CLINICAL REVIEWS

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Pearls for Ablation in Congenital Heart Disease

RONALD J. KANTER, M.D.

From the Duke University School of Medicine, Department of Pediatrics, Division of Cardiology, North Carolina, USA

Ablation in Congenital Heart Disease. **Tachyarrhythmias occur in patients having congenital heart disease most commonly related to surgically created or naturally occurring conduction obstacles, and to postoperative hemodynamic effects on cardiac muscle. Less frequently, the underlying disease is associated with tachycardia substrates. Thorough knowledge of the patient's congenital anatomy and surgical procedures is required before considering catheter ablation. In many instances, procedural considerations should include meticulous hemodynamic surveillance, analogous to patients having structurally normal heart but cardiomyopathy. This includes careful selection of sedating and anesthetic agents. Congenital heart defects that have a higher than expected incidence of naturally occurring tachyarrhythmia substrates include Ebstein anomaly of the tricuspid valve, congenitally corrected transposition, and some of the heterotaxies. Intraatrial reentry tachycardia and atrial flutter are especially prevalent following the Mustard or Senning operations for d-transposition of the great arteries and the earlier Fontan type operations. Although these tachyarrhythmias are not as frequent following atrial septal defect repair, the high incidence of this defect also makes these patients germaine to this discussion. Focal atrial tachyacrdia and atrioventricular nodal reentry tachycardia also occur in these patient groups. Macroreentry ventricular tachycardia occurs most frequently following right ventricular outflow tract surgery, especially following repair of tetralogy of Fallot. This article focuses on the technical aspects of catheter ablation of these arrhythmias, due to the challenges presented by the underlying anatomy compared with patients having normal hearts.**
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catheter ablation, cardiomyopathy, congenital heart disease, Ebstein's anomaly, supraventricular tachycardia

Children and adults with congenital heart defects may have tachycardia substrates that are naturally part of the underlying heart disease, and, hence, may present prior to surgery; or, their tachycardia substrates may evolve postoperatively, related to surgical scar, artificial material, and secondary hemodynamic changes. Because the idea that "one size fits all" has no place when considering catheter ablation in congenital heart disease, this presentation will attempt to highlight a seemingly unrelated series of topics, all intended to prepare the reader for the wide range of structural–electrical abnormalities that may confront them. That said there are fundamental principles when approaching these patients, which, if ignored, will be to the patient's detriment. These too will be discussed. This paper is not intended to provide a sweeping summary of congenital heart defects and their palliative/corrective operations. For that, the reader is encouraged to consult standard textbooks.[1-3]

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Address for correspondence: Ronald J. Kanter, M.D., Duke University Medical Center, Box 3090, Durham, NC 27710. Fax: 919-668-6122; E-mail: kante001@mc.duke.edu

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Before the Patient Enters the Electrophysiology Laboratory

Many patients undergo electrophysiologic testing due to known congenital heart disease and a worrisome clinical event such as severe palpitations, sudden unexplained shortness of breath, or syncope. If a pathological tachycardia has not been documented, knowledge of the structural defect should provide the electrophysiologist a "head start" with respect to the most likely tachycardia mechanism. These are outlined in Table 1. However, this information represents a crude compass, and the operator should investigate the patient's heart disease more fully prior to the study. Less a "pearl" and more an inviolate principle, the electrophysiologist must have a firm understanding of the patient's congenital and operative anatomy. This includes structural nuances that vary according to congenital anatomy and surgical approach. Table 2 summarizes a number of these. This information should be accrued from all prior surgical reports, hemodynamic catheterization reports, angiography, and other imaging studies (echocardiography, cardiac computed tomography [CT], and cardiac magnetic resonance imaging [MRI]).

During the Study

Analogous to patients having dilated cardiomyopathy and other low cardiac output states, some patients having congenital heart disease will experience cardiovascular

TABLE 1

Tachyarrhythmias Most Commonly Seen with Specific Congenital Heart Defects

Defect	Pre- versus Postoperative (Type of Surgery)	Tachyarrhythmias (Less Common)
Ebstein anomaly	Both (tricuspid valve repair)	AVRT; AVNRT; (IART/AFl; VT)
l-TGA	Both (variable)	AVRT; (VT)
Tricuspid atresia	Pre-	(AVRT)
	Post (Fontan)	IART/AFl; (AVRT; FAT; AF)
HLHS	Pre-	AET
	Post-(Fontan)	IART/AFl; (AET; FAT; AF)
Other single ventricle	Post-(Fontan)	IART/AFl; (FAT; AF)
AVSD	Post-(repair)	IART/AFl; (AVRT; AVNRT)
Heterotaxy	Both	AVRT: Twin AV node- or AP-related
	Post-(Fontan)	IART/AFl; (FAT; AF)
ASD	Post-(repair)	IART/AFl; (FAT)
d-TGA	Post-(Mustard or Senning)	IART/AFl; (AVNRT; FAT)
Tetralogy of Fallot	Post-(repair)	VT; (IART/AFl)

