MANAGEMENT OF THE FAILING FONTAN CIRCULATION

Prepared by **Dr. Sidhartha Mullapudi**, DrNB (Cardiology), MD (General Medicine)
Last reviewed: October 2026

I THE FONTAN PHYSIOLOGY AND THE "FAILING FONTAN" DEFINITION

- **The Fontan Circulatory Paradox:** The Fontan operation (Total Cavopulmonary Connection [TCPC]) routes systemic venous return from the SVC and IVC directly into the pulmonary arterial tree without a subpulmonary ventricular pump. Consequently, the circulation operates under a fundamental physiological paradox: **chronic upstream systemic venous hypertension (CVP 12 to 20 mmHg)** required to drive passive transpulmonary flow, coupled with **chronic downstream systemic ventricular preload deprivation and low cardiac output**.
- **Definition of the "Failing Fontan":** A multi-organ clinical syndrome of circulatory attrition characterized by progressive decline in functional capacity, refractory systemic venous congestion (ascites, edema), worsening cyanosis, arrhythmias, or lymphatic/end-organ failure (**Protein-Losing Enteropathy, Plastic Bronchitis, or Fontan-Associated Liver Disease**)—which frequently occurs **even when single-ventricle systolic ejection fraction is completely preserved**.

II FOUR DISTINCT HEMODYNAMIC PHENOTYPES OF FONTAN FAILURE (ADAPTED FROM BOOK ET AL.)

- **Phenotype I — Reduced Systolic Function (Failing Single Ventricle with Low EF):** Classic systolic pump failure, most common when the systemic single ventricle is a **morphological Right Ventricle (e.g., Hypoplastic Left Heart Syndrome)** subjected to systemic afterload and chronic **systemic atrioventricular (tricuspid) valve regurgitation**.
- **Phenotype II — Preserved Systolic Function with Elevated Ventricular End-Diastolic Pressure (Diastolic / Restrictive Failure):** Progressive myocardial hypertrophy, interstitial fibrosis, and loss of ventricular compliance raise **systemic ventricular end-diastolic pressure (VEDP greater than 12 to 15 mmHg)**, which backs up across the lungs to mandate dangerously high Fontan venous pressures.
- **Phenotype III — Normal Heart with Elevated Pulmonary Vascular Resistance or Pathway Obstruction ("Pulmonary / Conduit Failure"):** Preserved ventricular systolic and diastolic function, but passive flow is blocked by **anatomical stenosis** in the Fontan conduit/baffle or branch pulmonary arteries, pulmonary vein compression, or **maladaptive pulmonary vascular remodeling (elevated PVR)** from non-pulsatile flow.
- **Phenotype IV — Abnormal Lymphatics with Near-Normal Hemodynamics ("Lymphatic Failure"):** Decompression of pressurized thoracic duct and hepatoduodenal lymphatics into low-resistance mucosal lumen spaces, manifesting as **Protein-Losing Enteropathy (PLE)** or **Plastic Bronchitis (PB)**.

Failing Fontan Phenotype	CVP / Fontan Pressure	Ventricular EDP (PAWP)	Cardiac Output	Primary Pathophysiological Driver
Type I: Systolic Failure	Elevated (>15 mmHg)	Elevated (>12 mmHg)	**Low**	Morphological RV systolic failure; severe AV valve regurgitation
Type II: Diastolic Failure	Elevated (>15 mmHg)	**Markedly Elevated (>15 mmHg)**	Low / Normal	Stiff, fibrotic non-compliant single ventricle
Type III: Fontan / PA Impedance	**Markedly Elevated (>18 mmHg)**	Normal (<10 mmHg)	**Low**	Conduit/PA stenosis; elevated **PVR** (wide transpulmonary gradient >6–8 mmHg)
Type IV: Lymphatic Failure	Mild-Moderately Elevated	Normal / Mildly Elevated	Normal / High	Thoracic duct hypertension -> **PLE** (enteric leak) or **Plastic Bronchitis** (airway leak)

III DIAGNOSTIC WORKUP (ESC 2020 ACHD / AHA 2019 GUIDELINES)

