Aortopulmonary Collaterals After Bidirectional Cavopulmonary Connection or Fontan Completion Quantification With MRI

Lars Grosse-Wortmann, MD; Abdulmajeed Al-Otay, MD; Shi-Joon Yoo, MD

Background—Aortopulmonary collaterals (APCs) have been associated with increased morbidity after the Fontan operation. We aimed to quantify APC flow after bidirectional cavopulmonary connections and Fontan completions, using phase-contrast MRI, and to identify risk factors for the development of APCs.

Methods and Results—APC blood flow was quantifiable in 24 of 36 retrospectively analyzed MRI studies. Sixteen studies were performed after the bidirectional cavopulmonary connections (group A) and 8 after the Fontan operation (group B). APC blood flow was calculated by subtracting the blood flow volume through the pulmonary arteries from that through the pulmonary veins. The ratio of pulmonary to systemic blood flow (Qp/Qs) was 0.93 ± 0.26 in group A and 1.27 ± 0.16 in group B. APC flow was 1.42 (0.58 to 3.83) L/min/m² and 0.82 (0.50 to 1.81) L/min/m² in groups A and B, respectively. The mean inaccuracies corresponded to 7.9 ± 14.5% and 7.1 ± 13.6% of ascending aortic flow in groups A and B, respectively. Qp/Qs was negatively correlated with a younger age at the time of the bidirectional cavopulmonary connections operation (r = -0.62, P = 0.01) and positively correlated with the age at the time of the Fontan completion (r = 0.81, P = 0.01). Patients with a previous right-sided modified Blalock-Taussig shunt had more collateral flow to the right lung than those without.

Conclusions—APC blood flow can be noninvasively measured in bidirectional cavopulmonary connections and Fontan patients, using MRI in the majority of patients and results in a significant left-to-right shunt. (Circ Cardiovasc Imaging. 2009;2:219-225.)

Key Words: collateral circulation ■ Fontan procedure ■ MRI

Whereas there is general agreement that aortopulmonary collaterals (APCs; Figure 1) develop frequently with a bidirectional cavopulmonary connections (BCPC) or Fontan circulation, controversy persists regarding their hemodynamic and clinical consequences.1–5 Some investigators have found them to be associated with prolonged pleural effusions and even an increased mortality rate after the Fontan operation, whereas others could not confirm a deleterious effect.2,4

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The contradicting nature of these reports may be related to the difficulty in assessing the magnitude of blood flow through collaterals angiographically.1 Conventional angiographic grading of APCs is neither objective nor quantitative as the extent of visualization of the APCs varies widely according to where, how fast, and how much contrast medium is injected.1,6 Ichikawa et al1 and Bradley et al6 quantified the blood flow through APCs during cardiopulmonary bypass at the time of the Fontan operation by measuring the amount of blood returning to the left atrium and relating it to the pump flow delivered via the aortic cannula. This approach, although of academic value in elucidating the magnitude and risk factors for APCs, is not helpful in the patients' clinical management before the Fontan operation and does not allow for follow-up after the operation. Furthermore, it requires intracardiac access, which is not otherwise necessary when extracardiac modifications or catheter laboratory completions of the Fontan procedure are performed. Inuzuka et al7 recently used nuclear imaging in combination with catheterization to quantify APC flow in BCPC patients. To date, no noninvasive and readily applicable method has been proposed to quantify APC blood flow in patients with functionally single ventricles, except for in a case report, using MRI.8 (Figure 1) It is conceptually possible to quantify APC blood flow by subtracting the pulmonary arterial from the pulmonary venous blood flow volumes.8–10 MRI is the gold standard for the quantification of arterial flow volumes,10,11 and it has recently been shown that pulmonary venous flow volumes can be measured accurately using phase-contrast MRI.12

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From The Labatt Family Heart Center and the Department of Diagnostic Imaging, The Hospital for Sick Children, The University of Toronto, Toronto, Ontario, Canada.
Correspondence to Shi-Joon Yoo, MD, Department of Diagnostic Imaging, The Hospital for Sick Children, 555 University Ave, Toronto, Ontario, Canada M5G 1X8. E-mail shi-joon.yoo@sickkids.ca
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the systemic to the pulmonary circulation. The demographic patient data and primary cardiac diagnoses are summarized in Table 1.

