Cardiac arrhythmias remain a major source of morbidity and mortality for patients with congenital heart disease (CHD). This is especially true for those who underwent successful operative correction or palliation during childhood, only to present decades later with a complex arrhythmogenic substrate caused by their abnormal anatomy, surgical scars, and suboptimal hemodynamics of long duration. Surgical advances are attenuating some of the hemodynamic and suture-related proarrhythmic factors; however, they cannot be eliminated totally and are only part of the puzzle. Inherent preoperative abnormalities and chronic remodeling continue to affect conduction properties of the myocardium, despite many intelligent modifications in surgical approach.

Although antiarrhythmic medications retain a role under certain circumstances, there is growing reliance on interventional procedures to address rhythm disturbances in children and adults with CHD. This article will provide an update on clinical data and technological advances in this field, with a focus on ablation and device therapy.

Atrial Tachycardias

Atrial tachycardias are the most common arrhythmias observed in patients with CHD. They are a major cause of symptomatology, and in some cases they can contribute to sudden cardiac death (SCD). Bouchardy et al found in a large cohort of adults with CHD that the prevalence of some type of atrial tachycardia was 15%, with a lifetime risk of ∼50% regardless of severity of the CHD lesion. Atrial arrhythmias were associated with a 50% increase in mortality, a 2-fold increased risk of heart failure or stroke, and a 3-fold higher risk of requiring some major cardiac intervention. Single ventricle patients with a Fontan circulation and those with transposition of the great arteries after Mustard or Senning operations have the highest burden of atrial tachycardia, but even simple atrial septal defect closure can be associated with atrial rhythm disturbances.

Most atrial tachycardias in patients with CHD involve macreentrant circuits localized to the right atrium (RA). These can include classic atrial flutter through the cavitricuspid isthmus, as well as atypical circuits related to atriotomy incisions and other scars. The general term intra-atrial reentrant tachycardia (IART) has been used to describe all these circuits in CHD. Focal atrial tachycardia can also be observed in a small number of patients, usually arising from locations mapped in close proximity to a scar zone.

Atrial fibrillation (AF) is also being reported with increasing regularity in patients with CHD, particularly those with left heart obstructive lesions, chronic left atrial dilation, and advanced age. There are some disturbing trends being recognized in this regard. It was recently reported that nearly a quarter of patients will develop AF many years after straightforward atrial septal defect closure. In addition, it has been noted that as many as 30% of patients with CHD who have undergone prior successful IART ablation may present with AF at some later time. Clearly, AF is emerging as a growing concern as more patients with CHD survive to older ages.

Ablation for IART in CHD

As a consequence of better equipment and expanded procedural experience, the acute success rate for catheter ablation of organized IART or focal atrial tachycardia in CHD has now improved to ≈85% (Table 1). Distinctive patterns for IART circuit location have been uncovered that vary according to underlying CHD lesion and method of repair, allowing one to predict with reasonable certainty the location of usual suspect sites for productive ablation. To take advantage of these patterns, intracardiac and venous anatomy must be well understood. Operative notes are mandatory reading because surgical techniques vary significantly among patients, surgeons, and eras. Femoral venous access can sometimes be challenging in patients with CHD who have undergone multiple prior catheterizations and may necessitate alternative approaches.

It is also important to ascertain the precise status of the atrial septum. If transseptal access to the left atrium becomes necessary for mapping and ablation in patients with CHD, entry can be challenging if the septum is thickened and calcified or has been reconstructed with synthetic material and occlusion devices. In such cases, if standard Brockenbrough technique fails, puncture will often be facilitated by delivering radiofrequency energy through the needle tip. However,
there may be residual defects or patch margin leaks in the septum that the operator can take advantage of for easy left atrial entry. Knowing details of this sort in advance of the procedure, or as the first step of a procedure, it significantly improves the likelihood of success.

Similar to any other mapping study for a complex tachycardia substrate, clear imaging of anatomy, and accurate integration of these images with electric activity, is crucial for IART ablation. Anatomic registration has been performed using a wide variety of techniques, beginning with standard angiography, moving to 3-dimensional (3D) spatial shells generated by the catheter tip, and now more detailed shells involving computed tomography/MRI scans and intracardiac echo. Preferences vary among institutions and operators, but our own center has come to rely most heavily on intracardiac echo. Modern 3D systems that register anatomy and display circuit location have greatly improved IART ablation in CHD, to localize and confirm the circuit according to standard methodology for macroreentry.

