2018 Guideline for the Management of Adults with Congenital Heart Disease

GUIDELINES MADE SIMPLE

A Selection of Tables and Figures

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2018 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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The 2018 ACHD guideline is a full revision of the "2008 ACC/AHA Guidelines for the Management of Adults with Congenital Heart Disease", which was the first U.S. guideline to be published on the topic. This revision uses the 2008 ACHD guideline as a framework and incorporates new data and growing ACHD expertise to develop recommendations.

The following resource contains tables and figures from the 2018 Guideline for the Management of Adults with Congenital Heart Disease. The resource is only an excerpt from the Guideline and the full publication should be reviewed for more tables and figures as well as important context.

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2018 Guideline for the Management of Adults with Congenital Heart Disease

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Physiological Variables as Used in ACHD AP Classification *(1 of 4)*

Variable	Description
Aortopathy	Aortic enlargement is common in some types of CHD and after some repairs. Aortic enlargement may be progressive over a lifetime. There is no universally accepted threshold or repair, nor is the role of indexing to body size clearly defined in adults, as is done in pediatric populations. For purposes of categorization and timing of follow-up imaging: • Mild aortic enlargement is defined as maximum diameter 3.5–3.9 cm • Moderate aortic enlargement is defined as maximum diameter 4.0–4.9 cm • Severe aortic enlargement is defined as maximum diameter ≥5.0 cm

Arrhythmia	Arrhythmias are very common in patients with ACHD and may be both the cause and consequence of deteriorating hemodynamics, valvular dysfunction, or ventricular dysfunction. Arrhythmias are associated with symptoms, outcomes, and prognosis, thus are categorized based on presence and response to treatment.
	• No arrhythmia–no documented clinically relevant atrial or ventricular tachyarrhythmias
	• Arrhythmia not requiring treatment–bradyarrhythmia, atrial or ventricular tachyarrhythmia not requiring antiarrhythmic therapy, cardioversion, or ablation
	• Arrhythmia controlled with therapy:
	  ◦ Bradyarrhythmia requiring pacemaker implantation
	  ◦ Atrial or ventricular tachyarrhythmia requiring antiarrhythmic therapy, cardioversion, or ablation
	  ◦ AF and controlled ventricular response
	  ◦ Patients with an ICD
	• Refractory arrhythmias:
	  ◦ Atrial or ventricular tachyarrhythmia not currently responsive to or refractory to antiarrhythmic therapy or ablation

Concomitant VHD	Severity defined according to the 2014 VHD guideline • Mild VHD • Moderate VHD • Severe VHD

Table 3 is continued in the next page. For abbreviations please refer to page 7.

Table 3

Physiological Variables as Used in ACHD AP Classification *(2 of 4)*

Variable	Description
End-organ dysfunction	Clinical and/or laboratory evidence of end-organ dysfunction including • Renal (kidney) • Hepatic (liver) • Pulmonary (lung
Exercise capacity	Patients with ACHD are often asymptomatic notwithstanding exercise limitations demonstrated as diminished exercise capacity when evaluated objectively. Thus, assessment of both subjective and objective exercise capacity is important (see NYHA classification system below). Exercise capacity is associated with prognosis. • Abnormal objective cardiac limitation to exercise is defined as an exercise maximum ventilatory equivalent of oxygen below the range expected for the specific CHD anatomic diagnosis. • Expected norms for CPET values should take into account age, sex, and underlying congenital diagnosis. Published studies with institution-specific norms can be used as guides, bearing in mind variability among institutional norms and ranges.
Hypoxemia/ hypoxia/ cyanosis	See Section 3.16 for detailed definition of cyanosis. • Hypoxemia is defined as oxygen saturation measured by pulse oximetry at rest ≤90%. • Severe hypoxemia is defined as oxygen saturation at rest <85%. • In patients with normal or high hemoglobin concentrations, severe hypoxemia will be associated with visible cyanosis (which requires ≥5g/L desaturated hemoglobin to be appreciated). • The terms cyanosis and hypoxemia (or hypoxia) are sometimes used interchangeably. Such interchangeability would not apply; however, in the presence of anemia, when severe hypoxemia can be present without visible cyanosis.

Table 3 is continued in the next page. For abbreviations please refer to page 7.

Table 3

Physiological Variables as Used in ACHD AP Classification *(3 of 4)*

Variable	Description
NYHA functional classification system	**Class** — **Functional Capacity** **I** — Patients with cardiac disease but resulting in no limitation of physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain. **II** — Patients with cardiac disease resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity results in fatigue, palpitation, dyspnea, or anginal pain. **III** — Patients with cardiac disease resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity causes fatigue, palpitation, dyspnea, or anginal pain. **IV** — Patients with cardiac disease resulting in inability to carry on any physical activity without discomfort. Symptoms of HF or the anginal syndrome may be present even at rest. If any physical activity is undertaken, discomfort increases.
Pulmonary hypertension	Pulmonary hypertension is a broad term that encompasses pulmonary arterial hypertension, which is pulmonary hypertension with increased pulmonary vascular resistance. This document defines PH and PAH as they are used in the field of pulmonary hypertension. Pulmonary hypertension is defined: • Mean PA pressure by right heart catheterization ≥25 mm Hg. PAH is defined: • Mean PA pressure by right heart catheterization ≥25 mm Hg and a pulmonary capillary wedge pressure ≤15 mm Hg and pulmonary vascular resistance ≥3 Wood units

Table 3 is continued in the next page. For abbreviations please refer to page 7.

Table 3

Physiological Variables as Used in ACHD AP Classification *(4 of 4)*

Variable	Description
Shunt (hemo-dynamically significant shunt)	An intracardiac shunt is hemodynamically significant if: • There is evidence of chamber enlargement distal to the shunt • And/or evidence of sustained Qp:Qs ≥1.5:1 • An intracardiac shunt not meeting these criteria would be described as small or trivial
Venous and arterial stenosis	• Aortic recoarctation after CoA repair • Supravalvular aortic obstruction • Venous baffle obstruction • Supravalvular pulmonary stenosis • Branch PA stenosis • Stenosis of cavopulmonary connection • Pulmonary vein stenosis

Table 3

ACHD indicates adult congenital heart disease; AF, atrial fibrillation; AP, anatomic and physiologic; CHD, congenital heart disease; CoA, coarctation of the aorta; CPET, cardiopulmonary exercise test; HF, heart failure; ICD, implantable cardioverter-defibrillator; NYHA, New York Heart Association; PA, pulmonary artery; PAH, pulmonary arterial hypertension; Qp:Qs, pulmonary–systemic blood flow ratio; and VHD, valvular heart disease.

