The Fontan operation has been the primary technique for surgical palliation of patients with single ventricle physi-ology since its eponymous description in 1971. Early modi-fications incorporated the morphological right atrium within the systemic venous-pulmonary circulation,1–3 although in its adopted role as the subpulmonary chamber the right atrium hindered rather than contributed to efficient pulmonary blood flow and provided an ideal electroanatomic substrate for atrial arrhythmia that contributed to long-term morbidity and mortality. To overcome these inherent limitations, in 1988, de Leval pioneered a novel surgical variant of the original Fontan procedure, channeling inferior vena caval blood flow directly to the pulmonary artery via an intracardiac baffle created within the posterolateral aspect of the right atrium,4 generating a pulmonary venous atrium from the remaining morphological right and left atria. Subsequently, the extracardiac connection has been increasingly used.5 Although these modifications have improved the hemodynamic profile and reduced arrhyth-mia burden in the short term, arrhythmia remain an important longer-term complication.6,7

By its nature, the total cavopulmonary connection (TCPC) limits access to a significant proportion of atrial myocardium using a standard percutaneous approach. Anterograde access to the pulmonary venous atrium via a fenestration in the intracardiac baffle or a transbaffle puncture is possible in some cases, but allow only a single catheter approach that may significantly hinder accurate determination of arrhythmia mechanism and ablation. The purpose of this study was to determine the varied arrhythmia mechanisms after the TCPC and the feasibility, safety, and success of ablation.

Background—The ability to identify and ablate different arrhythmia mechanisms after the total cavopulmonary connection has not been studied in detail.

Methods and Results—After obtaining Institutional Review Board approval according to institutional guidelines, consecutive patients after a total cavopulmonary connection undergoing electrophysiology study over a 6-year period were included (2006–2012). Arrhythmia mechanism was determined, and the procedural outcome was defined as complete, partial success, or failure. A 12-point arrhythmia severity score was calculated for each patient at baseline and on follow-up. Fifty-seven procedures were performed on 52 patients (18.4±11.8 years; 53.0±27.2 kg). Access to the pulmonary venous atrium was necessary in 33 procedures, via fenestration (16) or transbaffle puncture (17), and in 2 cases, an additional retrograde approach was used. In total, 80 arrhythmias were identified in 47 cases: macroreentrant (n=25) or focal atrial tachycardia (n=8), atrioventricular nodal reentry tachycardia (n=13), reentry via an accessory pathway (n=4) or via twin atrioventricular nodes (n=4), ventricular tachycardia (n=5), and undefined atrial tachycardia (n=21). Procedural outcome in 32 patients who underwent ablation was complete success (n=25), partial success (n=3), failure (n=3), or empirical ablation (n=1). After successful ablation, there was a significant decrease in arrhythmia score over 18.2 (4–32) months follow-up, with a sustained trend even in the face of arrhythmia recurrence (50%).

Conclusions—Arrhythmia mechanism post total cavopulmonary connection is highly varied, encompassing simple and more complex substrates, documentation of which facilitates a strategic approach to invasive arrhythmia management. Despite the anatomic limitations, successful and clinically meaningful ablation is possible.

Mechanism and Ablation of Arrhythmia Following Total Cavopulmonary Connection

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Keywords: arrhythmia ▪ catheter ablation ▪ congenital heart disease ▪ electrophysiology mapping ▪ Fontan procedure ▪ total cavopulmonary connection

Received May 6, 2014; accepted December 23, 2014.
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Circ Arrhythm Electrophysiol is available at http://circep.ahajournals.org DOI: 10.1161/CIRCEP.114.001758
WHAT IS KNOWN

- Atrial arrhythmias are a common source of morbidity in patients after the Fontan operation.
- Although recent surgical modifications decrease or delay the arrhythmia burden, it was unclear how anatomic constraints and surgical intervention affected arrhythmia mechanisms and the ability to map and ablate these arrhythmias.

WHAT THE STUDY ADDS:

- This is the largest published experience to date of catheter mapping and ablation in the total cavopulmonary connection population, demonstrating varying arrhythmia mechanisms.
- Specific mechanisms identified included varying forms of macroreentrant and focal atrial tachycardia, varying forms of atrioventricular nodal reentry, and twin nodal tachycardia.
- Despite the limited access to the pulmonary venous atrium, mapping and ablation are possible with an associated measurable improvement in arrhythmia symptoms, even though a high rate of recurrence was documented.

