



Up2Date

March 2013

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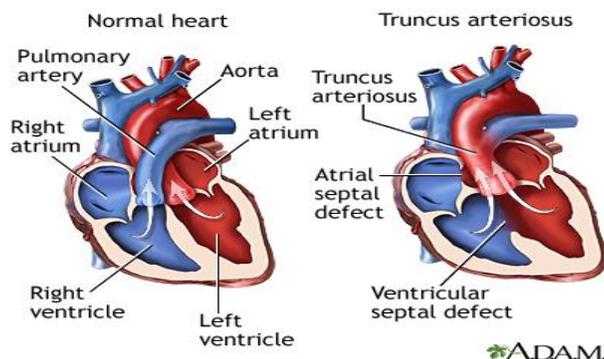
Cyanotic Congenital Heart Diseases in infants

You are on a flight with a HROB nurse, and the delivery is imminent. The HROB nurse will be caring for the mother, while you will be expected to care for the newborn infant. Here is one way to remember the top congenital heart diseases causing cyanosis, and what to do about them. These are scary situations, so if you ever find yourself here, hopefully this will help.

There are 5 main congenital defects you may see- you can remember them on one hand.

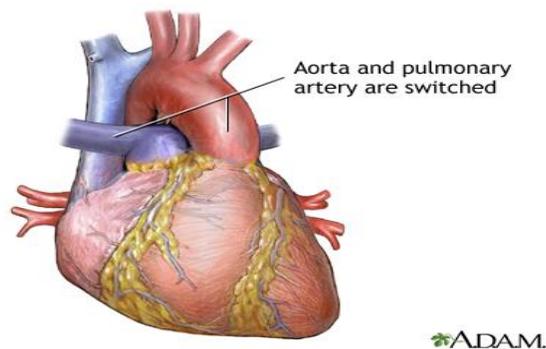


Truncus arteriosus: Occurs when the pulmonary artery and the aorta are joined together – creating one vessel instead of two. (Usually the pulmonary artery stems from the right ventricle and the aorta comes from the left ventricle.) The concern with Truncus arteriosus is that the oxygenated and deoxygenated blood are mixed, and there is too much blood being sent to the lungs, causing the infant to become short of breath.

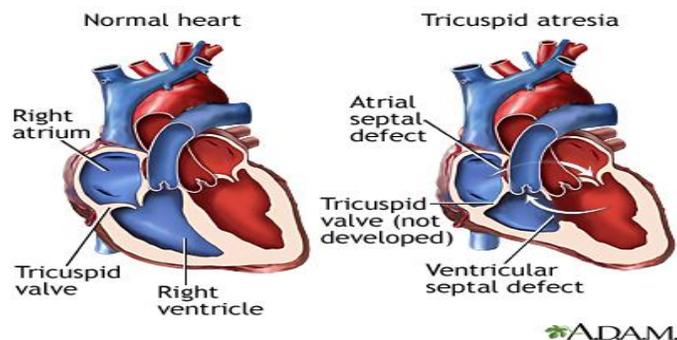




Transposition of the great vessels: In transposition of the great vessels, the aorta and the pulmonary artery are switched. This creates two distinct circulatory systems. The aorta arises from the right ventricle and the main pulmonary artery comes out of the left ventricle. Therefore, oxygenated blood is pumped into the lungs, and hypoxic blood is pumped to the rest of the body. These babies can only survive if they have a VSD, ASD, PDA (patent ductus arteriosus) so that some oxygenated blood gets into the systemic circulation. These babies are cyanotic and appear to be short of breath. These infants will need prostaglandin to keep a patent ductus arteriosus allowing some of the blood to mix until surgery can be performed.

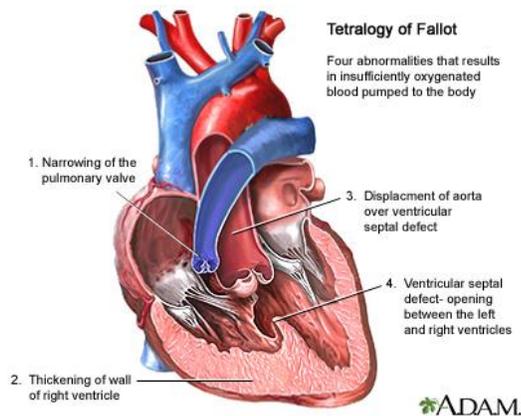


Tricuspid Atresia: The tricuspid valve is either missing or non-functioning. There is poor development of the right ventricle and pulmonary artery. The blood cannot continue its normal path to the lungs and the infant will become hypoxic and cyanotic.