ACHD Anatomic and Physiological (AP) Classification (CHD Anatomy + Physiological Stage = ACHD AP Classification)

(1 of 3)

CHD Anatomy*
I: Simple
Native disease • Isolated small ASD • Isolated small VSD • Mild isolated pulmonic stenosis **Repaired conditions** • Previously ligated or occluded ductus arteriosus • Repaired secundum ASD or sinus venosus defect without significant residual shunt or chamber enlargement • Repaired VSD without significant residual shunt or chamber enlargement
II: Moderate Complexity
Repaired or unrepaired conditions • Aorto-left ventricular fistula • Anomalous pulmonary venous connection, partial or total • Anomalous coronary artery arising from the pulmonary artery • Anomalous aortic origin of a coronary artery from the opposite sinus • AVSD (partial or complete, including primum ASD) • Congenital aortic valve disease • Congenital mitral valve disease • Coarctation of the aorta • Ebstein anomaly (disease spectrum includes mild, moderate, and severe variations) • Infundibular right ventricular outflow obstruction • Ostium primum ASD • Moderate and large unrepaired secundum ASD • Moderate and large persistently patent ductus arteriosus • Pulmonary valve regurgitation (moderate or greater) • Pulmonary valve stenosis (moderate or greater) • Peripheral pulmonary stenosis • Sinus of Valsalva fistula/aneurysm • Sinus venosus defect • Subvalvar aortic stenosis (excluding HCM; HCM not addressed in these guidelines) • Supravalvar aortic stenosis • Straddling AV valve • Repaired tetralogy of Fallot • VSD with associated abnormality and/or moderate or greater shunt

CHD Anatomy *will continue in the next page. For abbreviations please refer to page 10.*

*This list is not meant to be comprehensive; other conditions may be important in individual patients.

Table 4

ACHD Anatomic and Physiological (AP) Classification (CHD Anatomy + Physiological Stage = ACHD AP Classification)

(2 of 3)

CHD Anatomy*
III: Great Complexity (or Complex)
• Cyanotic congenital heart defect (unrepaired or palliated, all forms) • Double-outlet ventricle • Fontan procedure • Interrupted aortic arch • Mitral atresia • Single ventricle (including double inlet left ventricle, tricuspid atresia, hypoplastic left heart, any other anatomic abnormality with a functionally single ventricle) • Pulmonary atresia (all forms) • TGA (classic or d-TGA; CCTGA or l-TGA) • Truncus arteriosus • Other abnormalities of AV and ventriculoarterial connection (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)
Physiological Stage
A
• NYHA FC I symptoms • No hemodynamic or anatomic sequelae • No arrhythmias • Normal exercise capacity • Normal renal/hepatic/pulmonary function
B
• NYHA FC II symptoms • Mild hemodynamic sequelae (mild aortic enlargement, mild ventricular enlargement, mild ventricular dysfunction) • Mild valvular disease • Trivial or small shunt (not hemodynamically significant) • Arrhythmia not requiring treatment • Abnormal objective cardiac limitation to exercise

Physiological Stage *will continue in the next page. For abbreviations please refer to page 10.*

*This list is not meant to be comprehensive; other conditions may be important in individual patients.

Table 4

ACHD Anatomic and Physiological (AP) Classification (CHD Anatomy + Physiological Stage = ACHD AP Classification)
(3 of 3)

Physiological Stage
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

Table 4

ACHD indicates adult congenital heart disease; AP, anatomic and physiologic; ASD, atrial septal defect; AV, atrioventricular; AVSD, atrioventricular septal defect; CCTGA, congenitally corrected transposition of the great arteries; CHD, congenital heart disease; d-TGA, dextro-transposition of the great arteries; FC, functional class; HCM, hypertrophic cardiomyopathy; l-TGA, levo-transposition of the great arteries; NYHA, New York Heart Association; TGA, transposition of the great arteries; and VSD, ventricular septal defect.

Circumstances Where CMR, CCT, TEE, and/or Cardiac Catheterization May be Superior to TTE

- Assessment of RV size and function in repaired TOF, systemic right ventricles, and other conditions associated with RV volume and pressure overload
- Identification of anomalous pulmonary venous connections
- Serial assessment of thoracic aortic aneurysms, especially when the dilation might extend beyond the echocardiographic windows
- Accurate assessment of PA pressure and pulmonary vascular resistance
- Assessment for recoarctation of the aorta
- Sinus venosus defects
- Vascular rings
- Evaluation of coronary anomalies
- Quantification of valvular regurgitation

Table 11

Comparison of Imaging Modalities Useful in ACHD Evaluation

	Radiation Exposure	Relative Cost	Ventricular Volumes/ Function	Valvular Structure/ Function	Coronary Anatomy and Course	Extracardiac Vascular Anatomy
Echocardiography	No	$	++	+++	+/-	+/-
CMR	No	$$	+++	++	++*	+++
CCT	Yes	$$	+*	+	+++	+++
Cardiac Catheterization	Yes	$$	+	++	+++	++

*In specific gated imaging protocols.

Table 9

$ indicates less expensive; $$, more expensive; +/-, possible value; +, good; ++, very good; and +++, excellent.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; and CMR, cardiovascular magnetic resonance.

Specific Management Practices for Cyanotic CHD

- Recording clinical oxygen saturation at rest (>5 min) rather than immediately after effort (e.g., walking into a clinic examination room);
- 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;
- Supplemental oxygen as needed for symptom relief but not to a target oxygen saturation level and not if there is no demonstrable symptomatic benefit;
- Avoidance of or cautious use of therapies that may reduce the patient's hypoxia-mediated drive to ventilation, such as narcotics or, in rare circumstances, excess supplemental oxygen;
- Anesthesia by providers with expertise in anesthesia for patients with ACHD for any noncardiac surgery;
- 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 fly safely on commercial airlines without undue risk. Preflight simulation testing or mandated supplemental oxygen are not usually indicated, though adequate hydration and movement during the flight are appropriate;
- Measurement of coagulation parameters (e.g., activated partial thromboplastin time, international normalized ratio, 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.

