

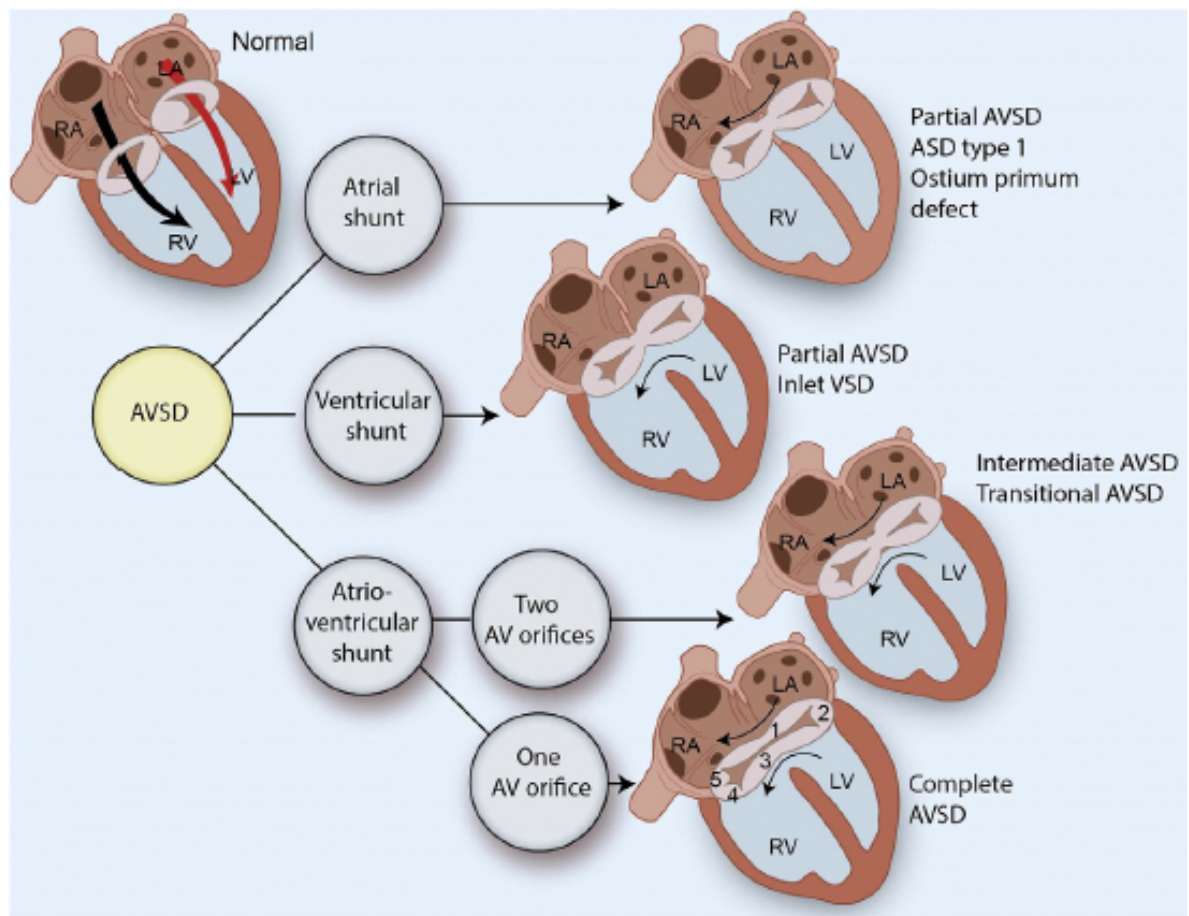


Figure 1. Schematic presentation of nomenclature of different types of AVSD. Defect can occur at atrial, ventricular or atrioventricular level. Black arrows indicate the defects, but do not necessarily represent blood flow direction.

RA=right atrium, LA=left atrium, RV=right ventricle, LV=left ventricle. In the complete AVSD the five leaflets are shown: superior bridging leaflet (1), left lateral (mural) leaflet (2), inferior bridging leaflet (3), right inferior leaflet (4) and right antero-superior leaflet (5).

Calkoen EE, et al. Atrioventricular septal defect: from embryonic development to long-term follow-up. *Int J Cardiol* 2016; 202:784-795.

