

Table 1. AHA/ACC 2025 GL on ACHD. Established Risk Factors for Sustained Monomorphic Ventricular Tachycardia/Sudden Cardiac Death in Repaired Tetralogy of Fallot

QRS duration $\geq$ 180 ms
Moderate or greater LV or RV dysfunction
Prior ventriculotomy
Late age at definitive repair
Symptomatic nonsustained ventricular tachycardia/arrhythmic symptoms
QRS fragmentation
Inducible sustained ventricular arrhythmia at invasive electrophysiology study

LV indicates left ventricular; and RV, right ventricular.

Table 2. AHA/ACC 2025 GL on ACHD. Published Risk Scores for Calculating Risk for Sustained Ventricular Tachycardia/Sudden Cardiac Death in Repaired Tetralogy of Fallot

PREVENTION-ACHD ¹⁷		Brompton, UK ¹⁸		PACES ¹⁹	
Case-control design		Observational design		Case-control design	
Outcome: SCD		Outcome: SCD, sustained VT, resuscitated VF		Outcome: SCD, sustained VT, appropriate ICD shock	
783 patients		550 patients		286 patients	
8 events		29 events		70 events	
Risk factor*	Points	Risk factor†	Points	Risk factor* or †	Points
				Surgical era	<1980 ≥1980
Coronary artery disease	1	RV LGE extent		Arrhythmic symptoms	3 3
NYHA II-III symptoms	1	Moderate	22	≥ Moderate LV dysfunction	3 3
Supraventricular tachycardia	1	Severe	24	≥ Moderate RV dysfunction	2 1
Systemic EF <40%	1	LV LGE present	11	≥ Moderate RV pressure	1 1
Subpulmonary EF <40%	1	RVEF		≥ Moderate RV volume	1 1
QRS duration $\geq$ 120 ms	1	36%-47%	8	Age at repair <6.5 years	2 0
QT dispersion $\geq$ 70 ms	1	≤35%	16	Shunt pre-repair	2 1
		LVEF		Complex repair	1 2
		36%-55%	5	QRS duration $\geq$ 180 ms	1 1
		≤35%	17	Sustained/nonsustained VT	1 1
		Peak VO ₂ $\leq$ 17 mL/kg/m ²	9		
		BNP $\geq$ 127 ng/L	9		
		Akinetic RVOT length $\geq$ 55 mm	9		
		RV pressure $\geq$ 47 mm Hg	5		
Total score	Annual event rate	Total score	Annual event rate	Total score	Risk category
0-2	<1%	0-20	0.20%	<3	Low
3	1%-2%	21-39	0.90%	3-6	Moderate
4	3%-4%	≥40	3.70%	7-9	High
5	5%-10%			>9	Very high
6	11%-25%				
7	>25%				

ACHD indicates adult congenital heart disease; BNP, B-type natriuretic peptide; CMR, cardiovascular magnetic resonance; EF, ejection fraction; ICD, implantable cardioverter-defibrillator; LGE, late gadolinium enhancement; LV, left ventricular; NYHA, New York Heart Association; RV, right ventricular; RVOT, right ventricular outflow tract; SCD, sudden cardiac death; VF, ventricular fibrillation; and VT, ventricular tachycardia.
*Echocardiographic values.
†CMR values

Table 3. AHA/ACC 2025 GL on ACHD. Recommendations for adults with ToF**Diagnostic**

