

Table 1. ACC/AHA 2025 GL on ACHD. Pulmonic Stenosis Severity*

Degree of stenosis	Doppler velocity and gradient across the right ventricular outflow tract
Mild	Peak gradient, <36 mm Hg (peak velocity <3 m/s)
Moderate	Peak gradient, 36–64 mm Hg (peak velocity 3–4 m/s)
Severe	Peak gradient, >64 mm Hg (peak velocity >4 m/s); Mean gradient >35–40 mm Hg

*Severity of pulmonary valve stenosis may be underestimated in the context of ventricular dysfunction.

Table 2. ACC/AHA 2018 GL on ACHD. Valvular Pulmonary Stenosis: 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	36–60	24	6–12	3–6
Electrocardiogram	36–60	24	12	12
Transthoracic echocardiogram	36–60	24	12	12

Table 3. ESC 2020 GL on ACHD. Recommendations for intervention in right ventricular outflow tract obstruction

In valvular PS, balloon valvuloplasty if anatomically suitable.	I C
RVOTO intervention at any level regardless of symptoms when the stenosis is severe (Doppler peak gradient is >64 mmHg ^a), provided that no valve replacement is required.	I C
If surgical valve replacement is the only option, it is indicated in patients with severe stenosis who are symptomatic. ^b	I C
If surgical valve replacement is the only option ^b in patients with severe stenosis who are asymptomatic, it is indicated in the presence of one or more of the following.	I C
• Objective decrease in exercise capacity. • Decreasing RV function and/or progression of TR to at least moderate. • RVSP >80 mmHg. • R_L shunting via an ASD or VSD.	
Intervention in patients with a Doppler peak gradient <64 mmHg in the presence of one or more of the following.	IIa C
• Symptoms related to PS. • Decreasing RV function and/or progressive TR to at least moderate. • R_L shunting via an ASD or VSD.	
Peripheral PS, regardless of symptoms, for catheter interventional treatment if $>50\%$ diameter narrowing, and RVSP >50 mmHg, and/or related reduced lung perfusion is present.	IIa C
ASD = atrial septal defect; PS = pulmonary stenosis; R_L = right-to-left; RV = right ventricle/ventricular; RVOTO = right ventricular outflow tract obstruction; RVSP = right ventricular systolic pressure; TR = tricuspid regurgitation; VSD = ventricular septal defect. a RVSP estimated from TR velocity should confirm severe PS. b The threshold for intervention is higher when a valve substitute is required because long-term risks, such as endocarditis and re-intervention for prosthetic valve failure, need to be taken into account.	

