

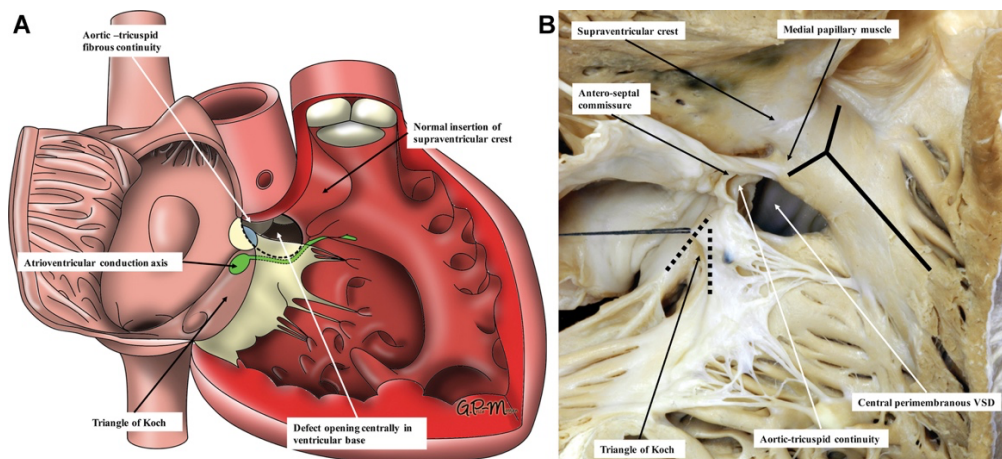


Figure 1. (A) Illustration of a central perimembranous defect. The hole is behind the septal leaflet of the tricuspid valve, below the right or noncoronary leaflets of the aortic valve, or both, and below the postero-inferior limb of the septal band. Notice the conduction pathway in green coursing along the postero-inferior rim. (B) Pathology specimen of a central perimembranous defect below the medial papillary muscle and posteroinferior limb of the septal band.

Lopez L, et al. Classification of Ventricular Septal Defects for the Eleventh Iteration of the International Classification of Diseases-Striving for Consensus: A Report From the International Society for Nomenclature of Paediatric and Congenital Heart Disease. *The Annals of thoracic surgery*. 2018;106:1578-1589.

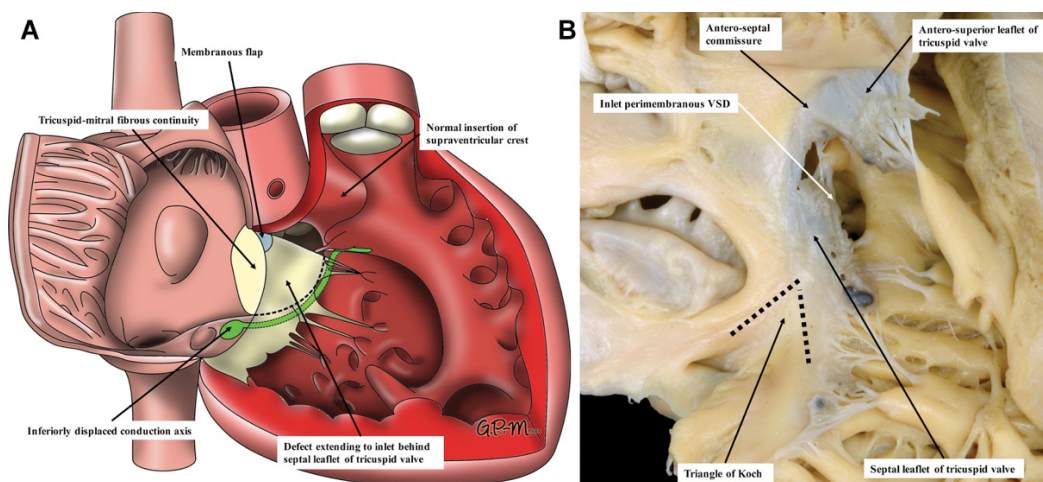


Figure 2. (A) Illustration of an inlet perimembranous defect with aligned atrial septum and ventricular septum. Notice the abnormally inferior atrioventricular node and conduction pathway coursing along the postero-inferior rim (dotted line). (B) Pathology specimen of an inlet perimembranous defect opening into the right ventricular inlet below the antero-septal commissure of the tricuspid valve; the defect extends along and is mostly covered by the septal leaflet of the tricuspid valve.