AET = atrial ectopic tachycardia; AF = atrial fibrillation; AFl = atrial flutter; AP = accessory pathway; ASD = atrial septal defect; AVNRT = atrioventricular nodal reentry tachycardia; AVRT = atrioventricular reciprocating tachycardia; AVSD = atrioventricular septal defect; d-TGA = d-transposition of the great arteries; FAT = focal atrial tachycardia; HLHS = hypoplastic left heart syndrome; IART = intraatrial reentry tachycardia; l-TGA = l-transposition of the great arteries; VT = ventricular tachycardia.

decompensation during protracted studies, possibly related to excessive unloading from anesthetic agents, long periods of pathological tachycardia (or rapid pacing used to induce tachycardia!), cumulative effect of intravenous crystalloid and contrast material, or unappreciated valve regurgitation from prolonged catheter positioning. Preelectrophysiologic testing hemodynamic measurements including filling pressures and mixed systemic venous saturation, continuous urine collection, and postelectrophysiologic hemodynamic measurements have become a standard part of our procedural strategy. Postprocedure recovery in an intensive care unit should be anticipated prior to the procedure. Congenital heart patients having reduced systemic ventricular systolic function and all patients having Fontan physiology are particularly preload dependent. Those having associated pulmonary hypertension are among the highest risk patients, and the risk-benefit considerations should be carefully discussed with the patient, their family, and the anesthesiologist before proceeding.

Amongst the general population of patients having congenital heart disease, we have found that general anesthesia, when administered by cardiac anesthesiologists well-versed in congenital heart disease, represents the safest venue for prolonged electrophysiologic testing and ablation procedures. These professionals may afford smooth transition between anesthetic agents when indicated by changes in the hemodynamic or electrophysiologic milieu. For example, the Ebstein patient with severe tricuspid regurgitation who develops progressive hypotension during prolonged periods of pacing and tachycardia may benefit from a transition from propofol- to ketamine-based anesthesia, allowing the procedure to continue.

Planning for and executing catheter access to the heart and to the chamber(s) of interest represent a vital pre- and intraprocedure task in congenital heart disease patients. Table 3 includes those conditions that are expected to pose vascular and/or cardiac chamber access challenges and methodologies that have been reported to allow catheter access. We do not include standard transseptal puncture to access the left atrium in this category, as this is now considered a standard procedure in most ablationists' armamentarium. Flexibility in planning extends beyond the mapping/ablation catheter in these patients. There is often limited opportunity to place the standard number of diagnostic electrode catheters; creative approaches to pacing and recording may include the esophagus, hepatic veins, and retrograde to the atria from the ventricles.

The modern ablationist has become comfortable with adjunctive real-time imaging equipment during ablation procedures for atrial fibrillation, especially intracardiac echocardiography (ICE). For the congenital heart disease patient population, landmark identification using ICE may be difficult due to percutaneous vascular limitations. In those cases, transesophageal echocardiography (TEE) may be equally useful. We also rely upon angiography in many instances: to help characterize venous structures (such as interrupted inferior vena cava in patients with left isomerism form of heterotaxy, occluded superior vena cava baffle following Mustard or Senning operations, and iliofemoral venous occlusion), to measure right atrial size in some Fontan patients, and to identify the plane of the atrioventricular (AV) valves (by ventriculography with specific angulation for complex heterotaxy and single ventricular patients and by coronary arteriography in Ebstein patients).

Electroanatomic mapping systems are now considered requisite for mapping and catheter ablation in this patient population. In multivariate analysis, Triedman *et al*. showed that failure to use such a system approached significance in identifying patients having an "unfavorable arrhythmia score" at follow-up from ablation for intraatrial reentry tachycardia following congenital heart surgery.[4] The two most

TABLE 2

Anatomical Details of Interest to the Ablationist

Defect/Pre- versus Postoperative	Observation	Obtained by	Importance to Ablationist
ToF and DORV/post-	Specific repair	Operative report	Potential corridors of slow conduction
d-TGA/post-Mustard	Relationship of inferior baffle line to CS ostium	Operative report	Biases approach to CTI
ASD (2° and venosus)/post-	Atriotomy line	Operative report	Potential corridors of slow conduction
Single ventricle/post-"lateral tunnel"	Relationship of anterior suture line to CT	Operative report	Potential corridors of slow conduction
Heterotaxy/pre-	Ventricular looping; AV valve anatomy	Imaging	Potential for twin AV node-related AVRT

ASD = atrial septal defect; AV = atrioventricular; AVNRT = atrioventricular reciprocating tachycardia; CS = coronary sinus; CT = crista terminalis; CTI = cavotricupsid isthmus; DORV = double outlet right ventricle; ToF = tetralogy of Fallot.