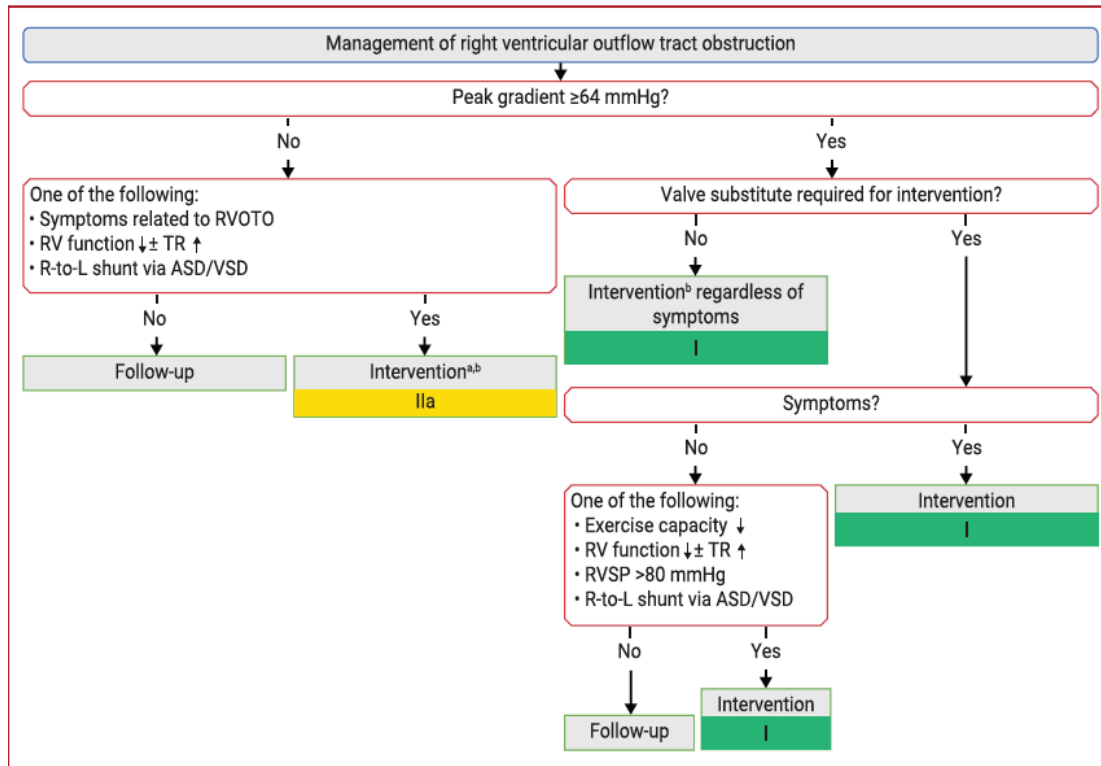


Figure 1. ESC 2020 GL on ACHD. Management of right ventricular outflow tract obstruction.

ASD = atrial septal defect; PS = pulmonary stenosis; R_L = right-to-left; RV = right ventricle/ventricular; RVOTO = right ventricular outflow tract obstruction; RVSP = right ventricular systolic pressure; TR = tricuspid regurgitation; VSD = ventricular septal defect.

a In peripheral PS, regardless of symptoms, catheter interventional treatment should be considered if $>50\%$ diameter narrowing and RVSP >50 mmHg and/or related reduced lung perfusion is present.

b In valvular PS, balloon valvuloplasty is the intervention of choice if anatomically suitable

Table 4. ACC/AHA 2025 GL on ACHD. Recommendations for Valvular Pulmonary Stenosis

In symptomatic adults with moderate or severe valvular pulmonary stenosis who have favorable valve anatomy, balloon valvuloplasty is recommended to improve symptoms.	1 B-NR
In symptomatic adults with moderate or severe valvular pulmonary stenosis who are ineligible for or have had suboptimal results after balloon valvuloplasty, pulmonary valve replacement is recommended to improve symptoms.	1 B-NR
In asymptomatic adults with moderate or severe valvular pulmonary stenosis and at least moderate tricuspid regurgitation or any RV dysfunction, pulmonary valve intervention is recommended to reduce pressure overload and improve valvular function.	1 C-LD
In asymptomatic adults with severe valvular pulmonary stenosis, intervention is reasonable to reduce pressure overload and improve valvular function.	2a C-EO

Table 5. ACC/AHA 2025 GL on ACHD. Recommendations for Isolated PR After Repair of Pulmonary Stenosis

Diagnostic

In adults with moderate or greater pulmonary regurgitation resulting from treated isolated valvular pulmonary stenosis, CMR imaging is recommended to establish anatomy, quantify the degree of regurgitation, and measure RV volumes and systolic function.	1 B-NR
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Therapeutic

In patients with symptomatic moderate or greater pulmonary regurgitation resulting from treated isolated valvular pulmonary stenosis, pulmonary valve replacement is recommended to improve symptoms.	1 B-NR
In asymptomatic patients with moderate or greater pulmonary regurgitation resulting from treatment of isolated pulmonary stenosis with RV systolic dysfunction, pulmonary valve replacement is reasonable to restore valve function and preserve RV systolic function.	2a C-LD
In asymptomatic patients who have moderate or greater pulmonary regurgitation resulting from treatment of isolated pulmonary stenosis and at least moderate tricuspid regurgitation and/or progressive RV dilation or progressive decline in exercise capacity, pulmonary valve replacement may be reasonable to restore valve function, ameliorate RV size, and preserve RV systolic function.	2b C-LD

Table 6. ACC/AHA 2025 GL on ACHD. Isolated Pulmonary Regurgitation After Repair of Pulmonary Stenosis: 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	36-60	24	6-12	3-6
Electrocardiogram	36-60	24	12	12
Transthoracic echocardiogram	36-60	24	12	12

