Interventional Electrophysiology in Patients With Congenital Heart Disease

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“The treatment of congenital heart disease is unsatisfactory. As a rule, nothing can be done to improve patients symptomatically; in some instances digitalis may be of help.”

—L. Emmett Holt, MD, 1933

On August 26, 1938, Dr Robert Gross performed the first successful surgery for a congenital heart defect by closing a patent ductus arteriosus in a 7-year-old girl. The event marked the beginning of an interventional approach to congenital heart disease (CHD) that forever banished the sort of pessimism expressed by Dr Holt in his classic pediatric textbook only 5 years earlier.1 Subsequent diagnostic and operative innovations ultimately led to surgical solutions for nearly all anatomic heart defects, allowing the vast majority of infants born with CHD in the modern era to survive into adulthood. However, improved hemodynamic longevity has exposed alternate sources of morbidity and mortality for this population, central among which are cardiac rhythm disorders. In some instances, arrhythmias are intrinsic to the CHD lesion itself, but in most cases, they arise as an unintended consequence of prior corrective surgery whenever patches and suture lines function in conjunction with hypertrophy and fibrosis to create the substrate for reentrant tachycardia. It is fortunate that improved understanding of rhythm abnormalities in CHD has coincided with the emergence of interventional electrophysiological techniques. Catheter ablation, arrhythmia surgery, pacemakers, and implantable defibrillators have now become indispensable treatment options for this group. It is the purpose of this article to review the application of these tools to CHD with an emphasis on unique technical challenges.

Mechanism and Incidence of Arrhythmias in CHD

Congenital heart defects occur in roughly 0.8% of live births, and in 30% to 50% of cases, the malformations are severe enough to warrant ≥1 surgical procedures during early childhood.2 Arrhythmia mechanisms vary according to the underlying anatomic defect and method of surgical repair. The electric pathophysiology involves a complex interplay between gross cardiac anatomy, chamber enlargement from abnormal pressure and volume loads, cellular injury from hypoxia and cardiopulmonary bypass, fibrosis at sites of suture lines and patches, and direct trauma to the specialized conduction tissues. A more likely environment for arrhythmia generation can hardly be imagined. The principal forms of CHD are listed in order of prevalence in the Table, along with an estimate of the relative risk for specific arrhythmias in each condition.

Atrial Tachycardias

The most common arrhythmia mechanism in CHD patients involves a macroreentrant circuit within abnormal atrial muscle. This disorder is conventionally referred to as intra-atrial reentrant tachycardia (IART) to distinguish it from macroreentrant atrial flutter, which occurs in patients with normal anatomy. The P-wave contour during IART often differs from the familiar sawtooth appearance of flutter because the reentrant path does not necessarily propagate around the tricuspid valve ring but instead may rotate around patches, atriotomy incisions, and other atypical conduction obstacles.3–5 Furthermore, IART tends to be slower than flutter, with atrial rates typically in the range of 150 to 250 per minute. This can result in episodes of 1:1 conduction with resultant hypotension or even circulatory collapse in patients with limited myocardial reserve.6 Thromboembolic complications can also arise if episodes are protracted.7

Although IART can develop in the setting of nearly any CHD lesion,8 it is especially problematic after the Fontan operation for single ventricle and the Mustard or Senning operation for transposition of the great arteries (D-TGA). Up to 50% of single-ventricle patients who have undergone older variations of the Fontan procedure will develop IART within a decade of surgery,9,10 as will ≈30% of patients after Mustard or Senning repair.11 Apart from anatomic lesion and type of surgery, other risk factors for IART include concomitant sinus node dysfunction (tachy-brady syndrome) and older age at the time of heart surgery.8,9

Acute termination of IART episodes can be accomplished reliably with standard maneuvers such as electric cardioversion, but the far greater challenge is prevention of recurrence.6 Experience with antiarrhythmic drug therapy in this condition has been discouraging even when class Ic and III...
agents are used; hence, the contemporary approach to IART treatment has shifted largely to interventional procedures.

Although far less common than IART, atrial fibrillation can develop in a subset of CHD patients who have conditions that result in marked left atrial dilation such as un repaired atrial septal defects, left-sided valve disease, and dysfunction of their systemic ventricle.

