THE RELATIVELY BENIGN NATURE of paroxysmal supraventricular tachycardia (PSVT) in patients with normal cardiovascular systems stands in sharp contrast to the malignant nature of supraventricular arrhythmias encountered in patients with structural heart anomalies.\(^1\)-\(^3\) Risk is especially high after complex corrective or palliative heart surgery. Patients after Fontan palliation for functionally single ventricle, patients after atrial rerouting for complete transposition of the great arteries, and patients after other complex atrial surgery may develop potentially lethal intra-atrial reentrant tachycardia (IART). Although similar in nature to atrial flutter, IART is often more difficult to manage, in part due to the presence of multiple reentrant circuits in the atria.

Abnormal ventricular myocardium is frequently a part of congenital cardiovascular anomalies, especially in older patients. The abnormal myocardium and post-myotomy scars produce a substrate for malignant ventricular tachyarrhythmias. Sudden cardiac death has been well documented in patients with congenital heart disease. It is imperative that arrhythmia in children, adolescents, and adults with congenital heart disease be treated aggressively using all possible therapeutic options. In many cases of postoperative arrhythmias, medical or surgical improvement in hemodynamic function will rectify rhythm disturbances. Antiarrhythmic agents are, in general, the first line of therapy following measures to improve hemodynamic function, although they may further worsen ventricular dysfunction and sinus node and atrioventricular (A V) abnormalities. Antiarrhythmic medications may also induce life-threatening arrhythmia because of their proarrhythmic properties. Antitachycardia pacing and radiofrequency (RF) or surgical ablation of arrhythmic substrates are other valuable adjuncts in the arrhythmia management.

This article reviews present experience in the application of RF ablation in congenital heart disease. It has to be stressed that RF ablation should be attempted in referral centers with expertise in arrhythmia management and RF ablation in the young as well as with experienced surgical backup.

PSVT with Structural Heart Disease

As in children and adolescents with normal cardiovascular system, PSVT in patients with congenital structural heart disease may be produced by the presence of an A V bypass tract or dual A V node conduction pathways. In fact, the incidence of Wolff-Parkinson-White (WPW) syndrome is higher in congenital heart disease compared to patients with normal hearts. As much as 20% to 37% of infants and children with WPW syndrome were shown to have a cardiovascular defect.\(^4\)-\(^7\) Ebstein anomaly of the tricuspid valve is the defect most frequently associated with the presence of an accessory A V bypass tract. In a large international study, the prevalence of ventricular pre-excitation in patients with Ebstein anomaly was 10%, and another 28% gave a history of paroxysmal palpitations.\(^8\) Rapid PSVT in patients with cardiovascular anomalies is
likely to produce hemodynamic decompensation because of reduced cardiac reserve in this group of patients. Moreover, ventricular fibrillation is a known complication in patients with ventricular pre-excitation, and it may be more likely in a setting of stressed and abnormal myocardium. Once safe pharmacological treatments have proven to be ineffective, RF ablation should be attempted as it is already considered a treatment-of-choice in older children, adolescents, and adults with most types of PSVT.

The approach to electrophysiologic testing in patients with congenital heart defects and PSVT follows general guidelines used for these procedures in their counterparts with normal hearts. However, the anatomic obstacles and variations in the conduction system distribution add to the challenge of RF ablation. It is imperative that the operator is knowledgeable about congenital heart disease when attempting RF ablation in these patients. Solid experience in cardiac catheterization in congenital heart disease and the ability to improvise and adapt techniques for the particular situation encountered will prove to be an asset to the operator. The electrophysiologic study should be preceded by meticulous echocardiographic and angiographic evaluation. Angiography will not only allow one to learn about the patient's anatomy but will also aid in positioning a fluoroscopic camera in parallel and orthogonal views of the A V valves. The electrode catheters are advanced into the right atrium in proximity to the sinus node and into the venous ventricle. Localization of the bundle-of-His may be difficult in heterotaxy syndromes and in other defects with abnormal anatomy of the conduction system. No particular difficulties were described with regard to the position of the electrode catheter in the coronary sinus except for patients with heterotaxy syndromes and A V discordance. The delineation of the right A V groove may be difficult, especially in patients with Ebstein anomaly. If the right coronary artery is well developed, one may attempt to cannulate it with a 2-French multipolar electrode catheter in order to outline the right A V ring. Epicardial mapping of the right A V ring will aid in localization of accessory A V pathways. In Ebstein anomaly, A V bypass tracts may be obscured by abnormal, fractionated ventricular electrograms, particularly along the posterior half of the right A V ring where they occur most frequently. The presence of abnormal ventricular potentials does not correlate with the extent of ventricular atrialization or tricuspid valve displacement. A right-sided bypass tract may also be found in cases with tricuspid atresia where A V ring definition is not clear.