Methods

Data from the case records of all patients after TCPC palliation undergoing invasive electrophysiology studies (EPS) over a 6-year period (March 2006–March 2012) was collected. This study period was based on a stable operator and institutional procedural experience in invasive electrophysiological assessment of the TCPC patients and familiarity with mapping and ablative technologies, thereby minimizing these as potential confounders in our analysis. All studies were performed on clinically defined indications at Boston Children’s Hospital.

Electrophysiology Study

Electrophysiology studies were performed using standard electrophysiological mapping system (Prucka, GE Medical Systems, Milwaukee, WI) with electroanatomic mapping (CARTO, Biosense Webster, Diamond Bar, CA) used where indicated by clinical need. Intra cardiac echo was used on 18 (32%) cases being used in the majority of the EPS performed after 2009 (Video 1 in the Data Supplement).

CARTOSOUND (CARTO; Biosense Webster, Diamond Bar, CA) was used creating anatomic shells on most cases where intra cardiac echo was used. Catheters were advanced from the femoral veins to the TCPC baffle and when needed retrograde via the femoral artery to the systemic ventricle. Anticoagulation with Heparin was used routinely targeting activating clotting times ≥250 seconds throughout the case. Patients receiving prior oral anticoagulation (9) were either bridged with heparin (7) or the procedure was performed with a therapeutic international normalized ratio (2). Those not formally anticoagulated who required ablation received intravenous heparin until the following day. Venous pressure was measured within the cavopulmonary circulation and angiography used if necessary to identify residual leaks or the position of the fenestration. Access to the pulmonary venous atrium and when necessary the systemic ventricle was anterograde via a baffle leak, a fenestration fashioned at the time of TCPC surgery, retrograde, or by transbaffle puncture using either a standard or radiofrequency (Baylis, Montreal, Canada) needle. Atrial plus/minus ventricular programmed stimulation were used for definition of baseline electrophysiological parameters and arrhythmia induction, including isoproterenol when necessary. Entrainment mapping was used at the discretion of the operator to (i) identify reentrant circuits within either atrium or (ii) to define electrogram (EGM) sequence after cessation of ventricular pacing with 1:1 ventriculoatrial (VA) conduction and atrial entrainment to determine arrhythmia mechanism.

Single ventricular premature beats were introduced during tachycardia at decreasing coupling intervals to assess for atrial reset.

EPS and Arrhythmia Definitions

In analysis of arrhythmia mechanism and underlying electroanatomic substrate, the following definitions were used:

- Macroreentrant atrial tachycardia: organized atrial tachycardia with stable cycle length (CL), EGM morphology, and consistent pattern of atrial activation. In each case, a complete loop of atrial activation was demonstrable on electroanatomic activation maps supported by contact EGM.
- Focal atrial tachycardia: organized atrial arrhythmia with stable CL, EGM morphology, and consistent pattern of centrifugal activation from a focal source.
- Atrial fibrillation: irregular pattern of atrial activation with beat-to-beat variability in CL and morphology with no visually demonstrable area of organized activity.
- Atrioventricular nodal reentry tachycardia (AVNRT): tachycardia with a regular CL and either a 1:1 or 2:1 atrioventricular (AV) relationship, an A-V response on cessation of ventricular pacing with atrial entrainment, and no atrial reset with ventricular premature beats. The VA interval was defined as the onset of the earliest site of ventricular activation to the earliest atrial EGM recorded within the pulmonary venous atrium. In the context of the variable anatomy and limited access to the pulmonary venous atrium, AVNRT was classified as typical if the VA interval was <70 ms and atypical if >70 ms.
- Atrioventricular reentry tachycardia (AVRT): tachycardia with a regular CL and a 1:1 atrioventricular relationship, an A-V response to cessation of ventricular pacing with atrial entrainment, and atrial reset with ventricular premature beats.
- Twin AV nodal tachycardia was defined by the presence of 2 nonpreexcited QRS morphologies each with a distinct His EGM, decremental anterograde and retrograde conduction, and regular tachycardia CL with 1:1 atrioventricular relationship anterograde over 1 AV node and retrograde via the alternate node. An A-V response was seen with cessation of ventricular pacing with atrial entrainment.