Most MRI studies were performed on a GE signa CV/H 1.5-T MRI scanner (General Electric Medical Systems, Milwaukee, Wis). After July 2007, 2 studies were carried out on a 1.5-T Avanto magnet (Siemens Medical Solutions, Erlangen, Germany). The phase-contrast MRI data were obtained using the following imaging parameters: minimum echo and repetition times, flip angle 20°; bandwidth, 31.25 kHz; 1 to 2 k-space lines per segment; number of excitations, 1 to 2; slice thickness, 4 to 5 mm; minimum field of view; matrix 256×128 to 256; upper velocity limit, 150 cm/s; 20 or more reconstructed phases per cardiac cycle. Measurements were made across the ascending aorta at the level of the right PA, the descending aorta at the level of the diaphragm, the superior vena cava, branch Pas, and the individual pulmonary veins. The imaging plane for each vessel, including the pulmonary veins, was based on 2 localizer images that were parallel to the vessel long axis and orthogonal to each other. Patients younger than 6 years of age were examined under general anesthesia.

A commercially available work station (CV Flow, Medical Imaging Systems, Leiden, The Netherlands) was used for quantification of the flow volumes through the ascending aorta (QAAA), the descending aorta at the diaphragm (QDAA), the right and left PAs (QRPA and QLPA), the individual pulmonary veins (QPV), and the superior vena cava (QSVC). In 3 studies showing sizable systemic venous collaterals between the superior and inferior vena cava systems, the volume of the systemic venous collateral runoff (Qrunoff) was also measured. All flow volumes were indexed to the body surface area of the patient. The following parameters were calculated from the measured flow volumes:

- Blood flow volume through the PAs (QPA) = QRPA + QLPA
- Total pulmonary blood flow volume (QPV) = QRPA + QLPA
- Total systemic blood flow volume (QS)
- = QSVC + QDA (when there were no significant systemic venous collaterals) or = QSVC + QDA + Qrunoff (when there were obvious systemic venous collaterals)
- Ratio between the pulmonary and systemic blood flow volumes (Qp/Qs)
- APC flow volume (QAFC)
- Method A: QAFC = Qp - QPA
- Method B: QAFC = QAA - QPA
- APC flow volume fraction (APC fraction) = QAFC/QP

Oxygen saturations were retrieved from the documentation of an outpatient visit within 3 months of the MRI. A chart review confirmed that there had been no interventions or deteriorations between the oxygen saturation recording and the MRI. Mean PA and atrial and end-diastolic ventricular pressures were recorded if there had been no clinical or surgical change between the times of MRI and cardiac catheterization. Pulmonary vascular resistance (PVR) was calculated by dividing the transpulmonary pressure gradient obtained at the time of cardiac catheterization by Qp (sum of QPV), measured by phase-contrast MRI.

Statistical Analysis
Continuous variables that follow a Gaussian distribution are presented as mean±SD. Nonnormally distributed variables are given as medians and ranges. Proportions are presented as a frequency (%). Paired t tests were performed to compare demographic data and the results of the flow measurements between the 2 patient groups. The intraclass correlation coefficient was used to compare the 2 methods of calculating APC blood flow. Parametric (Pearson correlation analysis) and nonparametric (Mann–Whitney U test) correlation analysis were performed as appropriate.

Results
The results of the measurements are summarized in Table 2. There was a good intraclass correlation between the APC

We report the systematic use of MRI in the quantification of APC blood flow in patients after BCPC or modified Fontan procedure. We sought to investigate the risk factors for and hemodynamic consequences of these APCs.

Methods
We reviewed the MRI data of patients after BCPC as well as after a modified Fontan operation or completion of the Fontan circuit in the catheterization laboratory. We identified 36 studies between May 2003 and October 2007. Among these, APC flow was quantifiable in 24 studies performed in 19 patients. In the remaining 12 studies, APC quantification was not possible. It was precluded by susceptibility artifacts caused by pulmonary artery (PA) stents, vascular coils and clips, and septal occluder devices in the Fontan fenestration in 5 studies. In 7 studies, complex pulmonary venous anatomy prevented adequate flow measurements and hence APC quantification.