Isolated Pulmonary Regurgitation After Repair of Pulmonary Stenosis

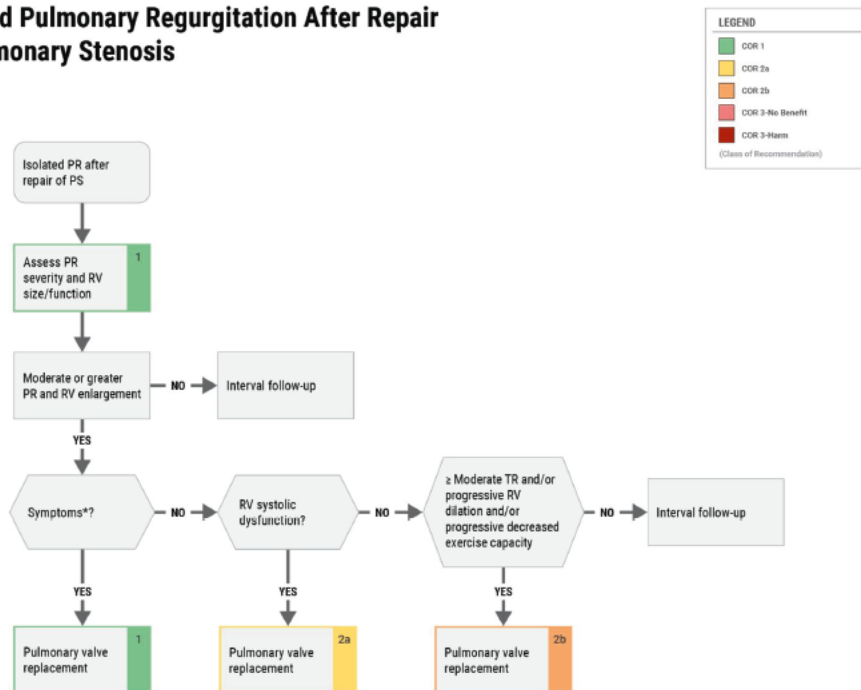


Figure 2. ACC/AHA 2025 GL on ACHD. Isolated PR After Repair of PS

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

Table 7. ACC/AHA 2025 GL on ACHD. Recommendations for Isolated Branch Pulmonary Stenosis

Diagnostic	
In adults with peripheral or branch PA stenosis, echocardiography and cross-sectional imaging (cardiac CT or CMR) are recommended to evaluate RV pressure and systolic function, degree of tricuspid regurgitation, and degree and location of stenosis. ¹	1 B-NR
Therapeutic	
In adults with symptoms related to peripheral or branch PA stenosis, pulmonary branch balloon angioplasty and/or stent implantation is recommended to improve hemodynamics and symptoms.	1 B-NR
In asymptomatic adults with isolated peripheral PA stenosis* and any of the following—RV hypertension that is more than half the systemic pressure, RV dysfunction, moderate or greater degree of tricuspid regurgitation, or worsening exercise performance—pulmonary branch balloon angioplasty and stent implantation can be useful to improve hemodynamics, reduce tricuspid regurgitation, and improve pulmonary blood flow.	2a B-NR
*Angiographic narrowing and/or asymmetric pulmonary blood flow with a reduction >10% in expected perfusion to the affected lung.	

Table 8. ACC/AHA 2025 GL on ACHD. Isolated Branch Pulmonary Artery Stenosis: 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	24-36	24	6-12	3-6
Electrocardiogram	24-36	24	12	12
Transthoracic echocardiogram	24-36	24	12	12

Table 9. ACC/AHA 2018 GL on ACHD. Recommendations for Management of Right Ventricle-to-PA Conduit

Diagnostic	
In patients undergoing RV-to-PA conduit stent implantation and/or transcatheter pulmonary valve implantation, preprocedural assessment of the risk for coronary artery compression should be performed to prevent coronary artery obstruction.	1 B-NR
In patients with a stent implanted within an RV-to-PA conduit who have unanticipated progression in conduit dysfunction, imaging should be performed to rule out conduit stent fracture.	1 B-NR
In patients with an RV-to-PA conduit, with or without transcatheter pulmonary valve implantation, unexpected conduit dysfunction with worsening pulmonary stenosis or pulmonary regurgitation should prompt an evaluation to rule out infective endocarditis and/or thrombus.	1 B-NR
In adults with an RV-to-PA conduit and unexplained sustained arrhythmia, heart failure symptoms, ventricular dysfunction, or cyanosis, cardiac catheterization is reasonable to assess the hemodynamics to guide further management.	2a C-LD
In adults with an RV-to-PA conduit being considered for transcatheter intervention, preprocedural coronary CT is reasonable to assess conduit calcification, anatomic characteristics, and coronary artery proximity to guide procedural planning.	2a C-LD
Therapeutic	
For adults with an RV-to-PA conduit, severe stenosis and/or severe regurgitation, and symptoms or worsening functional capacity or a sustained arrhythmia, conduit intervention should be performed to improve cardiovascular status. *	1 B-NR
For adults with an RV-to-PA conduit, moderate stenosis and/or moderate regurgitation, and symptoms or worsening functional capacity or a sustained arrhythmia, conduit intervention is reasonable to improve cardiovascular status. *	2a B-NR
For asymptomatic adults with an RV-to-PA conduit and severe stenosis and/or severe regurgitation, and reduced RV ejection fraction, RV dilation, or progressive tricuspid valve regurgitation to at least moderate, conduit intervention is reasonable to improve cardiovascular status. *	2a B-NR
*For criteria for interventions in patients with repaired TOF and an RV-to-PA conduit, refer to “Tetralogy of Fallot.”	

