Top Ten Things to Know
Diagnosis and Management of Noncardiac Complications in Adults with Congenital Heart Disease

1. In 2002, there were an estimated 650,000-1,300,000 Americans with congenital cardiovascular defects in the U.S. Also known as congenital heart defects, these structural problems arise from abnormal formation of the heart or major blood vessels.
2. Many patients with repaired simple CHD can live into their 60s or 70s, whereas the patient population with more complex CHD such as Fontan circulation is relatively younger. While the quality of life and life expectancy have both increased for adults with congenital heart defects (CHD), noncardiac complications (e.g., renal dysfunction, restrictive lung disease, anemia, and cirrhosis) significantly contribute to the morbidity and mortality of adults with CHD.
3. This Scientific Statement provides an overview of prevalence, etiology, impact, and management of several noncardiac complications in adults with congenital heart defects.
4. Renal dysfunction is common in adults with all forms of CHD; this is associated with 2-5-fold increase in 6-year mortality for mild to moderate/severe dysfunction. Renal function can be assessed at regular intervals and considered in planning for interventional or surgical procedures.
5. Several studies have reported lower pulmonary function and higher pulmonary complications in Adults with CHD compared to healthy controls. Treatments such as pulmonary rehabilitation, supplemental oxygen, and others are discussed.
6. The prevalence of liver disease among patients with CHD is poorly characterized and difficult to estimate because the prevalence is likely to vary between types of CHD and liver disease is in many cases subclinical and undiagnosed. Contributing causes of liver disease in the adult CHD population can be divided broadly into two categories: those related to hemodynamic derangements (i.e. venous congestion, ischemic injury) and those related to non-hemodynamic factors (i.e. viral hepatitis, drug-induced liver injury).
7. The statement provides a detailed overview of several immunology and infectious disease, hematology, endocrine, vascular, and psychosocial complications that can occur in adults with CHD.
8. Cancer is the fourth leading cause of mortality after heart failure, pneumonia, and sudden cardiac death in a large registry of adults with CHD, with an increasing proportion of death from noncardiac causes with older age. The use of low-dose ionizing radiation from medical diagnostics and procedures is a risk factor for carcinogenesis and its use in the management of CHD has increased steadily over time. The most common types of cancer in adults with CHD mirror those of the general population: breast, colorectal, and uterine for women and prostate, colorectal, and bladder for men.
9. CHD encompasses a heterogenous group of conditions, often with multiorgan involvement, which can lead to unique risk factors for surgical complications. Regional ACHD centers have specialists familiar with and experienced in the management of those with CHD.
10. As adults with CHD grow in number and age, preventative strategies with intervention at an earlier age may help to mitigate the development of later non-cardiac complications. Adults with CHD often have complex multi-systemic disease for which both cardiac outcomes and noncardiac complications warrant clinical attention and empirical investigation; an interdisciplinary approach is required across the lifespan of these patients.

Lui GK, Saidi A, Bhatt AB, Burchill LJ, Deen JF, Earing MG, et al; on behalf of the American Heart Association Adult Congenital Heart Disease Committee of the Council on Clinical Cardiology and Council on Cardiovascular Disease in the Young; Council on Cardiovascular Radiology and Intervention; and Council on Quality of Care and Outcomes Research. Diagnosis and management of noncardiac complications in adults with congenital heart disease: a scientific statement from the American Heart Association [published online ahead of print October 9, 2017]. *Circulation*. doi: 10.1161/CIR.0000000000000535.