What a General Cardiologist Should Know About Adult Congenital Heart Disease

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Disclosures

• I have no relevant financial disclosures or conflicts of interest
Cardiology Boards Blueprint

• Congenital Heart Disease 5%
  • Congenital malformations of cardiac chambers and connections <2%
  • Congenital malformations of cardiac septa <2%
  • Congenital malformations of pulmonary and tricuspid valves <2%
  • Congenital malformations of aortic and mitral valves <2%
  • Other congenital malformations of the heart <2%
  • Congenital malformations of the great arteries <2%
  • Congenital malformations of the great veins <2%
  • Congenital disorders with cardiovascular implications <2%
  • Eisenmenger syndrome <2%
2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines

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Key Words: AHA Scientific Statements • arrhythmias • cardiac catheterization • cardiac defects • congenital heart disease • congenital heart surgery • unrepaired/repair defect

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e698 April 2, 2019
Outline

• Adult Congenital Heart Disease (ACHD) burden
• ACHD Anatomic and Physiological (AP) Classification

• Pregnancy and Contraception
• Cyanosis and Eisenmenger syndrome
• Endocarditis Prevention
• Exercise and Sports
• Mental Health and Neurodevelopmental Issues
• Concomitant Syndromes
• Noncardiac Medical Issues
• Noncardiac Surgery
• Heart Failure and Transplant
• Important congenital coronary anomalies
Congenital Heart Disease (CHD)

• Most common congenital disorder of newborns
  • ~1% of all live births

• Critical CHD (requiring intervention in first year of life)
  • ~25% of all CHD

• Advances in cardiovascular medicine and surgery
  • 85-95% survive to adulthood
Changing Mortality in CHD
Mortality in CHD 1987-2005 in Quebec

CHD Burden

Modified from Webb G et al. Int J Cardiol. 2015;195:326-333
Changing prevalence of CHD in the European Union by age group

Numbers European Union (Population 497 Mill. in 2008)

- **Prevalence** 5.1 %* - 5.2 % ** (2012/13)
  - Estimated prevalence 11% (2030)

- **Prevalence** 0.5 %* (2000)

- **ACHD Patients > 60 years**
- **ACHD Patients < 60 years**
- **Children with CHD**

Extrapolation

** German Competence Network for Congenital Heart Disease (data on file)
U.S. Estimates of CHD

- More adults with CHD than children with CHD
- >1.3 million adults with CHD living in the US in 2010
- 20,000 new adult CHD patients per year. 5% increase per year
Trends in ACHD

Annual ACHD Admissions in the US
## Comparative Frequencies of Pediatric and ACHD

<table>
<thead>
<tr>
<th>Pediatric Congenital Heart Disease</th>
<th>Adult Congenital Heart Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Defect</strong></td>
<td><strong>Defect</strong></td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>Atrial or ventricular septal defects</td>
</tr>
<tr>
<td>35%</td>
<td>22%</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>Tetralogy of Fallot</td>
</tr>
<tr>
<td>9%</td>
<td>14%</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>Complex disease (eg, Fontan)</td>
</tr>
<tr>
<td>8%</td>
<td>13%</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>Obstruction of the left ventricular outflow tract</td>
</tr>
<tr>
<td>8%</td>
<td>12%</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>Transposition of the great arteries</td>
</tr>
<tr>
<td>6%</td>
<td>10%</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>Obstruction of the right ventricular outflow tract</td>
</tr>
<tr>
<td>6%</td>
<td>8%</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>Coarctation of the aorta</td>
</tr>
<tr>
<td>5%</td>
<td>7%</td>
</tr>
<tr>
<td>Transposition of great vessels</td>
<td>Marfan syndrome</td>
</tr>
<tr>
<td>4%</td>
<td>5%</td>
</tr>
<tr>
<td>Atrioventricular septal defect</td>
<td>Corrected transposition of the great vessels</td>
</tr>
<tr>
<td>3%</td>
<td>4%</td>
</tr>
<tr>
<td>Other rarer conditions (eg, tricuspid atresia, bicuspid valves, univentricular heart)</td>
<td>Atrioventricular septal defect</td>
</tr>
<tr>
<td>16%</td>
<td>3%</td>
</tr>
<tr>
<td>Eisenmenger syndrome</td>
<td></td>
</tr>
<tr>
<td>2%</td>
<td></td>
</tr>
</tbody>
</table>

5-year mortality rates in ACHD patients compared with general UK population

<table>
<thead>
<tr>
<th>Patient's age (years)</th>
<th>20</th>
<th>25</th>
<th>30</th>
<th>35</th>
<th>40</th>
<th>45</th>
<th>50</th>
<th>55</th>
<th>60</th>
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</thead>
<tbody>
<tr>
<td>ASD</td>
<td>25</td>
<td>26</td>
<td>32</td>
<td>38</td>
<td>42</td>
<td>47</td>
<td>52</td>
<td>57</td>
<td>61</td>
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<tr>
<td>Valvar disease</td>
<td>29</td>
<td>31</td>
<td>36</td>
<td>40</td>
<td>45</td>
<td>49</td>
<td>54</td>
<td>59</td>
<td>63</td>
</tr>
<tr>
<td>VSD</td>
<td>28</td>
<td>30</td>
<td>36</td>
<td>40</td>
<td>44</td>
<td>49</td>
<td>53</td>
<td>59</td>
<td>63</td>
</tr>
<tr>
<td>Aortic Coarctation</td>
<td>32</td>
<td>33</td>
<td>38</td>
<td>43</td>
<td>47</td>
<td>52</td>
<td>56</td>
<td>62</td>
<td>66</td>
</tr>
<tr>
<td>AVSD</td>
<td>33</td>
<td>34</td>
<td>39</td>
<td>44</td>
<td>48</td>
<td>52</td>
<td>57</td>
<td>62</td>
<td>66</td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>37</td>
<td>38</td>
<td>42</td>
<td>46</td>
<td>50</td>
<td>54</td>
<td>59</td>
<td>64</td>
<td>68</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>37</td>
<td>38</td>
<td>42</td>
<td>47</td>
<td>50</td>
<td>54</td>
<td>60</td>
<td>65</td>
<td>69</td>
</tr>
<tr>
<td>Ebstein anomaly</td>
<td>42</td>
<td>43</td>
<td>47</td>
<td>51</td>
<td>54</td>
<td>59</td>
<td>63</td>
<td>68</td>
<td>72</td>
</tr>
<tr>
<td>Systemic RV</td>
<td>46</td>
<td>48</td>
<td>51</td>
<td>55</td>
<td>59</td>
<td>63</td>
<td>67</td>
<td>72</td>
<td>76</td>
</tr>
<tr>
<td>Eisenmenger syndrome</td>
<td>57</td>
<td>58</td>
<td>62</td>
<td>65</td>
<td>69</td>
<td>73</td>
<td>77</td>
<td>81</td>
<td>84</td>
</tr>
<tr>
<td>Complex CHD</td>
<td>58</td>
<td>59</td>
<td>63</td>
<td>67</td>
<td>70</td>
<td>74</td>
<td>78</td>
<td>82</td>
<td>85</td>
</tr>
<tr>
<td>Fontan</td>
<td>64</td>
<td>65</td>
<td>68</td>
<td>72</td>
<td>75</td>
<td>78</td>
<td>82</td>
<td>86</td>
<td>91</td>
</tr>
</tbody>
</table>

