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# The Role of Atrial Arrhythmia Ablation in Adolescent and Adult Congenital Heart Disease

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## Abstract

**Purpose of Review** Atrial arrhythmias cause significant morbidity in patients with congenital heart disease (CHD). Catheter ablation remains one of the most effective treatment modalities for atrial arrhythmias. However, patients with congenital heart disease present unique challenges for catheter ablation.

**Recent Findings** Recent expert consensus guidelines inform physicians about treating arrhythmias in patients with CHD. These guidelines outline appropriate selection criteria for ablation and highlight treatment alternatives. The authors also suggest electrophysiology laboratory and physician standards for performing these complex procedures. Recent studies report that 51% of atrial arrhythmias in CHD involve the cavo-tricuspid isthmus (CTI), 28% were non-CTI related and two types of IART were present in 21%. These studies link recurrence of tachycardia after ablation to CHD complexity, non-CTI-related arrhythmias, and patients with prolonged intra-atrial conduction. An analysis of patients with displaced AV nodes showed that cryoablation is a safe and effective technology to perform ablation in CHD with perinodal substrates. Changes in surgical Fontan palliation away from intracardiac baffles to extracardiac conduit has hopefully decreased arrhythmia burden in single ventricle patients. However, in those with atrial arrhythmias, access to the atria is complicated by no direct systemic venous access to the heart. Recent single-center and multicenter studies evaluated the success of ablation in these patients and outline safe approaches to transbaffle puncture. Acute success was 83% with similar complication profile to other CHD patients.

**Summary** The anatomic variations of congenital heart disease create special problems for catheter ablation. Teams performing ablation need pre-procedural preparation and specialized understanding of a vast anatomic variation and surgical repairs. This understanding coupled with the knowledge of the pathophysiology of arrhythmia disorders and the biophysics of catheter ablation technology is required to perform successful and safe ablation procedures.

**Keywords** Congenital heart disease · Intra atrial reentrant tachycardia · Catheter ablation · Ablation biophysics · Transbaffle puncture

## Introduction

Congenital heart disease (CHD) affects approximately 1 out of 100 children born worldwide [1]. The success of surgical palliation, repair, and medical management of patients with CHD as infants and children has led to increasing numbers of patients now living into adulthood. It is now estimated that > 90% will live past 18 years of age [2]. In 2010, the number of adult patients living with repaired or palliated congenital heart disease surpassed the number of children [3]. This number is expected to continue to grow until the year 2050 when equilibrium may be reached.

Arrhythmia burden continues to be one of the highest morbidities of patients with CHD. Overall patients with CHD have a 50% chance of developing atrial arrhythmias in their lifetime [4] and ventricular arrhythmias are likely the leading cause of sudden death in patients with CHD with a 100-fold increase in risk compared to age-matched controls [5, 6, 7••].

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## Ablation Guidelines

Recent expert consensus guidelines outline the treatment recommendations for arrhythmias in patients with CHD

[7••, 8••]. Treatment options included ablation procedures in addition to medical management with antiarrhythmic medications and device management with pacemakers and ICDs. Catheter ablation is indicated for CHD patients with atrial tachycardia and preferred over long-term antiarrhythmic medications [7••, 8••]. The authors also recommend using a 3-dimensional (3D) mapping system to perform the ablation procedure as well as irrigated or large tip radiofrequency ablation catheters for ablation of postoperative atrial tachycardia. For the first time, atrial fibrillation was included in the guidelines and pulmonary vein isolation was considered useful (grade IIa (level C)) for drug refractory atrial fibrillation in adults with CHD. Physicians should also consider ablation of tachycardia substrate prior to congenital heart disease surgery that will limit venous or chamber access to the heart postoperatively [8••]. Medical management of postoperative atrial arrhythmias for 3–6 months is preferred over catheter ablation; however, in drug refractory postoperative atrial arrhythmia, ablation is considered a useful alternative [8••].

The technical challenges of ablation in CHD patients require modification of the usual techniques and special biophysical considerations of ablation technology. Physicians and staff should be familiar with the nuances of different congenital heart disease anatomy and physiology. Surgical backup should be trained in congenital heart surgery at centers performing ablation in patients with moderate to complex CHD [8••]. Often, hemodynamic catheterization and interventions should be coupled with an electrophysiology procedure with consideration of hemodynamic intervention that can improve arrhythmia burden.

