Decreased Incidence of Supravalvar Pulmonary Stenosis After Arterial Switch Operation

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Background—Supravalvar pulmonary stenosis (SVPS) is frequently observed after arterial switch. Traditionally the coronary arteries are removed from the neopulmonic root by excising the entire sinus of Valsalva. As a result, reconstruction of the neopulmonic root requires a pericardial patch encompassing two-thirds of the anastomosis between the neopulmonic root and pulmonary artery. We present a technique where the coronary arteries are removed as limited buttons of sinus tissue, leaving the transected edge of the neopulmonic root intact. We hypothesize that maintaining native arterial tissue in the anastomosis between the neopulmonic root and the pulmonary artery bifurcation reduces postoperative SVPS.

Methods and Results—We performed a retrospective review of neonates with D-transposition of the great arteries undergoing arterial switch procedure from 1996 to 2009. Charts were reviewed, and clinical outcomes recorded for each patient. Most recent echocardiograms were evaluated for right ventricular outflow tract obstruction. A total of 120 patients received arterial switch using this technique. There was 99% survival and no injuries to the coronary arteries regardless of anatomy. Total follow-up was 564 patient-years. Mean follow-up at last clinical visit was 66±46 months. Evaluation of the most recent outpatient echocardiogram revealed an average peak instantaneous gradient across the neopulmonic root of 22.5±5 mm Hg. Only 7 (5%) patients required reintervention (balloon dilation, n=5; surgery, n=2).

Conclusions—Our technique of removing the coronary arteries as limited buttons, and anastomosis of the pulmonary artery using only native arterial tissue provides excellent midterm results with minimal SVPS. (Circulation. 2012;126[suppl 1]:S118–S122.)

Key Words: transposition of the great vessels ■ congenital heart disease

The arterial switch operation is the preferred surgical treatment for transposition of the great arteries. However, despite its improved outcomes over the mustard and atrial switch procedures, postoperative supravalvar pulmonary stenosis (SVPS) is common. Supravalvar pulmonary stenosis occurs in 17% to 55% of patients and is related to surgical technique, as well as age at operation. Historically, the coronary arteries are removed from the neopulmonic root by excising the entire associated sinus of Valsalva. Reconstruction of the neopulmonic root therefore requires a pectus pericardial patch encompassing two-thirds of the anastomosis between the neopulmonic root and pulmonary artery.

We have modified our surgical approach to remove the coronary arteries as limited buttons of sinus tissue, leaving the transected edge of the neopulmonic root intact. We hypothesize that maintenance of native arterial tissue in the anastomosis between the neopulmonic root and the pulmonary artery bifurcation reduces postoperative incidence SVPS.

Methods

Surgical Technique

Before initiation of cardiopulmonary bypass, the aorta, aortic arch, and great vessels are dissected free. Adequate mobilization of the aorta is necessary for allowing transection of the ascending aorta, which is more distal than the conventional approach. Figure 1A demonstrates the location for aortic transection using our technique compared with traditional methods. The branch pulmonary arteries are dissected free to the hilum of the lung, allowing optimal mobility after the LeCompte maneuver. Cardiopulmonary bypass was initiated after aortic and bicaval cannulation, unless bicaval cannulation was not technically feasible (from 1996 to 2000, single venous cannulation was performed, and the patient was cooled to 18°C for intracardiac repair). After 2000, nearly all patients, regardless of size and intracardiac anatomy, had bicaval venous cannulation and mild hypothermia (≈35°C). After myocardial arrest with cold blood cardioplegia, the ascending aorta is transected midway between the sinotubular junction and the origin of the innominate artery (Figure 1A). The main pulmonary artery is completely mobilized and transected as distally as possible. A LeCompte maneuver is performed. The individual coronary ostium are excised as a button, leaving the majority of the coronary sinus intact (Figure 1B). This is...
easily accomplished in nearly every case, even in those children with a single coronary ostium. In the event of an intramural coronary artery, the single button is excised, along with the intramural portion of the coronary artery. Every effort is made to not interrupt the original transection point of the aorta. Coronary artery mobilization is commensurate with the goal of translocating the coronary artery in a tension free manor. Coronary artery translocation is accomplished with a medial flap technique using 8–0 Prolene suture. The distal ascending aorta is then anastomosed to the neopulmonic root, and a second dose of cold-blood cardioplegia is administered. The defects in the neopulmonic facing sinuses are patched with autologous pericardium. In the case of an intramural coronary artery, the patch may be extended not only to cover the defect from the coronary button excision but also patched in the trailing portion of the aorta. After the neopulmonic root has been patched, any intracardiac repair is accomplished and the aortic cross-clamp is removed (Figure 1C).

In contrast to our technique, Figure 1D demonstrates the traditional surgical approach (this has already been described). During rewarming, the neopulmonic root is anastomosed to the pulmonary artery bifurcation using 7–0 Prolene suture with the anastomosis between the neopulmonic root and main pulmonary artery entirely composed of arterial tissue and no autologous tissue. The patient is weaned from cardiopulmonary bypass and transferred to the pediatric cardiac intensive care unit for postoperative management.

