Clinical Guideline

RIGHT VENTRICULAR OUTFLOW TRACT OBSTRUCTION (TO INCLUDE PERIPHERAL PULMONARY ARTERY STENOSIS AND DOUBLE-CHAMBERED RIGHT VENTRICLE)

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:

- Mild PS – discharge - (N.B. sub and supravalvar may progress)
- Moderate - 2 yearly
- Severe – annual
- Peripheral artery stenosis - 2-3 yearly, depending on severity

Associated lesions:

- Sub-infundibular stenosis (DCRV) - commonly associated with VSD.
- Infundibular stenosis - usually occurs with VSD, Fallot, and secondary to valvar PS (reactive myocardial hypertrophy).
- Valvar PS - usually isolated. Dilation of main PA can occur, independent of severity. Thin and doming; dysplastic valve less common (15-20%) and associated with Noonan Syndrome.
- Supravalvular PS (pulmonary arterial stenosis) - seldom occurs in isolation, may occur in Williams, Noonan, Alagille and, congenital rubella syndromes.

Inheritance: maternal 5%

Long-term complications:

- progressive obstruction post valvotomy/vavuloplasty (more common if residual gradient > 30 mmHg, seldom with subvalvar, supravalvar and DCRV repair)
- PR
- atrial arrhythmias

At each visit:

History: dyspnoea, chest discomfort, dizziness, or syncope during exertion.

Exam: consider syndromes above
pulmonary ESM, increasing with inspiration, radiating to back (heard laterally in peripheral PS)

	elevated JVP with prominent A wave RV heave wide splitting of S2 cyanosis possible if septal defect and right to left shunt
ECG:	if severe P pulmonale, right-axis deviation, and RVH
Echo:	thin doming valve in PS with dilated MPA or dyplastic level of obstruction (sub [infundibular], valvular or supravalvular) severity of obstruction by Doppler image branch PAs PR post valvuloplasty/valvotomy RV size/RVH/function (systolic interventricular septal flattening if severe obstruction) tricuspid regurgitation (TR velocity a more reliable estimation of RVSP) intracardiac shunting, especially right-to-left
Drugs:	if dynamic outflow tract obstruction, beta-blockers may improve filling time judicious diuretics if fluid overload or elevated right-sided pressure

Further investigations:

CXR:	RA may be large calcification of valve or conduit dilated MPA
CPET:	at baseline if >moderate and if considering intervention if asymptomatic
Holter:	not routine
TOE:	not usually required
Catheter:	to assess severity/level of obstruction if other imaging not adequate
EP study:	not routine
MRI/CT:	to assess RV size/RV volumes/function in PR imaging of the main, branch, and peripheral pulmonary arteries to identify the level of obstruction/conduits pulmonary perfusion (differential pulmonary flow)
Pregnancy:	low risk unless very severe PS, right to left shunt through ASD or RV failure
Contraception:	any
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
Exercise/sports:	avoid high-intensity and static sports if moderate PS, low-intensity sports only in severe PS

Discuss if:

Symptomatic severe (PG ≥64mmHg) or moderate PS
Aymptomatic severe PS if
catheter intervention is possible or
≥ one of:

1) objective decrease in exercise capacity
2) decreasing RV function and /or progression of TR to **≥** moderate
3) RVSP >80mmHg
4) R-L shunting via an ASD or VSD

Aymptomatic moderate PS if one of:

1) Symptoms
2) Decreasing RV function and /or progression of TR or at least moderate
3) R-L shunting via and ASD or VSD

Peripheral PA stenosis (regardless of symptoms)
If 50% diameter narrowing and RVSP >50 mmHg and/or reduced lung perfusion

Unrepaired double-chambered RV

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
Jan 2021	2	Consultant Cardiologist	Minor	Updated contacts and related documents. Follow-up changed to: - Mild PS – discharge - (N.B. sub and supravalvar may progress) - Moderate - 2 yearly - Severe – annual - Peripheral artery stenosis - 2-3 yearly, depending on severity. "Maternal 5%" added under inheritance.