Table 12

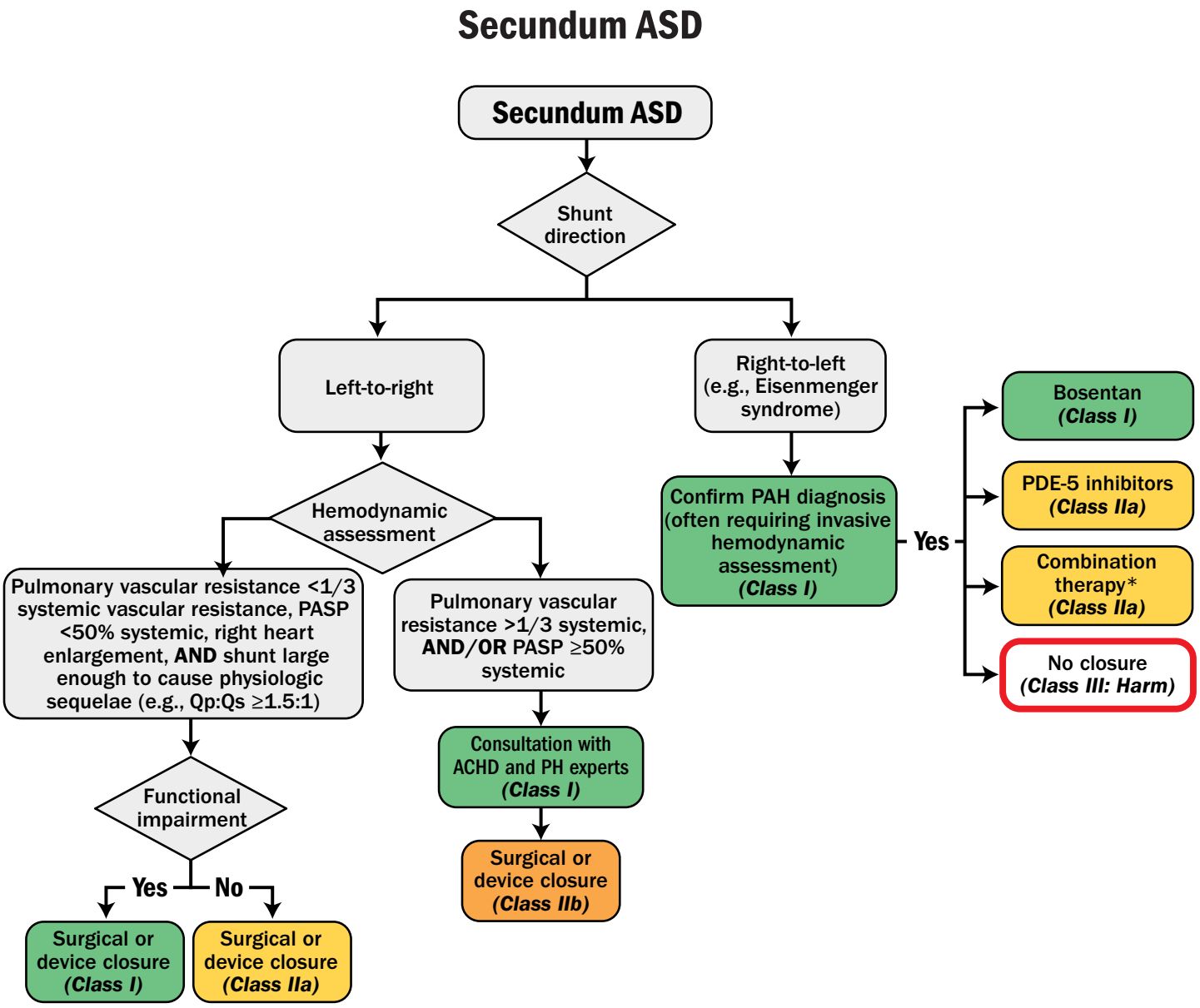


Figure 1

*Combination therapy with bosentan and PDE-5 inhibitor if symptomatic improvement does not occur with either alone.

ACHD indicates adult congenital heart disease; ASD, atrial septal defect; PAH, pulmonary artery hypertension; PASP, pulmonary artery systolic pressure; PDE-5, phosphodiesterase type-5 inhibitors; PH, pulmonary hypertension; and Qp:Qs, pulmonary–systemic blood flow ratio.