Four patients had more than 1 MRI examination. Eight patients had a Fontan-type circulation with an even distribution of male and female patients in this group. Twelve patients, 9 of them male, had undergone a BCPC but not a Fontan operation at the time of the MRI. One patient was in both groups because she underwent an MRI examination under general anesthesia.
flow volumes measured by the 2 different methods ($r=0.73$) (Figure 2). The APC flow volumes as calculated using method A were subsequently used for analyses.

In the BCPC group, the APC flow index was 1.42 (0.58 to 3.83) L/min/m². This accounted for 46.0% of Qp and led to a Qp/Qs of 0.93/0.26. In the Fontan group, the APC flow index was 0.82 (0.50, 1.81) L/min/m². This made up 19.7% of Qp and led to a Qp/Qs of 1.27/0.16.

Table 3 compares the extent of collateral flow between patients with and without a previous modified Blalock-Taussig shunt.

In the BCPC patients, there was a strong correlation between APC flow volume and Qp as well as Qp/Qs ($r=0.83$, $P<0.0001$ and $r=0.82$, $P<0.0001$, respectively). Qp/Qs as well as APC flow volume index correlated with the arterial oxygen saturation ($r=0.59$, $P=0.02$ and $r=0.49$, $P=0.05$, respectively). The percentage of Qp that was supplied via collaterals (APC flow volume fraction) was inversely correlated with QPA in BCPC patients ($r=-0.68$, $P=0.004$). Increased mean PA pressure was associated with a lower QPA ($r=0.53$, $P=0.03$). Qp/Qs showed a significant positive correlation with PVR ($r=0.62$, $P=0.01$). Ventricular end-diastolic pressure and atrial pressures were not associated with APC flow or Qp/Qs. APC flow volume did not correlate with PVR or PA pressure. A higher Qp/Qs was associated with a younger age at the time of the BCPC ($r=0.62$, $P=0.01$). The

### Table 1. Demographic Data and Primary Diagnoses of Patients Who Were Studied With a BCPC and After the Fontan Completion

<table>
<thead>
<tr>
<th></th>
<th>BCPC Studies (n=16)</th>
<th>Fontan Studies (n=8)</th>
<th>$P$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, mo</td>
<td>44.4 (14.2, 154.9)</td>
<td>91.7 (41.0, 215.2)</td>
<td>0.03</td>
</tr>
<tr>
<td>Weight, kg</td>
<td>13.9 (9.5, 63.0)</td>
<td>23.1 (14.6, 76.9)</td>
<td>0.0068</td>
</tr>
<tr>
<td>Body surface area, m²</td>
<td>0.61 (0.45, 1.71)</td>
<td>0.87 (0.63, 1.91)</td>
<td>0.0068</td>
</tr>
<tr>
<td>Age at BCPC, mo</td>
<td>6.0 (2.4, 22.9)</td>
<td>6.7 (2.3, 56.0)</td>
<td>0.15</td>
</tr>
<tr>
<td>Time between BCPC and MRI, mo</td>
<td>36.1 (11.3, 144.8)</td>
<td>85.6 (32.8, 192.0)</td>
<td>0.03</td>
</tr>
<tr>
<td>Age at Fontan, mo</td>
<td>N/A</td>
<td>33.8±11.9</td>
<td>N/A</td>
</tr>
<tr>
<td>Time between BCPC and Fontan, mo</td>
<td>N/A</td>
<td>18.0±11.3</td>
<td>N/A</td>
</tr>
<tr>
<td>Time between Fontan and MRI, mo</td>
<td>N/A</td>
<td>80.1±69.9</td>
<td>N/A</td>
</tr>
<tr>
<td>HLHS</td>
<td>3</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>AVSD, small left ventricle</td>
<td>2</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Double-inlet left ventricle</td>
<td>1</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Double-inlet left ventricle, TGA</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>AVSD, TGA, small right ventricle</td>
<td>0</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>2</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Tricuspid atresia, TGA</td>
<td>3</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>DORV</td>
<td>4</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>TGA, subpulmonary stenosis</td>
<td>0</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

AVSD indicates atrioventricular septal defect; DORV, double-outlet right ventricle; HLHS, hypoplastic left heart syndrome; TGA, transposition of the great arteries.

