

Table 1.1 ACC/AHA 2025 guideline on ACHD.

ACHD AP Classification

(CHD Anatomy + Physiological Stage = ACHD AP Classification)

CONGENITAL HEART DISEASE ANATOMY	
I: Simple	
Ostium secundum ASD	
Patent ductus arteriosus	
Ventricular septal defect	
Pulmonic stenosis	
II: Moderate Complexity	
Anomalous aortic origin of a coronary artery from the opposite sinus	
Anomalous coronary artery arising from the pulmonary artery	
Anomalous pulmonary venous connection, partial or total	
Atrioventricular septal defect (partial or complete)	
Congenital aortic valve disease	
Congenital mitral valve disease	
Coarctation of the aorta	
Cor triatriatum sinister	
Dextro-TGA after arterial switch operation	
Ebstein anomaly	
Infundibular right ventricular outflow obstruction/double-chamber right ventricle	
Peripheral pulmonary artery stenosis	
Sinus venosus defect	
Subvalvular aortic stenosis (excluding hypertrophic cardiomyopathy, which these guidelines do not address)	
Supravalvar aortic stenosis	
Tetralogy of Fallot	
Vascular ring or sling	
III: Great Complexity (or Complex)	
Double-outlet ventricle	
Fontan physiology	
Interrupted aortic arch	
Pulmonary atresia (all forms, including tetralogy of Fallot with pulmonary atresia)	
Single-ventricle anatomy (including double-inlet left ventricle, tricuspid/mitral atresia, hypoplastic left heart, any other anatomic abnormality with a functionally single ventricle)	
Congenitally corrected TGA (CCTGA, levo-TGA)	
Dextro-TGA after atrial switch (Mustard/Senning) operation	
Dextro-TGA after Rastelli operation	
Truncus arteriosus	
Unrepaired or partially palliated cyanotic congenital heart defect	
PHYSIOLOGICAL STAGE	
Stage A	
No cardiac symptoms	
No hemodynamic or anatomic sequelae	
No sustained arrhythmias*	
Normal exercise capacity*	
Normal pulmonary pressure	

PHYSIOLOGICAL STAGE**Stage B**

Arrhythmia* not requiring new treatment or a change in therapy in the past 12 months

Mild native valve dysfunction* or prosthetic valve with normal function

Mild ventricular dysfunction

Mild ventricular enlargement

Presence of a permanent pacemaker or ICD, without need for ICD therapy in the past 12 months

Trivial or small shunt* (not hemodynamically significant)

Stage C

BNP or NT-proBNP level ≥ 2 times the upper limit of normal

Hemodynamically significant shunt*

Mild or moderate chronic hypoxemia* (baseline resting oxygen saturation 86%-92%)

Moderate or greater valvular dysfunction*

Moderate or severe ventricular dysfunction (systemic, pulmonic, or both)

Pulmonary arterial hypertension (low-risk)*

Sustained or high-burden tachyarrhythmia in the past 12 months requiring treatment with antiarrhythmic drugs, ablation, cardioversion, or ICD therapy

Stage D

Hospitalization for heart failure in the past 12 months^{10,16-18}

Endocarditis in the prior 1 year¹⁹⁻²¹

Eisenmenger syndrome

NYHA functional class* III or IV symptoms

Recurrent arrhythmias* that are hemodynamically significant and/or refractory to treatment

Severe hypoxemia* (baseline oxygen saturation $\leq 85\%$)

Pulmonary arterial hypertension* (intermediate- or high-risk)

ACHD indicates adult congenital heart disease; AP, anatomic and physiological; ASD, atrial septal defect; BNP, B-type natriuretic peptide; ICD, implantable cardioverterdefibrillator; NT-proBNP, N-terminal prohormone of B-type natriuretic peptide; NYHA, New York Heart Association; PDA, patent ductus arteriosus; TGA, transposition of the great arteries; and VSD, ventricular septal defect.

PLEASE DELETE * and Nos in blue

Table 2. EHRA/HRS/APHRS/LAHS 2022 Expert Consensus Statement on Genetic Testing.

Non-exhaustive list of high confident genes for nonsyndromic human CHD

Familial non-syndromic CHD	Gene	Inheritance
Atrial septal defect	<i>GATA4</i>	AD
Atrial septal defect (with or without atrioventricular conduction block)	<i>NKX2.5</i>	AD
	<i>TBX5</i>	AD*
Atrioventricular septal defect	<i>CRELD1</i>	AD
	<i>NR2F2</i>	AD
Supravalvar aortic stenosis	<i>ELN</i>	AD
Aortic valve stenosis	<i>NOTCH1</i>	AD
	<i>TAB2</i>	AD**
Tetralogy of Fallot	<i>NOTCH1</i>	AD
	<i>FLT4</i>	AD
Patent ductus arteriosus	<i>TFAP2B</i>	AD***
Heterotaxy	<i>ACVR2B</i>	AD
	<i>CFC1</i>	AD
	<i>NODAL</i>	AD
	<i>CCDC11</i>	AR
	<i>CFAP53</i>	AR
	<i>PKD1L1</i>	AR
	<i>ZIC3</i>	XL

Pathogenic variants in *TBX5*, *TAB2*, and *TFAP2B* can cause non-syndromic CHD, or may be associated with *hand anomalies, **connective tissue disorder, or ***facial dysmorphism.

Table 3. ACC/AHA 2025 guideline on ACHD.

Genetic Syndromes and Their Associations With Congenital Heart Disease

Syndrome	Genetic Abnormality	Clinical Features	Percentage of Affected Patients With Congenital Heart Disease	Common Cardiac Findings
22q11.2 deletion syndrome ^{12,16-18} (DiGeorge syndrome, velocardiofacial syndrome)	22q11.2 deletion	Facial dysmorphism, palate and speech abnormalities, thymic and parathyroid hypoplasia, hypocalcemia, immunodeficiency, neurodevelopmental and psychiatric disorders	60%-80%	Interrupted aortic arch type B, other aortic arch abnormalities, truncus arteriosus, tetralogy of Fallot, ventricular septal defect/pulmonary atresia
Down syndrome ^{26,27}	Trisomy 21	Facial dysmorphism, developmental disability, atlantoaxial instability, obstructive sleep apnea, craniofacial abnormalities	40%-50%	Atrial septal defect, ventricular septal defect, atrioventricular septal defect, tetralogy of Fallot
Holt–Oram syndrome ^{28,29}	<i>TBX5</i>	Upper-limb skeletal abnormalities, conduction abnormalities	75%	Atrial septal defect, ventricular septal defect, valvular disease (mitral valve prolapse, pulmonary stenosis)
Klinefelter syndrome ³⁰	47,XXY	Tall stature, hypoplastic testes, delayed puberty and developmental disability	50%	Patent ductus arteriosus, atrial septal defect, mitral valve prolapse
Noonan syndrome ^{31,32}	<i>PTPN11</i> , <i>KRAS</i> , <i>SOS1</i> , <i>RAF1</i> , <i>NRAS</i> , <i>BRAF</i> , <i>MAP2K1</i>	Facial anomalies, webbed neck, chest deformity, lymphatic abnormalities, bleeding abnormalities	80%	Pulmonary stenosis, atrial septal defect, hypertrophic cardiomyopathy, atrioventricular septal defect
Turner syndrome ³³⁻³⁵	45,X	Short stature, webbed neck, lymphedema, primary amenorrhea	30%	Bicuspid aortic valve, coarctation of the aorta, aortopathies, partial anomalous pulmonary venous connection, atherosclerosis risk
Williams syndrome ^{36,37}	7q11.23 deletion	“Elfin” facial features, social personality, hearing loss, developmental delay, infantile hypercalcemia	50%-80%	Supravalvar aortic stenosis, branch pulmonary artery stenosis, supravalvar pulmonary stenosis, coronary ostial stenosis, QTc prolongation, mid-aortic narrowing syndrome
Alagille syndrome ^{38,39}	<i>JAG1</i>	Facial dysmorphism, hepatic dysfunction (chronic cholestasis, elevated liver enzymes, biliary atresia, hypercholesterolemia, or liver failure), neurovascular accidents, renal anomalies (renal artery stenosis, hypertension, renal tubular acidosis, uteropelvic obstruction, small, hyperechoic kidneys)	70%-75%	Two-thirds with peripheral or branch pulmonary artery stenosis or other arterial narrowing (coarctation of the aorta, renal artery stenosis, middle cerebral artery, Moyamoya); also can have tetralogy of Fallot, aortic stenosis, septal defects, mid-aortic narrowing syndrome
Heterotaxy syndrome ¹²⁻¹⁴	<i>ZIC3</i> , <i>SHROOM3</i> , <i>GRK5</i> , <i>ANKS3</i> , <i>NODAL</i> , <i>CFCT1</i> , <i>LEFTY2</i> , <i>GDF1</i> , <i>SMAD2</i> , <i>ACVR2B</i>	Situs abnormalities, ciliary dysfunction, functional asplenia, conduction abnormalities, biliary atresia	50%-95%	Atrioventricular septal defect (balanced, unbalanced), malposed great vessels, interrupted inferior vena cava, bilateral superior vena cava, atrial isomerism

