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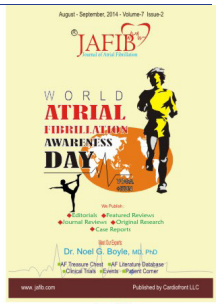

Anatomic Challenges In Pediatric Catheter Ablation

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Abstract

Pediatric patients present unique anatomic challenges for catheter ablation. Small patient size requires special adaptation and understanding to perform safe procedures when clinically indicated. The anatomic variations of congenital heart disease also create problems that require pre-procedural preparation for each case in addition to a 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 are required to perform successful and safe ablation procedures in this special population.

Introduction

The first pediatric radiofrequency ablations for cardiac arrhythmias were performed in the early 1990s.[1,2,3,4] Since then the safety and efficacy of catheter ablation has led to it being considered a first line treatment for many pediatric arrhythmia disorders.[5] Ablation offers a high acute and chronic success rate with low risk of complications.[6,7] While the technology used in pediatric ablation procedures is the same as for adult procedures, pediatric patients come with unique challenges. Smaller patient size is an obvious challenge and patients with congenital heart disease have varied intracardiac and venous anatomy creating additional unique challenges. Arrhythmia substrates in pediatrics are similar to those found in adult patients, however the presentation and distribution of the specific substrates are different.[8] Supraventricular tachycardia accounts for the majority of arrhythmias in children and accessory pathways make up about 75% of cases in older children and 95% in infants. AV nodal reentry tachycardia and ectopic atrial tachycardia account for the remainder with only rare occurrences of typical atrial flutter and ventricular tachycardia.[8] In patients with repaired or palliated congenital heart disease scar related macro reentrant tachycardia predominates usually occurring in the right atrium and right ventricle. These differences coupled with anatomic challenges require special care and caution.

Small Patient Size

A large pediatric ablation registry reported that the complication rate of pediatric ablation procedures was highest for patients less than 15 kg in weight.[4,6] Given this data the North American Society of Pacing and Electrophysiology (NASPE) Expert Consensus Conference writing group recommended that pediatric electrophysiologists use a cut off of approximately 5 years of age and 15 kg for elective ablation procedures.[5] More specifically ablation is reasonable (Class IIB) for patients > 5 years old as an alternative to medical therapy. Some younger patients however may have arrhythmias that are challenging to control with antiarrhythmic medications. In such cases the complication rate of the ablation procedure must be weighed against the morbidity and frequency of tachycardia for each case. The NASPE writing group stated that ablation was reasonable (class IIB) in patients < 5 years old (including infants) "when antiarrhythmic medications, including sotalol and amiodarone, are not effective or associated with intolerable side effects".[5] However, the technical challenges of ablation in these patients require modification of the usual techniques and special biophysical considerations of the technology.

Small Children 1-5 Years

In smaller patients vascular access is challenging and procedures are usually modified and performed with fewer catheters (2-3) than for larger patients.[9,10,11] Several alternatives include 1. Utilizing a transesophageal catheter to pace the left atrium and provide atrial and ventricular electrograms. 2. Using smaller 5 French radiofrequency ablation catheters. 3. Using alternative ablation catheter entry and course in reaching challenging substrates. Examples include RIJ access for right atrial anterior and anterior septal substrates and prolapsing a loop of the ablation catheter across the tricuspid valve to facilitate better catheter stability for right free wall substrates.[12] Long sheaths are utilized for trans-septal access and also act as a platform for catheter stability and manipulation; however, precurved sheaths designed for larger hearts are often difficult to utilize in small patients.

The most common serious complications from pediatric radiofrequency ablation reported to the above mentioned registry are complete heart block, thrombus formation and cardiac perforation.[4,5,6] Death is very rare (0.12 %) and more frequent with small patient size, left sided procedures and greater number of ablation lesions, but was not found to be related to operator experience.[13] Over the course of

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None.

