Clinical Guideline

TGA (PREVIOUS MUSTARD OR SENNING PROCEDURE)

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
Associated lesions:	(sub) pulmonary stenosis, VSD, LVOTO and coarctation
Inheritance:	rare
Long-term complications:	Failure of systemic right ventricle Tricuspid regurgitation (systemic AV valve) Supraventricular tachyarrhythmia – most commonly cavotricuspid isthmus dependent flutter, then macro re-entrant tachycardia due to scar Bradyarrhythmias requiring pacing (15-20%) Ventricular arrhythmias (if RV dysfunction)- polymorphic VT, or VF, if poor RV function, monomorphic VT if secondary to scar SVC/IVC baffle obstruction (superior more common) SVC/IVC baffle leak (up to 25%) (causing L-R or R-L shunt) Less commonly PAH, residual VSD, dynamic subpulmonic stenosis, pulmonary venous obstruction (rare), and SCD
Annually:	
History:	sustained palpitations presyncope exertional dyspnoea
Exam:	right parasternal heave loud A2 tricuspid regurgitation ejection systolic murmur if subpulmonic outflow tract obstruction pan systolic murmur if VSD SVC syndrome if SVC baffle obstruction leg oedema, hepatomegaly, varices, cirrhosis if IVC baffle obstruction signs of heart failure
ECG:	sinus node dysfunction/junctional rhythm right-axis deviation and RV hypertrophy QRS duration

Echo:	RV size and function, including strain degree of tricuspid regurgitation baffle leak or stenosis gradient across LVOT (subpulmonic) pulmonary hypertension VSD LV size and function
Drugs:	Latest ESC guidelines – 'no data to support use of ACEI, ARB, B blocker or aldosterone antagonist in systolic dysfunction of systemic RV' *ACE inhibitors and beta-blockers benefit controversial* diuretics if clinical evidence of heart failure beta-blockers may precipitate heart block if sinus node dysfunction
Further investigations:	
CXR:	not routine narrow mediastinal shadow
CPET:	at baseline, if change in symptoms and if referring for transplant, to assess chronotropic response, functional capacity and for tachyarrhythmias on exercise. Desaturation on exercise may imply baffle leak in patient who is asymptomatic at rest.
Holter:	if clinically indicated
Contrast echo:	to look for baffle leak
TOE:	to look for baffle obstruction/leak
Catheter:	to assess haemodynamics (including PVR), baffle leak/obstruction
EP study:	for refractory atrial arrhythmias (N.B. baffles will complicate access to atria)
MRI:	at baselines and every 3-5 years, to assess volumes, function and baffles (CT or catheter if pacemaker). Quantification of shunt related to baffle leak.
Pregnancy:	risk depends on RV function. Risk of prematurity and low birth weight. Long-term consequences on RV function not known
Contraception:	avoid combined pill if baffle leak or obstruction
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:

- New symptoms

- Significant /progressive tricuspid regurgitation (regardless of symptoms)
- Severe right or left ventricular dysfunction
- Symptomatic bradycardia, tachyarrhythmias or sick sinus syndrome
- Baffle leak resulting in a significant left-to-right shunt, any right-to-left shunt
- Baffle obstruction (more common in Mustard)
- Heart failure

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 only