Excellent Long-Term Outcomes of the Arterial Switch Operation in Patients With Intramural Coronary Arteries

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Background. Intramural coronary arteries may complicate coronary artery transfer during the arterial switch operation. We sought to determine the long-term outcomes of 28 patients with intramural coronary arteries who underwent an arterial switch operation at a single institution.

Methods. All patients who had intramural coronary arteries and underwent an arterial switch operation were identified from the hospital database and retrospectively reviewed.

Results. From 1983 to 2009, 720 patients underwent an arterial switch operation at our institution. Twenty-eight (3.9%, 28 of 720) had intramural coronary arteries. Patients with intramural coronary arteries had transposition of the great arteries (96%, n = 27) or Taussig-Bing anomaly (4%, n = 1). There were no deaths. Follow-up was 100% complete. Mean follow-up was 16.3 years (median, 15.5 years; range, 5.6 to 26.9 years). No patient required reoperation or catheter reintervention on the coronary arteries. Freedom from reoperation was 93% at 10 years. No patient had more than mild aortic regurgitation at last follow-up. Nine (32%, 9 of 28) patients had coronary angiograms at median 16 months (range, 14 months to 17 years) after arterial switch operation. All patients were asymptomatic at the time of angiogram. One patient had mild stenosis of the circumflex coronary artery demonstrated on a routine coronary angiogram 14 months postoperatively. All 28 patients were asymptomatic and in New York Heart Association functional class I at last follow-up.

Conclusions. Patients with intramural coronary arteries are not at increased risk of death or coronary reinterventions and have excellent late outcomes after the arterial switch operation.


The arterial switch operation (ASO) has excellent early outcomes with low mortality and morbidity rates in the modern era [1–5]. Intramural coronary arteries (IMCAs) may complicate coronary transfer and contribute to a mortality of up to 28% in such patients (6). A metaanalysis of 1,942 children demonstrated that an IMCA was associated with a 6.5-fold increased risk of mortality following ASO [7]. Recent studies on the outcomes of children with IMCAs who undergo the ASO demonstrated variable results [3, 6, 8] (Table 1). We have previously reported that the IMCA was not a risk factor for mortality in 618 children who underwent an ASO for transposition of the great arteries (TGA) over a 25-year period at our institution (2). Herein we describe the surgical management of IMCA and the long-term outcomes for this subgroup of patients.

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Material and Methods

Patients

The study was approved by the Royal Children’s Hospital Human Research Ethics Committee. Between May 1983 and January 2009, a total of 720 patients underwent an ASO at the Royal Children’s Hospital. An ASO was performed for TGA (n = 618), Taussig-Bing anomaly (TBA; n = 57), congenitally corrected TGA (n = 21), for atrial to ASO conversion (n = 15) and TGA with univentricular physiology (n = 9). All operation reports were reviewed and all patients with IMCA were identified. There were 28 (3.9%, 28 of 720) patients with IMCA. Twenty-seven (96%, 27 of 28) patients had TGA and 1 (4%, 1 of 28) patient had TBA. Twenty (74%, 20 of 27) of the patients with TGA had an intact interventricular septum (TGA-IVS).

Definitions

An IMCA was defined as any coronary pattern in which at least one coronary artery coursed through the aortic wall for a variable distance.

Early death or reoperation was defined as death or reoperation occurring prior to hospital discharge or within
30 days of ASO. Late death or reoperation was defined as any death or reoperation that occurred after discharge and more than 30 days after ASO. Reoperation was defined as an operation on the heart or great vessels performed after the ASO excluding exploration for bleeding, wound debridement, mechanical circulatory support and pacemaker replacement. Reintervention encompassed reoperation and catheter reintervention performed after ASO.

Functional status was described according to the New York Heart Association (NYHA) classification.

Statistical Analysis

Data were analyzed using Stata Version 10 (Stata Corp, College Station, Texas). Continuous variables were reported as a mean or median with an accompanying range. Kaplan-Meier curves were constructed to display freedom from the study’s outcomes.