### Table 2. Results of Transcutaneous Oximetry, Invasive Pressure Measurements, and MRI

<table>
<thead>
<tr>
<th></th>
<th>BCPC Studies (n=16)</th>
<th>Fontan Studies (n=8)</th>
<th>$P$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oxygen saturation, %</td>
<td>79.0 (73, 88)</td>
<td>94.0 (77, 99)</td>
<td>0.004</td>
</tr>
<tr>
<td>Mean pulmonary artery pressure, mm Hg</td>
<td>12 (8, 18) (n=16)</td>
<td>13 (13, 15) (n=3)</td>
<td>0.06</td>
</tr>
<tr>
<td>Ventricular end-diastolic pressure, mm Hg</td>
<td>8 (6, 13) (n=14)</td>
<td>7 (7, 9) (n=3)</td>
<td>0.85</td>
</tr>
<tr>
<td>Pulmonary vascular resistance, Wood units/m²</td>
<td>7.6±6.2 (n=12)</td>
<td>10.4±2.8 (n=3)</td>
<td>0.47</td>
</tr>
<tr>
<td>Qp, L/min/m²</td>
<td>2.76±0.90</td>
<td>3.37±0.82</td>
<td>0.12</td>
</tr>
<tr>
<td>Qs, L/min/m²</td>
<td>3.00±0.65</td>
<td>2.70±0.74</td>
<td>0.32</td>
</tr>
<tr>
<td>Qs to head and upper extremities, %</td>
<td>49±12%</td>
<td>45±14%</td>
<td>0.47</td>
</tr>
<tr>
<td>Qp/Qs</td>
<td>0.93±0.26</td>
<td>1.27±0.16</td>
<td>0.002</td>
</tr>
<tr>
<td>APC flow by method A, L/min/m²</td>
<td>1.42 (0.58, 3.83)</td>
<td>0.82 (0.50, 1.81)</td>
<td>0.05</td>
</tr>
<tr>
<td>APC flow by method B, L/min/m²</td>
<td>0.78 (−0.07, 2.76)</td>
<td>0.63 (−0.17, 1.90)</td>
<td>0.53</td>
</tr>
<tr>
<td>Contribution of collateral flow to Qp</td>
<td>46.0 (31.7, 90.3)</td>
<td>19.7 (18.4, 60.9)</td>
<td>0.0059</td>
</tr>
<tr>
<td>Ventricular output toward collaterals, %</td>
<td>36.4±14.0 (15.3, 68.0)</td>
<td>26.3±9.1 (17.3, 39.1)</td>
<td>0.046</td>
</tr>
</tbody>
</table>

APC indicates aortopulmonary collateral.
correlations of a younger age at the time of the MRI and increased APC flow volume almost reached statistical significance \( r = 0.47, P = 0.07\). A higher \( Q_p/Q_s \) was associated with an older age at the time of the Fontan completion \( r = 0.81, P = 0.01\).

Patients with a previous right-sided modified Blalock-Taussig shunt had more APC flow to the right lung than those without a previous shunt on the right side (Table 3, patients with pulmonary venous obstruction were excluded from this part of analysis). Only 2 patients had a history of left-sided shunts, so this could not be assessed as a risk factor for APCs. There was no difference in the magnitude of \( Q_p/Q_s \) or the degree of collateral formation in male versus female patients, right versus left aortic arch, and unilateral versus bilateral BCPC.

We previously reported the case of 1 patient, included in this analysis, who underwent 3 studies and developed massive APCs on the side of progressive and finally total occlusion of the left pulmonary veins.8 One patient underwent an MRI before and after her Fontan completion: After the operation, the APC flow volume index decreased from 1.38 to 0.50 L/min/m². The APC flow volume fraction decreased from 61.9% to 20.0%. After the Fontan operation, the oxygen saturation increased from 73% to 98%.