A lifetime of multiple interventional procedures and congenital heart surgery makes CHD patients at particular risk for venous occlusion. Others have surgical palliation or venous anatomy that makes access to the heart challenging. Using alternative ablation catheter entry is often necessary. Internal jugular and transhepatic access may be useful in patients with variations in IVC anatomy or venous occlusion. Transbaffle puncture or retrograde aortic access is required for patients with intracardiac or extracardiac baffles. Long precurved and adjustable sheaths can act as a platform for catheter stability and manipulation. Operators with experience, patience, and willingness to try multiple approaches will likely have the best success.

## Atrial Arrhythmias in Congenital Heart Disease

Patients with CHD can have usual forms of SVT including AVRT from manifest or concealed accessory pathways, ectopic atrial foci, and AVNRT. The approach to ablation is similar as in patients with normal anatomy but can be more challenging given the anatomic changes in CHD. Special circumstances of displaced AV node and Ebstein's Anomaly are considered below.

Congenital heart surgery often involves an atriotomy of the right-sided atrium and may include septal patches or intracardiac baffles with long suture lines that can create areas of delayed conduction and arrhythmia substrate. CHD patients often live for years with abnormal hemodynamics and varying degrees of atrial-ventricular valve leak creating a volume overload and higher pressure in the atria. The atria are often dilated and thick and contain areas of fibrosis all contributing to arrhythmia mechanism. Atrial tachycardia circuits in patients with CHD is most often located in the right-sided atrium. These arrhythmias are usually reentrant but can be triggered or micro-reentrant with a focal origin (also known as non-automatic focal atrial tachycardia NAFAT) [7••, 8••, 9]. Reentrant tachycardia is termed intra-atrial reentrant tachycardia or incisional atrial reentrant tachycardia (IART) to distinguish it from typical right atrial flutter in which there is a similar mechanism but more predictable origin. Typical atrial flutter involves a reentrant circuit with counterclockwise conduction around the tricuspid valve and is treated by creating a line a block in a critical portion of the circuit at the cavo-tricuspid isthmus (CTI). IART may also involve the CTI but often involves other areas of slow conduction to create a reentrant circuit [7••, 8••, 9].

IART is often slower than typical atrial flutter with cycle lengths of 270–450 ms and usually presents as a 2:1 tachycardia. The p waves may appear more discrete rather than the typical saw-tooth pattern of atrial flutter [9, 10]. Clinicians should hold a high index of suspension for these arrhythmias when treating patients with CHD. Often, a mild increase in baseline heart rate and mild symptoms are the only clinical indicator of the arrhythmia. Figure 1 is an ECG of a patient in IART with 2:1 conduction and ventricular rate of 111.

Ablation of IART requires an in-depth knowledge of CHD anatomy including surgical scars and suture lines. Thorough review of the patient's clinical data and operative reports is essential. Special attention should be dedicated to the location of the conduction system that can be located in atypical locations. Care should be taken in each ablation case to find and mark the location of the AV node and His signals. Special care should also be taken on the right lateral atrial wall to find and mark locations of phrenic nerve stimulation. Post-surgical adhesions often fix the phrenic nerve to the lateral wall of the atrium making the phrenic nerve more vulnerable to ablation injury. If phrenic nerve capture is identified at perspective ablation sites, attempted cryoablation can be performed while continuous phrenic pacing is performed from a location cranial to the ablation site such as the right subclavian vein [11•]. If the freeze begins to affect the phrenic nerve, ablation can be terminated with expected return of normal phrenic function similar to the return of AV node function with cryoablation of perinodal substrates.

While arrhythmia circuits in CHD patients can be complex, 51–67% of IART circuits involve the CTI [9, 12, 13••].

