Clinical Guideline

BICUSPID AORTIC VALVE DISEASE AND ASSOCIATED AORTOPATHY

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

Follow-up: depending on degree of AS/AR as per BSE guidelines; at least annual
4 yearly if BAV with no functional disease

Associated lesions: usually occurs in isolation but associated with:
coarctation of the aorta/arch hypoplasia (common)
other left-sided obstructive lesions, i.e. subaortic stenosis, parachute mitral valve, supramitral ring, known as Shone's syndrome
aortopathy (40-70%, of sinuses/ascending/arch, and dissection
VSD, PDA
Turner syndrome (associated with BAV and coarctation)

Inheritance: affects 1% population; 10-20% BAV in first degree relatives (screen). Maternal inheritance 5%, paternal 3%. Consider autosomal dominant aortopathies (e.g. mutations in ACTA2, MYH11) and also mutation in NOTCH1 if premature calcification.

Long-term complications: aortic stenosis/regurgitation
left ventricular hypertrophy/dilation/impairment
aortic root or ascending aortic dilation (may occur without functional valve disease)

Post-surgical intervention: recurrent aortic stenosis/regurgitation
damage to aortic or mitral valve
heart block
iatrogenic VSD

Post Ross: RV to PA conduit degeneration
pulmonary autograft degeneration
coronary abnormalities

At each visit:

History: usually asymptomatic
dyspnoea in severe AR - surgery
dyspnoea, syncope, angina in AS (heralds poor prognosis – urgent surgery)

Exam: ejection click
heaving apex in AS, thrusting in AR

ejection systolic murmur over aortic valve, radiating to carotids, +/- thrill at suprasternal notch in AS
early diastolic murmur in AR

ECG: left ventricular hypertrophy +/- strain
look for ischemic changes or arrhythmia

Echo: 3-5 yearly if mild AS/AR, 2 yearly if moderate AS/AR, 6-12 monthly if peak velocity ≥4m/s or severe AR with dilating LV
assess morphology, level of obstruction and associated lesions
peak and mean gradient
valve area (indexed)
measurements of aorta: annulus, sinus of Valsalva, sinotubular junction, ascending aorta, transverse arch
LV size/volume, systolic and diastolic function, strain (prognostic value)

Further investigations:

CXR: dilated aorta or calcification of aortic valve may be visible

CPET: to assess functional capacity, symptoms and physiology for risk stratification and timing of surgery
in borderline severe asymptomatic aortic stenosis in young adult when advising on athletic participation or pregnancy
dobutamine stress testing in low flow low gradient AS

Holter: not routine

TOE: to assess likelihood of valve repair in AR (rarely AS)

Catheter: rarely needed unless pre-Ross to assess coronary arteries if CT is inadequate

EP study: not usually indicated

MRI: mainly for assessment of aorta outside echo windows (whole aorta needs to be imaged lifelong); if < 40 mm, reimage every 5 years; if > /= 40 mm, 2-3 yearly
alternative method of quantifying AR

Bloods: BNP if severe AS (prognostic value)

Drugs: nothing slows progression of AS (including statins)
vasodilators not indicated for long-term therapy in AR, consider careful use if BP >140 mmHg (calcium channel blocker or ACE inhibitor)
◦ consider beta-blockers to delay/prevent aortic root dilatation/progression (benefit only shown in Marfan's or acute dissection)

Pregnancy: pre-pregnancy counselling unless mild disease
high risk in severe AS, aortopathy >50mm, or moderate LV impairment and mechanical prosthetic valves on warfarin
balloon dilation of severe AS during pregnancy can be considered

Contraception: avoid combined pill if on warfarin for mechanical valve

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

Discuss for surgery if:

Symptomatic with severe AS:
mean gradient ≥ 40mmHg or
mean gradient < 40 mmHg with reduced EF and evidence of flow reserve (low-flow/low-gradient severe AS)

Asymptomatic with severe AS
Abnormal exercise test with symptoms or drop in BP, LVEF<50%, abnormal strain or elevated BNP not due to another cause

Discuss anyway if:

- very severe AS (peak velocity ≥ 5.5 m/s)
- severe valve calcification and an increase in aortic velocity ≥0.3 m/s per year.
- BNP x3 normal
- sPAP >60mg on cath

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Severe AR with:

- Symptoms
- LV systolic dysfunction (LVEF ≤ 50%) if no other cause
- Severe LV dilatation (LVEDD >70mm or LVESD >50 mm (or 25 mm/m2)

Ascending aortic aneurysm with:

a. aorta diameter ≥5.5 cm
b. aortic diameter ≥50 mm with risk factors (family history of dissection, HT, coarctation, growth >3mm/yr, desire for pregnancy)
c. Turner syndrome if ASI ≥ 25mm/m2 if elongated transvers aorta, BAV, coarctation or hypertension, ≥27mm/m2 otherwise.

Other issues: Consider avoidance of mechanical valve in women of reproductive age
Counsel against competitive and strenuous isometric exercise in moderate to severe AS

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, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, Lung B, Kluin J, Lang IM, Meijboom F, Moons P. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2020. 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, 25255; DOI: 10.1016/j.jacc.2018.08.1029 Baumgartner H et al. 2017 ESC/EACTS Guidelines for the management of valvular heart disease. Eur Heart J 2017 38(36), 2739–2791 Erbel R et al. 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases. European Heart Journal (2014) 35, 2873–2926
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 Management of Aortopathy in Pregnancy ManagementOfAortopathyInPregnancy-3.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
Nov 2020	2	Consultant Cardiologist	Minor	Updated contacts and related documents. “Screen” added under inheritance. Follow up changed to 4 yearly for BAV with no functional disease. Otherwise as per BSE guidelines.