Hemodynamically Significant Ventricular Level Shunt

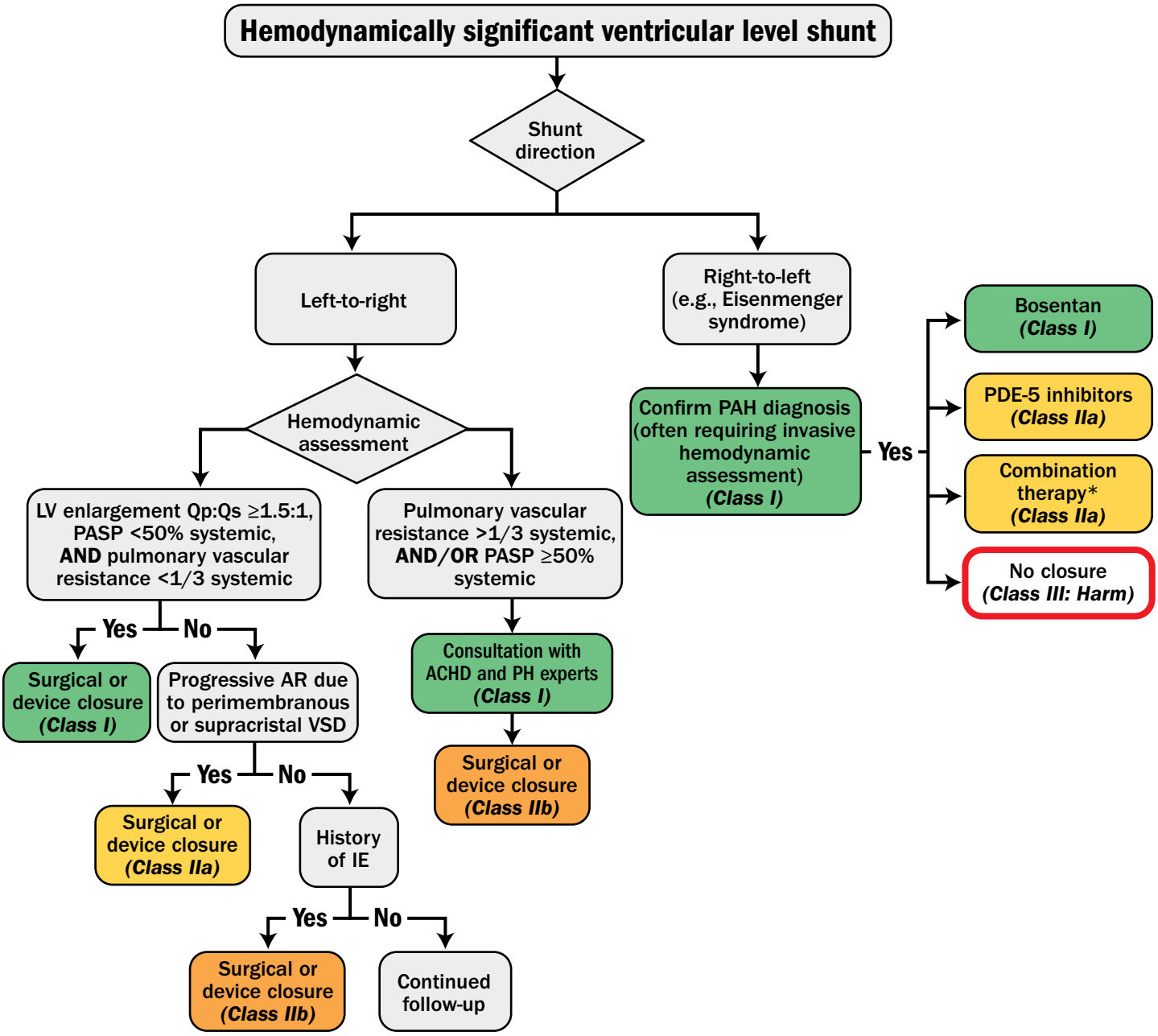


Figure 2

*Combination therapy with bosentan and PDE-5 inhibitor, if symptomatic improvement does not occur with either alone.

ACHD indicates adult congenital heart disease; AR, aortic regurgitation; IE, infective endocarditis; LV, left ventricular; PAH, pulmonary artery hypertension; PASP, pulmonary artery systolic pressure; PDE-5, phosphodiesterase type-5 inhibitors; PH, pulmonary hypertension; Qp:Qs, pulmonary–systemic blood flow ratio; and VSD, ventricular septal defect.