Table 4. ESC 2020 GL on ACHD. Recurrence rates for various congenital heart lesions according to the sex of the affected parent

	Recurrence rate (%)*	
	Women	Men
ASD	4–6	1.5–3.5
VSD	6–10	2–3.5
AVSD	11.5–14	1–4.5
PDA	3.5–4	2–2.5
CoA	4–6.5	2–3.5
Marfan/HTAD	50 ^b	
LVOTO	8–18	3–4
RVOTO (PS)	4–6.5	2–3.5
Eisenmenger syndrome	6	NR
TOF	2–2.5	1.5
Pulmonary atresia/VSD	NR	NR
TGA	2 ^b	
ccTGA	3–5 ^b	
UVH (HLHS)	21 ^b	

ASD = atrial septal defect; AVSD = atrioventricular septal defect; ccTGA = congenitally corrected transposition of the great arteries; CHD = congenital heart disease; CoA = coarctation of the aorta; HLHS = hypoplastic left heart syndrome; HTAD = heritable thoracic aortic disease; LVOTO = left ventricular outflow tract obstruction; NR = not reported; PDA = patent ductus arteriosus; PS = pulmonary stenosis; RVOTO = right ventricular outflow tract obstruction; TGA = transposition of the great arteries; TOF = tetralogy of Fallot; UVH = univentricular heart; VSD = ventricular septal defect.

*Except for Marfan syndrome, rates apply to patients with isolated cardiac lesions in whom known genetic/syndromic entities have been excluded.

^b Sex-specific data not available or not relevant.

Type of CHD	Supraventricular arrhythmias			Ventricular arrhythmias and SCD		Bradycardia			
	AVRT	IART/ EAT	AF	Sustained VT	SCD	SND		AV block	
						Congenital	Acquired	Congenital	Acquired
Secundum ASD		++	++			(+)	+		(+)
Superior sinus venous defect		++	+				+		
AVSD/primum ASD		++	++	(+)		(+)		(+)	++
VSD		+	(+)	+	(+) ^a				+
Ebstein anomaly	+++	++	+	(+)	++ ^b		++		
TOF		++	++	++	++		+		+
TGA									
Atrial switch		+++	+	++ ^c	+++ ^b		+++		+
Arterial switch		+		+ ^c	(+)		(+)		
ccTGA	++	+	+	(+)	++ ^b			+	++
Fontan operation									
Atriopulmonary connection		+++	++		+ ^b		++		
Intracardiac lateral tunnel		++	+		+ ^b		++		
Extracardiac conduit		+	+		+ ^b		+		
Eisenmenger physiology Incompletely palliated CHD		++	++		++ ^d				

Empty cells indicate that although not specifically indicated, arrhythmic events may occur (no symbol).

(+) = minimal risk + = mild risk ++ = moderate risk +++ = high risk

Figure 1. ESC 2020 GL on ACHD. Risk estimates for arrhythmic events and bradycardias in ACHD

a Considering the high prevalence of VSD, the overall risk in unselected patients with VSD is considered to be minimal.

b SCD may be due to supraventricular arrhythmias with rapid AV conduction.

c VT higher estimated risk in complex dextro-TGA.

d Non-arrhythmic

AF = atrial fibrillation; ASD = atrial septal defect; AV = atrioventricular; AVRT = atrioventricular reentrant tachycardia; AVSD = atrioventricular septal defect; ccTGA = congenitally corrected transposition of the great arteries; CHD = congenital heart disease; EAT = ectopic atrial tachycardia; IART = intraatrial reentrant tachycardia; SCD = sudden cardiac death; SND = sinus node dysfunction; TGA = transposition of the great arteries; TOF = tetralogy of Fallot; VSD = ventricular septal defect; VT = ventricular tachycardia

Table 5. ACC/AHA 2025 guideline on ACHD. Assessment for Pulmonary Arterial Hypertension in Adults With Congenital Heart Disease

Assessment Type	Findings
Clinical history, examination	■ Reduced functional capacity ■ Symptoms: ■ Functional: dyspnea, fatigue ■ Right heart failure: congestion, abdominal distention, early satiety ■ History of near-syncope ■ Desaturation at rest ■ Cyanosis ■ Clubbing ■ Accentuated S2 heart sound ■ Jugular venous distention, distended abdomen, peripheral edema
Exercise capacity	■ Reduced 6-minute walk distance ■ Possibly reduced resting or exercising oxygen saturation ■ Abnormal lung function, at rest and during exercise
Imaging	■ Electrocardiogram: right ventricular enlargement ■ Transthoracic echocardiography: RVE, flattened IVS, RAE, peak TR jet >2.9 m/s, RVOT acceleration time <105 ms, TAPSE <18 mm
Biomarkers	■ Elevated BNP or NT-proBNP
Hemodynamics	■ mPAP >20 mm Hg and ■ PVR >2 Wood units and ■ PCWP ≤15 mm Hg

BNP indicates B-type natriuretic peptide; CT, computed tomography; IVS, interventricular septum; JVD, jugular venous distention; mPAP, mean pulmonary artery pressure; NT-proBNP, N-terminal pro-hormone of B-type natriuretic peptide; PAH, pulmonary arterial hypertension; PCWP, pulmonary capillary wedge pressure; PVR, pulmonary vascular resistance; RAE, right atrial enlargement; RVE, right ventricular enlargement; RVOT, right ventricular outflow tract; TAPSE, tricuspid annular plane systolic excursion; and TR, tricuspid regurgitation.

Table 6. ACC/AHA 2025 GL on ACHD. Isolated Shunt Lesions and Pulmonary Arterial Hypertension

PAH-ACHD Subtype	Patient Characteristics		Evidence for Use of Risk Assessment Factors/Tools	Evidence for Use of PAH-Specific Therapy
	Resting hypoxemia/cyanosis	Typical defect size		
A. Eisenmenger syndrome	Usually	Large	Patient age, pretricuspid shunt, resting oxygen saturation, loss of sinus rhythm, pericardial effusion, ⁷ BNP, ⁸ C-reactive protein, ⁹ 6MWD, ^{7,10} echocardiography, ¹¹ renal dysfunction, or other clinical stratification tools. ¹²⁻¹⁴	Endothelin-receptor antagonists, ¹⁵⁻¹⁷ phosphodiesterase-5 inhibitors, ¹⁸ prostacyclin analogs ¹⁹⁻²³ †
B. Prevalent systemic to pulmonary shunts (left-to-right shunts with elevated PVR, yet not meeting criteria for Eisenmenger syndrome)	No	Moderate to large	Review using the 3-strata risk-prediction model for newly diagnosed patients, the 4-strata risk-prediction model for patients with established PAH, ¹ and/or any of the Eisenmenger syndrome tools. ^{‡1-7}	No direct studies on this isolated cohort; treatment has typically followed recommendations for idiopathic PAH ¹
C. Coincidental defects with elevated PVR	Rare	Small	Review using the 3-strata risk-prediction model for newly diagnosed patients, the 4-strata risk-prediction model for patients with established PAH, ¹ and/or any of the Eisenmenger syndrome tools. ^{‡1-7}	No direct studies on this isolated cohort; treatment has typically followed recommendations for idiopathic PAH ¹
D. Postcorrection defects with elevated PVR	Rare (with small residual shunts and severe pulmonary vascular disease)	Corrected	Review using the 3-strata risk-prediction model for newly diagnosed patients, the 4-strata risk-prediction model for patients with established PAH, ¹ and/or any of the Eisenmenger syndrome tools. ^{‡1-7}	Phosphodiesterase-5 inhibitors, ²⁴ endothelin receptor antagonists, ²⁵⁻²⁷ oral prostacyclin receptor analog, ²⁸ activin signaling inhibitors ^{§29}

*No validated score available.

†See Section 4.4.6 for details on the order of PAH-directed drug initiation.

