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ELECTROPHYSIOLOGY ISSUES IN ADULT CONGENITAL HEART DISEASE

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Introduction

Improved surgical outcomes in children have led to a growing population of adults with congenital heart disease. Rhythm disturbances in the adult congenital heart disease (ACHD) patient can be intrinsic to the anomaly or acquired after palliation. Tachyarrhythmias, either supraventricular or ventricular, and bradyarrhythmias, either sinus node dysfunction or atrioventricular block, may occur frequently. Technological advances in intervention and surgical approaches have led to prophylactic and therapeutic reduction in arrhythmias. In order of escalation, this article addresses medical management, catheter ablation, device therapy for antitachycardia pacing and defibrillation, and surgical intervention.

There are now an estimated one million-plus ACHD patients living in the United States. An estimated 45% of those have simple defects (e.g., atrial septal defects), 40% have moderately complex defects (e.g., tetralogy of Fallot, or ToF), and 15% have severely complex defects (e.g., single ventricle anatomy or surgical palliations for transposition of the great arteries [TGA]).[1] The moderate and severe categories have a high incidence of arrhythmia. Of older repaired ToF patients, 34% develop symptomatic atrial or supraventricular tachycardias,[2] 8.5% develop high-grade ventricular tachycardia (VT),[3] and there are an increasing number of implantable defibrillators due to a sudden-death estimate of 2% per decade.[4] Thus, an estimated 50,000 adults with repaired ToF will require electrophysiology follow-up with 100 sudden deaths per year nationally.[5] Roughly 3% of all congenital heart surgeries requiring cardiopulmonary bypass have post-operative AV block, and 1% will require permanent pacing.[6] Older atriopulmonary Fontans have up to an estimated 50% incidence of atrial tachycardia within a decade of palliation due to suture lines and elevated atrial pressures.[7] Patients who have undergone an atrial switch operation (e.g., Mustard or Senning operations) are rarely exclusively in sinus rhythm a decade after repair, thus posing problems for rhythm control and anticoagulation.[8]

Catheter ablation in congenital heart disease is often more challenging than structurally normal hearts because of abnormal anatomy (congenital and/or post-surgical) and thicker chamber walls due to unfavorable hemodynamics. In preparation, review of noninvasive imaging, previous catheterization angiography, and surgical palliation reports are paramount prior to a procedure. Fortunately, to meet the growing challenge, newer software with 3-D packages allow for improved mapping. In addition, advances with larger-tipped and irrigated-tip catheters allow for deeper, more effective lesions to be placed.[9]

Several advances in surgical approach have led to a reduction in arrhythmias. For example, the arterial switch operation for d-transposition of the great arteries instead of the aforementioned atrial switch operation eliminates the use of a systemic right ventricle that can later develop intra-atrial reentrant tachycardia and sinus node dysfunction. Other advances include paying careful attention to minimizing ventriculotomies in ToF and ventricular septal defect (VSD) repairs, earlier complete repairs, and valve sparing to reduce pulmonary insufficiency. Finally, completion of the extra-cardiac Fontan procedure (e.g., total cavopulmonary connection) for single ventricles avoids extensive suture lines in the right atrium, thereby reducing scarring and higher pressures that lead to IART and sinus node dysfunction.[5] Extracardiac (EC) conduits and lateral tunnel (LT) Fontans are preferred today, and the Fontan conversion procedure (converting prior atriopulmonary Fontans to the EC or LT type) can be performed to reduce arrhythmia and thromboembolic events.[10]

Tachyarrhythmias

Atrioventricular reentrant tachycardia[5, 9]

Atrioventricular (AV) reentrant tachycardias result from accessory pathways that are sometimes intrinsic to congenital lesions. Classically, Ebstein's anomaly of the tricuspid valve is associated with Wolff-Parkinson-White (WPW) syndrome in 20% of cases, most often in the posterior septal location where the valve is most displaced. Almost half of these have multiple pathways. L-TGA (Levo-TGA, also known as congenitally corrected TGA) can also be associated with WPW syndrome when the left-sided tricuspid valves are Ebsteinoid. With atrial dilation over time, atrial flutter and atrial fibrillation may become complicated by rapid ventricular response via the accessory pathway. Catheter ablation is now the standard of care, but short-term success is less than in structurally normal hearts (~85%), with higher rates of tachyarrhythmia recurrence (~25%) due to right atrial enlargement, poor catheter contact with tricuspid regurgitation, fractionated electrograms, multiple pathways, and a less aggressive approach near a thinner right AV groove. Right coronary angiography or thin-caliber electrode

catheters down the right coronary artery can help map the appropriate location to ablate. Non-Ebstein CHD has an acute success rate of 80% with 15% late recurrence. Cryoablation allows closer proximity to the AV node with more time and margin of error to detect AV block before permanent damage is done.