It is now well established that the culprit circuits for IART in simple 4-chamber CHD, for example, atrial septal defect closure and tetralogy of Fallot (TOF) repair, involve the cavoatrial isthmus but will frequently take a figure-of-eight or dual loop configuration, with an outer loop around an atriotomy scar on the lateral RA wall. Thus, effective ablation in these patients typically requires interruption of both loops by creation of conduction block at the isthmus, as well as at the narrow corridor between the lower edge of the atriotomy scar and the inferior vena cava.

The classic circuit for IART in patients after Mustard or Senning operations for transposition also involves the cavoatrial isthmus, but in this case the isthmus is located on the pulmonary venous side of the atrial baffle. Access to the pulmonary venous atrium is almost always necessary for ablation of IART in these hearts. This can be accomplished via a retrograde aortic approach through the tricuspid valve or by transbaffle puncture.

The most difficult IART circuits to map and ablate involve patients with single ventricle physiology who have undergone the Fontan procedure. The surgical design for this operation has evolved over the years, and the different designs vary in terms of both arrhythmogenic potential and IART circuit location(s). The original Fontan with RA-to-pulmonary artery anastomosis usually resulted in RA dilation, wall thickening, large areas of low-voltage or fractionated potentials, and an atrial tachycardia incidence as high as 20% to 40%. The IART circuits in such patients are often multiple and can occur just about anywhere within the dilated RA chamber. Ablation success and freedom from recurrence tend to be lower in this group than in any other CHD category, prompting some centers to advocate operative intervention for conversion to one of the newer Fontan designs, along with excision of redundant RA tissue and a surgical maze. Newer Fontan designs bypass the RA to a large extent, using either a lateral tunnel or an extracardiac conduit, thereby avoiding RA dilation and resulting in significant reduction in IART incidence to 2% to 7%. However, for the occasional patient who does develop IART in a modern Fontan design, circuit locations are typically on the pulmonary venous side of the tunnel or conduit, requiring complex techniques for transseptal access.

Modern 3D systems that register anatomy and display circuit location have greatly improved IART ablation in CHD,
Pathological specimens comparing right atrial wall thickness between normal heart (A) and postoperative single ventricle patient who had undergone a Fontan operation (B).

Ablation for AF in CHD

As mentioned, AF is emerging as a more significant problem as patients with CHD attain older ages. There is now a small but growing literature describing catheter ablation of AF for this population. Most cases have involved fairly straightforward lesions, such as atrial septal defects, although a few reports of more complex anatomy also exist. One item that may set these cases apart from more conventional AF ablation in a structurally normal heart is the challenge of achieving left atrial access across patches and septal occlusion devices, but there is now ample evidence that this can be accomplished safely.

Catheter ablation for AF will almost certainly become a more common intervention for CHD in the near future.

Ablation of Other Supraventricular Tachycardias in CHD

The recent clinical success of the Cone procedure has resulted in a growing number of patients with Ebstein anomaly being referred for surgical reconstruction of the abnormal tricuspid valve. In light of the well-known association of accessory pathways (APs) with this condition, electrophysiological testing and AP ablation are becoming a more routine preoperative intervention. Early reports of AP catheter ablation in Ebstein from the 1990s had suggested a lower rate of acute success and higher recurrence than in structurally normal hearts. More recent publications confirm that these cases remain challenging, despite advances in imaging, mapping, and ablation energy delivery. The difficulty stems from the presence of multiple APs in >50% of cases, trouble defining the true atrioventricular groove (Figure 4), and dramatic RA dilation that can confound catheter navigation and tip stability. Nonetheless, preoperative testing remains an important exercise. Not only can APs be mapped and (in most cases) ablated, but a high percentage of patients will also be identified to have secondary arrhythmia substrates, including atrial flutter, atriofascicular fibers, and even ventricular tachycardia (VT). These secondary mechanisms can often be addressed with catheter ablation during the same session in patients with Ebstein anomaly. Should catheter ablation fail, mapping data can still be used to guide surgical ablation in the operating room as part of the Cone procedure.

Supraventricular tachycardia caused by atrioventricular nodal reentry can also be identified occasionally in patients with CHD. Even though this arrhythmia substrate is relatively conventional, procedures can be challenging in malformations.
that are associated with displacement of the compact node, especially when surgical patches and baffles confound catheter tip navigation to the target site.\textsuperscript{53,54}