Patients undergoing both right- and left-sided RF ablation should receive systemic heparinization. Accessory pathways are located by standard techniques involving identification of the Kent's bundle potential, recording the earliest ventricular activation signal during sinus rhythm in patients with ventricular pre-excitation, and recording of the earliest atrial activation electrogram at the time of orthodromic reciprocating A V tachycardia or during ventricular pacing. RF ablation of right-sided pathways is attempted from the venous approach, and ablation of left-sided tracts is done either through a patent foramen ovale, via a transseptal puncture or retrograde, across the aortic valve.

Difficult catheter manipulation in an abnormal heart, lack of standard anatomical landmarks, and unusual location of accessory A V bypass tracts render the time spent in the electrophysiology laboratory and the fluoroscopic exposure much longer than usual. Reported mean and median fluoroscopy times are well in excess of 60 minutes. Frequent occurrence of multiple bypass tracts in patients with congenital heart disease and particularly in Ebstein anomaly adds to the x-ray exposure and length of the procedure. Pulsed fluoroscopy units should be used to minimize radiation exposure. The number of RF pulses required to destroy the arrhythmic substrate is also higher than usual. An average of 9 to 22 RF pulses were required to interrupt the bypass tract in 2 studies compared to the average of 8 pulses in our experience in patients with normal hearts. Procedural difficulties and documented higher recurrence rate of tachycardia in patients with congenital heart anomalies frequently require more than one RF session to achieve successful ablation. In 47 patients reported, 68% required 1 procedure, 30% required 2 procedures, and 1 patient underwent 3 procedures. It has to be stressed that despite technical difficulties, long procedure time, and frequent need for repeat sessions, there were no serious complications. A surgically-treated arteriovenous fistula without any long-term sequelae complicated femoral cannulation in one patient.

In our experience, RF ablation of a bypass tract in
one patient with Ebstein anomaly resulted in ventricular perforation and tamponade. In a 13-year-old boy, endocardial mapping localized a bypass tract to the lateral aspect of the A V ring. A less than 10-s RF pulse produced atrial fibrillation and rapid development of clinical signs of cardiac tamponade. Atrial wall perforation was documented at emergent thoracotomy. A paper-thin wall of the atrialized right ventricle in Ebstein anomaly may predispose to such a complication.

The difficulties encountered in RF ablation in this patient group contribute to a lower success rate and more frequent recurrence rate compared to young patients with normal cardiovascular anatomy. In patients with A V reentrant tachycardia, complete cure is reported in 60% to 76% of patients compared to more than 80% in children and adolescents with normal hearts. Although not completely cured of the tachyarrhythmia, several more patients improved clinically after the procedure. Thirty-three patients with Ebstein anomaly represent the majority of cases with bypass tracts in whom RF ablation was attempted. A single pathway was found in 45% of patients, 2 pathways were documented in another 45%, and 3 or 4 pathways were found in 3 patients. Concealed pathways represented 27% of all documented bypass tracts. RF ablation was initially successful in 75% of A V bypass tracts. Recurrence of conduction over a bypass tract, as judged by recurrent pre-excitation or tachycardia, occurred in 15% of accessory pathways. Lower success rate of RF ablation and higher recurrence rate are blamed on several factors in Ebstein anomaly. Mentioned-above ventricular potential fractionation makes electrogram timing difficult, especially for posteriorly located pathways which constitute the majority of abnormal connections. Kent's bundle potentials cannot be identified either. Seemingly excellent signals may be recorded away from the bypass tracts. Delineation of the A V groove is difficult, especially in older patients with big hearts. Catheter stabilization becomes a problem in big hearts as well and the use of long sheaths may not improve it. Finally, complex geometry of accessory pathways may further hinder successful ablation.

