

# Catheter Ablation to Treat Supraventricular Arrhythmia in Children and Adults With Congenital Heart Disease: What We Know and Where We Are Going

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**Background:** Catheter ablation has been used to manage supraventricular arrhythmia in children since 1990. This article reviews the history of catheter ablation used to treat arrhythmia in children and discusses new frontiers in the field. We also address ablation in adult patients with a history of congenital heart disease (CHD) that was diagnosed and initially treated in childhood.

**Methods:** We conducted an evidence-based literature review to gather available data on ablation for supraventricular tachycardia in children and adult patients with CHD.

**Results:** Ablations can be performed safely and effectively in children. Complication rates are higher in children <4 years and <15 kg. In one study, the overall success rate of radiofrequency ablation in pediatrics was 95.7%, with the highest success rate in left free wall pathways (97.8%). Recurrence was higher in septal pathways. Cryoablation has been reported to have a 93% acute success rate for atrioventricular (AV) nodal reentrant tachycardia and septal pathways with no risk of AV block and a 5%-9% risk of recurrence. Three-dimensional mapping, intracardiac echocardiography, remote magnetic navigation, and irrigated catheter ablation are new technologies used to treat pediatric and adult patients with CHD. The population of adult patients with CHD is growing, and these patients are at particularly high risk for arrhythmia. A paucity of data is available on ablation in adult patients with CHD.

**Conclusion:** Electrophysiology for pediatric and adult patients with CHD is a rapidly growing and progressing field. We benefit from continuous development of ablation techniques for adults with structurally normal hearts and have the unique challenge and responsibility to ensure the safe and effective application of these techniques in the vulnerable population of pediatric and adult patients with CHD.

**Keywords:** Catheter ablation, child, heart diseases, tachycardia–supraventricular

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## INTRODUCTION

Catheter ablation has been used for management of supraventricular arrhythmias in children since 1990. In this paper, supraventricular tachycardia (SVT) refers to accessory pathway–mediated tachycardia and atrioventricular (AV) nodal reentrant tachycardia (AVNRT); these conditions account for the majority of ablations performed in children. Catheter ablation in children has been proven to be safe and effective. Since the initial ablations in 1990,<sup>1</sup> the indication for catheter ablation has gradually shifted from treatment of medically refractory arrhythmia to first line treatment and patient/parent preference. We review the history of catheter ablation used to treat arrhythmia in children and discuss new frontiers in the field. Our main focus is catheter ablation of SVT in children with structurally

normal hearts. We also discuss ablation in adult patients with a history of congenital heart disease (CHD) that was diagnosed and initially treated in childhood.

## HISTORY OF CATHETER ABLATION IN CHILDREN

Catheter ablation became available as a treatment for SVT in children in 1990. Initially, radiofrequency ablation (RFA) was the only catheter-based technology available for treatment of arrhythmia. RFA uses heat generated by alternating electrical current to scar dysfunctional conduction tissue in the heart. The Pediatric Radiofrequency Catheter Ablation Registry was developed by the Pediatric Electrophysiology Society in 1990 to determine the efficacy and safety of RFA in children.<sup>1</sup> The registry included data on 2 major groups of patients. The first group had ablations

performed in 1991-1995, and the second group had ablations performed in 1996-1999. No significant difference in patient demographics or type of SVT was noted between the 2 groups. Significant improvement was noted in failure rate (50% decrease), fluoroscopy time (20% decrease), and complication rate (25% decrease) in the 1996-1999 group compared to the 1991-1995 group.<sup>1</sup> These data also demonstrated a lower chance of success in right-sided pathways and in patients with increased body weight (>80 kg) in both study groups. The data also showed that the chance of success increased as the number of cases done at the center increased. A body weight <15 kg was associated with a higher complication rate. Age <4 years was also found to be an independent predictor of complications.

Kugler et al showed that a lower risk of AV block correlated with free wall accessory pathways (both right-sided and left-sided) and increased body weight and reported a total death rate of 0.11% (4/3653 patients),<sup>1</sup> which is comparable to similar studies in adult patients.<sup>2-4</sup> One 13-year-old child died 3 hours after successful ablation of a left lateral pathway initially attempted by a retrograde approach and eventually by a transseptal approach. The patient developed chest pain followed by AV block. An autopsy showed thrombosis of the left coronary artery orifice. Two infants died during follow-up. One was a 3-week-old infant who had successful RFA of the left lateral accessory pathway because of incessant SVT despite multiple medications. This patient died 2 weeks after the procedure and was found to have a torn mitral valve leaflet, but the cause of death was unclear. The second infant who died had 2 ablations at 4 and 6 months of age for a left posteroseptal pathway. Death occurred 17 months after the second ablation, and the patient was found to have a posterior left ventricular wall myocardial infarction during autopsy. The fourth death was an 11-year-old child who died 3 months after successful RFA of a left lateral pathway and was found to have ventricular perforation during autopsy.