TABLE 3
Conditions Associated with Challenges for Vascular and Cardiac Chamber Access

Occlusion or Access Challenge	Associated Conditions	Alternate Strategies
Iliofemoral venous occlusion	• d-TGA undergoing BAS as newborn prior to 1985 • Any complex patient with history of multiple catheterizations	• Internal jugular, subclavian veins • Transhepatic venous (especially for transseptal access)
Interrupted IVC (above renal veins)	• Heterotaxy (left atrial isomerism)	• Internal jugular, subclavian veins • Transhepatic venous (especially for transseptal access) • Femoral vein to azygos vein
Access to pulmonary venous atrium	• "Lateral tunnel," TCPC types of Fontans	• Transbaffle puncture (consult interventionalist?) • Via baffle fenestration, if present • Hybrid procedure (CT surgeon performs limited thoracotomy, atrial pursestring)
Access to supra-annular rim of systemic venous atrium	• Fontan patients with supra-annular patch (especially in double inlet ventricle)	• Retrograde from aorta
Access to pulmonary venous atrium	• D-TGA after Mustard or Senning	• Transbaffle puncture (consult interventionalist?) • Retrograde from aorta
Access to atrial mass	• Fontan patients with extracardiac conduit	• Transthoracic puncture (consult CT surgeon) • Hybrid procedure (CT surgeon performs limited thoracotomy, atrial pursestring) • Retrograde from aorta

BAS = balloon atrial septostomy; CT = cardiothoracic; d-TGA = d-transposition of the great arteries; IVC = inferior vena cava; TCPC = total cavopulmonary connection.

widely used systems, Carto (Biosense Webster/Johnson & Johnson, USA) and NavX (Endocardial Solutions/St. Jude Medical, USA) each permit point-to-point acquisition of endocardial electrograms coupled to anatomic reconstructions. The anatomy can then be interpolated into a DICOM-stored CT- or MRI-generated chamber of interest. While widely used for the left atrium in persons having atrial fibrillation and normal anatomy, this technology is just now being applied to the congenitally malformed and reconstructed heart. Endocardial Solution's Enguide system generates an endocardial surface voltage map from unipolar electrograms that are recorded from an intracavitary array mounted on a balloon catheter. This system has the advantage of recording from a single beat of tachycardia, but the disadvantages of limited accuracy beyond a 4 cm radius, contamination by farfield electrograms, and inability to "see around geometric corners." This system will be discussed in more detail below.

A String of Pearls

Ebstein Anomaly of the Tricuspid Valve

Ebstein anomaly of the tricuspid valve is associated with accessory AV connections (and, rarely, atriofascicular fibers) in approximately 20% of cases. These are nearly all right-sided, with the majority located in the inferior half of the tricuspid valve annulus, especially right posteroseptal and right posterolateral.[5] Among patients with manifest preexcitation, estimating the accessory pathway location from the standard electrocardiogram (ECG) is fraught with difficulty. First, about one-half of affected patients have multiple pathways.[6] Second, the malformed inlet portion of the right ventricle (between the normally located tricuspid valve annulus and the coaptation plane of the leaflets) often has slow conduction properties, meaning that the hallmark ventricular fusion during sinus rhythm in patients with manifest preexcitation, tips in favor of the AV node. Hence, preexcitation may be unexpectedly minimal.[7] Pre-procedure commitment to pathway location is not a good idea!

At the start of the study, fluoroscopic identification of the tricuspid valve annulus should be recorded in about 20° right anterior oblique projection. Although the fat pad represents this structure well, selective right coronary arteriography is preferable and may be a useful "roadmap." Mapping the accessory pathway(s) insertions is performed by standard methods, but complex and fractionated electrograms from the ventricular side of the annulus, and unrelated to the accessory pathway *per se,* may complicate the process. Authors have touted the placement of a 2 Fr multipole electrode catheter into the right coronary artery for direct electrogram identification to ease this task,[5] even applying color-coded activation mapping techniques.[8] However, in cases where the local electrograms are very fractionated, we find that there is no substitute for such fundamental techniques as placement of a ventricular electrogram-synchronous atrial extrastimulus during orthodromic AVRT[5] to sort out the chamber–electrogram associations.

Because the right atrium may be enormous and the tricuspid annulus very dilated, access to the annulus by the mapping/ablation catheter usually requires a specialized long sheath. We have had success with the site-specific Swartz sheaths (SR series, St. Jude Medical) and with the deflectable Agilis sheath (St. Jude Medical). Catheter stability can be difficult in this very dynamic portion of the heart, and it may be monitored by either ICE or TEE. Further complicating successful accessory pathway ablation using radiofrequency energy in this portion of the heart is the occasional difficulty in achieving adequate catheter tip temperatures. The use of cryothermal energy has been shown to ameliorate both issues by virtue of "cryoadherence." Along the tricuspid annulus, it has been shown that successful long-term ablation can be expected if the pathway disappears within 9 seconds of cryoadherence.[9]

Congenitally Corrected Transposition of the Great Arteries (levo- or l-TGA)

In this disease, there is AV and ventriculoarterial discordance, resulting in a subaortic right ventricle, complete with

outflow chamber. Approximately 2–5% of patients having l-TGA have one or more accessory AV connections. Although these pathways have been long thought to exist with almost equal prevalence along both AV valves, in our experience, they are mostly associated with the left-sided tricuspid valve. (The morphologic AV valve remains with the morphologic ventricle that it enters.) These pathways may truly be located anywhere along the tricuspid valve annulus, as there is no tricuspid valve-aortic fibrous continuity. Left anteroseptal and midseptal pathways should be anticipated. Fortunately, in the typical forms of this disease, the specialized conduction system is relatively remote from the tricuspid valve.

