Congenital Heart Disease

A BRIEF REVIEW

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Congenital Heart Disease
A BRIEF REVIEW

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I have no actual or potential conflict of interest in relation to any product or service mentioned in this presentation.

Congenital Heart Disease
A Brief Review

Objectives

- Review some of the more common congenital heart problems seen in infants
- Understand the normal circulation of blood through the heart
- Explain the terms Oxygenated and Unoxygenated blood
- Understand the term Cyanosis
- Recognize that infants with congenital heart problems grow up to be adults with heart problems
- Acquire the basic information and terminology needed to assist patients and their caregivers

Congenital Heart Disease
A BRIEF REVIEW

Why is this topic important to this audience?

Congenital Heart Disease
A Brief Review

- Congenital Heart Defects (CHD) are the most common type of birth defects
- CHDs affect nearly 1% of births per year in the United States.
  - About 40,000 babies
- Approximately 1 in 4 babies with a CHD have a **Critical CHD**
  - These infants need surgery or other interventions in their first year of life

Congenital Heart Disease
A Brief Review

- You must be educated in order to help the families with whom you have contact
- Example:
  - -If a child has and elevated blood sugar, would you not suggest to the family that he/she have a visit with the PCP or an endocrinologist?
- Recognition of the signs of Congenital Heart Disease enables you to consider appropriate referrals also.

Congenital Heart Disease
A Brief Review

Congenital Heart Disease Can Be Missed

- Congenital Heart Disease can be missed at birth
- Congenital Heart Disease can be diagnosed at birth, but no follow up is done
- Congenital Heart Disease may not show up until later in the first year of life or thereafter
- Surgical and medical care of infants and children with congenital heart disease has dramatically improved
  - -Over 85% will reach adulthood; they will require ongoing knowledgeable care

Congenital Heart Disease
A Brief Review

Critical Heart Defects

Congenital Heart Disease
A Brief Review

- Critical Heart Defects (CHD)
  - Critical Congenital Heart Disease
  - 1 in every 4 babies born with a heart defect has a Critical CHD
  - Critical CHD needs surgery or other intervention(s) in the first year of life
  - Most will be Cyanotic

Congenital Heart Disease
A Brief Review

- Critical Heart Defects
  - In the United States,
  - 7,200 babies born every year have Critical CHDs
  - Babies with these heart defects have low levels of oxygen in the blood

Congenital Heart Disease
A Brief Review

- Critical Heart Defects
  - -Some CHDs may be diagnosed during pregnancy
  - -Diagnosis is with an ultrasound called a fetal echocardiogram
  - -Creates pictures of the heart

Congenital Heart Disease
A Brief Review

- Critical Heart Defects
  - -Newborn screening for CHDs
  - -Bedside test
    - uses a pulse oximeter
  - -Measures the amount of oxygen in a baby's blood
  - -Low oxygen in the blood may be sign of a Critical CHD.

Congenital Heart Disease
A Brief Review

- Critical Heart Defects
  - Screening
    - Done at least 24 hours of age
    - Or as late as possible if the baby will be discharged early
  - Babies with a Critical CHD need intervention ASAP

Congenital Heart Disease
A Brief Review

The Story of Oxygen Transport by the Blood

- Understanding Cyanosis
- Knowing the physiology of oxygen transport is necessary to fully understand the impact a poorly functioning heart has on the body.

Congenital Heart Disease
A Brief Review

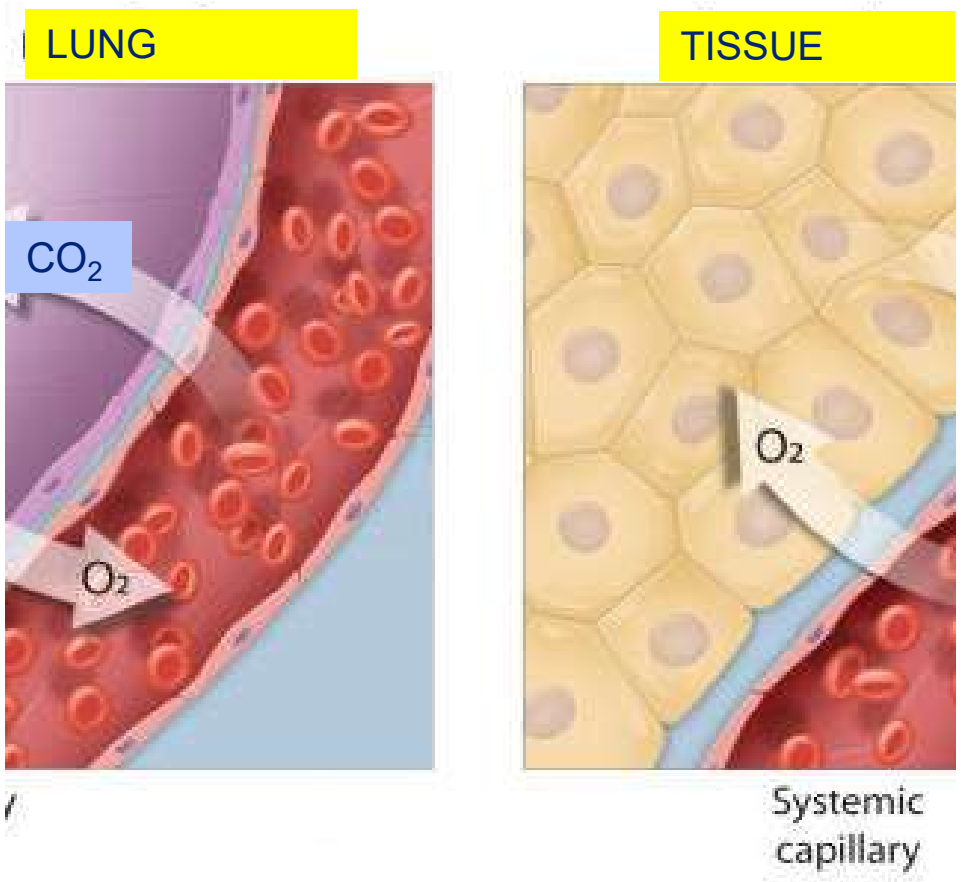
The Story of Oxygen Transport by the Blood

- Hemoglobin is the molecule in the Red Blood Cell that carries the Oxygen in the Blood

Congenital Heart Disease
A Brief Review

The Story of Oxygen Transport by the Blood

- In the Lungs, the Hemoglobin absorbs Oxygen from the inhaled air
  - -It becomes “saturated” with Oxygen



Congenital Heart Disease
A Brief Review

The Story of Oxygen Transport by the Blood

- Saturated Hemoglobin is RED in color
- When the Blood leaves the Lungs, it is pumped to the rest of the Body

Congenital Heart Disease
A Brief Review

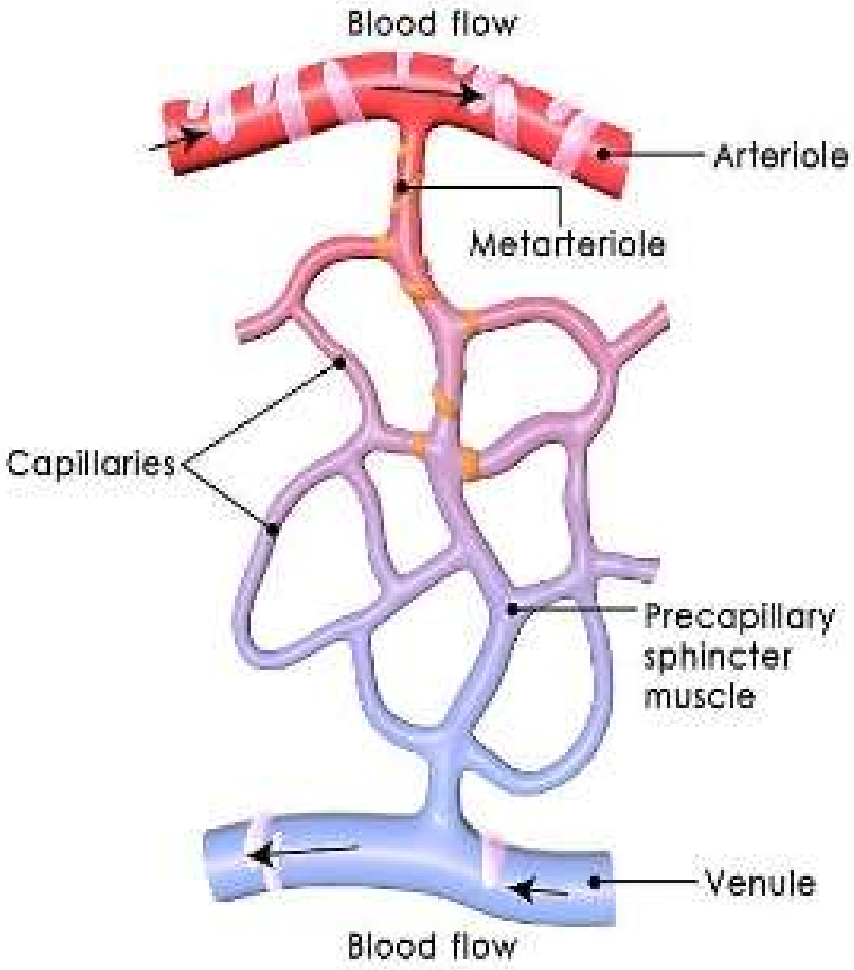
The Story of Oxygen Transport by the Blood