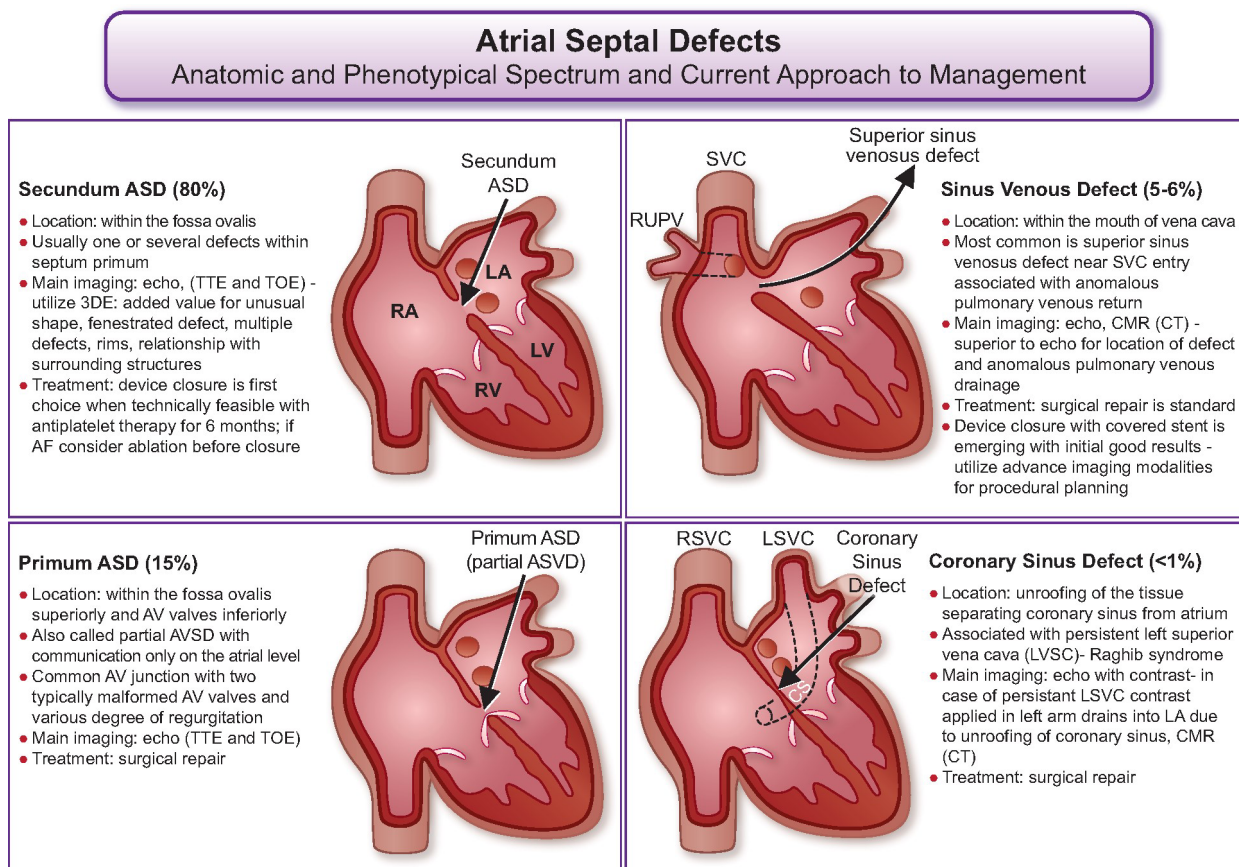


Figure 2. Spectrum of atrial septal defects.

ASD, atrial septal defect; AF, atrial fibrillation; AVSD, atrioventricular septal defect; CMR, cardiac magnetic resonance; CT, computed tomography; LSVC, left superior vena cava; RSVC, right superior vena cava; RUPV, right upper pulmonary vein; SVC, superior vena cava; TOE, transoesophageal echocardiography; TTE, transthoracic echocardiography; 3DE, three-dimensional echocardiography.

Brida M, et al. Atrial septal defect in adulthood: a new paradigm for congenital heart disease. *Eur Heart J*. 2022;43:2660-2671.

Table 1. ACC/AHA 2025 GL on ACHD. Recommendations for adults with atrioventricular septal defect.

Diagnostic

Adults with an unrepaired AVSD or those with residual shunts after prior repair should be assessed for the presence of PAH to guide medical therapy and assess suitability for repair. 1 C-LD

Adults with an AVSD (repaired or unrepaired) and PAH should be managed by clinicians with pulmonary hypertension expertise to improve outcomes. 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	24–36	12–24	6–12	3–6
Electrocardiogram	24–36	12–24	12	12
Transthoracic echocardiogram	24–36	12–24	12	12

Table 2. ACC/AHA 2025 GL on ACHD. Recommendations for adults with atrioventricular septal defect.