To assess the accuracy of our flow measurements, we compared the flow volume in the ascending aorta with the total venous return to the heart, that is, with the sum of pulmonary and systemic venous flows in patients with a BCPC situation and to the total pulmonary venous flow volume in patients with a Fontan circulation. If all the flow measurements were perfectly accurate, then the discrepancy between the total venous return to the heart and the ascending aortic flow would be zero. The discrepancy was \( 0.26 \pm 0.48 \) L/min/m² or 7.9 ± 14.5% (0 to 44.0%) of flow through the ascending aorta (\( Q_{AA} \)) in the BCPC and 0.19 ± 38 L/min/m², corresponding to 7.1 ± 11.3% (0 to 25.1%) of \( Q_{AA} \) in the Fontan group (excluding the patient with the fenestrated Fontan conduit).

**Table 3. Pulmonary Arterial and Systemic-to-Pulmonary Arterial Collateral Blood Flow to the Right Lung in Patients With and Without a Previous Right-Sided Modified Blalock-Taussig Shunt**

<table>
<thead>
<tr>
<th></th>
<th>No Right mBTS</th>
<th>Right mBTS</th>
<th>( P )</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right pulmonary artery flow, L/min/m²</td>
<td>1.32 ± 0.83</td>
<td>0.84 ± 0.42</td>
<td>0.16</td>
</tr>
<tr>
<td>Collateral flow to right lung, L/min/m²</td>
<td>0.47 ± 0.21</td>
<td>0.88 ± 0.44</td>
<td>0.02</td>
</tr>
<tr>
<td>Contribution of collateral flow to right pulmonary blood flow, %</td>
<td>23.8 (16.3, 65.4)</td>
<td>51.2 (9.1, 100)</td>
<td>0.06</td>
</tr>
<tr>
<td>( Q_p/Q_s )</td>
<td>1.03 ± 0.28</td>
<td>1.07 ± 0.24</td>
<td>0.73</td>
</tr>
</tbody>
</table>

Four studies were excluded from this analysis because of left-sided pulmonary vein obstruction. mBTS indicates modified Blalock-Taussig shunt.

**Discussion**

The current study is the first to report the systematic use of cardiac MRI in the quantification of APC flow. This was successful in most patients. In almost one fourth, however, a complex venous anatomy, consisting of early branching and/or a tortuous course, flow measurement in at least 1 pulmonary vein was not possible. The internal validation of flows in this cohort confirms that the measurements of APC flow are accurate with an acceptable margin of error in the majority of patients. Sources of inaccuracy include the fact that several flow measurements were combined in the calculation of APC flows, each of which introducing a slight inaccuracy, but also, once again, by complex pulmonary vein anatomy. The 2 proposed methods of APC flow volume calculation (\( Q_p \) minus \( Q_{PA} \) versus \( Q_{AA} \) minus \( Q_s \)) showed good agreement. Figure 2B shows that the 3 patients with the
The greatest differences in the APC flow volumes calculated by methods A and B all had lower flow volumes from method B than from method A, and all 3 had undergone a Norwood operation. Method B uses Q_{AA}, Q_{SVC}, and Q_{DA}. We have recently demonstrated the accuracy of phase-contrast MRI measurements of these flows. This, however, may not be the case for Q_{AA} after the Norwood procedure. In fact, the discrepancy of venous return to the heart and cardiac output in these outliers was much greater (up to 44.0% of Q_{AA}) than the overall mean in this cohort. A plausible explanation for the underestimation of APC flow by method B in these 3 patients is the inaccuracy of flow measurements in the reconstructed ascending aorta. Turbulence from competing flows at the aortopulmonary anastomosis and swirling flow in the large reconstructed aortic arch undermine the physical prerequisites for accurate phase-contrast flow velocity measurements. We postulate that method A (Q_{p}-Q_{PA}) should be the preferred method for APC flow volume calculation, especially in the presence of a previous Norwood or Damus-Kaye-Stansel procedure.