‡For patients in groups B, C, and D, the 3- and 4-strata models have been applied, and evidence for use has included enrollment of patients in this category; however, validation for this specific population in isolation from other forms of PAH has not been directly studied.

§Trials that included patients with PAH after congenital defect correction.

6MWD indicates 6-minute walk distance; ACHD, adult congenital heart disease; BNP, B-type natriuretic peptide; N/A, not applicable; PAH, pulmonary arterial hypertension; and PVR, pulmonary vascular resistance.

Table 7. ESC 2020 GL on ACHD. Indications for cardiovascular magnetic resonance imaging in ACHD patients

- Quantification of RV volumes, EF (including subpulmonary RV, systemic RV, and single ventricle)
- Evaluation of RVOTO and RV_PA conduits
- Quantification of PR
- Evaluation of PAs (stenoses, aneurysms) and the aorta [aneurysm, dissection, coarctation (CCT may be superior)]
- Evaluation of systemic and pulmonary veins (anomalous connection, obstruction, coronary venous anatomy pre-procedure, etc.)
- Collaterals and arteriovenous malformations (CCT may be superior)
- Coronary anomalies and CAD (CCT is superior for intramural course, slit-like course, acute angle take-off, myocardial bridging, and plaque assessment)
- Detection and quantification of myocardial ischaemia by CMR stress perfusion
- Evaluation of intra- and extracardiac masses
- Quantification of myocardial mass (LV and RV)
- Detection and quantification of myocardial fibrosis/scar (late gadolinium enhancement, T1 mapping) tissue characterization (fibrosis, fat, iron, etc.)
- Quantification of systemic and pulmonary blood flow to calculate Qp:Qs
- Quantification of perfusion distribution to the right/left lung
- Measurement of pulmonary blood flow in patients with multiple sources of blood supply (i.e. with major aorto-pulmonary collateral arteries)

ACHD = adult congenital heart disease; CAD = coronary artery disease; CMR = cardiovascular magnetic resonance; CCT = cardiovascular computed tomography; EF = ejection fraction; LV = left ventricle/ventricular; PA = pulmonary artery; PR = pulmonary regurgitation; Qp:Qs = pulmonary to systemic flow ratio; RV = right ventricle/ventricular; RVOTO = right ventricular outflow tract obstruction.

Table 8. ACC/AHA 2025 guideline on ACHD.**ACHD Program and Cardiologists**

Patients in ACHD AP classes IB–D, IIA–D, and IIIA–D should be evaluated and managed by or in collaboration with an ACHD cardiologist and, when indicated and accessible, an ACHD program to improve outcomes. 1 B-NR

Patients in ACHD AP class IA should have at least 1 evaluation by an ACHD cardiologist to develop a plan of care. 1 B-NR

ACHD Program Components

ACHD board-eligible/-certified cardiologists
Congenital cardiac surgeons
Advanced practice providers
Registered nurses
Advanced imagers: echocardiography/CT/CMR*
Cardiac intensivist*
Advanced heart failure and transplant cardiologist*
Cardiac catheterization interventionalist*
Electrophysiologist*
Cardiac anesthesiologist*
Multidisciplinary specialists
Maternal fetal medicine
Hepatology
Pulmonary hypertension
Cardiovascular genetics
Palliative medicine
Social work
Psychology

*Personnel and service lines supervised, performed, and interpreted by clinicians with expertise and/or training in congenital heart disease.

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

Mental Health and Neurocognitive Assessment

Adults with congenital heart disease should be assessed for mental health conditions, including depression, anxiety, and post-traumatic stress disorder, to identify those who may benefit from mental health services. 1 C-LD

For adults with congenital heart disease who have mental health concerns, referral to a mental health clinician can be beneficial to improve psychological symptoms and quality of life. 2a C-LD

For adults with congenital heart disease, neuropsychological testing may be considered to guide therapies that enhance academic, behavioral, psychosocial, and adaptive functioning. 2b B-NR

For adults with congenital heart disease, evaluation for cognitive decline and frailty may be considered to identify patients who can benefit from additional support 2b C-LD

Ionizing Radiation Exposure

In adults with congenital heart disease, processes to limit radiation exposure during imaging and cardiovascular procedures are recommended to reduce the lifetime risk for cancer. 1 B-NR

Exercise Testing

In patients with ACHD, CPET can be useful for establishing a baseline functional assessment, guiding clinical care, stratifying risk for major adverse cardiovascular events, and ongoing surveillance. 2a B-NR

In patients with ACHD and established PAH, or in those unable to perform CPET yielding reliable diagnostic information, a 6-minute walk test can be useful to objectively assess symptom severity, functional capacity, and response to therapy. 2a B-NR

Expertise for Cardiac Surgery and Invasive Cardiac Procedures

In patients with ACHD who undergo invasive cardiac procedures, preprocedural involvement of an ACHD cardiologist is recommended to riskstratify the patient, determine appropriate procedure location, guide periprocedural planning and support, and assist in preprocedural clinical optimization to improve outcomes. 1 B-NR

In patients with ACHD, invasive cardiac procedures involving congenital lesions should be performed by operators with expertise in congenital heart disease procedures to ensure appropriate periprocedural care. 1 B-NR

In patients with ACHD undergoing invasive cardiac procedures, postprocedural care should involve collaboration with an ACHD cardiologist to incorporate expert opinion, provide guideline-based therapy, and improve outcomes. 1 C-LD

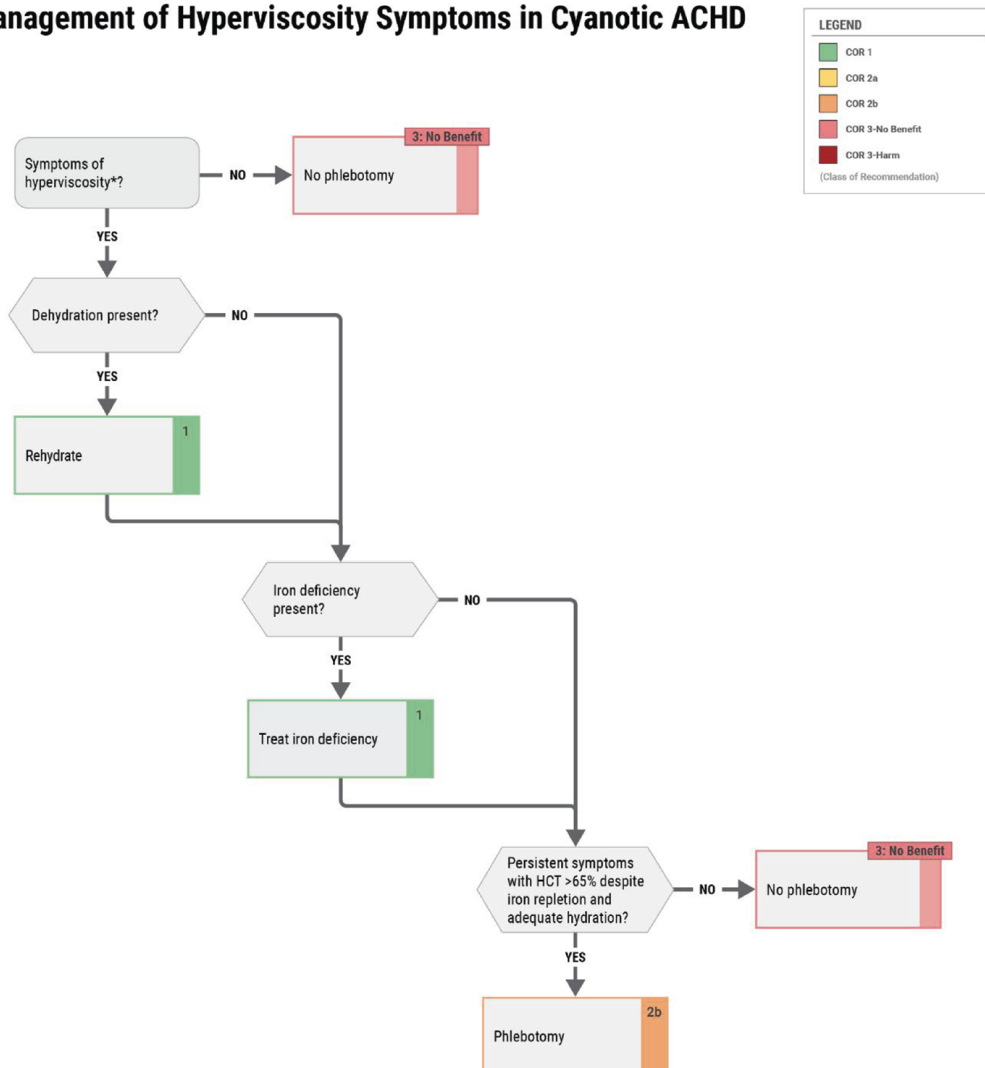
In patients with ACHD AP classifications IC–D, IIA–D, and IIIA–D, anesthesia for invasive cardiac procedures should be administered by, or in collaboration or consultation with, an anesthesiologist who has congenital heart disease expertise to reduce the likelihood of perioperative complications. 1 C-LD

