FONTAN SURGERY: INDICATIONS, CURRENT GUIDELINES & COMPLICATIONS

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I THE FONTAN PHYSIOLOGY AND INDICATIONS

- **The Core Concept:** The Fontan operation is not an anatomical cure; it is the final staged surgical palliation for patients born with a **functionally univentricular heart** (where biventricular repair is precluded by hypoplasia of one ventricle or non-septatable atrioventricular/ventriculoarterial connections).
- **The Hemodynamic Physiology:** Separates the pulmonary and systemic circulations in series so that the single functional ventricle pumps oxygenated blood exclusively into the systemic aorta, while 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])** without a subpulmonary ventricular pump. Consequently, **pulmonary blood flow is entirely passive**, driven solely by the transpulmonary pressure gradient between **Central Venous Pressure (CVP, typically 10 to 15 mmHg)** and **Systemic Atrial / Ventricular End-Diastolic Pressure (typically 5 to 8 mmHg)**.
- **Primary Anatomical Indications:**

Tricuspid Atresia.

Hypoplastic Left Heart Syndrome (HLHS).

Double Inlet Left Ventricle (DILV).

Unbalanced Atrioventricular Septal Defects (AVSD).

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

Complex Double Outlet Right Ventricle (DORV) with remote/non-routable VSD.

II THE MODERN THREE-STAGE SURGICAL PATHWAY

- **Physiological Rationale for Staging:** A complete Fontan cannot be performed in a neonate because **neonatal Pulmonary Vascular Resistance (PVR) is physiologically high** (preventing passive cavopulmonary flow) and the single ventricle must gradually adapt from volume overload to normal preload.
- **Stage 1 (Neonatal Period — First Weeks of Life):** Optimizes balanced pulmonary and systemic blood flow while protecting the pulmonary vascular bed from overcirculation:
- **For Decreased Pulmonary Flow (e.g., Tricuspid Atresia with PS/PA): Modified Blalock-Thomas-Taussig (mBTT) Shunt** or transcatheter **PDA stenting**.
- **For Unobstructed High Pulmonary Flow (e.g., DILV or Tricuspid Atresia without PS): Pulmonary Artery (PA) Banding** to restrict pulmonary pressure and prevent PVOD.
- **For Systemic Outflow Obstruction (HLHS): Norwood Procedure** (reconstructing the neoaorta using the main PA + either a **Sano RV-to-PA conduit** or mBTT shunt + atrial septectomy).
- **Stage 2 (4 to 6 Months of Age): Bidirectional Glenn (BDG) Shunt** (superior cavopulmonary anastomosis) or **Hemi-Fontan**. Once PVR has fallen after infancy, the **SVC is anastomosed end-to-side to the right pulmonary artery** (and the Stage 1 shunt is taken down), routing upper-body venous return (approx. 50 percent of venous return in infants) passively to both lungs and **unloading the single ventricle**.
- **Stage 3 (2 to 4 Years of Age / Weight 10 to 15 kg): Fontan Completion**. The **IVC (and hepatic venous return)** is routed to the pulmonary artery via a **Lateral Tunnel** or **Extracardiac Conduit**, completing the Total Cavopulmonary Connection. (Routing hepatic venous blood containing **hepatic factor** to both lungs is also essential to prevent or reverse **pulmonary arteriovenous malformations [PAVMs]** that can develop after a Glenn shunt.)

III CURRENT SURGICAL TECHNIQUES AND GUIDELINE STANDARDS (ESC 2020 / AHA 2019)

- **The Extracardiac Conduit (ECC) vs. Lateral Tunnel (LT):** Both forms of **Total Cavopulmonary Connection (TCPC)** have completely replaced the classical **Atriopulmonary Connection (APC)**. The **Extracardiac Conduit** uses an **18 to 20 mm Gore-Tex (PTFE) tube** outside the heart connecting the IVC to the right PA; it can be performed without aortic cross-clamping, preserves optimal laminar flow, leaves the entire atrium at low pressure, and minimizes sinus node injury and late atrial arrhythmias.
- **The Conduit Fenestration ("Pop-Off Safety Valve"):** A calibrated **4 mm communication** created between the Fontan baffle/conduit and the pulmonary venous atrium. In the presence of post-operative PVR spikes, the fenestration allows controlled right-to-left shunting that **decompresses systemic venous pressure (reducing pleural effusions)** and **maintains systemic ventricular preload and cardiac output** at the expense of mild arterial desaturation (SpO2 88 to 92 percent).

