Clinical Guideline

Tetralogy of Fallot (Repaired)

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, two yearly if good repair (good RV function and mild PR/PS)

Associated lesions: branch PA stenosis/hypoplasia, right arch (25%), additional muscular VSDs, PDA, aortopulmonary collaterals (mainly in pulmonary atresia/VSD), left SVC, ASD (pentalogy of Fallot), AVSD (usually Downs'), coronary artery anomalies (most often LAD from RCA in 3-4%).

Inheritance: maternal inheritance 5%; 22q11 is autosomal dominant (especially consider if right arch, pulmonary atresia/MAPCAs), Alagille, Noonan, Williams, and Klippel Feil

Long-term complications:
residual PR with RV dilation/dysfunction
associated TR
residual RVOT obstruction
branch PA stenosis or hypoplasia
LV dysfunction
aortic root dilatation and progressive AR
residual VSD

Arrhythmias:
atrial arrhythmias (prevalence 20%)
atrial tachycardias (IART due to RA incision, RA dilatation)
atrial flutter/fibrillation (in 1/3 due to LA dilatation)
ventricular arrhythmias: polymorphic VT/VF (typically related to RV/LV dysfunction), and monomorphic VT.
Risk of SCD 1-3.5% per decade of follow-up

Predictors of VT/SCD:
QRS fragmentation and QRSd > 180 ms
LV systolic and diastolic dysfunction
Ventricular and atrial arrhythmias
Inducible VT at EP testing
RVH
RV dysfunction
long-standing palliative shunts
older age at the time of repair

At each visit:

History:
palpitations
pre-syncope, syncope

	exertional dyspnoea/fatigue
Exam:	Split second heart sound ESM if residual RVOTO ESM and EDM if significant PR, may be EDM of AR PSM if residual VSD absent P2 component of S2 absent arm pulses on the side of classical BT shunt
ECG:	SR, RAD and RBBB, may have RVH QRS duration > 180ms a risk factor for VT and SCD
Echo:	RV size and function, including strain degree of PR (PHT <100ms severe PR) gradient across PV and RVOT separately systolic PA pressure from TR, PAEDP from PR residual VSD LV function aortic root/AR
Drugs:	beta blockers reasonable first line treatment for all tachyarrhythmias.

Further investigations:

CXR:	right sided aortic arch in 25%
CPET:	At baseline. If deteriorating symptoms or asymptomatic but nearing criteria for intervention. To look for exercise induced arrhythmias.
Cardiac rhythm analysis:	for selected patients (if suspected or clinical arrhythmia)
Catheter:	To assess haemodynamics when non-invasive evaluation is non-conclusive, assess for PPVI, coronary angiogram pre-cardiac surgery
MRI:	At baseline and every 3-5 years (if not nearing surgery), to assess RV volumes and function, degree of PR and split flows to branch PAs and anatomy of PAs, residual shunts and LGE for scar. Annually if nearing criteria for intervention.
CT:	to assess suitability for PPVI and check for coronaries crossing RVOT; can image MAPCAs
EP study:	For refractory atrial arrhythmias. ILR and consider VT stim/ablation if syncope.
ICD:	Secondary prevention (IC) Primary prevention can be considered if very high risk of SCD.
Genetics:	Check 22q11 at baseline (blood sample to Southmead)
Pregnancy:	If unrepaired, pregnancy high risk

In repaired, risk depends on RV function/PR and TR. If haemodynamics good and no residual lesion, low risk.

Contraception: no restrictions

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: No restrictions if asymptomatic repaired TOF and good haemodynamics
Low intensity sports and avoid isometric exercise if high risk for clinical arrhythmias/SCD, advanced biventricular dysfunction.

Discuss if:

1. Pulmonary regurgitation
   - Severe PR and symptoms,
   - If asymptomatic but with moderate to severe dilatation RVEDV>160ml/m^2, RVESV>80ml/m^2, RVEF< 47% on MRI, progression of TR to at least moderate, progression of RV dysfunction, sustained arrhythmias or decreased exercise performance on CPET.

2. RVOT obstruction
   - RVOTO with RVSP >80mmHg
   - with RV dysfunction
   - arrhythmias
   - symptoms (low CO, syncope, dizziness, right heart failure)/deteriorating exercise performance
   - Branch PA obstruction with unbalanced pulmonary blood flow

3. Arrhythmias
4. Heart failure
5. Aortic root dilatation > 50mm or progressive growth (rare)
6. Severe AR with symptoms or LV enlargement/dysfunction (as per ESC guidelines)
7. Large RV outflow tract aneurysm
8. Residual VSD with a left-to-right shunt >1.5:1

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, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, Lung B, Kluin J, Lang IM, Meijboom F, Moons P. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2020.

	• Geva, Tal, et al. "Preoperative predictors of death and sustained ventricular tachycardia after pulmonary valve replacement in patients with repaired tetralogy of Fallot enrolled in the INDICATOR cohort." Circulation 138.19 (2018): 2106-2115. • 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) • Khairy P, Van Hare GF, Balaji S, Berul CI, Cecchin F, Cohen MI, et al. PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease. Heart Rhythm.11:e102-e65.
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. Maternal inheritance changed to 5%.