In patients with ACHD who undergo invasive cardiac procedures for acquired cardiovascular disease, it can be beneficial for operators with expertise in procedural treatment of acquired disease to collaborate with ACHD experts 1 B-NR

Table 9. ACC/AHA 2025 guideline on ACHD. Cyanotic ACHD**Management of Cyanosis**

Patients with ACHD and cyanosis should receive annual screening for and treatment of iron deficiency to improve exercise capacity and functional status.	1 B-NR
In patients with ACHD and cyanosis who present with persistent, new, or worsening neurologic deficits, urgent brain imaging to exclude cerebral abscess or stroke should be performed.	1 B-NR
In patients with ACHD and cyanosis who present with symptoms of hyperviscosity, rehydration with oral or intravenous fluids should be performed to improve symptoms and reduce the risk for vascular complications.	1 B-NR
Pregnant patients with ACHD and cyanosis should be closely followed for the entire duration of the pregnancy by a multidisciplinary cardioobstetrics team of experts in maternal-fetal medicine, ACHD, and obstetric anesthesia to reduce the risk for pregnancy-related cardiovascular and obstetric complications.	1 B-NR
Patients with ACHD and cyanosis who are of childbearing age should receive comprehensive family planning consultation, including recommendations on safe and reliable contraceptive methods, from a multidisciplinary team of experts in family planning and ACHD.	1 C-EO
In patients with ACHD and cyanosis who experience recurrent unexplained hypertension or tachycardia, evaluation to detect pheochromocytomas and paragangliomas is reasonable	2a B-NR
In patients with ACHD, cyanosis, and recurrent episodes of joint pain, it is reasonable to check serum uric acid levels and evaluate for gout.	2a B-NR
In patients with ACHD and cyanosis, the use of supplemental oxygen during exercise may be considered to improve exercise capacity.	2b B-R
In patients with ACHD, hematocrit >65%, and hyperviscosity symptoms that persist despite adequate rehydration and treatment of iron deficiency, phlebotomy may be considered.	2b C-EO
In patients with ACHD and cyanosis, prophylactic phlebotomy is not recommended.	No Benefit B-NR

Management of Hyperviscosity Symptoms in Cyanotic ACHD



*All patients should have at least annual screening for iron deficiency. ACHD indicates adult congenital heart disease; and HCT, hematocrit.

Figure 2. ACC/AHA 2025 guideline on ACHD. Management of Hyperviscosity Symptoms in Cyanotic ACHD

Table 10. ESC 2021 GL on pacing and CRT. Recommendations for cardiac pacing in patients with congenital heart disease

In patients with congenital complete or high degree AVB, pacing if one of the following risk factors is present: a. Symptoms b. Pauses >3_ the cycle length of the ventricular escape rhythm c. Broad QRS escape rhythm d. Prolonged QT interval e. Complex ventricular ectopy f. Mean daytime heart rate <50 b.p.m.	I C
In patients with congenital complete or high degree AVB, permanent pacing even if no risk factors are present.	IIb C
Permanent pacing in patients with persistent post-operative bifascicular block associated with transient complete AVB.	IIb C
In patients with complex CHD and asymptomatic bradycardia (awake resting heart rate <40 b.p.m. or pauses >3 s), permanent pacing on an individual basis.	IIb C
AVB = atrioventricular block; BBB = bundle branch block; b.p.m. = beats per minute; CHD = congenital heart disease; ECG = electrocardiogram.	

Table 11. ESC 2022 GL on VA and SCD. Therapy of arrhythmias in ACHD

Recommendations for risk stratification and primary prevention of sudden cardiac death in congenital heart disease

Risk stratification and primary prevention of SCD

All CHD patients

ICD in adults with CHD with biventricular physiology and a left systemic ventricle presenting with symptomatic heart failure (NYHA II/III) and EF ≤35% despite ≥3 months of OMT.	I C
ICD in patients with CHD with presumed arrhythmic syncope and with either at least moderate ventricular dysfunction or inducible SMVT on PES,	IIa C
ICD in patients with advanced single ventricle or systemic RV dysfunction with additional risk factors, ^a	IIb C
Tetralogy of Fallot	
In patients after repair of TOF with arrhythmia symptoms and NSVT, electrophysiologic evaluation including PES.	IIa C
ICD in patients after repair of TOF with arrhythmia symptoms and a positive PES, or a combination of	IIa C

other risk factors^b and a positive PES.

In patients after repair of TOF without arrhythmia symptoms,
but with a combination of other risk factors,^c electrophysiologic evaluation,
including PES. I Ib C

In patients with repaired TOF undergoing surgical or transcutaneous pulmonary
valve replacement, pre-operative catheter mapping and transection
of VT-related anatomical isthmuses before or during the intervention. I Ib C

AV, atrioventricular; CHD, congenital heart disease; CMR, cardiac magnetic resonance;
EF, ejection fraction; ICD, implantable cardioverter defibrillator; LV, left valve; NSVT,
non-sustained monomorphic ventricular tachycardia; NYHA, New York Heart
Association; OMT, optimal medical treatment; PES, programmed electrical stimulation;
RV, right ventricular; SCD, sudden cardiac death; SMVT, sustained monomorphic
ventricular tachycardia; TOF, tetralogy of Fallot; VT, ventricular tachycardia.

^aData are sparse and risk factors may be lesion-specific, including non-sustained VT,
NYHA II/III, severe AV valve regurgitation, and wide QRS ≥ 140 ms (transposition of
the great arteries).

^bOther risk factors include moderate RV or LV dysfunction, extensive RV scarring on
CMR, QRS duration ≥ 180 ms and severe QRS fragmentation.

Recommendations for secondary prevention of sudden cardiac death and treatment of ventricular arrhythmia in congenital heart disease

Secondary prevention of SCD and treatment of VA

All CHD patients

In patients with CHD presenting with sustained VAs, evaluation for residual lesions
or new structural abnormalities. I B

In patients with CHD with not tolerated VT/ aborted CA due to VF, ICD implantation
is indicated after exclusion of reversible causes. I C

In patients with CHD and recurrent, symptomatic
SMVT or ICD shocks for SMVT not manageable
by medical therapy or ICD reprogramming,
catheter ablation performed in specialized centres.^a IIa C

In selected patients with CHD (including atrial
baffle repair for transposition of the great arteries,
Fontan operation and Ebstein anomaly)
presenting with CA, evaluation and treatment of
SVT with rapid ventricular conduction. IIa C

Tetralogy of Fallot

In patients with repaired TOF who present with
SMVT or recurrent, symptomatic appropriate
ICD therapy for SMVT, catheter ablation
performed in specialized centres. I C

In patients with repaired TOF with SMVT who are undergoing surgical or transcutaneous pulmonary valve replacement, pre-operative catheter mapping and transection of VT-related anatomical isthmuses before or during the intervention. IIa C

In patients with repaired TOF with a preserved biventricular function and symptomatic SMVT, catheter ablation or concomitant surgical ablation performed in specialized centres as an alternative to ICD therapy. IIb C

CA, cardiac arrest; CHD, congenital heart disease; ICD, implantable cardioverter defibrillator; SCD, sudden cardiac death; SMVT, sustained monomorphic ventricular tachycardia; SVT, supraventricular tachycardia; TOF, tetralogy of Fallot; VA, ventricular arrhythmia; VF, ventricular fibrillation; VT, ventricular tachycardia.

^aSpecific recommendations for TOF (Class I-B).