Just as in patients with normal hearts and left-sided accessory pathways, tricuspid annulus-related accessory pathways may be mapped and ablated from either transseptal or retroaortic approaches. However, negotiating the tricuspid annulus from the aorta requires that the catheter traverse the right ventricular outflow tract (RVOT) and wind through the chordae tendinae of the tricuspid tensor apparatus. Unlike the mitral valve, there are chordal attachments to the ventricular septum, and there is greater risk of chordal damage or entanglement in l-TGA patients. We favor the transseptal approach; however, there is even a caveat for this procedure: The fossa ovalis tends to be located more posteriorly within the atrial septum in these patients, having reduced distance between its posterior rim and the back of the heart. ICE or TEE guidance may be helpful.

When the tachycardia substrate is related to the mitral valve, either AV nodal reentry or AV reciprocating tachycardia, knowledge of the anatomy of the specialized conduction system is of greater importance. Except in the presence of pulmonary valve atresia or severe hypoplasia (in which case the AV node is normally located at the apex of the triangle of Koch),[10] the AV node is displaced anteriorly at the right atrial–mitral annulus junction due to atrial and ventricular septal malalignment. Rare right-sided accessory pathways should be considered in that light. Ablation targets for AV nodal reentry have been reported to be in the usual posteroseptal locations, related to the ostium of the coronary sinus. L-TGA is associated with situs inversus with a much greater than expected frequency. The AV node in those patients tends to be located in a normal but mirror-image location. This may be due to the very high association with pulmonary valve atresia or hypoplasia.

Heterotaxy and Twin-AV Node-Related Tachycardias

The combination of ventricular l-looping (as described above in classic l-TGA) and AV septal defect may be associated with both an anterior AV node at the anterior mitral annulus–atrial junction and a posterior AV node, just anterior to the putative location of the coronary sinus ostium. Although this combination of structural defects is not specific, it is most commonly seen in heterotaxy patients. As a caveat to the electrophysiologist, most congenital heart disease experts (who concentrate on the functional hemodynamic abnormalities in a given patient) will emphasize the *other* structural abnormalities in these patients in their descriptions, namely, single (usually right) ventricle (hence, with "unbalanced AV septal defect"), double outlet of that right ventricle, and pulmonary atresia or severe stenosis. The nuance of ventricular looping must often be sought out.

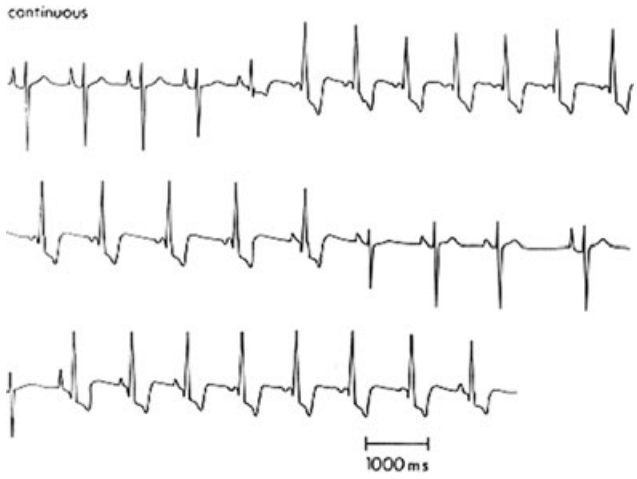

Figure 1. *Continuous rhythm strip from a 16-year-old male with heterotaxy (including unbalanced AV septal defect with dominant right ventricle, ventricular l-looping, double outlet right ventricle, and pulmonary atresia) who had only undergone aortopulmonary shunt (modified Blalock-Taussig shunt). Illustrated is changing P wave morphologies, accompanied by changing PR intervals and QRS morphologies. This patient was demonstrated to have twin AV nodes and His bundles, 2 bundle branch systems, and AV reciprocating tachycardia utilizing the posterior conduction system as the ventriculoatrial limb and the anterior system as the anterograde limb.*

Twin-AV nodes and their associated His bundles may each give rise to a separate bundle branch system. In such cases, sudden changes in the QRS may occur when the atrial pacemaker changes—another phenomenon highly associated with the heterotaxies (Fig. 1). This observation during routine rhythm monitoring in a patient also having pathological tachycardia should make the operator suspect AVRT using twin-AV nodes.

Initial diagnostic electrophysiologic testing should focus on functional anterograde and retrograde conduction properties in the two AV conduction systems. His bundle electrograms should be mapped by scanning the single large AV valve, aided by prior ventriculograms and coronary arteriograms and by real-time ultrasound. The posterior AV node/His bundle may be mapped by scanning the most proximate portions of the inferior AV valve from the vantage of the inferior vena cava (or, if congenitally interrupted, from the vantage of the hepatic vein when using a transhepatic approach). The anterior AV node/His bundle may be mappable just superior to the superior-most portion of the AV valve. In cases where there is residual anterior atrial septum, it may be within that structure. This often requires a long sheath, if approached from an inferior approach. Once identified, the structures should be "tagged". In combination with initial functional testing, differential pacing from adjacent atrial sites, atrial mapping during tachycardia, and His-synchronous (i.e., anterograde His) placement of premature ventricular beats during tachycardia permit identification of the AV conduction system having the most robust anterograde conduction properties. The anterogradedly "weaker" AV node/His bundle should be targeted for ablation. This is usually the system which comprises the retrograde limb of AVRT.