### Accessory Atrioventricular Pathways

The incidence of accessory pathways in CHD is not appreciably higher than in the general population, with the 2 notable exceptions of Ebstein’s anomaly of the tricuspid valve and “congenitally corrected” transposition of the great arteries (L-TGA) in which an Ebstein’s-like malformation of the systemic atrioventricular (AV) valve often exists. The high incidence of accessory pathways in these 2 conditions suggests a probable developmental link between the valve deformity and the abnormal conduction tissue. As many as 20% of patients with Ebstein’s will have Wolff-Parkinson-White syndrome, and multiple accessory pathways will be found at mapping studies in nearly half of these cases. The situation can be aggravated further by atrial enlargement, leading to IART or atrial fibrillation with the potential for rapid anterograde conduction over the abnormal pathways. Catheter ablation has become the preferred treatment of this condition, although, as discussed later in this review, procedures tend to be more difficult than ablation in a normal heart.

### Ventricular Tachycardia and Sudden Cardiac Death

Sudden arrhythmic death occurs with disturbing frequency in certain forms of CHD as patients reach adolescence and adulthood. Rapidly conducted IART or bradycardia may be the culprits in rare cases, but most of these events appear to involve ventricular tachycardia (VT). Patients at highest risk for VT are those who have undergone a ventriculotomy or patching of a ventricular septal defect (eg, tetralogy of Fallot repair) in which a macroreentrant circuit develops near the region of surgical scar. Alternatively, VT can develop in the setting of a more generalized myopathic process without discrete ventricular scars (eg, chronic aortic valve disease or a failing right ventricle after the Mustard and Senning procedures) if long-standing hemodynamic stress causes advanced degrees of ventricular hypertrophy or global dysfunction.

Most published experience with VT in CHD involves tetralogy of Fallot. The prevalence of VT in this condition has been measured in the range of 3% to 14% in several large series, with a risk for sudden death estimated at 2% per decade of follow-up. Studies seeking risk factors for VT have identified clinical variables with modest predictive value, including: (1) older age at surgery, (2) older age at follow-up, (3) history of prior palliative shunts, (4) high-grade ventricular ectopy on Holter monitoring, (5) inducible VT at electrophysiological study, (6) poor right-heart hemodynamics, and (7) prolonged QRS duration on standard ECG. This list provides good sensitivity but poor specificity for identifying the tetralogy patient at risk.

Similar to the interventional approach that has been adopted for sudden death prevention in ischemic heart disease and cardiomyopathy, treatment for CHD patients with documented or suspected VT has moved away from drug therapy to focus predominantly on implantable defibrillators and ablation procedures. It should perhaps be emphasized that many CHD patients will also require hemodynamic interventions in the course of VT treatment such as valve replacement, relief of conduit stenosis, or closure of residual septal defects. Optimal management obviously requires attention to the broad cardiovascular picture, not just the arrhythmia in isolation.

### Sinus Node Dysfunction and AV Block

Direct injury to the sinus node can occur with surgical incisions or suturing in the high right atrium. The Fontan, Glenn, Mustard, and Senning operations are notorious in this regard. More than 50% of survivors from the Mustard operation, for instance, have lost reliable sinus rhythm by adulthood. The resultant chronotropic incompetence limits exercise capacity, aggravates AV valve regurgitation, and

### Relative Risk for Specific Arrhythmias in Common Congenital Heart Defects

<table>
<thead>
<tr>
<th>Relative Risk for Specific Arrhythmias in Common Congenital Heart Defects</th>
<th>IART</th>
<th>AF</th>
<th>WPW</th>
<th>VT/SCD</th>
<th>SA Node Dysfunction</th>
<th>Spontaneous AV Block</th>
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AF indicates atrial fibrillation; WPW, Wolff-Parkinson-White syndrome; SCD, sudden cardiac death; SA, sinoatrial; VSD, ventricular septal defect; ASD, atrial septal defect; TOF, tetralogy of Fallot; AS, aortic stenosis; M&S, after the Mustard or Senning operation; CAVC, common AV canal defect; SING V (F), single ventricle after the Fontan operation; ++, high risk; +++, moderate risk; and +, slight risk.