- As Blood Vessels get further from the Heart, they become progressively smaller

Congenital Heart Disease
A Brief Review

The Story of Oxygen Transport by the Blood

- In all tissues, the smallest vessels are called Capillaries



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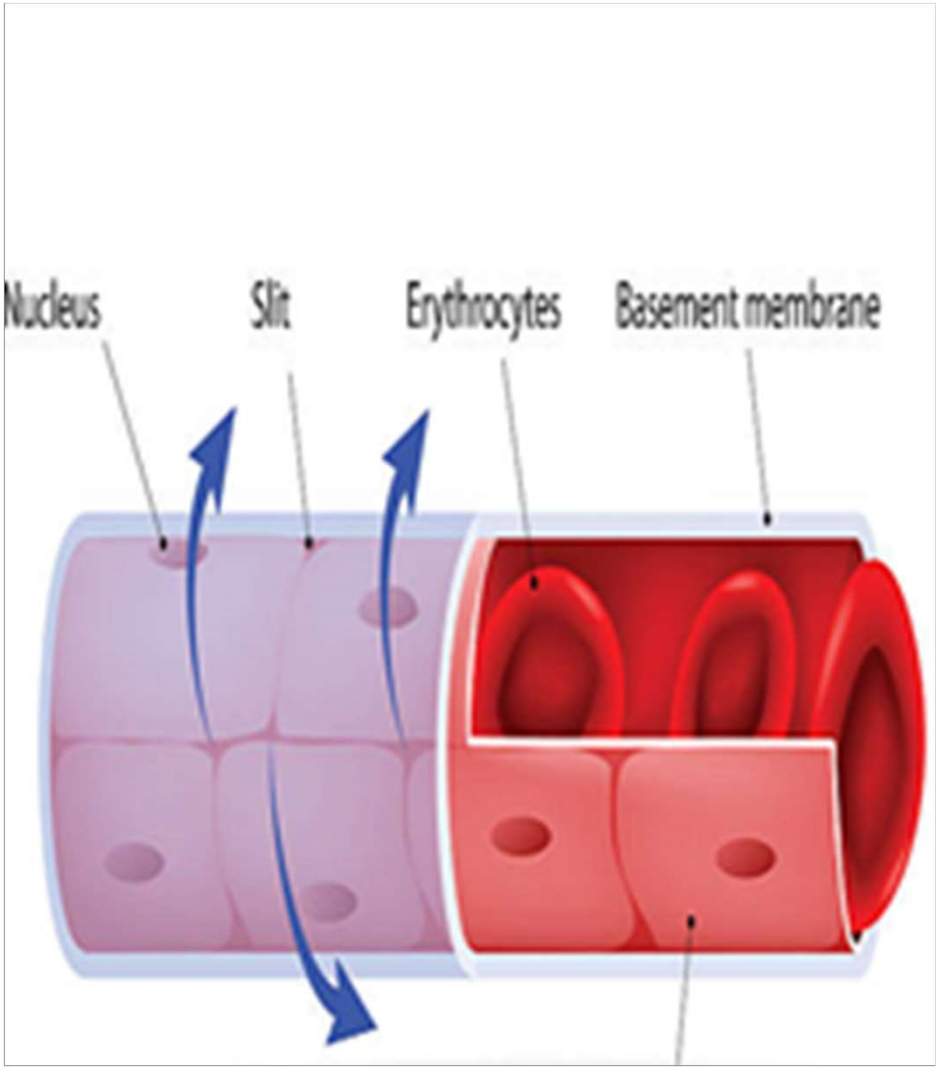
A Brief Review

The Story of Oxygen Transport by the Blood

- Capillaries are just slightly larger than the diameter of a Red Blood Cell



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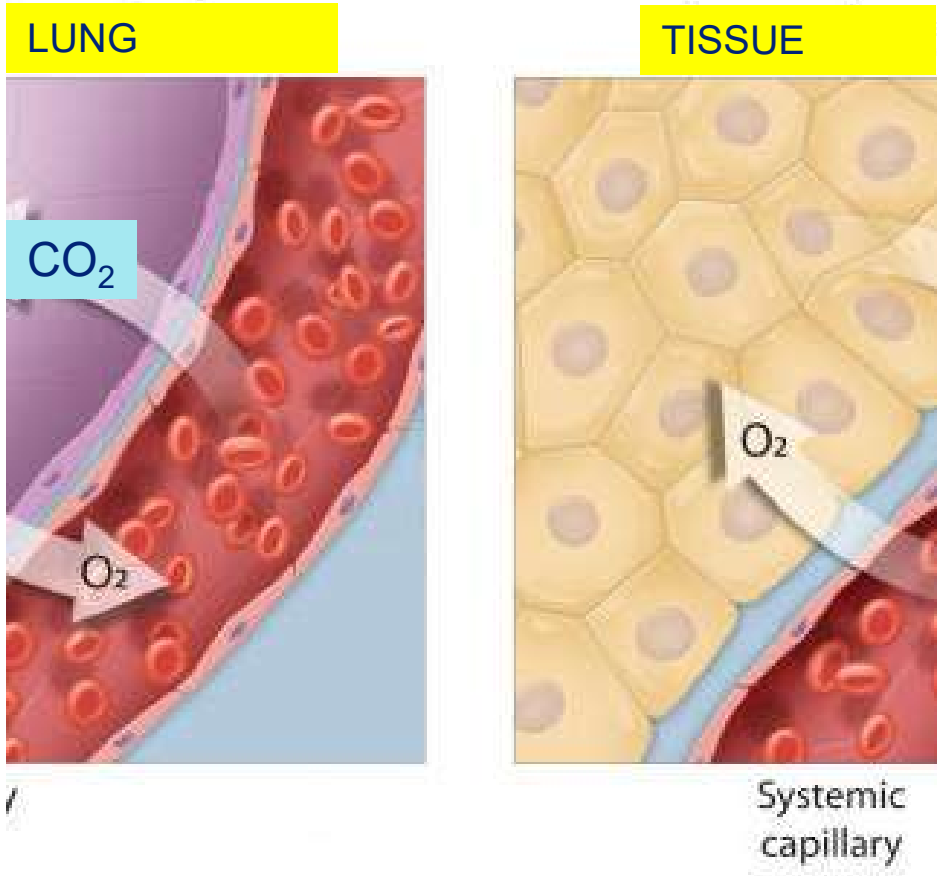
A Brief Review

- The Story of Oxygen Transport by the Blood

- In the **Tissue** Capillary, the Red Blood Cell is very close to the tissue cells
- The tissue cells have a greater attraction for Oxygen than does Hemoglobin