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Figure 1: Important Considerations for Ablation Procedures in Congenital Heart Disease

1. Understand the anatomy of the particular congenital lesion
   a. Atrial situs and conduction system location
   b. Ventricular situs
   c. Relation of the great arteries
2. Understand the operative procedures the patient has had with review of clinical data such as operative reports, catheterization reports, echocardiograms, cardiac MRI and CT scans
   a. Atriotomy location
   b. Ventriculotomy and/or transanular incisions
   c. Septal patches and locations
   d. Intracardiac or extracadiac baffles
      i. Mustard, Senning and Double Switch
      ii. Fontan
         1. Classic
         2. Intracardiac (lateral tunnel)
         3. Extracardiac
3. Understand the venous and arterial access to the heart
   a. Vessel occlusion (may require transhepatic access or alternative ablation catheter entry sites such as RIJ or subclavian vein)
   b. Abnormal locations of venous connections in heterotaxy syndromes may limit the catheter access to the heart

Figure 1: Important Considerations for Abalation Procedures in Congenital Heart Disease.

the registry these complications became rarer with a rate of (4.2%) in the first 4193 patients and (3%) in the later 3407 patients.[6] Complications were found to be more common in younger smaller patients for both eras. Additional complications not described by the registry have also come to light and again are more common in small patients. Coronary artery stenosis is reported after RF ablation on the right posterior septum and the left mitral annulus.[14,15,16] A smaller but prospective and later era registry reported a similar complication rate but no deaths in 1776 patients.[7] Given the higher rate of complications in the smallest patients with radiofrequency ablation we feel cryoablation, which was not available at the time of these registries, should be considered and available for procedures in small patients.

Cryoablation 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 or shrinking of the tissue and can be placed within venous structures such as the coronary sinus with no reported risk of stenosis.[17] 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.[18] Essentially operators can see what the catheter is going to do before it is permanent. If the freeze is terminated at this point the conduction nproperties 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. While there have been reports of late AV block after combination cryoablation and RF ablation for AVNRT and reports of first degree AV block, temporary second degree AV block, and permanent right bundle branch block, there have been no reported cases of permanent inadvertent AV block from cryoablation alone.[19,20,21] A multicenter study of cryoablation in patients less than 5 years old and 15kg confirmed that cryoablation can be performed safely in these patients.[22]

Biophysics Of Catheter Ablation

Understanding the biophysics of catheter ablation can aid in trouble shooting difficult cases. Radiofrequency ablation remains the mainstay of catheter ablation in pediatrics except for right septal and coronary sinus substrates. In patients with normal cardiac anatomy standard 4mm tip catheters are all that is usually required. Large tip RF catheters and irrigated tip catheters are rarely needed in small patients however special circumstances may require their use. Given the possibility of lesion growth[23] in small patients extra caution should be used and trial of other techniques such as different catheter orientation, long sheaths or cryoablation should be utilized first. Cryoablation has been shown to create larger lesions in low blood flow situations such as in the coronary sinus and in the atrial appendages.[24,25,26,27] Accessory pathway substrates and ectopic foci in these locations may be more amenable to cryoablation than standard 4mm tip RF ablation when power delivery is limited.

Infants

Ablation procedures have been performed successfully in small infants but with a higher morbidity and mortality directly related to the procedure. During infancy ablation should be reserved for dire circumstances in which adequate arrhythmia control cannot be obtained with a sufficient trial of multiple and combinations of antiarrhythmic medications. [9,10,11,28]

Radiation Exposure

Ionizing radiation exposure is of particular importance for pediatric patients especially when considering a lifetime of exposure and the possible increased risk of developing malignancy or genetic defect.[5,29,30] Radiation safety is also of great importance for procedural staff and the ablation operator. With these risks in mind, many pediatric centers are now performing ablations with very little and even no fluoroscopy by utilizing 3D mapping systems. Full utilization of 3D mapping systems allows routine ablation of right sided substrates without fluoroscopy.[31,32,33] Left sided substrates require either brief fluoroscopy during transeptal puncture or other imaging modalities such as intracardiac echocardiography (ICE) or trans-esophageal echocardiography (TEE) and can be performed with either no fluoroscopy or very little fluoroscopy used only during the transeptal puncture.[34]

Congenital Heart Disease

Palliated and repaired congenital heart disease is associated with a significant risk of developing arrhythmias. These patients are at risk for both atrial and ventricular arrhythmias and while the treatment options available are the same as for those with morphologically normal hearts there are unique differences that present challenges. The full breadth of congenital heart disease anatomy is beyond the scope of this article however there are arrhythmia associations between various congenital anomalies that can be discussed and general principles of an approach to catheter ablation in congenital heart disease is outlined. Figure 1 gives an outline of important considerations for each patient.

