Clinical Guideline

CONGENITALLY CORRECTED TRANSPOSITION (CCTGA) ALSO KNOWN AS ATRIOVENTRICULAR DISCORDANCE AND VENTRICULOARTERIAL DISCORDANCE, DOUBLE DISCORDANCE, OR L-TGA

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: common (80-90%) - VSD (70%), usually perimembraneous, pulmonary or subpulmonary stenosis (40%) and systemic (tricuspid) valve anomalies (up to 90%, Ebstein-like in 30%)
Congenital complete heart block in 5%. Dextrocardia in 20%

Inheritance: rare

Long-term complications: 1) Systemic RV dysfunction and failure
2) Progressive TR (systemic AV valve)
3) LVOTO
4) Complete AV block (2% loss of AV conduction per year)
5) VT (extremely rare)
50% alive aged 40 with associated lesions, 50% aged 60 without

Annually:

History:
fatigue
dyspnoea
palpitation
syncope
progressive cyanosis if VSD/PS

Exam:
consider dextrocardia
RV heave
palpable S2 and loud A2)
pansystolic murmur at apex or lower left sternal border
ejection systolic murmur at left upper sternal border if PS/subPS
pansystolic murmur left sternal edge if VSD
may be cyanosis if VSD and PS

ECG: long PR interval

	complete heart block right and left bundle branches inverted, septal activation occurs right to left, so absent Q waves in left precordial leads, often present in inferior leads III and AVF, as well as V1 accessory pathways common, especially if Ebstein-like tricuspid valve
Echo:	reversed offset of AV valves in four chamber view great arteries difficult to see, parallel if seen (aorta anterior and to the left, PA posterior and to the right) tricuspid valve anomalies, may be Ebstein-like may be perimembranous VSD, may extend into the inlet septum may be pulmonary valve/subpulmonary obstruction (LVOTO) carefully assess tricuspid regurgitation and RV function (including strain) estimate PAP from MR jet
Further investigations:	
CXR:	narrow mediastinal shadow ascending aorta not visible on the right descending aorta and pulmonary artery may not be visible on the left ventricular silhouette has a “humped” appearance may be dextrocardia (20%) or mesocardia (relatively common)
CPET:	baseline, if symptoms change or if considering transplant to assess functional capacity and for chronotropic incompetence
Holter:	annual
TOE:	if need to assess tricuspid regurgitation further for potential surgery
Catheter:	to assess haemodynamics if pulmonary hypertension suspected
EP study:	for refractory atrial arrhythmias. If pacing required, consider CRT to preserve RV systolic function.
MRI:	at baseline to establish situs and connections 3-5 yearly to assess volumes, function and TR
Drugs:	management of arrhythmias and treatment of ventricular dysfunction little evidence to support the use of ACE inhibitors/ARB/beta-blockers caution with beta-blockers in view of risk of complete AV block
Pregnancy:	low risk unless impaired right ventricular function
Contraception:	no limitations
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: Symptomatic patients and preserved RVEF (>40%) - avoid high intensity; no more than moderate static and moderate –intensity. If low RVEF (<40%) and associated lesions - limit to low-static and low-intensity.

Discuss if:

- Moderate or greater systemic (tricuspid) AV valve regurgitation (intervention must be performed before EF<40% for benefit)
- New brady or tachyarrhythmia (urgent pacing if complete heart block)
- Heart failure (transplantation referral may be required)
- Significant cyanosis (< 90%) if VSD/PS -in the absence of severe pulmonary hypertension should be an indication for intracardiac repair

---

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	None	Updated contacts and related documents only