Isolated PR after Repair of PS

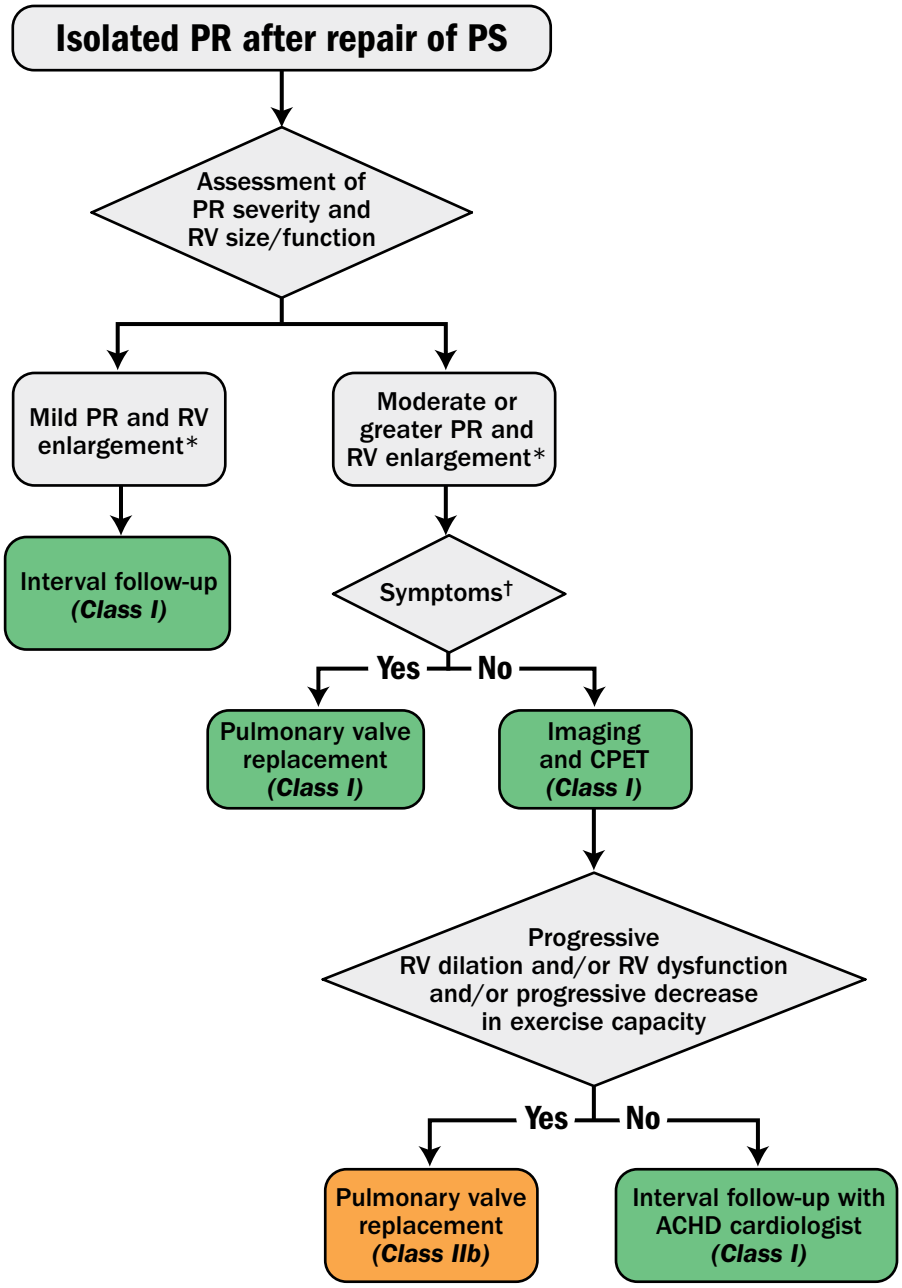


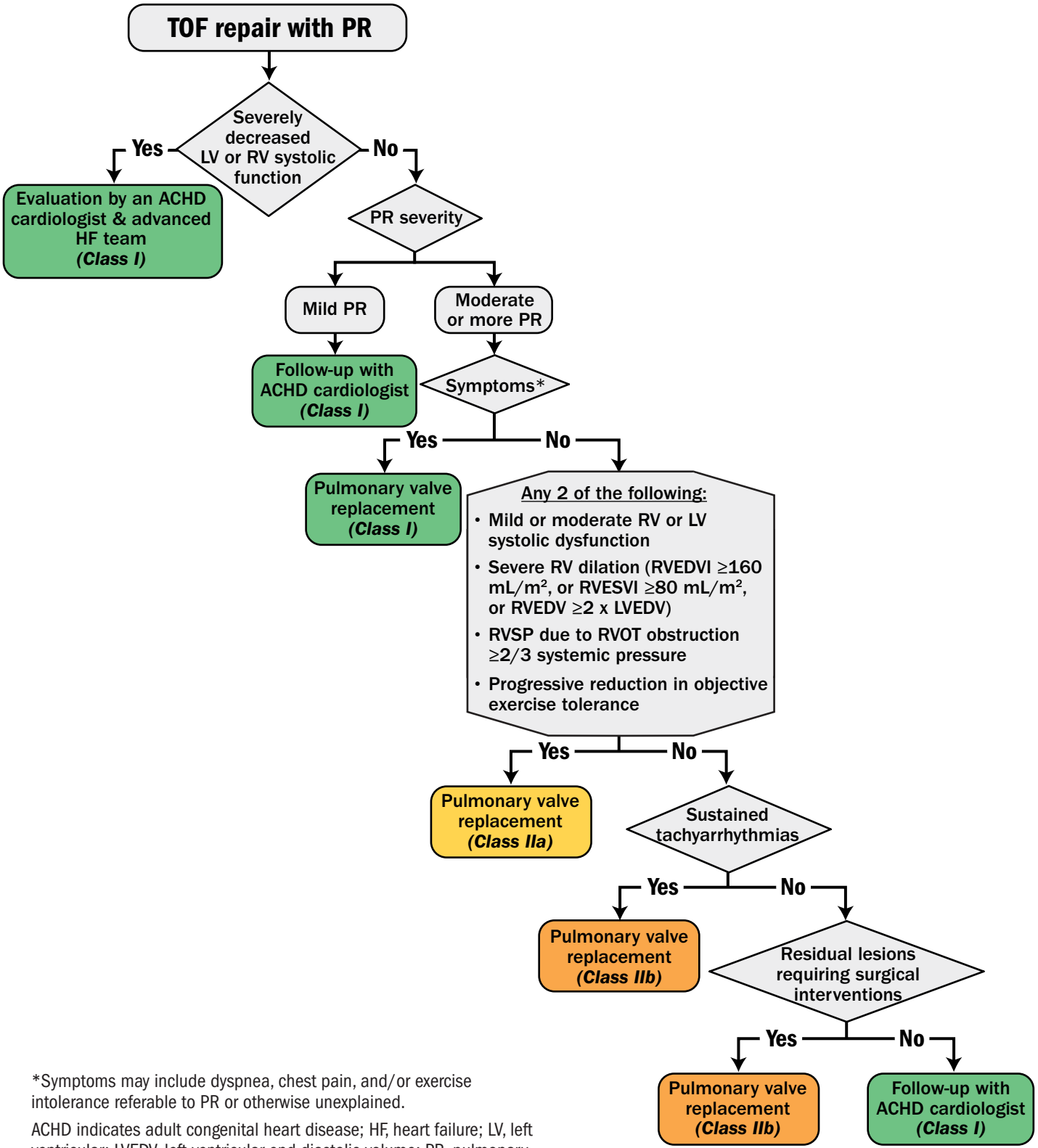
Figure 3

*Significant PR causes RV dilation. If a patient has moderate or greater PR and normal RV size, most likely the estimation of PR severity is inaccurate or there may be restrictive RV physiology, which would warrant further investigation.

†Symptoms may include dyspnea, chest pain, and/or exercise intolerance referable to PR or otherwise unexplained.

ACHD indicates adult congenital heart disease; CPET, cardiopulmonary exercise test; PR, pulmonary regurgitation; PS, pulmonary stenosis; and RV, right ventricular.

Pulmonary Valve Replacement in Patients With TOF Repair and PR



*Symptoms may include dyspnea, chest pain, and/or exercise intolerance referable to PR or otherwise unexplained.

ACHD indicates adult congenital heart disease; HF, heart failure; LV, left ventricular; LVEDV, left ventricular end diastolic volume; PR, pulmonary regurgitation; RV, right ventricular; RVEDV, right ventricular end diastolic volume; RVEDVI, right ventricular end