Ablation and Definition of Success

Conventional (50 W/70°C) and irrigated radiofrequency (48 W; 30 mls/min) ablation were standardly used. If the ablation site was close to the native conduction system, then cryoablation (−80°C) was used at the discretion of the operator to (i) identify reentrant circuits within either atrium or (ii) to define electrogram (EGM) sequence after cessation of ventricular pacing with 1:1 ventriculoatrial (VA) conduction and atrial entrainment to determine arrhythmia mechanism.

Table. Clinical Arrhythmia Severity Score

<table>
<thead>
<tr>
<th>Category</th>
<th>Score</th>
<th>Category</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Documented arrhythmia</td>
<td>None</td>
<td>Frequency of cardioversion</td>
<td>0</td>
</tr>
<tr>
<td>Nonsustained only</td>
<td>1</td>
<td>One cardioversion</td>
<td>1</td>
</tr>
<tr>
<td>Sustained arrhythmia</td>
<td>2</td>
<td>AAT conversion</td>
<td>2</td>
</tr>
<tr>
<td>Incessant</td>
<td>3</td>
<td>Two or more cardioversions</td>
<td>3</td>
</tr>
<tr>
<td>Arrhythmia severity</td>
<td></td>
<td>Antiarhythmic medications</td>
<td></td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>0</td>
<td>None or Digoxin only</td>
<td>0</td>
</tr>
<tr>
<td>Palpitations only</td>
<td>1</td>
<td>Class II or Class IV</td>
<td>1</td>
</tr>
<tr>
<td>Syncope/CHF/thrombosis</td>
<td>2</td>
<td>Class I or Class III</td>
<td>2</td>
</tr>
<tr>
<td>Cardiac arrest</td>
<td>3</td>
<td>Amiodarone toxicity</td>
<td>3</td>
</tr>
</tbody>
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AAT indicates anti-tachycardia cardioversion; and CHF, congestive heart failure.
during the application of energy in the absence of atrial ectopy and subsequent failure to reinroduce the previously identified tachycardia or noninducibility alone if ablation was performed during sinus rhythm.

Procedural case success was considered complete if all inducible arrhythmia mechanisms were successfully ablated and partial if some, but not all, arrhythmia substrates were eliminated. Arrhythmia recurrence was defined by documented arrhythmia on either ECG or pacemaker telemetry.

Clinical Arrhythmia Severity Score
Arrhythmia burden during the 3 months before EPS was assessed using a clinically relevant 12-point scoring system subdivided into 4 categories (Table). An arrhythmia score was calculated for each patient at baseline and on each clinical encounter during follow-up where data were available.

Follow-Up and Adverse Events
Adverse events were defined as any anticipated or unanticipated event for which injury could or did occur as a consequence of performing the electrophysiological study and assigned a severity level as trivial, minor, moderate, major, or catastrophic. Pre- and postprocedure pulse oximetry data were obtained from clinic visits and follow-up encounters when available, and patient or physician concerns regarding worsening cyanosis were collected.

Statistical Analysis
The Kolmogorov–Smirnov test was used to assess normal distribution. Variables with approximately normal distribution were expressed using mean (standard deviation) and those with nonparametric distribution using median (interquartile range). Comparison between clinical arrhythmia scores at baseline and follow-up was made using the Wilcoxon signed-rank test.

Results
Patient Demographics
In total, 57 electrophysiological procedures were performed in 52 patients (age 18±11.8 years; weight 53±27.2 kg), of whom 48 had an intracardiac and 4 an extracardiac TCPC. Two patients had undergone prior surgical conversion from an atrio pulmonary Fontan with concomitant arrhythmia surgery, and 5 patients had a repeat EPS during the study period. Arrhythmia had been documented before in 45 patients, and 21 antiarrhythmic agent had been used in 38, including amiodarone (n=6). The clinical indications for EPS were palpitations (n=33), syncope with (n=4) or without palpitations (n=2), documented arrhythmia (n=10), cardiac arrest (n=2), and asymptomatic ventricular preexcitation (n=1). Eleven patients had permanent atrial (n=10) or dual chamber (n=1) pacemakers implanted. Overall follow-up of >3 months was available in 42 patients (81%) with no follow-up in 10 (19%).

Catheterization Data
The mean cavopulmonary pressure measured in 46 patients was 16.1±4.1 mm Hg. Access to the pulmonary venous atrium was required on 33 occasions and was achieved via a fenestration (n=16), using a transbaffle needle (n=14) or radiofrequency puncture (n=3). A retrograde aortic approach was used in combination with the transbaffle approach in 2 cases.

