**Transcaval Puncture for Access to the Pulmonary Venous Atrium After the Extracardiac Total Cavopulmonary Connection Operation**

Jeremy P. Moore, MD, MS; Benjamin Hendrickson, MD; Daniel Z. Brunengraber, MD; Kevin M. Shannon, MD

**Background**—Patients with surgically palliated total cavopulmonary connection are at risk for recurrent atrial arrhythmia requiring catheter ablation. Transcatheter procedures for those with extracardiac conduits (extracardiac-total cavopulmonary connection) are perhaps the most challenging because of exclusion of the venous circulation from the arrhythmia substrate. Puncture through the inferior vena cava to the pulmonary venous atrium may be an effective route for access in these patients.

**Methods and Results**—The pediatric and adult congenital surgical databases were explored for patients with extracardiac-total cavopulmonary connection and postoperative computed tomography imaging to assess for the presence of clinically relevant (>3 mm) apposition between the inferior vena cava and pulmonary venous atrium (cavoatrial overlap). The degree of overlap between the structures was measured by 2 blinded reviewers. Patients were stratified by surgical repair in childhood versus adult congenital heart disease. Thirty-seven patients were identified, with cavoatrial overlap observed in 9 (36%) of pediatric and 1 (9%) of adult congenital heart disease–repaired patients. Time elapsed after surgery was associated with cavoatrial overlap in the pediatric cohort (P=0.034) and was identified in all pediatric patients with computed tomography imaging ≥8 years after surgery. Three patients underwent successful transcaval puncture during the study period without complication.

**Conclusions**—Puncture through a region of overlap between the inferior vena cava and pulmonary venous atrium is feasible. Cavoatrial overlap is present in a substantial proportion of patients undergoing extracardiac-total cavopulmonary connection in childhood and is associated with a longer time elapsed since surgery. (Circ Arrhythm Electrophysiol. 2015;8:824-828. DOI: 10.1161/CIRCEP.115.002969.)

**Key Words:** arrhythmias, cardiac □ catheter ablation □ Fontan procedure □ heart diseases □ vena cava, inferior

Supraventricular tachycardia is prevalent after the total cavopulmonary connection (TCPC) operation and is independently related to morbidity and mortality. Although modern surgical techniques such as the lateral tunnel and extracardiac variants are thought to be associated with decreased arrhythmia burden, the issue remains problematic. Electrophysiology procedures and catheter interventions requiring access to the pulmonary venous atrium (PVA) in patients with extracardiac conduits (E-TCPC) may be the most challenging, as the material used to create the conduit completely excludes this chamber from the venous circulation.

Current techniques to access the PVA after E-TCPC include hybrid surgical, retrograde aortic, and direct puncture approaches through synthetic conduits. Direct puncture is less invasive than surgical approaches, yet is a challenging procedure that may require multiple attempts, catheter balloon dilation, and occasionally prolonged procedure times. Transcatheter entry to the PVA through an area of native tissue below the extracardiac conduit has been previously reported and may represent a safe and technically straightforward alternative to the transconduit approach.

We hypothesized that a significant proportion of patients with prior E-TCPC performed in childhood would possess favorable anatomy for puncture from the inferior vena cava (IVC) into the PVA (cavoatrial approach), and that this finding would be more prevalent with time after operation.

**Methods**

After approval from the Institutional Review Board, the Pediatric Cardiology and Ahmanson/UCLA Adult Congenital Heart Disease databases were searched for all patients who had undergone prior E-TCPC between 2005 and 2015. Those patients without computed tomography (CT) imaging after the E-TCPC operation were excluded from further study. Patient characteristics including baseline demographics, congenital diagnosis, surgical history, arrhythmia history, and hemodynamic data were recorded.

All postoperative CT studies were reviewed in blinded fashion by 1 electrophysiologist and 1 radiologist. Three-dimensional reconstructions of CT examinations were performed using DICOM viewer.
WHAT IS KNOWN

- Access to the pulmonary venous atrium for electrophysiological procedures after the extracardiac total cavopulmonary connection is challenging.
- Existing approaches include direct surgical exposure, hybrid catheter-surgical, retrograde aortic, and transconduit puncture techniques, all of which require considerable time and effort and add to procedural complexity.

WHAT THE STUDY ADDS

- A technique involving puncture from the inferior vena cava just below the extracardiac conduit may allow facile access to the pulmonary venous atrium.
- A region of contact between the walls of the inferior vena cava and the pulmonary venous atrium can be demonstrated by computed tomography imaging in approximately one-third of patients undergoing childhood conduit placement. This finding seems to become more frequent with time after surgery.

