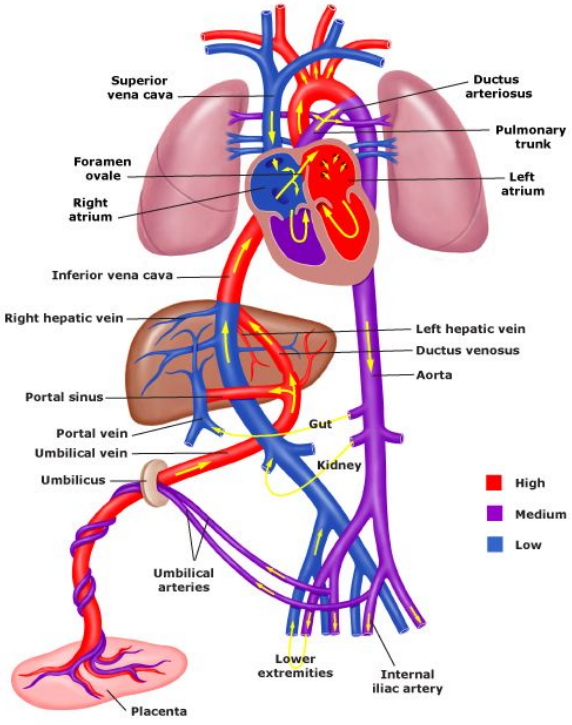
Congenital Pediatric Heart Disease

Basic Pathophysiology

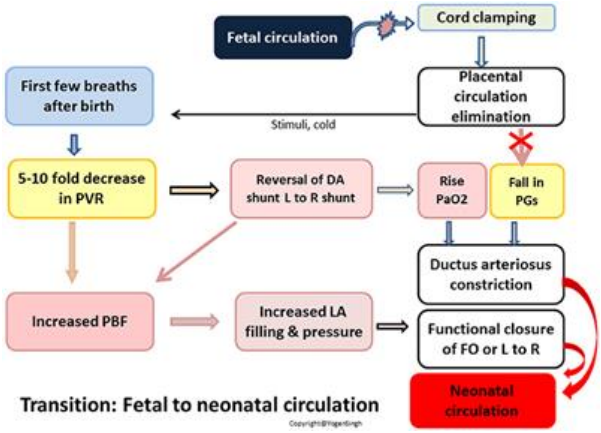
Fetal Circulation

- Have shunts to bypass liquid filled lungs
- Oxygenated blood from mom goes through placenta into baby via umbilical vein
    - Half the blood goes to liver
    - Half goes to ductus venosus to inferior vena cava where it mixes with deoxygenated blood returning from lower half of body
        - IVC and SVC (this is deoxygenated blood returning from upper half of body) meet at right atrium
            - Blood then goes to either:
                - LA via foramen ovale to LV to aorta
                - RV which then goes to pulmonary artery where most is shunted away from lungs via ductus arteriosus into aorta
                - Small amount goes to lungs to provide oxygen and nutrients for growth



Transitional Circulation

- When baby is born, lungs expand and fill with air with gradual reabsorption of fetal lung fluid – this increases PaO2 of blood flowing in the lungs which in turn mediates transition from fetal to neonatal lung patterns
- Flow through umbilical arteries stops and venous flow slows and stops eventually too
- Pulmonary vascular resistance falls and pulmonary blood flow increases – this continues for the first 30-45 days of life
- Ductus venosus and arteriosus close, this helps to continue decrease pulmonary vascular resistance, increases systemic resistance
- LA pressures increase, resulting in closure of foramen ovale



Neonatal Cardiac Physiology

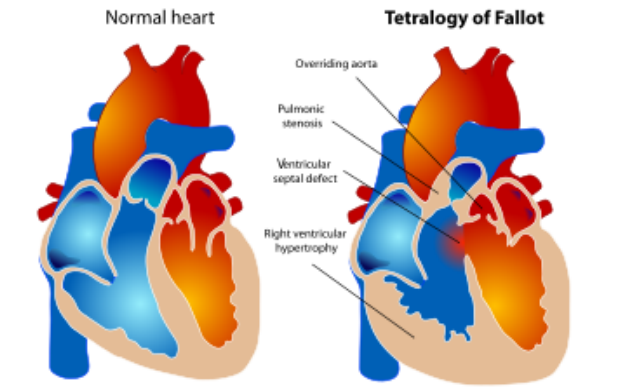
- They have non-compliant ventricular walls, so they can't increase SV, rely on increasing HR to increase CO – so sinus tachycardia is first response to stress (**DO NOT EVER BLOW OFF TACHYCARDIA!!!**)
    - Remember CO = SV x HR
    - Due to this, they are more susceptible to CHF as there is but so much one can increase HR before it is not effective
- Ductus arteriosus closes within first 15 hours of life
- Foramen ovale closes within first 3 months of age