Atrial Arrhythmias

Atrial tachycardia in patients with congenital heart disease is most often found in the morphological right atrium. These arrhythmias are usually reentrant but can rarely be triggered or micro-reentrant with a focal origin.[35] 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 por-

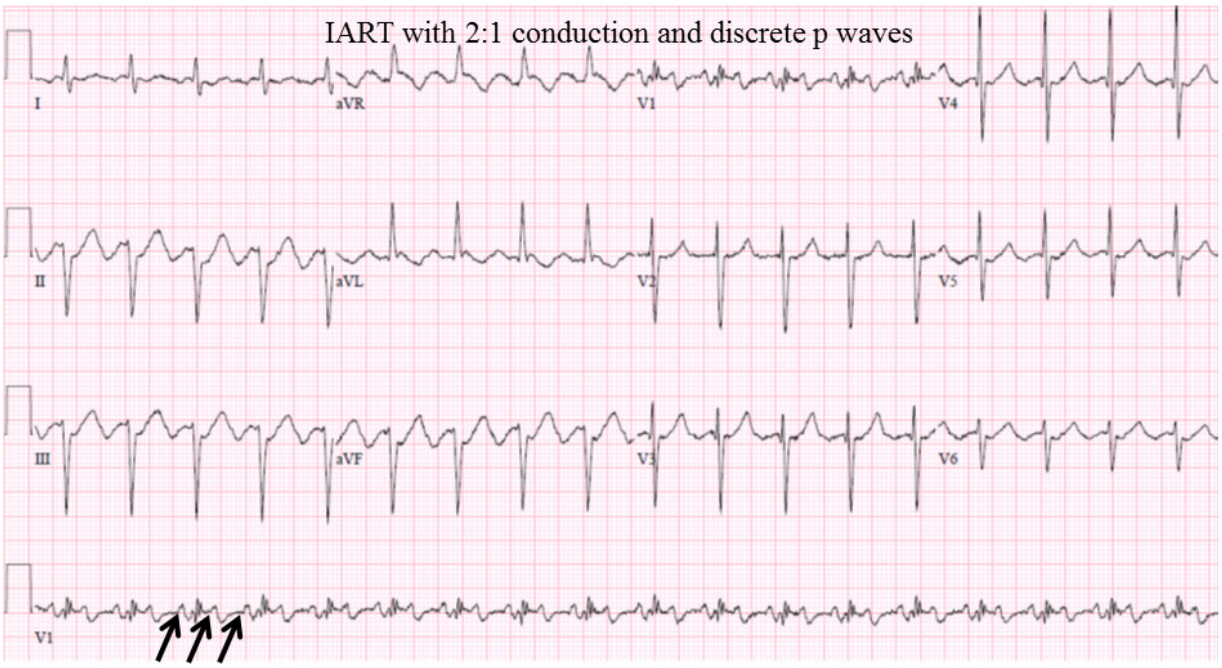

Figure 2: **ECG of a 28y/o with Noonan's syndrome ASD and pulmonary valve stenosis post repair in 2:1 IART. Note the discrete p waves and relatively slow tachycardia p-p interval of 270 msec.**

tion 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.[36] Congenital heart surgery usually involves an atriotomy and may include septal patches or intracardiac baffles with long suture lines that can create other areas of delayed conduction and arrhythmia substrates. Moreover, congenital heart patients often have abnormal hemodynamics or varying degrees of atrial-ventricular valve leak creating a higher pressure atrium that is often dilated, thick and with varying degrees of fibrosis all contributing to arrhythmia mechanism.

IART is usually slower than typical atrial flutter with cycle lengths of 270-450msec and often presents as a 2:1 tachycardia with what appear to be discrete p waves rather than the typical saw-tooth pattern of atrial flutter. [35,36] 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. ECG will often appear to be of an ectopic atrial rhythm or sinus tachycardia when the true underlying rhythm is 2:1 conducted IART with the second p wave buried in the QRS complex or T wave. Figure 2 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 the patients anatomy including surgical scars and suture lines and thorough review of the patient's clinical data and operative reports is essential. Special attention should be paid 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.

While the arrhythmia circuit in these patients can be quite complex, 67% of IART circuits involve the CTI in patients with straight forward and repaired CHD such as Tetralogy of Fallot, ASDs and VSDs.[36,37] In these cases ablation requires similar techniques as typical atrial flutter. Other more 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.[36,37] 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 however the presence of the baffle from Inferior vena cava to the mitral valve creates further complexity often requiring ablation on both sides of the baffle.[36,38] In addition to finding and marking the AV node special care should be made on the right lateral atrial wall to find and mark locations of phrenic nerve stimulation.