Age difference:
- >40
- 30-40
- 20-30
- 10-20
- 5-10
- 2-5
- <2
Physiological Classification of CHD

Acyanotic
- With L-R shunt e.g. ASD
- Without L-R shunt e.g. valvular disease

Cyanotic
- Ductal-dependent pulmonary blood flow e.g. pulmonary atresia
- Ductal-dependent systemic blood flow e.g. HLHS
Severity of ACHD

- Severity depends on
  - Native Anatomy
  - Surgical Repair
  - Current Physiology

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease): Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

AHA/ACC Guideline

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: Executive Summary: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines

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Predictability of Mortality from Different ACHD Classifications

**All-cause mortality**

![Graph A](image)

- **ACHD AP**: 0.71, 0.0378
- **CHD anatomy**: 0.67, 0.0353
- **NYHA**: 0.71, 0.0404
- **CHDFI**: 0.74, 0.0254
- **CHD anatomy+NYHA**: 0.76, 0.0344
- **CHD anatomy+CHDFI**: 0.78, 0.0282

**Cardiac mortality**

![Graph B](image)

- **ACHD AP**: 0.75, 0.0375
- **CHD anatomy**: 0.64, 0.0445
- **NYHA**: 0.65, 0.0515
- **CHDFI**: 0.76, 0.0268
- **CHD anatomy+NYHA**: 0.72, 0.0453
- **CHD anatomy+CHDFI**: 0.78, 0.0327

*P-values for pairwise comparisons adjusted for false discovery rate*
# ACHD AP Classification: Anatomic

## I: Simple

<table>
<thead>
<tr>
<th>Native disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isolated small ASD</td>
</tr>
<tr>
<td>Isolated small VSD</td>
</tr>
<tr>
<td>Mild isolated pulmonic stenosis</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Repaired conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Previously ligated or occluded ductus arteriosus</td>
</tr>
<tr>
<td>Repaired secundum ASD or sinus venosus defect without significant residual shunt or chamber enlargement</td>
</tr>
<tr>
<td>Repaired VSD without significant residual shunt or chamber enlargement</td>
</tr>
</tbody>
</table>

*ACHD Guidelines 2018 Circulation. 2019;139:e698–e800*
**ACHD AP Classification: Anatomic**

<table>
<thead>
<tr>
<th>III: Great Complexity (or Complex)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cyanotic congenital heart defect (unrepaired or palliated, all forms)</td>
</tr>
<tr>
<td>Double-outlet ventricle</td>
</tr>
<tr>
<td>Fontan procedure</td>
</tr>
<tr>
<td>Interrupted aortic arch</td>
</tr>
<tr>
<td>Mitral atresia</td>
</tr>
<tr>
<td>Single ventricle (including double inlet left ventricle, tricuspid atresia, hypoplastic left heart, any other anatomic abnormality with a functionally single ventricle)</td>
</tr>
<tr>
<td>Pulmonary atresia (all forms)</td>
</tr>
<tr>
<td>TGA (classic or d-TGA; CCTGA or l-TGA)</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
</tr>
<tr>
<td>Other abnormalities of atrioventricular and ventriculoarterial connection (ie, crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)</td>
</tr>
</tbody>
</table>
# ACHD AP Classification: Anatomic

## II: Moderate Complexity

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Repaired or unrepaired conditions</td>
<td></td>
</tr>
<tr>
<td>Aorto-left ventricular fistula</td>
<td></td>
</tr>
<tr>
<td>Anomalous pulmonary venous connection, partial or total</td>
<td></td>
</tr>
<tr>
<td>Anomalous coronary artery arising from the pulmonary artery</td>
<td></td>
</tr>
<tr>
<td>Anomalous aortic origin of a coronary artery from the opposite sinus</td>
<td></td>
</tr>
<tr>
<td>AVSD (partial or complete, including primum ASD)</td>
<td></td>
</tr>
<tr>
<td>Congenital aortic valve disease</td>
<td></td>
</tr>
<tr>
<td>Congenital mitral valve disease</td>
<td></td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td></td>
</tr>
<tr>
<td>Ebstein anomaly (disease spectrum includes mild, moderate, and severe variations)</td>
<td></td>
</tr>
<tr>
<td>Infundibular right ventricular outflow obstruction</td>
<td></td>
</tr>
<tr>
<td>Ostium primum ASD</td>
<td></td>
</tr>
<tr>
<td>Moderate and large unrepaired secundum ASD</td>
<td></td>
</tr>
<tr>
<td>Moderate and large persistently patent ductus arteriosus</td>
<td></td>
</tr>
<tr>
<td>Pulmonary valve regurgitation (moderate or greater)</td>
<td></td>
</tr>
<tr>
<td>Pulmonary valve stenosis (moderate or greater)</td>
<td></td>
</tr>
<tr>
<td>Peripheral pulmonary stenosis</td>
<td></td>
</tr>
<tr>
<td>Sinus of Valsalva fistula/aneurysm</td>
<td></td>
</tr>
<tr>
<td>Sinus venosus defect</td>
<td></td>
</tr>
<tr>
<td>Subvalvar aortic stenosis (excluding HCM; HCM not addressed in these guidelines)</td>
<td></td>
</tr>
<tr>
<td>Supravalvar aortic stenosis</td>
<td></td>
</tr>
<tr>
<td>Straddling atrioventricular valve</td>
<td></td>
</tr>
<tr>
<td>Repaired tetralogy of Fallot</td>
<td></td>
</tr>
<tr>
<td>VSD with associated abnormality and/or moderate or greater shunt</td>
<td></td>
</tr>
</tbody>
</table>
Physiological Variables

