Mechanism and Ablation of Arrhythmia Following Total Cavopulmonary Connection

Running title: Correa et al.; Arrhythmia post TCPC

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Abstract:

**Background** - The ability to identify and ablate different arrhythmia mechanisms following the total cavopulmonary connection (TCPC) has not been studied in detail.

**Methods and Results** - After obtaining IRB approval according to institutional guidelines, consecutive patients following a TCPC 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 ± 27.2kg). Access to the pulmonary venous atrium was necessary in 33 procedures, via fenestration (16) or transbaffle puncture (17) and in two 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 empiric ablation (n=1). Following 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 TCPC is highly varied, encompassing simple and more complex substrates, documentation of which facilitates a strategic approach to invasive arrhythmia management. Despite the anatomical limitations successful and clinically meaningful ablation is possible.

**Key words:** congenital heart disease, Fontan procedure, catheter ablation, electrophysiology mapping, arrhythmia, total cavopulmonary connection
Introduction

The Fontan operation has been the primary technique for surgical palliation of patients with single ventricle physiology since its eponymous description in 1971. Early modifications incorporated the morphological right atrium within the systemic venous-pulmonary circulation\(^1\)\(^-\)\(^3\), although in its adopted role as the sub-pulmonary 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” (PVA) from the remaining morphological right and left atria. Subsequently the extra-cardiac connection has been increasingly used\(^5\). While these modifications have improved the hemodynamic profile and reduced arrhythmia burden in the short term, arrhythmia remain an important longer-term complication\(^6\)\(^-\)\(^7\).

By its very 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 are 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 following the TCPC and the feasibility, safety and success of ablation.

Methods

Data from the case records of all patients following 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 EP 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 (ICE) was used on 18 (32%) cases being employed in the majority of the EPS performed after 2009 (video 1). CARTOSOUND (CARTO, Biosense Webster, Diamond Bar, CA) was utilized creating anatomic shells on most cases were ICE was employed. 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 (ACT) >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 INR (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 (VPBs) 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:

- **Macreentrant 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 (AF)**: 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 relationship, an AV response on cessation of ventricular pacing with atrial entrainment, and no atrial reset with VPBs. 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 PVA, 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 AV response to cessation of ventricular pacing with atrial entrainment, and atrial reset with VPBs.

Twin AV nodal tachycardia was defined by the presence of two non-preexcited QRS morphologies each with a distinct His EGM, decremental anterograde and retrograde conduction, and regular tachycardia CL with 1:1 atrioventricular relationship anterograde over one AV node and retrograde via the alternate node. An AV response was seen with cessation of ventricular pacing with atrial entrainment.

**Ablation and definition of success**

Conventional (50W/70°C) and irrigated radiofrequency (48W; 30mls/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 individual operator. Ablation was considered successful for any given mechanism if termination of arrhythmia was seen during the application of energy in the absence of atrial ectopy and subsequent failure to reinduce the previously identified tachycardia, or non-inducibility 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 three months prior to EPS was assessed using a clinically relevant 12-point scoring system sub-divided into four categories (table 1). An arrhythmia score was calculated for each patient at baseline and on each clinical encounter during follow-up where
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 EP study, and assigned a severity level as trivial, minor, moderate, major or catastrophic. Pre and post procedure pulse oximetry data was 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 non-parametric 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 of 57 EP procedures were performed in 52 patients (age 18.4 ± 11.8 years; weight 53 ± 27.2kg), of whom 48 had an intracardiac and 4 an extracardiac TCPC. Two patients had undergone prior surgical conversion from an atriopulmonary 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 at least one anti-arrhythmic 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 pre-excitation (n=1). Eleven patients had permanent atrial (n=10) or dual chamber (n=1) pacemakers implanted. Overall follow-up of greater than 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 ± 4.1mmHg. 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 two cases.