Therapeutic

In adults with repaired AVSD and left ventricular outflow tract (LVOT) obstruction with symptoms attributable to the obstruction, or LV systolic dysfunction (LV ejection fraction <50%), surgical repair is recommended to improve functional status and preserve ventricular function. 1 B-NR

In adults with repaired AVSD with a) symptomatic severe left atrioventricular valve regurgitation or b) asymptomatic severe left atrioventricular valve regurgitation with LV dilation (LV end-systolic diameter ≥ 40 mm) or LV systolic dysfunction (LV ejection fraction <60%), valve surgery is recommended to improve functional status and prevent worsening ventricular function. 1 B-NR

In adults with an unrepaired AVSD or those with residual shunts after previous repair, significant left-to right shunt ($Q_p:Q_s \geq 1.5$), and/or significant or progressive atrial/ventricular dilatation with no PAH ($PVR \leq 2$ Wood units), closure of the AVSD or residual shunt is recommended to improve functional class and clinical outcomes. 1 C-LD

In adults with an unrepaired AVSD or those with residual shunts after previous repair, significant left-to right shunt ($Q_p:Q_s \geq 1.5$), and/or 2a C-EO

significant or progressive atrial/ventricular dilatation with mild PAH (PVR >2 to <5 Wood units), closure is reasonable to improve functional class and clinical outcomes.

In asymptomatic adults with AVSD, LVOT obstruction, and progressive greater than mild aortic regurgitation, surgical repair may be reasonable to prevent worsening ventricular function and progressive aortic valve disease. 2b C-LD

In adults with an unrepaired AVSD or those with residual shunts after previous repair demonstrating Eisenmenger physiology or PVR >10 Wood units, shunt closure should not be performed to avoid additional morbidity and mortality. 3 Harm C-LD

Table 3. ESC 2020 GL on ACHD. Recommendations for intervention in atrioventricular septal defect

Complete AVSD

Surgical repair is not recommended in patients with Eisenmenger physiology, and patients with PAH (PVR > 5 WU) presenting with desaturation on exercise.^a

(see also recommendations for intervention in VSD)

Partial AVSD (primum ASD)

Surgical closure in patients with significant RV volume overload and should only be performed by a congenital cardiac surgeon. I C

(see also recommendations for intervention in ASD)

AV valve regurgitation

Valve surgery, preferably AV valve repair, in symptomatic patients with moderate to severe AV valve regurgitation, and performed by a congenital cardiac surgeon. I C

Valve surgery in asymptomatic patients with severe left-sided AV valve regurgitation, when LVESD > 45 mm and/or LVEF < 60% provided other causes of LV dysfunction are excluded. I C

Intervention in asymptomatic patients with severe left-sided AV valve regurgitation, preserved LV function (LVESD < 45 mm^b and/or LVEF > 60%), high likelihood of successful valve repair, and low surgical risk, when atrial fibrillation or systolic PAP > 50 mmHg is present. IIa C

Left ventricular outflow tract obstruction

See recommendations for intervention in Subaortic AS

ASD = atrial septal defect; AV = atrioventricular; AVSD = atrioventricular septal defect; LV = left ventricle/ventricular; LVEF = left ventricular ejection fraction; LVESD = left ventricular end systolic diameter; LVOTO = left ventricular outflow tract obstruction; PAH = pulmonary artery hypertension; PAP = pulmonary artery pressure; PVR = pulmonary vascular resistance; RV = right ventricle/ventricular; SubAS = subaortic stenosis; VSD = ventricular septal defect; WU = Wood units.

a 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%.

b Cut-off refers to average-sized adults and may require adaption in patients with unusually small or large stature.

Table 4. ESC 2025 GL on Pregnancy

ACHD	Maternal Risk	Obstetric and foetal risk	Monitoring	Pregnancy management and delivery
AVSD	• Low risk in repaired AVSD without significant residual lesions • Arrhythmia and ↑ AV valve regurgitation and HF if residual left AV valve regurgitation^{459,460} • ↑ Paradoxical emboli risk in unoperated (partial) AVSD	• Offspring mortality in 6% primarily due to recurrence of the congenital heart disease⁴⁵⁹	<i>Unrepaired and uncomplicated AVSD:</i> consider TTE at 28–32 weeks ↑ FU frequency in significant valve regurgitation, PAH, ↓ ventricular function, or ↑ NYHA class⁴⁶¹	• Residual shunt: see ASD and/or VSD • ↑ AV valve regurgitation and/or HF^f • PAH^e • Delivery: see ASD and VSD