Table 10. ACC/AHA 2025 GL on ACHD. RV to PA Conduits: 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	N/A	12	6-12	3-6
Electrocardiogram	N/A	12	12	12
Transthoracic echocardiogram†	N/A	12	12	6-12

Table 11. ESC 2020 GL on ACHD. Recommendations for intervention in right ventricular to pulmonary artery conduits

Intervention with preference for catheter intervention (TPVI) if anatomically feasible for symptomatic patients with RVSP >60 mmHg (may be lower in case of reduced flow) and/or severe PR^a I C

Intervention, preferably catheter intervention (TPVI) if anatomically feasible, for asymptomatic patients with severe RVOTO and/or severe PR. when at least one of the following criteria is present: IIa C

- Decrease in objective exercise capacity (CPET).
- Progressive RV dilation to RVESVi > 80 mL/m², and/or RVEDVi > 160 mL/m², and/or progression of TR to at least moderate.
- Progressive RV systolic dysfunction.
- RVSP >80 mmHg.

CPET = cardiopulmonary exercise testing; CMR = cardiovascular magnetic resonance; PR = pulmonary regurgitation; RV = right ventricle/ventricular; RVEDVi = right ventricular end diastolic volume indexed; RVESVi = right ventricular end systolic volume indexed; RVOTO = right ventricular outflow tract obstruction; RVSP = right ventricular systolic pressure; TPVI = transcatheter pulmonary valve implantation; TR = tricuspid regurgitation.

a Regurgitant fraction by CMR >30-40%.

Table 12. ACC/AHA 2025 GL on ACHD. Management of Double-Chambered Right Ventricle (DCRV)

Diagnostic

In adults with RV dysfunction or symptoms attributed to DCRV, cardiac catheterization is reasonable to characterize anatomy and severity. 2a C-LD

In adults with DCRV and moderate or greater RVOT obstruction, diagnostic assessment with CMR (or, if contraindicated, cardiac CT) can be beneficial to characterize the anatomy. 2a C-EO

Therapeutic

In adults with DCRV, moderate or greater RVOT obstruction, and otherwise unexplained symptoms of heart failure, cyanosis, or exercise limitation, surgical repair is recommended to improve symptoms. 1 C-LD

In asymptomatic adults with DCRV and severe RVOT obstruction, surgical repair is reasonable to prevent symptoms and adverse RV remodeling.

2a C-LD

Table 13. ACC/AHA 2025 GL on ACHD. Double-Chambered Right Ventricle: 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	24–36	24	6–12	3–6
Electrocardiogram	24–36	24	12	12
Transthoracic echocardiogram	24–36	24	12	6–12

Table 14. ESC 2025 GL on Pregnancy.

ACHD	Maternal Risk	Obstetric and foetal risk	Monitoring	Pregnancy management and delivery
RVOTO/PV stenosis	- Mild to moderate: low risk - Severe: RV failure and arrhythmia²¹	Very low complication risk^{462,463}	- Mild to moderate: TTE at 28–32 weeks - Severe stenosis:	<i>Pre-pregnancy severe RVOTO (Doppler peak gradient > 64 mmHg) or/and any signs of right HF:</i>
			(Bi)monthly TTE (focused on RV function)	- Intervention (at any level of the RVOT)²¹ <i>Severe symptomatic PV stenosis (not responding to bed rest and conservative management):</i> - Consider transcatheter balloon valvotomy²¹ <i>Consider caesarean section in severe RVOTO/PV stenosis⁴⁶³</i>
PV regurgitation	↑ Risk when impaired RV function⁴⁶²	Premature birth⁴⁶³	Bimonthly TTE if severe PV regurgitation and ↓ RV function	
Post pulmonary valve replacement (surgical or transcatheter without severe stenosis/regurgitation)	Low risk^{463–466}	Very low complication risk^{462,463}	TTE at 28–32 weeks	Vaginal delivery is preferred