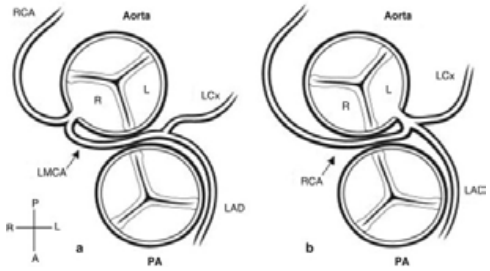
Coronary Artery Anomalies	
Anomalous Left Coronary Artery off the Pulmonary Artery (ALCAPA)	
Workup	CXR w/ cardiomegaly, pulmonary edema**.** EKG w/ signs of anterolateral ischemia manifest as pathologic Q waves (often very deep, but fairly narrow), inverted T waves and ST-segment elevation in leads I, aVL and V4-V6. Prolonged QTc may also be seen. **Echo is definitive**, may confirm w/ MR/ CT/angiography
Treatment	Surgery to reimplant LA to aorta and patch pulmonary artery
Anomalous Aortic Origin of a Coronary Artery (AAOCA)	
Presentation	Range from asymptomatic→ massive ischemia and **sudden death**
Pathophys	Variation in the number, shape or location of the ostia (origin) of the coronary arteries, usually non pathologic. LCA or LAD arising from the right coronary cusp leads the anomalous vessel to course anteriorly around the aortic valve, placing the vessel between the aorta and pulmonary artery and at risk for compression during times of peak cardiac output. Anomalous LCA from the right coronary cusp (picture on L) is always trx w/surgery, even if asymptomatic, due to high risk of sudden death Anomalous RCA from the left coronary cusp (picture on R) is also associated w/ increased frequency of sudden death, though to a lesser extent. Treatment is debated. 

Pulmonary Hypertension	
Presentation	Acute→ Sx of right heart failure. CHronic→ dyspnea w/ exertion and fatigue. Can lead to hemoptysis and sudden death from arrhythmias. Exam w/ RV heave, +/- TR murmur, cyanosis, clubbing, RHF signs such as JVD, hepatomegaly, peripheral edema
Pathophys	**Mean pulmonary atrial pressure >25 mmHg at rest**. Causes are 1. Pulmonary arterial HTN 2. Left heart dysfunction/obstruction 3. Lung pathology or hypoxemia 4. Chronic thromboembolism 5. Multifactorial
Workup	• **EKG**: **RV hypertrophy** often w/ accompanying strain (excessive right-sided forces for age w/ QRS-T angle > 90 degrees) In children, upright T-waves in V1 after 7-10 days of life suggests this diagnosis as can a qR pattern in V1. • **CXR**: may show mildly enlarged cardiac chambers, underlying lung disease and **prominent proximal pulmonary arteries** w/ diminished distal pulmonary vasculature. • **Echo**: may show enlarged or **hypertrophied right-sided chambers**. Position of the interventricular septum (which should bow into the usually low pressure RV) may flatten or bow into the LV. If present, the TR jet can estimate RV pressure using the Bernoulli equation (upper limit of normal is ~25mmHg). Septal defects may also be used in this manner. • Definitive diagnosis of pulmonary hypertension is done via **cardiac catheterization**. Mean PA pressures greater than 25 mmHg are diagnostic. This often performed w/ pulmonary vasodilator testing to assess response to potential therapies.
Treatment	Correct underlying cause! Counseling to avoid strenuous activity esp. Isometric exertion, avoid alpha adrenergic meds. Pulmonary vasodilators can be used→ Remodulin (IV infusion of trepostinil), Bosentan (endothelin receptor antagonist), Sildenafil (phosphodiesterase inhibitor), nifedipine (calcium channel blocker), iNO