\textbf{Ventricular Tachycardias}

VT remains an uncommon but potentially lethal complication of CHD. Fortunately, detection techniques and interventional treatment options have been improving steadily. It can be helpful to draw a distinction between two categories of VT in the population with CHD. The first involves developmental and surgical abnormalities within ventricular muscle that creates discrete corridors of slow conduction capable of supporting \( \geq 1 \) monomorphic reentrant VT circuits. Specific CHD lesions that exemplify this anatomic predisposition to macroreentrant VT include: (1) TOF, (2) ventricular septal defects (VSD) closed through a ventriculotomy incision, and (2) Ebstein anomaly. These are the sort of patients most likely to benefit from catheter or surgical ablation. The second category involves more disorganized polymorphic VT and ventricular fibrillation that occur in diffusely abnormal myocardium, similar to arrhythmias seen in other forms of hypertrophic or dilated cardiomyopathy. The myopathic substrate in patients with CHD is generated by long-standing pressure and volume loads, often complicated by cyanosis, that ultimately result in advanced degrees of hypertrophy, fibrosis, and ventricular dilation. Lesions and conditions associated with VT of this type include (1) congenital aortic outflow obstruction, (2) transposition of the great arteries after Mustard or Senning operations where the right ventricle (RV) has been recruited as the systemic ventricle, (3) TOF with advanced ventricular dilation and dysfunction, (4) unrepaired VSD with Eisenmenger physiology, and (5) palliated single ventricle. In general, these are ill patients with poor hemodynamics in whom the index of suspicion for unstable VT may already be quite high. As a consequence, treatment with an implantable cardioverter defibrillator (ICD) is usually the preferred treatment approach,\textsuperscript{65} although ablation may still be considered as secondary therapy as a way to reduce the shock burden from the device. Naturally, there is the potential for the coexistence of these 2 categories of VT in a given patient whenever there is a combination of underlying anatomic substrate and ventricular dysfunction.

Most literature on VT in CHD has centered on TOF. Occasional patients with TOF with relatively slow monomorphic VT may be stable at presentation, but syncope or cardiac arrest is a more typical presenting symptom. The overall incidence of SCD in TOF has been estimated at 0.2% per year of follow-up, but this risk is not linear. Sustained VT and SCD is in fact quite rare in children and adolescents with TOF, but the risk then accelerates to reach as high as 1% per year in adulthood by the fourth or fifth decades of life.\textsuperscript{56–58} Predicting VT and SCD in patients with TOF has been a topic of intense investigation. Numerous studies have now identified risk factors with modest prognostic value,\textsuperscript{59–63} although none provides perfect predictive accuracy. Studies examining the risk for VT and SCD in conditions other than TOF are even more limited by smaller patient numbers, but case reports and clinical series have succeeded in establishing a level of concern in certain lesions, including aortic stenosis, Ebstein anomaly, and others.\textsuperscript{64–66} It should be emphasized that many patients with CHD may require hemodynamic interventions in the course of VT treatment, such as valve replacement, relief of conduit stenosis, closure of residual septal defects, or aggressive medical treatment of congestive failure. Optimal management obviously requires attention to the broad cardiovascular picture and not just the arrhythmia in isolation.

\textbf{Ablation for Macroreentrant VT in TOF}

Mapping and ablation techniques for VT in CHD are similar to those used for other forms of heart disease, with a few important differences related to distorted ventricular geometry and surgical scars that influence reentrant propagation patterns. As previously mentioned for atrial tachycardia ablation, one must also be aware of the possibility for variations in location of the atrioventricular node and His bundle to avoid inadvertent damage.\textsuperscript{37}

The anatomy of TOF and its predilection for macroreentrant VT can be appreciated by examining the heart specimen shown in Figure 5, where the anterior wall of the RV has been removed to reveal the VSD created by anterior displacement of the conal septum that in turn impinges on the outflow tract to create the hallmark pulmonary stenosis. It should be apparent that the conal septum (an isolated band of muscle between the pulmonary valve and the VSD) represents a potential protected corridor for macroreentrant conduction using the VSD as the central obstacle. In addition, if surgery was performed using a nontransannular patch, there is also the potential a second corridor through the small rim of tissue between the outflow tract incision and the pulmonary valve. The outflow incision could also contribute to a third protected conduction zone between the right lateral patch margin and the tricuspid valve annulus. A landmark article from Zeppenfeld et al\textsuperscript{67} confirmed the importance of these 3 corridors in patients with TOF by showing that ablation at the sites could effectively eliminate monomorphic VT.

All contemporary reports of ablation in this condition stress the need for a careful baseline RV substrate map focused on scar areas that correspond to the VSD patch and the RV outflow patch.\textsuperscript{67–69} Assuming patient tolerance and circuit stability,
detailed activation mapping can then be performed during induced VT to outline the propagation pattern (Figure 6). Diastolic potentials with fractionated activity of low amplitude will typically be recorded in relevant conduction corridors, and pacing maneuvers within these corridors should result in a pattern of concealed entrainment with a postpacing interval identical to the tachycardia cycle length. In cases where VT is too rapid for the patient to tolerate, alternate techniques, such as noncontact mapping and pace mapping, have been used effectively to localize macroreentrant circuits in TOF. If it proves difficult to induce and sustain the patient’s clinical VT, an empirical lesion set directed at the 3 common conduction corridors in the RV may be a feasible strategy even when detailed circuit mapping is impossible.