**Review Methods**

Institutional Review Board approval was obtained at The University of Rochester Medical Center, Rochester, NY, and Upstate Medical Center, Syracuse, NY, and consent was waived for this review. A retrospective review of all patients with the diagnosis of D-transposition of the great arteries repaired by arterial switch from June 1, 1996, to December 31, 2009, was performed. Inpatient chart review allowed description of perioperative events. Patient follow-up and need for reintervention was determined from outpatient clinic charts. The most recent postoperative echocardiograms were independently reviewed by 1 of 2 echocardiographers (N.A.-Y., C.M.). Postoperative echocardiograms were analyzed for incidence, severity and location (valvar, supravalvar, and branch pulmonary artery) of right ventricle outflow tract obstruction as estimated by the velocity of blood flow. This gradient was classified as either trivial (<20 mm Hg), mild (20–40 mm Hg), moderate (41–60 mm Hg), or severe (>61 mm Hg). In addition the amount of aortic insufficiency was determined by the latest echocardiogram as either none, trace, mild, moderate, or severe.

**Statistical Analysis**

Data are presented as mean±SD for continuous variables, and number (%) for noncontinuous variables. A 2-sample t test accounting for unequal variances was used to compare between groups. Kaplan-Meier freedom from intervention curves were constructed using GraphPad Prism version 5.0, GraphPad Software, San Diego, CA. In all cases, a probability value of <0.05 was considered significant.

**Results**

From June 1, 1996, to December 31, 2009, 120 patients underwent the arterial switch operation. Patient characteristics are depicted in the Table. Mean age at repair was 13±33 days (range, 1–306 days). A total of 12 patients were older than 14 days, and 6 patients were older than 30 days at the time of the operation. Whereas 59% of patients (n = 71) patients had isolated transposition, 41% of patients (n = 49) had additional defects including coarctation of the aorta (n = 9), ventricular septal defect (n = 34), and Taussig-Bing (n = 6) (Table). According to the Leiden classification,11 the majority of patients had 1LCx-2R pattern. Three patients had intramural had coronary artery anatomy and 9 patients had coronary arteries that originated from a single sinus.

**Operative Course**

No coronary ostial injuries occurred as a result of removing both the left and right coronary ostia from the sinus as a small
button, even in those patients with both coronaries ostia arising from a single sinus. One patient required internal mammary artery bypass of the left anterior descending artery secondary to myocardial ischemia of the left ventricular anterior and lateral wall. This patient had a single intramural left main coronary artery supplying the entire coronary circulation. There were no intraoperative mortalities; however, 1 patient died on postoperative day 2 of hypoxia. There were 2 deaths, both at 2 months after discharge. Each patient had an echocardiogram demonstrating good ventricular function within the prior 30 days, and therefore both deaths were thought to be unrelated to the arterial switch procedure.

**Outcome**

Follow-up was available for 109 (90%) patients, at a mean duration of 66±46 months. Total follow-up was 564.2 patient-years. There were 10 patients who were transferred from the same referring hospital as neonates for arterial switch who did not follow-up with a cardiologist. Attempts to contact these patients directly were unsuccessful. From the most recent transthoracic echocardiogram, aortic insufficiency was absent in 40% of patients, trace in 38% of patients, and mild in 22% of patients. There were no patients with moderate or severe aortic insufficiency, and no patient required a surgical intervention on the aortic valve. A total of 61 (56%) patients had trivial stenosis, 41 (38%) had mild stenosis, and 7 (6%) patients had greater than mild SVPS (Figure 2A). All 7 patients with greater than mild SVPS have undergone reintervention. Five (4%) have undergone catheter-based balloon dilation, and 2 patients (2%) have required reoperation for complex right ventricular outflow tract reconstruction. As demonstrated in Figure 2B, the majority of patients had the highest gradient across the supravalvar region of the pulmonary artery.

**Interventions**

Patients requiring any reintervention (n=7) for SVPS after arterial switch were significantly younger at the time of operation than patients (n=102) who did not require an intervention for SVPS (6.6±3.1 versus 33±33 days; P=0.05). However, despite differences in age at operation, there were no differences in preoperative weight, cardiac diagnoses, or coronary artery pattern (data not shown). The 3 patients who required balloon dilation were reintervened at 3, 3.3, 4, 5, and 11 years after arterial switch. All patients had trivial or mild stenosis that progressed over time. Interestingly, in 1 patient, it took 11 years before the development of significant SVPS requiring catheter-based intervention. In all patients, balloon dilation reduced supravalvar gradients from moderate/severe to trivial/mild. Freedom from balloon dilation was 86.3% at 14.75 years (Figure 3).

Only 2 patients (1.6%) required surgical reconstruction for SVPS at 9 and 24 months, respectively. One patient who had surgical reconstruction 9 months after arterial switch had the diagnosis of Taussig-Bing anatomy with side-by-side great vessels, and, necessarily, the neopulmonic root was placed to the right pulmonary artery, not the distal main pulmonary artery at the time of arterial switch. The second patient underwent arterial switch as a neonate for transposition of the great arteries with ventricular septal defect, subvalvar pulmonary stenosis, and L-malposition of the great arteries. At the time of arterial switch, relief of the subvalvar pulmonary artery.