- **Invasive Cardiac Catheterization (Class I Mandatory at Low Threshold):** Any Fontan patient presenting with unexplained decline, edema, ascites, PLE, plastic bronchitis, worsening cyanosis, or arrhythmia requires complete right and left heart catheterization:
- **Diagnostic Hemodynamic Cut-offs:** A **mean Fontan/cavopulmonary pressure of 15 mmHg or greater** (and especially **greater than 18 to 20 mmHg**) or a **Transpulmonary Gradient (TPG = mean PA pressure minus LA/wedge pressure) greater than 6 to 8 mmHg** confirms a failing circuit.
- **Zero-Tolerance for Anatomical Gradients:** In a passive non-pulsatile circuit, **even a 1 to 2 mmHg pressure gradient** across the Fontan conduit, Glenn anastomosis, or branch pulmonary arteries represents a severe hemodynamically significant obstruction that mandates percutaneous stenting.
- **Collateral Mapping:** Identifies **venovenous collaterals** (from SVC/innominate/IVC to pulmonary veins/LA, causing severe cyanosis) and **aortopulmonary collaterals (APCs)** (causing competitive pulmonary flow and single-ventricle volume overload).
- **Cross-Sectional Imaging (CMR and MDCTA): CMR** quantifies single-ventricle volumes, ejection fraction, AV valve regurgitant fraction, collateral flow fraction, and **T2-weighted Dynamic Contrast MR Lymphangiography** (mapping thoracic duct leak pathways); **MDCTA** evaluates stents, conduit thrombus, and pulmonary embolism.
- **End-Organ Biomarkers and Liver Surveillance:** Serum **Albumin, Total Protein, IgG, lymphocyte count, and Fecal Alpha-1 Antitrypsin clearance** (for PLE); **NT-proBNP**; and annual **FALD surveillance (LFTs, GGT, Platelet count, Alpha-Fetoprotein [AFP], and Liver Ultrasound / Transient Elastography [FibroScan])**.

IV MECHANISM-SPECIFIC MANAGEMENT STRATEGIES (ESC 2020 / AHA 2019)

A Hemodynamic and Pharmacological Optimization

- **Judicious Diuretic Therapy: Loop diuretics (Furosemide/Torsemide)** combined with **Mineralocorticoid Receptor Antagonists (Spironolactone 25 to 100 mg/day)** are the cornerstone for controlling ascites and peripheral edema; however, **over-diuresis must be avoided** because excessive reduction in central venous pressure abolishes the passive transpulmonary gradient and precipitates acute low cardiac output and prerenal azotemia.
- **Pulmonary Vasodilators (Lowering the "Critical Bottleneck" PVR — Class IIb in ESC 2020):** Because pulmonary vascular resistance is the primary determinant of single-ventricle preload in a Fontan circuit, **PDE-5 Inhibitors (Sildenafil, Tadalafil, or Udenafil [studied in the Phase III FUEL Trial, 2020])** and **Endothelin Receptor Antagonists (Bosentan [TEMPO Trial] or Macitentan)** are used to lower PVR and improve submaximal exercise capacity in patients with elevated Fontan pressures and low PAWP.
- **Systemic Ventricular GDMT:** While empirical ACE inhibitors showed no benefit in single-ventricle and preserved-EF Fontan cohorts (e.g., the Pediatric Heart Network infant single-ventricle enalapril trial), **ACEi/ARBs/ARNI, Beta-blockers, MRAs, and SGLT2 Inhibitors (Empagliflozin/Dapagliflozin)** are utilized in **Phenotype I (systolic dysfunction)**.

B Targeted Management of Lymphatic Failure (PLE and Plastic Bronchitis)

- **Medical Therapy for Protein-Losing Enteropathy (PLE):**
  - **Dietary Modification: High-protein (greater than 2 g/kg/day), low-long-chain-fat, high-Medium-Chain Triglyceride (MCT) diet** (because MCTs are absorbed directly into the **portal venous bloodstream** rather than intestinal lacteal lymphatics, bypassing and decompressing engorged gut lymphatics).
  - **Targeted Anti-Inflammatory and Endothelial Therapy: Oral Controlled-Ileal-Release Budesonide (3 to 9 mg/day)** suppresses enteric mucosal inflammation and restores tight-junction integrity (requires monitoring for systemic cushingoid effects because hepatic congestion impairs first-pass steroid metabolism); **High-Dose Spironolactone**; **Subcutaneous Unfractionated Heparin** (which restores enterocyte membrane **heparan sulfate proteoglycans**); **Octreotide** (reduces splanchnic blood and lymphatic flow); and intravenous **Albumin + Furosemide** infusions with IVIG replacement for severe hypogammaglobulinemia.
  - **Acute and Chronic Management of Plastic Bronchitis (PB):**
  - **Airway Cast Clearance:** Nebulized **Recombinant Tissue Plasminogen Activator (tPA / Alteplase)** (which lyses the fibrin matrix of bronchial casts), nebulized heparin, **dornase alfa**, hypertonic saline, vest physiotherapy, and urgent **rigid/flexible bronchoscopic cast extraction** for airway obstruction.
  - **Interventional Lymphatic Decompression (Game-Changer for PLE & PB):**
  - **Percutaneous Lymphatic Embolization:** Using intranodal/intrahepatic lymphangiography and thoracic duct cannulation to selectively embolize abnormal **peribronchial lymphatic leaks (in Plastic Bronchitis, >85% resolution)** or **hepatoduodenal/periduodenal lymphatic fistulae (in PLE)** using lipiodol, cyanoacrylate glue, and microcoils.
  - **Innominate Vein Turn-Down (Hraska Procedure):** Surgically disconnecting the thoracic duct-draining left innominate vein from the high-pressure SVC/Glenn and anastomosing it directly to the low-pressure **pulmonary venous atrium**, decompressing the thoracic duct from 18 mmHg down to 6 mmHg.