Arrhythmia Mechanism and Ablation
Eighty distinct arrhythmia substrates (CL, 312±78ms) were induced in 47 patients further defined as follows:

Macroleientrant Atrial Tachycardia
Twenty-five different circuits (CL, 277±55ms) were identified in 17 patients (range 1–4 per patient) using activation mapping alone (13) or in combination with entrainment mapping (12). Using activation mapping, 92%±5% of the tachycardia CL could be accounted for. The cavotricuspid isthmus or CTI (cavomtricial in L-looped ventricles) was critical to 14 circuits (56%), all of which were successfully ablated at this site, either within both the intracardiac baffle and pulmonary venous atrium (10) or in the pulmonary venous atrium alone (4; Figure 1). A further 11 circuits (44%) were independent of the CTI, of which 8 were located and successfully ablated within the intracardiac baffle and 3 were confined to the pulmonary venous atrium. Of the latter, 2 were successfully ablated at the mitral isthmus and posterolateral chamber, respectively, and a third circuit appeared dependent on an inaccessible section of the anterior morphological right rim. In the 5 patients without a CTI-dependent circuit, 2 had empirical CTI ablation as part of the overall ablative strategy.

Focal Atrial Tachycardia
Eight focal tachycardias (CL, 380±57ms) were identified in 5 patients using activation mapping of sustained tachycardia in 7 cases and P-wave morphology during atrial ectopy in 1 case where tachycardia could not be sustained. Six foci were within the intracardiac tunnel and 2 in the pulmonary venous atrium (Figure 2). All 7 sustained tachycardias were successfully ablated.
AVNRT

Thirteen different AVNRT patterns were seen in 8 patients, of which 7 were typical (VA interval 33–65ms) and 6 were atypical (VA interval 176–260ms). Four patients had both typical and atypical patterns, 1 had 2 atypical patterns (Figure 3A) and 1 had typical AVNRT in the upper of twin AV nodes. During programmed atrial stimulation, dual AV nodal physiology as defined by recognized criteria was seen in only 1 patient. Using single ventricular premature beats in tachycardia at progressively shorter coupling intervals to ventricular refractoriness (maximal V1–V2 interval achieved 110–180 ms), atrial reset was not encountered in any patient. Transient 2:1 AV block was seen in 2 patients during tachycardia without change in the VA interval. Ablation was attempted in 7 of 8 patients using radiofrequency alone (n=2), cryotherapy alone (n=3), or both (n=2), with successful modification of AV nodal conduction properties in 6 patients.

Accessory Pathway Mediated AVRT

Orthodromic AVRT was seen in 4 patients. Two patients had manifest preexcitation on surface ECG, 1 of whom was asymptomatic with rapid pathway conduction demonstrable on a prior limited study. The pathway refractory period was 280 ms in both cases. Both cases with concealed pathways showed VA conduction that was nondecremental and resistant to adenosine. AVRT was induced in each case, although the preexcitation index was calculated in only 1 case (70 ms) because of hemodynamic instability or CL variability in the others. A single accessory pathway was seen in each patient (2 posteroseptal and 2 left posterior), and ablation was successful in all 4 cases. Of note, both patients with concealed pathways had undergone prior ablation of manifest pathways located at different anatomic sites.

Twin AV Nodal Tachycardia

Twin AV nodes facilitated 4 patterns of reentrant tachycardia in 3 patients who all had the anatomic triad of atrial isomerism, atrioventricular septal defect, and discordant VA connections. In 2 cases, anterograde conduction was exclusively via the superior AV node during tachycardia and via either the superior or inferior AV node in a single case (Figure 3C). Ablation was successfully performed in 1 patient and deferred in 2 because of the risk of atrioventricular block in minimally symptomatic patients.

Other Atrial Arrhythmias

An additional 21 atrial arrhythmias were induced in 14 patients, which led to hemodynamic instability (n=2), were not considered clinically relevant (n=6), degenerated to atrial fibrillation necessitating cardioversion (n=1), or were nonsustained (n=12). These were not mapped in sufficient detail to define mechanism nor were ablation performed.

Ventricular Tachycardia

Ventricular tachycardia was recorded in 5 patients, 2 of them with previously documented ventricular tachycardia. The 3 remaining had a history of syncope and were inducible via ventricular drive trains with 3 extrastimuli. One patient underwent ablation for repeated episodes of monomorphic ventricular tachycardia, despite amiodarone therapy, which was ultimately unsuccessful. The clinical relevance of ventricular tachycardia in the other 4 patients remains uncertain. None of these patients had atrial arrhythmias identified during the study.