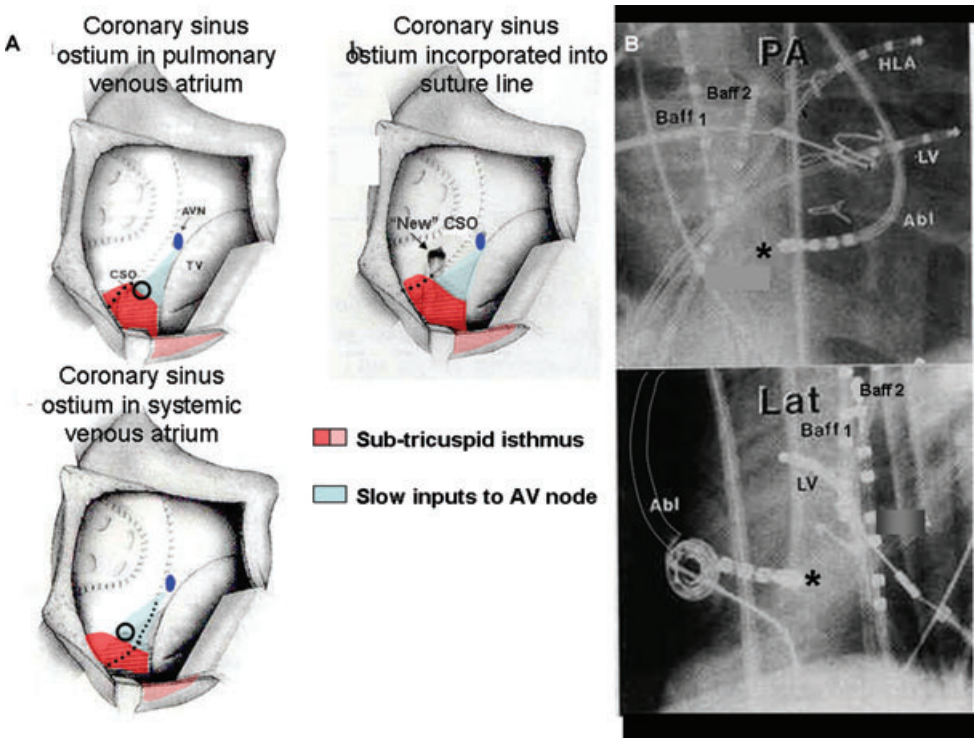

Figure 2. *(A) Diagrams of the pulmonary venous atrium following the Mustard operation for d-transposition of the great arteries, illustrating the location of the compact AV node, the slow inputs to the AV node, and the subtricuspid isthmus in relation to the inferior baffle suture line depending upon the surgical handling if the coronary sinus ostium. As can be seen at the bottom, when the suture line is placed anterior to the ostium, so that coronary sinus return remains in the systemic venous atrium, very little of the subtricuspid isthmus remains on the pulmonary venous side. This is also very destructive to the slow inputs to the AV node. CSO = coronary sinus ostium; TV = tricuspid valve. (B) Posteroanterior (PA) and lateral (Lat) radiographs from a patient having undergone Mustard operation, showing diagnostic catheter positions and position of the mapping/ablation catheter (Abl) during ablation of the slow AV nodal inputs for typical variety of AV nodal reentry tachycardia. The catheter tip (∗) is retrogradedly placed just across the tricuspid valve. Baff 1&2 = multipole electrode catheters within the systemic venous baffle; HLA = high lateral morphologically but left atrium (systemic venous); LV = morphologically (but subpulmonic) left ventricle. (Adapted from: Kanter RJ, et al, Radiofrequency catheter ablation of supraventricular tachycardia substrates following Mustard and Senning operations for d-transposition of the great arteries. J Am Coll Cardiol 2000;35:428-441, American College of Cardiology, with permission).*

Supraventricular Tachycardias Following Mustard and Senning Operations

These operations were performed from the early 1960s until about 1985 as the major long-term surgical palliations for young children having d-transposition of the great arteries. Hence, there is a population of persons in their late 20s to early 50s who have undergone these operations and who are at great risk of having supraventricular tachycardia, especially intratrial reentry tachycardia (IART) and cavotricuspid isthmus (CTI)-dependent atrial flutter.

