By the time patients with congenital heart disease (CHD) reach adolescence and early adulthood, rhythm status has often become an active issue, if not the central issue, in their cardiac management. For some, arrhythmias are intrinsic to the structural malformation itself, as is the case with Wolff-Parkinson-White syndrome in the setting of Ebstein’s anomaly, or atrioventricular (AV) block in the setting of "congenitally corrected" transposition of the great arteries (L-TGA). For most other CHD patients, arrhythmias represent an acquired condition related to the unique myocardial substrate created by surgical scars in conjunction with cyanosis and abnormal pressure/volume loads of long duration. This review will attempt to address the major rhythm difficulties faced by adults with CHD, concentrating on electrophysiological features that distinguish CHD from more conventional forms of adult heart disease. A listing of specific physiological features that distinguish CHD from more conventional forms of adult heart disease. A listing of specific physiological features that distinguish CHD from more conventional forms of adult heart disease.

Prevalence

As of 2001, there were estimated to be 800,000 adults with CHD living in the United States, a number that has grown steadily as more survivors of childhood surgery reach maturity. Of these, roughly 45% are considered to have mild forms of CHD (eg, atrial septal defect, valvar pulmonary stenosis), 40% are classified as having moderate disease (eg, tetralogy of Fallot, Ebstein’s anomaly), and 15% are considered to have complex disease (eg, single ventricle, transposition of the great arteries). Although arrhythmias can develop within any of the 3 subgroups, the incidence is highest for patients in the moderate and severe categories. Tetralogy of Fallot is an instructive example of moderate CHD with a large arrhythmia burden. As many as one third of patients with repaired tetralogy develop symptomatic atrial tachycardias by adulthood, ~10% develop high-grade ventricular arrhythmias, and ~5% require pacemaker implantation for surgically acquired AV block or sinus node dysfunction. In addition, a small but increasing number are now receiving implantable cardioverter defibrillators (ICDs) for treatment of ventricular tachyarrhythmias. Clearly, serious arrhythmias develop in a sizable segment of the population with repaired tetralogy, all of which contribute to a sudden death risk that is currently measured at ~2% per decade of follow-up. Even when using conservative population assumptions, this translates on a national level to more than 50,000 adult patients with tetralogy who require close electrophysiological attention, and perhaps as many as 100 sudden arrhythmic deaths per year if patients were left untreated. Although the magnitude of this sudden death risk may seem small compared with the threat faced by older patients with ischemic heart disease, the emotional impact of unexpected death in a young adult who has struggled with CHD throughout his or her abbreviated lifetime is nonetheless profound.

The situation is equally concerning for patients falling into the category of severe CHD. The Fontan operation for single ventricle, as well as the atrial switch operations (Mustard or Senning) for transposition of the great arteries, results in extensive suture lines and abnormal hemodynamics that predispose patients to atrial tachycardias and sinus node dysfunction. Up to 50% of patients with single ventricle who have undergone older-style Fontan operations will develop atrial tachycardia within a decade of surgery, and hardly any transposition patients remain in reliable sinus rhythm 10 years after Mustard or Senning repairs. These patients pose challenges in terms of both rhythm control and prevention of thrombembolic complications. Any center with a sizable adult CHD program is now likely to be involved with ongoing care for hundreds of such cases.

The rhythm prognosis for older CHD patients is sobering, but in some ways, it might be viewed as an indirect triumph for pediatric cardiology and cardiac surgery. Patients who in past years would have perished during infancy are now surviving thanks to accurate anatomic diagnosis and early surgical intervention. Late-onset arrhythmias in adulthood appear to have become the price to pay for decades of effective hemodynamic palliation, and fortunately, treatment options for the arrhythmias themselves are improving rapidly to meet the emerging demand. Perhaps the most optimistic facet of the arrhythmia experience in CHD involves modern refinements in surgical strategy that have arisen in response to dissatisfaction with earlier electrophysiological outcomes. Nowadays, transposition is managed with direct arterial switch rather than large atrial baffles, Fontan connections are constructed in such a way as to minimize right atrial dilation,

From the Electrophysiology Division, Department of Cardiology, Children’s Hospital Boston, and Department of Pediatrics, Harvard Medical School, Boston, Mass.
Correspondence to Edward P. Walsh, MD, Cardiac Electrophysiology, Children’s Hospital Boston, 300 Longwood Ave, Boston MA 02115. E-mail epwalsh@cardio.tch.harvard.edu
(Circulation. 2007;115:534-545.)
© 2007 American Heart Association, Inc.
Circulation is available at http://www.circulationaha.org
DOI: 10.1161/CIRCULATIONAHA.105.592410
and tetralogy of Fallot is being repaired at younger ages with smaller right ventricular (RV) incisions and attempts to preserve competence of the pulmonary valve. Modifications of this sort have resulted in a promising reduction in the incidence of late arrhythmias11–14 for the younger generation of CHD patients.