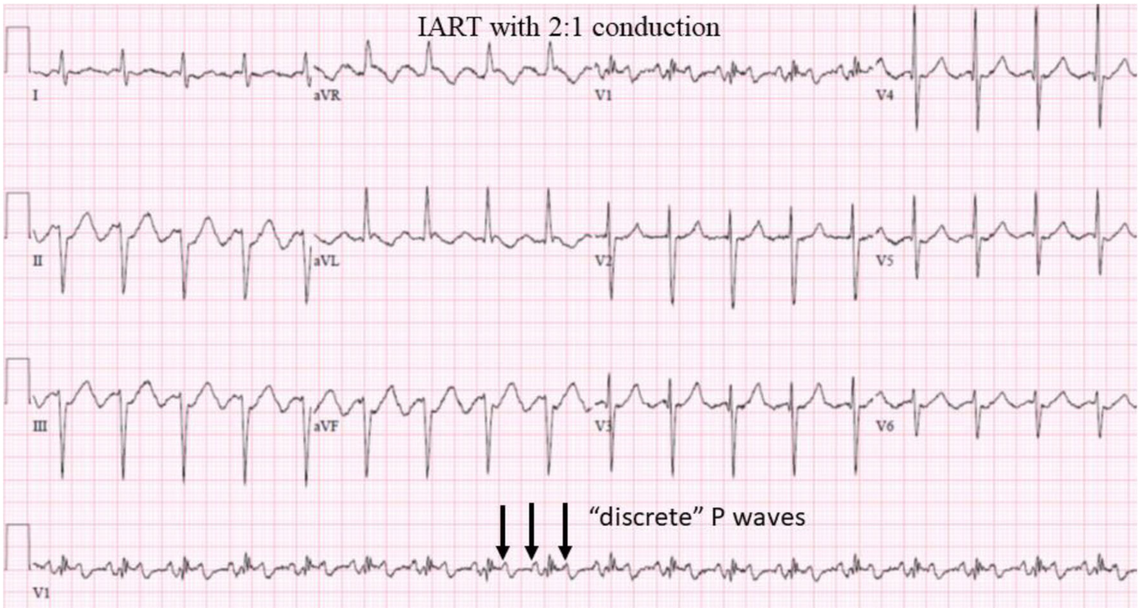


**Fig. 1** ECG of a 28-year-old male with Noonan's syndrome, ASD, and pulmonary valve stenosis. ECG performed during 2:1 IART and ventricular rate of 111 bpm. Note the discrete p waves and relatively slow tachycardia p-p interval of 270 ms

In these cases, ablation requires similar techniques as typical atrial flutter. However, dilation of the atrium can make completion of an isthmus line difficult and thicker atrial wall tissue associated with CHD can make transmural lesions and bidirectional block difficult to achieve. Persistence coupled with the willingness to utilize different long sheaths and higher power ablation is often necessary to achieve success.

A recent analysis of 94 patients undergoing ablation of IART showed that 51% of patients had arrhythmia that involved the CTI, 28% were non-CTI related, and two types of IART were present in 21% [13••]. Complex congenital heart disease and atypical ECG pattern were associated with non-CTI dependent IART. However, 32% of patients with an atypical ECG pattern had CTI-related tachycardia [13••]. In earlier studies, complex congenital heart lesions such as single ventricle patients with Fontan palliation were found to have circuits throughout the atrium with a large proportion (51%) on the lateral right atrial wall and (25%) anterior right atrial wall [9, 14]. Patients with Mustard and Senning baffles for transposition of the great arteries were found to have a large proportion (57%) of circuits involving the CTI [9, 15].

3D mapping systems have become an important resource for ablations in congenital heart disease and are now recommended for use in ablation of moderate to complex CHD [7••, 14]. These systems allow for better collection, assimilation, and visualization of arrhythmia data. 3D mapping not only facilitates success but also simplifies ablation procedures. The newest iterations of contact mapping collect geometry, timing, and voltage data simultaneously. New high-density mapping catheters can be useful in collecting more rapid maps of complex circuits. It is particularly important to make sure the geometry and map collected in the 3D mapping system completely represents the chamber of interest. A segmented CT or MRI scan can be merged with the geometry or angiography and can be performed to facilitate mapping insuring all areas of the atrium are included in the map. This is of particular importance in dilated atria where typical catheter curves do not easily reach the lateral atrial wall and TV annulus. Recently, 3D printing was utilized to make a heart model of a patient with Senning palliation of dTGA prior to ablation. The operators used the heart model to evaluate areas of likely ablation and practice catheter manipulation. This same group also used virtual reality simulation to visualize the heart and plan the ablation [16•].

While 3D mapping systems have improved the ability to assimilate information and identify corridors of arrhythmia conduction, traditional techniques of entrainment pacing aid in identifying critical areas of arrhythmia circuits. Potential areas for ablation should be evaluated and marked with the mapping system. For NAFAT, successful ablation sites often include locations of low amplitude fractionated electrograms with local timing preceding a discrete p wave by 50–80 ms. When a macroreentrant IART is suspected, entrainment pacing should be used to verify that the ablation site is critical to the circuit (post-pacing intervals − tachycardia cycle length ≤ 30 ms, perfect concealed entrainment, or electrogram to p time = stimulus to p time) [10, 17]. These techniques provide reassurance of arrhythmia mechanism and verify locations of appropriate ablation targets. This is important to provide the confidence to persist and achieve a successful line of block especially in challenging cases that may require multiple passes and higher power ablation of the desired area.