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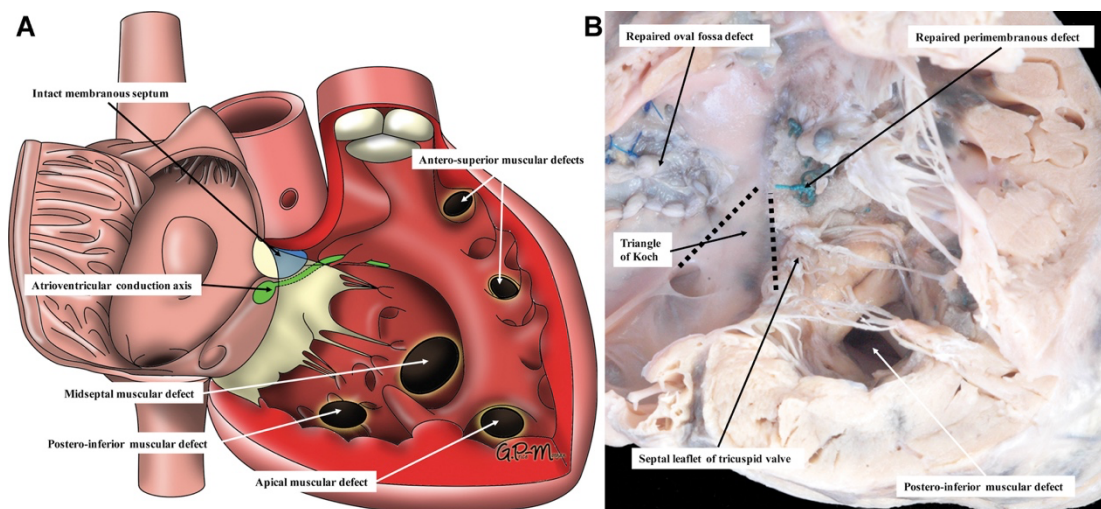


Figure 3. (A) Illustration of trabecular muscular defects located in the midseptal, postero-inferior, apical, and anterosuperior aspects of the muscular trabecular septum. (B) Pathology specimen of a large postero-inferior muscular trabecular defect with the inferior margin at the diaphragmatic wall of the ventricle.

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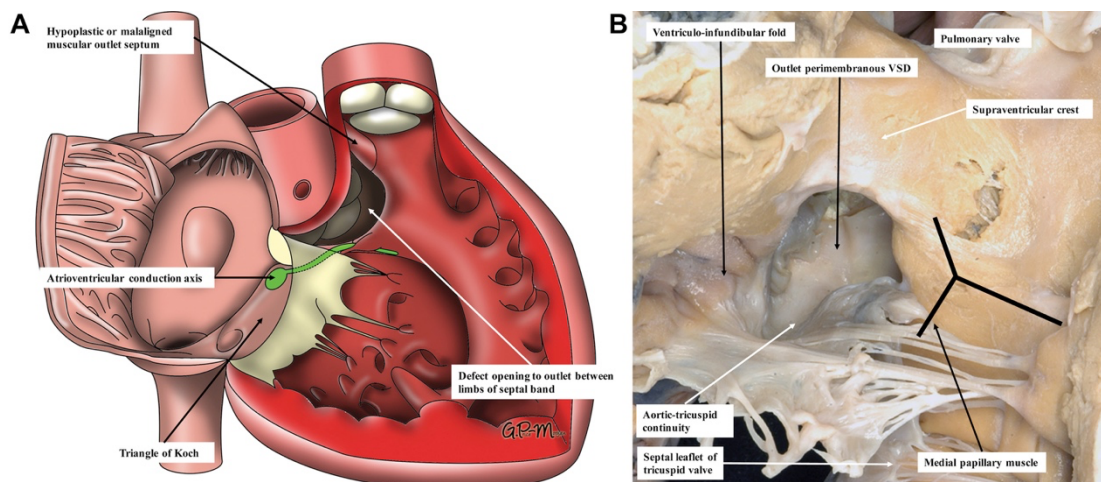


Figure 4. (A) Illustration of an outlet perimembranous defect located between the limbs of the septal band and extending to the area of fibrous continuity between the tricuspid and aortic valves. Notice the malaligned muscular outlet septum. (B) Pathology specimen of an outlet perimembranous defect opening into the right ventricular outlet between the limbs of the septal band.