Intra-atrial reentrant tachycardia (IART)[5,9]

Intra-atrial reentrant tachycardia is the most common symptomatic tachyarrhythmia in adults with palliated congenital heart disease. IART is a macro-reentrant circuit within atrial muscle caused by progressive right-atrial dilation from elevated pressures, hypertrophy, and scarring around suture lines. It is most often seen in older Fontans (up to 50%)[7] and atrial baffles for TGA (~30%)[8] but can also occur with repair of simple cardiac defects, such as an atrial septal defect.[11]

IART has a slower atrial rate (150–250 bpm) than flutter, often allowing 1:1 conduction. Unlike flutter, the electrocardiogram usually does not have classic sawtooth P waves, and 2:1 or 3:1 conduction (Figure 1) may have P waves masked in the QRS and T waves. AV nodal blockade with vagal maneuvers or adenosine can assist in unmasking the abnormal atrial activity. Like atrial fibrillation or flutter, this can lead to hemodynamic instability including cardiac arrest or, in the subacute setting, cause heart failure from loss of AV synchrony and thromboembolic phenomena. When a tricuspid valve is present, the cavotricuspid isthmus is often part of the circuit as it is in atrial flutter. Caval orifices, valve orifices, the crista terminalis, and suture lines can complete the circuit. Thus, multiple coexisting IART pathways are possible. Circuits are less predictable when there is tricuspid atresia or right ventricular hypoplasia. The tachy-brady syndrome, consisting of IART and sinus node dysfunction, and older age of surgical palliation are risk factors.

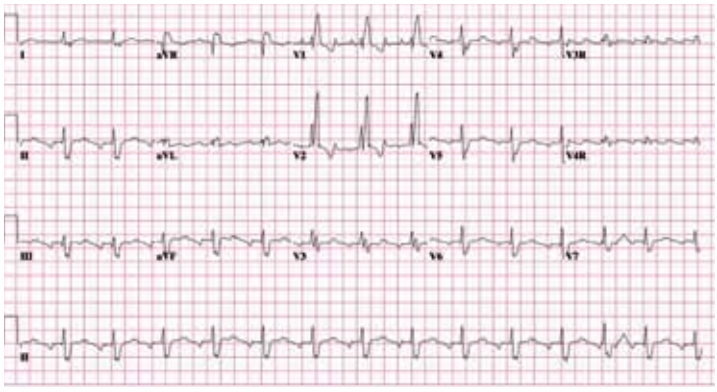

Figure 1. 3:1 intra-atrial reentrant tachycardia in repaired tetralogy of Fallot.

Acute treatment of IART by electrical means (direct current cardioversion or overdrive pacing) or chemical means (class I or III antiarrhythmics) is effective, but maintenance of sinus rhythm is challenging. Infrequent episodes require intermittent cardioversion. Escalation with more aggressive therapy depends on individual risk-benefit profiles and includes potent antiarrhythmics such as amiodarone, radiofrequency ablation using large-tip and irrigated-tip catheters with 3-D mapping, permanent pacemaker implantation for bradycardia with antitachycardia pacing for IART, and surgical revision with modified atrial maze procedure. Antiarrhythmics alone tend not to be effective, with 70% still having breakthrough IART in first two years of medication. Antitachycardia pacing can be successful in half the patients but can also shift the cycle length to faster IART circuits or degenerate into atrial fibrillation. Ablation can have ~80% early success rates, but recurrence can be 20% in biventricular circulations and up to 40% in Fontans due to multiple circuits and thicker atrial tissue. In atrial switch palliations for D-TGA, the cavotricuspid isthmus is only accessible prograde from a transbaffle puncture or retrograde arterial approach. Mapping the activation sequence is critical for postulating a rational reentry model to ablate. Benefits include decreasing IART frequency to allow cessation of antiarrhythmics.[12] If surgery is necessary for hemodynamic reasons, older atriopulmonary Fontans can be converted to more contemporary extracardiac conduit or lateral tunnel Fontans using the maze procedure, with 12.5% IART recurrence and only 10% on long-term antiarrhythmics.[13] However, there are estimates of 5% mortality or postoperative need for orthotopic heart transplantation with these procedures, making risk-benefit discussion paramount.[10]