Four-dimensional phase-contrast imaging, the acquisition of flow information in all directions within an imaging volume containing the heart and great vessels, was not available when the data that were used in this retrospective analysis was taken. This method holds promise particularly when flow at many different locations is to be measured as well as when the target is difficult since the optimal cut can be sought offline. The value of this approach has yet to be proven for small vessels: If the in-plane resolution in the resulting measurement plane is to be sufficient for pulmonary veins in young children, the scan time becomes quite long and quickly exceeds 15 minutes during which the patient must not move. Owing to extensive data processing, the results cannot be viewed until after the examination. The calculated APC flow in our BCPC cohort (1.42, 0.58 to 3.83 L/min/m²) was comparable to that by Inuzuka et al.7 who used a combined approach with nuclear imaging and catheterization (1.75 ± 0.43 L/min/m²).7 Their method, however, relies on a number of assumptions, is invasive, requires ionizing radiation, and cannot be used in Fontan patients.

Our analysis revealed a surprisingly high degree of APC blood flow, leading to important left-to-right shunts, in some cases within only a few months of the BCPC operation. On average, the Qp/Qs in our BCPC cohort was much greater than that reported by Salim et al.15 (0.93 ± 0.26 versus 0.61 ± 0.07). This difference in the degree of left-to-right shunting between their study and ours may be partly related to a patient referral bias as, particularly in the earlier studies in our cohort, MRI tended to have been requested because of PA or pulmonary venous obstruction, which probably promotes APC development.

Using the formula (Q_{p}-Q_{PA}) in normal volunteers, MRI showed that approximately 6.6% of the estimated cardiac output reaches the lungs via systemic arteries.12 In contrast, all patients in the current study showed a substantial amount of APC flow, with the minimum APC flow constituting 15.3% and 17.3% of the ascending aortic flow in BCPC and Fontan patients, respectively (Table 2).

The causes, risk factors, and the pathophysiology of the development of such collateral arteries in patients with single-ventricle physiology are not entirely clear. APC flow volumes, normalized to the patient’s body surface area, were significantly smaller in patients with a Fontan circuit than in those with a BCPC (0.82 L/min/m² versus 1.42 L/min/m², P=0.05). These data suggest that APCs regress spontaneously after the Fontan operation as was the case in the patient who was investigated both before and after the Fontan operation. APCs have been reported to occur more commonly after a longer interval between the BCPC and Fontan operation.3,4 Likewise, in our study, patients who proceeded to BCPC at a younger age were more likely to develop APCs, and patients who were older at the time of the Fontan completion had a greater degree of left-to-right shunting. It is, therefore, tempting to speculate that a factor unique to the BCPC circulation promotes the formation of APCs. What exactly mediates the development of APCs is uncertain. They may be an adaptive mechanism during prolonged periods of cyanosis, decreased volume and velocity of flow, or promoted by absent pulsatility, a high transpulmonary gradient, low flow of blood to the lungs, or lack of hepatic venous effluent.3,4 Experimental studies showed that reduced blood flow to only 1 region of the lung results in localized collateralization limited to the hypoperfused area.16 Similarly, in our study, a distribution of APCs that is limited to the obstructed lung could be seen in 2 patients with pulmonary vein obstruction (Figure 2). Therefore, systemic undersaturation or humoral factors17,18 alone cannot serve to explain the pathophysiology of APC development because they would affect both lungs. Furthermore, if hypoxemia was the principal stimulus, an association of APC flow with lower oxygen saturations would be expected. The opposite was true in our study, as well as in that by Triedman et al.1 We speculate that the velocity, pulsatility, and volume of flow are more important determinants of the development of APCs than oxygen saturation.

McElhinney et al.1 and Triedman et al.4 found an association between APCs and a previous Blalock-Taussig shunt.1,4 In agreement with these reports, patients in this cohort with a previous right-sided shunt had significantly less PA and more APC flow to that lung than those without a shunt. The mechanism by which Blalock-Taussig shunts promote the development of APCs is unclear but may be related to distortion of PA geometry. Pleural scarring and adhesions to the chest wall after placement of a Blalock-Taussig shunt may facilitate the recruitment of chest wall vessels into the network of APCs into the lung on that side.