In adults with repaired TOF, echocardiography is recommended for assessment of pulmonary and tricuspid valve dysfunction, right atrial size, ventricular size and function, residual VSDs, and estimation of RV systolic pressure to characterize residual hemodynamic sequelae.	1 B-NR
In adults with repaired TOF and residual hemodynamic sequelae, CMR imaging is recommended for quantifying pulmonary valve function, ventricular size and function, and branch PA size and flow; characterizing the right and LVOT anatomy; quantifying residual shunts or aortopulmonary collateral flow; and assessing fibrosis of the ventricular myocardium.	1 B-NR
In adults with repaired TOF undergoing right ventricle-topulmonary artery (RV-to-PA) conduit stenting or transcatheter pulmonary valve placement with a balloon-expandable platform, assessment of coronary artery compression risk is indicated to ensure absence of coronary artery compression from stent/valve placement.	1 B-NR
In adults with repaired TOF and native RVOT anatomy being considered for transcatheter pulmonary valve replacement, cardiac CT is recommended to determine anatomic suitability.	1 B-NR
In adults with repaired TOF and moderate risk for future sustained ventricular tachycardia and/or SCD, invasive electrophysiology evaluation is reasonable to inform clinical management.	2a B-NR
In adults with repaired TOF and moderate risk for future sustained ventricular tachycardia and/or SCD, invasive electrophysiology evaluation is reasonable to inform clinical management in adults with repaired TOF in whom adequate data cannot be obtained noninvasively in the setting of an arrhythmia, heart failure, unexplained ventricular dysfunction, suspected pulmonary hypertension, residual hemodynamic lesions, or cyanosis, cardiac catheterization with angiography is reasonable for assessing hemodynamics to direct management and therapies.	2a C-EO
In adults with repaired TOF undergoing pulmonary valve placement (surgical or transcatheter), preprocedural invasive electrophysiology evaluation with programmed ventricular stimulation and 3-dimensional mapping may be considered to identify and treat latent monomorphic ventricular tachycardia substrates.	2a C-LD
In adults with repaired TOF, noninvasive evaluation (12-lead electrocardiography, contrast-enhanced MR imaging, and/or CT angiography) may be considered to characterize ventricular tachycardia substrates.	2b C-LD

Routine Follow-up and Testing Intervals

Type of Follow-Up or Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12-24	12-24	12	3-6
Electrocardiogram	24	12-24	12	12
Transthoracic echocardiogram	24	12-24	12	6-12

Table 4. AHA/ACC 2018 GL on ACHD. Recommendations for adults with ToF

Therapeutic

In symptomatic adults with repaired TOF and moderate or greater pulmonary valve dysfunction,* pulmonary valve replacement (surgical or transcatheter) is recommended for relief of symptoms.	1 B-NR
In asymptomatic adults with repaired TOF, moderate or greater pulmonary valve dysfunction,* and at least 2 of the following—RV endsystolic volume index >80 mL/m ² , RV end-diastolic volume ≥2X LV end-diastolic volume, RV ejection fraction ≤46%, LV ejection fraction ≤50%, or progressive decline in exercise capacity—pulmonary valve replacement (surgical or transcatheter) is reasonable for improving outcomes, including lowering risks for ventricular arrhythmias and death.	2a B-NR
In adults with repaired TOF and high risk (by clinical markers and/or multivariable score†) for SCD, placement of an ICD is reasonable to prevent SCD.	2a B-NR
In adults with repaired TOF and appropriate ICD therapies for monomorphic ventricular tachycardia, adjunctive catheter ablation is reasonable to reduce ventricular tachyarrhythmia burden.	2a B-NR
In adults with repaired TOF with moderate or greater pulmonary valve dysfunction* and progressive ventricular systolic dysfunction, pulmonary valve replacement (surgical or transcatheter) is reasonable to preserve ventricular function.	2a C-LD
In adults with repaired TOF with at least moderate pulmonary valve dysfunction* and progressive functional tricuspid regurgitation of a moderate or greater severity associated with RV dilation, pulmonary valve intervention may be reasonable to prevent worsening tricuspid valve dysfunction and RV dilation.	2b C-LD
For carefully selected patients with repaired TOF and hemodynamically tolerated monomorphic ventricular tachycardia related to a welldefined anatomical isthmus, ablation monotherapy may be considered	2b

in lieu of ICD placement at centers with ACHD electrophysiology expertise.²

In adults with repaired TOF and moderate or greater pulmonary regurgitation and ventricular tachyarrhythmia requiring treatment,[‡] pulmonary valve replacement (surgical or percutaneous), in addition to electrophysiological intervention, may be considered. 2b C-LD

*Pulmonary valve dysfunction defined as moderate PR (CMR-derived RF $\geq 25\%$) or RVSP $> 2/3$ systemic pressure due to RVOT obstruction. †Refer to Tables for individual risk scores.
‡Antiarrhythmic classes I/III or amiodarone, catheter ablation, electrical cardioversion, defibrillation, pace termination, or ICD shock for sustained ventricular arrhythmia.