Three-dimensional (3D) mapping systems have become an important resource for ablations in congenital heart disease.[37] These systems include both contact and noncontact mapping and allow for better collection, assimilation, and visualization of arrhythmia data. 3D mapping not only facilitates success but simplifies ablation procedures. The newest iterations of contact mapping collect geometry, timing and voltage data simultaneously. Noncontact mapping can facilitate further simplification of the procedure in select patients however the severely dilated atrium of many congenital heart patients is a limiting factor as illustrated in figure 3. Future improvements of noncontact mapping may eliminate this problem with directable sensing electrodes that incorporate contact mapping technology. It is particularly important to make sure the geometry created in the 3D mapping system represents the chamber of interest. Often an angiogram is useful to make sure that all areas of these complex and dilated chambers are included in the geometry. In the most difficult cases a segmented CT or MRI scan can by fused or merged with the geometry collected by the 3-D mapping system.

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 can be evaluated and marked with the mapping system and include locations of low amplitude fractionated electrograms, local timing preceding a discrete p-wave by 50-80 msecs, or areas that demonstrate entrain-

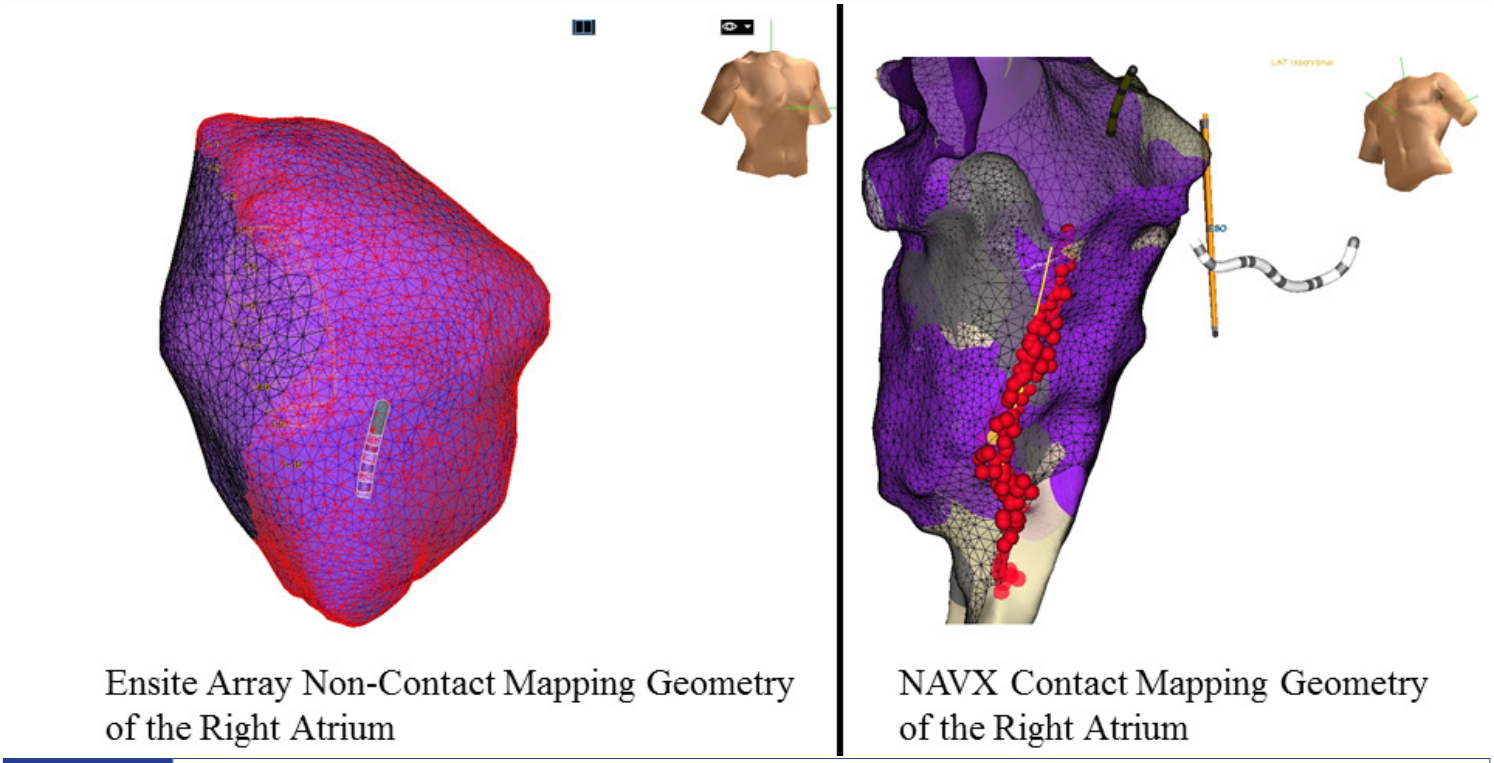

Figure 3: **3D mapping of a severely dilated right atrium in a 28 year old male with Ebstein Anomaly who is post tricuspid valve replacement and right atrial maze. The first panel shows a noncontact mapping image displaying the areas out of the 4cm accuracy range in red. The second panel shows a contact mapping geometry. In this case contact mapping revealed that the low voltage area and scar likely created by a right atrial maze procedure was a target for ablation that was out of range for noncontact mapping. Ablation in this area with a large (8mm) tip RF catheter resulted in arrhythmia termination, non-inducibility and bidirectional conduction block.**

ment (post pacing intervals – tachycardia cycle length <= 30 msec, perfect concealed entrainment, or electrogram to p time = stimulus to p time).[35,39] Without these techniques success can be limited in all but the simplest cases.

Special Anatomic Considerations

AV Node

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 congenital heart patients. Examples of abnormal location include a posterior deviation in patients with AV septal (AV canal) defects, anterior deviation and double AV nodes in congenitally corrected transposition of the great arteries, and positioned in the floor of the blind right atrium in tricuspid atresia.[40] 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. Location can be identified and 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.

Ebstein Anomaly

Ebstein anomaly is a specific 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.[41] 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 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.[8,41] 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.[8,42] 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.[43] However this electrode wire is not currently manufactured and is of limited use in patients with small right coronary diameter. Even with careful mapping successful ablation in these patients may require persistence and multiple test ablations to find a successful site.[8]

Baffles And Fontans

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. In some of these patients components of an arrhythmia circuit may be located on the pulmonary venous or both sides of 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 approach 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 patients anatomy is needed and review of imaging studies such