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A Brief Review

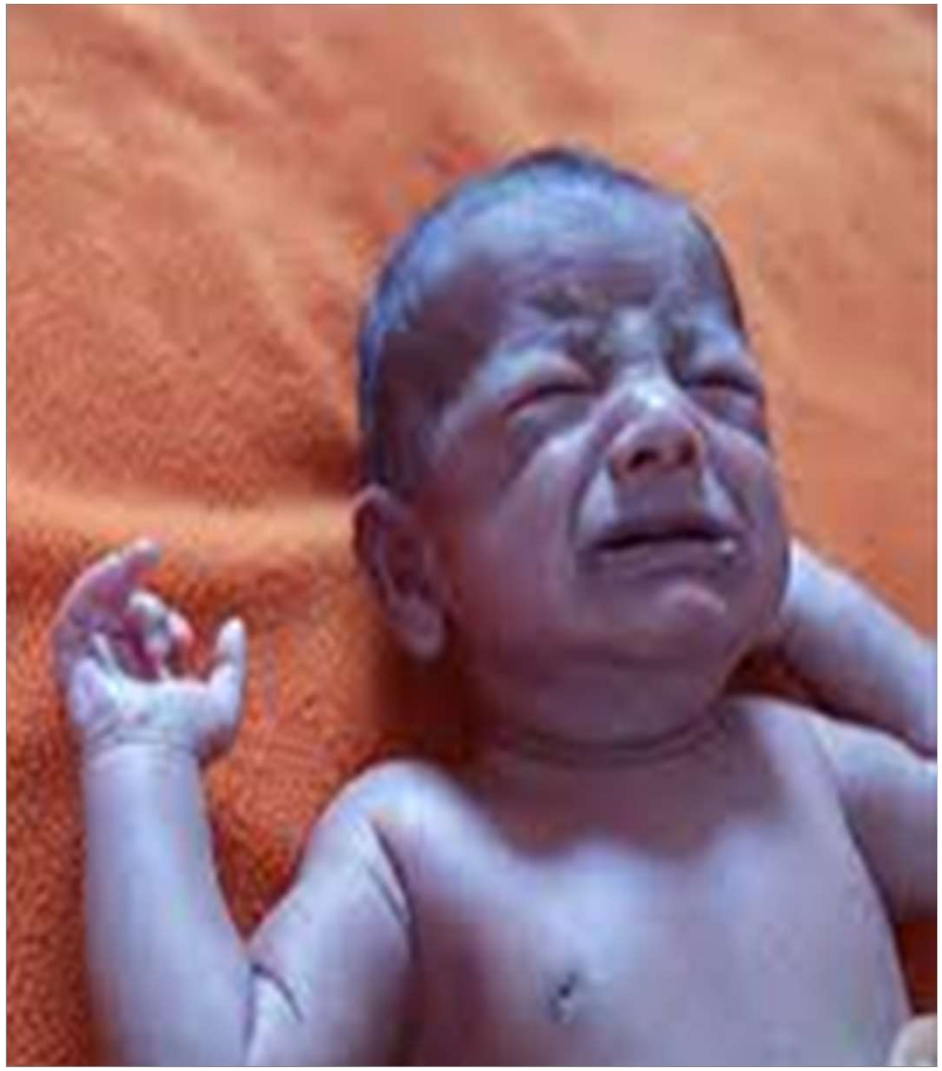
The Story of Oxygen Transport by the Blood

- The Red Blood Cell Hemoglobin gives up the Oxygen to the tissue cells
- Without Oxygen, the Hemoglobin turns Blue
- If a large percent of the hemoglobin in the arteries is Blue, it shows up in the skin as

Cyanosis

Congenital Heart Disease
A Brief Review
Cyanosis

- Cyanosis refers to a bluish-purple hue to the skin.
- It is most easily seen where the skin is thin,
- Lips
- Mouth
- Earlobes
- Fingernails
- Caused by not enough oxygen in the blood



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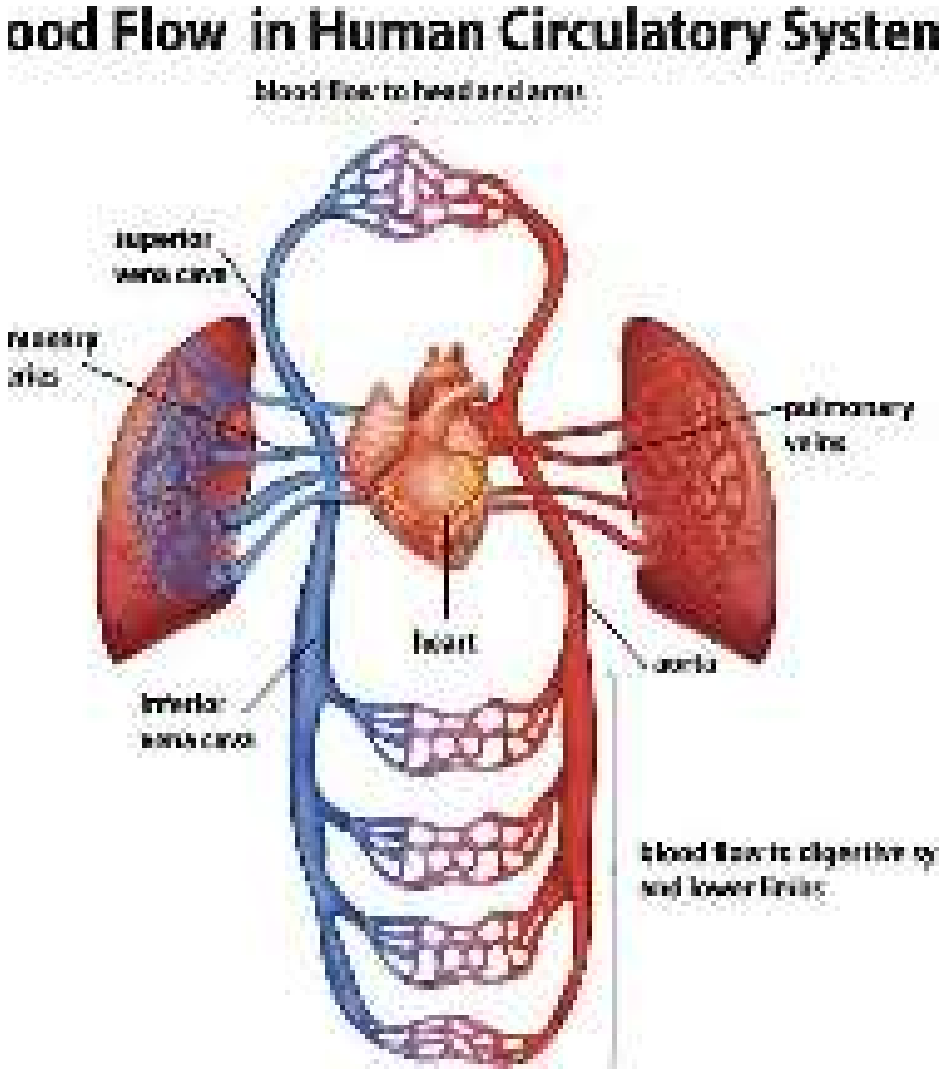
A Brief Review

The Story of Oxygen Transport by the Blood

- On the other side of the Capillary is a very small Vein
- Veins get progressively larger as they connect to form larger veins
- They return the blood to the Heart



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A Brief Review

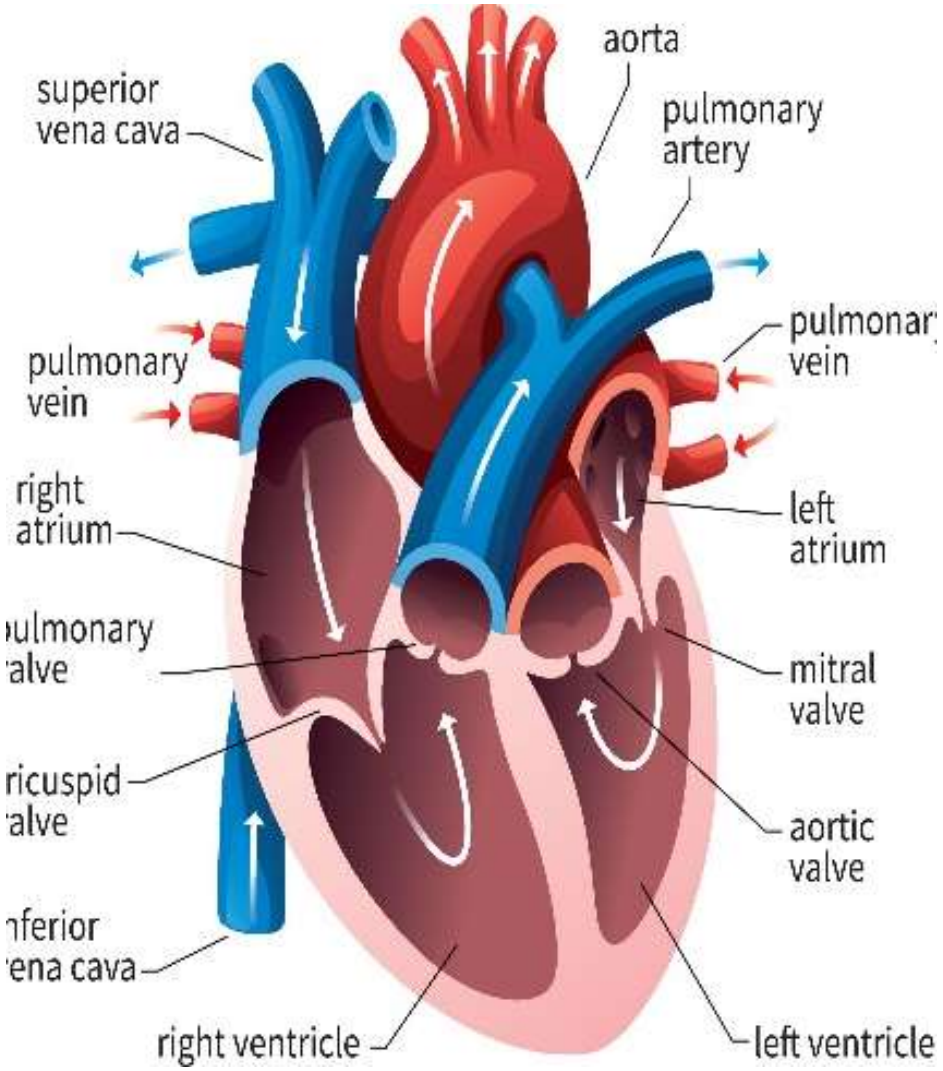
The Story of Oxygen Transport by the Blood

