

Mechanism and Therapy of Cardiac Arrhythmias in Adults with Congenital Heart Disease

STEVEN K. FURER, M.D., J. ANTHONY GOMES, M.D., BARRY LOVE, M.D., AND DAVENDRA MEHTA, M.D., PH.D.

Abstract

Over the past few decades, surgical advances have helped to prolong the lives of many young patients with congenital heart disease (CHD). However, as these patients reach adulthood, they are at risk for many late sequelae of their disease or of their corrective surgery. One of the unique challenges associated with CHD is the high incidence of cardiac arrhythmias that arise from the myocardial substrate created by abnormal pressure/volume changes, septal patches, and suture lines. Medical therapy has proven to be disappointing in treating a majority of these cases. Nonetheless, radiofrequency catheter ablation (RFA), an effective tool in treating atrial and ventricular arrhythmias in structurally normal hearts, has been used to treat arrhythmias in adults with congenital heart disease. This review will discuss some of the common congenital heart diseases in adults and the arrhythmias associated with them, as well as the therapeutic modalities used to treat them. Finally, it will present Mount Sinai Hospital's experience in using RFA for the management of cardiac arrhythmias in adults with congenital heart disease.

Key Words: Congenital heart disease, radiofrequency ablation, arrhythmia.

Introduction

AN INCREASING NUMBER OF PATIENTS with congenital heart disease are reaching adulthood, due to the success of corrective surgical procedures. But one of the problems associated with longer survival is cardiac rhythm disturbances. The high incidence of cardiac arrhythmias in the adult with congenital heart disease (Table 1) is a result of the irritable foci caused by abnormal pressure/volume changes, and reentrant circuits created by septal patches and suture lines that are characteristic of this population.

Intracardiac shunting, leading to chronic volume and pressure overload, varying degrees of pulmonary hypertension, and ventricular dysfunction all contribute to arrhythmogenesis. In response to atrial volume or pressure overload, atrial stretch prolongs

atrial refractoriness, making the atria vulnerable to the induction of atrial flutter and atrial fibrillation.

Atrial flutter is usually caused by a reentrant circuit. The circuit revolves around one or more electrical barriers such as the tricuspid valve or an atrial suture line. Injured or scarred atrial tissue from surgical incisions, such as an atriotomy site or an atrial septal defect (ASD) patch repair, creates zones of slow conduction that allow the circuit to propagate. Atrial premature beats lead to the initiation of reentry (atrial flutter) by further slowing impulse conduction in damaged tissues around area of blockage.

Ventricular tachycardia (VT) is seen in a smaller number of patients, most notably in those with tetralogy of Fallot (TOF). Progressive myocardial fibrosis of the right ventricle, with corresponding slowing of conduction, increases the incidence of reentrant ventricular arrhythmias. The surgical procedure for a right ventriculotomy scar and ventricular septal defect (VSD) patch repair also contributes to the arrhythmia substrate. The VSD patch repair provides a fixed anatomic obstacle around which reentrant arrhythmias may occur. In addition, isolated muscle bundles surrounded by fibrosis in the margin of the junction between the right ventricular outflow tract patch-right ventricular free wall will slow conduction and further con-

From the Division of Cardiac Electrophysiology, Cardiovascular Institute, Mount Sinai Hospital and Medical School, New York, NY.

Address all correspondence to Davendra Mehta, M.D., Ph.D., F.A.C.C., Director, Electrophysiology Section and Cardiac Arrhythmia Service, Cardiovascular Institute, Box 1054, Mount Sinai School of Medicine, One East 100th Street, New York, NY 10029; e-mail: Davendra.Mehta@mssm.edu

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TABLE 1*Congenital Heart Defects and Their Associated Arrhythmias*