Tachycardias in Adults With CHD

Accessory Pathways and Twin AV Nodes

The embryological accidents responsible for congenital heart defects can have a direct impact on development of the conduction system. Sometimes this takes the form of simple displacement of the AV node and His bundle away from the usual septal position in Koch’s triangle,15 but occasionally, the malformation results in accessory or duplicated AV connections with the potential for reentrant tachyarrhythmias. The most familiar example involves Ebstein’s anomaly of the tricuspid valve, which is complicated by Wolff-Parkinson-White syndrome in ~20% of cases.16 The accessory pathways in Ebstein’s anomaly are typically located along the posterior and septal aspect of the tricuspid ring where the valve leaflets are most abnormal,17 and nearly half of these patients will be found to have multiple accessory pathways.18,19 The same observations hold true for patients with L-TGA, who not infrequently have Ebstein’s malformation in association with accessory pathways along their left-sided tricuspid valve.

Tachycardia events for Ebstein’s patients become increasingly problematic in adolescent and adult years when atrial dilation increases the likelihood of recurrent atrial flutter or atrial fibrillation with potentially rapid anterograde conduction over the accessory pathway(s). Definitive therapy with catheter ablation is now viewed as the standard of care for Ebstein’s patients with Wolff-Parkinson-White syndrome. However, compared with ablation for simple accessory pathways in a structurally normal heart, it must be recognized that the short-term success rate appears lower, and the risk of recurrence higher, in Ebstein’s anomaly.20,21 These differences result from the challenges of distorted anatomic landmarks, difficulty identifying the true AV groove,22 extremely fractionated electrograms, and the high incidence of multiple pathways.

Table 1. Specific Arrhythmias and Associated Defects in Adults With Congenital Heart Disease

<table>
<thead>
<tr>
<th>Arrhythmias</th>
<th>Associated Defects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tachycardias</td>
<td></td>
</tr>
<tr>
<td>Accessory pathways</td>
<td>Ebstein’s anomaly; L-TGA</td>
</tr>
<tr>
<td>Twin AV nodes</td>
<td>Heterotaxy syndrome</td>
</tr>
<tr>
<td>Intra-atrial reentrant tachycardia</td>
<td>Postoperative Mustard; postoperative Senning; postoperative Fontan; others</td>
</tr>
<tr>
<td>Atrial fibrillation</td>
<td>Mitral valve disease; aortic stenosis; un repaired single ventricle</td>
</tr>
<tr>
<td>Ventricular tachycardia</td>
<td>Tetralogy of Fallot; congenital aortic stenosis; others</td>
</tr>
<tr>
<td>Bradycardias</td>
<td></td>
</tr>
<tr>
<td>Congenital sinus node dysfunction</td>
<td>Heterotaxy syndrome</td>
</tr>
<tr>
<td>Acquired sinus node dysfunction</td>
<td>Postoperative Mustard; postoperative Senning; postoperative Fontan; postoperative Glenn; others</td>
</tr>
<tr>
<td>Congenital AV block</td>
<td>Endocardial cushion defects; L-TGA</td>
</tr>
<tr>
<td>Acquired AV block</td>
<td>VSD closure; sub aortic stenosis relief; AV valve replacement</td>
</tr>
</tbody>
</table>

VSD indicates ventricular septal defect.

Figure 1. Representation of twin AV nodes with a Mönckeberg sling. The cardiac anatomy in this sketch includes a large septal defect in the AV canal region, shown in a right anterior oblique projection. Both an anterior and posterior AV nodal node are depicted (each with its own His bundle) along with a connecting “sling” between the 2 systems. This conduction arrangement can produce 2 distinct non preexcited QRS morphologies (depending on which AV node is engaged earliest by the atrial activation wave front), and a variety of reentrant tachycardias.
A more exotic type of redundant AV connection can occasionally be identified in patients with a single ventricle of the heterotaxy variety, when there is a specific combination of anatomic defects involving discordant AV alignment and a large septal defect in the AV canal region. In this situation, patients can actually have 2 separate AV nodes (so-called twin AV nodes) with 2 discrete His bundles, often with evidence of a connecting fiber between these 2 systems (so-called Mönckeberg sling). A wide variety of reentrant tachycardias can arise within this complicated network (Figure 1), most of which can be eliminated by strategic ablation of 1 limb of the duplicated system. Although twin AV nodes are very rare, the condition epitomizes the peculiar conduction arrangements that are possible in CHD.