- Because the blood in the veins has lost its Oxygen, it is “unsaturated”
- The Hemoglobin in the Veins is Blue

Congenital Heart Disease
A Brief Review

Normal Heart Circulation

- This is why the pictures of vessels on the Right side of the heart are Blue and those on the Left side are Red



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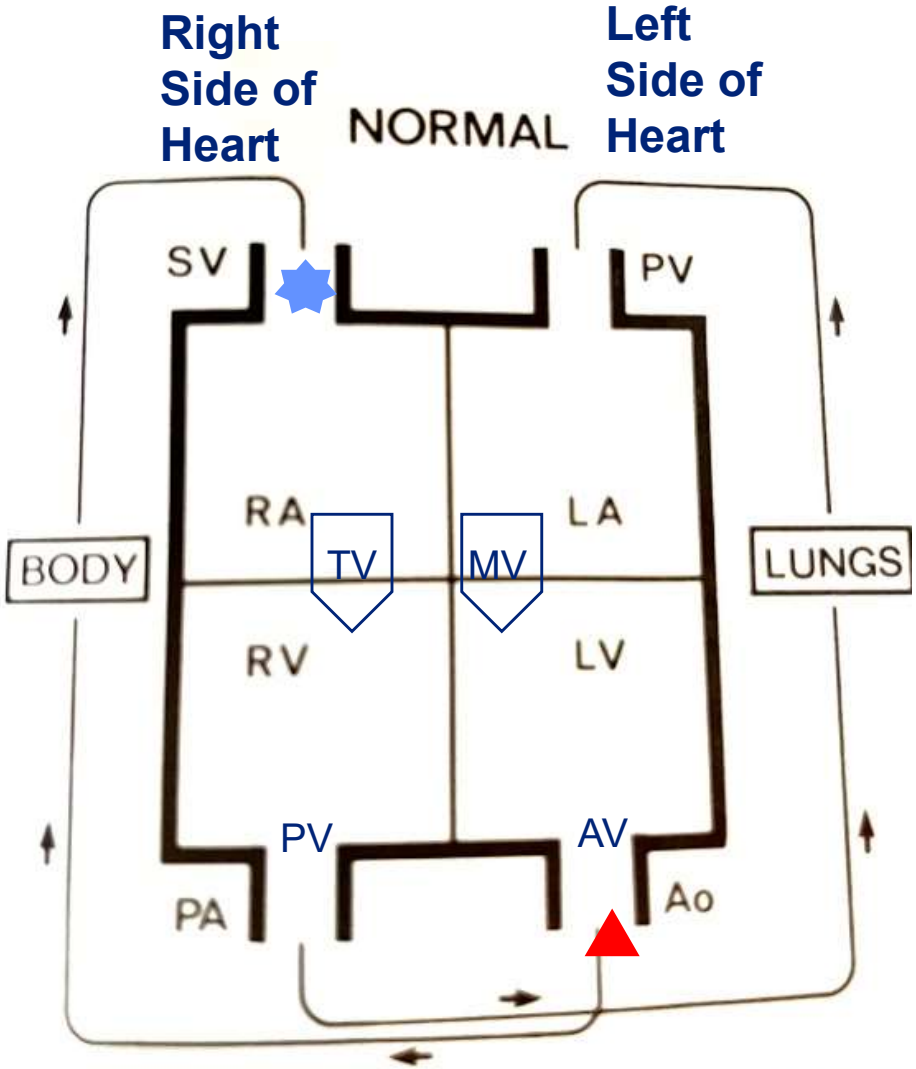
Congenital Heart Disease
A Brief Review

Normal Heart Circulation

- The pictures of the Heart will always have the Right side on the Left and the Left side on the Right
- Blood pumped to the body through the AORTA (Ao)
- Blood returns from the body through VENA CAVAS (SV)



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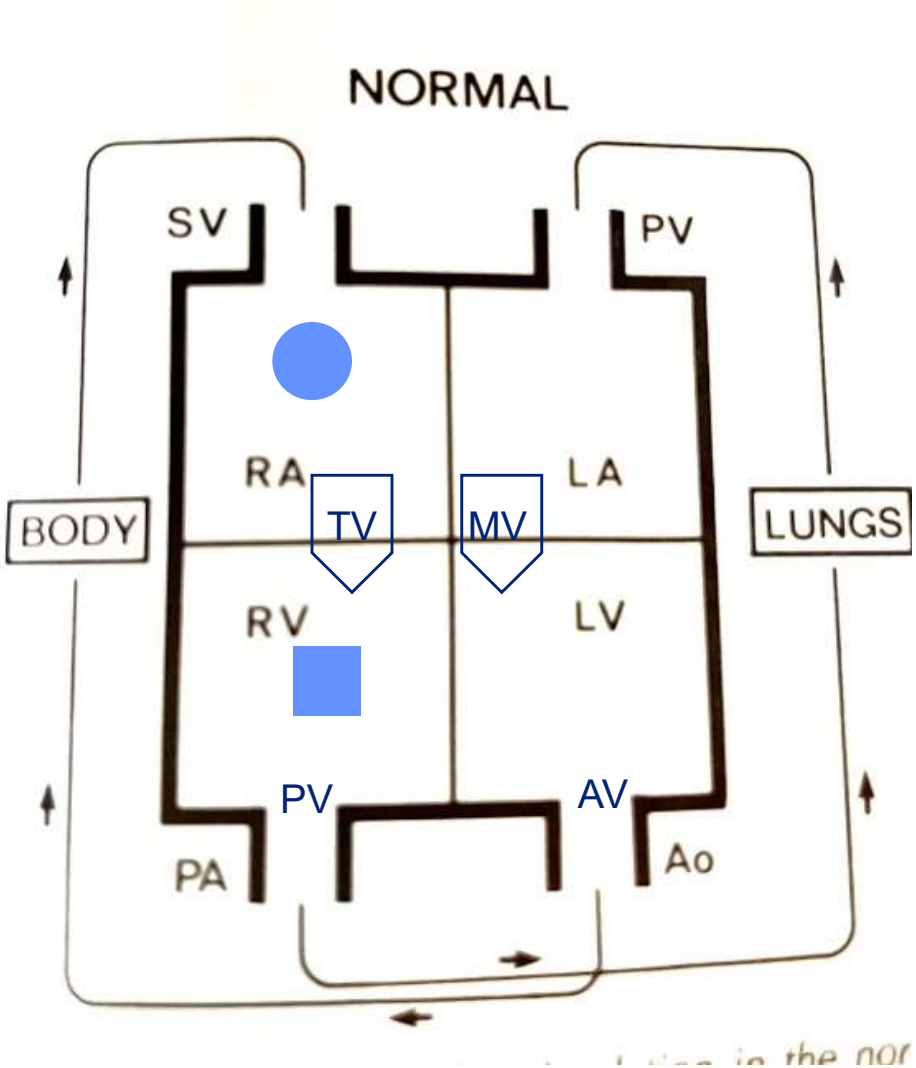
A Brief Review