Recurrence of IART is reported to be between 20 and 45% depending on multiple factors including complexity of CHD [18••, 19–21]. From a cohort of 94 patients, authors report a recurrence of 38% after the first ablation procedure. Sixty-seven percent of those were the same arrhythmia and 37 were a different arrhythmia than the first ablation [18••]. This population included a large number of moderate and complex CHD patients. Non-CTI-related IART was the highest predictor of recurrence in multivariate analysis (HR 5.06). The authors utilized logistic regression analysis to devise a point system based on independent predictors weighted to their regression values to predict recurrence. The point system is as follows: non-CTI IART = 16 points, PR interval > 200 ms = 14 points, AF induction = 11 points, and previous AF = 11 points. Three groups were identified: low risk (0 to 22 points), medium risk (23 to 36 points), and high risk (> 36 points). Risk of recurrence was 5.8%, 20.0%, and 58.8% for low, medium, and high-risk groups in their cohort, respectively ($p < 0.0001$) [18••]. The authors postulate that the factors of the point system and recurrence of tachycardia are related to the degree of atrial scarring and intra- and inter-atrial conduction delay.

## Special anatomic considerations

### AV node displacement

The location of the AV node is tied to the tricuspid valve and right atrial septum, but can be altered into seemingly unusual positions in CHD patients. Examples of abnormal location include posterior deviation along the atrial septum and anterior to the coronary sinus ostium in patients with AV septal defects. In congenital corrected transposition, the AV node is deviated anterior lateral to a position just below the atrial appendage and can also include double AV nodes with a second posterior node. The AV node is usually positioned in the floor of the blind right atrium near the ostium of the coronary sinus in tricuspid atresia. In double inlet left ventricle, the AV node is usually displaced anterolateral and in double inlet right ventricle, it can be positioned more posterior although often is in a normal location [22–27]. Because of these unusual locations, it is important to locate the bundle of His and the location of the AV node prior to proceeding with ablation. The location can be marked on the 3D mapping geometry. This is most important in patients with single ventricle palliation where inadvertent heart block and the need for long-term pacing would require epicardial pacing lead placement. Cryoablation should be available and considered when arrhythmia substrates are located in the peri-nodal region. With cryoablation, successful ablation can be performed with minimal risk of permanent AV block and reasonable ablation success.

A report of 12 perinodal substrates in 10 patients with CHD anatomy and expected abnormal AV node position showed that cryoablation was acutely successful in 9 (75%) of the 12 substrates. In the three cases that cryoablation was not successful, crossover ablation with RF was successful but attempted only after cryoablation showed no AV node affect in the locations of desired ablation. No AV block occurred in this series and one of the cryoablation substrates recurred 1 month after ablation with an overall follow-up period of 26 months [28••].

### Ebstein's Anomaly

Ebstein's anomaly is a congenital heart defect that consists of an inferior displacement of the septal and posterior leaflets of the tricuspid valve and results in an atrialized portion of the right ventricle [29]. The range of severity extends from mild tricuspid displacement to severe displacement resulting in little right ventricular myocardium. In the more severe forms of Ebstein's anomaly, there is usually associated severe right atrial dilation and AV valve regurgitation. Manifest accessory pathways (WPW) are seen in approximately 21% with multiple pathways common [29, 30]. The supraventricular tachycardia may be poorly tolerated with hemodynamic instability in moderately and severely affected patients. The atrialized portion of the AV grove complicates catheter ablation. This anatomy makes it difficult to locate the true AV annulus and results in fractionated electrical signals around pseudo annulus that are difficult to differentiate from areas of true accessory pathway conduction. The use of 3D mapping system is essential and can be utilized to create a detailed geometry of the AV groove. If necessary, this can be correlated with right coronary angiography to identify the right AV annulus [30, 31]. There are also reports of using a small multipolar electrode wire within the right coronary to mark the AV groove and provide electrogram data similar to a coronary sinus catheter for left-sided accessory pathways [31]. Even with careful mapping, successful ablation in these patients may require persistence and multiple test ablations to find a successful site [30].