**Arrhythmia mechanism and ablation**

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

**a) Macroreentrant 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 (cavomitrail 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, two 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 atrium. In the 5 patients without a CTI-dependent circuit, 2 had
empiric CTI ablation as part of the overall ablative strategy.

b) 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.

c) 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, one had two atypical patterns (figure 3a), and one 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 one patient. Using single VPBs in tachycardia at progressively shorter coupling intervals to ventricular refractoriness (maximal V1-V2 interval achieved 110-180ms), 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 RF alone (n=2), cryotherapy alone (n=3) or both (n=2), with successful modification of AV nodal conduction properties in 6 patients.

d) Accessory pathway mediated AVRT

Orthodromic AVRT was seen in 4 patients. Two patients had manifest pre-excitation on surface ECG, one of whom was asymptomatic with rapid pathway conduction demonstrable on a prior limited study. The pathway refractory period was 280ms in both cases. Both cases with concealed pathways showed VA conduction that was non-decremental and/or resistant to adenosine. AVRT was induced in each case, although the preexcitation index was calculated in
only one case (70ms) due to 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 anatomical sites.

e) Twin AV nodal tachycardia

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

f) 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 AF necessitating cardioversion (n=1) or were non-sustained (n=12). These were not mapped in sufficient detail to define mechanism nor was ablation performed.

g) Ventricular tachycardia

Ventricular tachycardia (VT) was recorded in 5 patients, two of them with previously documented VT. The three 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 VT despite amiodarone therapy, which was ultimately unsuccessful. The clinical relevance of VT in the other 4 patients remains uncertain. None of these patients had atrial arrhythmias identified during the study.
h) Non-inducible patients

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

Patient outcomes:

Ablation & 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 empiric 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 due to induction of only non-specific atrial arrhythmia. Including patients who underwent two procedures, of the 28 patients who had an initial ablation with either complete or partial success, follow-up data was 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 pre-procedure 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 following 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, one patient who had previously suffered an out-of-hospital cardiac arrest prior to EPS died from multi-organ failure 1 month later, having undergone an empiric cavotricuspid isthmus ablation during that admission.

Minor adverse events included two cases of transient atrioventricular block following AVNRT and AVRT ablations respectively, and one case each of groin hematoma, and femoral venous bleeding post-procedure.

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

**Discussion**

These findings represent the largest published experience of catheter ablation following 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 anatomical 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 utilizing 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 following 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 extra-stimuli 15, or was initiated spontaneously by atrial extra-systoles 16. 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 17. The data presented here would support the clinical relevance of peri-tricuspid circuits traversing the CTI, which was the most common single mechanism seen in 14 of 25 cases, suggesting this is a site to target using activation and/or entrainment mapping. In all CTI-dependent circuits, successful ablation required access to the pulmonary venous atrium to create the line between the tricuspid annulus and inferior vena cava; no complications related to transbaffle puncture were encountered.

Interestingly, 8 of the 11 non-CTI dependent reentrant circuits and 6 of 8 focal tachycardias were located within the intracardiac baffle, suggesting that the hemodynamic
burden imposed by the pulmonary circulation has the ability to remodel a small section of atrial myocardium including the crista terminalis sufficiently to facilitate arrhythmia.

More simple substrates including AVNRT and AVRT via either an accessory pathway or a second AV node were prevalent, accounting for 21 different arrhythmia in 15 patients. Those with relatively slow cycle length and 1:1 AV relationship may appear similar to focal atrial tachycardia, although accurate differentiation can be performed using simple pacing maneuvers such as the AV response on cessation of pacing.\(^8,9\). The emergence of more common arrhythmia substrates and specifically AVNRT may reflect the natural history of congenital cardiac surgery over the last three decades, where palliative surgery was offered to infants with increasingly complex cardiac disease potentially involving the intrinsic conduction system. In this study 4 of 8 patients with AVNRT had both typical and atypical variants, much higher than would be typically seen in patients with a normal heart\(^18\). Such mechanisms should be actively considered in the presence of a 1:1 AV relationship, and may be highlighted by those with recurrent, spontaneously terminating arrhythmia.

Empiric ablation at sites of slow conduction and/or low electrogram amplitude with a view to prevent further arrhythmia recurrence was not routinely performed during this study. It was our practice to address all inducible arrhythmias where possible. The decision to undertake such an ablative strategy should be balanced between the true potential for future arrhythmia reduction and the potential for incomplete, pro-arrhythmia lesions, and should be performed in a highly individualized manner.

