17th Symposium on Cardiac Arrhythmias

Electrophysiology Updates in Adult Congenital Heart Disease

Wilson W. Lam, MD

The adult congenital heart disease (ACHD) population is growing. In the United States, there are well over 1.5 million ACHD patients living today, and the estimated rate of increase is 5% per year. Advances in medical management and surgical technique have enabled survival to adulthood to approach 90%, and the relative percentage of patients with moderate (for example, Ebstein anomaly and tetralogy of Fallot) and complex CHD (such as Fontan circulation and transposition of the great arteries) has increased in union with improved survival.[1,2] For newly diagnosed ACHD in adults older than 40 years of age with simple disease or disease that presents no hemodynamic concern, evaluation by a general cardiologist in consultation with an ACHD cardiologist is recommended. Moderate-to-complex disease should have at least annual follow-up at an ACHD center to ensure the best possible long-term outcomes.[3]

The 2014 American College of Cardiology/American Heart Association/Heart Rhythm Society (ACC/AHA/HRS) guideline for management of patients with atrial fibrillation (AF) summarizes the pathophysiology of atrial arrhythmias.[4] The "fire and fuel" include a combination of scar, stretch, and stress, with triggers that initiate arrhythmias and postulated rotors that sustain them. Treatment should be targeted at the various inputs and mechanisms of action.[4] Three recent areas that pertain both to structurally normal hearts and to ACHD include obesity reduction, sleep apnea management, and influenza vaccination. Weight reduction of 14.3 kg (compared with 3.6 kg in control patients) from a mean body mass index of 33 kg/cm^2 led to less burden (in symptom severity, number of episodes, and cumulative duration) and to smaller interventricular septal thickness and left atrial area.[5] Results from multiple studies have shown that continuous positive airway pressure to manage sleep apnea reduces AF recurrence.[6,7] Just earlier this year, investigators in Taiwan showed a correlation between influenza vaccination and an 18% lower risk of AF.[8] Therefore, reducing the burden of AF begins with the primary care provider and the general cardiologist, before an electrophysiologist is consulted.

The 2014 ACC/AHA/HRS guidelines for AF[4] propose several oral agents for rhythm control. In general, we avoid Vaughn-Williams class IC agents (such as flecainide or propafenone) in structural heart disease because the slowing of conduction can have proarrhythmic effects. In scar tissue, this can promote atrial flutter or ventricular tachycardia via reentry. When flecainide was studied in a pediatric population with cardiomyopathy or structural heart disease, it was not found to significantly increase complications, but it has not been studied in the ACHD population.[9] Two series[10,11] using dofetilide for atrial tachyarrhythmias showed rhythm control but a high percentage of torsade de pointes or excessive QTc prolongation (8%–15%). However, both of these series were small (n=13 and 20). No studies exist for dronedarone in the ACHD population. Therefore, sotalol and amiodarone remain the tried-and-true therapies until more data are gathered.

The 2014 Pediatric & Congenital Electrophysiology Society (PACES)/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease[12] addresses itself to anticoagulation as a treatment for atrial arrhythmias. The guidelines recommend vitamin K antagonists in moderate-to-complex cases of CHD because of the lack of safety and efficacy data for the target-specific novel oral anticoagulants (NOACs). The writers suggest that these newer agents could be used in simple forms of CHD (without prosthetic valves or hemodynamically

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From: *Department of Cardiology, Baylor–St. Luke's Medical Center and Texas Heart Institute, Houston, Texas 77030*

Address for reprints: *Wilson W. Lam, MD, 6621 Fannin St., MC 19345-C, Houston, TX 77030*

E-mail: *wlam@bcm.edu*

significant valve disease) but endorse a class III contraindication to the use of NOACs in adults who have undergone Fontan surgery.[12]

In 2015, the European Heart Rhythm Association became more progressive in defining the eligibility for NOAC use in valvular AF.[13] They suggest vitamin K antagonists for patients with mechanical prosthetic valves or moderate-to-severe mitral stenosis (often rheumatic), but patients who have mild-to-moderate native-valve disease, severe aortic stenosis, bioprosthetic valves, mitral valve repairs, transcatheter aortic valve replacement, or hypertrophic cardiomyopathy (HCM) are considered eligible for NOACs.[13]