Figure 4

Anomalous Aortic Origin of the Coronary Artery

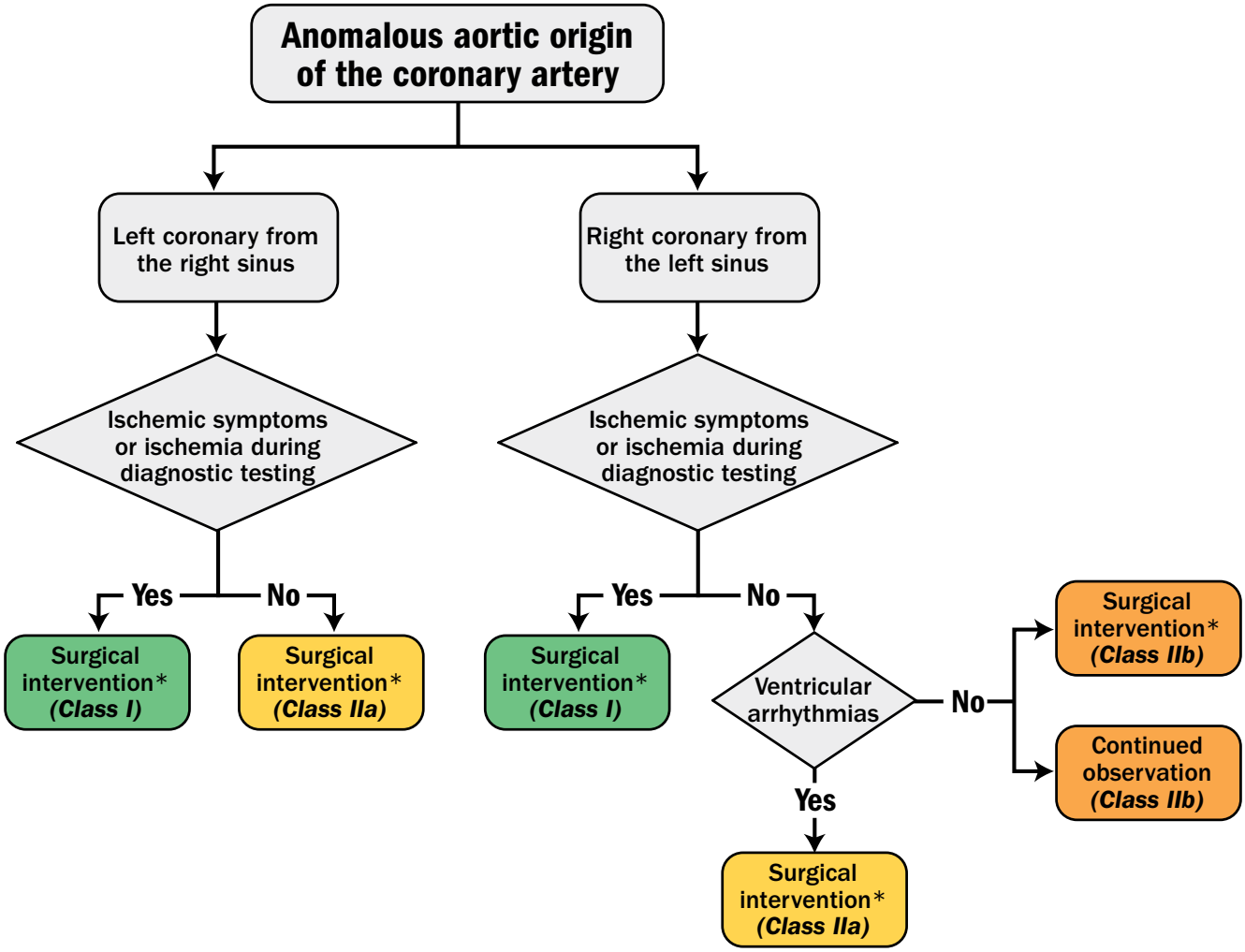


Figure 5

*Surgical intervention to involve unroofing or coronary revascularization for patients with concomitant fixed obstruction.

Routine Follow-Up and Testing Intervals for Specific Conditions

Frequency of Routine Follow-Up and Testing	Physiologic Stage A* (months)	Physiologic Stage B* (months)	Physiologic Stage C* (months)	Physiologic Stage D* (months)
ASD				
Outpatient ACHD Cardiologist	36–60	24	6–12	3–6
ECG	36–60	24	12	12
TTE	36–60	24	12	12
Pulse Oximetry	As needed	As needed	Each visit	Each visit
Exercise Test†	As needed	As needed	12–24	6–12
VSD				
Outpatient ACHD Cardiologist	36	24	6–12	3–6
ECG	36	24	12	12
TTE	36	24	12	12
Pulse Oximetry	As needed	As needed	Each visit	Each visit
Exercise Test†	As needed	As needed	12–24	6–12
AVSD				
Outpatient ACHD Cardiologist	24–36	24	6–12	3–6
ECG	24–36	24	12	12
TTE	24–36	24	12	12
Pulse Oximetry	As needed	As needed	Each visit	Each visit
Exercise Test†	As needed	As needed	12–24	6–12
PDA				
Outpatient ACHD Cardiologist	36–60	24	6–12	3–6
ECG	36–60	24	12	12
TTE	36–60	24	12	12
Pulse Oximetry†	As needed	As needed	Each visit	Each visit
Exercise Test‡	As needed	As needed	12–24	6–12

*See Tables 3 and 4 for details on the ACHD AP classification system.

ASD (Table 13), VSD (Table 14), and AVSD (Table 15):
†6-minute walk test or cardiopulmonary exercise test, depending on the clinical indication.

PDA (Table 16):
†Upper and lower extremity.

‡6-minute walk test or cardiopulmonary exercise test, depending on the clinical indication.

Abbreviations are listed on page 25.

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Routine Follow-Up and Testing Intervals for Specific Conditions

Frequency of Routine Follow-Up and Testing	Physiologic Stage A* (months)	Physiologic Stage B* (months)	Physiologic Stage C* (months)	Physiologic Stage D* (months)
Congenital Mitral Stenosis				
Outpatient ACHD Cardiologist	24	24	6–12	3–6
ECG	24	24	12	12
TTE	24	24	12	12
Exercise Test†	As needed	24	24	12
SubAS				
Outpatient ACHD Cardiologist	24	24	6–12	3–6
ECG	24	24	12	12
TTE	24	24	12	12
Exercise Test†	As needed	24	24	12

*See Tables 3 and 4 for details on the ACHD AP classification system.

†6-minute walk test or cardiopulmonary exercise test, depending on the clinical indication.

Congenital Mitral Stenosis (Table 17), SubAS (Table 18)

Abbreviations are listed on page 25.

