

# IM Care for Patients with Congenital Heart Defects

## Point-of-Care Tool

### OVERVIEW

---

- ♥ Most children with congenital heart defects (CHDs) live into adulthood.
- ♥ Although mortality has improved significantly, morbidity remains a considerable problem.
- ♥ Since 2000, more adults than children are living in the United States with CHDs: (<https://www.cdc.gov/ncbddd/heartdefects/features/kf-chd-estimates-us.html>)
- ♥ Many patients with CHDs are lost to congenital cardiology care before they are teenagers; a majority of adults with CHDs have gaps in cardiology care.
- ♥ Ask your patients with CHDs if they are regularly seeing a cardiologist. If not, refer them to an adult congenital cardiologist (<https://www.achaheart.org/your-heart/resources/clinic-directory/>).

### CLASSIFICATION OF CHDs: 2018 AHA/ACC Adult congenital heart disease guidelines (abbreviated list) is based on CHD types and symptoms.

---

- ♥ Simple
  - Patients typically have a normal lifespan compared to the general population.
  - Associated with these CHDs: mild pulmonary stenosis, small unrepaired atrial septal defects (ASDs), small unrepaired ventricular septal defects (VSDs), small unrepaired patent ductus arteriosus (PDA), repaired ASDs, repaired VSDs, and repaired PDA.
- ♥ Moderate complexity
  - Ensure that patients are monitored regularly by an adult congenital cardiologist to identify and treat potential long-term complications which may arise in adulthood.
  - Associated with these CHDs: coarctation of the aorta, anomalous venous return, Tetralogy of Fallot (ToF), and atrioventricular canal.
- ♥ Complex
  - Ensure that patients are monitored frequently by an adult congenital cardiologist to identify and treat morbidity secondary to their CHD.
  - Associated with these CHDs: Transposition of the Great Arteries (TGA), cyanosis, Fontan palliation, and single ventricle physiology.
- ♥ 2018 AHA/ACHD Guideline for the Management of Adults with Congenital Heart Disease: Executive Summary provides details on specific types of CHDs (<https://www.ahajournals.org/doi/10.1161/CIR.000000000000602>)



## COMMON PRESENTATIONS

---



### Repaired or palliated patients

- Utilize social work, case management, and contact adult and pediatric cardiology specialists at major regional centers to ensure expedited follow-up.
- Note that some patients may present with exercise intolerance, arrhythmias, or heart failure.



### Unrepaired patients

- Although most patients with CHDs are identified in childhood, a small number of patients are diagnosed as adults.
- Be aware of patients who present with upper extremity hypertension, murmurs or abnormal heart sounds, cyanosis or clubbing, arrhythmias, or heart blocks.
- Also look for patients with partial anomalous venous return or bicuspid aortic valve with dilated ascending aorta identified incidentally on CT or MRI.

## LONG-TERM CARDIAC COMPLICATIONS

---



Arrhythmias – Adults with CHDs are at higher risk of developing arrhythmias, a major source of morbidity.

- Intraatrial reentrant tachycardia (IART) or atypical atrial flutter are the most frequent types of arrhythmia in adults with CHDs. Tachycardia may be scar-mediated and can be faster or slower than typical atrial flutter.
  - Anticoagulation – The complexity of the patient's CHD is a better tool than CHAD-VASC score to risk-stratify a patient's need for anticoagulation.
  - Use this approach unless the patient has a simple CHD. For these patients, CHAD-VASC scores are best for decision-making.
- Ventricular tachycardia and heart block are also common arrhythmias in this population.
- CHD type determines common related arrhythmias. See the 2020 European Society of Cardiology Adult Congenital Heart Disease Guidelines for more information (<https://academic.oup.com/eurheartj/article/42/6/563/5898606>)



Heart failure – Patients with CHDs and heart failure are younger and have longer admissions than the general heart failure population. Heart failure is the number one cause of death in patients with CHDs older than 40 years.



### Increased risk of endocarditis

- Subacute bacterial endocarditis (SBE) prophylaxis is recommended in specific ACHD populations.
- Ensure that all patients with CHDs have regular dental care as a preventive measure.









### Pulmonary hypertension

- Patients with CHDs may develop pulmonary hypertension from large or moderate unrepaired shunts.
- Make sure that patients with residual shunts avoid air in their IVs to prevent paradoxical emboli and stroke.






## NON-CARDIAC COMPLICATIONS

---

-  Note that many patients with CHDs have long-term comorbidities that are not cardiac-related. The Diagnosis and Management of Non-cardiac Complications in Adults with CHD: A Scientific Statement From the AHA summarizes these complications (<https://www.ahajournals.org/doi/10.1161/CIR.0000000000000535>)
-  Recommend a preoperative evaluation for patients with CHDs. Depending on the severity of their CHD, you may need to refer them to a cardiac anesthesiologist who is familiar with their CHD.
-  Note that patients with CHDs may need procedures performed at an ACHD referral center.
-  Perform age-appropriate cancer screening and provide referrals when needed.
-  Primary care physicians should screen patients with CHDs for depression and anxiety; young adults with CHDs are 4-5 times more likely to have mental health-related concerns.
-  Many patients need a multidisciplinary approach with multiple adult subspecialists. The 2020 European Society of Cardiology Adult Congenital Heart Disease Guidelines describe the need for multidisciplinary care (<https://academic.oup.com/eurheartj/article/42/6/563/5898606>)




## REPRODUCTIVE HEALTH

---

-  Encourage patients with certain CHDs who are at increased risk of blood clots to avoid estrogen-containing contraceptive methods (ie, combined birth control pills, patches, or rings). Provide contraceptive counseling for all women of childbearing age.
-  Ensure all patients of childbearing age receive prepregnancy counseling with an ACHD physician and OB/MFM physician to assess cardiac health, determine whether any procedures are needed before pregnancy, and discuss the safety of procedures or medications.
-  See the related AAP course for more information: **OBGYN Care for Patients with Congenital Heart Defects** (<https://aap.arche.services/curriculum/9c8d8c19-0420-4e5a-8518-0e4644063doc/activity/90c0e05d-1122-43e1-8bdf-1a3f500e8f95>).

## SPECIALIZED CARE CENTERS

---

-  Recommend care at specialized care centers when appropriate; research shows that referral centers specializing in adult CHD care have improved survival rates.
-  If advanced cardiac imaging or interventions are required, refer patients to specialized care centers.
-  Review the following site for a list of adult CHD centers (<https://www.achaheart.org/your-heart/resources/clinic-directory/>)

This project is supported by the Centers for Disease Control and Prevention of the U.S. Department of Health and Human Services (HHS) as part of a financial assistance award totaling \$400,000 with 100 percent funded by CDC/HHS. The contents are those of the author(s) and do not necessarily represent the official views of, nor an endorsement, by American Academy of Pediatrics, CDC/HHS, or the U.S. Government.