Table 12. ACC/AHA/HRS 2017 GL on VA/SCD. Adult Congenital Heart Disease

Evaluate for potential residual anatomic or coronary abnormalities patients with repaired complex congenital heart disease presenting with frequent, complex, or sustained VA, or unexplained syncope.	I-B-NR
In complex or sustained VA in the presence of important residual hemodynamic lesions, treatment of hemodynamic abnormalities with catheter or surgical intervention is indicated prior to consideration of ablation or an ICD.	I-B-NR
ICD in hemodynamically unstable VT, after evaluation and appropriate treatment for residual lesions/ventricular dysfunction.	I-B-NR
ICD in SCA due to VT or VF in the absence of reversible causes	I-B-NR
Electrophysiological study to evaluate the risk of sustained VT/VF in repaired tetralogy of Fallot physiology with high-risk characteristics and frequent VA.	IIa-B-NR
ICD in repaired tetralogy of Fallot physiology and inducible VT/VF or	IIa-N-NR
Catheter ablation for recurrent sustained monomorphic VT or recurrent ICD shocks for VT.	IIa-B-NR
Beta blocker can be beneficial to reduce the risk of SCA in frequent or complex VA.	IIa-B-NR

ICD or an electrophysiological study with ICD implantation for inducible sustained VA.	IIa-B-NR
ICD implantation or an electrophysiological study with ICD implantation for inducible sustained VA in repaired moderate or severe complexity congenital heart disease with unexplained syncope and at least moderate ventricular dysfunction or marked hypertrophy.	IIa-B-NR
ICD for severe ventricular dysfunction (LVEF <35%) and symptoms of heart failure despite GDMT or additional risk factors.	IIb-B-NR
Prophylactic antiarrhythmic therapy with class Ic medications (i.e., flecainide, propafenone) or amiodarone for asymptomatic VA is potentially harmful.	III-B-NR Harm
ICD is recommended if meaningful survival of greater than 1 year is expected.	

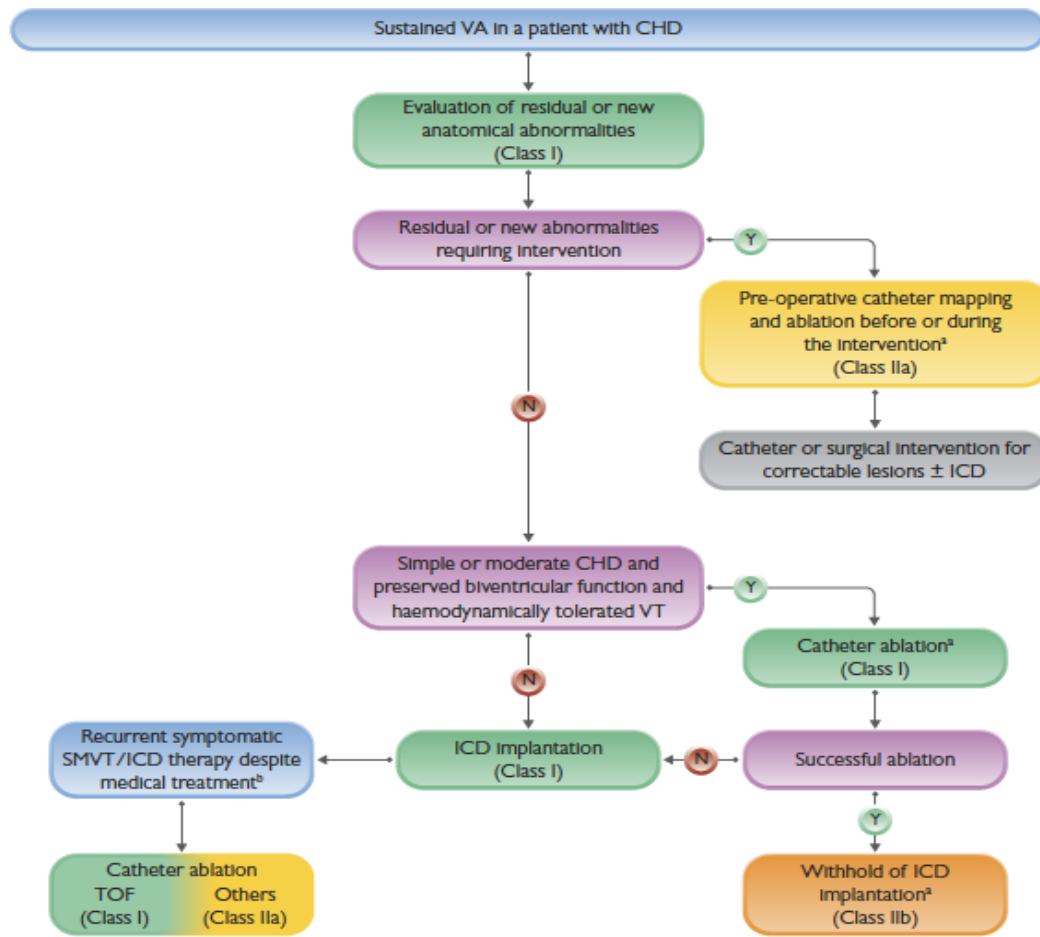


Figure 3. ESC 2022 GL on VA and SCD. Algorithm for the management of sustained ventricular arrhythmia in patients with congenital heart disease.

a Data derived from patients with TOF and related lesions.

b In TOF, anti-arrhythmic drug failure is not required.

CHD, congenital heart disease; ICD, implantable cardioverter defibrillator; N, No; TOF, tetralogy of Fallot; VA, ventricular arrhythmia; VT, ventricular tachycardia; Y, Yes.

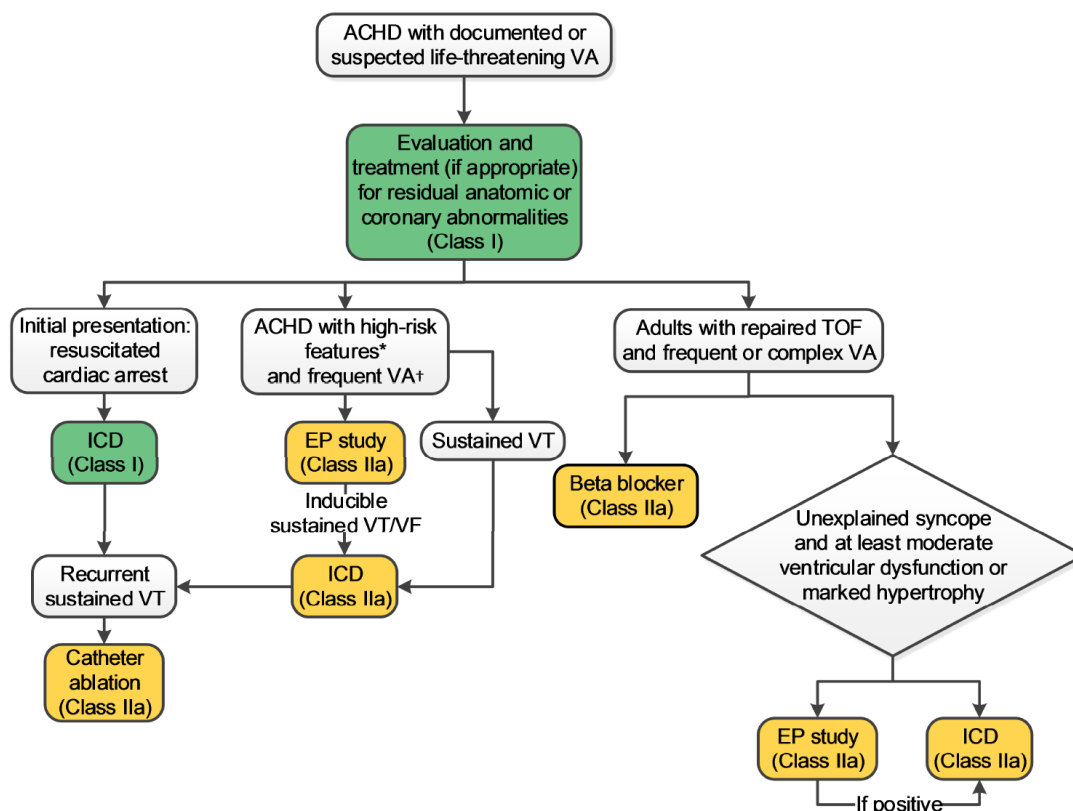


Figure 4. ACC/AHA/HRS 2017 GL on VA/SCD. Prevention of SCD in Patients With Adult Congenital Heart Disease.

*High-risk features: prior palliative systemic to pulmonary shunts, unexplained syncope, frequent PVC, atrial tachycardia, QRS duration ≥ 180 ms, decreased LVEF or diastolic dysfunction, dilated right ventricle, severe pulmonary regurgitation or stenosis, or elevated levels of BNP.

†Frequent VA refers to frequent PVCs and/or nonsustained VT.