End-Organ Dysfunction
- Liver
- Lungs
- Kidney

Pulmonary HTN
- PH: Mean PAP >25mmHg
- PAH: Mean PAP >25 mmHg
- PCWP <15mmHg, PVR >3 WU

Exercise Performance
- Max ventilatory equivalent of O2 below range expected for the specific CHD

Shunt
- Chamber enlargement
- Qp:Qs>1.5:1
- If none, then mild

Venous/Arterial Stenosis
- Recoarctation, supravalvarAS/PS
- Venous baffle obstruction, Branch PA stenosis, pulm vein stenosis
- Cavopulmonary stenosis
## Physiological Variables

### Arrhythmia
- None
- Not requiring Treatment
- Controlled with Treatment
- Refractory

### Aortopathy
- Mild: 3.5 - 3.9 cm
- Moderate: 4.0 - 4.9 cm
- Severe: > 5.0 cm

### Valvular Heart Disease
- Mild, Mod, Severe
- 2014 VHD guidelines

### Hypoxemia/Cyanosis
- pOx < 90%
- Severe hypoxemia < 85%
- Cyanosis: > 5g/dl of de-saturated Hb

### NYHA Class
- I – no limitations
- II – mild limitations with ordinary activities
- III – marked limitations with ordinary activities
- IV – limitation at rest
## ACHD AP Classification: Physiological Stage

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>A</strong></td>
<td></td>
</tr>
<tr>
<td>NYHA FC I symptoms</td>
<td></td>
</tr>
<tr>
<td>No hemodynamic or anatomic sequelae</td>
<td></td>
</tr>
<tr>
<td>No arrhythmias</td>
<td></td>
</tr>
<tr>
<td>Normal exercise capacity</td>
<td></td>
</tr>
<tr>
<td>Normal renal/hepatic/pulmonary function</td>
<td></td>
</tr>
<tr>
<td><strong>B</strong></td>
<td></td>
</tr>
<tr>
<td>NYHA FC II symptoms</td>
<td></td>
</tr>
<tr>
<td>Mild hemodynamic sequelae (mild aortic enlargement, mild ventricular enlargement, mild ventricular dysfunction)</td>
<td></td>
</tr>
<tr>
<td>Mild valvular disease</td>
<td></td>
</tr>
<tr>
<td>Trivial or small shunt (not hemodynamically significant)</td>
<td></td>
</tr>
<tr>
<td>Arrhythmia not requiring treatment</td>
<td></td>
</tr>
<tr>
<td>Abnormal objective cardiac limitation to exercise</td>
<td></td>
</tr>
</tbody>
</table>
## ACHD AP Classification: Physiological Stage

<table>
<thead>
<tr>
<th>Stage</th>
<th>Symptoms</th>
</tr>
</thead>
</table>
| **C** | NYHA FC III symptoms  
Significant (moderate or greater) valvular disease; moderate or greater ventricular dysfunction (systemic, pulmonic, or both)  
Moderate aortic enlargement  
Venous or arterial stenosis  
Mild or moderate hypoxemia/cyanosis  
Hemodynamically significant shunt  
Arrhythmias controlled with treatment  
Pulmonary hypertension (less than severe)  
End-organ dysfunction responsive to therapy |
| **D** | NYHA FC IV symptoms  
Severe aortic enlargement  
Arrhythmias refractory to treatment  
Severe hypoxemia (almost always associated with cyanosis)  
Severe pulmonary hypertension  
Eisenmenger syndrome  
Refractory end-organ dysfunction |
Most frequent non-cardiovascular admission diagnoses in ACHD

- Blood-forming organs
- Endocrine diseases
- Respiratory system
- Neoplasms
- External injuries
- Genitourinary system
- Nervous system
- Musculoskeletal system
- Digestive system
- Pregnancy / delivery
Normal Cardiovascular Changes in Pregnancy

Mehta LS et al. Circulation. 2020;CIR00000000000000772. doi:10.1161/CIR.00000000000000772
Maternal Cardiovascular Risk Stratification Score

- Canadian Cardiac Disease in Pregnancy (CARPREG) score

- Zwangerschap bij Aangeboren HARtAfwijkingen (ZAHARA; translated in English as “Pregnancy and Congenital Heart Disease”) score

- **Modified WHO** Classification of Maternal Cardiovascular Risk
  - most reliable predictor of maternal CV complications
    - arrhythmias and heart failure
# Modified WHO Classification of Maternal Cardiovascular Risk

<table>
<thead>
<tr>
<th>WHO Pregnancy Risk Category</th>
<th>Risk Description</th>
<th>Maternal Risk Factors</th>
</tr>
</thead>
</table>
| I                          | No detectable increase in maternal mortality and no/mild increase in morbidity risk | Uncomplicated small/mild pulmonary stenosis, PDA, mitral valve prolapse  
Successful repair of simple lesions (ASD, VSD, PDA, anomalous pulmonary venous drainage)  
Atrial or ventricular ectopic beats, isolated |
| II                         | Small increase in maternal mortality and moderate increase in morbidity risk | If otherwise well and uncomplicated:  
Unoperated ASD, VSD  
Repaired TOF  
Most arrhythmias |
| II-III                     | Moderate increase in maternal mortality morbidity risk |  
Mild LV impairment  
Hypertrophic cardiomyopathy  
Native or tissue valvular disease (not considered risk category I or II)  
Marfan syndrome without aortic dilation  
Aortic dilation <45 mm in bicuspid aortic valve aortopathy  
Repaired coarctation |
|                            |                  | Significantly increased maternal mortality or severe morbidity risk. Expert counseling required. In the event of pregnancy, intensive specialist cardiac and obstetric monitoring needed throughout pregnancy, childbirth, and the puerperium. |