These operations have in common the performance of an atrial septectomy and longitudinal bisection of the subtricuspid isthmus by a baffle of artificial material or pericardium (Mustard) or right atrial free wall (Senning), for the purpose of redirecting venous returns to the opposite AV valves. In the case of the Mustard operation, surgical strategies varied as regards the precise location of the baffle suture line within the CTI. Inclusion of the coronary sinus into the systemic venous side (posterior to the suture line) relegates relatively less of the mid- and medial-CTI between the suture line and the tricuspid valve annulus than if the coronary sinus ostium is incorporated into the suture line or remains anterior to the suture line (Fig. 2). Especially in the former instance, there is substantial damage to the posterior inputs to the AV node.[11] The original surgical procedure note should be carefully read for this important detail. Also, the portion of the suture line along the anterior atrial septal remnant (and superior to CTI) may have damaged the fast inputs to the AV node. The surgical note will not be helpful for this detail, but first degree AV block and complete absence of ventriculoatrial conduction may be clues to that circumstance. If present, catheter ablation of the slow inputs may result in AV block.[12]

Because these patients routinely underwent balloon atrial septostomy via femoral venous cannulation by very large sheaths as newborns (initially, 8 Fr!) and because they often underwent multiple cardiac catheterizations, this patient group is especially likely to have unilateral and even bilateral iliofemoral vein occlusion. The conduct of electrophysiological testing in these patients requires a minimum of an atrial, a ventricular, and a map/ablation electrode catheters and femoral arterial access. For the reason provided above, we believe that demonstration of ventriculoatrial conduction, even if this requires isoproterenol, is reassuring that lesion placement in the medial and mid-CTI will not result in AV block.[12] A His bundle electrogram can only rarely be identified along the anterior suture line on the systemic venous side; more often, it can only be observed from the pulmonary venous side (see below).

Most atrial tachycardias in this patient group are CTI-dependent atrial flutter, but IART with critical zones of slow conduction between a suture line and the superior vena caval

orifice, mitral valve annulus, and pulmonary vein orifice have all been described. Focal atrial tachycardias (FAT) adjacent to suture lines are also not uncommon. We have been impressed by the number of IART circuits whose critical zones have been between the original right atrial incision (in the new pulmonary venous atrium) and the tricuspid valve annulus. That incision may have been longitudinal or transverse, and knowledge of that detail may help direct mapping in a very difficult location. Critically, CTI-dependent atrial flutter often requires placement of lesions on both sides of the baffle suture line. This suture line does not necessarily represent an obstacle to conduction. Lesions placed in the systemic venous side can only be relegated to the medial and mid-CTI (if safe), because the baffle usually hugs the inferior vena cava orifice anteriorly, whereas lesions in the pulmonary venous side will naturally be more lateral, as explained below. Even using electroanatomic mapping systems, complete delineation of the tachycardia circuit is not necessary or practical in many of these patients, although reconstruction of multiple segmental maps now makes that possible. We prefer point-to-point mapping in these patients, as their tachycardias tend to be very stable. Although the Enguide array (ESI) has been successfully deployed in these patients, we have found it especially unwieldy in the systemic venous baffles. Principles of identification of atrial diastole (with adenosine or ventricular pacing), concealed entrainment mapping, and continuous diastolic potentials are useful in this patient group.

Access to the tricuspid valve annulus and pulmonary venous atrium is necessary for mapping and ablating the substrates of many IARTs and CTI-dependent atrial flutters and for most AV nodal reentry tachycardias. This may be achieved by needle perforation of the interatrial baffle from a systemic venous atrial approach or by retro-aortic approach. For the former, the needle must be aimed anteriorly and slightly rightward and using ultrasound guidance. Access to the posterior pulmonary venous atrium is difficult with this approach. We prefer the retroaortic approach, wherein a bidirectional deflectable electrode catheter is advanced across the aortic valve into the right ventricle. Tip flexion naturally assumes an anterior attitude. Clockwise torque and marked flexion rotates the distal portion to the patient's right, where the catheter may be partially opened. Two motions may then be employed: (1) Further clockwise rotation (and some withdrawal) will allow the tip to engage the septal tricuspid valve annulus, where the His electrogram can be identified. Additional tip extension will engage the slow pathway portion of the juxta-annular atrial rim (Fig. 2), or (2) The catheter may be advanced across the tricuspid valve and into the pulmonary venous atrium. Further advancement will negotiate the pulmonary vein ostial region, whereas opening the curve, providing slight counterclockwise rotation, and withdrawal, sequentially, will allow mapping of the posterior and then lateral native right atrial wall, lateral CTI, and posterolateral-to-posterior tricuspid valve annulus.

Atrial Tachycardias Following the Fontan Operations

These operations are designed to direct systemic venous return to the pulmonary arteries without an interposed ventricle (with the exception of the Bjork modification). The right atrium or a portion of it is included in that circuit in all of the early styles of Fontan operation. That portion of the atrial mass is necessarily at elevated pressure, resulting in hypertrophy. The combination of hypertrophy, fibrosis, suture lines, and natural conduction barriers (venous orifices, valve annulus, and crista termninalis) results in a highly proarrhythmic milieu.