Atrial Septal Defect	<ul style="list-style-type: none"> • Atrial flutter • Atrial fibrillation
Ebstein's Anomaly	<ul style="list-style-type: none"> • Accessory pathways • Atrial flutter • Atrial fibrillation • Ventricular tachycardia
Fontan Patients	<ul style="list-style-type: none"> • Atrial flutter
Tetralogy of Fallot	<ul style="list-style-type: none"> • Ventricular tachycardia • Atrial flutter • Atrial fibrillation
Transposition of the Great Arteries	<ul style="list-style-type: none"> • Atrial flutter • Atrial tachycardia • Atrioventricular nodal reentrant tachycardia • Ventricular tachycardia

tribute to the initiation of reentry. Right ventricular dilatation and stretch with slowed ventricular activation contribute to the creation of reentrant circuits within the right ventricle, whereas impaired hemodynamics, predominantly right ventricular dilatation due to pulmonary regurgitation, are responsible for sustaining VT, once initiated.

Atrial Septal Defect

Atrial arrhythmias, primarily atrial flutter and atrial fibrillation, are the most frequently seen arrhythmias in an adult patient with an ASD. Age at surgical repair is an important predictor of persistent or new arrhythmias after ASD closure. While ASD closure often leads to improved functional status and reduces the risk of right-sided heart failure and progressive pulmonary hypertension, it is not curative or totally preventative for arrhythmias. Older age at surgery, the presence of preoperative atrial flutter or atrial fibrillation, and the presence of postoperative atrial flutter, atrial fibrillation or junctional rhythm, are predictive of late postoperative atrial flutter or atrial fibrillation (1). Several years after surgical closure, 60% of patients in one study continued to have atrial flutter or atrial fibrillation, particularly those older than 40 years of age (1). This data suggests that early closure of an ASD reduces the long-term occurrence of atrial arrhythmias.

Atrial flutter is right sided and caused by a reentrant circuit usually found around the tricuspid ring or the atriotomy scar. Catheter ablation of the area of slow conduction is the treatment of choice. Delivered from the tip of a catheter to the endocardial surface, radiofrequency energy produces con-

trolled focal tissue ablation. Recent data on entrainment mapping (Fig. 1) combined with three-dimensional electro-anatomic mapping showed 81% successful ablation of macro-reentrant circuits in adult congenital heart disease patients, half of whom had an ASD repair (2). In older patients with atrial fibrillation, the standard maze procedure has been performed at the time of ASD closure with excellent results and no recurrence of atrial fibrillation (3). (The maze procedure employs several small incisions in the right and left atria to interrupt the potential reentrant pathways required for atrial fibrillation maintenance.)

Ebstein's Anomaly

This condition involves the apical displacement of one or more tricuspid valve leaflets from the atrioventricular (AV) ring into the right ventricle, associated with a diminution in size of the functioning right ventricle. The deformity and displacement of the tricuspid valve can result in severe tricuspid insufficiency and right atrial dilatation. These abnormalities, along with the frequent coexistence of an ASD, predispose to the development of atrial arrhythmias. In addition, there is a high prevalence of accessory pathways, usually located on the right side or in the posteroseptal region. This is presumed to be a consequence of the discontinuity of the central fibrous body and septal AV ring,



Fig. 1. Intracardiac electrograms showing entrainment mapping during scar-related atrial flutter in a patient with an ASD repair. Arrows indicate activation sequence. Top 2 channels show surface EKGs. CS channels represent recordings from the coronary sinus, 1, 2 being distal and 9, 10 proximal electrograms. T1 – 10 represent recordings from multipolar catheter placed around the tricuspid ring, T1 near the inferior vena cava and T10 in the upper septal region of the right atrium. Activation sequence is counterclockwise going from T10 to T1. Tachycardia cycle length was 295 ms. Pacing was performed at 270 ms. Post-pacing interval (PPI) was same as tachycardia cycle length (295 ms), indicating entrainment. This confirms that pacing was performed in the tachycardia reentry circuit.

resulting in persistence of fetal accessory AV pathways. Accessory pathway variants, such as Mahaim fibers, are more common in this condition.

About one third of patients with Ebstein's anomaly have an arrhythmia, most commonly atrioventricular reentrant tachycardia or Wolff-Parkinson-White, and to a lesser extent, atrial tachycardia, atrial flutter, atrial fibrillation, and VT. Atrial arrhythmias are encountered with increased frequency, with increasing age and duration of follow-up.