ACHD indicates adult congenital heart disease; BNP, B-type natriuretic peptide; EP, electrophysiological; ICD, implantable cardioverter-defibrillator; LVEF, left ventricular ejection fraction; PVC, premature ventricular complexes; SCD, sudden cardiac death; TOF, tetralogy of Fallot; VA, ventricular arrhythmia; and VT, ventricular tachycardia.

Table 13. ACC/AHA 2025 GL on ACHD. Heart Failure

In patients with ACHD and heart failure symptoms who have systemic left ventricular (LV) dysfunction and biventricular circulation, and who otherwise meet accepted criteria for cardiac resynchronization therapy (CRT), use of CRT is reasonable to improve heart failure symptoms.	2a B-NR
In patients with ACHD and heart failure symptoms who have systemic LV systolic dysfunction and biventricular circulation, use of GDMT for heart failure is reasonable to improve heart failure symptoms.	2a C-LD
In patients with ACHD and heart failure symptoms who have systemic LV dysfunction and biventricular circulation, who otherwise meet accepted criteria, use of a primary prevention implantable cardioverter-defibrillator (ICD) is reasonable to prevent sudden cardiac death (SCD).	2a C-LD

Table 14. ESC 2021 GL on HF. Treatment of adult congenital heart disease and heart failure in specialized centres

ACHD patients with chronic HF should be referred to specialized centres.
Specific guidelines for medical treatment of chronic HF in ACHD are lacking, and practitioners should follow the current guidelines for medical treatment of HF. It remains unknown whether the long-term use of neurohormonal modulators affects clinical outcomes and prognosis in ACHD.
Sacubitril/valsartan may decrease morbidity, ^{991–993} however, no recommendation can be made at this moment based on the retrospective or anecdotal nature of these observations.
Co-morbidities in HF such as diabetes mellitus, AF, CSA, iron deficiency, and cachexia should be treated according to specific recommendations reported in this document.
In a biventricular circulation, patients with an impaired systemic LV should be treated with conventional HF therapy; this may also be considered in symptomatic patients with a failing systemic right ventricle.
Diuretics are recommended to control symptoms of fluid retention.
Treatment of symptomatic patients with a failing single ventricle in a Fontan circulation, or in the case of a persistent right-to-left shunt, should always be carefully initiated, taking the labile balance of ventricular preload and systemic afterload into account.
CRT may be a therapeutic option in ACHD patients with HF, but evidence in this specific setting is still lacking. Efficacy of CRT will depend on the underlying structural and functional substrate, such as anatomy of the systemic ventricle (left, right, or functionally single), presence and degree of structural systemic AV valve regurgitation, primary myocardial disease or scarring, and type of electrical conduction delay. ⁹⁸⁸
Treatment of acute HF in ACHD patients should be in an expert centre, with proper knowledge of inotropes, the availability of extracorporeal membrane oxygenation, and advanced bridging techniques. ^{988,994}
Timely evaluation for transplantation by ACHD HF specialists in a transplant centre with ACHD expertise is recommended.
Ventricular assist devices can bridge patients to transplantation, or in a subgroup of patients, may be an option as destination therapy.
ACHD = adult congenital heart disease; AF = atrial fibrillation; AV = atrioventricular; CRT = cardiac resynchronization therapy; CSA = central sleep apnoea; HF = heart failure; LV = left ventricular.

Minor risk	Patients with small, uncorrected defects, and no need for medication or any other treatment Patients with successfully corrected CHD with no symptoms, no relevant residua, and no need for medication
Intermediate risk	Patients with corrected or uncorrected conditions with residual haemodynamic abnormality, with or without medication
Severe risk	Patients with uncorrected cyanotic heart disease, pulmonary hypertension, other complex CHD, ventricular dysfunction requiring medication, and patients listed for heart transplantation

Table 15. ACC/AHA 2025 GL on ACHD. Heart Transplantation and Mechanical Circulatory Support (MCS)

In patients with ACHD and heart failure, evaluation by specialists with medical and surgical experience in ACHD, as well as by specialists with expertise in advanced heart failure, is recommended to assess appropriateness for advanced heart failure therapies, such as heart transplantation or MCS.	1 B-NR
In patients with ACHD who are considered for heart transplantation, multiorgan transplantation (heart–lung, heart–kidney, or heart–liver) is reasonable based on center-specific assessment of the degree of respective lung, kidney, or liver dysfunction.	2a B-NR
In patients with ACHD and advanced heart failure that is refractory to optimal medical and device-based therapy, the use of durable MCS devices is reasonable to improve quality of life or as a bridge to transplantation.	2a B-NR

Table 16. Noncardiac surgery

ESC 2022 Guidelines on noncardiac surgery. Risk stratification for non-cardiac surgery in adults with congenital heart disease

Minor risk	Patients with small, uncorrected defects, and no need for medication or any other treatment Patients with successfully corrected CHD with no symptoms, no relevant residua, and no need for medication
Intermediate risk	Patients with corrected or uncorrected conditions with residual haemodynamic abnormality, with or without medication
Severe risk	Patients with uncorrected cyanotic heart disease, pulmonary hypertension, other complex CHD, ventricular dysfunction requiring medication, and patients listed for heart transplantation

**ACC/AHA 2025 guideline on ACHD.
Expertise for Noncardiac Surgery**

In patients with ACHD being considered for noncardiac procedures, preprocedural involvement of an ACHD cardiologist is recommended to risk-stratify the patient, determine appropriate procedure location, guide periprocedural planning and support, and assist in preprocedural clinical optimization to improve outcomes.	1 B-NR
In patients with ACHD AP classifications IC–D, IIB–D, and IIIA–D who undergo noncardiac procedures, postprocedural care should involve collaboration with an ACHD cardiologist to incorporate expert opinion, provide guideline-based therapy, and improve outcomes.	1 B-NR
In patients with ACHD AP classifications IC–D, IIC–D, and IIIA–D,	1 C-LD

anesthesia for noncardiac procedures should be administered by, or in collaboration or consultation with, an anesthesiologist with ACHD expertise to reduce the likelihood of perioperative complications.

Preoperative Checklist for Noncardiac Surgery in Patients With ACHD

Clarify clinical information related to congenital heart disease

- Clarify prior procedures, residua, sequelae, and current status, including ACHD anatomic-physiological classification.
 - Develop management strategies for preoperative optimization to minimize risk and improve postprocedural outcome.
-

Factors associated with increased risk for perioperative morbidity and mortality^{2,12,26}:

- Cyanosis
 - Complex congenital heart disease
 - Frailty
 - Heart failure
 - Operations on the respiratory and nervous systems
 - Pulmonary hypertension
 - Urgent/emergency procedures
-

Issues to consider:

- Arrhythmias, including bradyarrhythmias
 - Abnormal venous and/or arterial anatomy affecting venous and arterial access and blood pressure monitoring
 - Adjustment of anticoagulant volume in tubes for some blood work in patients with cyanosis
 - Developmental disability
 - Differential dependence on preload/afterload and impacts on ventilation and/or insufflation strategies
 - Endocarditis prophylaxis
 - Erythrocytosis
 - Monitoring of renal and liver function
 - Need for meticulous line care (consider air filters for intravenous lines) to reduce risk for paradoxical embolism in patients with reduced oxygen saturations because of right-to-left shunts
 - Periprocedural anticoagulation
 - Persistent shunts
 - Prevention of venous thrombosis
 - Pulmonary vascular disease
 - Valvular disease
 - Ventricular function (systemic and/or subpulmonary)
-

Table 17. Table 3. ACC/AHA 2025 guideline on ACHD. Genetic Syndromes Screening

For adults with conotruncal defects, screening for 22q11.2 deletion syndrome is reasonable to assess the risk of transmission to offspring and to screen for associated comorbidities.¹ 2a B-NR

In adults with congenital heart disease and a family history of congenital heart disease or clinical features associated with a genetic syndrome, referral for genetic evaluation is reasonable to guide management. 2a C-EO

Table 18. EHRA/HRS/APHRS/LAHRs 2022 Expert Consensus Statement on Genetic Testing.**State of genetic testing for congenital heart disease**

Genetic testing for patients with congenital heart disease (CHD) should prioritize the personal goals and preferences of the patient, parents or guardian, require pre-test genetic counseling and should be coordinated by multidisciplinary teams with expertise in genetics of CHD.

Antenatal testing**Should do**

When foetal congenital heart disease (CHD) is identified on antenatal ultrasound examinations, a chromosomal microarray (CMA) or CNV sequencing (CNV seq) of foetal tissue [amniocentesis or chorionic villous sample (CVS)] should be offered.