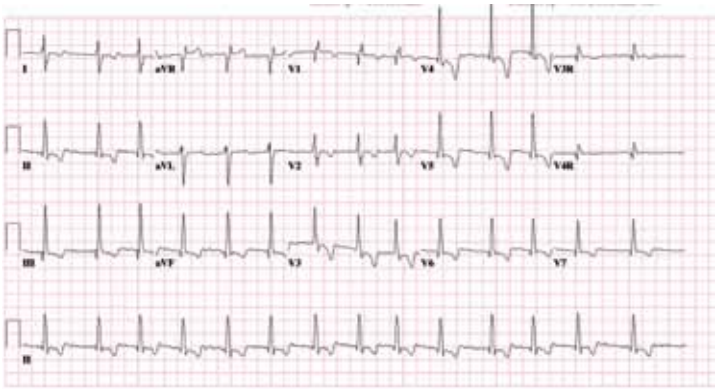

Figure 2. Atrial fibrillation in Fontan circulation.

Atrial fibrillation

Atrial fibrillation (Figure 2) can be seen commonly with left-sided lesions such as congenital aortic stenosis or mitral valve abnormalities where the left atrium receives hemodynamic stress. Hemodynamic instability can occur with rapid ventricular response. Heart failure and stroke are complications as they are in structurally normal hearts. Acute treatment with electrical or chemical cardioversion and chronic treatment with rate control and anticoagulation should be utilized. As with IART above, antiarrhythmics, catheter ablation (not yet case reported), pacemaker implantation for tachy-brady syndrome, and Cox-MAZE procedure at time of surgery are all examples of therapeutic options.[5,9]

In a population-based CHD database study of atrial tachyarrhythmias in congenital cardiac anomalies, 11% of 71,467 patients were adults with a diagnosis of atrial arrhythmia (flutter, tachycardia, IART, or fibrillation). Isolated right-sided lesions (mostly ASDs at 73%) accounted for 3.1%, isolated left-sided lesions were 2.4% (31% mitral insufficiency, 26% aortic stenosis, 23% VSDs), and 5.3% were mixed or unspecified. Patients with right-sided lesions were on average five years younger (63 versus 68) than left-sided patients and included 41% men compared to 54%. In this study, an 18 year old had an 18% 30-year risk of developing an atrial arrhythmia (comparable to a 55-year-old structurally normal heart). There was no statistical significance between mortality, stroke, or heart failure between right- and left-sided lesions, but left-sided lesions had a higher rate of catheterization, cardiac surgery, or arrhythmia surgery. This may be due to lack of standardized therapies or limitations of population-based database studies.[14] An example of a wide complex tachycardia — atrial tachycardia with aberrant conduction — is depicted in Figure 3.

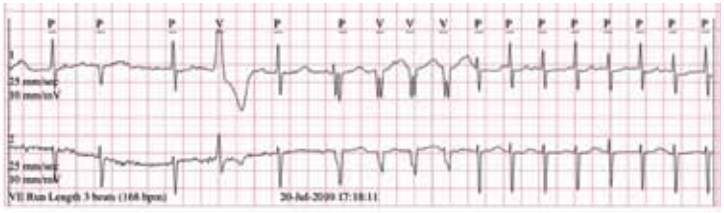

Figure 3. Atrial tachycardia with aberrancy in a D-TGA status post-Mustard operation.