Radiofrequency ablation (RFA) is used to treat atrial flutter, incisional atrial tachycardia and accessory pathway conduction in patients with Ebstein's anomaly, with a success rate of 76% (4), compared to 95% of patients without the anomaly. Factors that contribute to decreased success include the complex geometry of the accessory pathway, location of the pathway along the atrialized portion of the right ventricle, abnormal endocardial activation potentials confounding identification of the accessory pathway, distorted anatomy of the AV ring, and the presence of multiple pathways. In addition to a low success rate, RFA in Ebstein's anomaly is associated with a 25% risk of recurrence (4).

Refractory arrhythmias are an indication for surgical repair. For patients undergoing tricuspid valve repair or replacement, therapeutic options include surgical cryoablation, as well as concomitant right atrial maze for chronic atrial flutter or biatrial maze for atrial fibrillation. Surgical intervention for accessory pathway-mediated tachycardia and atrioventricular node reentrant tachycardia (AVNRT) has had excellent results, without recurrence from these arrhythmias (5). In contrast, surgical intervention for atrial fibrillation/atrial flutter is less effective, with a recurrence rate of 40% (5).

The Fontan Operation

The Fontan operation is a palliative surgical procedure used for tricuspid atresia, pulmonary atresia, complex single ventricle and double-inlet ventricle. Older variants of the procedure such as the right atrium-pulmonary artery connection, have given way to newer modifications such as the lateral tunnel and external conduit, which reduce the distention of the right atrium. In response to chronic stretching secondary to persistent pressure overload, the fontan right atrium remodels and dilates. This occurs in parallel to the change in electrophysiological properties manifested by the atrial conduction delay and the imbalance of the conduction heterogeneity.

Risk factors for early postoperative arrhythmias (Table 2) include preoperative AV valve regurgitation and an abnormal AV valve (6). Older age at operation is also a risk factor, because these

patients will have experienced long periods of hypoxia, volume overload, resultant ventricular hypertrophy and abnormalities of diastolic filling prior to surgery. Risk factors for late atrial arrhythmias include poor preoperative functional status, previous atrial septectomy, preoperative atrial tachyarrhythmias, older age at operation, need for AV valve replacement, pulmonary artery reconstruction, atriopulmonary anastomosis, early postoperative atrial tachyarrhythmias, postoperative sinus node dysfunction, and length of follow-up (7, 8).

Atrial arrhythmias develop in 41–61% (9, 10) of Fontan patients. The median time from fontan operation to the first arrhythmia is 7 years, with 50% having recurrent arrhythmia despite treatment (11). Fontan patients with arrhythmias have been shown to have longer P-wave duration and greater P-wave dispersion than those who do not have arrhythmias (10). Patients who develop atrial arrhythmias are more likely to develop heart failure, right atrial thrombus, left atrial enlargement, right atrial enlargement, and moderate-severe systemic valve regurgitation. But there is no difference in survival time (9). Sustained atrial arrhythmias may present with hemodynamic compromise in patients with atrial flutter and 1:1 or 2:1 AV conduction. Slowed atrial conduction results in slower flutter rates, with P wave morphology reflecting the location of reentry.

Sinus node dysfunction is common, occurring either as a consequence of the operation or due to compromise of sinus node blood supply, and is associated with the development of atrial arrhythmias. Approximately 10–15% of patients will exhibit sinus bradycardia and/or junctional escape rhythm on follow-up.

Management of the resulting atrial arrhythmias generally entails a combination of antiarrhythmic drugs, pacemakers, RFA, and reoperation. Antiarrhythmic drugs must be used cautiously, as many of these patients have underlying sinus node disease. Further, the slowing of the atrial rate facilitates 1:1 conduction during atrial reentry. Finally, proarrhythmia in a patient with ventricular dysfunction is a concern.