### Pulmonary Venous Atrium Substrates, Baffles, and Fontans

Atrial tachycardia in CHD is most often found in the systemic venous atrium (usually morphologic right atrium). The pulmonary venous atrium, however, is an important source of arrhythmia in many patients with complex CHD. In patients with biventricular repair, the left atrium is an uncommon source for atrial arrhythmias unless prior left atrial surgery was performed. A recent single-center analysis of CHD ablation found that there was a negative association with PVA tachycardia without surgical incision in the PVA (OR 0.19, CI 0.05–0.64, $p = 0.10$) and a positive association with PVA tachycardia in patients with prior PVA incision (OR 15.7, CI

4.8–59.9, $P < .001$). In this study, patients with biventricular repair excluding DTGA with intra-atrial baffles most often had NAFAT in the LA (6 out of 11) while patients with intracardiac baffles, i.e., dTGA Mustard or Senning patients most often had macroreentrant IART (10 out of 15) [32••].

Intracardiac baffles found in patients with Mustard and Senning palliation of transposition of the great arteries and in variations of the Fontan procedure present an artificial barrier to catheter manipulation. Components of the arrhythmia circuit are often located in the pulmonary venous atrium and may be transected by the baffle. In these cases, there are two options for catheter placement on the other side of the baffle including the retrograde approach and a transbaffle puncture similar to performing a transeptal puncture for left atrial access in patients with normal cardiac anatomy. Depending on the anatomy, one or both of these approaches may best facilitate success. For both techniques, a detailed understanding of the patient's anatomy is needed and review of imaging studies such as cardiac MRI, CT, or previous catheterization angiograms can be of great benefit. Intra-procedure imaging with TEE, ICE, and angiography facilitate transbaffle procedures which have been performed extensively by interventional congenital cardiologist and electrophysiologists [33–36]. In a recent multicenter report of ablation in patients with extracardiac conduit Fontan, 63% ($n = 46$) were performed via transconduit puncture, 26% across an open fenestration, 13% were performed retrograde. One patient access was obtained via transpulmonary artery puncture and in another access was percutaneous chest wall. Transconduit puncture required balloon dilation to facilitate long sheath placement after needle puncture in 9 pts. Figure 2 shows several approaches for transbaffle puncture for ablation in patients with extracardiac Fontan. In this study, acute success was achieved in 83% and the majority of patients had IART [37••].

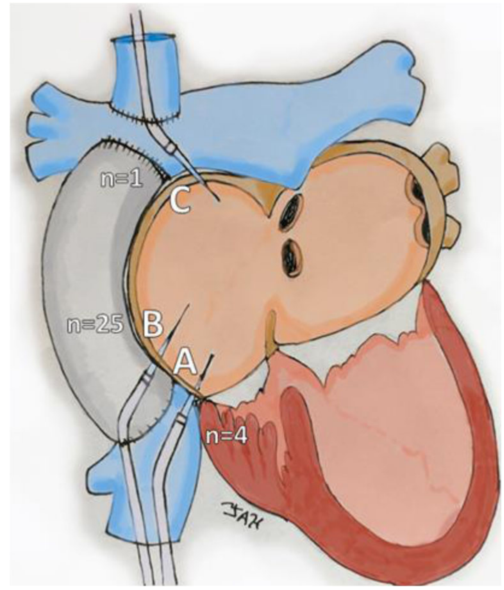


**Fig. 2** Illustration of potential target puncture sites to access the pulmonary venous atrium after the extracardiac total cavopulmonary connection operation. (A) For patients with substantial overlap between the inferior vena cava and the pulmonary venous atrium, a transcaval approach can be performed. (B) The standard transconduit approach is best performed at the lower portion of the conduit, where needle purchase is greatest and forward force can be transmitted without distortion of the needle shape or dissection into the conduit wall. (C) Transpulmonary puncture may rarely be required, for instance, for patients with absent intrahepatic portion of the inferior vena cava as seen with left atrial isomerism (data from Moore JP, Shannon KM, Fish FA, Seslar SP, Garnreiter JM, Krause U, et al. "Catheter ablation of supraventricular tachyarrhythmia after extracardiac Fontan surgery." Heart Rhythm 2016;13:1891–1897 with permission)