One quarter of the patients in ARISTOTLE (Apixaban versus Warfarin in Patients with Atrial Fibrillation Trial)[14] had moderate or severe valve disease (most often mitral regurgitation). A German series[15] that was presented at the 11th International Congress of Update in Cardiology and Cardiovascular Surgery (2015) had 55 ACHD patients on NOACs, half of whom had greatly complex cases (3 with anatomy that suggested Fontan circulation). In only 1 patient was the NOAC discontinued for bleeding associated with muscular rupture.[15] Ongoing studies should focus on demonstrating the safety and efficacy of these novel medications in this problematic population so that they too might reap the benefit of medical advances.

The 2008 ACC/AHA Guidelines for the Management of Adults with Congenital Heart Disease[16] suggest that pacemaker implantation and catheter ablation should be performed primarily in centers experienced with complex anatomy and surgical repairs. Surgically placed epicardial pacemakers are preferred in the presence of intracardiac shunts. Two interesting guidelines from this document include the role of antitachycardia pacing in possibly reducing tachy-brady episodes in this population, and a IIa indication for cardioverter-defibrillator implantation (ICD) for aborted sudden death or hemodynamically significant ventricular tachycardia. Most comparable guidelines give the latter a class I indication.[16] As examples of challenging anatomy, we reviewed a case of refractory ventricular tachycardia after Medtronic Melody® transcatheter pulmonary valve replacement and a case of superior vena cava baffle obstruction that needed recannulation and stenting before dual-chamber pacemaker placement for sinus node dysfunction. The 2014 PACES/HRS guidelines recommend a IIb indication extrapolating 2008 ACC/AHA/HRS guidelines to the ACHD population using Sudden Cardiac Death in Heart Failure Trial (SCD-HeFT) criteria (that is, single or systemic right ventricle with ejection fraction ≤0.35 and risk factors such as unexplained syncope or New York Heart Association class II or III symptoms).[17]

However, there are still unanswered questions about antitachycardia pacing for ventricular arrhythmias. In 2012, the Multicenter Automatic Defibrillator Implantation Trial–Reduce Inappropriate Therapy (MADIT-RIT) suggested survival benefit and reduced inappropriate shock without increasing syncope when anti-tachycardia pacing and defibrillation therapies were delayed—this in the same year in which a small series (n=79) in children and CHD patients had shown that anti-tachycardia pacing reduced appropriate and inappropriate shocks.[18,19] To avoid the complications of transvenous leads, subcutaneous ICD remains an option. In the Evaluation of Factors Impacting Clinical Outcome and Cost Effectiveness of the S-ICD (EFFORTLESS) registry, 7% of patients had CHD, 13% had an inherited "channelopathy," and 12% had HCM.[20]

The 2011 American College of Cardiology Foundation/AHA guideline for the diagnosis and treatment of HCM moves through a basic algorithm looking at maximal wall thickness, unexplained syncope, family history of sudden death, nonsustained ventricular tachycardia, and hypotension on exercise testing.[21] With these contemporary management strategies, the clinical course of HCM has been altered, resulting in a low disease-related mortality rate of 0.5% per year.[22] Other investigators report that too many defibrillators are being implanted and that primary prevention has a low appropriate-therapy rate (2.4%/yr), outweighed by the 3.8%/year inappropriate-shock rate.[23] Recent suggestions to help guide the ICD-implantation decision include late gadolinium enhancement on cardiac magnetic resonance imaging,[24] age >60 years and a low annual risk of sudden death (0.2%/yr),[25] and a European Sudden Cardiac Death Risk prediction tool.[26] For a risk of greater than 6% over 5 years, the Europeans consider implanting a defibrillator.

In conclusion, growth of the ACHD population has given new importance to our familiarity with guidelines that can advise us when to refer patients to an ACHD center of excellence. Preventive care (in regard to such factors as obesity, sleep apnea, and influenza vaccination) remains central to arrhythmia management. Although the data are still limited, medical management is expanding our indications for ACHD patients. Controversy remains in regard to who will benefit from a defibrillator, but newer tools, we hope, will sharpen our acumen.

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