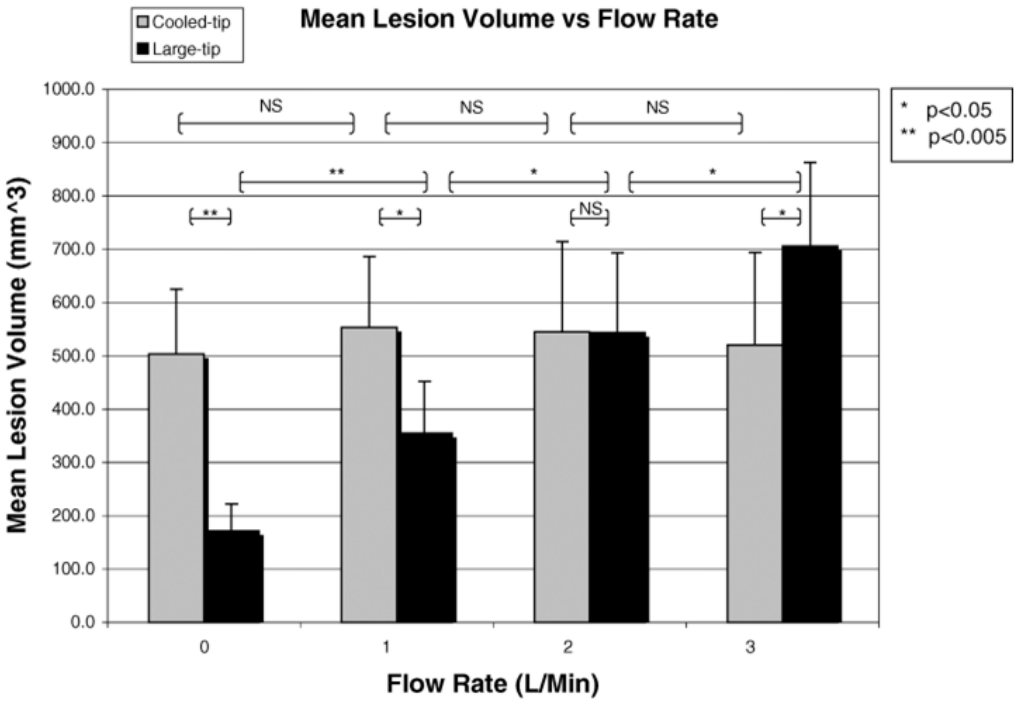

Figure 4: **Relationship between mean lesion volume and flow rate for irrigated and large tip RF catheter types. Highest flow rate (3 L/min) corresponds to a flow velocity of 15.5 cm/s. (Data from Pilcher TA, Sanford AL, Saul JP, Haemmerich D. Convective cooling effect on cooled-tip catheter compared to large-tip catheter radiofrequency ablation. Pacing Clin Electrophysiol. 2006; 29: 1368-1374. With permission.)**

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.[44,45] There are recent case reports of transbaffle puncture for ablation in patients with extracardiac fontan.[46,47]

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.[48,49] 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 knows as the Monckeberg sling.[50] 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.[48,49] 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 and cryoablation may act as an important tool for safe ablation attempts.[8,48,49]

Ablation Biophysics

The atrium of many congenital heart 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 make 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 with in the tissue. Studies show that both large tip and irrigated tip ablation were superior to standard 4mm tip catheters in ablation of IART.[51,52] Irrigated tip catheters are likely to give superior lesion size in areas of low blood flow[53] but 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. Figure 4 shows a comparison of irrigated tip ablation to large tip ablation at various flow rates.