- Mechanical valve  
- Systemic RV  
- Fontan circulation  
- Cyanotic heart disease (unrepaired)  
- Other complex CHD  
- Aortic dilation 40–45 mm in Marfan syndrome  
- Aortic dilation 45–50 mm in bicuspid aortic valve aortopathy |

- Pulmonary arterial hypertension (of any cause)  
- Severe systemic ventricular dysfunction (LV ejection fraction <30%, NYHA class III–IV)  
- Previous peripartum cardiomyopathy with any residual impairment of LV function  
- Severe mitral stenosis, severe symptomatic aortic stenosis  
- Aortic dilation >45 mm in Marfan syndrome  
- Aortic dilation >50 mm in bicuspid aortic valve aortopathy  
- Native severe coarctation |
# Recommendations for Pregnancy

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>RECOMMENDATIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>C-LD</td>
<td>1. Women with CHD should receive <em>prepregnancy counseling</em> with input from an ACHD cardiologist to determine maternal cardiac, obstetrical and fetal risks, and potential long-term risks to the mother.</td>
</tr>
<tr>
<td>I</td>
<td>C-LD</td>
<td>2. An <em>individualized plan of care</em> 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.</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>3. Women with CHD receiving <em>chronic anticoagulation</em> should be counseled, ideally before conception, on the risks and benefits of specific anticoagulants during pregnancy.</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>4. Women with ACHD AP classification IB-D, IIA-D, and IIIA-D* should be <em>managed collaboratively</em> during pregnancy by ACHD cardiologists, obstetricians, and anesthesiologists experienced in ACHD.</td>
</tr>
<tr>
<td>I</td>
<td>C-EO</td>
<td>5. In collaboration with an ACHD cardiologist to ensure <em>accurate assessment of pregnancy risk</em>, patients at <em>high risk</em> of maternal morbidity or mortality, including women with pulmonary arterial hypertension (PAH), Eisenmenger syndrome, severe systemic ventricular dysfunction, severe left-sided obstructive lesions, and/or ACHD AP classification ID, IID, IIDD should be <em>counseled against becoming pregnant</em> or be given the option of terminating pregnancy.</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>6. Men and women of childbearing age with CHD should be counseled on the risk of CHD recurrence in offspring.</td>
</tr>
<tr>
<td>IIA</td>
<td>B-NR</td>
<td>7. <em>Exercise testing</em> can be useful for risk assessment in women with ACHD AP classification IC-D, IIA-D, and IIIA-D* who are considering pregnancy.</td>
</tr>
<tr>
<td>IIA</td>
<td>B-NR</td>
<td>8. When either parent has CHD, it is reasonable to perform <em>fetal echocardiography</em>.</td>
</tr>
</tbody>
</table>

COR: Class of Recommendation; LOE: Level of Evidence; *Circulation*. 2019;139:e698–e800
Assessment and Management
Women with Complex Congenital Heart Disease

Preconception maternal–fetal risk assessment
Complete history and physical exam
Prior health/surgical records obtained and reviewed
Baseline 12-lead electrocardiogram, echocardiogram, laboratory studies
Cardiopulmonary exercise tolerance and functional class
Medical or surgical repair as indicated
Genetic referral for patients with a heritable cardiac lesion
Discuss discontinuation of teratogenic drugs prior to conception
Appropriate contraception provided until pregnancy desired

Prenatal Assessment upon confirmation of pregnancy
Ensure teratogenic medications: ACE, ARB, have been discontinued;
If pregnancy was unplanned conduct full maternal–fetal risk assessment and diagnostic evaluation; discuss plan of care (POC) including where to be delivered

Pregnancy in patient who is clinically stable
Joint management by ACHD and MFM team; establish POC for patients geographically distant from tertiary center
Regular cardiac and OB follow-up
Repeat echocardiogram per trimester as indicated
Fetal echocardiogram 18-22 weeks

Pregnancy in patient with severe ventricular dysfunction (NYHA III or IV), cyanosis, pulmonary hypertension:
Discuss maternal–fetal morbidity and mortality risk and outline plan of care including possible need for early hospitalization.
Joint decision regarding pregnancy by patient, family cardiologist, MFM team

Written delivery plan prepared and distributed to team by 28-32 weeks
Vaginal delivery with epidural preferred; C-section reserved for obstetric reasons
Induction of labor as indicated ~39 weeks
Antibiotic prophylaxis as indicated

Terminate Pregnancy
Appropriate contraception or sterilization

Joint management by experienced MFM and ACHD teams at tertiary center
Multidisciplinary conference after 26 weeks to discuss L&D management
Close team follow-up increasing as indicated by clinical status
Delivery date determined by clinical status; C-section may be indicated

Circulation. 2017;135:e50-e87. DOI: 10.1161/CIR.0000000000000458
**Normal Electrocardiographic Changes Associated With Pregnancy**

<table>
<thead>
<tr>
<th><strong>Observation</strong></th>
<th><strong>Description</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Left axis shift is seen, with the greatest shift in the third trimester</td>
<td>caused by elevation of the diaphragm.</td>
</tr>
<tr>
<td>Shortening of the PR, QRS, and QT intervals may accompany the increase in</td>
<td>resting heart rate.</td>
</tr>
<tr>
<td>Nonspecific ST abnormalities, including segment depression or flattened and</td>
<td>inverted T waves in lead III, occur frequently.</td>
</tr>
<tr>
<td>inverted T waves in lead III, occur frequently.</td>
<td></td>
</tr>
</tbody>
</table>
Fetal Risk with Maternal CHD

