THE FONTAN HEART (SINGLE VENTRICLE PALLIATION)

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I PART 1 — THE FONTAN HEART: DEFINITION & ANATOMICAL CONFIGURATION

- **Definition (ESC 2020 ACHD Guidelines):** The Fontan operation is the final definitive palliation for congenital single-ventricle physiology (e.g., Tricuspid Atresia, Hypoplastic Left Heart Syndrome). It completely bypasses the right heart by directly connecting systemic venous return to the pulmonary arteries.
- **Modern Configurations (Total Cavopulmonary Connection - TCPC):**
  - **Lateral Tunnel:** The SVC is anastomosed to the RPA (Glenn shunt), and an intra-atrial baffle directs IVC blood to the pulmonary arteries.
  - **Extracardiac Conduit (Current Gold Standard):** A Gore-Tex tube connects the IVC directly to the pulmonary artery, entirely bypassing the right atrium. *Advantage:* Drastically lowers the incidence of atrial arrhythmias and saves atrial kinetic energy.

II THE "FONTAN PARADOX" (UNIQUE HEMODYNAMICS)

- **The Paradox:** There is no sub-pulmonary ventricular pump. Pulmonary blood flow is completely passive, driven exclusively by a chronically elevated Central Venous Pressure (CVP, usually 12-15 mmHg) and actively hindered by Pulmonary Vascular Resistance (PVR).
- **The Result:** The patient lives in a state of chronic systemic venous congestion (high CVP) and chronic diminished cardiac output (decreased ventricular preload).

III SEQUELAE OF THE FAILING FONTAN

- **Natural History:** The Fontan circuit is a palliative time-bomb; it inherently fails over decades.
- **Fontan-Associated Liver Disease (FALD):** Universal in adult patients. Chronic hepatic congestion leads to cardiac cirrhosis and Hepatocellular Carcinoma (HCC). Routine hepatic surveillance (ultrasound/elastography and AFP) is recommended.
- **Protein-Losing Enteropathy (PLE):** Severe intestinal lymphatic congestion causes massive enteric protein loss (low albumin, severe edema). Highly lethal.
- **Plastic Bronchitis:** Lymphatic failure causing dense, cast-like secretions in the bronchial tree, leading to asphyxiation.
- **Arrhythmias:** Intra-atrial Re-entrant Tachycardia (IART) is the most common cause of sudden decompensation. Catheter ablation in an experienced ACHD centre is preferred.

IV PART 2 — MANAGEMENT OF A CYANOTIC ("TET") SPELL: DEFINITION & PATHOPHYSIOLOGY

- **Definition:** A life-threatening hypercyanotic paroxysm classic to Tetralogy of Fallot (TOF), peaking between 2 to 4 months of age.
- **The Mechanism:** Triggered by crying, feeding, or defecation. A sudden surge in catecholamines causes violent spasm of the Right Ventricular Outflow Tract (RVOT) infundibulum. This drops pulmonary blood flow to near zero and forces a massive right-to-left shunt of deoxygenated blood across the VSD into the aorta.
- **The Clinical Trap:** The classic TOF murmur *disappears* during a spell because flow across the stenotic RVOT has completely ceased.

V ACUTE PHARMACOLOGICAL & MECHANICAL MANAGEMENT (THE ALGORITHM)

- **Goal:** Strictly hemodynamic: increase Systemic Vascular Resistance (SVR) and decrease RVOT spasm.

A Mechanical Maneuvers (First-Line)

- **Knee-Chest Position (Infants) / Squatting (Older Children):** Kinking the femoral arteries massively and instantly increases SVR, forcing blood back across the VSD and into the pulmonary bed.

B Pharmacological Rescue

- **100% Oxygen:** Acts as a potent pulmonary vasodilator to reduce PVR.
- **Morphine Sulfate (0.1 mg/kg IV/SC):** The ultimate sedative. It depresses the hyperpneic respiratory drive, breaks the panic cycle, and blunts the catecholamine surge driving the RVOT spasm.
- **Intravenous Fluid Bolus (Isotonic Saline):** Rapidly increases RV preload to physically pry open and force blood across the spasming RVOT.
- **Beta-Blockers (IV Esmolol or Propranolol):** The specific antidote to the infundibular spasm. Reverses the catecholamine-driven hypercontractility.
- **Vasopressors (IV Phenylephrine):** A pure alpha-1 agonist. It massively spikes SVR without any beta-1 chronotropic effects (which would worsen the spasm). Reduces the right-to-left shunt.
- **Sodium Bicarbonate:** Administered to correct severe metabolic acidosis, because acidosis directly increases PVR and worsens the right-to-left shunt.

VI LANDMARK ADVANCES (ESC 2020 ACHD / AHA 2019)

- **The Fontan Anticoagulation Debate:**
  - *Current Guideline (ESC 2020):* Routine prophylactic anticoagulation in a well-functioning Fontan is not universally recommended (antiplatelet or anticoagulant therapy is individualized). However, if there is *any* history of atrial arrhythmia, thrombus, or a prior stroke, lifelong systemic anticoagulation (Warfarin preferred over DOACs due to lack of robust DOAC data) is a strict **Class I, LOE B** mandate.
- **Pulmonary Vasodilators in the Fontan:**
  - *The Advance:* Off-label use of Endothelin Receptor Antagonists (Bosentan) or PDE-5 inhibitors (Sildenafil) is increasingly used to actively lower PVR and effectively "suck" blood through the passive Fontan circuit, significantly improving exercise capacity.
- **Emergency TOF Palliation:**
  - If a Tet Spell is completely refractory to medical therapy, emergency surgical palliation (modified Blalock-Taussig-Thomas shunt) or urgent transcatheter RVOT stenting is mandated as a life-saving bridge to total repair.

VII EXAM TIPS & LEXICON

- **Catchwords to underline in your exam:**
- "Extracardiac conduit (Total Cavopulmonary Connection)"
- "Fontan Paradox (High CVP, Low Cardiac Output)"
- "Protein-Losing Enteropathy (PLE) & FALD"
- "Infundibular spasm (Tet Spell)"
- "Knee-chest position (Increases SVR)"
- "IV Phenylephrine (Pure Alpha-1 agonist to reverse shunt)"

VIII EXECUTION TIP

- **Draw the "Hemodynamic Seesaw of a Tet Spell" on your exam sheet:** Draw a fulcrum (seesaw).
- **Left Side (Down):** Write **SVR**. (Trigger: Crying/Vasodilation).
- **Right Side (Up):** Write **PVR / RVOT Spasm**.
- **Arrow:** Draw a massive arrow pointing from the Right Ventricle to the Aorta (Right-to-Left Shunt).
- **Therapy Box:** Draw an arrow pushing SVR *UP* (Knee-chest, Phenylephrine) and an arrow pushing RVOT Spasm *DOWN* (Beta-blockers, Morphine). This visual proves to the examiner that you are not just memorizing drugs, but actively manipulating the physics of congenital shunts.