Routine Follow-Up and Testing Intervals for Specific Conditions

Frequency of Routine Follow-Up and Testing	Physiologic Stage A* (months)	Physiologic Stage B* (months)	Physiologic Stage C* (months)	Physiologic Stage D* (months)
Supravalvular Aortic Stenosis				
Outpatient ACHD Cardiologist	24	24	6–12	3–6
ECG	24	24	12	12
TTE†	24	24	12	12
CMR‡/CCT§	36–60	36–60	36–60	36–60
Exercise Test‖	As needed	24	24	12
CoA				
Outpatient ACHD Cardiologist	24	24	6–12	3–6
ECG	24	24	12	12
TTE†	24	24	12	12
CMR‡/CCT§	36–60	36–60	12–24	12–24
Exercise Test‖	36	24	24	12
Valvular PS				
Outpatient ACHD Cardiologist	36–60	24	6–12	3–6
ECG	36–60	24	12	12
TTE	36–60	24	12	12
Exercise Test†	As needed	24	24	12

*See Tables 3 and 4 for details on the ACHD AP classification system.

†Routine TTE may not be necessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.

‖6-minute walk test or cardiopulmonary exercise test, depending on the clinical indication.

Supravalvular Aortic Stenosis (Table 20):
‡CMR may be indicated for assessment of aortic anatomy. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§If CCT is utilized instead of CMR imaging, the frequency should be weighed against radiation exposure.

CoA (Table 21):
‡CMR may be indicated for assessment of aortic size and aortic arch/coarctation repair site anatomy. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§CCT may be utilized if CMR is not feasible and to evaluate cross-sectional imaging status-post stent therapy for coarctation of the aorta; the frequency should be weighed against radiation exposure.

Valvular PS (Table 23):
†6-minute walk test or cardiopulmonary exercise test, depending on clinical indication.

Abbreviations are listed on page 25.

Routine Follow-Up and Testing Intervals for Specific Conditions

Frequency of Routine Follow-Up and Testing	Physiologic Stage A* (months)	Physiologic Stage B* (months)	Physiologic Stage C* (months)	Physiologic Stage D* (months)
Branch and Peripheral PS				
Outpatient ACHD Cardiologist	24–36	24	6–12	3–6
ECG	24–36	24	12	12
TTE†	24–36	24	12	12
CMR‡/CCT§	36–60	36–60	24–36	24–36
Exercise Test‖	36	24	24	12
Double-Chambered Right Ventricle				
Outpatient ACHD Cardiologist	24–36	24	6–12	3–6
ECG	24–36	24	12	12
TTE	24–36	24	12	12
Exercise Test†	As needed	24	24	12
Ebstein Anomaly				
Outpatient ACHD Cardiologist	12–24	12	6–12	3–6
ECG	12–24	12	12	12
CXR	As needed	As needed	12–24	12–24
TTE†	12–24	12–24	12	12
Pulse Oximetry	24	12	Each visit	Each visit
Holter Monitor	As needed	As needed	24	12–24
CMR‡/CCT§	60	36	24–36	12–24
Exercise Test‖	36	24–36	24	12

*See Tables 3 and 4 for details on the ACHD AP classification system.

Branch and Peripheral PS (Table 24) and Ebstein Anomaly (Table 26):

†Routine TTE may not be necessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.

‖6-minute walk test or cardiopulmonary exercise test, depending on the clinical indication.

Branch and Peripheral PS (Table 24):

‡CMR may be indicated for assessment of branch PS. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§CCT may be utilized if CMR is not feasible and to evaluate cross-sectional imaging status-post stent therapy for peripheral PS; the frequency should be weighed against radiation exposure.

Double-Chambered Right Ventricle (Table 25):

†6-minute walk test or cardiopulmonary exercise test, depending on clinical indication.

Ebstein Anomaly (Table 26):

‡CMR may be indicated for assessment of right ventricular size and function. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§CCT may be utilized if CMR is not feasible; the frequency should be weighed against radiation exposure.

Abbreviations are listed on page 25.

Routine Follow-Up and Testing Intervals for Specific Conditions

Frequency of Routine Follow-Up and Testing	Physiologic Stage A* (months)	Physiologic Stage B* (months)	Physiologic Stage C* (months)	Physiologic Stage D* (months)
TOF				
Outpatient ACHD Cardiologist	12–24	12	6–12	3–6
ECG	24	12	12	12
TTE†	24	12–24	12	6–12
Pulse Oximetry	As needed	As needed	Each visit	Each visit
Holter Monitor	As needed	As needed	12–24	12–24
CMR‡/CCT§	36	24–36	12–24	12–24
Exercise Test‖	36–60	24–60	12–24	12–24
Right Ventricle-to-PA Conduit				
Outpatient ACHD Cardiologist	12–24	12	6–12	3–6
ECG	12–24	12	12	12
TTE†	12–24	12	12	12
CMR‡/CCT§	36–60	36–60	12–24	12–24
Exercise Test‖	As needed	As needed	12–24	12–24

*See Tables 3 and 4 for details on the ACHD AP classification system.

†Routine TTE may not be necessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.

‖6-minute walk test or cardiopulmonary exercise test, depending on the clinical indication.

TOF (Table 27):

‡CMR may be indicated for assessment of right ventricular size and function, pulmonary valve function, pulmonary artery anatomy and left heart abnormalities. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§CCT may be utilized if CMR is not feasible and to evaluate origin and course of the coronary arteries, and cross-sectional imaging status-post stent therapy. If cardiac CCT is utilized instead of CMR imaging, the frequency should be weighed against radiation exposure.

Right Ventricle-to-PA Conduit (Table 28):

‡CMR may be indicated for assessment of right ventricular size and function and valvular function, conduit anatomy and pulmonary artery anatomy. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§CCT may be utilized if CMR is not feasible and to evaluate cross-sectional imaging status-post stent therapy. If CCT is utilized instead of CMR imaging, the frequency should be weighed against radiation exposure.

Abbreviations are listed on page 25.

Routine Follow-Up and Testing Intervals for Specific Conditions

Frequency of Routine Follow-Up and Testing	Physiologic Stage A* (months)	Physiologic Stage B* (months)	Physiologic Stage C* (months)	Physiologic Stage D* (months)
d-TGA With Atrial Switch				
Outpatient ACHD Cardiologist	12	12	6–12	3–6
ECG	12	12	6–12	6–12
TTE†	12–24	12–24	12	12
Pulse Oximetry	12	12	Each visit	Each visit
Holter Monitor	24	24	12	12
CMR‡/CCT§	24–36	24	12–24	12–24
Exercise Test‖	36	36	24	12
d-TGA With Arterial Switch				
Outpatient ACHD Cardiologist	12–24	12	6–12	3–6
ECG	12–24	12–24	12	6
TTE†	12–24	12–24	12	12
CMR‡/CCT§	36–60	24–36	12–24	12–24
Exercise Test‖	36–60	36–60	24–36	12–24

†Routine TTE may not be necessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.