<table>
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<th>Table. Subject Characteristics</th>
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<tr>
<td>Age at repair, d</td>
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<tr>
<td>Male sex, n (%)</td>
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<td>Weight, kg</td>
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<tr>
<td>Cardiac diagnoses, n (%)</td>
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<tr>
<td>Simple TGA</td>
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<tr>
<td>TGA+coarctation</td>
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<td>TGA+VSD</td>
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<td>Taussig-Bing</td>
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<td>Coronary type, n (%)</td>
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<td>1LCx-2R</td>
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<td>1L-2RCx</td>
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<td>1R-2 LCx</td>
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<tr>
<td>1LR-2Cx</td>
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<tr>
<td>Intramural CA</td>
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<td>CA from 1 sinus</td>
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TGA indicates transposition of the great arteries; VSD, ventricular septal defect; and CA, coronary artery.

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**Figure 2.** A, Number of patients with either trivial (<20 mm Hg), mild (20 to 40 mm Hg), moderate (41 to 60 mm Hg), and severe (>61 mm Hg) pulmonary artery stenosis. B, Number of patients with either valvar, supravalvar, or branch pulmonary artery stenosis.
stenosis was accomplished through the pulmonary valve and the neopulmonic root was placed to the left main pulmonary artery. Both patients underwent a pulmonary artery arterioplasty using pulmonary homograft, which reduced the degree of SVPS to mild. At follow-up at 37 and 72 months, respectively, each patient had mild pulmonary stenosis. The overall freedom from reoperation for SVPS after arterial switch was 97.1% at 14.75 years.

Discussion
Despite evolution of pulmonary artery reconstruction during the arterial switch operation, supravalvar pulmonary stenosis continues to be the most common long-term sequelae. Although often amenable to catheter-based intervention, approximately 30% of patients with SVPS require surgical intervention. Although the cause of SVPS is unknown, many authors point to scar tissue formation at the anastomosis site, inadequate somatic growth of the pulmonary artery, and inadequate mobilization of both the neopulmonic root and pulmonary artery, resulting in tension at the anastomosis site, as well as nonviable tissue at the anastomotic site. Our technique attempts to address these hypothetical causes of SVPS. Traditionally the anastomosis between the neopulmonic root and pulmonary artery is two-thirds circumferential pericardium when using a pantaloon patch. Clearly, pericardial tissue, after fixation, will not grow along with the somatic growth of the patient. Therefore, with only one-third of the anastomosis using autologous vascular tissue, we believe this can result in an eccentric stenosis at the site of the anastomosis. Quaegebeur first described using a double patch and excising the coronary arteries as 2 “U”-shaped patches, rather than a single pantaloon-style patch. However, this technique still involves excision of the entire coronary sinus and utilizes pericardium within the pulmonary artery suture line. Ullmann et al described their technique for pulmonary artery reconstruction where the right coronary artery was removed similar to our technique with a rim of native pulmonary artery tissue above the sinus. However, the left coronary button as well as the surrounding tissue was removed in a more traditional manner, and autologous pericardium was used within the main pulmonary artery suture line. They reported a very low incidence (1.7%) of supravalvar PS, however, only described a 67% follow-up. Prifti proposed creating an anastomosis entirely of vascular tissue, thereby preventing the utilization of pericardial tissue. However, these results demonstrated the significant progression of SVPS when compared with the patch technique. More recently Moll et al reported a technique to reconstruct the neopulmonary artery without using patch material by creating a scalloped anastomosis. Their rate of SVPS requiring surgical intervention was 2.6%; however, they only included patients with simple transposition. Comparatively, the only 2 patients within our series who had SVPS and required surgical reconstruction had complex transposition. Last, utilization of the spiral anastomosis has been favored due to its ability to restore the great vessels to their natural position; however, this has not been shown to significantly reduce the incidence of SVPS.

Recently, the importance of aortic insufficiency has been noted in patients after arterial switch. The incidence of aortic insufficiency within our patients is similar to what has been previously reported during an intermediate-term follow-up. Further, our technique should not influence the amount of aortic insufficiency as the coronary buttons are translocated and sewn in a standard manner as reported previously.

We believe that anastomosis between the pulmonary artery and neopulmonic root must be constructed using only native arterial tissue in a tension free setting to reduce the incidence of SVPS. We extensively mobilize the main and branch pulmonary arteries as well as the aorta to ensure a tension free anastomosis. Tension may create excessive collagen formation at the anastomosis site. Especially in the low-pressure pulmonary vasculature, this may have more of a critical role in the outcome of the operation.

Conclusions
We present our technique of removing the coronary arteries from the neopulmonic root as limited buttons of sinus tissue, which leaves the transected edge of the neopulmonic root intact, which has produced excellent midterm results. There were no coronary artery injuries and a low incidence of supravalvar pulmonary stenosis. Further follow-up will be necessary to validate this approach over a longer period of time.

Disclosures
None.

References
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_Circulation_. 2012;126:S118-S122
doi: 10.1161/CIRCULATIONAHA.111.082867

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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