Basics of Congenital Heart Disease

- Congenital heart defects can present at any age in a spectrum from cyanosis, cardiovascular collapse to CHF
- Occur in 1 in 1000 births
- Cyanotic vs Acyanotic
    - **Cyanotic – Terrible Ts**
        - Tetralogy of Fallot
        - Tricuspid anomalies – tricuspid atresia and Ebstein's anomaly
        - Truncus arteriosus
        - Total anomalous pulmonary venous return
        - Transposition of great arteries
    - **Acyanotic**
        - VSD – ventricular septal defect
        - ASD – atrial septal defect
        - PDA – patent ductus arteriosus
        - Atrioventricular canal
        - Coarctation
        - Pulmonary stenosis
        - Aortic stenosis
- Cyanosis
    - In order for cyanosis to be present, 3-5 mg/dL has to be deoxyhemoglobin – this corresponds to 70-80% O2 sat
    - Here we are referring to central cyanosis – best places to look mucous membranes in mouth, under tongue, not just lips

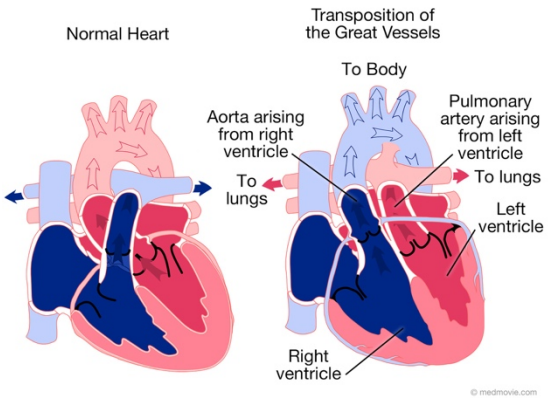
Tetralogy of Fallot

- Most common cyanotic lesion – 10% of all pediatric cardiac anomalies
- Four characteristics: large VSD, right ventricular outflow obstruction (due to pulmonic stenosis), overriding aorta, right ventricular hypertrophy
- Intensity of cyanosis depends on the amount of obstruction of right ventricular outflow tract
    - This also determines the shunting through VSD
        - Severe pulmonic stenosis → R -> L shunt → this will lead to cyanosis
        - Mild pulmonic stenosis → L -> R shunt → these are the ones you may discover later, will have stories of TET spells and episodic cyanosis, and can be helped by flexing hips and knees to increase pre-load
- CXR – boot shaped heart with decreased pulmonary markings

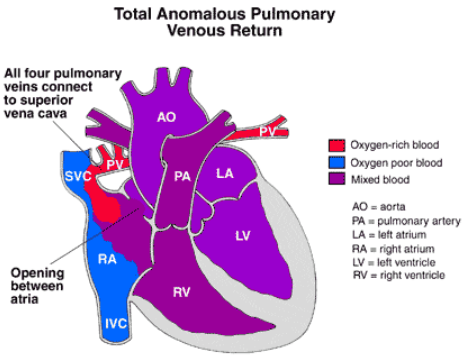


Transposition of Great Vessels

- Most common to present in newborn period – 6-8% of all pediatric cardiac anomalies
- Aorta arises from R ventricle and pulmonary artery arises from L ventricle
    - So you need communicating systems as this is not compatible with life – need VSD, ASD, PDA or any combination of those
- CXR – egg on a string, narrow mediastinum, increased pulmonary markings

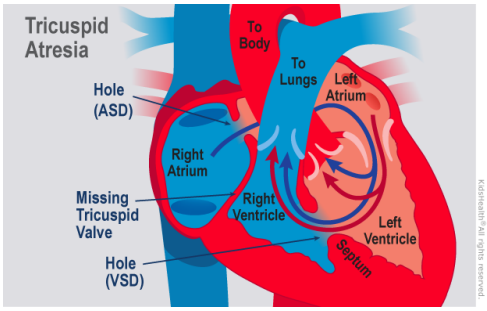


Total Anomalous Pulmonary Venous Return



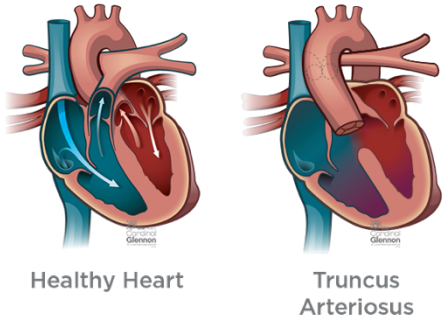
- 1% of all pediatric cardiac anomalies
- Pulmonary veins empty into R ventricle after returning from lungs instead of emptying into L ventricle
- Four types: depending on where the veins are emptying
    - Supracardiac (empty into SVC) – 50%
    - Cardiac (empty into coronary sinus) – 20%
    - Infracardiac (empty into portal vein, hepatic artery, IVC, ductus venosus) – 20%
    - Mixed lesions – 10%
- Again, not compatible with life without communications – need VSD or PDA or both
- CXR – Snowman sign, cardiomegaly, increased pulmonary markings

Tricuspid Atresia



- 1-2% of all pediatric cardiac anomalies
- Absence of tricuspid valve resulting in underdeveloped RA, RV and decreased flow to pulmonary vasculature
- Again, not compatible with life without communications – need VSD or PDA or both
- CXR – Normal heart with decreased pulmonary markings

Truncus Arteriosus



- <1% of all pediatric cardiac anomalies
- All pulmonary, systemic and coronary circulations arrive from one arterial trunk
- Have to have various other abnormalities to be compatible with life such as VSD, ASD, PDA, coronary artery irregularities
- CXR – Cardiomegaly with increased pulmonary markings