## Prosthetic Valves

Some patients with congenital heart disease receive prosthetic valves to alleviate often-long standing atrioventricular or semilunar valve insufficiency or stenosis. These patients are at risk of developing tachycardia with a portion of the critical ablation location covered by the prosthetic valve sewing ring. Successful ablation has been reported by ablating on both the atrial and ventricular aspect of the sewing ring and may also be aided by contact force-sensing catheters [38, 39]. Another more extreme technique recently reported was utilizing a transeptal needle to puncture into the tissue below the sewing ring of a prosthetic TV in a patient with Ebstein's anomaly and performing successful ablation [40•]. The authors caution that in this case, the patient had a planned transcatheter valve replacement intervention and appropriate surgical back up available. As others, they conclude that preoperative EPS and empiric surgical ablation should be performed during prosthetic valve placement surgery.

## Twin AV Node

Twin AV node is a unique anatomic variation found in patients with congenitally corrected transposition of the great arteries especially when associated with AV septal defect or heterotaxy syndrome [41, 42]. One AV node is located in the more usual anterior position found in patients with congenitally corrected transposition and the other in a more inferior and posterior location near the traditional area of the triangle of Koch. These two AV nodes may be connected by a sling of conducting tissue also known as the Monckeberg sling [43]. These patients often have two non-preexcited QRS morphologies suggesting the AV node anatomy and the two morphologies can be elicited with different site atrial pacing at EP study [41, 42]. Typically, a complex reentrant arrhythmia utilizing both AV nodes can be induced and successful ablation is performed by modifying one of the nodes. Care should be taken to identify the AV node that is dominate during sinus rhythm as inadvertent ablation of the wrong node may result in AV block. Cryoablation may act as an important tool for safe ablation attempts [30, 41, 42].

## Ablation Biophysics

The atrium of many CHD patients is abnormal in aspects other than general anatomy. Abnormal hemodynamics often results in severely dilated chambers with thick walls. The lower than usual blood flow of these dilated atria coupled with the thicker walls makes achieving transmural ablation lesions more difficult. Irrigated tip and large tip radiofrequency catheters were designed to create larger ablation lesions by allowing greater convective cooling of the catheter tip to facilitate increased power delivery for a given tip temperature. Increasing ablation power increases the current density within the tissue creating more resistive tissue heating and pushes the zone of ablation deeper within the tissue. Studies show that both large tip and irrigated tip ablation were superior to standard 4-mm-tip catheters in ablation of IART [44, 45]. Irrigated tip catheters are likely to give superior lesion size in areas of low blood flow [46]. Careful fluid balance should be maintained with external irrigation catheters in congenital heart patients that may be more susceptible to the deleterious effects of volume overload. Recent biophysical ablation data suggests that circuit impedance should be considered in titrating power for a given ablation lesion. Locations of high impedance will have lower current draw decreasing lesion size and vice versa locations of lower impedance will have higher current draw and increase lesion size. Figure 3 illustrates this principle and the authors provide a correction facture to consider for a given ablation application. They suggest rather than using constant power for each lesion that the power should be adjusted using the formula power = 40 W × (measured impedance / 120 Ω) [47•].

Cryoablation is a useful technology for ablation in CHD in select cases and provides several advantages that improve safety. First, the cryoablation catheter tip adheres to the myocardium and eliminates the risk of dislodgement. Second, cryoablation lesions are well demarcated and cause little distortion of the tissue and can be placed within venous structures such as the coronary sinus with no reported risk of stenosis. Third, and perhaps the most useful aspect of cryoablation, is the phenomenon of "cryomapping." As the temperature of the tissue cools, the conduction properties of the tissue terminate while the tissue remains viable. There is a longer safety window between the reversible electrical effect and loss of tissue viability than with RF ablation. Essentially, operators can evaluate electrical effect before it is permanent. If the freeze is terminated at this point, the conduction properties of the tissue return to what appears to be normal. This property is most useful for substrates near the AV node and greatly diminishes the risk of inadvertent AV block. An important aspect of cryoablation is that good catheter tip tissue contact is essential. While the ice forming around the catheter tip will likely contact with tissue and adhere to it, heat transfer from the tissue to the catheter will be diminished significantly if the gold tip is not touching the tissue. Ikeda and colleagues