C Catheter and Surgical Structural Rescue

- **Relief of Pathway Obstruction (Class I):** Balloon angioplasty and **endovascular stenting** of any Fontan conduit, baffle, or branch pulmonary artery stenosis, and **coil embolization of large aortopulmonary collaterals**.
- **Creation or Enlargement of a Fontan Fenestration (Class IIa/IIb):** Transcatheter needle puncture and stenting to create a **4 to 6 mm fenestration** between the Fontan conduit and the systemic atrium decompresses CVP (improving PLE/PB) and increases cardiac output at the cost of mild cyanosis. (Conversely, if severe cyanosis is driven by large **venovenous collaterals**, transcatheter coil/plug occlusion restores saturation provided baseline Fontan pressure is not prohibitively high.)
- **Arrhythmia Control and Surgical Fontan Conversion:** Immediate **DC cardioversion** and **3D catheter ablation** for Intra-Atrial Reentrant Tachycardia (IART). Patients with an obsolete dilated **Atriopulmonary Fontan**, refractory IART, and preserved ventricular function undergo **Surgical Fontan Conversion to an Extracardiac TCPC + Right Atrial Reduction + bi-atrial Cryo-Maze + epicardial dual-chamber pacemaker**.

V DEFINITIVE THERAPY: MECHANICAL SUPPORT AND TRANSPLANTATION

- **Indications for Heart (or Combined Heart-Liver) Transplantation:** Refractory **PLE or Plastic Bronchitis** (which resolves completely in over 90 percent of patients once normal low-CVP biventricular physiology is restored by transplant), end-stage systemic ventricular failure, or uncorrectable Fontan hemodynamic failure.
- **Unique Transplant Challenges:** High surgical bleeding risk from **multiple redo sternotomies and high-pressure collateral vessels**, need for extensive **pulmonary artery and vena caval reconstruction**, **high HLA allosensitization** (from prior homograft patches and transfusions), and determining when advanced **FALD cirrhosis** mandates **en-bloc Combined Heart-Liver Transplantation (CHLT)** rather than isolated heart transplantation.

VI PRACTICAL CONSIDERATIONS IN THE INDIAN CLINICAL CONTEXT

- **Mandatory Annual FALD FibroScan and AFP Screening:** Because many Indian Fontan survivors were operated at an older childhood age (resulting in longer cumulative pre- and post-Fontan hepatic injury), annual **Transient Elastography (FibroScan)**, liver ultrasound, and **serum Alpha-Fetoprotein (AFP)** are essential to detect early cardiac cirrhosis and hepatocellular carcinoma.
- **Maximizing Native Fontan Survival:** Given the limited availability and high cost of ACHD Heart and Heart-Liver Transplantation in India, aggressive medical and transcatheter palliation—combining **Sildenafil + Spironolactone + MCT diet + oral Budesonide**, **stenting of minor branch PA stenoses**, and **transcatheter fenestration creation**—is the frontline strategy.
- **Nutritional Rehabilitation and Contraceptive Counseling:** In Indian children and young adults with PLE, correcting severe protein-calorie malnutrition, fat-soluble vitamin deficiencies (A, D, E, K), and hypocalcemia is critical, alongside proactive counseling against high-risk pregnancy (**mWHO Class IV** in any failing Fontan).

KEY REFERENCES

- **Baumgartner H, De Backer J, Babu-Narayan SV, et al.:** 2020 ESC Guidelines for the management of adult congenital heart disease, *European Heart Journal*, 2020.
- **Rychik J, Atz AM, Celermajer DS, et al.:** Evaluation and Management of the Child and Adult With Fontan Circulation: A Scientific Statement From the American Heart Association, *Circulation*, 2019.
- **Book WM, Gerardin JF, Saraf AP, Marie Valente A, Rodriguez FH III:** Clinical Phenotypes of Fontan Failure: Implications for Management, *Congenital Heart Disease*, 2016.
- **Goldberg DJ, Zak V, Goldstein BH, et al. (FUEL Investigators):** Results of the Fontan Udenafil Exercise Longitudinal (FUEL) Trial, *Circulation*, 2020.