Results

Early Outcomes

The intramural course involved the left main coronary artery in 24 (86%, 24 of 28) patients, the left anterior descending artery in 2 (7%, 2 of 28) patients, the right coronary artery in 1 (3.5%, 1 of 28) patient, and both left and right coronary arteries in 1 (3.5%, 1 of 28) patient (Fig 1). Our approach to the IMCA (Fig 2A) was detachment of the posterior commissure of the aortic (neopulmonary) valve in 25 (89%, 25 of 28; Fig 2B) patients and unroofing of the IMCA (Fig 2C) in 16 (57%, 16 of 28) patients. Then the coronary arteries were detached and transferred either as a single button (Fig 2D) facilitated with a pericardial hood (Fig 2, D1–D3) in the first 3 (11%, 3 of 28) patients or as 2 separate buttons (Fig 2E) using the trapdoor technique (Fig 2, E1–E2) in the next 25 (89%, 25 of 28) patients. In all patients the pulmonary artery was reconstructed with a single autologous pericardial patch and the posterior neopulmonary commissure was reattached to it.

Median age at operation was 9 days (mean, 15 days; range, 2 to 81 days) and median weight at operation was 3.5 kg (mean, 3.8 kg; range, 2.6 to 10 kg). Mean cardiopulmonary bypass time was 158 min (median, 143 min; range, 93 to 275 min) and mean aortic cross clamp time was 89 min (median, 83 min; range, 49 to 149 min). Circulatory arrest was used in 50% of the patients with a mean circulatory arrest time of 9 min (median, 8 min; range, 5 to 54 min).

There were no early deaths or early reinterventions for the IMCA. There was no mechanical circulatory support in patients with IMCA before ASO. Two patients (7.4%, 2 of 28) with IMCA had TGA-IVS with poor preoperative left ventricular function and were supported with Medtronic BioMedicus (Eden Prairie, Minnesota) centrifugal left ventricular assist device. The first patient was a 5-day-old boy operated in 1994. The second patient was a 44-day-old girl operated in 2004. Both were weaned off of the left ventricular assist device in 2 days. In comparison,
of the remaining 647 patients, with simple TGA (n = 591) and TBA (n = 56), 4.3% (28 of 647) required mechanical circulatory support postoperatively. There were no perioperative morbidities related to the IMCA. One patient with IMCA had congenital left pulmonary artery stenosis that required early pericardial repair of the left pulmonary artery (Table 2).

Late Outcomes
Follow-up was obtained for all patients. Mean follow-up was 16.3 years (median, 15.5 years; range, 5.6 to 26.9 years). There were no late deaths. All patients were in NYHA functional class I at last follow-up.

LATE REINTERVENTION. There were 12 reinterventions in 5 patients that occurred at a mean 3 years after ASO (range, 1 day to 22 years). There were no reoperations or catheter reinterventions on the coronary arteries. Four (14%, 4 of 28) patients required reintervention including 7 reoperations and 4 catheter reinterventions. Freedom from reintervention was 89% at 5 and 10 years postoperatively and 84% at 15 and 20 years postoperatively (Fig 3). Reinterventions are listed in Table 2. One patient had 2 reoperations for supraaortic stenosis and an aorticto-pulmonary artery fistula, 1 patient had mitral valve repair, and 1 patient had four reoperations for repair of left pulmonary artery stenosis.

CORONARY SURVEILLANCE. No patients required reoperation or catheter reintervention on the coronary arteries. Thirteen (46%, 13 of 28) patients had exercise electrocardiograms (ECGs). Exercise ECGs were normal in 9 (69%, 9 of 13) patients and abnormal in 4 (31%, 4 of 13) patients. Two patients with positive exercise ECGs were followed up with normal myocardial perfusion scans and normal coronary angiograms. In one patient minor ST segment depression in the inferior leads was demonstrated. The patient was otherwise asymptomatic and required no further follow-up. One patient with a positive exercise ECG was scheduled for repeat testing.

Four (14%, 4 of 28) patients had myocardial perfusion scans at a median 11.5 years (mean, 11 years; range, 7 to 16 years) after ASO. All scans were negative for inducible ischemia. Myocardial perfusion scans were performed in 2 patients as part of routine follow-up, in 1 patient after ischemic changes on exercise ECG, and in the 1 patient with a mild stenosis of the circumflex coronary artery.

Nine (32%, 9 of 28) patients had 10 coronary angiograms at median 16 months (mean, 3.8 years; range, 14 months to 17 years) after ASO. Eight of the coronary angiograms were performed as part of routine follow-up and 2 coronary angiograms were performed because of ischemic changes on exercise ECG. All patients were asymptomatic and had normal coronaries on angiogram except 1 patient who had mild stenosis of the circumflex coronary artery. This patient had an angiogram 14 months postoperatively and remained asymptomatic.

