Top Ten Things to Know
2018 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease

1. The prevalence of adult congenital heart disease (ACHD) is growing due to the success of pediatric cardiology and congenital cardiac surgery in diagnosing and treating congenital heart defects in children. Overall, cardiovascular reasons account for approximately 77% of all deaths in patients with ACHD, with approximately half due to chronic heart failure.

2. This guideline establishes an anatomic and physiologic classification for patients with ACHD to better describe their clinical status and guide follow up and therapeutic decisions, providing specific tables to help determine frequency and type of diagnostic testing.

3. The importance of transition from pediatric to ACHD care and the need to provide access and delivery of high quality lifelong specialized CHD care are described.

4. Women with CHD should receive pre-pregnancy counseling with input from an ACHD cardiologist to determine maternal cardiac, obstetrical and fetal risks, and potential long-term risks to the mother. An individualized plan of care that addresses expectations and contingencies should be developed for and with women with CHD who are pregnant or who may become pregnant and shared with the patient and all caregivers.

5. Therapeutic intervention criteria and algorithms for shunt lesions are established in the guideline. The recommendations consider all the important features of the patient’s anatomy and hemodynamics.

6. Indications for pulmonary valve replacement based on data that has emerged over the last 10 years are described.

7. Initial and follow-up aortic imaging is recommended in adults with coarctation of the aorta, including those who have had surgical or catheter intervention.

8. Primary prevention implantable cardioverter defibrillator (ICD) therapy for repaired Tetralogy of Fallot and patients with multiple risk factors for sudden cardiac death is recommended.

9. Procedures including cardiac surgery, catheter–based interventions and electrophysiologic procedures involving patients with ACHD should be performed by operators with expertise in CHD procedures and in collaboration with ACHD cardiologists.

10. There are multiple challenges to developing evidenced-based care for patients with ACHD. Comprehensive multicenter and population registries and databases are needed to have adequate numbers of patients to address clinical questions. Novel study methodologies are needed to ascertain effectiveness of diagnostic and therapeutic options.