

Clinical Guideline

CYANOSIS IN CONGENITAL HEART DISEASE AND EISENMENGER'S SYNDROME

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:	12 months
Associated lesions:	depends on underlying lesion
Inheritance:	depends on underlying lesion

Long-term complications:

- **Hematologic:** erythrocytosis → hyperviscosity symptoms, usually when HCT > 65%; iron deficiency common - may manifest as symptoms similar to hyperviscosity at HCT < 65%; bleeding (up to 20%, variable severity, correlating with erythrocytosis)- ↑ PT and PTT, ↓ F V, VII, VIII and IX, platelet disorder, epistaxis, gingival bleeding, menorrhagia, and pulmonary hemorrhage most common.
- **Neurological:** cerebral hemorrhage due to haemostatic defects and anticoagulants; paradoxical emboli – stroke [higher risk if Fe deficient but not necessarily associated with higher HCT] and brain abscess; embolic events in atrial tachycardia; cognitive/psychosocial issues prevalent.
- **Renal dysfunction:** renal glomeruli are abnormal, hypercellular, and congested and eventually become sclerotic. Reduction of eGFR, increased creatinine, and proteinuria.
- **Rheumatologic:** gout (abnormal urate clearance plus increased turnover of RBCs [don't treat asymptomatic hyperuricemia - doesn't prevent gout], hypertrophic osteoarthropathy (HPOA, arthralgia and bone pain, especially legs ≤30%), scoliosis.
- **Gallstones:** increased breakdown of RBCs → increased risk of calcium bilirubinate gallstones.
- **Skin:** acne on the face and trunk, potential source of infection.

Additionally, in Eisenmenger's

- **Arrhythmias** (atrial fibrillation/flutter) and sudden death (30%)
- **Congestive heart failure** (25%)
- **Haemoptysis** (usually self-limiting, may be life-threatening, mostly due to bleeding bronchial vessels or pulmonary infarction, may be precipitated by stress/excitement or chest infection; CXR/CT to look for pulmonary hemorrhage or secondary cause (15%))
- **Angina** (due to RV ischemia, coronary artery compression by a dilated pulmonary artery)

At each visit:

History:

- 1) Hyperviscosity symptoms: headache, faintness, dizziness, fatigue, altered cognition, visual disturbances, paraesthesia, tinnitus and myalgia.
- 2) Heart Failure
- 3) Haemoptysis/Bleeding/Ischaemic complications
- 4) Syncope in Eisenmenger
- 5) Acute gouty arthritis/arthritis

Exam: cyanosis (less if iron deficient) and clubbing (differential in Eisenmenger PDA)
oxygen sats
scoliosis
acne

Investigations:

ECG: aim to maintain SR

Echo: depends on lesion, see PH protocol for Eisenmenger patients

Bloods: U&E, LFTs, clotting (corrected citrate tube if HCT>55%), uric acid, glucose (may be reduced due to erythrocytosis).
FBC iron deficiency is common but Hb should be high: FBC (MVC), iron levels (serum iron, iron saturation), ferritin, transferrin sats, folic acid, Vitamin B12 (if elevated/ normal MCV and low ferritin) WBC normal, plt low
Consider BNP/NT-pro-BNP, troponins, CRP

Further investigations:

CXR/MRI/CT: depends on lesion

CPET/6MWT: 6 minute walk test with sats more appropriate to assess functional capacity. Guides the initiation of PH medication and follow-up.

Holter: based on symptoms

Catheter: Right catheterization with compartmental oximetry is required for major decisions such as to start pulmonary vasodilators or surgery.