• Higher frequency of
  • spontaneous abortions (15% and 25%) and intrauterine fetal demise

• fetal CHD - fetal echocardiography at 18 to 22 weeks

• higher preterm birth rate (10%–12%), especially in those with complex CHD (22%–65%)

• neonatal events, for example, small for gestational age, respiratory distress syndrome, interventricular hemorrhage, and neonatal death (27.8%)

• perinatal mortality
  • >4-fold higher than in the general population
  • common with premature delivery or recurrence of CHD
  • highest in patients with Eisenmenger syndrome (27.7%)
AHA SCIENTIFIC STATEMENT

Management of Pregnancy in Patients With Complex Congenital Heart Disease

A Scientific Statement for Healthcare Professionals From the American Heart Association

*Circulation*. 2017;135:e50-e87. DOI: 10.1161/CIR.0000000000000458
## Recommendations for Contraception

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>RECOMMENDATIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>C-LD</td>
<td>1. Women of childbearing potential with CHD should be <strong>counseled</strong> about the risks associated with pregnancy and appropriate contraceptive options</td>
</tr>
<tr>
<td>III: Harm</td>
<td>B-NR</td>
<td>2. <strong>Estrogen-containing contraceptives</strong> are potentially harmful for women with CHD who are at high risk of thromboembolic events (e.g., cyanosis, Fontan physiology, mechanical valves, prior thrombotic events, PAH)</td>
</tr>
</tbody>
</table>

- Low-dose combination oral contraceptive (≤20 mcg of ethinyl estradiol) is an option except in women who are at increased risk of thrombosis
- Medroxyprogesterone acetate: less effective, fluid retention
- Intrauterine devices: highly effective, may experience vasovagal reactions at the time of implant
- Tubal ligation: generally safe, risks associated with anesthesia and abdominal insufflation
- Vasectomy for the male partner: long-term prognosis of the female CHD patient must be considered and discussed
- Unplanned pregnancy with desire for termination: the morning-after pill (levonorgestrel) is safe for women, but acute fluid retention is a risk
Cyanosis

• “blueish discoloration of the skin and/or mucous membranes resulting from inadequate oxygenation of the blood.”

Cardiac causes:

• Insufficient pulmonary blood flow
• Contamination of arterial blood by venous blood
  • right-to-left shunt
  • increased venous admixture

Eur Heart J 2014;35(21):1410
Chronic Cyanosis: Effects on CV System

- Myocardial ischemia risk (myocardial perfusion, micro-occlusion)
  - HCT>65% may decrease DO2
  - Hyperviscosity increase myocardial work

- Endothelial injury → imbalance of vasodilators/vasoconstrictors (Nitric oxide, Prostacylin) → BP, myocardial work, perfusion

- Myocardial gene expression alterations
  - Up-regulation of genes assoc with apoptosis, remodeling
  - Down-regulation of genes assoc with myocardial contractility

- Risk of systolic and diastolic dysfunction
Chronic Cyanosis: Effects on Other Organs

• Coagulation
  • Bleeding risk:
    • Decrease von Willebrand factor
    • Relative thrombocytopenia, decrease platelet life & function
  • Thrombosis risk – stiff RBC

• Neurologic
  • Cerebral abscess (R→L shunt)

• Respiratory
  • Blunted hypoxic ventilatory drive
  • Hyperventilation
  • ETCO2 may underestimate PaCO2 (R-to-L shunts)
Chronic Cyanosis: Effects on Other Organs

- Renal
  - Chronic hypoxemia proliferative lesions in glomeruli
  - Thickening of basal membrane
  - Proteinuria, increase uric acid

- Hematological:
  - Secondary erythrocytosis
  - Iron deficiency
Symptomatic Erythrocytosis

• Secondary erythrocytosis: a physiological increase in red blood cell mass in response to hypoxemia

• Hematocrit > 65% may be associated with hyperviscosity

• Hyperviscosity symptoms
  • Headache
  • faintness or dizziness, visual disturbance
  • fatigue, muscle aches, joint aches
  • paraesthesia
  • easy bruising, epistaxis, gingival bleeding
  • gout
  • poor mental function

Can mimic iron deficiency complicating treatment
Iron Deficiency in Cyanotic Individuals

- One-third of cyanotic patients are iron deficient.

- Microcytosis and hypochromia are unreliable markers of iron deficiency.

- Iron deficiency exacerbated by phlebotomies, GI bleed and menorrhagia.

- Annual assessment of serum iron, ferritin, and transferrin levels.

- Treatment of transferrin saturation <20% with iron supplementation until iron stores are replete can be done safely.
Treatment for Hyperviscosity

• **Routine** prophylactic phlebotomy to maintain an arbitrary hematocrit **should not be done**
  • Worsens iron deficiency
  • Increases risk of strokes
  • Worsens exercise capacity
  • Stimulates erythropoietin
  • Increases viscosity over time
  • Inhibits O2 delivery to tissues
  • Does not reliably improve symptoms
  • attack/stroke)
Treatment for Hyperviscosity

- Patients with suspected hyperviscosity
  - rehydrated either with oral fluids or intravenous normal saline solution as a first-line therapy

- evaluated for iron deficiency, and treated if appropriate

- phlebotomy (with equal volume fluid replacement) is sometimes performed in special cases wherein, after adequate hydration, hematocrit remains higher than the patient’s baseline and symptoms persist, or there is evidence of end-organ damage attributable to hyperviscosity (e.g., myocardial ischemia, transient ischemic attack/stroke)
Specific Management Practices for Cyanotic CHD