The 3-year freedom from recurrence rates in the 1997 study by Kugler et al was 71% for accessory pathways and 77% for AVNRT.<sup>5</sup> The rate of follow-up was 76%, thought to be because of asymptomatic patients not following up. Initially, this recurrence result seems disappointing, but if many asymptomatic patients were noncompliant with follow-up vs patients with recurrence who were very motivated to follow up, the data may be skewed toward a seemingly higher recurrence rate than the all-inclusive recurrence rate would be.

The Prospective Assessment after Pediatric Cardiac Ablation (PAPCA) database further demonstrated the safety and efficacy of RFA.<sup>6</sup> This prospective study involved all patients with AVNRT and accessory pathway-mediated tachycardia who underwent electrophysiology study (EPS) and RFA. The 504 cohort-eligible registry participants aged 0-16 years had an overall success rate of 95.7%, with the highest success rate being 97.8% for left-sided free wall pathways. The complication rate was 4%, with hematoma at the catheter site being the most common complication for patients receiving EPS and AV block being the most common complication for patients receiving RFA. AV block was only found in ablations of AVNRT (2.1%) and septal

pathways (3%).<sup>6</sup> A similar study in pediatric patients reported an incidence of 3% of AV block in septal pathway ablation. Overall, the recurrence rate was reported as 12%-14% for septal pathways.<sup>7</sup>

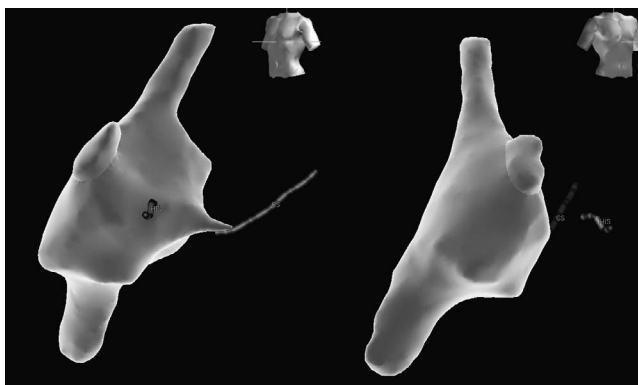
A new technology called cryoablation became available as another option for ablations near the normal conduction system. Cryoablation uses nitrous oxide circulating through the tip of the catheter to cool the tissue. As the tissue cools, if an undesirable effect is seen, the refrigerant flow can be stopped, allowing the tip to rewarm before creating a permanent effect. This important development essentially eliminated the risk of AV block when performing ablations near the AV node. Another advantage of cryoablation is the stability of the catheter as it becomes adherent to the endocardium while frozen. In 2000, the first case of catheter-based cryothermal ablation to eliminate slow pathway conduction for AV nodal reentry tachycardia was reported.<sup>8</sup> Since then, the safety and efficacy of cryoablation have been well established. In a retrospective multiinstitutional study of cryoablation for pediatric AVNRT by Kirsh et al, the acute success rate was reported as 93% without any permanent AV block.<sup>9</sup> In other data reported by McDaniel et al, a 5% recurrence rate of cryoablation is reported in nearly 100 patients (most with AVNRT) during an 18-month period.<sup>10</sup> The only predictor of recurrence was a cryoablation lesion application <4 minutes or catheter tip size <6 mm. A similar study showed a 5% recurrence rate with cryoablation for AVNRT.<sup>11</sup>

The Pediatric Radiofrequency Catheter Ablation Registry was the precursor to the North American Society of Pacing and Electrophysiology expert consensus conference in 2002 that led to development of official indications for RFA in pediatric patients.<sup>11</sup> These guidelines are still applicable in 2016 and have been expanded with specific statements such as management of asymptomatic patients with Wolff-Parkinson-White syndrome.<sup>12</sup>

During the first years of RFA in pediatrics, follow-up data were gathered on patients to evaluate long-term complications.<sup>5</sup> However, to our knowledge, no recent study of long-term complications has been conducted. After 25 years of performing RFA in children, an evaluation of coronary abnormalities, new arrhythmia, and other potential concerns in adult patients treated with RFA as children would be useful. This concept also applies to cryoablation, especially in regard to long-term AV conduction abnormalities.