Walsh Interventional EP in CHD
contributes to a higher incidence of IART. Pacemaker therapy becomes necessary for many of these patients, although distorted anatomy can make standard transvenous implant challenging.

Disorders of AV conduction can arise in CHD patients from either developmental abnormalities of the conduction tissues or direct trauma. In 2 very specific malformations, AV canal defect and L-TGA, the AV node is anatomically displaced outside the normal triangle of Koch, and functional conduction properties are depressed. About 5% of L-TGA patients will exhibit complete AV block at birth, and a number of others have a progressive decline in conduction over the first 2 decades of life, so about 20% of this group will require pacing for complete block by adulthood. Furthermore, the conduction tissues are very susceptible to injury during catheter and surgical procedures in canal defects and L-TGA. Even in conditions in which the conduction tissues are developmentally normal and properly situated in Koch’s triangle, a small number of patients will suffer traumatic AV block during closure of standard ventricular septal defects, surgery for left-heart outflow obstruction, or AV valve replacement. Surgically induced heart block that is not expected to resolve or persists >7 days after surgery is viewed as a class I indication for permanent pacing.

Catheter Ablation in CHD

Catheter ablation has been applied successfully to most forms of tachycardia in CHD patients. The major challenge during these cases is distorted anatomy that invalidates customary fluoroscopic landmarks and complicates catheter manipulation. Details of the underlying structural disease and all surgery need to be understood in advance of the intervention by reviewing past catheterization and operative data, along with prior noninvasive imaging studies. Angiography or echocardiography can be used to further clarify chamber orientation and to locate the true level of the AV groove at the time of the procedure (Figure 1A through 1C). To minimize the likelihood of inadvertent damage to the normal conduction tissues with ablation, it is also essential to recognize the atypical conduction patterns in certain forms of CHD and to carefully locate a high-quality His potential. Most laboratories performing ablation in CHD patients are now equipped with systems for 3-dimensional mapping, which has improved success rates for complex targets such as IART and VT. In addition, new software permitting 3-dimensional maps to be merged with magnetic resonance or computerized tomography images adds a level of sophistication to anatomic definition during ablation in CHD (Figure 2). Because thickened atrial and ventricular chamber walls are common in this population, irrigated ablation catheters and

Figure 1. Series of angiograms used for orientation during catheter ablation of a left-lateral accessory pathway in a 3-month-old child with complex CHD and intractable tachycardia. The infant had ventricular inversion with L-looped ventricles and Ebstein’s-like malformation of the left AV valve. A, An injection into the right-sided left ventricle (LV) shows the connection to the pulmonary artery (PA). B, A retrograde injection into the left-sided right ventricle (RV) shows severe AV valve regurgitation into a massively dilated left atrium (LA). The mobile edge of the displaced valve can be seen. C, An aortic root (Ao) injection fills the coronary artery (CA) to mark the level of the true AV groove.

Figure 2. A “merge” image showing the segmented anatomy of the right atrium (RA), left atrium (LA), pulmonary artery (PA), and a right-sided aortic arch (Ao) that was imported from a magnetic resonance study in an adult with repaired tetralogy of Fallot. The anatomic shell is combined with a 3-dimensional electroanatomic map of an IART circuit localized to the lateral RA wall. The propagation pattern for IART proceeds according to the color code (red→yellow→green→blue→purple). A series of RF applications along a narrow corridor in the circuit (small red dots) eliminated IART.
large-tip catheters with high-output radiofrequency (RF) generators are often required for effective lesion creation.48

**Catheter Ablation for IART in CHD**

Atrial mapping experience with IART in CHD has uncovered clear patterns for circuit propagation that depend on the underlying anatomy and type of repair.49 As a general rule, IART is restricted to right atrial tissue, and if a tricuspid valve is present, the isthmus between the valve ring and the inferior vena cava is a common component of such circuits.50 Patients with 4 cardiac chambers in normal orientation (eg, atrial septal defect, tetralogy of Fallot) can therefore be expected to have relatively straightforward circuits that resemble common atrial flutter, traveling around the tricuspid valve ring through the cavotricuspid isthmus (Figure 3A). An additional circuit around an atriotomy scar on the lateral right atrium4 also can be present in some of these cases (Figure 3B).