- Recording clinical **oxygen saturation at rest (>5 min)** rather than immediately after effort.
- Meticulous **intravenous care to avoid air** or particulate matter, which may include use of air/particulate filters on all intravenous access lines, when feasible, and careful de-airing of all lines.
- Cerebral imaging for any **new headache or neurologic sign** to assess for possible cerebral abscess, hemorrhage, or stroke.
- Measurement of serum uric acid and treatment with allopurinol in a patient with a history of **gout**.
- **Non-estrogen–containing birth control** for women of child-bearing potential (intrauterine device may be a preferred option). Avoidance of birth control entirely is not a safe, acceptable option.
- Patients can travel safely on commercial airlines without undue risk, although **adequate hydration and movement during the flight** are appropriate.
- Measurement of **coagulation parameters** (e.g., activated partial thromboplastin time, INR, thrombin time) in a patient with an elevated hematocrit >55% requires adjustment of anticoagulant volume in the blood collection vials to account for reduced plasma volume in the draw.
Pulmonary Hypertension

- Pulmonary hypertension
  - Mean PA pressure by right heart cath ≥ 25 mmHg at rest
- Pulmonary arterial hypertension
  - mean PA pressure by right heart cath ≥ 25 mmHg at rest AND
  - pulmonary capillary wedge pressure ≤ 15 mmHg
  - pulmonary vascular resistance ≥ 3 Wood units

- Pulmonary venous congestion
  - Obstruction
  - High left atrial pressures

- Large, unrestrictive L-to-R shunt
  - Increase flow through the pulmonary vasculature
  - Exposure of pulmonary vasculature to systemic pressures
Pulmonary Hypertension and ACHD

• Poorer prognosis in presence of PH

• PAH may develop years after shunt closure in patients with ACHD

• Predictors for the development or presence of PAH include:
  a. Anatomic defects: complete AVSD, sinus venosus defect, large non-restrictive defect (ASD >2 cm, VSD >1 cm, PDA >0.6 cm), and concomitant ACHD AP classification II or III abnormalities
  b. Preintervention Qp:Qs ≥ 3 and/or PASP >40 mm Hg
  c. Presence of associated syndrome (e.g., Down syndrome)
  d. Older age at repair
  e. Female sex
  f. Otherwise unexplained symptoms potentially attributable to PAH (decreased exercise capacity, syncope, chest pain, hemoptysis)
  g. clinical examination findings: hypoxia, elevated systemic venous pressure, fluid retention, loud P2, new TR or PR, new arrhythmia, decreased exercise capacity, EKG changes
# Recommendations for Severe PAH

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>RECOMMENDATIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>B-NR</td>
<td><strong>Diagnostic</strong></td>
</tr>
<tr>
<td>1.</td>
<td></td>
<td>Patients with ACHD with pulmonary vascular resistance 2.5 Wood units or greater (≥4 Wood units × m²) should be assessed collaboratively by an ACHD cardiologist and an expert in pulmonary hypertension to develop a management plan.</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>3. Adults with septal or great artery shunts should undergo periodic screening for pulmonary hypertension with TTE.</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>4. Cardiac catheterization to assess pulmonary vascular hemodynamics is recommended for adults with septal or great artery shunts and clinical symptoms, signs, or echocardiographic findings suggestive of pulmonary hypertension.</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>5. In adults with septal or great artery shunts, cardiac catheterization with hemodynamics (performed before or at time of closure) is beneficial to assess suitability for closure.</td>
</tr>
<tr>
<td>I</td>
<td>C-EO</td>
<td>6. BNP, chest x-ray, 6-minute walk test, and cardiac catheterization are useful for initial and follow-up evaluation of patients with ACHD with PAH.</td>
</tr>
</tbody>
</table>

**COR:** Class of Recommendation; **LOE:** Level of Evidence; *J Am Coll Cardiol* 2019;73:e81–192
Eisenmenger Syndrome and ACHD

- Caused by elevated pulmonary vascular resistance driving right-to-left intracardiac or great arterial shunting leading to systemic arterial desaturation.

- Should become more rare

- Natural course and outcomes remain incompletely defined
# Recommendations for Eisenmenger Syndrome

<table>
<thead>
<tr>
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<th>RECOMMENDATIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>C-EO</td>
<td>1. When evaluating adults with presumed Eisenmenger syndrome, clinicians should confirm diagnostic imaging and cardiac catheterization data accuracy and exclude other potential contributors to right-to-left shunting or pulmonary hypertension.</td>
</tr>
<tr>
<td>I</td>
<td>A</td>
<td>2. <strong>Bosentan</strong> is beneficial in symptomatic adults with Eisenmenger syndrome with ASD or VSD.</td>
</tr>
<tr>
<td>Ila</td>
<td>B-R</td>
<td>3. In symptomatic adults with Eisenmenger syndrome, <strong>bosentan and PDE-5 inhibitors</strong> are reasonable in combination if symptomatic improvement does not occur with either medication alone.</td>
</tr>
<tr>
<td>Ila</td>
<td>C-EO</td>
<td>4. <strong>Bosentan</strong> is a reasonable therapy to treat symptomatic adults with Eisenmenger syndrome with 1 of the following: shunts other than ASD/VSD (e.g., PDA, aortopulmonary window) (Level of Evidence C-EQ), or complex congenital heart lesion or Down syndrome (Level of Evidence B-NR).</td>
</tr>
<tr>
<td>Ila</td>
<td>B-NR</td>
<td>6. It is reasonable to use <strong>PDE-5 inhibitors</strong> (e.g., sildenafil, tadalafil) to treat symptomatic adults with Eisenmenger syndrome with ASD, VSD, or great artery shunt.</td>
</tr>
</tbody>
</table>

**Diagnostic**

**Therapeutic**

COR: Class of Recommendation; LOE: Level of Evidence; J Am Coll Cardiol 2019:73:e81–192
ACHD Heart Failure


ACHD Heart Failure

• Heart failure:
  • significant issue in patients with ACHD
  • common
  • multifactorial
  • associated with morbidity and mortality
  • anticipated to increase in prevalence

• However, there are no/limited data to support treatment recommendations

<table>
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<tr>
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</thead>
<tbody>
<tr>
<td>I</td>
<td>C-LD</td>
<td>1. Consultation with ACHD and HF specialists is recommended for patients with ACHD and HF or severe ventricular dysfunction.</td>
</tr>
</tbody>
</table>

COR: Class of Recommendation; LOE: Level of Evidence; Circulation. 2019;139:e698–e800
ACHD Heart Failure - Systemic Right Ventricle