OsiriX version 6.0 64-Bit software on a dedicated workstation with a display resolution of 2560×1440. The region below the conduit was evaluated for apposition between the wall of the IVC and the PVA. Cavoatrial overlap (clinically relevant apposition of the IVC and PVA walls) was considered to be present if the distance between these 2 structures was not appreciably different than the distance between the conduit and PVA wall and extended >3 mm inferiorly from the lower aspect of the conduit. Quantitative measurement of the region of overlap is measured in a plane orthogonal to a line transecting the center of both structures. Images were aligned with the axis of the conduit before measurement (Figure 1). Other values derived from the CT study included conduit length and diameter.

Statistical Analysis

Patients were stratified by E-TCPC in childhood (<18 years) versus those undergoing surgery in the setting of adult congenital heart disease (ACHD) (≥18 years). Interobserver variability for the degree of overlap was assessed using the method of Lin and illustrated by Bland–Altman plot. A priori acceptable 95% limits of agreement for Bland–Altman analysis were considered 3 mm. Subsequent values for overlap were considered to be the mean of the 2 observations. The prevalence of cavoatrial overlap was reported as a proportion (%) for each cohort. Characteristics for those patients with and without overlap were compared by Wilcoxon rank-sum method or the Student t test for continuous variables and Fisher exact test for categorical variables as appropriate. The relationship between overlap and time elapsed from surgery was illustrated with Kaplan–Meier curves. Values are reported as the median and the interquartile range or the number (%) as appropriate. A 2-sided P value of <0.05 is considered significant. All statistics were performed with JMP software (SAS Inc, Cary).

Results

A total of 37 patients with CT imaging of sufficient quality for analysis were included in the study. Twenty-six patients underwent E-TCPC in childhood and 11 in the setting of ACHD. Cavoatrial overlap was identified in 9 (36%) patients undergoing surgery during childhood versus only 1 (9%) patient for ACHD (P=0.097) despite similar follow-up duration (Table 1). The level of agreement for the cavoatrial dimension as calculated by Lin concordance correlation coefficient was 0.95 (confidence interval, 0.84–0.99), consistent with substantial agreement. Blinded observations were in agreement in all cases with respect to presence versus absence of overlap. Bland–Altman analysis demonstrated a mean bias of 0.02±0.57 mm with limits of agreement of −1.3 to 1.3 mm (Figure 2).

For children undergoing E-TCPC, there was an association between the interval from surgery to CT imaging and the presence of cavoatrial overlap (median, 4.9 versus 0.4 years for those with and without overlap, respectively; P=0.034). The median time from surgery to the observation of overlap by CT imaging was 8.2 years (confidence interval, 3.5–10.6) and was detected in all pediatric patients who had CT imaging >8 years after conduit placement (Figure 3). The overall median length of cavoatrial overlap was 11.7 mm (6.8–14.0 mm) with 5 (56%) patients demonstrating a dimension ≥10 mm. There was no significant association between prior conversion surgery (P=1.0), atrial pressure (P=0.163), or history of atrial arrhythmia (P=0.143) and cavoatrial overlap in the pediatric cohort.

Patients with E-TCPC in the setting of ACHD were more likely to undergo a conversion strategy than those operated in
childhood (P<0.001) and were more likely to have had prior atrial arrhythmia (P=0.028). The single ACHD patient found to have subsequent cavoatrial overlap had been previously palliated with bilateral aortopulmonary shunts before E-TCPC at the age of 44 years. The postoperative course was complicated by recurrent atrial arrhythmia and ventricular dysfunction with chronically elevated wedge pressure by cardiac catheterization. CT imaging performed 2.5 years later demonstrated a 12.5-mm region of overlap between the IVC and PVA.