Ventricular tachycardia

Ventricular arrhythmias occur primarily in patients with ventriculotomies, with or without patch closure of VSDs, and ventricular dysfunction from pressure or volume overload. With ventriculotomies, there may be macroreentrant circuits related to surgical scar, septal defect rims, and valve annulus edges. Examples of ventricular dysfunction include pressure-loaded right ventricles (systemic RVs, Eisenmenger syndrome, or unrepaired tetralogy), aortic valve disease, and severe Ebstein's anomaly.[5]

The majority of literature for VT in adult congenital heart disease surrounds repaired tetralogy, with a prevalence ranging from 3 to 14%.[2-4, 15-17] It can lead to syncope or cardiac arrest and accounts for a majority of the well-cited 2% per decade sudden death risk.[4] Risk factors include older age at complete repair, older age at follow-up, history of palliative shunts, high-grade ventricular ectopy, inducible VT on electrophysiology study, abnormal RV hemodynamics due to abnormal mechano-electric interaction,[15, 16] and wide QRS greater than 180 ms. However, no single criterion is highly predictive.

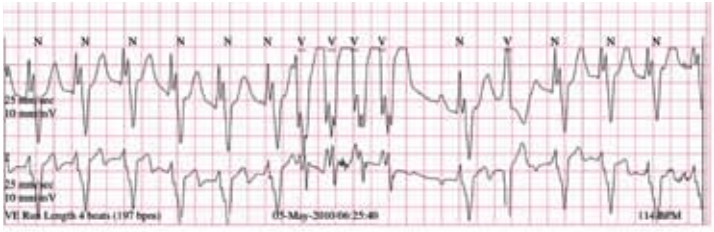

Figure 4. Nonsustained ventricular tachycardia in repaired tetralogy of Fallot.

Various routes of management may be conservative, such as observation of asymptomatic patients with yearly follow-up and EKG supplemented by Holter (Figure 4), or exercise treadmill testing for high-grade ventricular ectopy. Alternating echocardiograms and cardiac MRI to evaluate pulmonary regurgitation, RV size, and RV function might help with timing of pulmonary valve replacement. Worsening function or high-grade ectopy might prompt antiarrhythmics or placement of a primary implantable cardioverter-defibrillator (ICD). There are no standardized guidelines for the asymptomatic patient with repaired tetralogy.[5] Evaluation of symptoms (palpitations, dizziness, syncope) with hemodynamic catheterization and EP study may lead to ablation of stable monomorphic VT circuits or IART.[18] In two small series with a total of 30 patients (17 ToF, 8 VSD, 5 other CHD), mapping was used in 28 subjects and 25 of those (89%) had acutely successful ablation of VT with recurrence in 5 (20%).[18, 19] Primary prevention with an ICD may be necessary for more hemodynamically worrisome rhythms. Survived cardiac arrest or sustained VT almost universally warrant secondary prevention with an ICD.[20] Given the numbers of repaired ToF and other ACHD patients, multicenter collaboration can study, optimize, and refine guidelines for management.

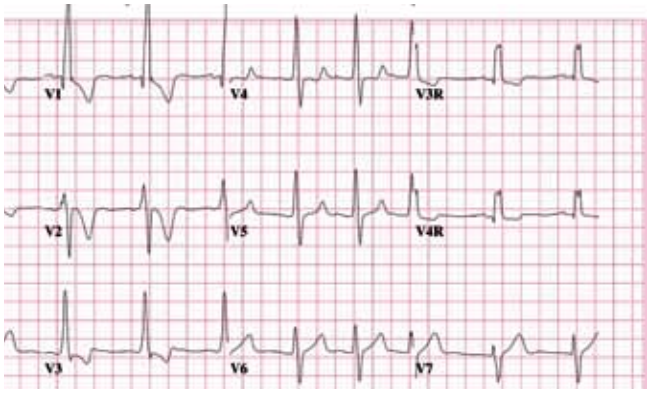

Figure 5. Atrial pacemaker in D-TGA status post-Senning operation. Note atrial pacing spikes and right ventricular hypertrophy.

Bradyarrhythmias

Sinus Node Dysfunction

Heterotaxy with right atrial isomerism may present with dual sinus and AV nodes, and left atrial isomerism may present without a true sinus node and rely on junctional escape or slower atrial ectopic rhythms, eventually requiring pacemaker placement. More commonly, sinus node dysfunction occurs in palliated single ventricles (Glenn, Fontan) and atrial switch operations for D-TGA (Figure 5). IART and atrial fibrillation are more frequent with sinus node dysfunction as part of the tachy-brady syndrome and warrant permanent pacing, as chronotropic incompetence is poorly tolerated with single ventricles, reduced cardiac function, and suboptimal anatomy such as worsening AV valve regurgitation.[5] ACC/AHA guidelines recommend permanent pacing for symptomatic bradycardia, tachy-brady syndrome requiring antiarrhythmics other than digoxin, and sinus bradycardia with average heart rate less than 35 bpm or greater than 3-second pauses.[21]