Intra-atrial Reentrant Tachycardia After Heart Surgery

First paroxysmal and later incessant, IART is a known complication of atrial rerouting in patients with complete transposition of the great arteries, in children with single ventricle circulation after Fontan-type palliation, and following repair of other heart defects requiring atriotomy. Extensive atrial cuts and suture lines put patients at higher risk of later IART. Hemodynamic derangements which increase atrial pressure and atrial wall stress might also contribute to the etiology of IART.

IART encountered in patients after correction of congenital heart disease is a different entity from type I atrial flutter. The classic type I atrial flutter is produced by the reentrant counterclockwise activation of the right atrium. The essential role of the continuous anterior barrier provided by the tricuspid annulus and the role of the slow conducting isthmus have been clarified recently. The narrowest isthmus of the flutter wave is bordered by the inferior tricuspid annulus and the Eustachian ridge. It is at this isthmus that the flutter wave may be amenable to RF ablation. The upper link of the flutter wave is located superior to the superior vena cava. On the superior and
lateral right atrial aspects the flutter wave is broad, inferiorly directed, and constrained between the crista terminalis posteriorly and the tricuspid annulus anteriorly. On the septal aspect the wave front continues superiorly from between the coronary sinus ostium and the tricuspid annulus. The flutter wave proceeds towards the superomedial portion of the crista terminalis and is limited by the tricuspid annulus anteriorly, leaving the foramen ovale outside of the circuit. In contrast to the well-defined and invariable type I atrial flutter wave, IART may be a result of a variety of intra-atrial circuits.28-31 Programmed electrical stimulation may induce a multitude of IART configurations in a single patient as documented by variable P-wave morphologies on the surface electrocardiogram and by intracardiac mapping. Only a minority of published cases had only one IART induced. Although the clinical significance of induced multiple tachycardias is unclear, they attest to the difficulties encountered in ablation procedures.

Management of IART after previous atrial surgery is fraught with difficulties. IART is often refractory to standard medical therapies. The success rate of pharmacological therapy is limited. Digoxin and beta-blockers are still considered the first line medications, although their efficacy, alone and in combination, is less than 50%.3 Propafenone is a preferred class IC agent because of the concern about the proarrhythmic effect of flecainide and encainide.32,33 Amiodarone has proven to be the most efficacious agent in the management of IART, although it still fails to control the tachyarrhythmia in more than 20% of patients.3,34 Other class III antiarrhythmics such as sotalol might also prove effective.35 In our experience amiodarone has been the most effective agent, even in cases in which sotalol failed. When amiodarone fails to control the tachyarrhythmia, it may still improve clinical status. By slowing the A V conduction and the IART rate, it may provide adequate heart-rate control. Alone or in combination with medical therapy, antitachycardia pacing is an attractive option in those cases where IART is not completely controlled with medications or pharmacological therapy associated with unacceptable side effects.36,37 When considering anti tachycardia pacing, one must document that the tachycardia can indeed be terminated with anti tachycardia pacing. Different pacing algorithms, available in the anti tachycardia devices on the market, have to be tested. RF ablation of the IART reentrant circuit is another viable option discussed in detail below. Finally, surgical interruption of the reentrant wave or modification of atrial anatomy may have to be considered in cases refractory to other treatments.29 It has been frequently stressed that IART must be treated vigorously since it may result in patient demise.

The critical role of nonconductive surgical or functional barriers in the reentrant wave propagation has been documented in animal experiments.38-41 In a recent publication, Rodefeld et al38 showed reentrant circuits around both caval orifices in dogs after a lateral tunnel Fontan procedure. With two exceptions, there were no breakthroughs across the surgical lines. Invasive electrophysiologic testing in patients after atrial surgery suggests a vital role of atrial scars in clinical IART as well.28,30,31 Frequently, an isthmus of conductive tissue between a surgical barrier and an anatomical barrier (superior vena cava, inferior vena cava, tricuspid valve annulus) is inadvertently created, allowing for successful RF ablation. Baker et al30 accomplished successful ablation in an isthmus between a presumed atrial scar and the inferior vena cava, the superior vena cava, and the tricuspid annulus. His 14 reported patients underwent a variety of intracardiac operations and included 4 patients after Fontan procedures. Triedman et al28 and Kalman et al31 localized IART exit points in similar areas of the right atrium in patients after Fontan operation, Mustard and Senning procedures for transposition of the great arteries, closure of atrial septal defects, and after correction of ventricular septal defects. The authors commented that the exit points most likely reflected outlet from the area of slow conduction for the reentry wave. They were located near the site of atrio-pulmonary Fontan anastomosis, at the lateral junction of the right atrium and the superior vena cava, at the junction of the right atrium and the inferior vena cava, and near the triangle of Koch. Other sites of successful RF ablation were between the atrial septal patch and the inferior vena cava, between the atrial septal patch and the tricuspid annulus, between the intracardiac baffle and the tricuspid annulus, and at the lateral right atrium.