Table 5. ESC 2020 GL on ACHD. Recommendations for intervention after repair of tetralogy of Fallot

PVRep in symptomatic patients with severe PR ^a and/or at least moderate RVOTO. ^b	I C
In patients with no native outflow tract, ^c catheter intervention (TPVI) if anatomically feasible.	I C
PVRep in asymptomatic patients with severe PR and/or RVOTO when one of the following criteria is present.	Ila C
• Decrease in objective exercise capacity. • Progressive RV dilation to RVESVi > 80 mL/m², and/or RVEDVi > 160 mL/m²^d, and/or progression of TR to at least moderate. • Progressive RV systolic dysfunction. • RVOTO with RVSP > 80 mmHg.	
VSD closure in patients with residual VSD and significant LV volume overload or if the patient is undergoing pulmonary valve surgery.	Ila C
Pre-operative catheter mapping and transsection of VT-related anatomical isthmuses before or during the intervention, in patients with sustained VT who are undergoing surgical PVRep or transcatheter valve insertion	Ila C
Electrophysiologic evaluation, including programmed electrical stimulation, for risk stratification for SCD in patients with additional risk factors (LV/RV dysfunction; non-sustained, symptomatic VT; QRS duration > 180 ms, extensive RV scarring on CMR).	Ila
ICD implantation in selected TOF patients with multiple risk factors for SCD, including LV dysfunction, non-sustained, symptomatic VT, QRS duration > 180 ms, extensive RV scarring on CMR, or inducible VT at programmed electrical stimulation.	Ila C
Catheter ablation or concomitant surgical ablation for symptomatic monomorphic sustained VT in those with a preserved biventricular function as an alternative to ICD therapy, provided that the procedure is performed in highly experienced centres and that established ablation endpoints have been reached (e.g. non-inducibility, conduction block across ablation lines).	Ilb C

CMR = cardiovascular magnetic resonance; ICD = implantable cardioverter defibrillator; LV = left ventricle/ventricular; PR = pulmonary regurgitation; PVRep = pulmonary valve replacement; RV = right ventricle/ventricular; RVESVi = right ventricular end systolic volume indexed; RVEDVi = right ventricular end diastolic volume indexed; RVOT = right ventricular outflow tract; RVOTO = right ventricular outflow tract obstruction; RVSP = right ventricular systolic pressure; SCD = sudden cardiac death; TOF = tetralogy of Fallot; TPVI = transcatheter pulmonary valve implantation; TR = tricuspid regurgitation; VSD = ventricular septal defect; VT = ventricular tachycardia.

a Regurgitant fraction by CMR >30-40%.

b Peak velocity >3 m/s.

c Patients with previous RVOT surgery using homografts, bovine jugular vein grafts, bioprostheses/conduits.

d Confirmed by repeated measurements.

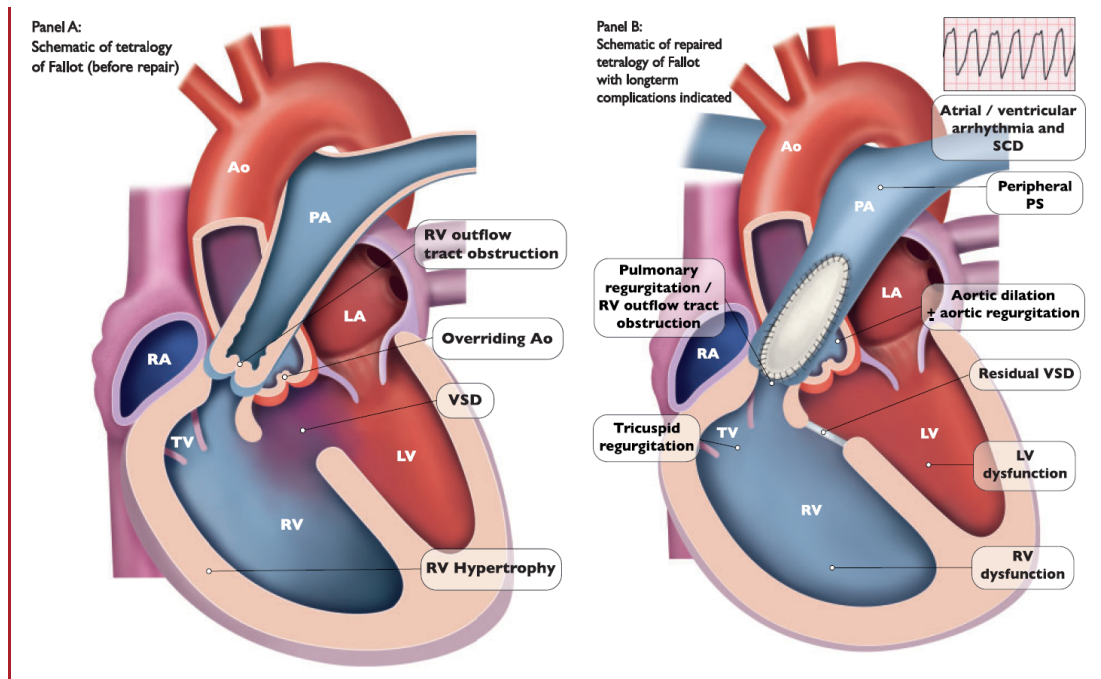


Figure 1. ESC 2020 GL on ACHD. Management of repaired tetralogy of Fallot: long-term complications to address during follow-up.

