

Clinical Guideline

# ATRIOVENTRICULAR SEPTAL DEFECT (AVSD) (ALSO KNOWN AS PRIMUM ASD WITH CLEFT MITRAL VALVE, ENDOCARDIAL CUSHION DEFECTS AND AV CANAL DEFECT)

<b>SETTING</b>	South West England and South Wales
<b>GUIDELINE FOR</b>	Cardiology teams in South West England and South Wales hospitals
<b>PATIENT GROUP</b>	Adult patients with congenital heart disease

## GUIDANCE

**Complete or partial. Most will be repaired. If unrepaired, partial, discuss for repair. If unoperated, complete, will have Eisenmenger's. See Cyanosis Guideline.**

**Follow-up:** 2-3 yearly if stable with mild LAVVR, annually if moderate LAVVR

**Associated lesions:** Down's syndrome in >75% complete AVSD, <10% partial Tetralogy of Fallot and other forms of complex CHD, isomerism, conotruncal abnormalities

**Inheritance:** 10–14% CHD recurrence if mother affected

**Long-term complications:** left AV valve regurgitation (less often stenosis, redo surgery needed in 5-10%)  
residual left-to-right shunt (uncommon)  
subaortic stenosis (5%) +/- AR, especially if left AV valve replacement  
atrial arrhythmias  
progression of AV block  
increased risk of PH in Down's, regardless of type

**Annually:**

**History:** usually asymptomatic  
exertional dyspnoea, fatigue  
palpitations

**Exam:** PSM at apex if LAVVR  
PSM at LLSE if VSD  
ESM at LUSE and split S2 if ASD  
ESM at LSE if LVOTO

<b>ECG:</b>	left axis deviation right bundle branch block first degree AV block common (may progress) sinus node dysfunction may occur atrial flutter/AF not uncommon with left AV valve regurgitation
<b>Echo:</b>	note lack of offset of AV valves residual VSD residual ASD morphology of left AV valve (may be abnormal lateral rotation of posteromedial papillary muscle) left AV valve regurgitation or stenosis LVOTO LV size and function estimated RVSP from right AV valve regurgitation
<b>Further Investigations:</b>	
<b>CXR:</b>	cardiomegaly if severe LAVVR and LV enlargement increased pulmonary vascular markings if significant left-to-right shunt if PH, prominent main pulmonary artery and pruning of distal pulmonary vessels
<b>CPET:</b>	baseline or to assess functional capacity
<b>Holter:</b>	if symptomatic only
<b>TOE:</b>	TEE to determine exact anatomy of left AV valve (if unclear after TTE) and severity/mechanism of LAVVR
<b>Catheter:</b>	to assess PVR if estimated sPAP >40mmHg non-invasively.
<b>EP study:</b>	for refractory atrial arrhythmias.
<b>MRI:</b>	to establish situs and connections, ventricular volumes can estimate Qp/Qs
<b>Drugs:</b>	none unless heart failure
<b>Pregnancy:</b>	well tolerated if repaired, unless impaired left ventricular function with severe left AV valve regurgitation. contra-indicated in Eisenmenger's anticoagulation management required in patients with mechanical valves
<b>Contraception:</b>	avoid combined pill in pulmonary hypertension and mechanical valves
<b>Endocarditis:</b>	antibiotic prophylaxis before high-risk dental work if prosthetic valve, previous endocarditis, residual defects at the site of or adjacent to the site of prosthetic material, or unrepaired/palliated cyanotic CHD and for first 6 months after procedure involving implanting prosthetic material

### Discuss if:

- unoperated with sustained atrial arrhythmias, impaired ventricular function, right ventricular volume overload, symptoms, heart failure, paradoxical embolism or reversible pulmonary hypertension
- symptomatic severe left AV valve regurgitation
- asymptomatic severe left AV valve regurgitation with LV enlargement/ deterioration in ventricular function (LVESD  $\geq$  45mm and/or LVEF  $\leq$  60% if no other cause of LV dysfunction)
- asymptomatic severe left AV valve regurgitation when LV is preserved but AF or systolic PAP > 50mmHg, if repair likely and low risk
- significant subaortic obstruction (see subaortic stenosis guideline)

**RELATED DOCUMENTS**      Regional Referral Guidance for Adult Patients with Congenital Heart Disease  
Regional Referral Pathway for Cardiac Disease in Pregnancy

**REFERENCES**

- Baumgartner H et al. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2020 00, 1-83.
- Stout et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease. Journal of the American College of Cardiology Aug 2018, 735-1097.  
<http://www.onlinejacc.org/content/early/2018/08/09/j.jacc.2018.08.1029>
- Canadian Adult Congenital Heart Network ([www.cachnet.org](http://www.cachnet.org))

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**SAFETY**                      None

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