The databases and registries have helped us understand and manage arrhythmia in extremely young patients.<sup>5,11</sup> Arrhythmia that presents during infancy typically either resolves or goes through a honeymoon period prior to recurrence later in childhood. For this reason and because of the higher complication rates noted in patients <15 kg and <4 years who undergo RFA,<sup>5</sup> pharmacologic therapy is generally preferred in our younger patients (usually <4 years of age) at Ochsner.

Children with CHD have a similar risk for SVT as the general population. However, some forms of CHD are prone to SVT, such as Ebstein anomaly, which is associated with Wolff-Parkinson-White syndrome and a high incidence of multiple pathways. Small case series have shown that RFA in patients with CHD is feasible; however, the acute success rate is lower.<sup>13-15</sup> These patients often have venous anomalies or vascular access issues that make access to



**Figure 1.** Map of the right atrium, superior vena cava, and inferior vena cava created by a 3-dimensional mapping system.

the heart difficult. Normal anatomic landmarks may be absent or abnormal, making an actual ablation challenging.<sup>11</sup> The reported complication and mortality rates for ablation in patients with CHD are slightly higher than in patients with structurally normal hearts.<sup>11</sup>

### MINIMAL FLUOROSCOPY WITH 3-DIMENSIONAL MAPPING

Exposure to radiation during medical procedures carries the known risks of dermatitis, cataracts, thyroid disease, birth defects, and malignancy.<sup>16-18</sup> Because of their long life expectancy, children have more risk from cumulative effects than adults. As with all other procedures involving radiation, the guiding principle for pediatric catheter ablation is as low as reasonably achievable (ALARA). A 2008 study estimated the increase in lifetime risk of fatal malignancy from a single ablation procedure to be 0.02%.<sup>19</sup> This estimate was based on a fluoroscopy time of 14.4 minutes and a highest median radiation dose of 43 mGy. However, it is difficult to justify even a small risk of fatal malignancy to treat a nonfatal disease such as SVT.<sup>20</sup>

The development of 3-dimensional (3-D) electrophysiologic mapping systems in the late 1990s has allowed us to significantly reduce the amount of radiation exposure to children during ablations. This exposure reduction is also advantageous to the physician and electrophysiology laboratory staff who are also exposed to radiation during ablations. Several 3-D mapping systems are available. The 2 used at our institution are the EnSite system (St. Jude Medical, Inc.) and the CARTO system (Biosense Webster, Inc.). EnSite functions by measuring electrical impedances and CARTO by measuring magnetic fields. Both render 3-D geometries of the heart chambers and allow for visualization of the ablation catheter on the mapping system. Figure 1 shows a right atrial map created by a 3-D mapping system. Maps are labeled with the location of normal conduction, valve annulus, and prior lesions. Smith and Clark reported a 95% reduction in fluoroscopy time when 3-D mapping was utilized, and a significant amount of their patients received no fluoroscopy.<sup>17</sup> The patients least likely to receive fluoroscopy were patients with right-sided accessory pathways or AVNRT. Fluoroscopy is still used to perform transseptal procedures and to perform ablations in the left

atrium. Long sheaths, often needed for catheter stability, are not seen on 3-D maps.

A decrease in total procedure time when using 3-D mapping has not yet been consistently reported, likely because of the learning curve associated with performing minimal fluoroscopy procedures.<sup>16,17,20</sup>

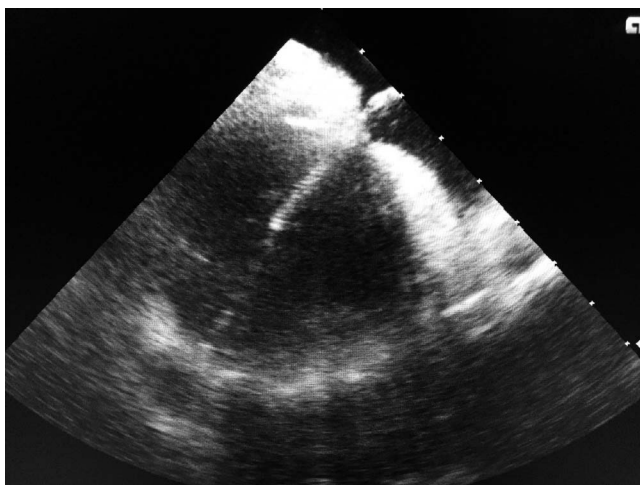
Although the goal of minimizing fluoroscopy or performing procedures without fluoroscopy is certainly ideal, patient safety should not be compromised to accomplish this goal. If the catheter course or location is questionable, fluoroscopy should be used because visualizing more than the tip of the catheter on a 3-D map is not possible.<sup>21</sup> No increase in complications has been reported from the use of a minimal fluoroscopy approach during 3-D mapping.<sup>22</sup>