Normal Heart Circulation

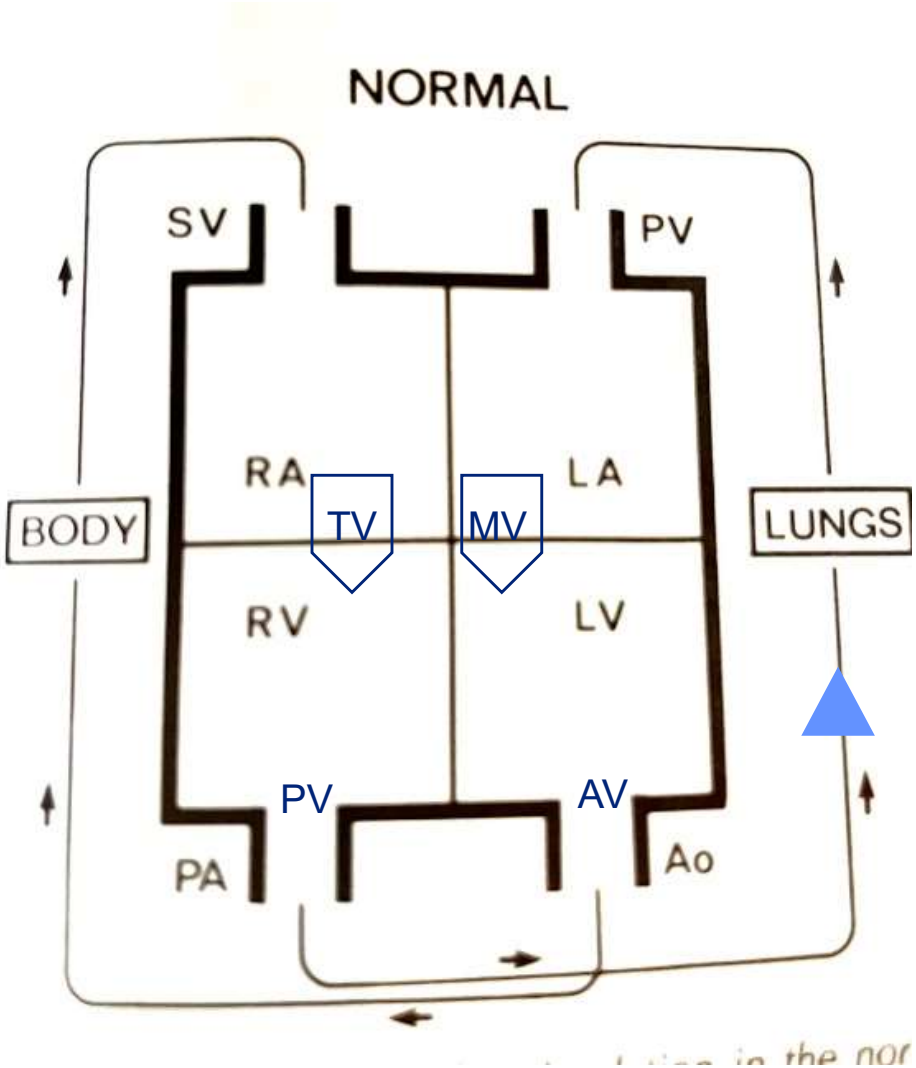
- VENA CAVA puts blood in the RIGHT ATRIUM (RA)
- Blood is pumped through the TRICUSPID VALVE (TV) into the RIGHT VENTRICLE (RV)



Congenital Heart Disease
A Brief Review

Normal Heart Circulation

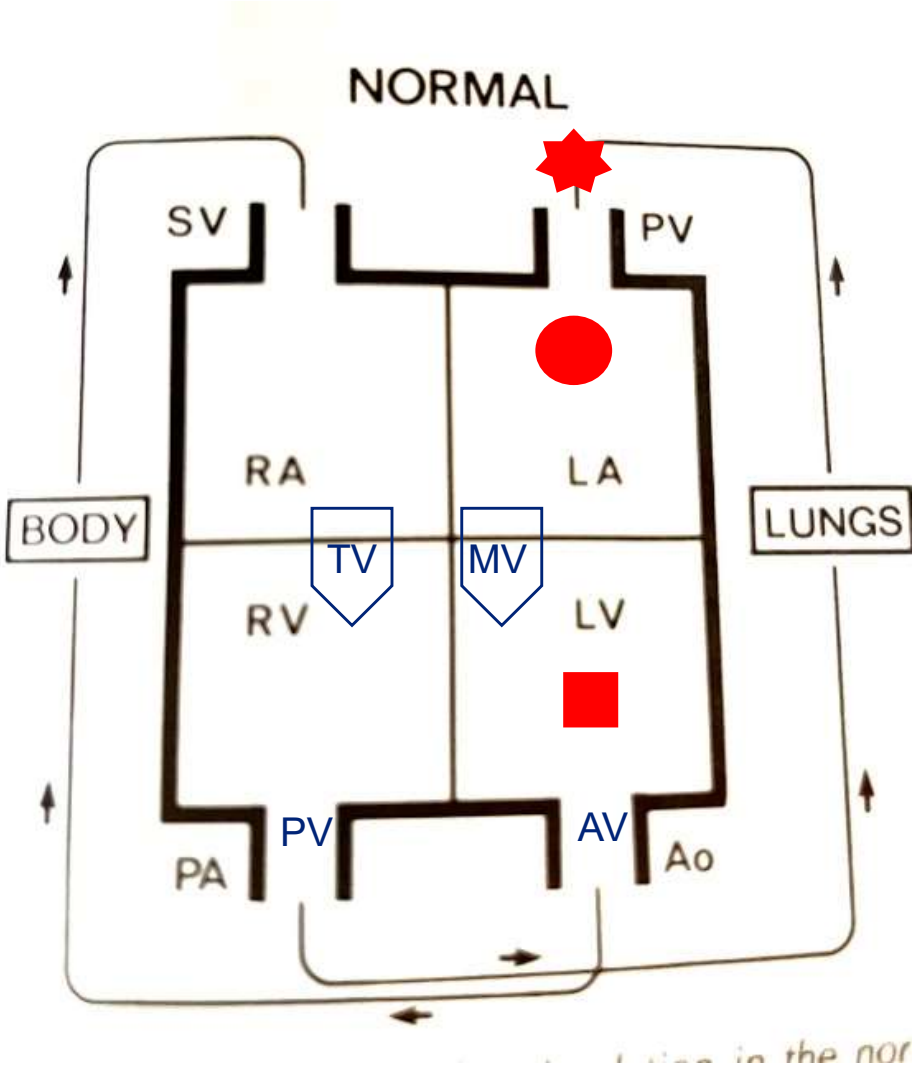
- Blood is pumped through the PULMONARY VALVE (PV) into the PUMONARY ARTERY
- The PULMONARY ARTERY carries blood to the LUNGS
- The blood receives oxygen in the lungs



Congenital Heart Disease
A Brief Review

Normal Heart Circulation

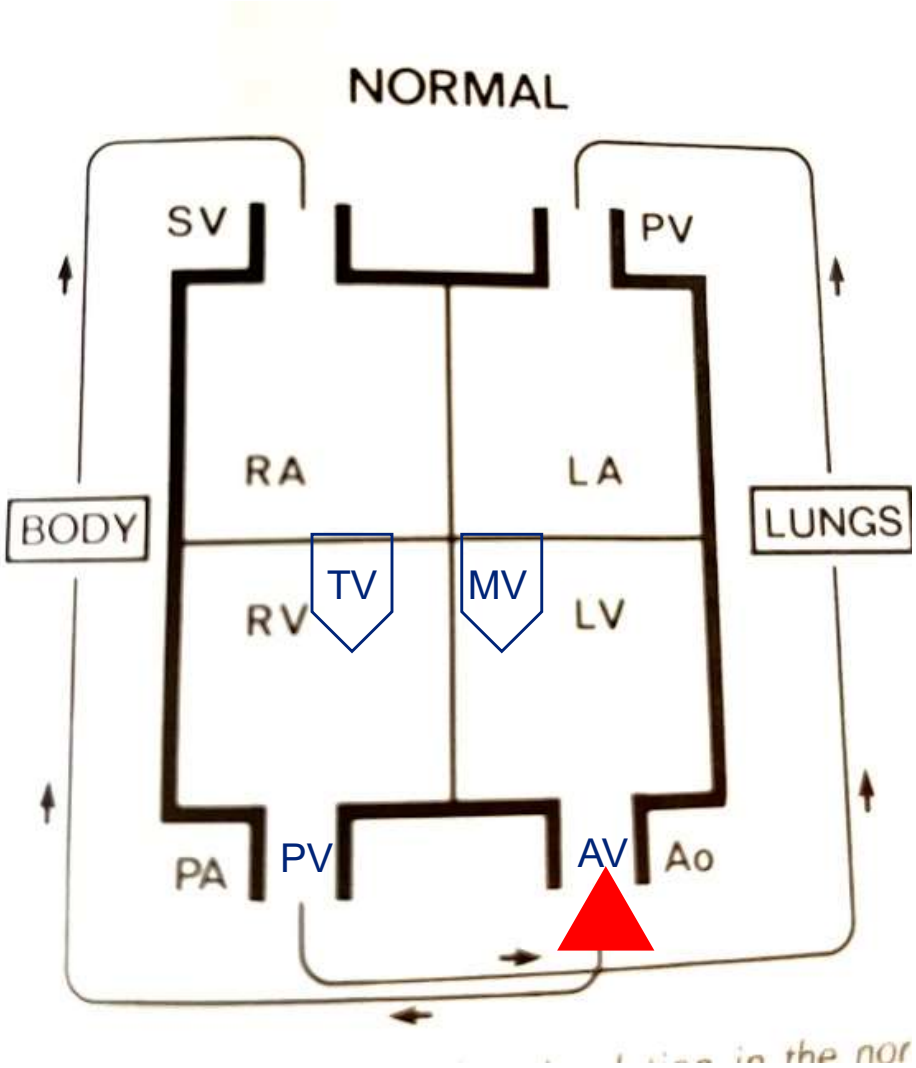
- The LUNGS send the oxygenate blood through the PULMONARY VEINS back to the heart; into the LEFT ATRIUM
- The LEFT ATRIUM pumps the blood through the MITRAL VALVE (MV) into the LEFT VENTRICLE