Atrioventricular block

Atrioventricular block may occur with L-TGA, increasing from 3 to 5% at birth to 20% by adulthood. The AV node and His bundle are more anteriorly displaced and more susceptible to AV block during catheter and surgical procedures. VSD closures, left ventricular outflow tract obstruction resections, and AV valve repairs and replacements are also fraught with post-op AV block due to stretch, edema, or severing of the conduction fibers. Stretch and edema may resolve within seven to ten days post-operatively.[5] Complete heart block or advanced second-degree AV block persisting after seven days post-operatively is a Class I indication for permanent pacing. Post-op AV block that reverts to sinus with bifascicular block is a Class IIb indication for permanent pacing.[21]

Pacemakers and Defibrillators[5, 9]

Permanent pacemakers and implantable defibrillators can have a transvenous or epicardial placement. The transvenous approach is less invasive to place with preferable long-term lead performance but may need to take into account systemic venous drainage or occlusion. Other anatomical considerations might be a left-sided SVC draining to a dilated coronary sinus; stenotic atrial baffles in d-TGA leading to small areas of left-atrial appendage close to the phrenic nerve or a thin-walled subpulmonic left ventricle less inclined for screwing in ventricular leads for pacing; and residual right-to-left shunting that may require device or surgical closure prior to lead placement (Figures 6A, 6B). Transvenous pacing may even be considered in atriopulmonary Fontans if there is access to ventricular pacing via the coronary sinus network. Although there are no published guidelines from the ACC or AHA, anticoagulation

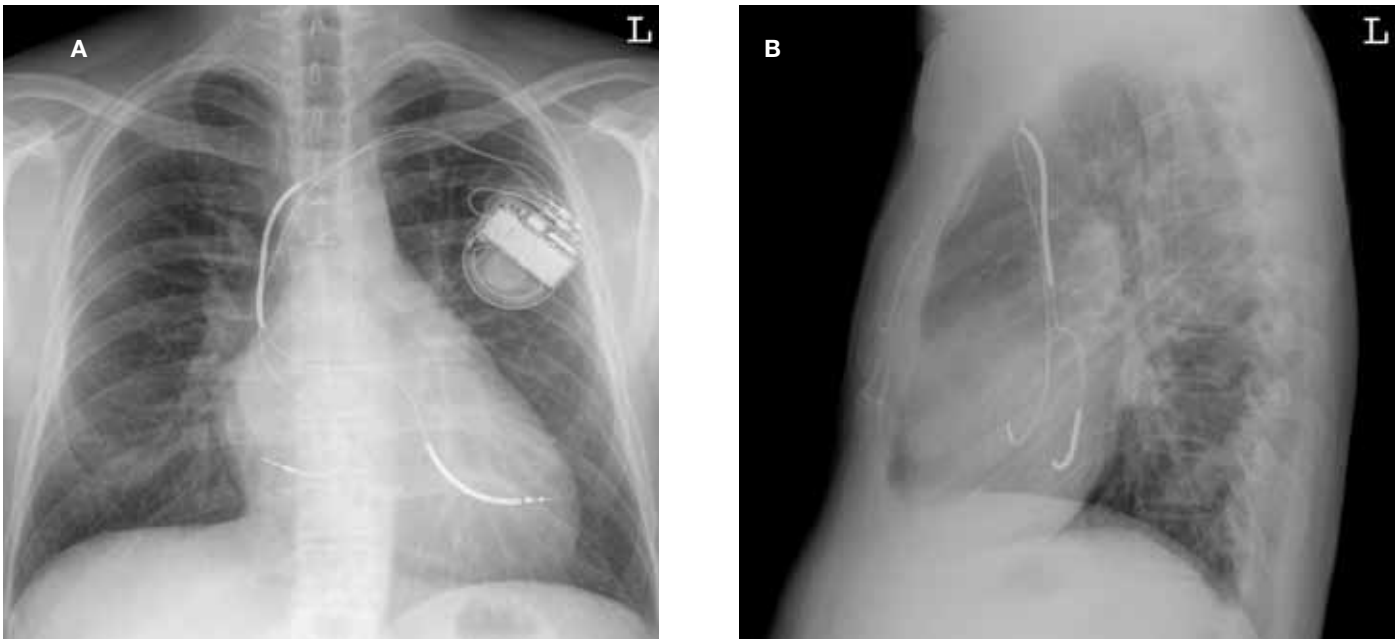

Figure 6 A & B. Posterior-anterior and lateral chest roenterogram of dual-chamber pacing leads in D-TGA status post-Mustard operation. Note the left-sided subpulmonic ventricular lead.