**Intra-Atrial Reentrant Tachycardia (Atrial Flutter)**

The most common mechanism for symptomatic tachycardia in the adult CHD population is macroreentry within atrial muscle. The terms “intra-atrial reentrant tachycardia” (IART) and “incisional tachycardia” have become customary labels for this arrhythmia in order to distinguish it from the typical variety of atrial flutter that occurs in structurally normal hearts. Generally, IART tends to be slower than typical flutter, with atrial rates in the range of 150 to 250 per minute. In the setting of a healthy AV node, such rates will frequently conduct in a rapid 1:1 A:V pattern that can result in hypotension, syncope, or possibly cardiac arrest. Even if the ventricular response rate is safely titrated, sustained IART can cause debilitating symptoms in some patients from loss of AV synchrony and may contribute to thromboembolic complications when the duration is protracted.

Usually, IART appears many years after operations that involved an atriotomy or other surgical manipulation of right atrial tissue. It can follow simple procedures such as closure of an atrial septal defect on occasion, but the incidence is highest among patients with advanced dilation, thickening, and scarring of their right atrium. Other risk factors for IART include concomitant sinus node dysfunction (tachy-brady syndrome) and older age at time of heart surgery. Hence, IART is especially problematic for older patients who have undergone the Mustard, Senning, or older-style Fontan operations, in which extensive suture lines and long-term hemodynamic stress result in markedly abnormal atrial myocardium.

The route of propagation for an IART circuit varies according to the anatomic defect and type of surgical repair. It is usually restricted to right atrial tissue (regardless of the surgical destination of this tissue) and is modulated by regions of fibrosis from suture lines or patches, which function in combination with natural conduction barriers to channel the wave front along a macroreentrant loop. If a tricuspid valve is present, the isthmus between the valve ring and the inferior vena cava is a common component of such circuits, but when the tricuspid valve is absent or otherwise deformed, the circuits follow less predictable paths that can only be deciphered by formal electrophysiological mapping (Figure 2). Quite often, multiple IART circuits can be present in the same patient.

**Figure 2.** Electroanatomic map of an IART circuit involving the anterolateral surface of the right atrium in a patient with a previous Fontan operation (cavopulmonary connection). A detailed anatomic shell was generated for the ablation procedure by merging high-resolution computed tomography data with real-time data gathered from the 3D mapping catheter. The propagation pattern for the IART circuit is shown by the black arrows and is also reflected by the color scheme (red > yellow > green > blue > purple). The critical component of the circuit appeared to be a narrow conduction channel through a region of scar (central gray area). A cluster of radiofrequency applications (maroon dots) was placed at the entrance zone to this narrow channel and permanently eliminated this IART circuit. IVC indicates inferior vena cava; LPA, left pulmonary artery; RPA, right pulmonary artery; SVC, superior vena cava; RA, right atrium; JXN, junction; and LAT, lateral.

The clinical diagnosis of IART can usually be made from the standard ECG. Uninitiated clinicians may occasionally misinterpret IART with 2:1 or 3:1 conduction as sinus rhythm if atrial activity happens to be obscured by the QRS and T-wave (Figure 3). The index of suspicion for IART should always be high in older CHD patients; if there is ever uncertainty about the underlying rhythm, vagal maneuvers ought to be performed in an effort to uncover hidden P waves. Once recognized, IART can be reliably terminated with electrical cardioversion, overdrive pacing maneuvers, or administration of certain class I or class III antiarrhythmic drugs. The far more difficult task is prevention of recurrence. Multiple strategies have been developed for IART prevention, all of which can have value in selected patients, but none of which represents the universal solution. If the episodes are infrequent, well tolerated, and recognized promptly, it may be sufficient to rely on periodic cardioversion before embarking on more involved therapy. However, if IART episodes become frequent, cause significant symptoms, or are associated with atrial thrombus formation, aggressive treatment is indicated.

The therapeutic options for IART include (1) antiarrhythmic drugs, (2) pacemaker implantation to correct bradycardia and/or provide automatic atrial antitachycardia pacing, (3) catheter ablation, and (4) surgical intervention with a modi-
fied atrial maze operation. The choice must be tailored to the hemodynamic and electrophysiological status of the individual patient. Chronic antiarrhythmic drugs are still prescribed in some cases, but the broad experience with pharmacological therapy for this condition has been discouraging, even when potent agents such as amiodarone are used. Pacemaker implantation may be reasonable for patients with a picture of tachy-brady syndrome, because correcting slow atrial rates to the physiological range often results in reduction of IART frequency. Pacemakers with advanced programming features that incorporate atrial tachycardia detection and automatic burst pacing to interrupt reentry may also be beneficial in select cases.