The atriopulmonary connection is the most common type of Fontan operation in adults. IART is the predominant tachycardia in these patients, although FAT and AV nodal reentry may occur. Regions of interest to the ablationist include the atriotomy site (usually low anterolateral atrium) and the CTI,[13] and, to a slightly lesser extent, the caval orifices, the crista terminalis, and the proximal anterior right atrial appendage (the edge of the roof of the atriopulmonary conduit). In the presence of tricuspid atresia and a well-formed right ventricle, a conduit from the anterior right atrium to the right ventricle (Bjork modification) represents another conduction barrier. When the floor of that conduit is a flap of native right atrial tissue, natural establishment of AV conduction across the suture line may result in functionally important "acquired" Wolff-Parkinson-White syndrome.[14] This connection may be extensive, but may nevertheless be successfully ablated. When the diagnosis is "double inlet ventricle," a patch is surgically sutured above the right atrium-associated AV valve annulus. This results in a rim of juxta-annular tissue, which may only be accessed from the aortic/ventricular side. In some instances, the leaflets had been sutured together, eliminating any access to that tissue.

Despite the above landmarks, critical corridors of slow conduction may exist anywhere in the right atrial mass, and multiple circuits are the rule (Fig. 3). Electroanatomic mapping is mandatory, and electrograms having amplitudes of 0.05 mV or less have been shown to be critical to IART circuits.[15] Concealed entrainment mapping is highly nonspecific for most circuits in these patients,[16] and we rely heavily on identification of atrial diastole and the Ensite array (ESI) for initial identification of the critical slow conduction corridor. The isovoltage map derived by ESI's system is unreliable beyond 4 cm radius. Some Fontan right atria have diameters exceeding 10 cm, meaning that, in those cases, the array must be repositioned to the portion of the atrium of greatest interest based upon supplementary information. Point-to-point mapping is then employed to help define the anatomic extent of slow conduction, often permitting identification of the entrance and exit of that region. Successful lesion generation requires radiofrequency energy through an irrigated tip catheter in most instances. This is due to both extraordinary wall thicknesses, sometimes exceeding 1 cm, and diminished dissipation of heat due to often very sluggish blood flow. Successful circuit ablation is not always associated with IART termination, especially when there are figure-of-eight circuits. We generally rely upon subtle shift in electrograms from an *in situ* multipole electrode catheter for just this reason. The ESI array is very useful to permit rapid identification of new or shifting circuits.

Particular challenges exist in ablation therapy of atrial tachycardia substrates in Fontan patients in whom part or all of the atrial mass is unavailable to venous (and even arterial) access. The "lateral tunnel," "total cavopulmonary connection," and extracardiac conduit styles of Fontan operation fall into this category. For these, collaboration with the cardiothoracic surgeon is necessary. After identification of the ipsilateral internal mammary artery, direct transthoracic puncture and placement of a deflectable sheath into the

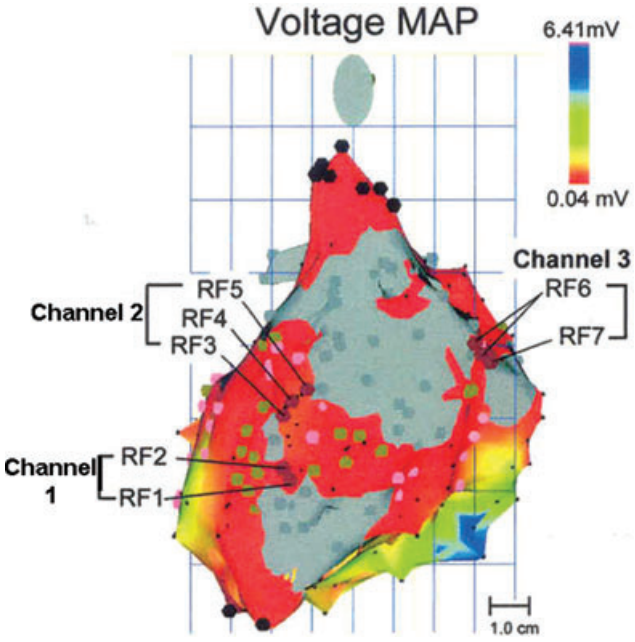

Figure 3. *Right posterolateral projection of an electroanatomic voltage map using Carto of the right atrium from a patient having undergone an atriopulmonary style of Fontan operation for pulmonary atresia. Illustrated are narrow conduction corridors between areas of scar (gray areas) that were critical to intratrial reentry tachycardia. Bipolar voltages in these regions were as low as 0.04 mV. Sites of radiofrequency energy delivery are denoted as RF. Red areas have lowest voltages and blue, highest. (Nakagawa, et al. Characterization of reentrant circuit in macroreentrant right atrial tachycardia after surgical repair of congenital heart disease: isolated channels between scars allow "focal" ablation. Circulation 2001:103:699–709, American Heart Association, with permission).*