Stage / Parameter	Timing & Procedure	Key Hemodynamic / Exam Target
Stage 1 (Neonatal)	**Norwood / mBTT Shunt / PA Band** (0–1 month)	Unobstructed systemic outflow + controlled low-pressure pulmonary flow
Stage 2 (Cavopulmonary)	**Bidirectional Glenn / Hemi-Fontan** (4–6 months)	SVC to RPA; unloads single ventricle volume load after neonatal PVR falls
Stage 3 (Completion)	**Extracardiac Conduit (18–20 mm) or Lateral Tunnel** (2–4 years)	Ideal pre-Fontan hemodynamics: **mPAP <15 mmHg, PVR <2–3 WU, VEDP <12 mmHg**
Failing Fontan Criteria	Late decompensation on RHC	**Fontan/CVP pressure >15–18 mmHg**, cardiac index <2.0 L/min/m2, PLE, PB, or cirrhosis

IV THE "FAILING FONTAN": MAJOR CARDIAC AND EXTRACARDIAC COMPLICATIONS

A Extracardiac Complications (Driven by Chronic Venous Hypertension [CVP 15 to 20 mmHg] & Lymphatic Congestion)

- **Fontan-Associated Liver Disease (FALD):** Universal time-dependent consequence of chronic non-pulsatile hepatic venous hypertension and low cardiac output, progressing from sinusoidal dilation to bridging fibrosis, **cardiac cirrhosis**, portal hypertension, and **Hepatocellular Carcinoma (HCC)**.
- **Protein-Losing Enteropathy (PLE):** Occurs in 5 to 12 percent of survivors. Elevated mesenteric venous pressure and intestinal lymphangiectasia cause severe enteric loss of albumin, immunoglobulins, calcium, and lymphocytes—presenting with **anasarca, ascites, chronic diarrhea, hypoalbuminemia, and elevated fecal alpha-1 antitrypsin**.
- **Plastic Bronchitis (PB):** Decompression of pressurized thoracic duct lymph into the bronchial tree produces **large rubbery, branching mucofibrinous airway casts**, presenting with chronic cough, wheezing, cast expectoration, and **life-threatening asphyxiation**.

B Cardiac and Vascular Complications

- **Progressive Systemic Ventricular Dysfunction:** Chronic preload deprivation, increased systemic arterial afterload, myocardial fibrosis, and atrioventricular valve regurgitation lead to systolic and diastolic failure (especially when a **morphological Right Ventricle** serves as the systemic chamber, as in HLHS).
- **Atrial Tachyarrhythmias: Intra-Atrial Reentrant Tachycardia (IART)** and **Atrial Fibrillation** occur in up to 50 percent of older APC Fontans. Loss of AV synchrony causes an immediate rise in atrial pressure and catastrophic drop in passive pulmonary flow; **sustained IART is a medical emergency requiring prompt cardioversion**.
- **Thromboembolism:** Sluggish, non-pulsatile cavopulmonary flow, dilated right atrium (in APC), prosthetic material, and hepatic coagulation factor abnormalities create a **10 to 20 percent lifetime risk of Fontan conduit/pulmonary artery thrombosis** or **paradoxical stroke** across a fenestration.

V LONG-TERM MANAGEMENT AND GUIDELINE MANDATES (ESC 2020 ACHD / AHA 2019)

- **Invasive Hemodynamic Evaluation (Class I): Cardiac catheterization** must be performed at a low threshold in any Fontan patient developing unexplained edema, exercise deterioration, cyanosis, new arrhythmia, hemoptysis, PLE, or plastic bronchitis to identify treatable **Fontan pathway/PA stenosis** (where even a **1 to 2 mmHg gradient** is hemodynamically significant and requires stenting), **aortopulmonary/venovenous collaterals** (requiring coil occlusion), or elevated PVR.
- **Antithrombotic Therapy: Lifelong Oral Anticoagulation (Warfarin, target INR 2.0 to 3.0; Class I)** is mandatory in patients with prior thromboembolism, atrial arrhythmias, right-to-left shunt/fenestration, or classical APC with atrial stasis; antiplatelet therapy or DOACs (studied in the **UNIVERSE Trial**) are used in low-risk uncomplicated ECC patients, while DOACs remain contraindicated in mechanical valves.
- **Mandatory FALD Surveillance (Class I):** Baseline and serial **every 1-to-2-year liver evaluation** using hepatic biochemical panels, **Alpha-Fetoprotein (AFP)**, **liver ultrasound / elastography (FibroScan)**, and hepatology co-management to screen for cirrhosis and HCC.
- **Surgical Fontan Conversion and Transplantation:** Patients with a failing classical **Atriopulmonary Fontan** and refractory atrial arrhythmias/thrombus with preserved ventricular function should undergo **Surgical Fontan Conversion to an Extracardiac TCPC with Right Atrial Reduction and Cryo-Maze Ablation** (plus epicardial dual-chamber pacemaker). Patients with end-stage ventricular failure, refractory PLE, or plastic bronchitis require **Heart Transplantation** (or **Combined Heart-Liver Transplantation** if advanced cirrhosis/HCC is present).
- **Pregnancy Risk Stratification (2018 ESC Pregnancy / 2020 ESC ACHD Guidelines):**
  - **Uncomplicated, Well-Functioning Fontan (mWHO Class III):** High risk (40 to 50 percent miscarriage rate, prematurity, postpartum hemorrhage, and atrial arrhythmias), requiring management in an expert multidisciplinary ACHD center.
  - **Complicated or Failing Fontan (mWHO Class IV — Pregnancy Strictly Contraindicated, Class III):** Pregnancy is **contraindicated** if the patient has **resting SpO2 less than 85 percent, depressed systemic ventricular function, moderate-to-severe AV valve regurgitation, refractory arrhythmias, or Protein-Losing Enteropathy / Plastic Bronchitis**.

KEY REFERENCES

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