Catheter ablation is now used at many centers as an early intervention for IART. The technique has evolved rapidly since the introduction of 3D mapping for improved circuit localization and irrigated-tip or large-tip ablation catheters for more effective lesion creation. When these technologies are combined with good anatomic definition and traditional electrophysiological mapping maneuvers, short-term success rates of nearly 90% can be achieved. Unfortunately, later tachycardia recurrence is still disappointingly common. The recurrence risk is particularly high (nearly 40%) among Fontan patients, who tend to have the largest number of IART circuits and the thickest/largest atrial dimensions. Although far from perfect, ablation outcomes for IART are likely to improve with continued experience and even now are far superior to the degree of control obtained with medications alone. Furthermore, even if IART episodes are not eliminated entirely by ablation, the procedure can often provide substantial improvement by reducing the frequency of episodes and eliminating the need for ongoing drug therapy.

If the above measures fail to prevent IART, or if the patient is returning to the operating room for hemodynamic reasons, consideration should be given to surgical ablation during a right atrial maze operation. This procedure is used most often for the Fontan population with the most refractory variety of IART and is usually combined with revision of the Fontan connection from an older atriopulmonary anastomosis to a modern cavopulmonary connection in the same setting. Results are encouraging, with relatively low rates of IART recurrence (≈12%), but surgical risks must be weighed against the electrophysiological benefit. A prospective trial aimed at comparing various IART management options would be welcome, particularly one that could address the much debated issue of catheter ablation versus surgical maze in Fontan patients.

**Figure 3.** ECG (simultaneous leads I, aVf, and V1) from a 49-year-old woman who had undergone uncomplicated closure of an atrial septal defect many years prior, now presenting with a 6-day history of intermittent palpitations. A, Regular rhythm with a ventricular rate of 118 bpm is seen, which at casual first glance could be mistaken for sinus tachycardia. After a Val-salva maneuver (B), the conduction pattern became more irregular, which allowed clearer demonstration of the underlying rapid atrial activity (arrows in lead V1) at a rate of ≈236 bpm. The diagnosis of IART can sometimes be difficult during periods of 2:1 conduction unless one is attuned to its high probability in CHD patients.

**Figure 4.** Macroreentrant VT in tetralogy of Fallot. A, An autopsy specimen of repaired tetralogy with the anterior RV surface opened to reveal the ventricular septal defect (VSD) patch and the patch-augmented RV outflow tract (RVOT; the outflow patch in this case is transannular). A hypothetical reentry circuit is traced onto this image (black arrows), with the superior portion of the loop traveling through the conal septum (upper rim of the VSD). B, Actual electroanatomic map of sustained VT from an adult tetralogy patient, showing a nearly identical circuit. The propagation pattern is shown by the black arrows and is reflected by the color scheme (red > yellow > green > blue > purple). A narrow conduction channel was found between the rightward edge of the outflow patch scar (gray area) and the superior rim of the tricuspid valve. A cluster of radiofrequency applications at this site (pink dots) closed off the channel and permanently eliminated this VT circuit. LV indicates left ventricle; MPA, main pulmonary artery; and TV, tricuspid valve.
Atrial Fibrillation

The principle hemodynamic derangement and site of surgical scarring in CHD tends to involve right heart structures, so that IART arising from the right atrium is far and away the most common form of atrial tachycardia. However, chronic hemodynamic stress is directed toward the left atrium in a subset of CHD patients, and atrial fibrillation may occur as a result. The CHD lesions commonly associated with atrial fibrillation include aortic stenosis, mitral valve deformities, and unpaired single ventricle. Management principles are similar to atrial tachycardia encountered in other forms of adult heart disease, beginning with medical therapy for anticoagulation and ventricular rate control, followed by electrical or medical cardioversion. As with IART, terminating an isolated atrial fibrillation episode in CHD is not difficult, but prevention of recurrence remains a challenge. Antiarrhythmic drugs may offer long-term protection against recurrence for some patients, but like IART, pharmacological therapy has been only marginally successful for this purpose. Pacemaker implantation may reduce atrial fibrillation recurrences in patients with sinus node dysfunction when fibrillation is part of the tachy-brady syndrome. Definitive elimination of atrial fibrillation can be achieved with a combined right and left atrial maze operation, which should be considered if a patient requires surgery to address other hemodynamic issues.