May do

Trio WES on amniocentesis or CVS samples may be performed in prenatal cases with syndromic and/or complex CHD.

Neonates and infants requiring investigation or procedures for complex CHD**Should do**

CMA or CNV seq is indicated in infants with CHD to identify pathogenic CNVs.

May do

Standard karyotype analysis in infants with CHD (or pathogenic CNVs) to identify balanced translocations.

Trio WES or WGS may be performed in infants with complex CHD to identify pathogenic variants/indels, contributing to prognostication for extracardiac outcome and for cardiac outcome.

Targeted CHD gene panels to identify variants in a set of CHD-related genes in infants with complex CHD.

Patients with CHD and extracardiac anomalies

Should do

CMA or CNV seq in patients with CHD and extracardiac anomalies to identify pathogenic CNVs.

Trio testing for de novo or inherited (autosomal or X-linked recessive) pathogenic variants with either WES or WGS should be performed in patients with CHD and extracardiac anomalies.

Familial forms of CHD

Should do

WES/WGS of the affected family members in families with at least two first degree relatives with heterotaxy or CHD.

May do

Targeted analysis of a specific gene if the CHD type is highly suggestive of a specific gene.

Sporadic non-syndromic CHD (excl. neonates or infants)

May do

CMA or CNV seq for pathogenic CNVs in older individuals with sporadic nonsyndromic CHD.

Do not do

Trio testing for de novo or inherited (autosomal or X-linked recessive) pathogenic variants with either WES or WGS in older patients with sporadic non-syndromic CHD has a low diagnostic yield and limited utility, and should not be performed routinely.

Heterotaxy

Should do

WES/WGS of the affected family members in families with at least two first degree relatives with CHD.

Trio testing for de novo or inherited (autosomal or X-linked recessive) pathogenic variants with either WES or WGS in patients with syndromic heterotaxy (e.g. primary ciliary dyskinesia).

Do not do

Routine trio testing for de novo or inherited (autosomal or X-linked recessive) pathogenic variants with either WES or WGS should not be performed in patients with sporadic non-syndromic heterotaxy.

Table 19. ACC/AHA 2025 GL on ACHD**Physical Activity, Exercise, and Sports Participation**

Patients with ACHD should have their activity levels assessed at regular intervals to receive counseling about the types and intensity of exercise appropriate to their clinical status.	1 B-NR
In patients with ACHD, exercise training and cardiac rehabilitation can be useful to increase exercise capacity.	2a B-R
In patients with ACHD, CPET can be useful to guide physical activity recommendations.	2a C-LD
In patients with ACHD, participation in competitive sports is reasonable following a comprehensive evaluation by an ACHD specialist that incorporates functional status and individualized assessment of risks and benefits.	2a C-EO

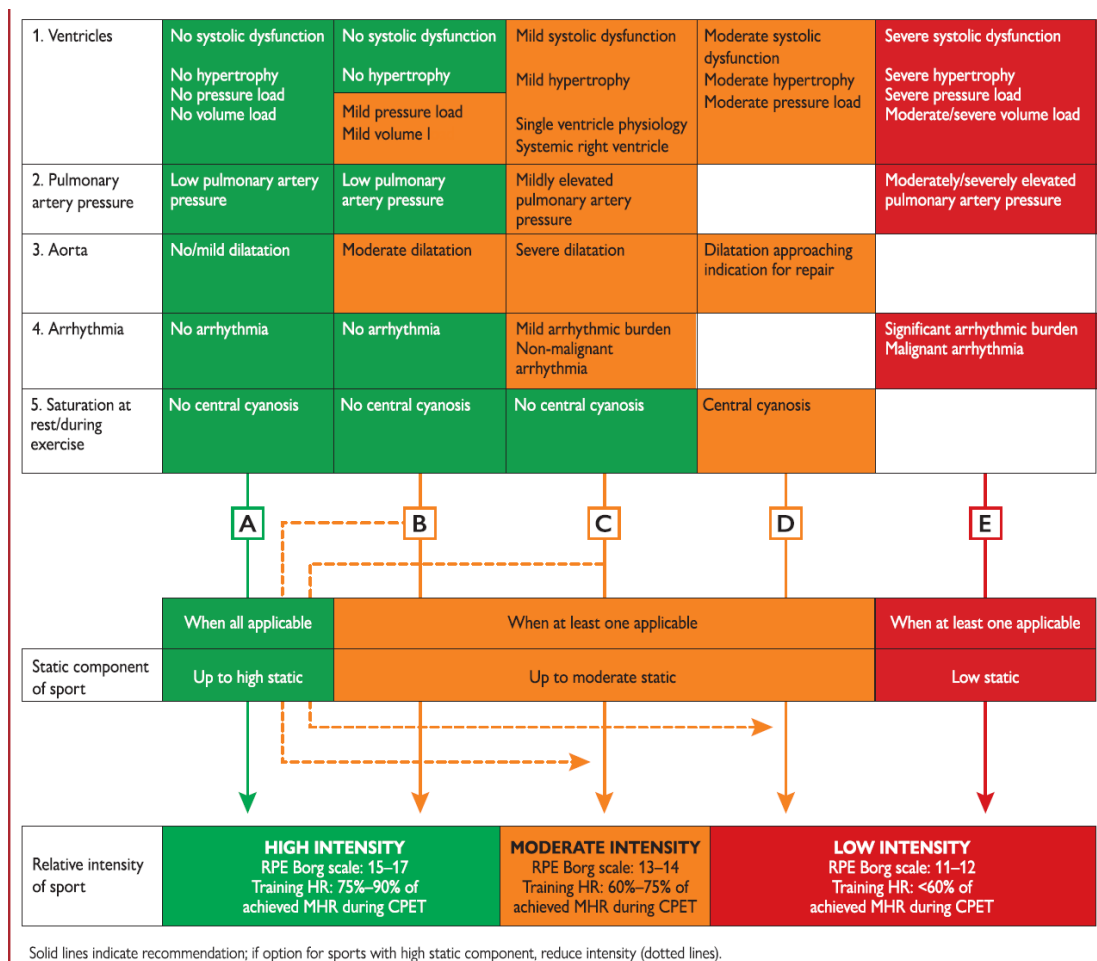


Figure 5. ESC 2020 GL on Sports. Pre-participation assessment of individuals with congenital heart disease.

CPET = cardiopulmonary exercise test; HR = heart rate; MHR = maximum heart rate; RPE = rate of perceived exertion. A_E represent pathways linking static and intensity components for each column. After assessment of CPET and the five variables, an individual recommendation can be given (solid arrow). If a higher static level sport is chosen, then a lower intensity level is advised (dotted arrow).

Table 20. Pregnancy in ACHD**ACC/AHA 2025 guideline on ACHD.****Reproductive Health, Pregnancy, and Contraception**

In adults with congenital heart disease of childbearing age, counseling on contraceptive options is recommended to inform pregnancy-related shared decision-making.	1 B-NR
Adults with congenital heart disease interested in pregnancy should receive preconception counseling, in collaboration with an ACHD cardiologist, about the maternal cardiac, obstetric, and fetal risks associated with pregnancy and the potential long-term risks to the pregnant individual.	1 B-NR
Screening fetal echocardiography is indicated to detect congenital heart disease if either parent has ACHD to optimize delivery planning and access to care.	1 C-LD
In pregnant patients with ACHD AP classifications IB–D, IIA–D, and IIIA–D, an individualized care plan, including mode of delivery, need for cardiac monitoring, and anesthetic support, should be documented in the medical record and communicated to the patient and care team, to reduce maternal risks for morbidity and mortality.	1 C-LD
In pregnant patients with ACHD AP classifications IB–D, IIA–D, and IIIA–D, management should be provided by, or in collaboration with, a cardio-obstetrics team with expertise in ACHD and maternal-fetal medicine, to improve clinical outcomes.	1 C-LD
Patients with ACHD who have high risk for maternal morbidity and mortality should be counseled to avoid pregnancy and given guidance on effective contraception to improve outcomes.	1 C-LD
For pregnant patients with ACHD who have high risk for morbidity and mortality, the option of terminating pregnancy should be discussed to reduce that risk.	1 C-LD
In patients with ACHD who are of childbearing age, referral to a genetic counselor for preconception planning is reasonable to facilitate understanding of the risks for congenital heart disease in their offspring.	2a C-LD
For patients with ACHD who are considering assisted reproductive technology, discussions involving an ACHD cardiologist can be beneficial to increase patients' knowledge about their options and the associated risks.	2a C-EO
Routine cesarean delivery has no benefit and may cause harm in pregnant patients with ACHD in the absence of an obstetric indication or high-risk cardiac condition.	3 No Benefit C-LD
In patients with ACHD who are at high risk for thromboembolic events, contraceptive agents that contain estrogen are potentially harmful.	3 Harm B-NR

Congenital Heart Conditions Considered Significantly High Risk for Maternal Complications in Pregnancy*

Congenital Heart Lesions

Severe, symptomatic, left-sided obstructive lesions (mitral stenosis, subaortic and aortic stenosis)

Severe native coarctation or recoarctation

Cyanotic congenital heart disease or Eisenmenger syndrome

Fontan physiology

Systemic right ventricle with at least moderately depressed ventricular function

Other Factors/Conditions

Advanced New York Heart Association functional class (III or IV)

Prior adverse cardiac event during pregnancy

Pulmonary arterial hypertension

Significantly decreased systemic ventricular dysfunction (ejection fraction <30%)

Aortopathy (diameter >5 cm or associated with connective tissue disorders)

Mechanical heart valves

Refractory ventricular arrhythmias

*Information adapted from the modified World Health Organization classification system.

