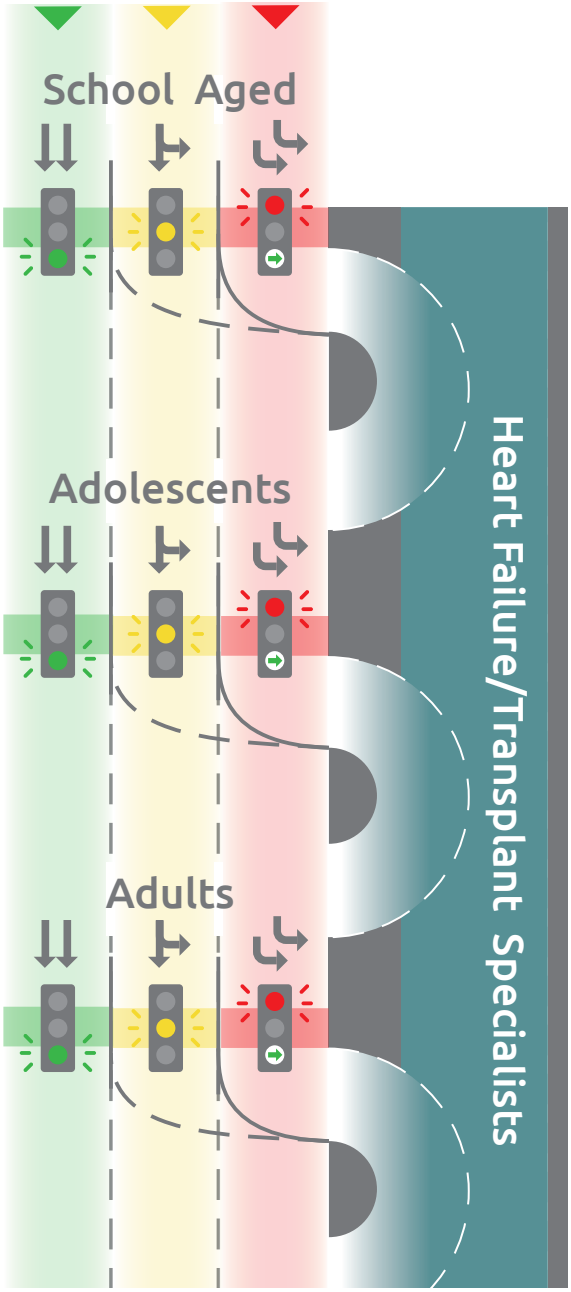
Fontan Surveillance:
When to Refer to Heart Failure Specialists

This roadmap provides recommendations on routine Fontan surveillance and when to refer for heart failure/transplant care. A referral does not necessarily mean a transfer of care, but often means a second set of eyes on the patient, with specific recommendations surrounding their management. In some cases, patients may require advanced therapies or heart transplantation. The goal of this roadmap is to decrease the percentage of patients who are referred too late to be eligible for advanced therapies.

To provide feedback on this tool, please scan the QR code.

Age Appropriate Surveillance



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- All individuals with SV heart disease should receive age-appropriate surveillance. ***See Page 2 for guidelines...***

Consider Referral for Heart Failure Care

- Moderate systolic dysfunction, or mild systolic dysfunction with moderate AVVR
- Mildly abnormal hemodynamics (ie, CI<2.5, EDP >12 mmHg, Fontan pressure >15 mmHg), with symptoms
- Subjective decrease in exercise tolerance or decline in NYHA class
- Progressive or symptomatic cyanosis
- New admission for arrhythmia, fluid overload, pleural effusion, ascites, PLE, or plastic bronchitis
- Evidence of end-organ dysfunction
  - Mild CKD (eGFR 60–89)
  - Progression of liver fibrosis with evidence of portal hypertension (varices, hemorrhoids, ascites, thrombocytopenia)

Stop & Refer for Advanced Therapies Evaluation[1]

- Severe systolic dysfunction (ie, LVEF <35% or RVEF <30%)
- Moderate systolic dysfunction with moderate AVVR
- Severe ventricular dilation (ie, EDV > 156 ml/m2)
- Severely abnormal hemodynamics (ie, CI <2, EDP >15, Fontan >20), with or without symptoms
- Sequential decrease in exercise tolerance (as measured by peak VO2 on CPET)
- Growth failure (ie, height for age percentile crossing 2 curves)
- Significant arrhythmia (ie, recurrent arrhythmias despite medical therapy, ICD need, aborted sudden cardiac death event)
- Chronic, symptomatic fluid overload despite medical therapies
- Recurrent/refractory pleural effusions or ascites
- Recurrent/refractory PLE or plastic bronchitis
- Significant end-organ dysfunction
  - Moderate CKD (eGFR<60)
  - Hepatic synthetic dysfunction
  - Hemoptysis

[1] *Lubert, A. M., Cedars, A., Almond, C. S., Amdani, S., Conway, J., Friedland-Little, J. M., Gajarski, R. J., Kindel, S. J., Lorts, A., Morales, D. L. S., O'Connor, M. J., Peng, D. M., Rosenthal, D. N., Smyth, L., Sutcliffe, D. L., & Schumacher, K. R. (2023). Considerations for Advanced Heart Failure Consultation in Individuals With Fontan Circulation: Recommendations From ACTION. Circulation. Heart failure, 16(2), e010123. https://doi.org/10.1161/CIRCHEARTFAILURE.122.010123*

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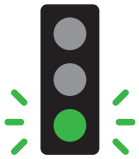

Suggested Tests for **Routine Surveillance**[2,3]

Additional studies may be clinically indicated

	Children (<12 years)	Adolescent (12–17 years)	Adults (>18 years)
Cardiology Clinic Visit (in-person)	Every 6–12 months	Every 6–12 months	Every 6–12 months
ECG	Every 6–12 months	Every 6–12 months	Every 6–12 months
Echo	Every 12 months	Every 12 months	Every 12 months
24-hr Rhythm Monitoring	Every 2–3 years	Every 1–2 years	Every 1–2 years
Exercise Test	6 min walk every 2–3 years	CPET every 1–3 years	CPET every 1–2 years
Cardiac MRI	Every 3 years	Every 2–3 years	Every 2–3 years
Cardiac Cath	As clinically indicated	Every 10 years	Every 10 years
Labs*	Every 3–4 years	Every 1–3 years	Every 1–2 years
Abdo U/S +/– Elastography	Every 3–4 years	Every 1–3 years	Every 1–2 years
Liver MRI with Elastography	N/A	Every 1–3 years	Every 1–2 years
Neurodevelopmental Evaluation	Prior to school entry	At middle school or high school entry	Once if never performed
Mental health screening	Yearly	Yearly	Yearly

** Basic labs include: CBC w/ differential, complete metabolic panel, GGT, cystatin C, vitamin D*
Advanced labs include: INR/PTT, alpha-fetoprotein, serum cholesterol, urinalysis, serum IgG, fecal alpha-1 antitrypsin level, PTH, serum iron/TIBC/Ferritin

[2] *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 Aug 6;140(6):e234-e284.*

[3] *Marino BS, Lipkin PH, Newburger JW, Peacock G, Gerdes M, Gaynor JW, Mussatto KA, Uzark K, Goldberg CS, Johnson WH Jr, Li J, Smith SE, Bellinger DC, Mahle WT; American Heart Association Congenital Heart Defects Committee, Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, and Stroke Council. Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management: a scientific statement from the American Heart Association. Circulation. 2012 Aug 28;126(9):1143-72.*