Ao = aorta; LV = left ventricle; RV = right ventricle; PA = Pulmonary artery; TV = tricuspid valve; RA = right atrium; RV = right ventricular; LA = left atrium; PA = pulmonary artery; PS = pulmonary stenosis; SCD = sudden cardiac death; VSD = ventricular septal defect.

Indications for Pulmonary Valve Replacement in Repaired Tetralogy of Fallot

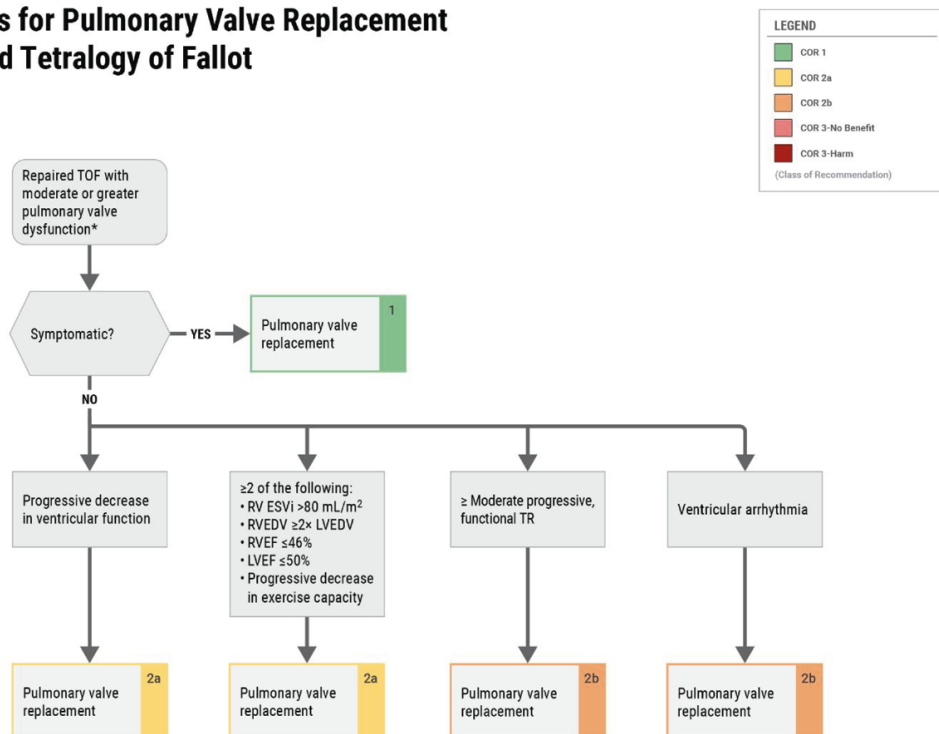


Figure 2. AHA/ACC 2025 GL on ACHD. Indications for Pulmonary Valve Replacement in Repaired Tetralogy of fallot

Table 6. 2025 ESC 2025 Guidelines on pregnancy.

ACHD	Maternal Risk	Obstetric and foetal risk	Monitoring	Pregnancy management and delivery
Repaired TOF				
	- Low risk if no residual lesions⁴⁶⁷ ↑ Risk of arrhythmia and HF (7%–10%) if pulmonary regurgitation, ↓ RV function, severe RVOTO^{467,468}	15% risk of foetal and obstetric complications, mainly pre-term delivery and low birth weight⁴⁶⁷ - Low foetal mortality (0.7%)⁴⁶⁷ - Recurrence risk in the offspring in 22q11 deletion syndrome: 50%	First trimester and at 28–32 weeks FU with TTE (increase FU depending on functional status)	- RV failure management: Bed rest and diuretics - Arrhythmia management⁴⁶⁸ - Severe PV stenosis/regurgitation: see above - Vaginal delivery is preferred - Consider caesarean section in severe RVOTO/PV stenosis