Electrophysiologic Therapeutics in Heart Failure in Adult Congenital Heart Disease

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KEYWORDS

• Arrhythmias • Electrophysiology • Heart failure • Adult congenital heart disease • Therapeutics • Antiarrhythmics • Device therapy • Resynchronization

KEY POINTS

- Antiarrhythmic therapy is an important component of atrial arrhythmia management in the adult with congenital heart disease.
- Device therapy including conventional pacing, antitachycardia pacing, cardioversion, and defibrillation can be useful for atrial and ventricular arrhythmias in patients with congenital heart disease.
- Ablation of atrial and ventricular arrhythmias can decrease morbidity and mortality in this patient population. Advanced technologies including three-dimensional navigation systems and new energy sources can aid in successful ablation.
- Surgical interventions to improve hemodynamics or to interrupt arrhythmia circuits can be a useful therapeutic option in selected adults with congenital heart disease.

HEART FAILURE AND ARRHYTHMIAS IN ADULT CONGENITAL HEART DISEASE: SCOPE OF THE PROBLEM

With improvement in medical, interventional, and surgical therapies for congenital heart disease (CHD), most patients with CHD are surviving into adulthood such that there are now more adults living with CHD in the United States and Canada than there are patients with CHD younger than 18 years old.[1] Survival of the patient with adult CHD (ACHD) continues to improve with decreasing mortality rates that parallel those of the general population.[2]

Despite these successes, heart failure remains one of the most common causes of morbidity and mortality in ACHD.[3–6] In 2007, heart failure accounted for 20% of all ACHD hospital admissions in the United States.[7] Patients with ACHD with heart failure had a threefold increase in hospital mortality compared to those without heart failure.

Not surprisingly, most patients with ACHD with heart failure die from cardiovascular causes, especially pump failure and arrhythmias.[8] Fifty-two percent of patients with ACHD admitted with heart failure in the United States in 2007 also had arrhythmias.[8] Management of arrhythmias, therefore, is a critical component of caring for the patient with ACHD with heart failure.

These arrhythmias often result from surgical scars, as well as chronic volume and pressure

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loads, cyanosis, and chamber enlargement. Electrophysiologic therapeutic strategies in this population can include control of arrhythmias and prevention of sudden cardiac death (SCD) as well as preservation of cardiac function.

BRADYARRHYTHMIAS

Sinus Node Dysfunction

Congenital sinus node dysfunction may be seen in patients with CHD, such as those with heterotaxy with left atrial isomerism. These patients may lack a true sinus node altogether, which makes their heart rate dependent on slower atrial or junctional escape rhythms. More commonly, however, sinus node dysfunction is a result of surgical trauma to the sinoatrial node or its artery, which may occur during the atrial switch procedure (Mustard or Senning procedures) for d-transposition of the great arteries (d-TGA) or single-ventricle palliation with a Glenn or Fontan procedure.[9–12] In patients with Mustard procedures for d-TGA, symptomatic sinus node dysfunction is observed in 64% and 82% at 5 and 16 years of follow-up, respectively.[13]

Chronotropic incompetence may be poorly tolerated in patients with ACHD with compromised hemodynamics, especially those with a single ventricle or significant atrioventricular (AV) valve regurgitation. The likelihood of a patient developing intra-atrial reentrant tachycardia (IART) or atrial fibrillation is also increased significantly in this setting, which can result in the induction of secondary ventricular tachycardia and SCD.[9,14]

American College of Cardiology (ACC)/American Heart Association (AHA)/Heart Rhythm Society (HRS) 2012 guidelines recommend permanent pacing for symptomatic age-inappropriate bradycardia (class I, level of evidence B), tachy-brady syndrome with recurrent IART (class IIa, level of evidence C), sinus bradycardia in the setting of complex CHD with resting heart rate less than 40 bpm or pauses in ventricular rate longer than 3 seconds (class IIa, level of evidence C), or CHD and impaired hemodynamics caused by sinus bradycardia or loss of AV synchrony (class IIa, level of evidence C).[15]

AV Node Dysfunction

Patients with AV discordance have a 2% incidence of developing spontaneous AV block on an annual basis.[16] AV nodal conduction defects, however, are more commonly the sequelae of intracardiac repair (1%–3% of congenital heart surgeries), typically involving the ventricular septum.[17–20] In a study of adult patients with heart failure with a single or systemic right ventricle, 72% of symptomatic patients had a history of heart block. Of those who died with heart failure, 76% had second-degree AV block or higher and 62% required a pacemaker.

ACC/AHA/HRS 2012 guidelines recommend permanent pacing for advanced second-degree or third-degree AV block associated with symptomatic bradycardia, ventricular dysfunction, or low cardiac output (class I, level of evidence C), postoperative advanced second-degree or third-degree AV block that is not expected to resolve or that persists at least 7 days after cardiac surgery (class I, level of evidence B), or congenital third-degree AV block with a ventricular rate less than 70 bpm in the setting of CHD (class I, level of evidence C).

When permanent pacing is indicated, challenges include lack of or obstructed venous access, obstructed baffles or conduits, baffle leaks, difficulties in lead positioning, high rates of lead complications, and coexisting intracardiac shunts.[21–23] Single-site ventricular pacing, even from the subpulmonary left ventricle (LV) in this setting, results in an obligatory dyssynchronous ventricular contraction that may be associated with a reduction in ventricular performance over time. Pacemaker implantation has been proposed as a risk factor for mortality after the first heart failure admission in adults with CHD.[8,24]

Patients with CHD and devices face a lifelong prospect of potential device and lead-related complications, often requiring multiple reinterventions.[22] Given the high incidence of lead failure in an aging patient population, lead extraction procedures are increasingly required in adults with CHD.[25] Particular challenges are encountered in this population. In a cohort of 175 adults with attempted laser extraction of 270 leads, those with CHD were younger at implantation, had older leads at extraction, more right-sided implants, a higher proportion of active fixation leads, and had particular anatomic features including intracardiac shunting, leads in subpulmonary LVs, left atrial (LA) appendages, severely dilated and/or dysfunctional subpulmonary right ventricles (RVs), and partially obstructed baffles.[26] Despite these complexities, success rates (91%) and complication rates (6%) were similar to those in patients without CHD, although a longer procedural time was required.[26]

ATRIAL TACHYARRHYTHMIAS

Intra-atrial Reentrant Tachycardia

IART is the most common symptomatic sustained tachyarrhythmia in the ACHD population.[11,27] The terms intra-atrial reentrant tachycardia and incisional tachycardia have become customary labels