Clinical Guideline

FONTAN CIRCULATION (TOTAL CAVOPULMONARY CIRCULATION (TCPC))

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: dependent on the underlying abnormality, note may be isomerism
Inheritance: dependent on the underlying abnormality

Long-term complications:

1. Deterioration of ventricular function
2. AV valve regurgitation
3. Fontan obstruction ('peel', Dacron>GoreTex)
4. Right pulmonary venous obstruction -by enlarged RA in patients with RA-PA Fontan
5. Bradyarrhythmia -sinus node dysfunction and heart block – unmask during exercise testing, pace via coronary sinus or surgically
6. Atrial tachyarrhythmia -affects > 50% with A-P Fontan vs. 10-20% of patients with lateral tunnel/TCPC; can result in profound haemodynamic compromise/clot formation. Aim to restore sinus rhythm. Often resistant to drugs/EP. May need Fontan conversion and MAZE.
7. Thromboembolism -systemic and pulmonary, may be associated with AF, slow flow and clotting abnormalities (e.g. protein C deficiency). RA clot especially common in atriopulmonary Fontan.
8. Fontant associated liver disease (FALD) -congestion, dysfunction, cirrhosis, hepatocellular carcinoma (AFP may be normal) and varices
9. PLE -in 10%, poor prognostic sign - 5-year survival 50%. If obstruction, may be cured my relieving this. Associated with ascites, peripheral edema, pleural and pericardial effusions, chronic diarrhoea and elevated stool α1 antitrypsin levels with low serum albumin.
10. Plastic bronchitis - very poor prognostic sign
11. Cyanosis -due to fenestration, veno-venous collaterals draining to pulmonary veins/ LA/ conduit dehiscence or development of pulmonary AVMs.
12. Narrowing of the proximal descending aorta (post Norwood)

Annually:

History: reducing exercise capacity/fatigue

	dyspnoea palpitations syncope haemoptysis/bleeding diarrhoea/recurrent infections think PLE, wt loss think HCC
Exam:	may depend on original anatomy non-pulsatile and high JVP should be quiet on auscultation single S2 PSM for AV valve regurgitation EDM for AR absent or weak radial pulse post BT shunt hepatomegaly (think obstruction/high pressure, if Glenn this may be the only sign as JVP will not be up) signs of chronic liver disease sats – compare with previous if ascites, peripheral oedema, pleural effusions (look for PLE)
ECG:	dependent on original anatomy confirm sinus rhythm (compare to old ECG- may be asymptomatic slow IART. Junctional rhythm common)
Echo:	Biphasic flow (increases with inspiration) in SVC and hepatic veins (vmax should be <1.5m/s) systemic ventricular function- TVI values, M-Mode, and strain AV valve regurgitation aortic root and valve thrombus in right atrium fenestration on CFM pulmonary venous return-assess for a gradient in visible veins
Bloods:	FBC, clotting, U+E, ferritin, LFTs, GGT, BNP, αFP, serum protein and albumin. If low albumin, stool α 1 anti-trypsin for PLE. Baseline viral hepatitis and hemochromatosis screen (once only)
Liver imaging:	annual USS for cirrhosis/masses/ spleen size, portal HT annual Fibroscan (email to hepatology outpatient coordinator) if endoscopy/MRI/CT as directed by hepatology (Dr James Orr)
Further investigations:	
CXR:	not routine. Normal heart size and pulmonary vascularity. May see calcification of TCPC. If pleural effusions, search for PLE.
CPET:	baseline and routinely every 5 years-otherwise if symptoms change or if considering transplant
Holter:	if palpitations, pre-syncope or syncope

TOE: to assess AV valve regurgitation further for potential surgery

Catheter: if well, every 5-10 years
if new symptoms, ventricular dysfunction, arrhythmias, cyanosis or suspected obstruction
to assess haemodynamics/ obstruction and cause for worsening cyanosis (AVMs, collaterals)
creation of a fenestration may be needed to decrease Fontan pressure

EP study: if documented atrial arrhythmias. If pacing required- to discuss.

MRI: at baseline to confirm anatomy and assess function/patency of Fontan pathway, collaterals and pulmonary vein obstruction by enlarged RA, thrombus, CO
repeat if change in symptoms or ventricular function. If well, every 3-5 years.

CT: if suspected thrombus in Fontan or if MRI not possible due to pacemaker.

Drugs: Anti-coagulate. No robust data on NOACs.
ACE/ARB reasonable if systemic ventricular dysfunction
If right atrial isomerism need to be on penicillin and receiving annual pneumovax
no hard evidence for pulmonary vasodilators

Pregnancy: Relatively low risk to woman if uncomplicated Fontan. Avoid if any complication. High risk of miscarriage/severe IUGR/prematurity/fetal death (up to 60%). Pre-pregnancy counselling mandatory.

Contraception: not for COCP/oestrogen containing preparations

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: moderate symptom-limited aerobic exercise recommended

Discuss if:

13. New ventricular dysfunction/heart failure
14. Worsening exercise capacity/cyanosis
15. Tachy- or bradyarrhythmias (Fontan conversion may be considered if resistant tachyarrhythmia)
16. ≥Moderate AV valve or aortic regurgitation
17. Fontan obstruction
18. Sub-aortic obstruction
19. Progressive aortic root dilatation/narrowing with hypertension
20. Pulmonary venous obstruction
21. PLE (admission, IV Furosemide, albumin solution, sc heparin, spironolactone, prednisolone/budesonide, ACEI, dietitian consultation for high protein low fat diet and salt restriction)

22. Any hepatic complication (be alert for weight loss, derangement of PFTs and abdominal distension (liver malignancy))

RELATED DOCUMENTS

Regional Referral Guidance for Adult Patients with Congenital Heart Disease
Regional Referral Pathway for Cardiac Disease in Pregnancy

REFERENCES

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SAFETY

None

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