may be used if transvenous systems are employed with residual intracardiac shunting. Risk factors for venous occlusion include multiple leads, prior thromboses, estrogen therapy, and lack of anticoagulation. Thus, epicardial pacing is often preferred for many ACHD patients. Epicardial leads have the advantage of being external to the bloodstream and not thrombogenic. However, gaining visualization through a scarred mediastinum can be challenging for the cardiothoracic surgeon, so anticipation and coordination with other surgical procedures is advised when treating patients who will eventually need pacing. Although there is concern about long-term performance of epicardial placement, 86% were functional at an average 252 days after placement.[22] Only ~20% of these epicardial leads were steroid eluting,[22] which may imply that with increasing use, a higher percentage might be functional at long-term follow-up.

Defibrillators are most often implanted for ToF, D-TGA, and aortic stenosis. At five years' post-implantation, 30% of patients had appropriate shocks, 25% received inappropriate shocks, and 21% had lead fractures, which may reflect a more vigorous lifestyle and younger patient profile.[20] Epicardial defibrillators used in intracardiac shunts, single ventricles, and small body habitus have transitioned from patches to pericardial and subcutaneous coils.

Cardiac resynchronization therapy (CRT) has been reported in a case series of 73 adults with congenital heart disease where the mean increase in ejection fraction was 12% and mean decrease in QRS was 39 ms. Single ventricles were unlikely to improve despite QRS shortening.[23] Pacing for right bundle branch block to improve contractility is also currently under investigation.[5]

EP Pearls

- In Ebstein's anomaly and L-TGA with an Ebstenoid left sided tricuspid valve, consider WPW for supraventricular tachycardias.
- For narrow complex regular tachycardias >100 bpm in older Mustards, Sennings, and Fontans, consider IART. Sinus node dysfunction leading to bradycardia is common, so tachycardia is often pathologic and atrial flutter waves may be masked.
- Ventricular tachycardia in repaired Tetralogy of Fallot is serious and requires careful consideration. Risk factors relate to chronology (older age of repairs), hemodynamics (large RV with pulmonary insufficiency), and conduction (ectopy, inducible VT, QRS >180 ms). Options include observation with stress tests, pulmonary valve replacement, or defibrillator implantation but should be facilitated by a center that specializes in adult congenital heart disease.
- Implantation of pacemaker and/or defibrillators for pacing or defibrillation requires understanding of the anatomy of complex congenital heart defects. To minimize major adverse cardiac events, expert ACHD consultation should be sought.
- Advances in medical therapies need to be tailored to the specific lesion and population type. For example, increased inappropriate shocks and lead fractures in a younger population may reflect a more physically-active lifestyle. Alternatively, Fontan patients with depressed ejection fraction may not benefit from cardiac resynchronization therapy due to their univentricular anatomy.
- Management of rhythm disturbances requires awareness of patient symptoms, risks of silent arrhythmias, and willingness to pursue therapy. Communication between the patient, the primary cardiologist, and an electrophysiologist with expertise in congenital heart disease is strongly recommended. Advanced management requires facilities with significant experience in catheter ablation, device implantation, and congenital heart surgery.

Conclusion

Adult congenital heart patients have various arrhythmias that require attention and electrophysiologic management. Due to the rapidly growing numbers of palliated ACHD patients, the incidence of arrhythmias will continue to increase. Many of the traditional published reports are based on data from smaller cohorts using older surgical techniques, prior to advances in catheter-based ablation and devices. More studies are needed to provide guidelines for managing common issues like ventricular tachycardia in repaired tetralogy or cardiac resynchronization therapy in ACHD. Management of tachyarrhythmias and bradyarrhythmias relies on therapeutic options that include antiarrhythmics, radiofrequency ablation, implantable devices, and surgical cryoablation.

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