

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

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’ <i>ACE inhibitors and beta-blockers benefit controversial</i> Diuretics if clinical evidence of heart failure Beta-blockers may precipitate heart block if sinus node dysfunction <i>digoxin may be used to reduce risk of fast ventricular rate in atrial tachyarrhythmia</i>
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: access to atria)	for refractory atrial arrhythmias (N.B. baffles will complicate
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

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 Committee

SAFETY No safety issues.

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