### INTRACARDIAC ECHOCARDIOGRAPHY

In catheter ablations performed on adult patients, echocardiography is often used as an adjunct to fluoroscopy to perform transseptal puncture to gain access to the left atrium. However, most institutions use fluoroscopy alone for ablations performed on pediatric patients. Daoud suggests that echocardiographic guidance should be the standard of care for all transseptal procedures.<sup>23</sup> The use of intracardiac echocardiography (ICE) in pediatric and adolescent patients has been reported during atrial septal defect and patent foramen ovale device closures.<sup>24</sup> One study reports the use of ICE in patients as young as 10 months of age.<sup>24</sup> To our knowledge, no studies report using ICE guidance for transseptal procedures in pediatric patients, perhaps because of the need for a relatively large additional venous access (9 French) to insert the ICE catheter. At our institution, we have accomplished ICE catheter placement by removing our catheter on the His bundle and replacing it with ICE. We have used this technique in patients as small as 30 kg with no known complications. Clark et al reported using transesophageal echocardiography (TEE) for transseptal procedures.<sup>25</sup> We believe TEE is a good option for small children (<25 kg), but it has several disadvantages. TEE requires equipment and personnel that often are not readily available in an electrophysiology laboratory. In addition, Clark et al report a 1-hour increase in procedure time on average to perform TEE while the patient is under general anesthesia vs using only fluoroscopy. Using ICE during our transseptal procedures has increased the safety of the procedure by giving us direct visualization of the needle/sheath crossing the atrial septum and entering the left atrium and reducing the amount of fluoroscopy used. ICE also has the additional benefit of monitoring for pericardial effusion after transseptal ablations and during left-sided ablations while the patient is heparinized. Figure 2 shows an ICE image of a transseptal needle crossing the foramen ovale from the right atrium to the left atrium.

### REMOTE MAGNETIC NAVIGATION

Remote magnetic navigation (RMN) (Stereotaxis, Inc.) is a technique being used selectively in pediatric patients. Special catheters with built-in magnets are guided in the body by external magnets similar to those used for magnetic resonance imaging (MRI) and are driven using a remote guidance system. A paucity of literature regarding the utility of RMN in pediatric patients is available. Kim et al evaluated 145 pediatric patients undergoing RFA for



**Figure 2. Intracardiac echocardiography shows a transseptal needle crossing the foramen ovale from right atrium to left atrium.**

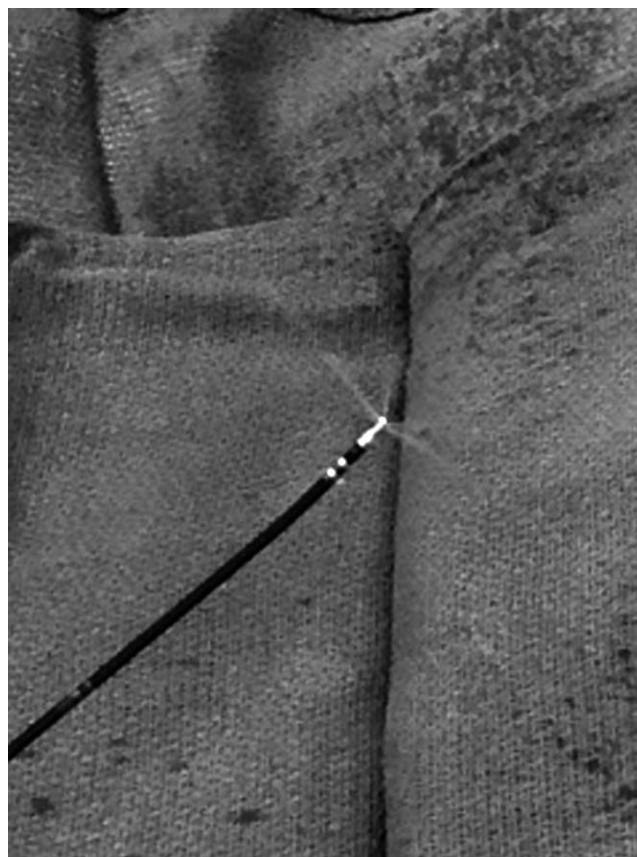
accessory pathway-mediated tachycardias.<sup>26</sup> Seventy-three patients received ablations with RMN, and 72 received manual catheter ablations. Acute success rates were equivalent, recurrence rates were not significantly different, no difference in total procedure time was observed, and no complications were noted in either group. The one advantage that the authors were able to identify was a decrease in fluoroscopy time with RMN compared to manual ablation. The authors considered this time reduction to be at least in part because the catheter is steered by magnets close to the tip of the catheter, leading to improved catheter stability and benefit in regions where catheter stability is an issue such as in right anterior accessory pathways.<sup>26</sup>