Noninducible Patients

Of the 5 patients with no inducible arrhythmias, 4 patients were symptomatic with atrial tachycardia pre-EPS, with tachycardia documented in 3 patients on prior noninvasive assessment. The fifth patient, who had no prior arrhythmia history but known severe ventricular dysfunction and prior cardiac arrest, underwent empirical CTI ablation based on the possibility of rapidly conducted atrial tachycardia.

Patient Outcomes

Ablation and Arrhythmia Recurrence

Overall, 32 of the 52 patients underwent ablation, complete procedural success was achieved in 25 patients (76%), partial success in 3 (9%), failure in 3 (9%), and empirical ablation in 1 (3%). Five patients had second procedures in this time. One patient did not undergo ablation on the first EPS, but subsequently had a successful ablation. Four had second procedures for arrhythmia recurrence at 8 days, 2, 11, and 39 months all following acutely successful ablation procedures. The same arrhythmia mechanism was seen in 1, but a different mechanism in 3. The second procedure was successful in 2, partially successful in 1, and no ablation performed in the final patient because of induction of only nonspecific atrial arrhythmia. Including patients who underwent 2 procedures, of the 28 patients who had an initial ablation with either complete
or partial success, follow-up data were available for 22 (79%), of whom 11 (50%) had documented arrhythmia recurrence.

Clinical Arrhythmia Severity Score
Over a median follow-up of 18.2 (4–33) months in 45 patients, 5 (2–8) arrhythmia scores were recorded per patient. Arrhythmia scores are displayed in Figure 4. Scores at follow-up in those with documented arrhythmia recurrence were higher than those without, although a trend in reduction from preprocedure scores was still evident at >24 months follow-up despite recurrence. In those with no arrhythmia recurrence,
the median arrhythmia score at 24 months remained at ≤1, with the loss of statistical significance likely relating to small numbers.

**Adverse Events**

Two major adverse events occurred during the study. A 47-year-old man died 6 days after an unsuccessful procedure after the onset of a wide complex tachycardia from which he could not be resuscitated. A 31-year-old male suffered a pulmonary embolus to the left lung diagnosed on thoracic CT scan, from which he made a full recovery. In addition, 1 patient who had previously suffered an out-of-hospital cardiac arrest before EPS died from multiorgan failure 1 month later, having undergone an empirical cavotricuspid isthmus ablation during that admission. Minor adverse events included 2 cases of transient atrioventricular block after AVNRT and AVRT ablations, respectively, and 1 case each of groin hematoma and femoral venous bleeding postprocedure.

There were 4 patients in whom an arbitrary drop in the pulse oximetry of >5% was recorded when the preprocedure saturation was compared with that during follow-up clinic encounters. Two of them had a transbaffle puncture performed during the procedure and 2 did not, and no study patients had worsening cyanosis recorded as 1 of their main follow-up complaints or as a concern from their referring cardiologist.

Figure 4. Clinical arrhythmia severity score at baseline and during follow-up. The P value for comparison between baseline and the postprocedure encounters is displayed under the time interval. EPS indicates electrophysiology studies.
Discussion
These findings represent the largest published experience of catheter ablation after the TCPC reported to date, providing novel information on both arrhythmia mechanism in a heterogeneous population with single ventricle physiology and the feasibility of detailed mapping despite the anatomic constraints. The major findings are as follows: first, arrhythmia mechanism is highly varied, encompassing reentrant circuits, focal atrial tachycardia, and the more common substrates, atrioventricular nodal, and atrioventricular reentry using either an accessory pathway or second AV node. Second, despite limited catheter access to the pulmonary venous atrium, successful mapping, mechanistic determination, and delivery of ablation lesions is possible. Third, using a clinically relevant arrhythmia scoring system, successful ablation is associated with a reduction in score with a trend that persisted even in the presence of arrhythmia recurrence.

