FONTAN SURGERY: INDICATIONS AND COMPLICATIONS

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I INTRODUCTION AND HEMODYNAMIC PHYSIOLOGY

- **The Core Principle:** The Fontan procedure (first performed by Francis Fontan in 1968 for tricuspid atresia) is the definitive **staged palliative operation** for hearts with a **single functional ventricle** where biventricular repair is anatomically impossible. It places the systemic and pulmonary circulations **in series** driven by a single ventricular chamber: all systemic venous return from the **Superior Vena Cava (SVC)** and **Inferior Vena Cava (IVC)** is routed **directly into the pulmonary arteries (Total Cavopulmonary Connection [TCPC])**, completely bypassing a subpulmonary ventricle.
- **Hemodynamic Consequences:** Separating the two circulations eliminates intracardiac mixing (resolving chronic arterial hypoxemia) and unloads the volume-overloaded single ventricle. However, because there is **no subpulmonary ventricular pump**, pulmonary blood flow is **entirely passive and non-pulsatile**, dependent on a **transpulmonary pressure gradient** between the **Central Venous Pressure (CVP / Fontan pressure, typically 10 to 15 mmHg)** and the **Left/Systemic Atrial Pressure (typically 5 to 8 mmHg)**.
- **Evolution of Surgical Modifications:**
  - **Classical Atriopulmonary Connection (APC — Obsolete):** Anastomosed the right atrium directly to the pulmonary artery; resulted in massive right atrial dilation, turbulent energy loss, mural thrombosis, and refractory atrial arrhythmias.
  - **Lateral Tunnel (LT) TCPC:** Intra-atrial prosthetic baffle directing IVC blood to the PA along the lateral RA wall (grows with the child, but places suture lines near the sinus node).
  - **Extracardiac Conduit (ECC) TCPC (Current Standard):** Interposes an **18 to 20 mm polytetrafluoroethylene (Gore-Tex) tube graft** outside the heart from the transected IVC to the undersurface of the right pulmonary artery, optimizing laminar hydrodynamic flow and keeping the entire atrium at low pressure free of suture lines.

II INDICATIONS AND SELECTION CRITERIA

- **Anatomical Indications (Functional Single Ventricle):**

Tricuspid Atresia (classic right-heart hypoplasia).

Hypoplastic Left Heart Syndrome (HLHS) (following Stage 1 Norwood and Stage 2 Bidirectional Glenn / Hemi-Fontan).

Double Inlet Left Ventricle (DILV) or Double Inlet Right Ventricle.

Unbalanced Atrioventricular Septal Defect (AVSD).

Pulmonary Atresia with Intact Ventricular Septum (PA-IVS) with severe bipartite/unipartite RV hypoplasia or RV-dependent coronary circulation.

Complex Double Outlet Right Ventricle (DORV) with non-routable VSD or straddling AV valves.

- **Choussat's "Ten Commandments" (1978) and Modern Selection Criteria:** While several original Choussat criteria (such as age 4 to 15 years, sinus rhythm, and normal RA volume) have been modified in the staged TCPC era (Fontan completion is now routinely performed at **2 to 4 years of age** or weight **10 to 15 kg**), three hemodynamic prerequisites remain mandatory for a successful Fontan:

Low Pulmonary Vascular Resistance (PVR less than 2 to 3 Wood Units·m2) and **Mean Pulmonary Artery Pressure (mPAP) less than 15 mmHg**.

Unobstructed, Well-Developed Pulmonary Arteries (Nakata Index greater than 200 to 250 mm2/m2; McGoon ratio greater than 1.8).

Preserved Systemic Ventricular Function (end-diastolic pressure less than 12 mmHg) without moderate-to-severe systemic atrioventricular valve regurgitation or subaortic/aortic arch obstruction.

III EARLY POST-OPERATIVE COMPLICATIONS

- **Acute Fontan Failure (Low Cardiac Output State):** Triggered by elevated PVR, pulmonary artery distortion, ventricular diastolic dysfunction, or positive-pressure mechanical ventilation (which impedes passive systemic venous return); manifests as high CVP (greater than 20 mmHg), systemic hypotension, lactic acidosis, and oliguria, requiring **early extubation** (spontaneous negative-pressure inspiration acts as a thoracic pump drawing blood into the lungs), **inhaled Nitric Oxide**, **Milrinone**, and **fenestration creation or takedown to Bidirectional Glenn**.
- **Persistent Pleural Effusions and Chylothorax:** The most common early morbidity (occurring in 20 to 40 percent of patients) due to acute elevation of superior/inferior vena caval pressure exceeding the lymphatic drainage capacity of the **thoracic duct**.
- **Acute Conduit / Pulmonary Artery Thrombosis:** Sluggish, non-pulsatile venous flow combined with post-bypass acquired protein C/S and antithrombin III deficiencies predisposes to circuit thrombosis or paradoxical systemic stroke across the fenestration.
- **Early Arrhythmias:** Sinus node dysfunction or **Junctional Ectopic Tachycardia (JET)**, which causes loss of AV synchrony and an immediate drop in ventricular preload and cardiac output.