Lopez L, et al. Classification of Ventricular Septal Defects for the Eleventh Iteration of the International Classification of Diseases-Striving for Consensus: A Report From the International Society for Nomenclature of Paediatric and Congenital Heart Disease. *The Annals of thoracic surgery*. 2018;106:1578-1589.

Table 1. ACC/AHA 20125GL on ACHD. Recommendations for ventricular septal defect.**Diagnostic**

Patients with an unrepaired VSD should be assessed for the presence of PAH to guide decisions about medical therapy and repair.	1 B-NR
In adults with a perimembranous VSD being considered for repair, evaluation to exclude right ventricular outflow tract (RVOT) obstruction or double-chambered right ventricle (DCRV) is recommended to improve management planning.	1 B-NR
Adults with a VSD (repaired or unrepaired) and PAH should be followed by specialists with expertise in ACHD and pulmonary hypertension to reduce morbidity and mortality	1 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	36-60	12-60	6-12	3-6
Electrocardiogram	36-60	12-60	12	12
Transthoracic echocardiogram	36-60	12-60	12	12

†Some physiological stage A patients with spontaneously closed muscular ventricular septal defects without residual anatomic or hemodynamic sequelae can be discharged from routine cardiology follow-up and be seen on an as-needed basis.

‡Stage B patients with a ventricular septal defect and a hemodynamically insignificant shunt who have normal chamber size without pulmonary hypertension can be seen every 36 to 60 months. Patients with ventricular dilation, dysfunction, or arrhythmia should be seen more frequently.

Table 2. ACC/AHA 2025 GL on ACHD. Recommendations for ventricular septal defect.**Therapeutic**

In adults with a VSD, a significant left-to-right shunt ($Q_p:Q_s \geq 1.5$), and significant or progressive LV dilatation with no evidence of PAH ($PVR \leq 2$ Wood units), closure of the defect is recommended to eliminate the shunt and preserve ventricular function.	1 B-NR
In adults with a VSD, LV volume overload, $Q_p:Q_s \geq 1.5:1$, and mild PAH ($PVR > 2$ but < 5 Wood units), closure of the VSD is reasonable to	2a C-LD

eliminate the shunt, in order to reduce the risk for progressive PAH and the burden of chronic volume overload on the LV.

In adults with an outlet or perimembranous VSD causing progressive moderate or greater aortic regurgitation, closure of the VSD is reasonable to preserve aortic valve function.	2a C-LD
In adults with a recent history of infective endocarditis involving a VSD, closure of the VSD may be reasonable to reduce the risk for recurrent endocarditis.	2b C-LD
In adults with a VSD, Qp:Qs ≥ 1.5 , and PAH (PVR 5 to 8 Wood units) without hypoxemia, if PVR drops to < 5 Wood units with PAH therapies, usefulness of closure of the VSD is uncertain to reduce the risk for progressive PAH and the burden of chronic volume overload on the left ventricle.	2b C-LD
In adults with a VSD, Qp:Qs < 1.5 , and no other indications, closure should not be performed to avoid unnecessary perioperative risks.	3 No benefit B-NR
In adults with a VSD and Eisenmenger physiology, VSD closure should not be performed to avoid escalating morbidity and mortality.	3 Harm B-NR

Table 3. ESC 2020 GL on ACHD. Recommendations for intervention in ventricular septal defect (native and residual)

VSD closure regardless of symptoms in patients with evidence of LV volume overload ^a and no PAH (no non-invasive signs of PAP elevation or invasive confirmation of PVR < 3 WU in case of such signs),	I C
VSD closure in patients with no significant L-R shunt, but a history of repeated episodes of IE.	IIa C
Surgeical closure in patients with VSD-associated prolapse of an aortic valve cusp causing progressive AR.	IIa C
VSD closure in patients who have developed PAH with PVR 3_5 WU, when there is still significant L_R shunt (Qp:Qs > 1.5).	IIa C
VSD closure in patients who have developed PAH with PVR > 5 WU, When there is still significant L_R shunt (Qp:Qs > 1.5), but careful individual decision in expert centres is required.	IIb C
VSD closure is not recommended in patients with Eisenmenger physiology and patients with severe PAH (PVR > 5 WU) presenting with desaturation on exercise. ^b	III C