Cryoablation may also be a useful tool for ablation in areas of low blood flow and several studies demonstrate that larger lesions are created in low blood flow situations.[24,25] However, the longer ablation times and inability to drag the cryo-catheter for ablation lines limits it use for substrates that require multiple lesions but focal arrhythmias may be more amenable to cryoablation in areas of low blood flow. Figure 5 shows comparative lesions of large tip cryoablation and large tip RF ablation at various flow rates.

Summary

Unique anatomic variation, small patient size, and unusual substrates for tachyarrhythmia make catheter ablation procedures in pediatrics and congenital heart disease challenging. It is important to understand these challenges and proceed with flexibility in trying alternative approaches and technology to perform safe and effective procedures.

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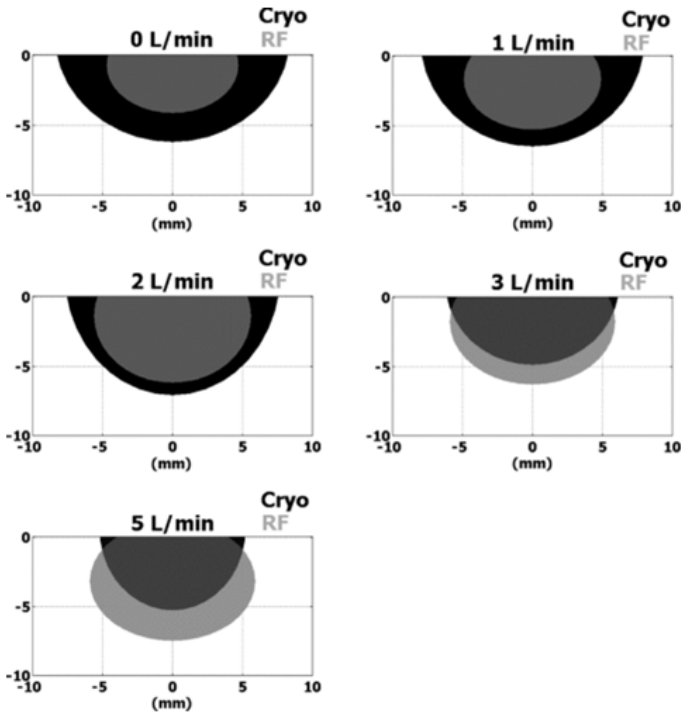

Figure 5: **Ellipsoid graphs comparing the average lesion dimensions for cryoablation and RF ablation at each respective flow rate. The series shows decreasing lesion dimensions with increasing flow rate for cryoablation (P < 0.005) and increasing lesion dimensions for increasing flow rate for RF ablation (P < 0.005). Note the partial prolate ellipsoid shape of cryoablation lesions and the partial oblate ellipsoid shape of the RF ablation lesions. (Data from Pilcher TA, Saul JP, Hlavacek AM, Haemmerich D. Contrasting effects of convective flow on catheter ablation lesion size: Cryo versus radiofrequency energy. Pacing Clin Electrophysiol. 2008; 31: 300-307. With permission.)**

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