Ventricular Tachycardia

Serious ventricular arrhythmias are rare among CHD patients during their first decade or 2 of life, but once adulthood is reached, the potential for ventricular tachycardia (VT) and sudden death becomes a looming concern in select cases. Patients at greatest risk for developing VT appear to be those who have undergone a ventriculotomy and/or patching for certain types of ventricular septal defects. In this scenario, the mechanism for VT is reminiscent of the macroreentrant circuits described earlier for IART, involving narrow conduction corridors defined by regions of surgical scar in conjunction with natural conduction barriers such as the rim of a septal defect or edge of a valve annulus (Figure 4). Less commonly, ventricular arrhythmias can develop independently of direct surgical scarring whenever a long-standing hemodynamic overload causes advanced degrees of ventricle dysfunction or hypertrophy. Examples of CHD lesions that can eventually lead to this myopathic variety of VT include (1) aortic valve disease, (2) L-TGA when the RV has been recruited as the systemic ventricle, (3) severe Ebstein's anomaly, (4) certain forms of single ventricle, (5) Eisenmenger's syndrome, and (6) unrepaired tetralogy of Fallot.

The bulk of literature and clinical experience regarding VT in CHD has centered on tetralogy of Fallot. The prevalence of VT after tetralogy repair has been estimated to be between 3% and 14% in several large clinical series. Some patients with slow VT may be hemodynamically stable at presentation, but VT tends to be rapid for the majority, causing syncope or cardiac arrest as the presenting symptom. Even though rare cases of abrupt AV block or rapidly conducted IART have been linked to catastrophic outcomes in tetralogy, sustained VT appears to be the single biggest contributor to the 2% per decade incidence of sudden cardiac death (Table 2).

Predicting VT events in tetralogy patients has been a topic of intense investigation for nearly 30 years. Selected articles from the long list of studies seeking risk factors are summarized in Table 3. To date, no perfect risk-stratification scheme has emerged, although several clinical variables with modest prognostic value have been identified, including (1) older age at time of definitive surgery, (2) history of palliative shunts, (3) high-grade ventricular ectopy, (4) inducible VT at electrophysiological study, (5) abnormal RV hemodynamics, and (6) wide QRS width (>180 ms). The recently appreciated correlation between QRS duration and VT is not surprising when one considers that the most dramatic degree of QRS prolongation tends to be seen among tetralogy patients with highly dysfunctional and dilated RVs (so-called mechanoelectric interaction). When viewed in the aggregate, this long list of variables helps to define a clinical profile for the tetralogy patient at risk, but no single item can be viewed as completely independent, and none provides perfect predictive accuracy.

If this generalized high-risk profile were applied to any contemporary population of adult tetralogy patients, the number who fit the mold could be impossibly large. To compensate for lack of specificity, the practical approach to VT risk stratification in older tetralogy patients usually incorporates attention to symptom status. Obviously, any patient who has survived a cardiac arrest or sustained VT is treated aggressively, usually with an ICD. But, in the absence of a serious clinical event, careful inquiry for more subtle symptoms is often relied on to determine whether additional testing or treatment is needed. At most centers, tetralogy patients who report concerning symptoms of palpitations, dizziness, or syncope usually undergo invasive eval-

### TABLE 2. Estimates of the Incidence of Sudden Death After Tetralogy of Fallot Surgery

<table>
<thead>
<tr>
<th>Study</th>
<th>Findings</th>
<th>Incidence per Decade, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Murphy et al</td>
<td>6% of 163 cases followed up for 30 years</td>
<td>2.0</td>
</tr>
<tr>
<td>Nollert et al</td>
<td>3% of 490 cases followed up for 25 years</td>
<td>1.2</td>
</tr>
<tr>
<td>Silka et al</td>
<td>~2 Deaths per 1000 patient-years</td>
<td>2.0</td>
</tr>
<tr>
<td>Norgaard et al</td>
<td>5.6% of 125 cases followed up for 25 years</td>
<td>2.2</td>
</tr>
<tr>
<td>Gatzoulis et al</td>
<td>6% of 793 cases followed up for 21 years</td>
<td>3.0</td>
</tr>
</tbody>
</table>

*References 2, 3, 6, 14, 48, 49, 54–58, and 61–84.
uation with hemodynamic catheterization and electrophysiology study. Programmed ventricular stimulation provides reasonably good predictive information on the risk of future clinical VT events. A positive study may prompt implantation of a primary prevention ICD, or if monomorphic VT can be induced and is tolerated long enough to permit mapping, catheter ablation of the VT circuit might be considered. An electrophysiology study might also uncover IART as a contributing or confounding factor for a patient’s symptoms, which could be addressed with ablation at the same setting. Correctable hemodynamic issues may also be identified at catheterization that could shift therapy toward a surgical solution, such as relief of valve regurgitation combined with formal intraoperative VT mapping and ablation.