AR = aortic regurgitation; IE = infective endocarditis; L_R = left-to-right; LV = left ventricle/ventricular; PAH = pulmonary arterial hypertension; PAP = pulmonary artery pressure; PVR = pulmonary vascular resistance; Qp:Qs = pulmonary to systemic flow ratio; VSD = ventricular septal defect; WU = Wood units.

a LV enlargement with increased stroke volume.

b There are limited data available for a precise cut-off, but by clinical experience, this would be given by a fall of arterial oxygen saturation $< 90\%$.

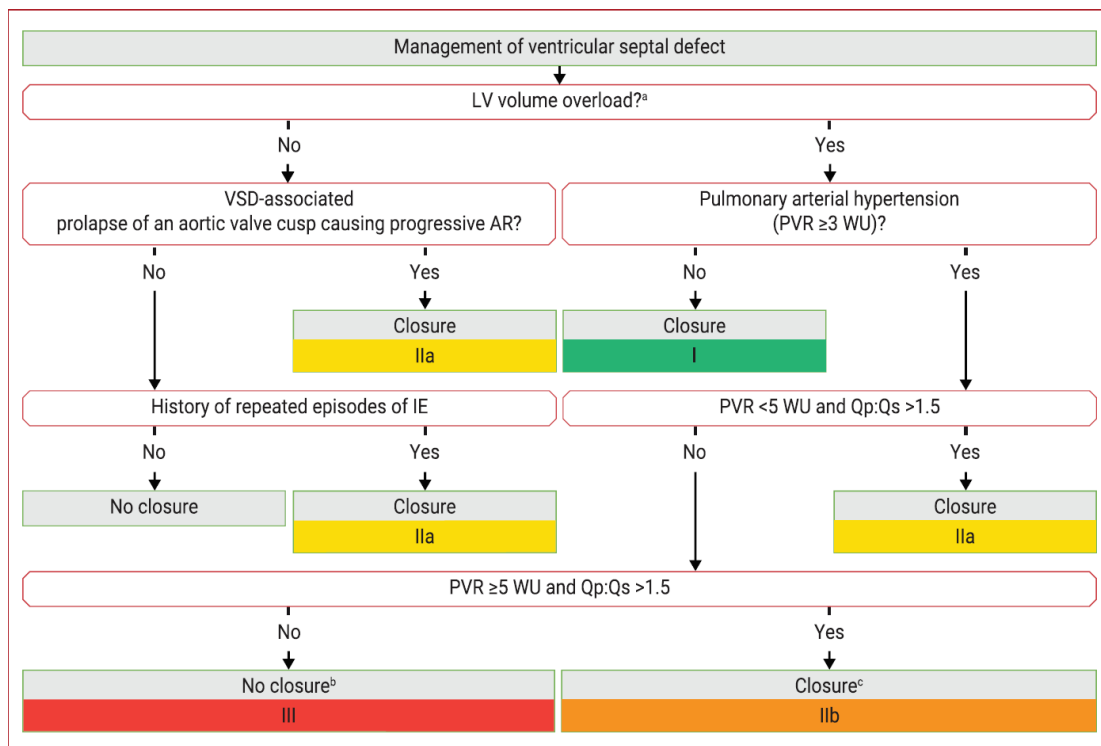


Figure 5. ESC 2020 GL on ACHD. Management of ventricular septal defect.

a LV enlargement with increased stroke volume.

b Includes all patients with desaturation at rest (Eisenmenger physiology) or on exercise.

c Careful individual decision in expert centres is required.

AR = aortic regurgitation; IE = infective endocarditis; LV = left ventricle/ventricular; Qp:Qs = pulmonary to systemic flow ratio; PAH = pulmonary artery hypertension; PVR = pulmonary vascular resistance; VSD = ventricular septal defect; WU = Wood units.