Attempts at RF ablation for IART have been reported in almost 50 patients.28-31 Anatomic and physiologic approaches have been employed. Anatomically-guided activation mapping during
Radiofrequency Ablation

IART has been reported to be successful in over 90%. In this approach, operative reports are reviewed and areas of previous surgical interventions are identified. Precise activation mapping during IART is done in the vicinity of neighboring anatomical barriers, and atrial activation electrograms preceding the surface P wave by more than 30 ms are presumed to represent impulse conduction through the isthmus. Multiple RF ablation lesions are produced in a linear way between the surgical and the anatomical barriers. Incomplete interruption of conduction across the isthmus is most likely responsible for IART recurrences. The authors of the anatomically-guided technique stress the importance of meticulous activation mapping to define the boundaries of nonconductive barriers.

Physiologic approach to RF ablation relies on identification of areas of unidirectional block and slow conduction. As in common atrial flutter, these areas usually lie in the anatomic isthmus for conduction. Although Baker et al used the anatomical approach and Triedman et al. attempted to identify reentrant segments of slow conduction, both groups targeted similar areas in the right atrium. In the physiologic approach, meticulous review of surgical reports and echocardiographic or even angiographic evaluation of the atrial anatomy precedes the procedure as well. The slow conducting isthmus is identified by concealed pace entrainment of the tachycardia. The post-pacing interval should be equal to the tachycardia cycle length, and the time from local atrial activation to the onset of the P wave should be close to the time from the pacing stimulus to the onset of the P wave during tachycardia entrainment. Atrial electrogram fractionation is not prerequisite for identification of the slow conducting isthmus. Precise recognition of an entry and an exit point from the isthmus is possible. As in anatomic approach, long, linear, and impenetrable RF lesions are necessary to prevent IART recurrences. The use of entrainment techniques allows to achieve an early success rate of 75%, and 50% of patients do not develop any recurrences in follow-up. As suggested by Baker et al., 30 RF ablation should be directed at those IART circuits which produce the same surface P wave configurations as the clinical arrhythmia or at those which are sustained in the electrophysiology laboratory. Long-term success rate of RF ablation for IART is still to be determined. Although much less effective, His-bundle ablation has also been used to control IART in selected patients.

Ventricular Tachycardia

Ventricular tachycardia (VT) is uncommon in children and adolescents with normal hearts. With the exception of preschool children where it may be life-threatening, VT is usually benign and does not require any treatment. More frequently, VT occurs in young patients with certain types of heart disease or it complicates surgery for congenital heart defects. Recurrent VT has been reported in myocarditis, Kawasaki disease, and in a variety of chronic cardiomyopathies. Ion channel disorders like the long QT syndrome and periodic paralysis may present with lethal VT. Malignant, recurrent VT may also accompany a number of congenital heart defects. VT is known to occur in mitral valve prolapse, tetralogy of Fallot, Ebstein anomaly, aortic valve stenosis, and in other anomalies. Life-threatening VT is a well-known early and late complication after repair of tetralogy of Fallot, ventricular septal defects, pulmonary and aortic valve stenosis, and a number of other defects. Sudden death may complicate VT after atrial rerouting for complete transposition of the great arteries.