TABLE 2

Risk Factors for Arrhythmias in Patients Who Underwent the Fontan Operation

Preoperative AV valve regurgitation
Older age at operation
Poor preoperative functional status
Previous atrial septectomy
Preoperative atrial tachyarrhythmias
Pulmonary artery reconstruction
Atriopulmonary anastomosis
Early postoperative atrial tachyarrhythmias
Postoperative sinus node dysfunction
Length of follow-up

AV = atrioventricular

Pacing may be required for sinus node disease and/or AV block. Due to anatomic constraints, atrial and ventricular leads have to be placed via an epicardial approach. Antitachycardia pacing has been combined with medical therapy with some success. Atrial rate-responsive pacemakers are preferred, as there is a high incidence of sinus node dysfunction. Rapid pacing prevents bradycardia and atrial extrasystoles, and may eliminate initiation of reentrant tachycardias.

RFA is associated with an immediate success rate of 83% but an at-least 20% incidence of recurrent tachycardia at short-term follow-up (12). Potential causes of recurrence include persistently abnormal hemodynamics, massive right atrial dilatation with distorted anatomic landmarks, stasis related to low flow, resulting in poor catheter tissue contact, and the inability to create deep lesions in these markedly thickened and fibrotic atrial walls. More common ablation sites often include the region of the fontan anastomosis, the lateral right atrial wall and the inferior right atrium.

Refractory atrial arrhythmias are an indication for reoperation. Surgical conversion from an atrio-pulmonary anastomosis to a total cavopulmonary anastomosis, along with electrophysiologically guided cryoablation, has shown excellent results in preventing recurrent arrhythmias and reducing symptoms. In a study by Deal et al., surgical cryoablation and antitachycardia pacing resulted in 83% of patients being arrhythmia-free without medications (13). Cryoablation is predominantly targeted at three locations: the inferomedial right atrium between the inferior vena cava os and coronary sinus, the superior rim of the ASD patch, and along the lateral right atrial wall corresponding to the length of the crista terminalis. For patients with atrial flutter, cryoablation is employed as part of the modified right-sided maze procedure, along with excision of the right atrial appendage and placement of an atrial pacemaker. In patients with atrial fibrillation, the maze-Cox III procedure combines the lesions of the right-sided maze with cryoablation from the pulmonary veins towards the posterior mitral or tricuspid valve annulus, and from the isolated pulmonary veins to the edge of the excised left atrial appendage.

Tetralogy of Fallot

This condition consists of four features: sub-pulmonary infundibular stenosis, VSD, overriding aorta and right ventricular hypertrophy. Patients with this condition are predisposed to both ventricular and atrial arrhythmias. Sustained VT has a prevalence of 4–7% (14) and is usually a right ventricular outflow tract reentrant tachycardia with left bundle branch morphology. Nonsustained ven-

tricular arrhythmias are present in up to 60% of Holter readings, but recent studies have demonstrated no predictive value for subsequent sustained VT or sudden cardiac death (SCD) (15).

There is a small but persistent risk of late SCD for patients with tetralogy of Fallot (TOF), with an estimated incidence rate of 0.5–6% (15–17). Initially thought to be due to conduction abnormalities and asystole, ventricular arrhythmias are now thought to be the primary cause. Older age at initial repair, moderate or severe pulmonary regurgitation, a history of sustained VT, moderate or severe left ventricular dysfunction, a QRS duration of 180 ms or more, and a rapid increase in QRS duration are predictive of risk for sudden cardiac death (Table 3) (15). The combination of moderate or severe left ventricular systolic dysfunction and QRS duration greater than 180 ms has a positive predictive value of 66% and negative predictive value of 93% for sudden cardiac death (18). QRS duration greater than 180 ms has been shown to have 100% sensitivity and 95% specificity (19) for sustained VT and SCD in tetralogy patients. QRS prolongation reflects initial damage to the right bundle branch during tetralogy repair and late progressive QRS prolongation, secondary to RV dilatation, invariably is the result of chronic pulmonary regurgitation. Moderate-to-severe pulmonary regurgitation and aneurysmal dilatation of the outflow tract are observed with greater frequency in patients with sustained VT (14). In addition to the above findings, electrophysiology study (EPS) is used to risk-stratify these patients further, as patients with inducible arrhythmias are probably at the highest risk.