Roy et al reviewed the published United Kingdom experience of RMN and noted the benefit of RMN in treating patients with congenital defects of complex anatomy.<sup>27</sup> RMN is particularly advantageous when retroaortic access to the heart is necessary or when the atrium or ventricle is large, allowing for distal tip mapping of all areas of a chamber. RMN also offered improved catheter stability for flutter lines in patients who had atypical scar lines. 3-D image integration on top of the stored fluoroscopy image was also helpful.<sup>27</sup> Given the complex anatomy of some patients with CHD, the RMN catheters are better able to traverse the complex angles than manual catheters that would have limited control in patients with an interrupted inferior vena cava and azygous continuation to the superior vena cava.<sup>27</sup>

A secondary benefit of RMN is the reduced exposure to radiation for all the staff and decreased amount of time the person manipulating the catheters needs to wear lead shielding. However, the cost relative to the perceived benefit for patients and staff makes adoption of this technology a complicated equation in a time of changing healthcare economics.

### IRRIGATED CATHETER ABLATION

Irrigated catheter ablation is a technique frequently used in adult patients. Irrigated tip ablation catheters were first approved for use in the United States in 1999.<sup>28</sup> In



**Figure 3. Irrigated catheter is flushed with saline prior to insertion into a venous sheath for an ablation.**

conventional ablation catheters, radiofrequency energy delivered from the tip of the catheter to the tissue causes resistive heating of the tissue and the surrounding blood stream that can lead to excessive heating at the tip and potential charring. Char is residue that can develop on the tip of the ablation catheter because of incomplete combustion of material exposed to high heat. Irrigated tip ablation catheters have small pores near the tip of the catheter that allow irrigation of the blood during an ablation lesion to create an effective lesion without excessive heat. Irrigated catheters are designed to increase the size and depth of the lesion while decreasing the risks of thrombus formation or char on the catheter tip.<sup>28</sup> Figure 3 shows an irrigated catheter being flushed with saline prior to insertion into a sheath for an ablation procedure. Interest in the use of irrigated ablation catheters in the pediatric population and in adult patients with CHD is increasing, but limited data support their use in these populations. At our institution, we have begun using irrigated tip catheters for many RFAs in children when the procedure is considered to be safe and appropriate. We are currently analyzing our data for publication.

Tanner et al studied the use of irrigated catheter ablation for intraatrial reentrant tachycardia (IART) in patients after surgical repair of CHD.<sup>28</sup> Of the 48 IART ablations studied, 45 (94%) were successful. At follow-up, 92% (33/36) of patients were recurrence free vs 64% of patients in the historical control group. Additionally, the number of lesions was higher and the mean power was lower in the control

group with conventional RFA vs irrigated ablation. Conventional thought is that the surgical scarring from congenital heart repair and the often associated atrial chamber enlargement make placing radiofrequency lesions with nonirrigated ablation more difficult.<sup>28</sup>

### **FLEXIBLE TIP AND CONTACT FORCE**

Two new ablation technologies that are currently being evaluated are flexible tip irrigated catheters and catheters that can demonstrate measured catheter tip contact.