Arrhythmia Mechanism
A mechanism for reentrant arrhythmias after the intracardiac TCPC was first proposed using animal models. Suture lines created along the posterolateral wall of the right atrium created a line of intercaval conduction block, permitting the development of a reentrant circuit using the tricuspid annulus as the anterior central barrier, analogous to classical atrial flutter where the crista terminalis acts as the posterior obstacle. Reentry could be induced acutely using programmed extrastimuli or was initiated spontaneously by atrial extrasystoles. Initiation of tachycardia could be prevented by creation of a further line of fixed block between the suture line and the tricuspid annulus to interrupt the circuit. The data presented here would support the clinical relevance of peri-tricuspid circuits and 6 of 8 focal tachycardias were located within the intracardiac baffle after the ablation and a fatal arrhythmia storm several days after a failed procedure. These patients, only 2 cases of transient AV block occurred. Synchronous dual chamber pacing in the single ventricle population, a major issue in prior iterations of the Fontan procedure. Five patients had a second procedure during the study period, with a different mechanism seen in 3. This suggests an ongoing process of remodeling and arrhythmogenesis secondary to the underlying hemodynamic limitations of the circulation, as noted in previous reports. However, the wisdom of using arrhythmia recurrence as the only marker of success for electrophysiology study and catheter ablation has been questioned previously, prompting the development of the arrhythmia scoring system used here. The persistent improvement seen in those who underwent ablation, and the sustained trend even in the face of arrhythmia recurrence, supports the notion that symptomatic improvement is not driven only by complete arrhythmia elimination.

Two serious adverse events occurred during the study; a pulmonary embolus presumably related to thrombus developing within the intracardiac baffle after the ablation and a fatal arrhythmic storm several days after a failed procedure. These serve as a reminder of the potential instability of patients after single ventricular palliation and the importance of vigilant peri-procedural care. Despite the potential issues related to catheter stability during ablation and potential for anomalous locations of the atrioventricular conducting system within these patients, only 2 cases of transient AV block occurred. Synchronous dual chamber pacing in the single ventricle population not only necessitates epicardial lead placement with the associated procedural morbidity, but may be associated with significant decrease in ventricular function and change in morphology secondary to the dysynchronous effects of single site pacing. Cryoablation with the benefits of catheter adhesion may limit the potential for AV block.

Study Limitations
This study is a retrospective analysis of 52 patients undergoing clinically necessitated electrophysiology studies and ablation. The specific approach to each case was based on operator preference rather than a predetermined protocol. Limited number of catheters within the pulmonary venous atrium during tachycardia prevented the use of certain pacing maneuvers that may have provided additional benefit in mechanistic
determination, for example, assessment of the postpacing interval corrected for AH delay to differentiate AVNRT from AVRT.20 The analysis of the arrhythmia score on the sub-group of patients with arrhythmia recurrence was limited by the fact that a documented arrhythmia itself would by definition increase ≥1 point and by ≤3 the total score. In this setting, the fact that there was a strong sustained trend of lower arrhythmia scores would suggest improvement in the overall clinical status that was reflected on the other measured components of the scale. Finally, because of the referral pattern to our institution, follow-up data are not uniform or complete.

Conclusions
Arrhythmia mechanism after the total cavopulmonary connection is varied and relates to the underlying anatomy, surgical procedures, and ongoing hemodynamic effects on atrial myo-cardium. Despite the anatomic limitations imposed, successful mapping and ablation are possible with sustained clinical benefit. Based on the findings reported here, a sequential approach to arrhythmia mechanistic determination may be adopted.

Sources of Funding
This study was possible because of generous support from the Sean Roy Johnson Research Fund

Disclosures
J.K. Triedman, MD, is a consultant for Biosense Webster, Inc. and Boehringer Ingelheim with a modest level of reimbursement (<10,000 US/year). D.J. Abrams, MD, MRCP, is a stock owner in Johnson and Johnson <50,000 US.

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*Circ Arrhythm Electrophysiol.* 2015;8:318-325; originally published online January 12, 2015; doi: 10.1161/CIRCEP.114.001758

*Circulation: Arrhythmia and Electrophysiology* is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 1941-3149. Online ISSN: 1941-3084

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http://circ.ep.ahajournals.org/content/8/2/318

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SUPPLEMENTAL MATERIAL

Video 1: Demonstration of most common uses of intracardiac echocardiography (ICE) during diagnostic and ablative procedures in patients following total cavopulmonary connection.

Intracardiac echocardiography (ICE) provides detailed definition of the cardiac anatomy, specifically the position of the atrioventricular valves, which may act as central obstacles to conduction in reentrant circuits. Additionally ICE may help identify the precise location of baffle fenestrations guiding catheter access to the pulmonary venous chamber, and demonstrate good tissue contact at during radiofrequency ablation.