**Patient outcomes**

Despite advances in arrhythmia mapping and ablative technology, arrhythmia recurrence remains significant in the TCPC 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 following 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 dyssynchronous 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 pre-determined 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 post-pacing interval corrected for AH delay to differentiate AVNRT from AVRT. The
analysis of the arrhythmia score on the subgroup of patients with arrhythmia recurrence was
limited by the fact that a documented arrhythmia itself would by definition increase at least 1
point and by up to 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, due to the referral pattern to
our institution, follow-up data is not uniform or complete.

Conclusions
Arrhythmia mechanism following the total cavopulmonary connection is varied, and relates to
the underlying anatomy, surgical procedures, and ongoing hemodynamic effects on atrial
myocardium. Despite the anatomical 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.

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Dominic J Abrams MD, MRCP: Stock owner in Johnson and Johnson less than 5,000 US.
References:


Table 1: Clinical Arrhythmia Severity Score

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<td>Non sustained only</td>
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<tr>
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<tr>
<td>Incessant</td>
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Figure Legends:

Figure 1: Activation map of the pulmonary venous atrium in a macroreentrant atrial tachycardia traversing the cavomitrail isthmus. The images depict an electroanatomic map of the pulmonary venous atrium (anteriorly) and the lateral tunnel (posteriorly) viewed from the left anterior oblique/caudal projection in a patient with L-looped transposition of the great arteries following the total cavopulmonary connection. Activation is color-coded, where red denotes early and purple late sites of local electrogram acquisition within a pre-determined time frame, and shows counterclockwise activation of an isthmus dependent circuit around the mitral valve annulus.
Activation within the lateral tunnel is not displayed.

**Figure 2:** Activation map of a focal tachycardia. An electroanatomic map of the the lateral tunnel demonstrates a typical centrifugal activation pattern of a focal atrial tachycardia spreading anteriorly and inferiorly away from an area of scar (grey).

**Figure 3:** Contact electrograms recorded during tachycardia. Surface electrograms recorded from leads I, aVF, V1 and V6 are displayed with contact bipolar intracardiac electrograms. Panel (a) depicts a patient with two atypical AVNRT circuits, with catheters positioned within the intracardiac baffle (REF), on the septum within the pulmonary venous atrium (MAP) and retrograde via the aorta to the systemic ventricle (RV). On the left hand image the cycle length is 400ms and VA interval 120ms, and on the right 363ms and 236ms, respectively. Successful slow pathway ablation was performed using radiofrequency and cryoablation. Panel (b) demonstrates a patient with atypical AVNRT (VA interval 230ms) with catheters positioned in the systemic ventricle via a transbaffle puncture (RV) and within the intracardiac baffle (REF). The return electrogram sequence on cessation of ventricular pacing with atrial entrainment shows a pseudo AAV response, where the last paced ventricular complex relates to the second atrial return electrogram (A*), which is then followed by another ventricular complex. Panel (c) shows two different circuits in a patient with twin AV nodes where catheters have been positioned within the intracardiac baffle (REF), at either HIS position (MAP), and retrograde via the aorta to the systemic ventricle (RV). The left and right hand images show a circuit using the upper and lower node, respectively, as the anterograde limb. Note the change in QRS polarity seen in aVF indicative of a switch from a superior to inferior pattern of ventricular activation. (A
denotes the atrial electrogram, V the ventricular, and H the HIS bundle).

**Figure 4:** Clinical Arrhythmia Severity Score at baseline and during follow-up. The p-value for comparison between baseline and the post-procedure encounters is displayed under the time interval.
Lateral tunnel

Mitral valve annulus

LAO/caudal
Arrhythmia Severity Score after EPS

No Ablation

- Initial: n=20
- 3-12 months: n=15
- 12-24 months: n=15
- >24 months: n=20

Ablation Attempted

- Initial: n=32
- 3-12 months: n=16
- 12-24 months: n=14
- >24 months: n=10

Statistical Significance:
- Initial: p=0.474
- 3-12 months: p=0.238
- 12-24 months: p=0.072
- >24 months: p=0.001 (Initial), p=0.002 (3-12 months), p=0.006 (>24 months)
Arrhythmia Severity Score After Successful Ablation

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<td>n=11</td>
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<tr>
<td>3-12 months</td>
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<td>12-24 months</td>
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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.