

Considerations for Advanced Heart Failure Consultation in Fontan Patients:

Guidance for primary cardiologists

BACKGROUND

To aid in decision-making on timing of referral of Fontan patients for advanced heart failure consultation with the aim of improving timely referral and facilitating collaborative care to enhance patient outcomes.

Patient population: Fontan patients

Considerations for referral by type of clinical Fontan dysfunction

(recognizing overlap exists between categories)

Cardiac/Systemic Ventricular Dysfunction

- 1) Severe¹ systolic dysfunction by echocardiogram, MRI, or cardiac catheterization.
- 2) Moderately depressed (by qualitative assessment) systolic function on imaging when accompanied by \geq moderate systemic AV valve regurgitation.
- 3) Significant growth derangement or failure to thrive including cachexia or linear growth failure
- 4) Decreasing exercise tolerance by patient report or as measured on sequential formal exercise testing or 6-minute walk
- 5) Significant electrophysiologic abnormalities, including recurrent arrhythmias despite therapy, implantation of a cardiac pacemaker, or aborted sudden cardiac death event

Fontan Pathway Dysfunction

- 1) Symptomatic, chronic fluid overload persisting despite new or increasing diuretic therapy
- 2) Occurrence of chronic pleural effusions or ascites, chylous or nonchylous, refractory to therapy and occurring outside the initial Fontan post-operative period
- 3) Major hemodynamic disturbance *resulting in symptoms* despite therapy including: low systemic cardiac output, diastolic ventricular failure, significantly elevated Fontan pressure, or symptomatic cyanosis

Lymphatic Dysfunction

- 1) Protein-losing enteropathy that has failed medical therapy and requires multiple hospital admissions in a 12-month period or PLE requiring repeated albumin infusions to treat symptoms despite standard PLE medical therapy
- 2) Plastic bronchitis requiring chronic therapy

Extra-cardiac Dysfunction

- 1) Hemoptysis requiring evaluation that is unrelated to an infection and persists after standard intervention
- 2) Liver disease with impaired synthetic function/abnormal liver function testing or undergoing evaluation for liver transplantation
- 3) Chronic kidney disease – Stage 3 or greater²

¹ Severe systolic dysfunction in a single ventricle can be graded by a qualitative assessment or by using calculated ejection fractions as follows: < 35% by echocardiogram or MR for a single LV; < 30% by MR for a single RV.

² Stage 3 CKD is an eGFR between 30 and 60 mL/min per 1.73 m²

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***Disclaimer:** The ACTION network is focused on quality improvement efforts such as harmonizing best practice protocols, disseminating them among institutions, and helping centers to improve care practices at the local level. This referral guideline was developed as a consensus tool for general cardiologists. The information in the guideline is based on center practices, individual opinions, experiences, and, where available, published literature. Providers & centers may choose to adapt this protocol to include in their center-specific protocols with reference to ACTION with the understanding that this is meant as a guideline and not as standard of care. (Revised: 05/30/2018)*