EP study: if arrhythmias unresponsive to drug treatment

Medical treatment

- 1) Social and psychological support
- 2) Flu vaccination
- 3) Avoid stressors: extreme heat, altitude, strenuous exercise, dehydration, surgery, GA, NSAIDs, radiopaque contrast unless pre-hydrated,
- 4) If hyperviscosity symptoms think 1) dehydration and 2) iron deficiency.
- 5) Iron supplementation if iron deficiency (MVC< 80fL, low iron stores) - be cautious of rebound - recheck Hb at 7–10days. Stop if dramatic ↑ RBCs. Single dose of oral iron daily

- is as good as tds dosing. Ferrinject as alternative.
- 6) Anticoagulation not indicated unless atrial flutter/AF (INR target 2-2.5). No data for NOACS.
 - 7) If haemoptysis:
 - stop aspirin/anticoagulation, treat hypovolemia and anaemia, rest
 - bronchoscopy (seldom useful), CTPA may reveal bleeding vessel
 - consider embolization of bronchial arteries if refractory
 - 8) Continuous O2 supplementation if pO2<60mmHg.
 - 9) In Eisenmenger with reduced exercise capacity (6MWT < 450m), endothelin receptor antagonist monotherapy should be considered followed by combination therapy if patient fails to improve (IIaB)
 - 10) Consider bisphosphonates if HPOA.

Important:

1) AVOID routine phlebotomy (depletes iron stores → iron-deficient red blood cells →increased risk of stroke)

2) Essential Surgery/intervention requires specialist advice

Pregnancy: high risk for fetus (viability unlikely if saturations <85%)
 contraindicated in Eisenmenger
 must be avoided in all cases of pre-capillary PH

Contraception: not for COCP/oestrogen containing preparations
 double contraception in Eisenmenger

Endocarditis: antibiotic prophylaxis recommended for all

Exercise/sport: gentle aerobic only recommended

Air travel: commercial air travel is well tolerated
 risk reduction strategies: avoid dehydration, alcoholic drinks
 and apply measures to prevent DVT

Exposure to high-altitude: acute exposure to > 2500m should be avoided.

Discuss if:

- Hyperviscosity symptoms, Hb > 200g/L, HCT > 65%, hydrated and iron replete (phlebotomy)
- Possibility of reducing cyanosis (see individual lesions)
- Refer all Eisenmenger's patients to the Pulmonary Hypertension Service
- Elective surgery needed

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
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n/a	
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Table A

REFERENCES	<p>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.</p> <p>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</p> <p>Rychik J, Atz AM, Celermajer DS, Deal BJ, Gatzoulis MA, Gewillig MH, Hsia TY, Hsu DT, Kovacs AH, McCrindle BW, Newburger JW, Pike NA, Rodefeld M, Rosenthal DN, Schumacher KR, Marino BS, Stout K, Veldtman G, Younoszai AK, d'Udekem Y; American Heart Association Council on Cardiovascular Disease in the Young and Council on Cardiovascular and Stroke Nursing. Evaluation and Management of the Child and Adult With Fontan Circulation: A Scientific Statement From the American Heart Association. Circulation. 2019 Jul 1.</p> <p>Greenway SC, Crossland DS, Hudson M, Martin SR, Myers RP, Prieur T, et al. Fontan-associated liver disease: Implications for heart transplantation. The Journal of Heart and Lung Transplantation 2016;35:26-33.</p>
RELATED DOCUMENTS AND PAGES	<p>Regional Referral Guidance for Adult Patients with Congenital Heart Disease RegionalReferralGuidanceAdultPatientsWithCongenita-3.pdf Regional Referral Pathway for Cardiac Disease in Pregnancy ClinicalGuidelineForCardiacDiseasePreExistingOrPre-1.pdf</p>
AUTHORISING BODY	Cardiac Executive Group, Bristol Heart Institute
SAFETY	None
QUERIES AND CONTACT	<p>Bristol: Contact any of the following via UHBW switchboard – 0117 923 0000 Dr S Curtis Dr G Szanthy Dr M Turner Dr R Bedair ACHD Specialist Nurse Team 0117 342 6599</p> <p>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</p>
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	Minor	Updated contacts and related documents. Follow up changed to 12 monthly. Under medical treatment: "Single dose of oral iron daily is as good as tds dosing. Ferrinject as alternative" and "Consider bisphosphonates if HPOA" added.