Table 1. Sample Population Characteristics Stratified by Age at Surgery

<table>
<thead>
<tr>
<th></th>
<th>Pediatric (n=26)</th>
<th>ACHD (n=11)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Subject characteristics</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Male, %</td>
<td>16 (61.5)</td>
<td>1 (9.1)</td>
<td>0.004</td>
</tr>
<tr>
<td>Congenital diagnosis</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>DILV</td>
<td>4</td>
<td>3</td>
<td>...</td>
</tr>
<tr>
<td>DORV</td>
<td>5</td>
<td>3</td>
<td>...</td>
</tr>
<tr>
<td>Heterotaxy</td>
<td>4</td>
<td>1</td>
<td>...</td>
</tr>
<tr>
<td>HLHS</td>
<td>5</td>
<td>1</td>
<td>...</td>
</tr>
<tr>
<td>PA/IVS</td>
<td>3</td>
<td>0</td>
<td>...</td>
</tr>
<tr>
<td>TOF/PA</td>
<td>1</td>
<td>0</td>
<td>...</td>
</tr>
<tr>
<td>TA</td>
<td>4</td>
<td>3</td>
<td>...</td>
</tr>
<tr>
<td>Fontan pressure, mm Hg</td>
<td>17.5 (15–19)</td>
<td>14 (10–18)</td>
<td>0.215</td>
</tr>
<tr>
<td>End-diastolic pressure, mm Hg</td>
<td>11 (8–14)</td>
<td>13 (7–15)</td>
<td>0.771</td>
</tr>
<tr>
<td>Atrial arrhythmia</td>
<td>6 (23.1)</td>
<td>7 (63.4)</td>
<td>0.028</td>
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<tr>
<td>Surgical characteristics</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at surgery, y</td>
<td>6.9 (4.8–12.8)</td>
<td>33.0 (23.4–36.6)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Fontan conversion</td>
<td>5 (19.2)</td>
<td>9 (81.8)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Conduit diameter, mm</td>
<td>18 (16–20)</td>
<td>20 (20–20)</td>
<td>0.001</td>
</tr>
<tr>
<td>Conduit length, cm</td>
<td>7.3 (6.3–7.9)</td>
<td>8.5 (7.9–9.0)</td>
<td>0.007</td>
</tr>
<tr>
<td>Cavoatrial overlap</td>
<td>9 (36)</td>
<td>1 (9.1)</td>
<td>0.097</td>
</tr>
<tr>
<td>Age at imaging, y</td>
<td>9.9 (6.0–16.4)</td>
<td>36.3 (30.9–36.6)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Time to imaging, y</td>
<td>1.4 (0.1–5.6)</td>
<td>1.7 (0.1–3.3)</td>
<td>0.572</td>
</tr>
</tbody>
</table>

ACHD indicates adult congenital heart disease; DILV, double inlet left ventricle; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; PA/IVS, pulmonary atresia with intact ventricular septum; TA, tricuspid atresia; and TOF/PA, tetralogy of Fallot with pulmonary atresia.

Three patient underwent transcaval puncture for access to the PVA to perform an electrophysiological procedure (n=2) or for creation of a fenestration (n=1) during the study period (Table 2). Prior CT imaging was only available for 1 electrophysiology procedure (Figure 4A–4C). There were no complications related to the transcaval puncture procedure in any patient.

Discussion

The main findings of the present study are that (1) a region of apposition between the IVC and the PVA can be identified by CT imaging in a significant percentage of patients who

Figure 2. Bland–Altman plot demonstrating mean bias and 95% limits of agreement between blinded measurements of cavoatrial overlap by computed tomography imaging.

Figure 3. Kaplan–Meier plot of cavoatrial overlap after pediatric extracardiac total cavopulmonary connection operation. White circles represent censored events at the time of computed tomography imaging.
undergo the E-TCPC operation in childhood and (2) the prevalence of this finding increases in this cohort with time, such that it may eventually be observed in all patients after surgery. Patients operated during adulthood do not seem to develop this region of overlap as commonly, although it may be still be encountered in individual cases.

Various approaches for achieving access to the PVA after the E-TCPC operation have been reported. These have included open surgical techniques,\textsuperscript{5,6,14} the retrograde aortic,\textsuperscript{7,8} and more recently transconduit puncture routes.\textsuperscript{9,15–17} Limitations of the retrograde aortic approach consist primarily in the difficulty of achieving catheter stability and the requisite crossing of 2 systemic valves, especially when dealing with complex arrhythmia substrates over extended periods of time. The transconduit puncture provides a more direct route to the PVA, but is a difficult procedure that often requires considerable manual force, radiofrequency energy, and often serial balloon dilations to create a large enough orifice for passage of a standard guiding sheath.\textsuperscript{9,11,17} Nevertheless, this technique has emerged as the preferred approach at many centers because of inherent advantages when compared with previous methods.\textsuperscript{10}

The presently described transcaval puncture technique is a modification of the transconduit approach and can be used in a significant proportion of patients after E-TCPC repair. Needle puncture through the native tissue just below the conduit results in a comparatively effortless entry into the PVA without the need for serial balloon dilation to advance the dilator and guiding sheath. This often results in a shorter period to achieve PVA access as well as an expected enhanced safety profile. As the prevalence of cavoatrial overlap is associated with time after surgery, it is possible that this technique can be performed in a much larger percentage of older patients than is reported in the present study, in which 18 patients (50%) had CT imaging performed within only a year of surgery.

