Clinical Guideline

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

SETTING	South West England and South Wales
GUIDELINE FOR	Cardiology teams in South West England and South Wales hospitals
PATIENT GROUP	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/moderate LAVVR, annually if severe
Associated lesions:	Down's syndrome in >75% complete AVSD, <10% partial Tetralogy of Fallot and other forms of complex CHD, isomerism, conotruncal abnormalities
Inheritance:	5% 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

ECG:	left axis deviation right bundle branch block first degree AV block common (may progress) sinus node dysfunction may occir atrial flutter/AF not uncommon with left AV valve regurgitation
Echo:	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
Further Investigations:	
CXR:	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
CPET:	baseline or to assess functional capacity
Holter:	if symptomatic only
TOE:	TEE to determine exact anatomy of left AV valve (if unclear after TTE) and severity/mechanism of LAVVR
Catheter:	to assess PVR if estimated sPAP >40mmHg non-invasively.
EP study:	for refractory atrial arrhythmias.
MRI:	to establish situs and connections, ventricular volumes can estimate Qp/Qs
Drugs:	none unless heart failure
Pregnancy:	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
Contraception:	avoid combined pill in pulmonary hypertension and mechanical valves
Endocarditis:	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 ≥ 45mm and/or LVEF ≤ 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 subaotic stenosis guideline)

Appendix 1 – Evidence of Learning from Incidents

The following table sets out any incidents/ cases which informed either the creation of this document or from which changes to the existing version have been made.

Incidents	Summary of Learning
n/a	

Table A

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. • Canadian Adult Congenital Heart Network (www.cachnet.org)
RELATED DOCUMENTS AND PAGES	Regional Referral Guidance for Adult Patients with Congenital Heart Disease RegionalReferralGuidanceAdultPatientsWithCongenita-3.pdf Regional Referral Pathway for Cardiac Disease in Pregnancy ClinicalGuidelineForCardiacDiseasePreExistingOrPre-1.pdf
AUTHORISING BODY	Cardiac Executive Group, Bristol Heart Institute
SAFETY	None
QUERIES AND CONTACT	**Bristol:** Contact any of the following via UHBW switchboard – 0117 923 0000 Dr S Curtis Dr G Szantho Dr M Turner Dr R Bedair ACHD Specialist Nurse Team 0117 342 6599 **Cardiff:** via UHWales switchboard - 029 2074 7747 Dr S MacDonald Dr H Wallis Dr DG Wilson Dr N Masani ACHD Specialist Nurse Team 02920 744 580

AUDIT REQUIREMENTS	Adherence to guideline will be audited periodically as part of ACHD departmental audit

Plan Elements	**Plan Details**
The Dissemination Lead is:	Dr Stephanie Curtis
Is this document: A – replacing the same titled, expired SOP, B – replacing an alternative SOP, C – a new SOP:	A
If answer above is B: Alternative documentation this SOP will replace (if applicable):	
This document is to be disseminated to:	South West and South Wales Congenital Heart Network
Method of dissemination:	Email
Is Training required:	No

Document Change Control

Date of Version	Version Number	Lead for Revisions	Type of Revision	Description of Revision
Oct 2020	2	Consultant Cardiologist	None	Updated contacts and related documents Recurrence risk changed to 5% Follow up interval changed