Circuits become more complicated whenever right atrial anatomy is distorted by abnormalities of atrial situs, atresia of an atrioventricular valve and ventricle, and surgical septation that sequesters portions of the right atrium into the pulmonary venous atrium. For example, when caval return is baffled toward the mitral valve as in D-TGA patients who have undergone the Mustard or Senning operation, the cavotricuspid isthmus is still likely to participate in reentry, but most of the critical isthmus tissue is located on the left side of the circulation. The catheter has to be delivered by a retrograde arterial approach or transbaffle puncture to reach this site for ablation.51–54

By far the most complex cases involve patients with a single ventricle after one of the many modifications of the Fontan operation in which the right AV valve and cavotricuspid isthmus are usually absent and the atrial chamber is massively enlarged. Multiple circuits can be present in this setting, with reentry around lateral wall atrial scars (Figure 4), atrial septal patches, or the region of anastomosis between the atrium and the pulmonary artery.3,54,55

The basic technique for IART mapping involves collection of electrograms from one or both atria during sustained tachycardia, incorporating enough data points to establish the activation sequence over the entirety of the IART cycle length. This pattern can then be viewed in the context of surgical scars and natural conduction barriers to arrive at a rational model for macroreentry. Entrainment pacing with analysis of postspacing intervals can be used to decipher complex circuits when uncertainty exists.56

Ablation can be attempted once the route for propagation has been established with reasonable certainty, but even when tachycardia location is firm, the task of creating an effective line of conduction block in CHD patients with thickened and dilated atrial chambers is demanding. Quite often, the target area is wider than the average lesion dimension of a single RF application, so multiple contiguous lesions have to be placed to establish permanent block in the target area. This leaves open the possibility of small gaps between application sites or...
variable depth for RF penetration that fails to produce transmural injury. This limitation has been addressed reasonably well with the widespread adoption of large-tip and irrigated designs for RF catheters.46 The combination of improved mapping with 3-dimensional technology and enhanced lesion creation has improved the acute success rates for IART ablation from ≈60% more than a decade ago to nearly 90% in the present era.47 Un fortunately, recurrence of the original IART or some new circuit remains disappointingly common. About 20% of patients with 4-chamber hearts will experience at least 1 episode of IART within 3 years of ablation, but the recurrence risk is particularly high (nearly 40%) in the Fontan population, who tend to have the largest number of IART circuits and the thickest atrial tissue.57 Although still far from perfect, outcomes are superior to the degree of control obtained with antiarrhythmic drugs alone, in which nearly 70% of patients have breakthrough IART within 2 years of starting a medication.52 Furthermore, even if IART episodes are not eliminated entirely by ablation, the procedure can provide effective palliation by reducing the frequency of episodes and eliminating the need for long-term drug therapy.47

Catheter Ablation of Accessory Pathways in CHD

Most published experience with accessory pathway ablation in CHD has involved Ebstein’s anomaly, in which the abnormal pathways are located almost exclusively in the region of the abnormal tricuspid valve.58,59 It is important to distinguish between the mobile edge of the valve leaflets in Ebstein’s (typically displaced down into the body of the right ventricle) and the true AV groove (marked by the course of the right coronary artery). The latter is the proper level for ablation but can be difficult to identify in this disease by standard fluoroscopy and electrogram characteristics alone (Figure 5). A right coronary angiogram will usually simplify this task and establish a useful roadmap for catheter navigation. Thin-caliber electrode catheters also have been used in some exceptional cases to map the right AV groove from within the lumen of the right coronary artery.60

Dilated right heart structures, fractionated electrograms, and a high incidence of multiple pathways all increase the challenge of accessory pathway ablation in Ebstein’s anomaly. Additionally, limits exist to how aggressive lesion creation can be because instances of coronary artery injury have been reported with standard RF applications along the exceptionally thin right AV groove in this condition.61 Thus, procedural outcomes fall short of what could be expected for a structurally normal heart, with acute success in ≈85% of Ebstein’s cases and pathway recurrence in as many as 25%.