The proper approach to an entirely asymptomatic adult with repaired tetralogy remains unsettled. Most clinicians rely on a yearly evaluation with history and ECG, supplemented regularly with Holter monitoring or exercise testing to screen for high-grade ventricular ectopy, along with periodic echocardiography or magnetic resonance imaging to monitor the

<table>
<thead>
<tr>
<th>Study</th>
<th>Age at Repair</th>
<th>Prior Shunt</th>
<th>VEA</th>
<th>EPS</th>
<th>PS</th>
<th>PR</th>
<th>RV Function</th>
<th>LV Function</th>
<th>ECG</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gillette et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Garson et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Horowitz et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Deanfield et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Garson et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kugler et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kavey et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dunnigan et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kobayashi et al</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Burns et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Deanfield et al</td>
<td>Yes</td>
<td>No</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Garson et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Deal et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Walsh et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chandar et al</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Zimmerman et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Downar et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Murphy et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cullen et al</td>
<td>Yes</td>
<td>No</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Jonsson et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Roos-Hesselink et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gatzoulis et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Balaji et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nollert et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Harrison et al</td>
<td>No</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Berul et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Daliento et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lucron et al</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gatzoulis et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Therrien et al</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hamada et al</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ghai et al</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dore et al</td>
<td>Yes</td>
<td></td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Russo et al</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Yes indicates study supports variable as predictive of malignant arrhythmias; No, study refutes variable as predictive of malignant arrhythmias; Age Repair, older age at time of definitive surgical correction; Prior Shunt, history of prior palliative shunt surgery; VEA, spontaneous high-grade ventricular ectopic activity on ECG or Holter monitoring; EPS, positive ventricular stimulation at electrophysiology study; PS, residual pulmonary stenosis or other outflow obstruction; PR, pulmonary regurgitation; LV, left ventricular; and ECG, ECG findings (QRS duration, QT and JT dispersion).
status of the RV. Should nonsustained VT be detected on surveillance monitoring in an asymptomatic patient, or should RV function appear to be deteriorating, opinions still vary widely as to the appropriate response. Some clinicians would advocate electrophysiology study to refine the arrhythmia risk, some would recommend surgery for pulmonary valve replacement, some would prescribe antiarrhythmic drugs, some would implant a primary prevention ICD, and some might refrain from any treatment as long as the patient remains symptom free. Therapy continues to be individualized for asymptomatic patients depending largely on institutional experience and philosophy.

The lack of objective guidelines for VT prediction and treatment in CHD patients is frustrating, but it would be hazardous to adopt a dogmatic stance based on the data at hand. With few exceptions, studies have been limited to single-center investigations with limited statistical power. Furthermore, because the event rate for sustained VT and sudden death in CHD is low compared with conditions such as ischemic heart disease, the duration of prospective follow-up necessary to answer questions in the CHD field would likely have to extend beyond 10 years. Now that the population of adults with CHD at risk for VT has reached such a substantial size, the opportunity may finally have arrived for an organized assessment of VT management. The ideal design for investigations of this type must involve close collaboration between pediatric and adult CHD programs to ensure continuity of data collection. It is incumbent on interested clinicians to design and cooperate in long-term multicenter studies of these arrhythmias.

**Bradyarrhythmias in Adults With CHD**

**Sinoatrial Node Dysfunction in CHD**

Developmental defects involving the caval-atrial junction can be associated with atypical anatomy and function for the sinoatrial node. This issue is most relevant to complex forms of heterotaxy syndrome in patients with a single ventricle. In the asplenia variety of heterotaxy, bilateral superior caval veins often exist, each with its own sinoatrial node, which results in an interesting ECG pattern of fluctuation between 2 discrete P waves at physiological rates. Apart from the usual ECG, the coexistence of 2 sinus nodes is of minimal clinical consequence. In contrast, patients with the polysplenia type of heterotaxy may lack a true sinus node altogether, which makes atrial depolarization dependent on slower atrial or junctional escape rhythms. Most patients with this rare condition will ultimately require pacemaker implantation.

A more common cause of sinus bradycardia in adults with CHD is surgical trauma to the sinoatrial node or its artery, as may occur during the Mustard, Senning, Glenn, and Fontan operations. Chronotropic incompetence is poorly tolerated in CHD patients with compromised hemodynamics, especially those with a single ventricle or AV valve regurgitation. The likelihood of a patient developing IART or atrial fibrillation is also increased significantly in this setting.

Implantation of an AAIR or DDR pacing system is currently recommended as a class I indication for any patient with CHD and sinoatrial node dysfunction who has symptoms directly attributable to slow heart rate. Pacemaker implantation is also advised as a class IIb indication for CHD patients with resting rates of <40 bpm or sinus pauses in excess of 3 seconds, even in the absence of symptoms. As will be discussed, decisions regarding device implantation cannot be made casually in CHD patients because of the need for epicardial implants and distorted venous pathways in many cases.