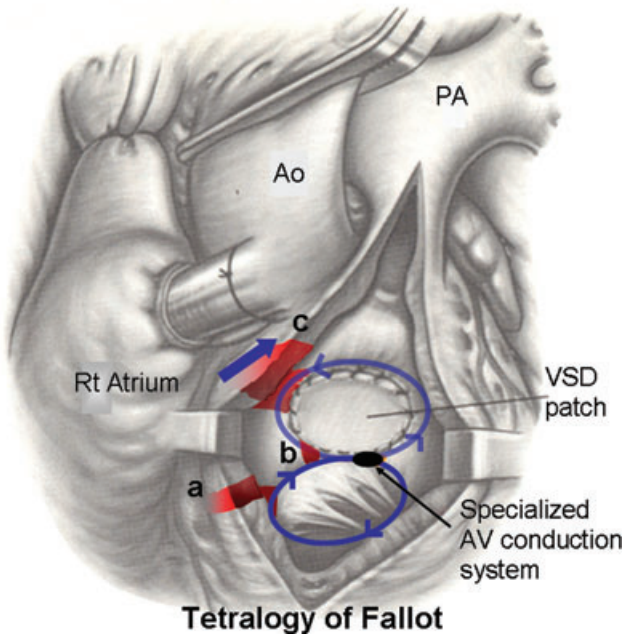

Figure 4. *Intraoperative view of the right ventricle from a patient having an older style repair of tetralogy of Fallot. Illustrated is the ventricular septum with ventricular septal defect (VSD) patch, visualized through an extensive transannular incision. Blue arrows indicate locations of potential late postoperative macroreentry ventricular tachycardia circuits. (The direction of arrowheads is arbitrary.) The brown shaded areas represent regions of possible slow conduction, amenable to catheter ablation. These are between the tricuspid valve annulus and the right ventricular outflow tract (RVOT) incision (A); between the tricuspid valve annulus and VSD patch (B); and between the VSD patch and the RVOT incision (C). Ao = aortic root; PA = main pulmonary artery. (Adapted from: Stark, deLeval. Surgery for Congenital Heart Defect, page 420, WB Saunders, 1994, with permission).*

atrium has been reported with successful outcomes. Risk of postprocedure tamponade is substantial,[17] mandating close hemodynamic monitoring and rapid surgical availability. A more controlled procedure, in which the surgeon enters the chest and helps with sheath entry into the atrium through a pursestring incision, has also been reported.[18]

Many of the above comments may be applied to the ablation of IART and atrial flutter in patients having undergone more simple congenital heart surgery, especially atrial septal defect (ASD) closure. In those patients, the CTI and the low anterolateral right atrium between the atriotomy scar and/or the crista terminalis constitute the vast majority of ablation targets.[19] The ASD patch, itself, is rarely a critical conduction obstacle. More so than the Fontan population (and using standard mapping techniques and energy sources), the great majority of these patients may be effectively and permanently treated.

Ventricular Tachycardia Following Repair of Tetralogy of Fallot

In this patient group, monomorphic ventricular tachycardia(s) (VT) using a macroreentry circuit(s) has been successfully and permanently ablated concomitant with elective pulmonary valve replacement in the operating room for decades. Energy sources have included cryothermal, microwave, and argon beam. The modern decision to attempt catheter ablation without placement of an implantable cardioverter defibrillator should only be considered if their clinical VT was hemodynamically well-tolerated and if their overall hemodynamics are reasonable. We routinely repeat programmed ventricular stimulation at an arbitrary 1–2 months following apparently successful VT ablation. Although not germaine to this topic, it is appropriate to note that inducible polymorphic ventricular tachycardia in this patient group is clinically important, is highly associated with future severe and even life-threatening ventricular arrhythmias, and has not been considered approachable with ablation strategies.[20]

As has been the case with all of the congenital heart operations heretofore mentioned, pre-procedure review of the specific surgical procedure is necessary. While it is true that there is always a patch placed over the ventricular septal defect (VSD), the precise handling of the RVOT muscle bundles and the valvular pulmonic stenosis is variable. Most commonly (about 75% of cases), these patients underwent placement of a transannular patch from the distal parietal RVOT onto the proximal anterior main pulmonary artery. In this instance, the VT circuits could theoretically encircle the VSD patch, encircle the tricuspid valve annulus, and/or could course between the RVOT and the superior tricuspid valve annulus (Fig. 4) in the region of the ventriculoinfundibular fold. Accordingly, and guided by electroanatomic mapping techniques, diastolic potentials and regions where concealed entrainment mapping with short postpacing intervals are demonstrated can be successfully targeted by radiofrequency energy in the following regions: between the right

inferior portion of the RVOT patch and the anterior tricuspid valve annulus; between the superior septal pulmonary valve annulus and the VSD patch; or between the anteroseptal tricuspid valve annulus and the rightward portion of the VSD patch (Fig. 4).[21] The normal specialized conduction system is related to the latter region and must be carefully avoided. Among patients not requiring a transannular patch (because their pulmonary valve annulus is of adequate size), their obstructive RVOT muscle bundles may be excised through an incision through the RVOT. This region may be then become an additional conduction obstacle and substrate for VT.

Radiofrequency energy is effective for these forms of VT but often requires high energy output and 8 mm catheters or irrigated tip catheters. Even in adult patients whose right ventricular pressure is normal following tetralogy of Fallot repair, portions of the right ventricular wall are at least 6 mm in thickness. Amongst patients having more complex RVOT surgery, such as those having d-TGA, VSD, and pulmonic stenosis repair (classic Rastelli operation); truncus arteriosus repair; complex repairs of certain forms of double outlet right ventricle; or pulmonary atresia and VSD repair, we have had less success using catheter-based techniques.

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