonary root translocation, mid-term surveillance demonstrates free pulmonary insufficiency without stenosis, mild pulmonary stenosis and insufficiency, and mild pulmonary insufficiency without stenosis respectively. For the first patient, the pulmonary valve was noted to be bicuspid with commissural fusion, which improved only marginally on commissurotomy prior to translocation. A decision was made in that case to utilize the translocated root as a conduit with transannular patch augmentation. These outcomes reflect our greater experience with pulmonary root translocation in more common lesions.

This overall experience demonstrates an approach to this complex lesion that is broadly applicable, promoting restoration of the morphologic left ventricle to the systemic circulation, while avoiding the shortcomings of a complete atrial-level switch. The partial offloading afforded by the superior cavopulmonary connection prolongs freedom from intervention for conduit failure and opens the possibility for salvage of the small but functional pulmonary valve, for which early experience is promising. A further delineation of the long-term performance of this circulation is warranted, and active investigation is ongoing to assess this cohort via physiologic testing.

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