ESC 2025 GL on Pregnancy.

Recommendations for congenital heart disease and pregnancy

Vaginal delivery is recommended in most women with ACHD. ^{161,163}	I	B
It is recommended that all women with Fontan circulation who wish to become pregnant receive counselling from the Pregnancy Heart Team regarding the high risk of pregnancy-related adverse events. ^{479,480,484,486,487}	I	C
It is recommended that women with a systemic RV (Mustard/Senning or congenitally corrected TGA), in NYHA class III/IV, systemic ventricular dysfunction (EF <40%), or severe TR wishing to become pregnant are counselled by the Pregnancy Heart Team regarding the high risk of pregnancy-related adverse events. ^{473–475}	I	C
In women with significant haemodynamic lesions, discussion about guideline-directed interventions is recommended prior to pregnancy.	I	C

ACHD, adult congenital heart disease; EF, ejection fraction; NYHA, New York Heart Association; RV, right ventricle; TGA, transposition of the great arteries; TR, tricuspid regurgitation.

ACHD	Maternal Risk	Obstetric and foetal risk	Monitoring	Pregnancy management and delivery
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Unrepaired cyanotic ACHD (without pulmonary hypertension)				
	HF, thrombosis, arrhythmia and endocarditis in $\geq 15\%$ ⁴⁸⁸	Degree of maternal hypoxaemia is the most important predictor of foetal outcome: 10% foetal loss if resting maternal blood saturation $>90\%$, chance of a live birth 12% if maternal oxygen saturation $<85\%$ ⁴⁸⁹	FU in expert centre	- Pre-pregnancy counselling about very high risk especially if maternal resting saturation $<85\%$⁴⁸⁹ - i.v. air filter

Table 21. ESC 2020 GL on ACHD. Congenital heart disease with high risk and extremely high risk for pregnancy

Significantly increased risk of maternal mortality or severe morbidity (mWHO class III) (cardiac event rate 19–27%)	Extremely high risk of maternal mortality or severe morbidity (mWHO class IV) ^a (cardiac event rate 40–100%)
Unrepaired cyanotic heart disease	Pulmonary arterial hypertension
Moderate LV impairment (EF 30–45%)	Severe LV impairment (EF $<30\%$ or NYHA class III–IV)
Systemic RV with good or mildly decreased ventricular function	Systemic RV with moderate or severely decreased ventricular function
Fontan circulation. If the patient is otherwise well and the cardiac condition uncomplicated	Fontan with any complication
Severe asymptomatic AS	Severe symptomatic AS
Moderate mitral stenosis	Severe mitral stenosis
Moderate aortic dilatation (40–45 mm in Marfan syndrome or other HTAD; 45–50 mm in BAV, 20–25 mm/m ² in Turner syndrome)	Severe aortic dilatation (>45 mm in Marfan syndrome or other HTAD, >50 mm in BAV, >25 mm/m ² in Turner syndrome)
Mechanical valve	Severe (re-)coarctation

AS = aortic stenosis; ASI = aortic size index; BAV = bicuspid aortic valve; CHD = congenital heart defect; EF = ejection fraction; HTAD = heritable thoracic aortic disease; LV = left ventricle/ventricular; mWHO = modified World Health Organization; NYHA = New York Heart Association; RV = right ventricle/ventricular; TOF = tetralogy of Fallot.

^aPregnancy should definitely be avoided in women with these conditions.

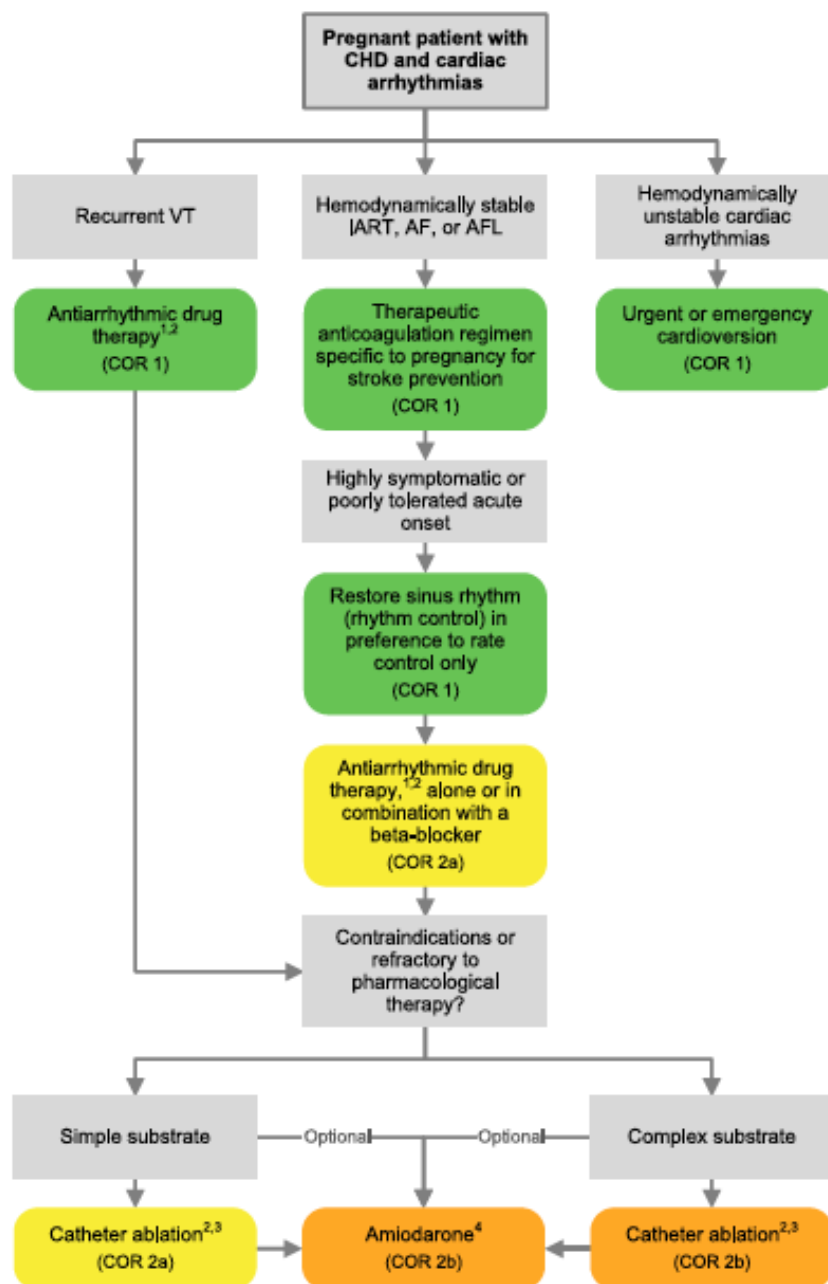


Figure 6. 2023 HRS expert consensus statement on the management of arrhythmias during pregnancy. Management algorithm for pregnant patients with congenital heart disease

1 Arrhythmic drug chosen according to the underlying cardiac substrate and potential impact of the drug on the developing fetus.

2 Preferably in conjunction with an electrophysiologist with expertise in CHD.

3 With attention to and techniques for eliminating or minimizing radiation exposure to as low as reasonably achievable.

4 After a risks-versus-benefits discussion with the patient.

AF: atrial fibrillation, AFL: atrial flutter, IART: intra-atrial reentrant tachycardia, VT: ventricular tachycardia.