The morbidity of TOF patients is also influenced by the presence of atrial arrhythmias. One third of TOF patients manifest atrial arrhythmias (Fig. 2; 20), with congestive heart failure and recurrence of atrial arrhythmia observed on follow-up. Risk factors include older age at operation, increased atrial size, tricuspid or pulmonary regurgitation and ventricular dysfunction. Atrial flutter and atrial fibrillation were more common in patients who had long-lasting pulmonary artery shunts, early operations for residual hemodynamic lesions, older age at repair, and mod-

TABLE 3
Risk Factors for Sudden Cardiac Death in Patients with Tetralogy of Fallot

Older age at initial repair
Moderate or severe pulmonary regurgitation
History of sustained ventricular tachycardia
Moderate or severe left ventricular dysfunction
QRS duration of 180 ms or more
Rapid increase in QRS duration

erate-to-severe tricuspid regurgitation (15). Tricuspid regurgitation leads to right atrial dilatation from volume or pressure overload, and prolongs atrial refractoriness, thus creating a substrate for atrial arrhythmogenesis. These patients usually present with palpitations and less often with syncope or presyncope. Atrial flutter with 1:1 AV conduction has been postulated as a cause of SCD. Sinus node dysfunction has been reported at 36% (20).

RFA has been successfully employed for atrial flutter (Fig. 3) and VT. A large area of low voltage, consistent with scarring, can usually be identified along the free wall of the right atrium, and successful ablation can be accomplished by creating a line between the lower margin of the scar and the inferior vena cava. Prerequisites for VT ablation include inducibility on EPS, hemodynamic stability during the tachycardia to enable adequate mapping, and monomorphic morphology. VT has been localized to the right ventricular outflow tract and the infundibulotomy scar or the septal surface of the VSD patch repair, with a high immediate success rate and low rate of recurrence.

Surgical reoperation to correct the hemodynamic substrate has been attempted. A significant reduction in preexisting VT and QRS duration stabilization has been reported following valve replacement for severe pulmonary regurgitation, while concomitant intraoperative electrophysiolog-

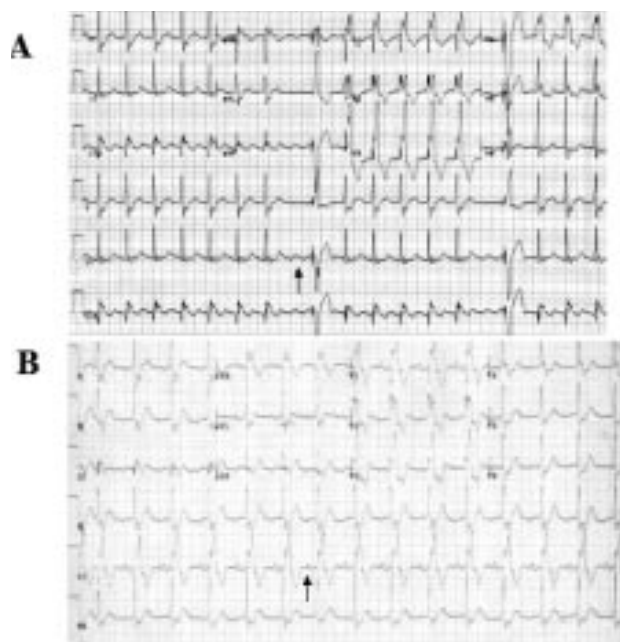


Fig. 2. Twelve-lead EKGs during 2 different tachycardias in a patient with tetralogy of Fallot. (A) shows p waves (arrow) suggestive of typical atrial flutter with a saw-tooth pattern in lead II. (B) shows a second tachycardia with p wave morphology suggestive of an atrial tachycardia. P waves are positive in lead II (arrow). This was mapped to be a focal atrial tachycardia originating from the high right atrium.