Accessory pathways can be seen sporadically in malformations other than Ebstein’s. The list of associated structural anomalies ranges from simple atrial septal defect to complex heterotaxy with single ventricle. The decision to ablate often is motivated by an impending cardiac operation involving patching or vascular redirection that might block future catheter access to the pathway region. Successful procedures once again demand clear anatomic definition, accurate localization of the AV groove, and knowledge of AV node location.46,59,62 Reported outcomes for non-Ebstein’s CHD patients leave room for improvement, with acute success in only ≈80% and recurrence in ≈15%. Many of the ablation failures involve small patients with complex anatomy and accessory pathways in close proximity to the AV node in whom RF applications pose a risk of heart block. The recent availability of cryocatheter ablation with an enhanced safety margin near the AV node should improve results for such cases.63

Catheter Ablation for VT in CHD

Published experience with catheter ablation for VT in CHD remains limited. Most data exist in the form of single case reports, confirming that monomorphic VT can be induced with programmed stimulation to replicate clinical tachycardia, that macroreentrant circuits can be mapped accurately in conditions like tetralogy, and that VT can be extinguished with RF ablation.64–73 Varied mapping strategies have been described, including activation sequence analysis, entrainment maneuvers, pace mapping, and 3-dimensional registration (Figure 6). Although these data are interesting and important, a case report format cannot account for ablation failures, nor is follow-up sufficiently uniform to examine the issue of VT recurrence. Only 2 small series involving a combined total of 30 subjects have been assembled to address these questions.74,75 Among these subjects, the underlying lesion was tetralogy of Fallot in 17, ventricular septal defect in 8, and miscellaneous CHD in the remaining 5. Acute RF ablation success was achieved during 25 of the 28 procedures when mapping was possible (89%), although later VT recurrence was documented in 5 cases (20%). Some degree of recurrence is not surprising, considering the difficulty with RF lesion creation in the often thick-walled chamber like the right ventricle of tetralogy. The obvious question thus becomes whether catheter ablation can ever function as exclusive therapy for CHD patients when the consequences of VT recurrence could be catastrophic. Carefully selected CHD patients with single circuits of well...
tolerated VT and otherwise good hemodynamics might be viewed as suitable candidates for primary catheter therapy, but if VT is rapid or causes serious symptoms, RF ablation usually is relegated to a secondary role as a way of reducing the shock burden in patients with frequent tachycardia episodes who already have a defibrillator in place.

Surgery for Arrhythmias in CHD
Cardiac surgery can serve either a preventive or a therapeutic role in arrhythmia management for CHD patients. Within the prevention category is a long list of modifications in technique and timing of corrective operations that have significantly reduced the incidence of late arrhythmias. A good example involves D-TGA, which is now managed with a direct arterial switch operation in the newborn period, rendering the complex atrial baffles of the Mustard and Senning operations nearly obsolete. This new approach not only offers hemodynamic advantages by retaining the left ventricle in the systemic circulation but also has effectively abolished the late complications of IART and sinus node dysfunction in this condition.76 Similarly, modern variations in the Fontan operation for single ventricle (such as the “lateral tunnel” and “extracardiac conduit”) now channel systemic venous return directly to the pulmonary arteries via a cavopulmonary connection, thereby averting the massive atrial enlargement seen with the old-style anastomosis between the right atrium and pulmonary circulation. The incidence of postoperative IART for single-ventricle patients has decreased substantially as a consequence.77,78 The risk of ventricular arrhythmias also has been reduced in conditions such as tetralogy of Fallot by performing surgery at younger ages with smaller right ventricular incisions and more attention to pulmonary valve competence.79,80

Therapeutic arrhythmia surgery is used predominantly in the older generation of CHD patients who were repaired with obsoleted techniques. These operations usually have the combined goals of tachycardia elimination and improvement of hemodynamics.