Closure Considerations for Ventricular Septal Defect

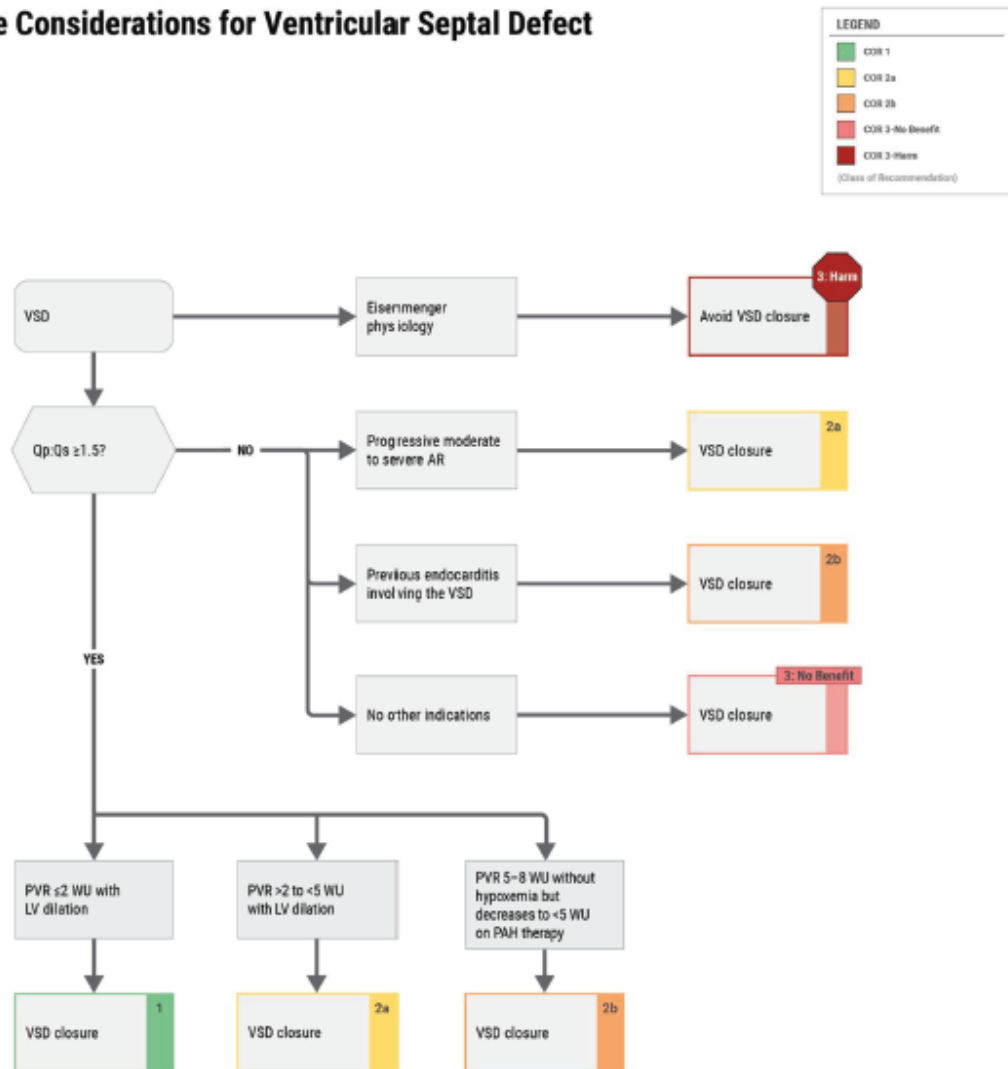


Figure 6. ACC/AHA 2025 GL on ACHD. Closure considerations for ventricular septal defect.

AR indicates aortic regurgitation; LV, left ventricular; PVR, pulmonary vascular resistance; Qp:Qs, pulmonary-to-systemic blood flow ratio; VSD, ventricular septal defect; and WU, Wood units.

Table 4. ESC 2025 GL on Pregnancy.

ACHD	Maternal Risk	Obstetric and foetal risk	Monitoring	Pregnancy management and delivery
VSD and patent ductus arteriosus	Low risk in small or repaired lesions with normal LV and no PAH ^a	No evidence for ↑ risk	<i>Unrepaired and uncomplicated VSD:</i> consider TTE at 28–32 weeks	Vaginal delivery is preferred