The natural history of APCs and their effect on the long-term outcome of single-ventricle patients has yet to be properly investigated.1,3,19 Whereas Ichikawa et al.19 reported APC flow rates of more than one third of total cardiopulmonary bypass flow to be predictors of early postoperative failure, patients within our cohort with as much as 52.4% of collateral flow went on to do well after the Fontan procedure. In a prospective, randomized study, Brown et al.19 did not find a different outcome within 3 months after the Fontan operation between patients who underwent coil occlusion of collaterals and those who did not. Nevertheless, other inves-
tigators have shown that APCs predispose the patient to postoperative complications after the Fontan completion, including prolonged pleural effusions, death, or the need for early “take-down” of the Fontan pathway.\(^2,3,20,21\) A number of potential pathophysiological pathways for this have been conjectured, including increased ventricular preload as a result of the left-to-right shunt.\(^21\) Although previous reports have found patients with APCs to have a higher Qp/Qs, this study is the first to unequivocally establish APCs as the origin of this left-to-right shunt. The recirculating blood through the collaterals imposes a volume load on the systemic ventricle, thereby counteracting one of the major goals of the BCPC, which is to volume-unload the ventricle.\(^5,6,8\)

Another potentially unfavorable consequence of APCs in BCPC and Fontan patients is the increase of PA and systemic venous pressures that must be low for a functioning single ventricle physiology. It has been shown that additional pulmonary blood flow through a surgically created systemic-to-PA shunt raises upper central venous pressure in BCPC patients.\(^22\) We were able to show that the APC blood flow competes with PA flow, likely via a rise in mean PA pressures, which was calculated taking into account the added pulmonary blood flow via APCs, correlated with the degree of left-to-right shunting. Our data indicate that PVR is elevated, at least partly, as a result of the chronic left-to-right shunt. Turbulent inflow through APCs competes with the passive laminar flow and results in energy loss within the Fontan pathway, which can be deleterious in a circulation that is powered by only 1 ventricle.\(^23\)

**Limitations**

Given the small number of patients in each group, the fact that correlation analyses produced statistically significant results is remarkable. Further associations, however, were potentially obscured by the small sample size, particularly in the Fontan group, with only 8 studies. A multivariate analysis for risk factors for APCs was not possible. Intraindividual longitudinal data collection from serial MRI examinations would be valuable in determining the time course of collateral formation and their natural history in patients throughout their career within the Fontan palliation.

Left-to-right shunts via APCs occur in most if not all patients with single ventricles both before and after the Fontan operation. The volume-unloading effect of BCPC disappears with development of APCs in a significant number of cases. The magnitude and hemodynamic consequences of these shunts can be reliably assessed with MRI and are greater than thus far presumed; they interfere with the BCPC and Fontan physiology. Proceeding to the BCPC at a younger age and to the Fontan completion at an older age is associated with a greater left-to-right shunt through APCs.

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**Disclosures**

None.

**References**


CLINICAL PERSPECTIVE

Aortopulmonary collaterals (APCs) are frequent in patients with a so-called single ventricle physiology. On the basis of previous reports that linked the presence of APCs to a higher incidence of complications after the Fontan completion, foremost prolonged pleural effusions, most institutions coil-occlude large APC. Recently, however, this practice has been challenged by studies that did not confirm a deleterious effect of these channels. The contradicting nature of these reports may be related to the difficulty in assessing the magnitude of blood flow through collaterals angiographically. We are showing that APC flow volumes can be measured accurately and noninvasively in the majority of patients before and after Fontan completion. An increasing number of institutions are obtaining an MRI in their patients before proceeding to the Fontan completion. Artifacts from metallic implants which prevented APC flow measurement in 5 of our patients, will become less important and less frequent as MRI compatible devices are introduced. Phase contrast MRI can quantify the degree of left-to-right shunting through APCs. From this pilot study, it is still unclear whether a large APC flow volume is associated with a worse clinical outcome. Even now, however, awareness of the hemodynamics, including the ratio of pulmonary to systemic blood flow and collateral blood flow, will help the clinician in obtaining a complete picture of the patient’s status. Confirmation about a large left-to-right shunt can potentially guide therapy in a patient with single ventricle physiology and heart failure. Therefore, we believe that APC flow assessment should be performed in every patient with a bidirectional cavopulmonary connections or Fontan circulation who undergoes an MRI.
Aortopulmonary Collaterals After Bidirectional Cavopulmonary Connection or Fontan Completion: Quantification With MRI
Lars Grosse-Wortmann, Abdulmajeed Al-Otay and Shi-Joon Yoo

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