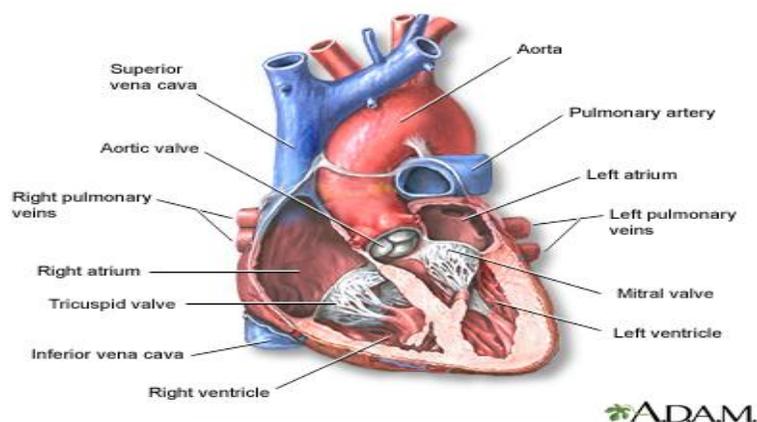




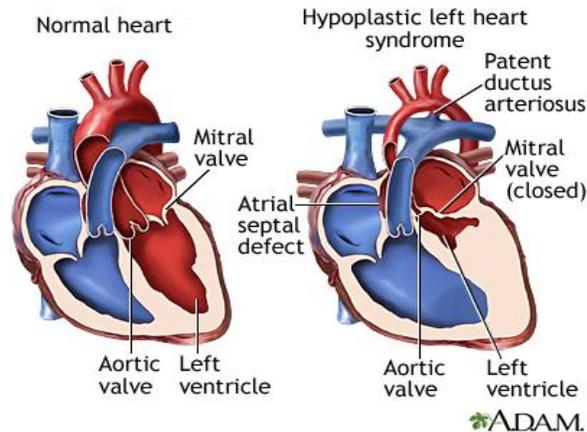
Tetralogy of Fallot: Combination of 4 cardiac defects that inhibit oxygenated blood reaching the body. These 4 defects include: a VSD, pulmonary outflow obstruction, an overriding aorta (the aorta pulls blood from both the right and left ventricle), and right ventricular hypertrophy. The amount of cyanosis depends on the amount of obstruction. Surgery is required but sometimes at a later age (3-5 years).



Total anomalous pulmonary venous return: (5 fingers because of the 5 components of the name) Occurs when the pulmonary veins that return circulation to the heart empty into the right atrium (with variances) rather than the left atrium. In order for the infant to survive, an atrial septal defect or a PFO must be present.



Hypoplastic Left Heart: This one does not have a fancy way to remember it, except that it too can cause the newly born infant to be cyanotic. In a hypoplastic left heart, the structures are essentially undeveloped resulting in poor perfusion to the body. These infants need surgery. To survive in the interim, they are given Prostaglandin to allow the ductus arteriosus to remain open. (The Ductus Arteriosus is a small opening allowing the blood to leave the pulmonary circulation and circulate through the body). The infant must also have a patent foramen ovale which will allow blood to move between the right and the left side of the heart which it would not otherwise do if the foramen ovale was closed.



General Treatment of Congenital Heart Disease:

Emergent treatment is supportive in nature. If the mother knows she has a neonate with a congenital heart disease, hopefully she will communicate that information. Otherwise, you will be left to find out rather quickly. Stick with your ABC's. If these kids need an airway, usually BVM support is sufficient. If not, move to an advanced airway that is secure. Ensure they are properly ventilated, and that they have circulation that is efficient as possible.

References:

Truncus Arteriosus: [http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002101/?log\\$=activity](http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002101/?log$=activity)

Transposition of the great vessels:

[http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002535/?log\\$=activity](http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002535/?log$=activity)

Tricuspid Atresia: [http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002100/?log\\$=activity](http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002100/?log$=activity)

Tetralogy of Fallot: <http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002534/>

Total Anomalous Pulmonary Venous Return:

[http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002104/?log\\$=activity](http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002104/?log$=activity)

Hyoplastic Left heart:

[http://www.ncbi.nlm.nih.gov/pubmedhealth/?term=hyoplastic+left+heart&log\\$=activity](http://www.ncbi.nlm.nih.gov/pubmedhealth/?term=hyoplastic+left+heart&log$=activity)