IV LATE COMPLICATIONS (THE "FONTAN PARADOX")

- **The Hemodynamic Paradox:** The Fontan circulation subjects the patient to **chronic systemic venous hypertension (CVP 12 to 20 mmHg)** upstream of the lungs and **chronic preload deprivation with low cardiac output** downstream in the systemic ventricle.
- **Fontan-Associated Liver Disease (FALD):** Universal time-dependent consequence of chronic non-pulsatile hepatic venous congestion and central perisinusoidal hypoxia. Progresses from congestive hepatopathy to bridging sinusoidal fibrosis, **cardiac cirrhosis**, hypervascular regenerative nodules, and **Hepatocellular Carcinoma (HCC)** (which can develop 15 to 25 years post-Fontan). **Surveillance Mandate (Class I):** Annual liver biochemical profile, **alpha-fetoprotein (AFP)**, and **abdominal ultrasound / transient elastography (FibroScan)**, with liver MRI for nodules.
- **Protein-Losing Enteropathy (PLE):** Occurs in **5 to 12 percent** of Fontan patients (5-year mortality historically 20 to 50 percent). Chronic mesenteric venous hypertension, low cardiac output, and intestinal **lymphangiectasia** cause enteric loss of albumin, immunoglobulins (IgG), and lymphocytes (CD4+ T-cells). Confirmed by **elevated fecal alpha-1 antitrypsin clearance** with hypoalbuminemia, anasarca, ascites, chronic diarrhea, and hypocalcemia/osteopenia.
- **Plastic Bronchitis (PB):** Rare (1 to 4 percent), life-threatening lymphatic complication in which elevated thoracic duct pressure causes decompression of proteinaceous/fibrinous chyle through fistulous peribronchial lymphatics into the bronchial lumen, forming **large rubbery, branching airway casts** that cause expectoration of casts, atelectasis, and **acute asphyxiation**.

- **Atrial Tachyarrhythmias: Intra-Atrial Reentrant Tachycardia (IART)** and **Atrial Flutter/Fibrillation** occur in over 50 percent of classical APC patients at 20 years (vs. less than 10 to 15 percent with modern ECC). Any sustained atrial arrhythmia in a Fontan patient is a **medical emergency**, as loss of atrial kick rapidly raises LA pressure, abolishes the transpulmonary gradient, and precipitates hemodynamic collapse and atrial thrombus.
- **Systemic Ventricular Failure:** Progressive diastolic stiffening and systolic failure, most rapid when the systemic single ventricle is a **morphological Right Ventricle (e.g., HLHS)** with systemic tricuspid regurgitation.

Late Complication	Primary Pathophysiological Mechanism	Diagnostic Hallmark & Targeted Management
Fontan-Associated Liver Disease (FALD)	Chronic passive hepatic venous hypertension + low cardiac output	Elevated GGT, FibroScan stiffness, hypervascular nodules; annual **US + AFP + MRI**; combined **Heart-Liver Transplant** if cirrhotic/HCC
Protein-Losing Enteropathy (PLE)	Mesenteric venous HTN + enteric lymphatic rupture + mucosal inflammation	Low serum albumin + **elevated fecal alpha-1 antitrypsin**; high-protein/MCT diet, **oral controlled-release Budesonide**, Sildenafil, Spironolactone, SC unfractionated heparin, lymphatic embolization
Plastic Bronchitis (PB)	Thoracic duct hypertension leaking fibrinous lymph into bronchial tree	Expectoration of branching **bronchial casts**; nebulized **tPA / dornase alfa**, sildenafil, **percutaneous thoracic duct / selective lymphatic embolization**
IART / Atrial Flutter	Atrial stretch + surgical atriotomy scars (especially classical APC)	Prompt **DC cardioversion**, **3D catheter ablation**, or surgical **Fontan Conversion to TCPC + Cryo-Maze**

V FENESTRATION, ANTICOAGULATION AND MODERN TRIAL EVIDENCE

- **Role of the Conduit Fenestration:** Creating a **4 mm communication** between the extracardiac conduit/lateral tunnel and the systemic atrium provides a right-to-left **decompression "pop-off"**, augmenting systemic ventricular preload and cardiac output while shortening post-operative pleural effusion duration, at the cost of mild arterial desaturation (SpO2 88 to 93 percent) and paradoxical embolism risk (the fenestration can later be closed percutaneously with an occlusion device if hemodynamics are favorable).
- **Thromboprophylaxis: Oral Anticoagulation (Warfarin, target INR 2.0 to 3.0; Class I)** is mandatory for any Fontan patient with prior thromboembolism, atrial arrhythmias, conduit stasis, or right-to-left shunt/fenestration; in uncomplicated Fontan patients, antiplatelet therapy (**Aspirin**) or anticoagulation is used (note: the randomized **UNIVERSE Trial** in children and observational adult registries support factor Xa DOACs in selected uncomplicated patients, though Warfarin remains standard in high-risk adult Fontans).
- **Pulmonary Vasodilator Trials (FUEL Trial):** The Phase III **FUEL Trial** (*Circulation* 2020) demonstrated that **Udenafil** (PDE-5 inhibitor) improved submaximal exercise performance at the ventilatory anaerobic threshold (though it missed the primary Peak VO2 endpoint), supporting PDE-5 inhibitors (**Sildenafil/Tadalafil**) and **Bosentan** (Class IIb in ESC 2020) in selected patients with elevated PVR/Fontan pressures.

VI PRACTICAL CONSIDERATIONS IN THE INDIAN CLINICAL CONTEXT

- **Late-Presenting Single-Ventricle Candidates:** In India, many single-ventricle children present beyond 5 to 10 years of age with longstanding cyanosis, abundant **aortopulmonary collaterals (APCs)**, and borderline elevated PVR. Pre-Fontan **cardiac catheterization with coil embolization of large APCs** and **routine surgical fenestration** are critical to prevent post-operative high-output pleural effusions and prolonged ICU stays.
- **Pragmatic Failing Fontan Management:** Because pediatric/ACHD heart and combined heart-liver transplantation have limited availability in India, aggressive medical optimization of **PLE** (high-protein **Medium-Chain Triglyceride [MCT]** diet, **oral enteric-coated Budesonide**, **Spironolactone**, and **Sildenafil**), transcatheter creation or dilation of a **fenestration**, and **percutaneous lymphatic embolization** are the primary strategies to prolong native Fontan survival.

KEY REFERENCES

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