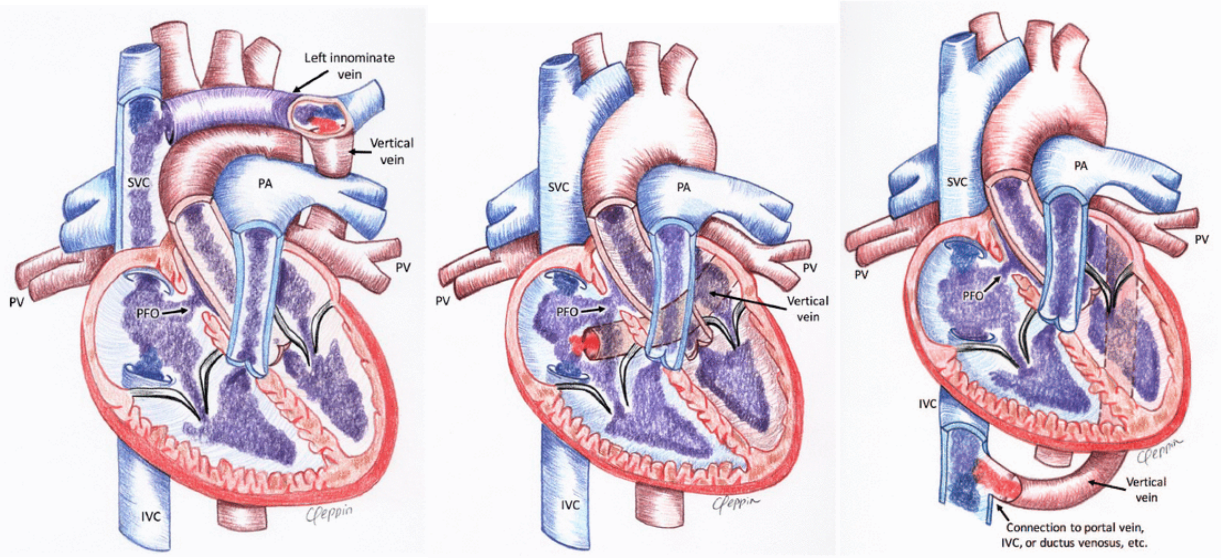
Total Anomalous Pulmonary Venous Return



1. Supracardiac (type 1, 45%): pulmonary vein confluence drains into the innominate vein/SVC via a vertical vein

2. Intracardiac (type 2, 25%): pulmonary vein confluence drains into the coronary sinus, sometimes directly into the right atrium

3. Infracardiac (type 3, 25%): pulmonary vein confluence drains into the portal vein, hepatic vein, or IVC via a vertical pulmonary vein passing through the esophageal hiatus (prone to obstruction)

4. Mixed (type 4, 5%): pulmonary venous drainage is some combination of the prior types

Pathophysiology:

- **Large L → R Shunt**
    - All pulmonary venous return is delivered to the RA
    - Pulmonary edema rapidly develops
- **ASD or PFO**
    - Essential for survival: only source of blood flow to the left atrium
- Pulmonary veins are often stenotic or obstructed → leads to severe pulmonary hypertension → supra-systemic RV/PA pressure → underfilled and flattened LV →shock. Quickly develops into a surgical emergency with cyanosis, severe pulmonary edema, and acidosis

Surgery:

- CPB with hypothermic circulatory arrest
- **Goals:** eliminate the anomalous connections, anastomose the pulmonary vein confluence directly to the left atrium, close intracardiac shunts
    - RA is opened to close the PFO or ASD

Anesthesia:

- **Goals:** Maintain age-appropriate HR (NSR), contractility, and preload to maintain CO—decreased CO will lead to decreased systemic venous saturation. With complete mixing lesions, arterial saturation will be reduced. TAPVR with pulmonary venous obstruction is a *surgical* emergency: attempts to increase pulmonary blood flow by decreasing PVR will worsen pulmonary edema.

  Drips

  - Dopamine, milrinone, epi, NTG, TXA

- Induction
  - Patients with obstructive pathology rarely tolerate any inhaled anesthetics
    - Induce with fentanyl and rocuronium, +/- ketamine
  - Expect highly reactive pulmonary vasculature, particularly in those with venous obstruction
    - Sufficiently blunt the stress response of intubation and surgical stimulation
  - Maintain HR, contractility, and preload to maintain CO
    - Support the RV function with inotropes (dopamine) as necessary

- Prior to Bypass
  - Avoid Nitric Oxide, as it worsens pulmonary edema in obstructive lesions
  - TEE should be done cautiously, as the mid esophageal position is directly behind the pulmonary venous confluence and can cause life-threatening pulmonary venous obstruction (remove if this occurs)

- Maintenance
  - Unstable neonates: high dose opioids (fentanyl 50mcg/kg) + NMBD
  - Stable infants: may consider volatile anesthetics

- Post-repair
  - Maintain age appropriate HR (NSR) as CO typically HR-dependent post-CPB
  - Diagnose and treat unresolved **Pulmonary HTN** (may require PAP, PAOP, and LA pressures to determine the etiology of post-repair pulmonary HTN)
    - Residual postoperative pulmonary venous obstruction may need to be surgically revised; attempts to decrease PVR in this setting will lead to pulmonary edema
    - Pulmonary arterial hypertension is treated by decreasing PVR and supporting RV: iNO, opioids, inotropes (dopamine, milrinone), hyperventilation, high FiO2
    - LV HTN may develop in the setting of adequate CO and a non-restrictive pulmonary anastomosis due to presence of small, non-compliant LA/LV or LV dysfunction