Medical Therapy for Systemic Right Ventricles: A Systematic Review (Part 1) for the 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines

• Standard guideline-directed management and therapy is ostensibly preferable to no treatment in
  • biventricular physiology
  • systemic left ventricular (LV) dysfunction
  • no repairable residual/reversible hemodynamic abnormalities
  • persistent heart failure symptoms

In conclusion, pooled results across the limited available studies did not provide conclusive evidence with regard to a beneficial effect of medical therapy in adults with systemic RV dysfunction. Randomized controlled trials or comparative-effectiveness studies that are sufficiently powered to demonstrate effect are needed to elucidate the efficacy of ACE inhibitors, ARBs, beta blockers, and aldosterone antagonists in patients with systemic RVs.
ACHD Heart Transplantation

• Specific criteria for timing of referral: universal recommendations cannot be made based on current data

• Recommend early ACHD transplant center

• Risks for poor outcomes: single ventricle anatomy, anatomic complexity, protein-losing enteropathy, or high titers of panel reactive antibodies

• Heart-lung transplants are performed internationally each year, with a median survival of 3.3 years and 10-year survival of 32%. Survival worse because of longer wait times.
# ACHD Heart Transplantation

<table>
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<td>I</td>
<td>C-LD</td>
<td>1. Consultation with ACHD and HF specialists is recommended for patients with ACHD and HF or severe ventricular dysfunction.</td>
</tr>
</tbody>
</table>
ACHD- Infective Endocarditis Prophylaxis

Cardiac Conditions Associated With the Highest Risk of Adverse Outcome From Endocarditis for Which Prophylaxis is Recommended:

• Those with previous IE;

• Patients with prosthetic valves (biological and mechanical, surgical and transcatheter);

• Patients within 6 months of placement of prosthetic material (whether placed by surgery or by catheter intervention);

• Patients with residual intracardiac shunts at the site of or adjacent to previous repair with prosthetic material or devices; or

• Patients with uncorrected cyanotic heart disease

• Cardiac transplantation recipients who develop cardiac valvulopathy
# Exercise and Sports

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>C-LD</td>
<td>1. Clinicians should <em>assess</em> activity levels at regular intervals and <em>counsel</em> patients with ACHD about the types and intensity of exercise appropriate to their clinical status.</td>
</tr>
<tr>
<td>Ila</td>
<td>C-LD</td>
<td>2. <em>CPET</em> can be useful to guide activity recommendations for patients with ACHD.</td>
</tr>
<tr>
<td>Ila</td>
<td>B-NR</td>
<td>3. <em>Cardiac rehabilitation</em> can be useful to increase exercise capacity in patients with ACHD.</td>
</tr>
</tbody>
</table>

COR: Class of Recommendation; LOE: Level of Evidence; *Circulation*. 2019;139:e698–e800
### Mental Health and Neurodevelopmental Issues

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>B-NR</td>
<td>1. Patients with ACHD should be evaluated for depression and anxiety.</td>
</tr>
<tr>
<td>IIA</td>
<td>B-NR</td>
<td>2. Referral for mental health evaluation and treatment is reasonable in patients with ACHD.</td>
</tr>
<tr>
<td>IIB</td>
<td>B-NR</td>
<td>3. Neurodevelopmental or neuropsychological testing may be considered in some patients with ACHD to guide therapies that enhance academic, behavioral, psychosocial, and adaptive functioning.</td>
</tr>
</tbody>
</table>

COR: Class of Recommendation; LOE: Level of Evidence; Circulation. 2019;139:e698–e800
### Underlying Genetic Syndromes Commonly Associated With CHD

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Genetic Abnormality</th>
<th>Clinical Features</th>
<th>Common Cardiac Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>DiGeorge syndrome (velocardiofacial syndrome)</td>
<td>22q11.2 deletion</td>
<td>Thymic and parathyroid hypoplasia, immunodeficiency, low-set ears, hypocalcemia, speech and learning disorders, renal anomalies, psychiatric disease</td>
<td>IAA type B, aortic arch anomalies, truncus arteriosus, TOF</td>
</tr>
<tr>
<td></td>
<td></td>
<td>25%–75% have CHD, depending on age studied&lt;sup&gt;53-9,7,11-9&lt;/sup&gt;</td>
<td></td>
</tr>
<tr>
<td>Down syndrome</td>
<td>Trisomy 21</td>
<td>Developmental disability, characteristic facial features, hypotonia, palmar crease</td>
<td>ASD, VSD, AVSD, TOF</td>
</tr>
<tr>
<td></td>
<td></td>
<td>40%–50% have CHD</td>
<td></td>
</tr>
<tr>
<td>Holt–Oram syndrome&lt;sup&gt;53-9,9&lt;/sup&gt;</td>
<td>TBX5</td>
<td>Upper limb skeletal abnormalities</td>
<td>ASD, VSD, MV disease</td>
</tr>
<tr>
<td></td>
<td></td>
<td>75% have CHD</td>
<td></td>
</tr>
<tr>
<td>Klinefelter syndrome</td>
<td>47 XXY</td>
<td>Tall stature, hypoplastic testes, delayed puberty, developmental disability</td>
<td>PDA, ASD, MV prolapse</td>
</tr>
<tr>
<td></td>
<td></td>
<td>50% have CHD</td>
<td></td>
</tr>
<tr>
<td>Noonan syndrome&lt;sup&gt;53-9,10&lt;/sup&gt;</td>
<td>PTPN11, KRAS, SOS1, RAF1, NRAS, BRAF, MAP2K1</td>
<td>Facial anomalies, webbed neck, chest deformity, short stature, lymphatic abnormalities, bleeding abnormalities</td>
<td>PS, ASD, HCM</td>
</tr>
<tr>
<td></td>
<td></td>
<td>80% have CHD</td>
<td></td>
</tr>
<tr>
<td>Turner syndrome</td>
<td>45X</td>
<td>Short stature, webbed neck, lymphedema, primary amenorrhea</td>
<td>Coarctation, BAV, aortic stenosis, hypoplastic left heart, ascending aortopathy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>30% have CHD</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Risk of aortic dissection</td>
<td></td>
</tr>
<tr>
<td>Williams syndrome</td>
<td>7q11.23 deletion</td>
<td>Elfin face, social personality, hearing loss, developmental delay, infantile hypercalcemia</td>
<td>Supravalvar aortic stenosis, peripheral PS</td>
</tr>
<tr>
<td></td>
<td></td>
<td>50%–80% have CHD</td>
<td></td>
</tr>
</tbody>
</table>