Descriptions of successful catheter ablation for VT in TOF first appeared in the early 1990s. These began as isolated case reports, and have grown to small clinical series. The reported acute success rate for radiofrequency catheter ablation now approaches 90% although VT recurrence or positive follow-up ventricular stimulation studies are still seen in ≈18% of patients (Table 2). Recurrences are not surprising given the dramatic thickness in RV muscle of some patients with TOF that may be hard to penetrate even when using irrigated radiofrequency catheters. The uncertainty surrounding recurrence has dampened enthusiasm for the use of catheter ablation as sole therapy for VT in TOF. This is particularly true for older patients with advanced RV dysfunction from long-standing pulmonary regurgitation, who may still be at risk for a myopathic-type VT even if monomorphic circuits are eliminated. Thus, for most patients with TOF, catheter ablation is primarily used in conjunction with ICD therapy as a way to minimize shock burden.

If catheter ablation is not successful, or if the patient is returning to the operating room for other reasons, surgical VT ablation can be considered. Reports of successful intraoperative VT mapping and ablation have been appearing since 1980, with the most productive ablation sites nearly identical to those reported for transcatheter therapy. The surgical ablation option remains important for patients with TOF because a sizable percentage are now being referred back to the operating room for replacement of regurgitant pulmonary valves. When valve replacement is combined with careful preoperative or intraoperative mapping, strategic cryoablation can be performed by the surgeon to reduce the likelihood of macroreentrant VT. However, the risk for myopathic-type VT might still remain if the patient has residual RV dysfunction, despite valve repair. Most available data suggest that the degree of RV remodeling seen after valve replacement in adult patients with TOF is insufficient to completely remove all VT risk.

Device Therapy in CHD

Pacemakers, ICDs, and cardiac resynchronization therapy (CRT) play an important role in the rhythm management of patients with CHD. The indications and techniques for device implant differ from patients with other forms of heart disease. The major distinctions in CHD relate to the young age/small size of many recipients, complex anatomy that may make transvenous implant difficult or impossible, thromboembolic risks if residual septal defect are present, and a higher lead failure rate that likely arises from the more active lifestyle of these relatively young patients.

CRT in CHD

Natural history and surgical intervention in CHD often leads to conduction delay in the RV because of either direct injury to the right bundle at time of surgery or delayed conduction within scarred regions of RV myocardium. In TOF, for instance, QRS duration relates largely to conduction delay within the RV muscle. The uncertainty surrounding recurrence has dampened enthusiasm for the use of catheter ablation as sole therapy for VT in TOF. This is particularly true for older patients with advanced RV dysfunction from longstanding pulmonary regurgitation, who may still be at risk for a myopathic-type VT even if monomorphic circuits are eliminated. Thus, for most patients with TOF, catheter ablation is primarily used in conjunction with ICD therapy as a way to minimize shock burden. If catheter ablation is not successful, or if the patient is returning to the operating room for other reasons, surgical VT ablation can be considered. Reports of successful intraoperative VT mapping and ablation have been appearing since 1980, with the most productive ablation sites nearly identical to those reported for transcatheter therapy. The surgical ablation option remains important for patients with TOF because a sizable percentage are now being referred back to the operating room for replacement of regurgitant pulmonary valves. When valve replacement is combined with careful preoperative or intraoperative mapping, strategic cryoablation can be performed by the surgeon to reduce the likelihood of macroreentrant VT. However, the risk for myopathic-type VT might still remain if the patient has residual RV dysfunction, despite valve repair. Most available data suggest that the degree of RV remodeling seen after valve replacement in adult patients with TOF is insufficient to completely remove all VT risk.

Table 2. Reported Outcomes for Catheter Ablation of VT in TOF

<table>
<thead>
<tr>
<th>Author</th>
<th>N</th>
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<th>F/U, m</th>
<th>Recur</th>
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<td>30</td>
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<td>Kriebel et al</td>
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<td>Schneider et al</td>
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<td>56</td>
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<td>Gonska et al</td>
<td>16</td>
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<td>16</td>
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<td>Morwood et al</td>
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<td>10/12</td>
<td>46</td>
<td>4/10</td>
</tr>
<tr>
<td>Total</td>
<td>56</td>
<td>50/56 (89%)</td>
<td>9/50 (18%)</td>
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</tbody>
</table>

F/U indicates follow-up duration; m, months; TOF, tetralogy of Fallot; and VT, ventricular tachycardia.
outflow tract rather than the RV body, a site not resynchronized using standard lead configurations. In numerous studies of ischemic and nonischemic cardiomyopathy, patients with right bundle branch block have not responded well to CRT.