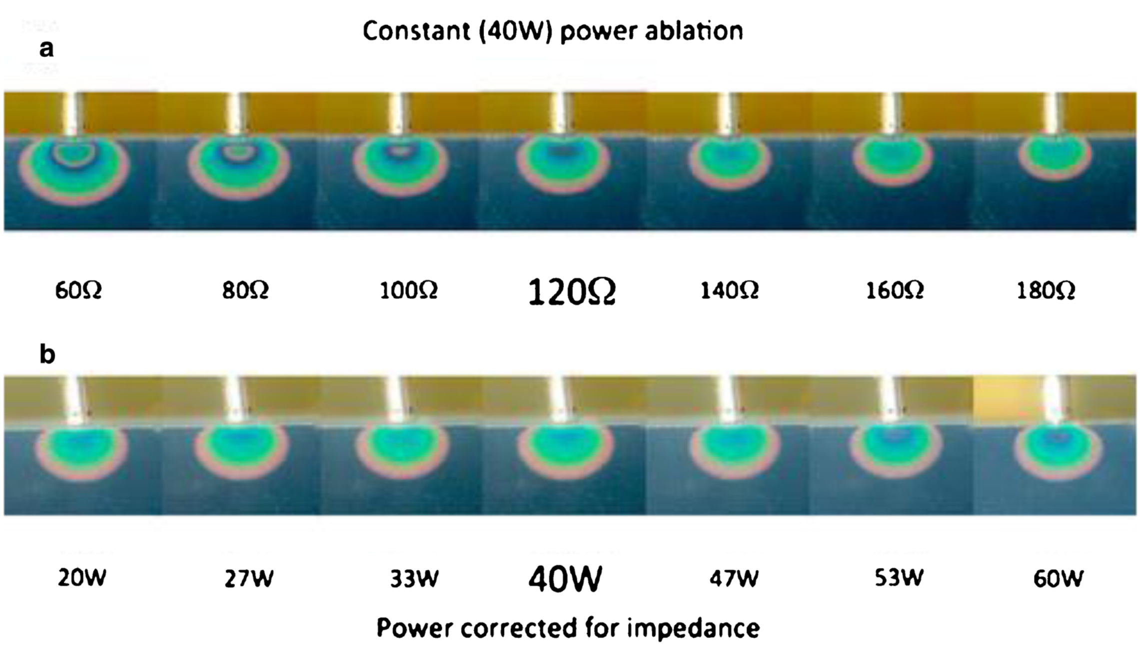


**Fig. 3** **a** Constant power ablation (40 W). RF lesions at circuit impedance from 60 to 180 Ω. The lesion volumes and overheated volumes were significantly higher at lower impedance values. **b** Power corrected for impedance. Lesion volumes were similar to that of 40 W at 120 Ω with no evidence of overheating (data from Bhaskaran A, Barry MA, Pouliopoulos J, Nalliah C, Qian P, Chik W, et al. "Circuit impedance could be a crucial factor influencing radiofrequency ablation efficacy and safety: a myocardial phantom study of the problem and its correction." J Cardiovasc Electrophysiol 2016;27:351-357 with permission)

showed that with the catheter tip about 1 mm above the tissue, very small or no lesions were formed [48]. This is a result of the 100-fold thermal conductivity difference between gold (catheter tip) (314 W/mK) and blood ice (3.19 W/mK) [49]. The ice forming between the catheter tip and tissue acts as an insulator shielding the tissue from heat transfer into the catheter decreasing tissue cooling. Several studies demonstrate that larger lesions are created with cryoablation in low blood flow situations while lesion size is decreased in high flow locations [50, 51]. Cryoablation may be particularly useful in low flow areas such as the coronary sinus and atrial appendages. However, longer ablation times and inability to drag the cryo-catheter for ablation lines limit its use for substrates that require multiple lesions.

## Summary

Unique anatomic variations and unusual tachyarrhythmia substrates found in CHD create special problems for catheter ablation. Teams performing ablation need pre-procedural preparation and specialized understanding of a vast anatomic variation and surgical repairs. This understanding coupled with the knowledge of the pathophysiology of arrhythmia disorders and the biophysics of catheter ablation technology is required to perform successful and safe ablation procedures. It is important to proceed with flexibility and patient persistence with the willingness to try alternative approaches to perform safe and effective procedures.

### Compliance with Ethical Standards

**Conflict of Interest** Thomas Pilcher declares that he has no conflict of interest.

**Human and Animal Rights and Informed Consent** This article does not contain any studies with human or animal subjects performed by any of the authors.



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