Fontan Conversion and Atrial Maze
Management of IART in the older Fontan population repaired with atriopulmonary connections requires a large arsenal of treatment options. Catheter ablation or pacemaker implantation can be effective in many cases, but if these measures fail or are otherwise deemed unsatisfactory, surgery can be considered that involves conversion to a modern cavopulmonary connection, combined with a modified maze procedure in the right atrium. It is clear from early surgical experience81 that hemodynamic conversion by itself is insufficient to eliminate IART. A formal right-sided maze must be incorporated into the operation, using a lesion set that consists of linear ablation at 3 regions: (1) the upper rim of the atrial septal defect toward the superior vena cava and right atrial appendage, (2) the posterior-inferior rim of the atrial septal defect toward the crista terminalis and edge of the atriotomy incision, and (3) the coronary sinus and right AV valve (when such a valve is present) toward the inferior vena cava.82 Placement of an epicardial atrial pacemaker typically is included for sinus node dysfunction, which will likely be present postoperatively if it was not already an issue preoperatively.83 For occasional Fontan patients who have atrial fibrillation in addition to IART, the maze can be extended to the left atrium with a variation of the Cox procedure.84

Fontan conversion with an atrial maze has a very good record for eliminating atrial tachycardia. Initial estimates of recurrence85 were on the order of 12% but are perhaps even lower now that the proper ablation lesion set has been better defined.86 Still, it is a major operation with a combined risk of mortality or need for postoperative heart transplant that exceeds 5%, and these risks must be balanced carefully against the potential electrophysiological benefit.84

Pulmonary Valve Replacement in Tetralogy of Fallot
Successful relief of right ventricular outflow obstruction during tetralogy repair often requires sacrifice of pulmonary valve competence. Regurgitation can be tolerated for many years, but the resultant volume load can eventually cause right ventricular dysfunction and increase the risk of VT. Many patients with advanced degrees of regurgitation are now undergoing pulmonary valve replacement in an effort to address this issue. Among those known to have reentrant VT preoperatively, simply replacing the valve does not appear to eliminate the underlying VT substrate, but if formal mapping
and surgical cryoablation of circuit tissue are performed, VT can be eliminated in most cases.\(^\text{36}\)

The hemodynamic benefit of valve replacement in tetralogy patients with right-heart dilation seems well established,\(^\text{87–89}\) but the exact risk of VT recurrence after surgical ablation is less clear. Estimates of VT recurrence range between 0% and 30% in published series.\(^\text{36,90,91}\) Whether the intervention can ever dismiss consideration of an implantable defibrillator in high-risk cases remains uncertain, but like catheter ablation, it could serve the role of reducing defibrillator shocks in patients who already have a device in place. Studies are now underway to test whether earlier valve replacement in tetralogy (ie, before the appearance of significant right ventricular failure) will prevent development of late VT.\(^\text{92,93}\)

Pacemaker and Implantable Defibrillator Therapy in CHD Patients

Indications for pacemaker and defibrillator implantation in CHD patients have been reviewed and updated as part of the guidelines published by the American College of Cardiology/American Heart Association/North American Society for Pacing and Electrophysiology task force.\(^\text{94}\) The most important considerations surrounding device therapy in this population relate to lead placement, which must take into account distorted anatomy and the risk for thromboembolic complications.

Lead Placement

Transvenous access for pacemaker or defibrillator leads in CHD often is difficult. The challenge is well demonstrated by patients after the Fontan operation, who usually lack a standard venous connection to their single ventricle and may be at risk for large thrombus formation on atrial leads unless chronically anticoagulated. Although transvenous atrial lead placement is possible in select cases, an epicardial approach typically is used for ventricular leads. Transvenous ventricular pacing can sometimes be accomplished with a lead advanced into a ventricular branch off the coronary sinus in patients with the old variety of atriopulmonary Fontan anastomosis, but this option is not available for those with modern cavopulmonary connections.\(^\text{95,96}\)

The venous route to the atrium and ventricle in Mustard or Senning patients is likewise challenging. Transvenous leads need to be advanced through the atrial baffle before fixation in the modified right atrium and the anatomic left ventricle, and it is not uncommon to encounter tight obstruction along the upper limb of the baffle that requires dilation and stent placement before leads can be passed. Baffle leaks with right-to-left shunting also might be present that necessitate closure with an occlusion device or surgery before a lead is inserted into the circulation.\(^\text{97}\) Finally, distorted atrial anatomy must be understood clearly to allow atrial lead fixation in the few areas of atrial muscle that are away from patches, scars, and the left phrenic nerve (Figure 7A through 7D).

The incidence of thromboembolic complications is significant when transvenous leads are used in CHD patients with residual intracardiac shunting from patch leaks or open septal defects.\(^\text{98}\) If such defects cannot be closed in advance of lead implantation, an epicardial approach may have to be considered.