**AV Block in CHD**

The AV conduction tissues may be congenitally abnormal in terms of both their location and function in specific forms of CHD, most notably L-TGA and endocardial cushion defects. In the former condition, the AV node and His bundle are displaced in an anterior direction away from the usual position in Koch’s triangle, whereas in the latter, the AV node and His bundle are displaced posterior to Koch’s triangle. The functional properties of these displaced conduction systems are often abnormal. In L-TGA, it is estimated that 3% to 5% of patients will have complete AV block at birth, and an additional 20% will develop spontaneous complete block by adulthood. Even when intrinsic conduction appears normal, these patients appear to be more susceptible to traumatic AV block during surgical or catheter procedures.

Surgical repair of certain forms of CHD can result in direct trauma to the AV conduction tissues. Although improved knowledge of the precise location for the AV node and His bundle in various forms of CHD has reduced the occurrence, closure of some ventricular septal defects, surgery for left-heart outflow obstruction, and replacement or repair of an AV valve may still be complicated by AV block. Fortunately, in more than half of cases, this injury is a transient affair related to myocardial stretch or edema rather than physical severing of the conduction tissues, and AV conduction recovers within 7 to 10 days of operation. But, for any patient with postoperative AV block that is not expected to resolve or that persists at least 7 days after cardiac surgery, permanent pacemaker implantation is advised as a class I indication. A pacemaker may be considered by some as a class IIb indication when surgical AV block recovers but the patient is left with permanent bifascicular block.

**Pacemaker and ICD Implantation in Adults With CHD**

**Indications**

An important consideration when one interprets the American College of Cardiology/American Heart Association/North American Society of Pacing and Electrophysiology task force guidelines for pacemaker implantation is that standard transvenous systems may be contraindicated or difficult in many CHD patients owing to complexities of venous anatomy or to the presence of significant intracardiac shunting that creates a thromboembolic risk from intravascular leads. Epicardial implantation is certainly possible in these circumstances, although the surgery is more involved, and long-term lead performance may be inferior to transvenous systems. Additionally, epicardial lead placement in an older CHD
TABLE 4. Special Considerations During Transvenous Pacemaker and ICD Lead Implantation in Adults With CHD

<table>
<thead>
<tr>
<th>Consideration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital venous anomalies</td>
</tr>
<tr>
<td>Acquired venous occlusion</td>
</tr>
<tr>
<td>Inability to access a chamber</td>
</tr>
<tr>
<td>Surgical obstacles (patches, conduits, and baffles)</td>
</tr>
<tr>
<td>Intracardiac shunts</td>
</tr>
<tr>
<td>Ventricular geometry and position</td>
</tr>
</tbody>
</table>

The patient who has undergone multiple prior cardiac operations presents the surgeon with a highly scarred mediastinum; dissection down to the myocardial surface must be performed carefully and deliberately to uncover sites with good sensing and pacing function. If a CHD patient is undergoing cardiac surgery for other hemodynamic reasons and it appears highly likely that epicardial pacing might be needed at some distant date, the surgeon may want to take advantage of wide intraoperative exposure to place leads for future use. Fortunately, 86% of leads placed at the time of cardiac surgery are found to function well when retrieved at a mean of 252 days after the operation.

The indications for ICD implantation in CHD patients are still evolving, but in general, they follow guidelines that are similar to other varieties of adult heart disease. The most common type of CHD that requires ICD implantation is tetralogy of Fallot, followed by transposition of the great arteries and left-sided obstructive diseases.

The experience with biventricular resynchronization pacing in CHD patients with depressed ventricular function is limited but promising. Clinical studies are now under way to refine selection criteria, as well as to investigate the clinical merits of RV resynchronization to offset right bundle-branch block after surgical closure of ventricular septal defects and tetralogy repair.

Technical Considerations

Careful preprocedural planning is essential for successful pacemaker or ICD implantation in adults with CHD (Table 4). Some of these patients can have anomalies of systemic venous return and the coronary veins, the most common of which is persistent left superior vena cava draining to the coronary sinus. In this situation, the right superior vena cava and innominate vein can vary in size from normal caliber to completely absent. Implantation is still possible through a left superior vena cava, although making the necessary turn across the tricuspid valve from the coronary sinus is difficult and may require use of a long sheath or a large right atrial loop to direct the tip toward the ventricle. If pacing within the coronary sinus is needed for cardiac resynchronization or any other purpose, anomalies of the coronary sinus need to be considered, including ostial atresia, unroofed coronary sinus, and extreme dilation due to persistent left superior vena cava.

Venous occlusion is a recognized complication of permanent transvenous pacing leads. Risk factors for venous thrombosis after lead placement include the absence of anticoagulant therapy, a history of prior venous thrombosis, use of female hormone therapy, and the presence of multiple pacing leads. Venous occlusion is therefore common in adult CHD patients with long-term pacing who may have multiple leads in place for several decades. A contrast injection into the antecubital vein(s) with follow-through venography is helpful to investigate acquired and congenital abnormalities. If venous access is needed in the setting of a complete venous occlusion, a variety of techniques for recanalization and venous dilation are now available.