Fig 1. (A–F) Intramural course of the left coronary artery (LCA) was most common (n = 27) and in most patients (n = 17) the intramural LCA came from sinus 2 (A and D). Intramural right coronary artery (RCA) was encountered in 2 children (E and G). (Ao = aorta; LAD = left anterior descending coronary artery; LCx = left circumflex coronary artery; PA = pulmonary artery.)
with a myocardial perfusion scan negative for inducible ischemia and a CT coronary angiogram demonstrating patent coronaries at 16 and 22 years postoperatively, respectively.

**Table 2. Reinterventions Following ASO**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Reintervention Type</th>
<th>Time Since ASO (years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Supravalvular aortic stenosis repair</td>
<td>2.1</td>
</tr>
<tr>
<td>2</td>
<td>Closure of aorta to RPA fistula</td>
<td>14.1</td>
</tr>
<tr>
<td>3</td>
<td>Mitral valve repair</td>
<td>13.4</td>
</tr>
<tr>
<td>4</td>
<td>Lengthen LPA pericardial tube</td>
<td>1 day</td>
</tr>
<tr>
<td>5</td>
<td>Reconnect LPA to MPA</td>
<td>3 days</td>
</tr>
<tr>
<td>6</td>
<td>Central shunt</td>
<td>1.0</td>
</tr>
<tr>
<td>7</td>
<td>LPA stenosis repair</td>
<td>2.7</td>
</tr>
<tr>
<td>8</td>
<td>Balloon angioplasty LPA</td>
<td>3.3</td>
</tr>
<tr>
<td>9</td>
<td>LPA stent</td>
<td>7.1</td>
</tr>
<tr>
<td>10</td>
<td>Balloon angioplasty LPA</td>
<td>15.3</td>
</tr>
<tr>
<td>11</td>
<td>Coiling of aortopulmonary collateral</td>
<td>1.2</td>
</tr>
</tbody>
</table>

ASO = arterial switch operation; LPA = left pulmonary artery; MPA = main pulmonary artery; RPA = right pulmonary artery.

**Comment**

Coronary transfer during the ASO in patients with IMCAs presents a surgical challenge. Few studies in the literature have looked specifically at the outcomes of this subgroup. A meta-analysis of 1,942 patients by Pasquali and colleagues [7] demonstrated that an IMCA had a 6.5-fold increased risk of mortality as compared to normal coronary anatomy. Metton and associates [6] reviewed 46 patients with IMCAs out of a cohort of 919 patients who underwent ASO between 1987 and 2008. They reported a mortality of 28%, including 11 deaths before discharge and 2 deaths at 51 and 105 days. Nine of the 11 deaths were deemed secondary to coronary complications.
Conversely, Thrupp and colleagues [3] reported 1 death (5.6%) in 18 patients with IMCAs out of 215 patients who underwent ASO between 1996 and 2006. They found that an IMCA was not a risk factor for mortality in their cohort of 215 patients and all survivors were asymptomatic after a median follow-up of 6.8 years. One death (14%) was reported in a smaller cohort of 7 IMCA patients operated by Chen and colleagues [8].

The results of our study demonstrate excellent early and late outcomes for patients with IMCAs who undergo an ASO. We report no deaths in 28 asymptomatic patients with a follow-up of more than 15 years. The first 3 patients in our series had a pericardial hood reconstruction of the coronaries and the next 25 underwent transfer using the trapdoor technique. The trapdoor technique for coronary transfer was first described by Brawn and Mee [9] at our institution and the modification of this technique for translocating IMCAs was first described at our institution by Asou and colleagues [10]. Our technique involves detaching the posterior commissure of the neopulmonary valve if the coronaries arise in close proximity, unroofing the IMCA if the ostium is stenotic and transferring the excised coronary button to a medially based trapdoor. We believe that a combination of wide unroofing and transfer of the IMCA using trapdoor technique is a key to successful outcomes. Thus, we advocate the use of this technique in patients with IMCAs who undergo the ASO because of its simplicity, reproducibility, and excellent long-term outcomes. Our results suggest that routine coronary angiograms in these patients during follow-up in childhood is unlikely to be useful in asymptomatic survivors.

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### References