

Clinical Guideline

SUBAORTIC STENOSIS (OPERATED OR UNOPERATED)

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:	3-5 yearly if mild, annually if moderate, 6-12 monthly if severe
Associated lesions:	VSD AVSD conotruncal anomalies left-sided obstructive lesions (e.g. subaortic stenosis and coarctation; multi-level obstruction = Shone syndrome) hypoplastic LVOT and small aortic root. rarely, abnormal insertion of mitral valve or accessory mitral leaflet after patch closure of malaligned perimembranous VSD/AVSD
Inheritance:	rarely familial. 3% paternal inheritance; 5% maternal inheritance
Long-term complications:	increasing obstruction (post op 20% over 10 years) development/progression of AR (>50%, regardless of resection, increased risk with increasing obstruction) left ventricular dysfunction arrhythmias post-op heart block, and iatrogenic VSD
At each visit:	
History:	often asymptomatic dyspnoea, chest pain and syncope with severe obstruction dyspnoea with AR palpitations
Exam:	systolic murmur at left lower sternal border, radiating to carotids, no ejection click early diastolic murmur at left lower sternal border if AR
ECG:	may be LVH
Echo:	visualise LVOT anatomy severity of subvalvular obstruction (do not use modified Bernoulli equation – quote velocity) AR

LV size and function
associated lesions

Further investigations:

CXR: not routine
usually normal

CPET: to assess functional capacity, symptoms, ECG changes or
arrhythmias, or to assist in timing of surgery

Holter: not routine

TOE: useful for assessing LVOT anatomy and relationship between
subAS and AR. Essential in pre-op planning.

Catheter: usually not required unless non-invasive imaging inconclusive

EP study: for refractory atrial arrhythmias

MRI/CT: may be helpful in assess LVOT anatomy if echo inconclusive
can assess degree of AR and LV volumes and function

Drugs: only if LV dysfunction

Pregnancy: only high risk in severe, symptomatic subAS

Contraception: no limitations

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 and for 6 months
following surgery

Discuss if:

Symptomatic and
severe subAS (mean ≥ 40 mmHg on echo) or severe AR

Asymptomatic and
mean < 40 mm Hg but LVEF $<50\%$; or
severe AR and LVESD >50 mm (or 25mm/m²) and/or EF $<50\%$; or
mean Doppler ≥ 40 mmHg and marked LVH or fall in BP on exercise.

Consider discussion if low surgical risk and mean Doppler ≥ 40 mmHg or progressive AR

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	

REFERENCES	<ul style="list-style-type: none"> 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	<p>Regional Referral Guidance for Adult Patients with Congenital Heart Disease RegionalReferralGuidanceAdultPatientsWithCongenita-3.pdf</p> <p>Regional Referral Pathway for Cardiac Disease in Pregnancy ClinicalGuidelineForCardiacDiseasePreExistingOrPre-1.pdf</p>
AUTHORISING BODY	Cardiac Executive Group, Bristol Heart Institute
SAFETY	None
QUERIES AND CONTACT	<p>Bristol: Contact any of the following via UHBW switchboard – 0117 923 0000 Dr S Curtis Dr G Szanthy Dr M Turner Dr R Bedair ACHD Specialist Nurse Team 0117 342 6599</p> <p>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</p>
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	South West and South Wales Congenital Heart

to:	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
Dec 2020	2	Consultant Cardiologist	Minor	Updated contacts and related documents Follow up interval