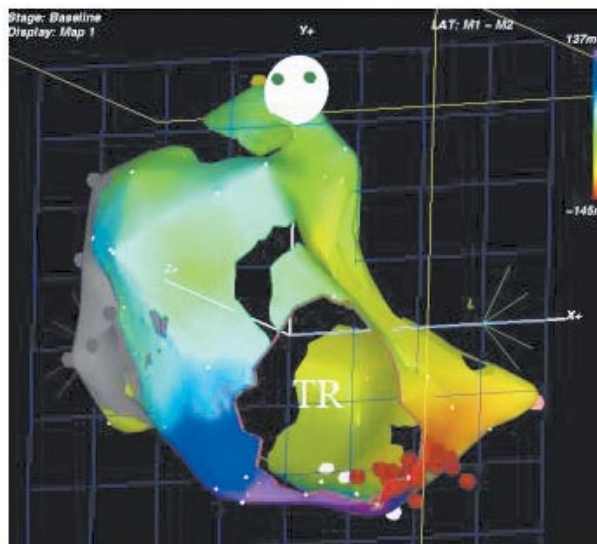


Fig. 3. Electroanatomic map of right atrium during atrial flutter in a patient with tetralogy of Fallot. Activation sequence is color-coded. Red areas represent early activation and purple areas late activation. This image depicts counterclockwise activation around the tricuspid ring (TR). Red spots represent areas of catheter ablation that lead to termination of atrial flutter. Gray areas represent areas with scarring with no electrogram. Reentry was not related to the scar.

ical-guided cryoablation prevents recurrence of preexisting tachyarrhythmias (21). For patients with atrial tachycardia requiring reoperation, a modified maze procedure should be considered. Patients with VT who cannot be ablated are treated with an implantable cardioverter defibrillator (ICD).

Transposition of the Great Arteries

Transposition of the great arteries (TGA) occurs when the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle. At birth, the patent foramen ovale and patent ductus arteriosus allow for mixing of blood to sustain life. The Mustard and Senning operations were developed to correct the physiologic abnormality by forming a baffle within the atria to switch the flow of blood at the inflow level. The arterial switch operation was subsequently created to enable the left ventricle to become the systemic ventricle. The Mustard repair requires extensive incisions and suture lines in the atria. This results in intra-atrial conduction delays and abnormalities in atrial refractoriness, creating the substrate for atrial flutter.

In the adult Mustard patient, there is a high incidence of systemic right ventricular dysfunction associated with late atrial arrhythmias. There is also a higher risk of SCD, reported at 7% on long-term follow-up (22), attributed to ventricular arrhythmias or atrial flutter with 1:1 AV conduction degenerating to

ventricular fibrillation, and asystole. Increased QT dispersion is a marker for heterogeneity of ventricular repolarization, with fast heart rates due to physical stress or atrial flutter, and ectopic ventricular beats serving as triggers to reentrant ventricular arrhythmias. Increased QT dispersion and loss of sinus rhythm have been associated with SCD.

At long-term follow-up of adult Mustard patients, only one third remained arrhythmia-free (23). A progressive loss of sinus rhythm has been observed at a rate of 2.4%/year with sinus rhythm present in 77% at 5 years and 40% at 20 years (24). The majority of supraventricular tachycardia are due to atrial flutter (23). Risk factors for supraventricular tachycardia include pulmonary hypertension, systemic ventricular dysfunction, and childhood junctional rhythm. Loss of sinus rhythm has been associated with previous septectomy, postoperative bradycardia, late atrial flutter, and preoperative arrhythmia. This loss of sinus rhythm is in contrast to the long-term maintenance of sinus rhythm in 95–98% (25) of arterial switch patients. The incidence of supraventricular tachycardia is also significantly lower, with a rate of 5% (25) in arterial switch patients, compared to 48% in Mustard patients. The arterial switch operation is likely to be associated with a lower incidence of long-term atrial arrhythmias, because there are no atrial scars.