The mechanism whereby cavoatrial overlap develops is likely related to the fixed size of a rigid extracardiac conduit in the setting of ongoing cardiac growth after surgery. As most pediatric patients currently undergo the E-TCPC operation at a relatively young age (6.9 years in the present study), the conduit length chosen for surgery is normally between 5 and 10 cm in length. After surgery, as the PVA continues to increase in size, the surrounding tissues, including those involving the IVC, must be pulled up around the atrium to accommodate the increased size. As a result, a region of contact may develop between the vena cava and atrium. Presumptive support for this process can be found in the present study findings that demonstrated a strong association between the elapsed time after surgery and the presence of cavoatrial overlap specific to the pediatric surgical population. It is likely that the development of tissue overlap after ACHD surgery can also occur, but as a result of other factors such as abnormal atrial dilation in the setting of ventricular dysfunction and recurrent atrial arrhythmia. Given the limited number of ACHD patients with E-TCPC in the present study, determination of these factors was not possible.

During the study period, 3 patients underwent PVA procedures via the transcaval route (Table 2; Figure 4). No complications were observed, suggesting that this approach can be both safe and effective. Residual shunting has been cited as a potential drawback to the puncture approach; however, this is unlikely to be a problem and we have therefore opted against the routine use of occlusion devices in our procedures.\textsuperscript{18} Techniques to insure a safe transcaval puncture that we have used at our center include (1) verification of cavoatrial overlap by angiography within the systemic venous circulation and during levophase in $\geq$2 fluoroscopic views, (2)

![Figure 4](image-url). Example of the transcaval puncture technique in a 29-year-old man with extracardiac-total cavopulmonary connection at the age of 17 years. A, Coronal view of computed tomographic scan demonstrating a region of overlap. B, The transseptal needle has been advanced through the inferior vena cava and into the pulmonary venous atrium (PVA). The septum was stained with contrast before entering the PVA to demonstrate compartmentalization. C, Transesophageal echocardiogram demonstrating the position of the transcaval puncture site below the conduit without evidence of pericardial effusion.

**Table 2. Characteristics of Transcaval Puncture Procedures**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Congenital Diagnosis</th>
<th>Age at EC Conduit, y</th>
<th>Age at Intervention, y</th>
<th>Intervention</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Heterotaxy, RAI</td>
<td>17.6</td>
<td>29.6</td>
<td>Catheter ablation</td>
</tr>
<tr>
<td>2</td>
<td>PA/IVS</td>
<td>12</td>
<td>16.7</td>
<td>Fenestration</td>
</tr>
<tr>
<td>3</td>
<td>Heterotaxy, RAI</td>
<td>9.1</td>
<td>18.3</td>
<td>Catheter ablation</td>
</tr>
</tbody>
</table>

EC indicates extracardiac; PA/IVS, pulmonary atresia with intact ventricular septum; and RAI, right atrial isomerism.
intraprocedural transesophageal echocardiography to assess the proposed puncture site and exclude pericardial effusion, and (3) staining of the space between the IVC and PVA with contrast to exclude significant extravasation before entering the PVA. Although extravasation of contrast into the pericardial or pleural space is unlikely to occur in the setting of the typically dense surgical adhesions in this region, the lack of more than minor contrast extravasation during performance of the puncture can be reassuring. Given the present results, we now also recommend preoperative CT angiography to assess the feasibility of this strategy and to guide preprocedural counseling. Either simultaneous upper and lower extremity or lower extremity contrast injection alone is most likely to demonstrate the IVC portion of the anatomy, although usually the conduit itself is readily visualized without the use of contrast.

As the population of E-TCPC patients continues to grow, techniques such as trans caval puncture may become increasingly important. The presently described approach may facilitate the interventional management of this population.

Limitations
This was a retrospective study at a single center and is therefore limited by the study design. For example, patients with prior E-TCPC surgery who underwent follow-up CT imaging during the study period did so for various clinical reasons rather than in a randomized manner. Therefore, the possibility that the indication for imaging was somehow related to the primary study end point (ie, presence of cavoatrial overlap) cannot be definitively excluded. In addition, the observation of clinically significant apposition of the IVC/PVA by CT imaging as defined in the present study may not necessarily equate to the successful performance of a transcaval puncture. However, the results of the present study are informative with respect to understanding the pathogenesis of this phenomenon and suggest that the technique may be useful in a significant proportion of patients. Further clinical study of the risks and benefits of this technique is warranted.

Conclusions
Apposition of the IVC and PVA (cavoatrial overlap) is observed in a significant proportion of patients undergoing the E-TCPC operation. The presence of this anatomy seems to be related to both childhood repair and time elapsed after surgery. If present, this finding may permit a transcaval approach through the region of overlapping tissue, representing a more direct and facile route of access to the PVA for electrophysiological procedures.

Disclosures
None.

References
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