

Clinical Guideline

## SUBAORTIC STENOSIS (OPERATED OR UNOPERATED)

<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

<b>Follow-up:</b>	annually
<b>Associated lesions:</b>	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
<b>Inheritance:</b>	rarely familial. 6-10% maternal inheritance
<b>Long-term complications:</b>	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
<b>At each visit:</b>	
<b>History:</b>	often asymptomatic dyspnoea, chest pain and syncope with severe obstruction dyspnoea with AR palpitations
<b>Exam:</b>	systolic murmur at left lower sternal border, radiating to carotids, no ejection click early diastolic murmur at left lower sternal border if AR
<b>ECG:</b>	may be LVH
<b>Echo:</b>	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

**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  $\geq 40$  mmHg on echo) or severe AR

Asymptomatic and  
mean  $< 40$  mm Hg but LVEF $<50\%$ ; or  
severe AR and LVEDD  $>50$ mm (or 25mm/m<sup>2</sup>) and/or EF $<50\%$ ; or  
mean Doppler  $\geq 40$  mmHg and marked LVH or fall in BP on exercise.

Consider discussion if low surgical risk and mean Doppler  $\geq 40$  mmHg or progressive AR

---

**RELATED DOCUMENTS** Regional Referral Guidance for Adult Patients with Congenital Heart Disease  
Regional Referral Pathway for Cardiac Disease in Pregnancy  
[Bicuspid Aortic Valve and Aortopathy Guideline](#)

**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, 25255; DOI: 10.1016/j.jacc.2018.08.1029

Canadian Adult Congenital Heart Network ([www.cachnet.org](http://www.cachnet.org))

**AUTHORISING BODY** Cardiac Executive Group, Bristol Heart Institute

**SAFETY** None

**QUERIES** **Bristol:** Contact any of the following via UHBristol switchboard – 0117 923 0000

Dr S Curtis, Consultant Cardiologist  
Dr G Szanthy, Consultant Cardiologist  
Dr M Turner, Consultant Cardiologist  
Dr R Bedair, Consultant Cardiologist

**Cardiff:** Specialist ACHD Team – Cardiff via UHWales switchboard - 029 2074 7747

Dr N Masani  
Dr H Wallis  
Dr DG Wilson  
ACHD Co-ordinator Elizabeth Corris – 02920 743 892  
ACHD Specialist Nurse Team Bethan Shiers / Sarah Finch 02920 744 580

**South Wales:** Lead Local Health Board Cardiologists:

Abertawe Bro Morgannwg LHB Dr H Wallis 01639 862049  
Dr C Weston & Dr M Heatley 01792 205666 ext 30836  
Aneurin Bevan LHB Dr P Campbell 01633 238863  
Cwm Taf LH Dr C Williams 01443 443642  
Hywel Dda LHB Dr H Wallis 01639 862049