Congenital Heart Disease
A Brief Review

Normal Heart Circulation

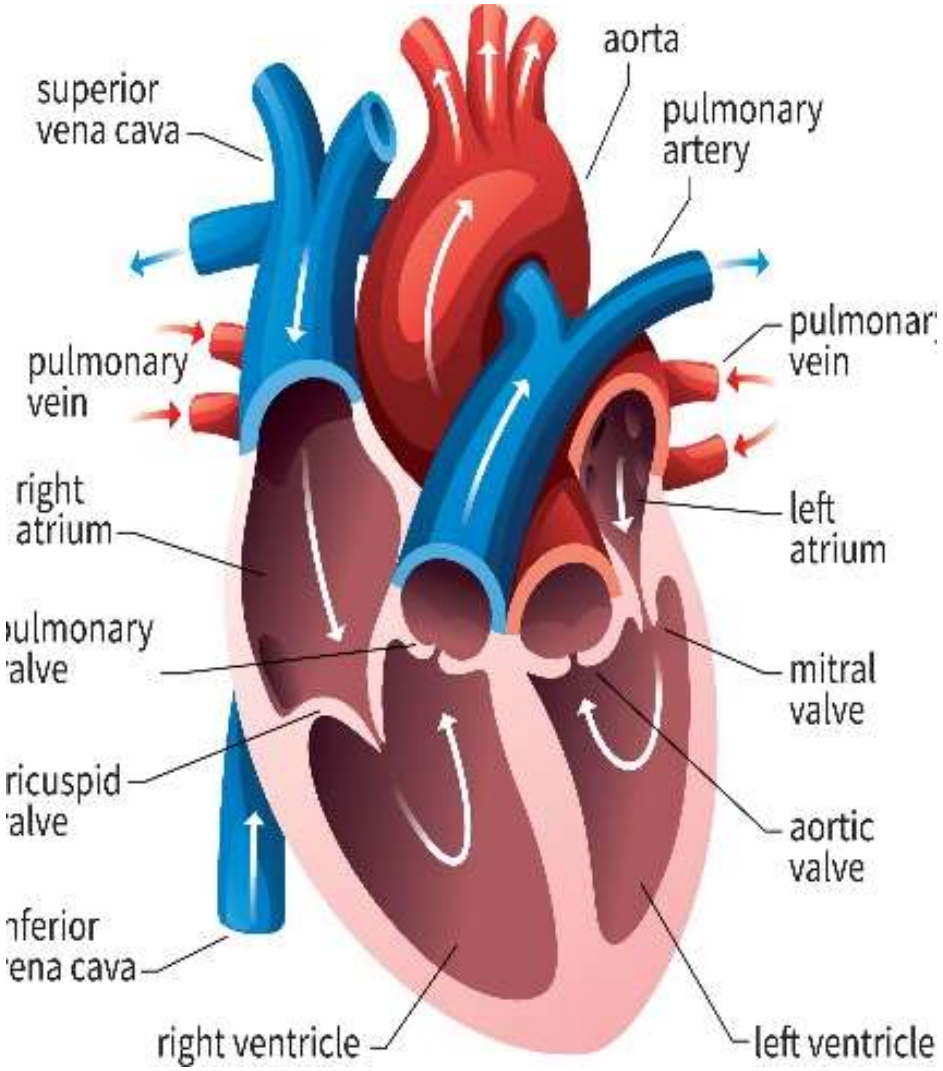
- The LEFT VENTRICLE pumps the blood through the AORTIC VALVE (AV) into the AORTA
- Then it is distributed to the entire body



Congenital Heart Disease
A Brief Review

Normal Heart Circulation

- This is the same circulation in a model of the heart



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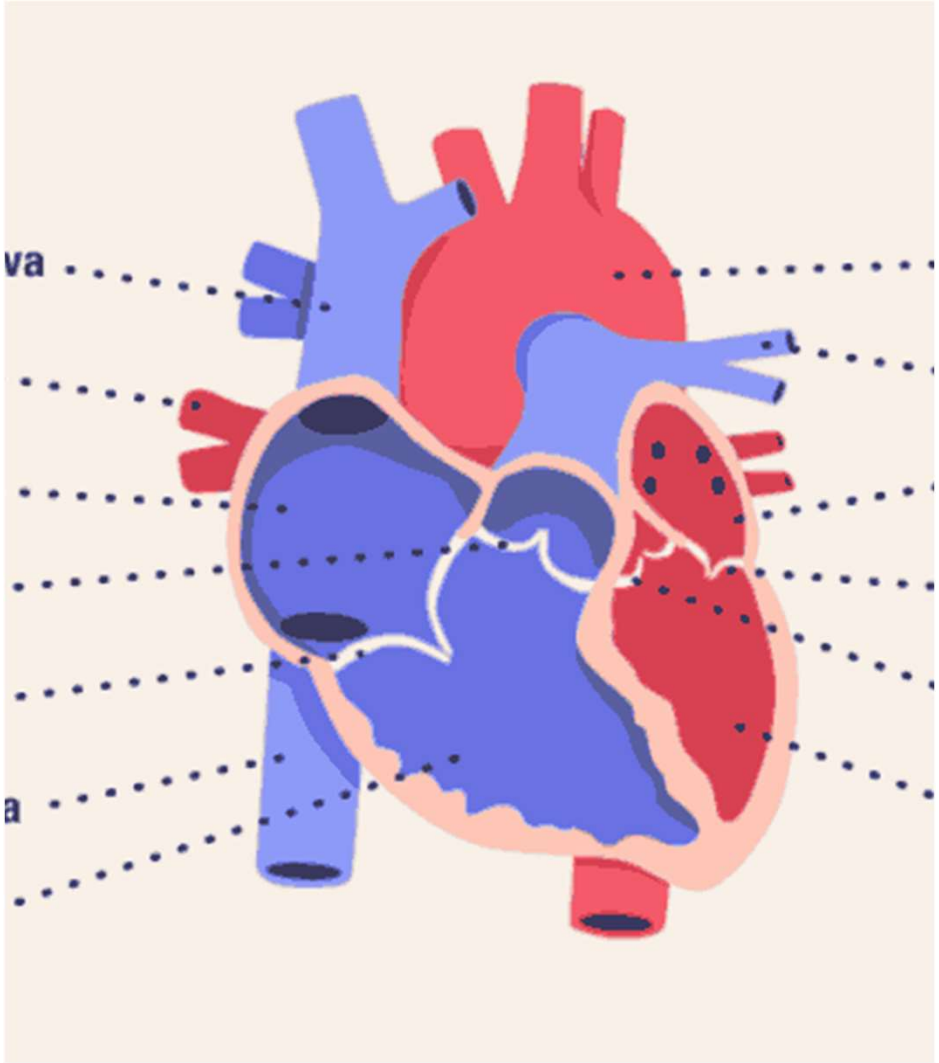
A Brief Review

Normal Heart Circulation

- This is how it looks in motion
- The pumping to the body and the pumping to the lungs happens at the same time.