‖6-minute walk test or cardiopulmonary exercise test, depending on the clinical indication.

d-TGA With Atrial Switch (Table 29):
*See Tables 3 and 4 for details on the ACHD AP classification system.

‡CMR may be indicated for assessment of ventricular size and function, systemic and venous baffle obstruction and leaks, and valvular function. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined anatomic and physiological findings.

§CCT may be utilized if CMR is not feasible and to evaluate cross-sectional imaging status-post stent therapy. If CCT is utilized instead of CMR imaging, the frequency should be weighed against radiation exposure.

d-TGA With Arterial Switch (Table 30):
*See ACHD AP classification Table 4.

‡CMR may be indicated for assessment of neoaortic size, the origin and proximal course of the coronary arteries, branch pulmonary arteries, ventricular function and valvular function. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§CCT or catheterization once to establish knowledge of coronary artery anatomy and then as warranted by clinical condition. CCT may be utilized if CMR is not feasible and to evaluate coronary artery anatomy and cross-sectional imaging status-post stent therapy. If CCT is utilized instead of CMR imaging, the frequency should be weighed against radiation exposure.

Abbreviations are listed on page 25.

Routine Follow-Up and Testing Intervals for Specific Conditions

Frequency of Routine Follow-Up and Testing	Physiologic Stage A* (months)	Physiologic Stage B* (months)	Physiologic Stage C* (months)	Physiologic Stage D* (months)
CCTGA				
Outpatient ACHD Cardiologist	12	12	6–12	3–6
ECG	12	12	12	12
TTE†	12–24	12	12	12
Pulse Oximetry	As needed	As needed	Each visit	Each visit
Holter Monitor	12–60	12–60	12–36	12
CMR‡/CCT§	36–60	36–60	12–24	12
Exercise Test‖	36–60	36–60	12–24	12
Fontan Palliation				
Outpatient ACHD Cardiologist	12	12	6	3–6
ECG	12	12	6–12	6
TTE†	12	12	12	12
Pulse Oximetry	12	12	Each visit	Each visit
Holter Monitor	12	12	12	12
CMR‡/CCT§	36	24	24	24
Exercise Test‖	36	24	12	12

*See Tables 3 and 4 for details on the ACHD AP classification system.

†Routine TTE may not be necessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.

‖6-minute walk test or cardiopulmonary exercise test, depending on the clinical indication.

CCTGA (Table 31):

‡CMR may be indicated for assessment of ventricular size and function and valvular function. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§CCT may be utilized if CMR is not feasible. If CCT is utilized instead of CMR imaging, the frequency should be weighed against radiation exposure.

Fontan Palliation (Table 32):

‡CMR may be indicated for assessment of the long-term sequelae of Fontan palliation: thrombosis, right-to-left shunts (e.g., fenestration. intrapulmonary AV malformation), obstructive lesion, systemic atrioventricular valve dysfunction, ventricular size and function, collateral burden, and branch pulmonary artery obstruction. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§CCT may be utilized if CMR is not feasible and to evaluate cross-sectional imaging status-post stent therapy. CCT with contrast injection in Fontan patients can be misleading; therefore, it should be done only when clinically indicated and when it can be appropriately protocoled and interpreted. If CCT is utilized instead of CMR imaging, the frequency should be weighed against radiation exposure.

Abbreviations are listed on page 25.

Routine Follow-Up and Testing Intervals for Specific Conditions

Frequency of Routine Follow-Up and Testing	Physiologic Stage C* (months)	Physiologic Stage D* (months)
Pulmonary Hypertension and Eisenmenger Syndrome		
Outpatient ACHD Cardiologist	6–12	3–6
ECG	12	12
TTE†	12	12
Pulse Oximetry	Each visit	Each visit
CMR‡	As needed	As needed
Exercise Test§	6–12	6–12
Cardiac Catheterization‖	As needed	As needed

*See Tables 3 and 4 for details on the ACHD AP classification system.

†Routine TTE may not be necessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.

‡CMR may be indicated for assessment of right ventricular function and CHD anatomy not clarified with TTE. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§6-minute walk test or cardiopulmonary exercise test, depending on clinical indication.

‖Cardiac catheterization should be performed at baseline and as needed.

Pulmonary Hypertension and Eisenmenger Syndrome (Tables 33)

Abbreviations for Tables 13–33

ACHD: adult congenital heart disease
ECG: electrocardiogram
TTE: transthoracic echocardiogram
ASD: atrial septal defect
VSD: ventricular septal defect
AVSD: atrioventricular septal defect
PDA: patent ductus arteriosus
SubAS: subaortic stenosis
V_{max}: maximum velocity
MRI: magnetic resonance imaging
CT: computed tomography
CMR: cardiovascular magnetic resonance imaging
CCT: cardiac computed tomography
CoA: coarctation of the aorta
PS: pulmonary stenosis
CXR: chest x ray
TOF: tetralogy of Fallot
PA: pulmonary artery
d-TGA: dextro-transposition of the great arteries
CCTGA: congenitally corrected transposition of the great arteries

Routine Follow-Up and Testing Intervals* for Congenital Aortic Stenosis

Stage	Frequency of Echocardiogram
Progressive (Stage B)	Every 3–5 y (mild severity, V_{max} 2.0–2.9 m/s) Every 1–2 y (moderate severity, V_{max} 3.0–3.9 m/s)
Severe (Stage C)	Every 6–12 mo (V_{max} ≥4.0 m/s)
Aortic Dilation >4.5 cm	Every 12 mo (echocardiogram, MRI or CT)

*Modified from existing GDMT for valvular heart disease (S4.2.4-5).

Table 19

V_{max} indicates maximum velocity; MRI, magnetic resonance imaging; CT, computed tomography.