ASD indicates atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CHD, congenital heart disease; HCM, hypertrophic cardiomyopathy; IAA, interrupted aortic arch; MV, mitral valve; PDA, patent ductus arteriosus; PS, pulmonary stenosis; TOF, tetralogy of Fallot; and VSD, ventricular septal defect.
## Concomitant Syndromes

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendation</th>
</tr>
</thead>
<tbody>
<tr>
<td>IIA</td>
<td>B-NR</td>
<td>1. Genetic testing for 22q11 deletions is reasonable for patients with conotruncal cardiac defects.</td>
</tr>
</tbody>
</table>

COR: Class of Recommendation; LOE: Level of Evidence; *Circulation*. 2019;139:e698–e800
### Noncardiac Medical Issues in ACHD

<table>
<thead>
<tr>
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<th>Recommendation</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>C-LD</td>
<td>1. Patients with ACHD at risk for hepatitis C should be screened and vaccinated for viral hepatitis and treated as appropriate.</td>
</tr>
</tbody>
</table>

- Universal screening for hepatitis C began in 1992
- Blood exposure during cardiac surgery
- Hepatitis vaccination and/or consultation with a hepatologist should also be offered where appropriate, particularly in patients with ACHD with concomitant liver disease (e.g., Fontan patients)
### Noncardiac Surgery in ACHD

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>C-LD</td>
<td>1. Optimization before and close surveillance after invasive procedures, regardless of the complexity of the anatomic defect or type of procedure is beneficial for patients with ACHD.</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>2. In patients with ACHD AP classification IB-D, IIA-D, and IIIA-D* noncardiac surgical and interventional procedures should be performed in a hospital with or in consultation with experts in ACHD when possible.</td>
</tr>
</tbody>
</table>

COR: Class of Recommendation; LOE: Level of Evidence; Circulation. 2019;139:e698–e800
Noncardiac Surgery in ACHD

Clarify CHD diagnosis

- Clarify prior procedures, residua, sequelae, and current status, including ACHD AP classification
- Be aware that history obtained from only the patient and family may be faulty or incomplete
- Obtain and review old records to ensure accurate understanding of past procedures and clinical course
- Complete additional investigations required to define ACHD AP classification
- Develop management strategies to minimize risk and optimize outcome

Factors associated with increased risk of perioperative morbidity and mortality

- Cyanosis
- Congestive HF
- Poor general health
- Younger age
- Pulmonary hypertension
- Operations on the respiratory and nervous systems
- Complex CHD
- Urgent/emergency procedures

Issues to consider:

- Endocarditis prophylaxis
- Complications related to underlying hemodynamics
- Abnormal venous and/or arterial anatomy affecting venous and arterial access
- Persistent shunts
- Valvular disease
- Arrhythmias, including bradyarrhythmias
- Erythrocytosis
- Pulmonary vascular disease
- Meticulous line care (also consider air filters for intravenous lines) to reduce risk of paradoxical embolus in patients who are cyanotic because of right-to-left shunts
- Adjustment of anticoagulant volume in tubes for some blood work in cyanotic patients
- Prevention of venous thrombosis
- Monitoring of renal and liver function
- Periprocedure anticoagulation
- Possible need for nonconventional drug dosing
- Increased prevalence of hepatitis C infection because of prior procedures and remote blood transfusions
- Developmental disability

ACHD indicates adult congenital heart disease; AP, anatomic and physiological; CHD, congenital heart disease; and HF, heart failure.
Anomalous Aortic Origin of the Coronary Artery

Anomalous Aortic Origin of the Coronary Artery

Anomalous aortic origin of the coronary artery

Left coronary from the right sinus

- Ischemic symptoms or ischemia during diagnostic testing
  - Yes: Surgical intervention* (Class I)
  - No: Surgical Intervention* (Class Ila)

Right coronary from the left sinus

- Ischemic symptoms or ischemia during diagnostic testing
  - Yes: Surgical intervention* (Class I)
  - No: Ventricular arrhythmias
    - Yes: Surgical intervention* (Class Ila)
    - No: Continued observation (Class IIb)
Anomalous Coronary Artery Arising From the Pulmonary Artery

Anomalous Origin of Left Coronary Artery from the Pulmonary Artery (ALCAPA)

Anomalous Coronary Artery Arising From the Pulmonary Artery

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
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<tr>
<td>Therapeutic</td>
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<tr>
<td>I</td>
<td>B-NR</td>
<td>1. Surgery is recommended for anomalous left coronary artery from the PA.</td>
</tr>
<tr>
<td>I</td>
<td>C-EO</td>
<td>2. In a symptomatic adult with anomalous right coronary artery from the PA with symptoms attributed to the anomalous coronary, surgery is recommended.</td>
</tr>
<tr>
<td>IIa</td>
<td>C-EO</td>
<td>3. Surgery for anomalous right coronary artery from the PA is reasonable in an asymptomatic adult with ventricular dysfunction or with myocardial ischemia attributed to anomalous right coronary artery from the PA.</td>
</tr>
</tbody>
</table>
Diverse ACHD Lesions and Associated Long-Term Complications

What a General Cardiologist Should Know About Adult Congenital Heart Disease

1. Track the medical history
2. Be aware of the outcome
3. Look for the expected
4. Be prepared for the unexpected
5. Use the right imaging technique
6. Deal with emergencies in the right way
7. Understand and discuss the implications and impact of pregnancy
8. Seek expert opinion


• Khairy P, et al. PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease: developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD). Heart Rhythm. 2014;11(10):e102-e165. doi:10.1016/j.hrthm.2014.05.009