Despite theoretical limitations and nonstandard indications, the clinical application of CRT to patients with CHD is showing promise. Biventricular pacing in TOF has been shown to decrease RV and left ventricular dyssynchrony and improve functional status. In 46 patients with more complex forms of CHD, CRT provided functional improvement in the majority, including single ventricle patients. Another study demonstrated similar findings in 71 patients with CHD, with mean change in ejection fraction of +10% and improvement in New York Heart Association status.

Lead placement for CRT in CHD offers anatomic challenges (Figure 7), including limited access to a ventricular chamber in Fontan patients, abnormal coronary venous anatomy, or exclusion of the coronary sinus from the right heart by surgical patches. As a result, many of these cases require epicardial lead placement that is not always easy in patients with multiple prior chest surgeries. Recent trials of transseptal left ventricular leads in patients with structurally normal hearts who fail to respond to standard CRT leads in the coronary sinus has potentially interesting implications for selected patients with CHD, although the safety of this option has yet to be tested for this population.

ICDs in CHD

Although the decision to implant an ICD in a patient with CHD after a cardiac arrest or life-threatening ventricular arrhythmia is not difficult, reliable risk factors to guide decisions for primary prevention remain elusive in this population. Risk stratification in CHD is inherently limited by the low incidence of malignant arrhythmic events, and the presence of an anatomically heterogeneous population that limits prospective analyses with sufficient power to address the issue. Evolving timing and nature of congenital cardiac surgery adds to the patient heterogeneity and alters factors that modify their risk of sudden death.

A recent analysis of 3 registries for several decades and totaling 25,790 adults with CHD found only 171 (0.66%) individuals who died suddenly from proven or presumed arrhythmic causes. In the absence of firm risk factors, the decision to implant a primary prevention ICD is still individualized to a large degree on the basis of electrophysiological, hemodynamic, and surgical factors, weighing the potential for a life-threatening arrhythmia against the documented rate of 25% to 30% for inappropriate therapies and device-related complications. In a group of high-risk patients with tetralogy undergoing primary or secondary prevention, the annual rate of appropriate ICD therapy was 7.7% and 9.8%, respectively (3.3% and 4.2% for polymorphic VT and ventricular fibrillation), suggesting benefit in carefully selected individuals. However, the absence of a control group prevents calculation of the relative and absolute risk reduction afforded by ICDs.

Many patients with CHD have anatomic considerations that impact ICD implantation, including venous occlusion from prior leads, anomalous systemic venous connections, mechanical or surgically repaired atrioventricular valves, and the risk of thromboembolic complications via residual intracardiac shunts. Such considerations often force novel ICD configurations (Figure 8) although lead function in these systems is reduced compared with that of standard transvenous devices. Therefore, newer technology for subcutaneous/leadless ICDs may be a promising option for patients with CHD, but experience is still limited, and instances of inappropriate shocks caused by improper sensing have been reported.

Figure 7. Hybrid lead system combining transvenous and epicardial leads for a patient with heart block after repair of an atrioventricular canal defect, chronic pacing for many years with an epicardial system. Left ventricular function was deteriorating, and the old epicardial atrial lead failed. The pacing system was upgraded with a new transvenous atrial lead and a transvenous lead in the coronary sinus (CS) for cardiac resynchronization therapy pacing. The old epicardial ventricular lead was retained and tunneled up to the new subclavicular pocket.

Figure 8. Epicardial implantable cardioverter defibrillator (ICD) system in a teenager with Ebstein anomaly, in whom a transvenous ICD lead across the newly reconstructed tricuspid valve was avoided.
Conclusions
With increased longevity for patients with CHD, arrhythmia management will continue to be a critical component of their care. Advances continue to be made in our understanding of arrhythmia mechanisms, in surgical techniques that decrease the arrhythmia burden, and in the technology that facilitates ablation and device therapy. Continued expert attention to the needs of this diverse and special population is essential.30,31

Disclosures
Dr Triedman is a consultant for Biosense Webster. The other authors report no conflicts.

References


Key Words: arrhythmias, cardiac | atrial fibrillation | atrial flutter | catheter ablation | Ebstein anomaly | implantable cardioverter-defibrillator | tachycardia, ventricular | tetralogy of Fallot | valvular heart disease, congenital
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