The need for epicardial pacemaker or defibrillator leads can sometimes be anticipated during a primary cardiac repair, in which case the cardiac surface is easily visualized in the scar-free mediastinum. Modern bipolar epicardial leads with steroid-eluting features function nearly as well as endocardial leads under these circumstances.\(^\text{99}\) Unfortunately, many epicardial implantations have to be done in older CHD patients who have undergone prior operations, which presents the surgeon with a scarred mediastinum that must be dissected carefully and deliberately to find suitable lead attachment points. It is much more of a chore to identify sites with adequate sensing function and capture thresholds when dense adhesions cover the heart, and long-term lead performance can suffer as a result.

Resynchronization Pacing in CHD

The optimistic results reported with biventricular resynchronization pacing in patients with normal cardiac anatomy and dilated myopathy have inspired parallel trials in the CHD population. The experience to date has been encouraging. In
a multicenter review of 73 patients with CHD undergoing resynchronization therapy,\textsuperscript{100} a mean increase in systemic ventricular ejection fraction of 12% was observed, along with a mean decrease in QRS duration of 39 milliseconds. The degree of improvement was similar with endocardial and epicardial leads. A good response did not seem to hinge on whether a left or right ventricle was the systemic pumping chamber, although patients with a single ventricle were unlikely to improve in this series despite shortening of the QRS width.

The concept of resynchronization also has also extended to CHD patients with right bundle-branch block and a failing right ventricle, as can occur in the older population with repaired tetralogy of Fallot. Selective placement of a ventricular pacing lead to a site that offsets right ventricular conduction delay seems to enhance contractile performance of the right ventricle,\textsuperscript{101,102} although long-term experience with this technique is limited.

**Atrial Antitachycardia Pacing in CHD**

The high incidence of tachy-brady syndrome in CHD has prompted trials of pacemakers equipped with automatic features to detect IART and attempt interruption with burst atrial pacing.\textsuperscript{103} In patients with a single circuit of relatively slow IART, this technology tends to be quite successful. However, difficulty may arise in those with multiple IART circuits because burst pacing can shift tachycardia to different cycle lengths or cause degeneration to atrial fibrillation, sometimes with a faster ventricular response and worsened symptoms.\textsuperscript{104} Overall, automatic antitachycardia pacing can be successful therapy for IART in \textasciitilde50% of patients.\textsuperscript{105,106}

**Implantable Defibrillators in CHD**

Implantable defibrillators are being used with increasing frequency in the CHD population.\textsuperscript{35,107} The most common CHD diagnosis among defibrillator recipients is tetralogy of Fallot, followed by D-TGA and aortic stenosis. Transvenous dual-chamber systems are possible in most cases, except for patients with significant intracardiac shunts, a single ventricle after the Fontan operation, or very small body size. Numerous configurations of coils and patches have been described for shock delivery (Figure 8) when epicardial implantation is the only option.\textsuperscript{108,109}

Defibrillation thresholds measured in CHD patients with either transvenous or epicardial systems are similar to those obtained in patients with conventional cardiac anatomy. Long-term data on defibrillator performance suggest appropriate shock therapy in \textasciitilde30% of CHD patients over 5 years of follow-up,\textsuperscript{35} with inappropriate shocks in 25% (mostly for sinus tachycardia) and lead failure (requiring revision) in 21%. The somewhat-high lead failure rate may reflect the vigorous lifestyle enjoyed by most adolescents and young adults with CHD compared with the older population with ischemic disease.

**Conclusions**

Older CHD patients who underwent repair or palliation with antiquated surgical techniques suffer from a wide variety of rhythm disturbances. Interventional electrophysiological techniques play a major role in their management. Although procedures often are complicated by atypical anatomy and the presence of intracardiac shunting, a high level of success can be achieved with careful attention to surgical history and modern imaging technology. Modifications in surgical technique have done much to reduce rhythm complications for the current generation of CHD patients.

**Disclosures**

None.

**References**


Key Words: arrhythmia • atrial flutter • catheter ablation • heart defects, congenital • pacemakers • Wolff-Parkinson-White syndrome