Clinical Guideline

EBSTEIN'S ANOMALY

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: 2 yearly if up to moderate TR and good right ventricular function
annual if severee TR or any right ventricular dysfunction

Associated lesions: PFO /ASD >90%, accessory pathways common (multiple in 50%), VSD, anatomic/physiologic RVOT obstruction, LVOTO, mitral valve prolapse, LV non-compaction

Inheritance: rare, increased incidence if maternal exposure to lithium/benzodiazepines

Long-term complications:
RV failure (RV myopathy)
severe TR
atrial arrhythmias (approx. 25%)
ventricular arrhythmias/SCD (more likely if accessory pathway)
cyanosis if shunt right to left through ASD/PFO
paradoxical embolus
LVOTO (due to either leftward displacement of the IVS, or attachments of the anterior MV leaflet to the LVOT)

At clinic visit:

History:
dyspnoea
fatigue/reducing exercise capacity (low CO, RV dysfunction)
palpitations
syncope
TIA/stroke

Exam:
may be minimal
cyanosis if right-to-left shunt through PFO or ASD (do O2 sats)
if low cardiac output - reduced pulse volume and peripheral cyanosis
JVP often normal because of large complaint RA
loud S1, 1 or more systolic clicks
pan-systolic murmur lower left sternal edge, increasing on inspiration
peripheral oedema/ascites if right heart failure
hepatomegaly

ECG:	tall, broad P waves 1st degree AV block may be pre-excited QR pattern often seen in V1, may extend to V4 commonly RBBB
Echo:	adherence of tricuspid valve leaflets to underlying myocardium (failure of delamination) redundancy/ tethering/fenestrations of anterior tricuspid valve leaflet (assess suitability for repair) apical displacement of septal and posterior leaflets below the AV junction into RV (>8mm/m2) and rotation towards RVOT resultant 'atrialization' and dilatation of inflow of RV enlargement of RA RV size and function, TV annulus size degree of TR - usually severe - flow reversal in hepatic veins rarely seen, ensure colour box 'apical' enough PA pressure underestimated in severe TR LV morphology and function LVOTO assess other valves look for ASD/PFO
Further investigations:	
CXR:	not routine RA enlargement CTR depends on severity globe shaped heart with narrow pedicle (aortic root small)
CPET:	at baseline or if change in symptoms and nearing criteria for surgical referral
Stress echo:	to look for LVOTO 2 yearly or if symptoms
Holter:	for recurrent palpitations and if any syncope
TOE:	if considering surgery and leaflets not seen well on TTE or to show PFO/ASD
Catheter:	not usually required unless concerns about pulmonary hypertension
EP study:	if pre-excited or for drug refractory supraventricular arrhythmias difficult/lower success rate to due to large RA and multiple accessory pathways
MRI:	at baseline and every periodically to assess RV volume and function
Drugs:	anticoagulation if previous paradoxical embolus or AF

diuretics if right-sided heart failure (will not affect fatigue and dyspnoea related to low CO)

Pregnancy: low risk if minimal TR/good RV
if mod-severe TR, increase TR/RV dysfunction likely
risk of arrhythmias and paradoxical embolism
higher risk if cyanotic or troublesome arrhythmias

Contraception: avoid oestrogen-containing contraceptive pill due to high likelihood of inter-atrial communication

Endocarditis: antibiotic prophylaxis before high-risk dental work if prosthetic valve, residual defects at the site of or adjacent to the site of prosthetic material

Exercise/Sports: Patients with more than mild TR, ventricular dysfunction, shunting, arrhythmias or other complications - avoid heavy isometric exercise.

Discuss if:

- Progressive > moderate TR and/or symptoms (≥NYHA Class II)
- Progressive RV dysfunction/dilatation
- Cyanosis (resting oxygen sats < 90%)
- Appearance/progression of arrhythmias /pre-excitation
- Paradoxical embolism
- Following previous repair/ bio prosthetic TVR:
    - stenosis - gradient > 12mmHG; or
    - progressive TR with symptoms/decreased exercise tolerance; or
    - new RV impairment

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