The diversity of VT etiologies in young patients renders its treatment especially difficult. Accurate data on the incidence and ideal treatment of VT after surgery for congenital heart disease are missing. Almost all known antiarrhythmic medications have been employed at some point in time for the treatment of VT in children, adolescents, and young adults. In general, the pediatric use of these medication has been based on the experience learned from adult patients, most of whom present with ischemic heart disease. Similarly, rejection of further use of some preparations followed lessons learned from large randomized trials in the adult population. Pharmacological agents that have proven to be effective and safe are similar to those used for complex supraventricular arrhythmias. Amiodarone has proven highly effective in preventing VT recurrences. Sotalol, a beta-blocker with class III properties, is another promising agent. As ventricular arrhythmias may
be facilitated by increased sympathetic tone, beta-blocking therapy might also prove efficacious.63 The role of propafenone, other class IC agents, and class IA and IB medications in long-term therapy is now limited.33,64,65 In several disorders VT is well known not to respond to pharmacological or surgical treatments. Diffuse myocardial abnormalities of the long QT syndrome, Andersen syndrome, arrhythmogenic right ventricular dysplasia, and other cardiomyopathies require protection from implantable defibrillating devices.66,67 Implantable cardioverters/defibrillators constitute the last resort in cases of malignant ventricular arrhythmia refractory to medical treatment. Recently, several reports confirmed efficacy of surgical resection of arrhythmogenic foci in selected cases of VT.43,68,69 RF ablation which targets small areas in the subendocardium is likely to be of very limited value in myocardial cell disorders or other generalized disorders of the myocardium. Today, RF ablation has proven effective in only a limited number of VTs. Excellent results have been achieved in bundle-branch reentry tachycardia and idiopathic ventricular tachycardia originating from the septal aspect of the left ventricle and from the right ventricular outflow tract.70-73 Experience in RF ablation for VT in ischemic heart disease is also relatively small.74 Experience with RF ablation in congenital heart disease is even more limited. Less than two dozen cases of attempted RF ablation have been reported till now in patients after repair of tetralogy of Fallot, ventricular septal defects, and infundibular pulmonary stenosis.75-79

In tetralogy of Fallot, VT has been documented to originate from areas of surgical myocardial scarring68,69,80 although progressive myocardial fibrosis may also play a role in the etiology of VT.81 In 14 patients with tetralogy of Fallot,75-79 successful RF pulses were delivered in the right ventricular outflow tract in twelve and in the proximity of the ventricular septal defect patch in two. Endocardial mapping in these and other reported cases suggests macro-reentry circuits in the proximity of an anatomical barrier, possibly produced by surgical scars. Theoretically, right ventricular fibrosis might also produce micro-reentry circuits. In reported RF ablation of VT in patients after closure of a ventricular septal defect and in a patient after repair of transposition of the great arteries and a ventricular septal defect, the RF pulses were delivered on the right side of the interventricular septum, possibly in the proximity to the patch. In 2 patients after relief of subpulmonic infundibular stenosis, RF ablation was successful on the interventricular septum and in the RV outflow tract. It is interesting that in all reported cases, successful ablation was carried out in the right ventricle even though some of these patients presented with right bundle-branch block VT morphology, especially after repair of tetralogy of Fallot.

RF ablation of the tachycardia exit point, corresponding to the area of slow conduction, is believed to be responsible for noninducibility of VT after the procedure. Mid-diastolic potentials, representing the slow conducting segment of the reentry circuit, were recorded at the successful RF site in some cases. It is not clear how large the area of ablation should be to destroy the reentry circuit. In some cases, VT was terminated with very few RF pulses. On the other hand, Chinushi et al18 had to produce a linear scar between the presumed upper edge of previous myotomy and the line just below the pulmonic valve, similarly to the way the procedure is done for atrial flutter or IART. Pace maps have proven most successful in defining successful RF sites.75,79 Recording mid-diastolic slow conduction area electrograms is also predictive of successful ablation. Although local ventricular electrograms at the successful site preceded QRS complexes during VT by more than 30 ms, there are no guidelines as to the time interval by which the intracardiac electrograms should be ahead of the QRS complex. In some cases it is difficult to estimate the point of QRS onset. Other tools that have proven to be effective in localizing successful RF sites were entrainment of the tachycardia and tachycardia resetting by single extrastimuli without changes in morphology.79,82

Conclusion

Limited experience with RF ablation in congenital heart disease does not allow one to draw broad generalizations or firm conclusions. Still, with few exceptions, RF ablation should be considered safe. Ablation of AV bypass tracts is feasible and very effective, although more difficult because of abnormal cardiac anatomy. In patients with corrected or palliated congenital heart disease,
medical or surgical optimization of hemodynamic function may improve or even abolish arrhythmia. Because of the frequent presence of several reentrant circuits in IART, RF ablation still has a limited role in the management of postoperative atrial tachycardia. Medical treatment and possibly antiarrhythmic therapy should be considered first. RF ablation may improve or even abolish arrhythmia. Guidelines as to how and when to target ablation sites are developing and should allow for increasingly effective procedures.

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