Approximately one fifth of adult Mustard patients will require pacing in long-term follow-up for symptomatic sinus node dysfunction or atrioventricular block, or to facilitate treatment of tachyarrhythmias (23, 26). Careful evaluation of individual anatomy and exclusion of baffle leaks must be performed prior to determining pacemaker location.

Given the high prevalence of arrhythmias in this population, RFA has been employed with a 73–83% success rate (27–29), and a 12% rate of recurrence (27). The isthmus of tissue between the tricuspid valve and inferior vena cava orifice, the area of the mouth of the coronary sinus, and the region extending from the mouth to the tricuspid annulus are critical components in the reentry circuit. Intra-atrial reentry may involve either atrium and may require a retrograde aortic approach to facilitate ablation in the pulmonary venous atrium. In addition, focal atrial tachycardia has been localized adjacent to baffle suture lines, while typical AVNRT can also occur.

Sudden Cardiac Death

A large population-based study estimated the incidence of late SCD in patients with congenital heart disease who have undergone surgery to be 0.9/1,000 patient-years (30). There is an increased incidence in patients with TOF, TGA, coarctation of the aorta and

aortic stenosis. The event rate for the previously mentioned group of patients with obstructive left heart lesions and cyanotic defects was found to be 2.2/1,000 patient-years compared to a rate of 0.14/1,000 patient-years in those with left-to-right intracardiac shunt lesions or pulmonic stenosis. While the most common cause of SCD is arrhythmic in origin, embolic events, aneurysm rupture, and acute ventricular failure have been reported as well.

The risk of SCD after repair of TGA starts early after repair and remains constantly high thereafter. The rate of SCD is 4% at 10 years and 9% at 20 years. In contrast, the risk of SCD after repair of TOF is less and occurs only late after repair. The risk in this group is 2.2% at 20 years, 4% at 25 years, and 6% at 30 years. Coarctation of the aorta also shows a higher risk of late SCD, 1% at 20 years, and 8% at 30 years, associated with profound left ventricular hypertrophy. Identification of patients with the highest risk, correction of hemodynamic defect, and use of ICDs will help prevent sudden death. The use of ICDs for the prevention of SCD in adults with congenital heart disease is relatively safe and effective.

Mount Sinai Data on Radiofrequency Ablation

Between 2001 and 2004, 28 EPSs were performed in 18 adult patients with surgically repaired congenital heart disease, to evaluate cardiac arrhythmias. Conventional activation mapping, electro-anatomical mapping, and entrainment mapping were performed to delineate each patient's arrhythmia. There were 13 female and 5 male patients, with a mean age of 40 (range 24–67 years) at EPS. There were 4 patients with TOF, 3 with TGA, 1 with a single ventricle treated with a Fontan operation, 1 with a VSD, 2 with Ebstein's anomaly, 2 with Ebstein's anomaly and an ASD, and 5 with an ASD. Patients were followed a mean of 22 months (range 3–44 months).

The arrhythmias documented on EPS included: 15 atrial flutters, 3 AVNRTs, 6 accessory pathways, 2 atrial tachycardias, and 2 atrial tachycardias/atrial flutters. RFA was performed on the induced arrhythmia in each patient. The overall immediate post-procedure success rate was 26/28 (93%). Six of the 18 patients (33%) had a recurrence of arrhythmia, 4 had a new arrhythmia, and 2 had an arrhythmia identical to the initial tachycardia. These data demonstrate that RFA is successful. Recurrence rates can be significant; however, the recurrent arrhythmia is often different from the one ablated and can be treated by repeat ablation procedure.

Eight patients with TOF presented with VT; all had ICD implants. These patients were followed for 6 months to 7 years. Four of the patients have

had recurrent VT appropriately treated by ICD. None of these patients have died.

Conclusion

Adults with congenital heart disease often experience cardiac complications many years after surgical repair of their defect. As these patients age, cardiac arrhythmias typically cause significant morbidity. These arrhythmias are treatable with hybrid therapy involving drug therapy, catheter ablation, implantable devices, and surgical repair of the underlying hemodynamic abnormality. Appropriate treatment is associated with long-term maintenance of sinus rhythm and decreased morbidity and mortality.

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