Transvenous chamber access can be an issue if complicated atrial baffling has been used to redirect venous return. One major challenge in this regard occurs when ventricular pacing is needed after a Fontan repair. An epicardial approach is used in most of these cases, but successful transvenous ventricular lead implantation after the Fontan operation has now been reported in several series, usually involving lead placement into a coronary venous branch vessel.

Combined challenges of surgical obstacles, chamber access, and unusual ventricular geometry are present in patients with transposition of the great arteries after an atrial switch (Mustard or Senning) procedure. Extensive atrial baffling limits sites of atrial capture to small regions in the left atrial appendage (where phrenic nerve stimulation may be hard to avoid), anterior left atrial roof, or superior cava–right atrial junction. Baffle obstruction is fairly common in these patients and may require endovascular stent placement before lead placement (Figure 5). In addition, the ventricular lead must be placed in a morphological left ventricle that is thin and non trabeculated, which requires close attention to tip fixation and sensing parameters.

Figure 5. Chest radiograph demonstrating transvenous ICD lead positions in a young adult with L-TGA after a Mustard procedure (A, posteroanterior projection; B, lateral projection). Complete obstruction of the superior limb of the atrial baffle was present and required recanalization, dilation, and stent placement (arrow and inset) to open a path to the left ventricle. The atrial lead was positioned in the superior vena cava limb of the baffle proximal to the stent. Note that the tip of the ICD lead is attached to the left ventricular apex. SVC/RA indicates superior vena cava and right atrial junction; LA, left atrium (which became the neo-right atrium after surgery); and LAA, left atrial appendage.
Intracardiac shunts can lead to embolic stroke via right-to-left shunting or inadvertent lead placement in the systemic circulation.114 These shunts can occur at either the atrial or ventricular level, ranging from a patent foramen ovale to large residual septal defects and patch leaks. In addition, patients with high central venous pressure (as often occurs after the Fontan repair) can develop direct connections between the supracardiac veins and pulmonary venous atrium through veno-venous collaterals.115,116 Trivial shunts that are predominately left-to-right are probably not absolute contraindications to transvenous leads, but larger shunts, particularly if right-to-left, need to be evaluated carefully by angiography or echocardiography before a final decision is made on the route for lead implantation. If transvenous leads are strongly preferred in such cases, shunt closure can be attempted beforehand with interventional techniques such as septal occluders, covered stents, or even surgery.117,118 If intracardiac shunting cannot be eliminated satisfactorily, epicardial lead placement is probably the wisest alternative. If there are still compelling indications for a transvenous system, anticoagulation is used after lead placement in patients with residual shunting at most centers, although no firm data currently exist to verify the efficacy of this measure.

Transvenous ICD lead implantation requires a thorough understanding of the ventricular anatomy and chamber positions to anchor the lead tip securely and ensure a suitable vector for defibrillation. When epicardial ICD leads are required, there has been a shift away from traditional patches to novel configurations involving coils (Figure 6) placed in a subcutaneous or pericardial position.119 The experience with this approach is limited, but it appears to offer increased flexibility that can accommodate a wide variety of ventricular geometries and heart sizes.

Conclusions

Arrhythmias in adults with CHD are a growing problem that requires unique solutions. The number of clinicians trained to provide electrophysiological care to this unusual patient group is still somewhat limited. Cardiology and electrophysiology training programs need to consider devoting more understanding of the ventricular anatomy and chamber positions.119 The experience with this approach is limited, but it appears to offer increased flexibility that can accommodate a wide variety of ventricular geometries and heart sizes.

Conclusions

Arrhythmias in adults with CHD are a growing problem that requires unique solutions. The number of clinicians trained to provide electrophysiological care to this unusual patient group is still somewhat limited. Cardiology and electrophysiology training programs need to consider devoting more curriculum time to this topic in an effort to improve care delivery into the future. The paucity of evidence-based management protocols for arrhythmias in adults with CHD is also a concern, and a need clearly exists for larger collaborative studies involving both pediatric and adult CHD centers to generate more objective treatment guidelines.

Disclosures

None.

References


KEY WORDS: ablation, atrial flutter, heart defects, congenital arrhythmia, pacemakers, electrophysiology, pediatrics
Arrhythmias in Adult Patients With Congenital Heart Disease
Edward P. Walsh and Frank Cecchin

Circulation. 2007;115:534-545
doi: 10.1161/CIRCULATIONAHA.105.592410
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2007 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/115/4/534

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/