The FlexAbility (St. Jude Medical, Inc.) flexible tip ablation catheter was designed to enhance tissue contact and allow for a deeper lesion compared to standard irrigated catheters. To our knowledge, no data exist to guide use in pediatric or adult patients with CHD. Winterfield et al compared the FlexAbility to the ThermoCool (Biosense Webster, Inc.) and the ThermoCool SF (Biosense Webster, Inc.) in an animal study.<sup>29</sup> The ThermoCool and ThermoCool SF catheters are both irrigated catheters with no flexible tip or contact force capabilities. The FlexAbility was noted to have a lower risk of steam pops than the ThermoCool catheters. Catheter type was not associated with a statistically significant difference in lesion size.<sup>29</sup>

Contact force is another emerging ablation catheter technology. Contact force catheters are designed to sense tissue contact on the distal tip of the catheter to allow for quantification of contact. The goal is to improve tissue contact, resulting in deeper and more accurate lesion placement compared to standard irrigated catheters. Because contact force catheters have only been approved in the United States since February 2014,<sup>30</sup> no significant body of data is available on their utilization in pediatric and adult patients with CHD. Bourier et al examined the accuracy of the technology in a 2016 article.<sup>31</sup> They found the technology to be highly accurate in nonparallel contact at 30 and 60 degrees but less so at 90 degrees. To date, studies on contact force catheters have focused on adult patients with atrial fibrillation. The EFFICAS II trial evaluated guidelines for effective use of contact force catheters and showed that 85% of pulmonary veins remained isolated after 3 months when the contact was maintained with an average of 20 g with a force-time integral (FTI) of 400 g/s per site, a significant improvement compared to the initial trial that had no defined guideline.<sup>32</sup> We have begun using contact force catheters in select circumstances for patients with accessory pathways, and anecdotally, contact force is a helpful adjunct to the electrogram as an additional marker of contact. Further study is warranted to better define the utility and the appropriate grams of force over time measured in FTI needed to successfully ablate accessory pathways. No data are available on average grams of force or FTI for accessory pathway ablations.<sup>32</sup>

### **CONGENITAL HEART DISEASE IN ADULT PATIENTS**

A growing consideration for ablation procedures is the treatment of adult patients who were diagnosed with CHD as children. As heart surgery to treat CHD continues to improve and patients survive longer, the population of adult patients with CHD continues to grow rapidly. More

than 1 million adult patients in the United States have CHD.<sup>33</sup>

As these patients age, they have an inherently higher risk of arrhythmia from the surgical scars combined with their abnormal anatomy and physiology. Ablations in this population are a challenge for electrophysiologists, requiring detailed understanding of the anatomy and of surgical correction to be effective. Cooperative partnerships between electrophysiologists who treat children and adults with CHD can help care for the full spectrum of arrhythmias these patients may encounter. In addition, the use of 3-D mapping and computed tomography (CT) or MRI overlays has some benefit in visualizing the anatomy.<sup>34</sup>

Because of concerns for radiation exposure with CT scans, ultrasound-guided 3-D imaging devices such as the CARTOSound (Biosense Webster, Inc.) have been used to similar effect. Kean et al reported a series of 13 patients with CHD treated with the assistance of CARTOSound devices with intracardiac echocardiography.<sup>35</sup> In patients with CHD with 2 ventricles, the procedural success was 100% compared to 86% in the control population.<sup>35</sup>

Noninvasive arrhythmia mapping may have particular benefit for adult patients with CHD. This technology uses noninvasive multielectrode electrocardiography mapping in conjunction with a CT scan overlay and allows for arrhythmia mapping without having to have catheters in the heart. Noninvasive arrhythmia mapping could be particularly helpful when access to the systemic atrium may be difficult such as in transposition or some single-ventricle hearts. Having an arrhythmia map ahead of time can drastically change the preprocedural planning. Ernst et al published a study of 14 adult patients with CHD in the United Kingdom who underwent noninvasive multielectrode electrocardiography mapping using ecVUE (CardioInsight Technologies, Inc.).<sup>36</sup> The technique was successful at localizing the origin of the arrhythmia and guiding procedural planning, but the patient cohort was small. To our knowledge, no data on similar patients in the United States are available at this time.

In our center, we use a team approach to treat adult patients with CHD, particularly for their complex arrhythmia management. We are fortunate to have the ability to provide comprehensive arrhythmia care with an integrated team of electrophysiologists experienced in treating pediatric and adult patients with expertise in both the structural abnormalities of CHD and the management of arrhythmias such as atrial fibrillation that often arise in adult patients. This team approach gives us the optimal chance of success and enhances the quality and safety in the care of our patients.

### **CONCLUSION**

The field of electrophysiology for pediatric and adult patients with CHD is rapidly growing and progressing. We benefit from continuous development of ablation techniques for adults with structurally normal hearts and have the unique challenge and responsibility to ensure the safe and effective application of those techniques in our more vulnerable population: pediatric and adult patients with CHD.

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