Clinical Guideline

SUPRAVALVAR AORTIC STENOSIS

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: annual, progression in adults rare

Associated lesions: feature of William's syndrome (deletion at chromosome 7q11.23, elastin gene): elfin facies, outgoing personality, neurodevelopmental and multisystem manifestations including obstructive arteriopathy

- hypoplasia of entire aorta
- renal artery stenosis
- stenoses of other major aortic branches
- long-segment peripheral pulmonary artery stenosis

Inheritance: William's syndrome is autosomal dominant
familial supraAS caused by mutation in elastin gene, also AD inherited (as are associated lesions of arteriopathy)
rarely in homozygous hypercholesterolaemia
test all non-syndromic for elastin mutations

Long-term complications: progressive (or recurrent) obstruction, rare
AR in 25%, (usually not progressive after surgical relief)
patch repair aneurysm
coronary ischaemia (origin of the coronaries is usually proximal to obstruction, subjecting them to high systolic pressure and limited diastolic flow)
systemic hypertension
mitral regurgitation

At each visit:

History: dyspnoea, syncope or chest pain due to outflow obstruction
angina due to ischemia

Exam: systemic hypertension
heaving apex
suprasternal thrill
ejection systolic murmur
right arm systolic pressure higher than left (due to into right brachiocephalic artery)

ECG:	LVH, ischaemia
Echo:	anatomy of the LVOT and supra-aortic area degree of obstruction (Doppler across supraAS may overestimate pressure drop) LV size and function main and branch pulmonary artery anatomy and flow aortic and mitral valves diameter of the aortic sinuses, sinotubular junction, ascending aorta origins of the coronary arteries (difficult)
Further investigations:	
CXR:	not routine may be hypoplasia of ascending aorta
CPET:	to assess functional capacity other methods of stress testing can investigate ischemia and need to be considered periodically
Holter:	not routine
TOE:	can show anatomy, though CT/MRI best
Catheter:	may need to show anatomy and measure gradients, though CT usually best caution with coronary angiography as ostial stenosis common
EP study:	not usually needed
MRI/CT:	shows anatomy of lesion/repair and additional lesions in aorta and branches (carotid and renal artery stenosis) and pulmonary arteries image whole aorta, including renal arteries
Drugs:	not usually indicated
Pregnancy:	avoid pregnancy if moderate or severe obstruction, coronary involvement, or aortic disease
Contraception:	no combined pill if hypertension
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 if:

- Symptoms and/or severe obstruction (echo mean gradient ≥ 40mmHg)
- Mean echo gradient < 40mmHg and:
  - symptoms
  - LV systolic dysfunction (EF<50% without other explanation)

- when surgery for significant coronary disease is required

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, 25255; DOI: 10.1016/j.jacc.2018.08.1029 Canadian Adult Congenital Heart Network (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 Szantho, 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