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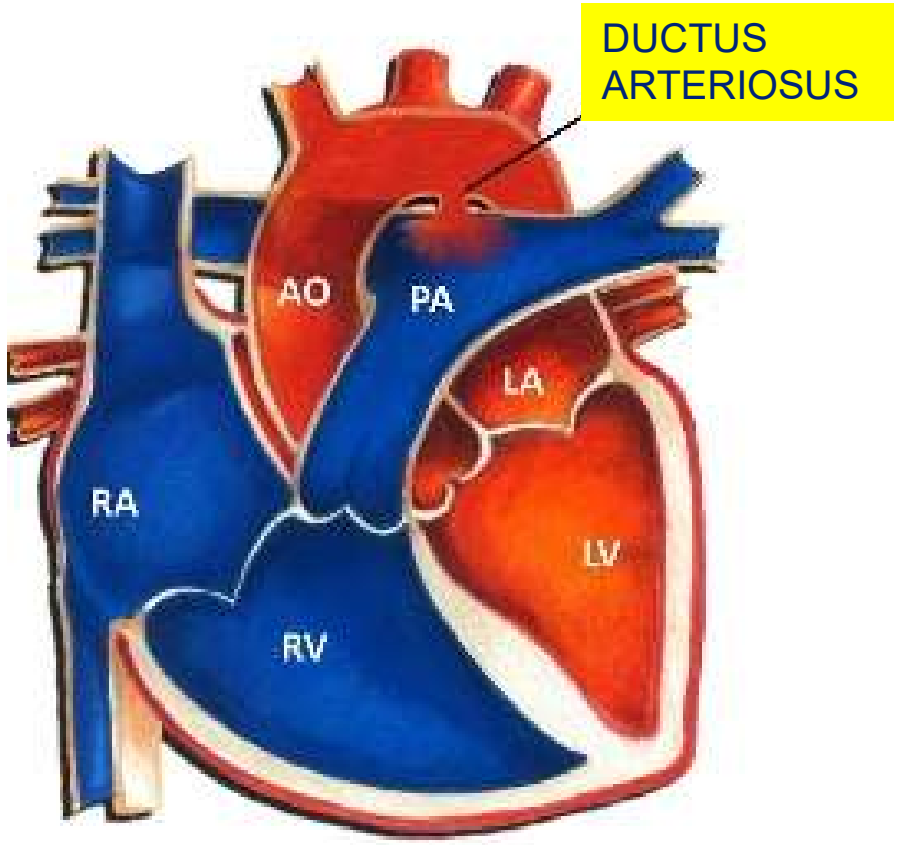
A Brief Review

Ductus Arteriosus

- The DUCTUS ARTERIOSUS (DA) is a NORMAL connection between the AORTA (AO) and the PULMONARY ARTERY (PA)
- Before birth it is open
- After birth it is normally closed



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Congenital Heart Disease
A Brief Review

Identification of

Congenital Heart Disease

- Most Congenital Heart Diseases are diagnosed using
  - -X-rays
  - -Ultrasound
  - -Catheterization with dye injection
  - -Some are identified *in utero*
    - Prenatal ultrasound

Congenital Heart Disease
A Brief Review

Types of Congenital

Heart Disease

There are 2 presentations of heart defects

Acyanotic

and

Cyanotic

Congenital Heart Disease
A Brief Review
Types of CHD

- Atrial Septal Defect
- Atrioventricular Septal Defect
- Coarctation of the Aorta*
- Double-outlet Right Ventricle*
- d-Transposition of the Great Arteries*
- Ebstein Anomaly*
- Hypoplastic Left Heart Syndrome*
- Interrupted Aortic Arch*
- Pulmonary Atresia*
- Single Ventricle*
- Tetralogy of Fallot*
- Total Anomalous Pulmonary Venous Return*
- Tricuspid Atresia*
- Truncus Arteriosus*
- Ventricular Septal Defect

Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

- Ventricular Septal Defect
- Atrial Septal Defect
- Atrioventricular Septal Defect
- Coarctation of the Aorta-**Critical CHD**

Congenital Heart Disease
A Brief Review
Types of CHD

Cyanotic Heart Defects- **Critical CHD**

- Ebstein Anomaly
- d-Transposition of the Great Arteries
- Double-outlet Right Ventricle
- Hypoplastic Left Heart Syndrome
- Interrupted Aortic Arch
- Pulmonary Atresia
- Single Ventricle
- Tetralogy of Fallot
- Total Anomalous Pulmonary Venous Return
- Tricuspid Atresia
- Truncus Arteriosus

Congenital Heart Disease
A Brief Review

Types of Congenital Heart Disease

We will review some of the more common heart defects.

Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

- Ventricular Septal Defect
- Atrial Septal Defect
- Atrioventricular Septal Defect
- Coarctation of the Aorta-**Critical CHD**

Congenital Heart Disease
A Brief Review
Types of CHD
Acyanotic Heart Defects
Presentation of Acyanotic CHD

- Shortness of breath
- Fast or heavy breathing
- Sweating
- Tiredness while feeding
- Weak pulse
- Poor weight gain
- Skipped heartbeats
- A heart murmur
- Swelling of legs, feet, or stomach area
- Pounding heart

Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

Ventricular Septal Defect (VSD)

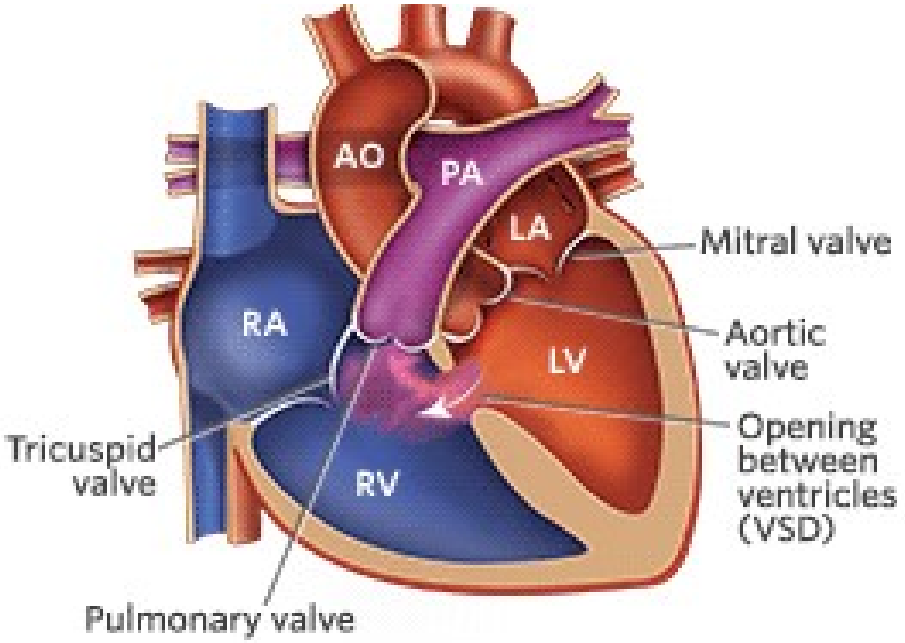
- Affects the 2 lower (large) chambers of the heart
- The septum (a wall) separates the two lower chambers
- The defect is a hole in the wall between them

Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

Ventricular Septal Defect

- Blood flows from the left ventricle to the right ventricle
- Extra blood is pumped into the lungs



Children's Hospital of Philadelphia

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A Brief Review
Types of CHD

Acyanotic Heart Defects

Ventricular Septal Defect

- Forces the heart and lungs to work harder
- Increases the risk for other complications if not repaired
  - -Heart failure
  - -High blood pressure in the lungs (pulmonary hypertension)
  - -Irregular heart rhythms (arrhythmia)
  - -Stroke.

Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

Ventricular Septal Defect

- The size and position of the hole is variable
- Treatment is to close the hole
  - -cardiac catheterization
  - -open-heart surgery
- Some VSDs close on their own
- If the VSD is closed, children live healthy lives

Congenital Heart Disease
A Brief Review
Types of CHD

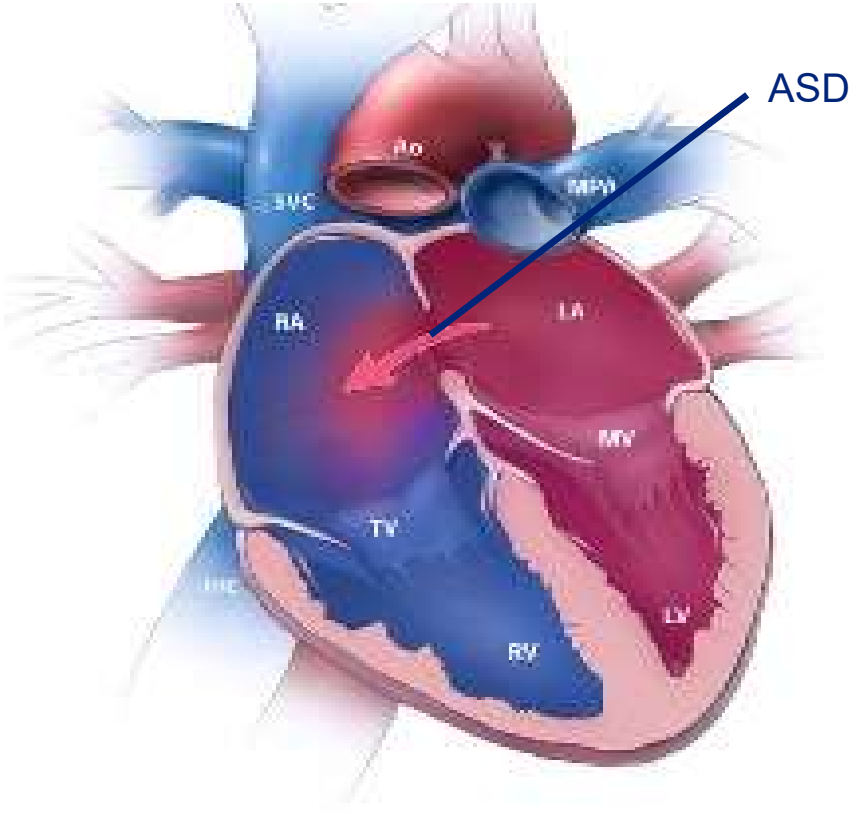
Acyanotic Heart Defects

Atrial Septal Defect (ASD)

- There is a hole in the wall (septum) between the 2 upper chambers (Atria) of the heart.
- Varies in size
- May be diagnosed during pregnancy, after birth, or not until adulthood.
- Some ASDs close on their own

Congenital Heart Disease
A Brief Review
Types of CHD
Acyanotic Heart Defects
Atrial Septal Defect (ASD)

- Blood flows from the Left Atrium to the Right Atrium through the ASD
- This causes increased blood flow though the Lungs
- The Lungs can be damaged from this



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A Brief Review
Types of CHD

Acyanotic Heart Defects

Atrial Septal Defect (ASD)

- A procedure may be necessary for persistent ASDs
  -Some close spontaneously
- Closure of the hole may be done during cardiac catheterization or open-heart surgery.
- Lung damage from Pulmonary Hypertension can occur
- With good care and follow up a normal life is possible

Congenital Heart Disease
A Brief Review
Types of CHD

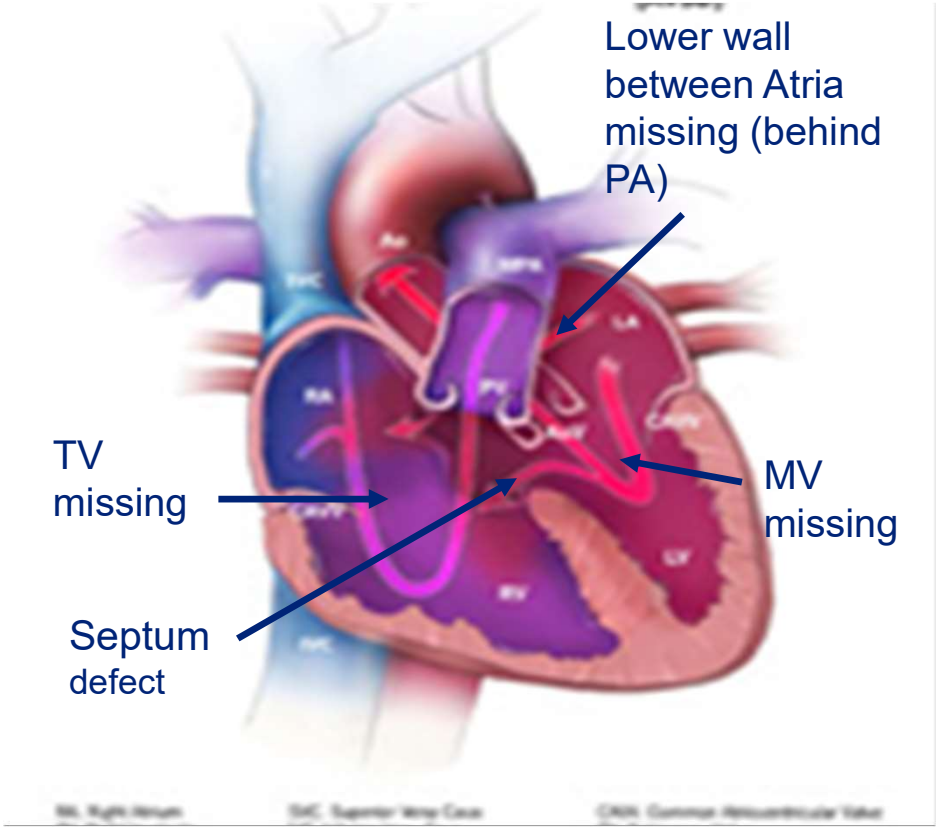
Acyanotic Heart Defects

Atrioventricular Septal Defect (AVSD)

- A defect that affects
  - The valves between the heart's upper and lower chambers
  - The walls between all 4 chambers
- Atrioventricular canal (AV canal) defect
- Endocardial cushion defect

Congenital Heart Disease
A Brief Review
Types of CHD
Acyanotic Heart Defects
Atrioventricular Septal Defect

- Defect may be complete or partial
- Mitral (MV) and Tricuspid (TV) valves are missing
- Lower wall of the Atria and part of the wall of the Ventricles (Septum) are missing



Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

Atrioventricular Septal Defect

- The defect in the center of the heart affects the conduction of signals for heart contraction from the SA node in the Atrium to the Ventricles
  - -Irregular heart rhythms (Arrhythmia) may occur
- Pulmonary Hypertension and Heart Failure occur

Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

Atrioventricular Septal Defect

- Surgery is required to fix the defects
- Valve repair or replacements may be needed
- Conduction arrhythmias require medication or ablation of conduction abnormal pathways
- Permanent damage to the lungs can occur if not repaired early
- There are life-long complications
- With good care and follow up a normal life is possible

Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

Coarctation of the Aorta-**Critical CHD**

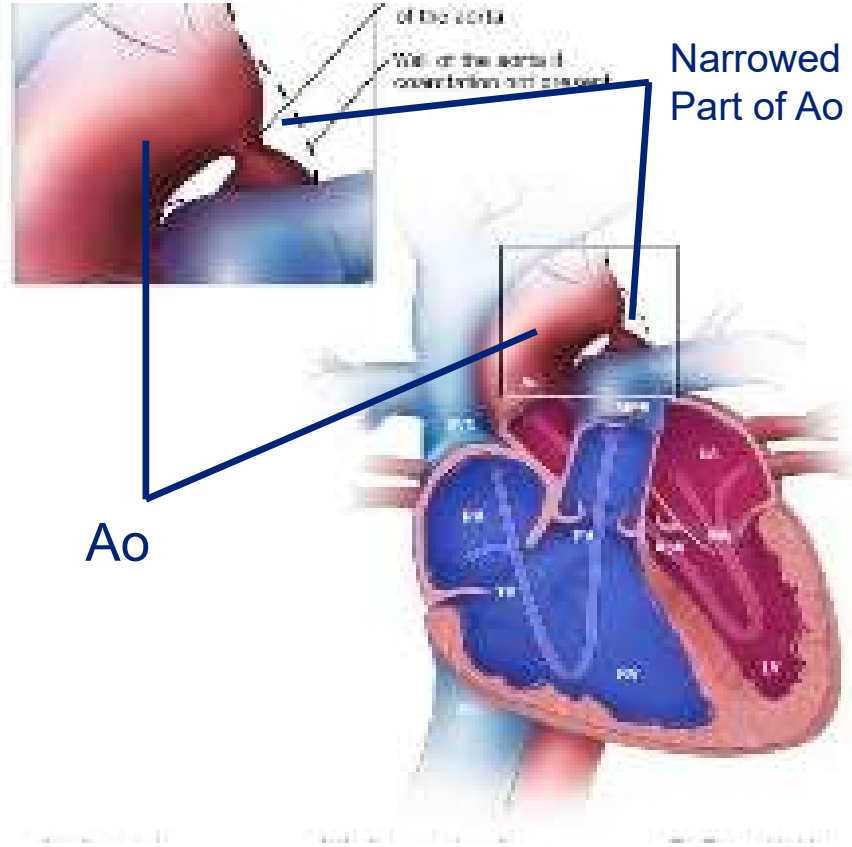
- Part of the aorta has a narrow segment
- With severe narrowing, emergency surgical repair may be necessary after birth
- Blocks normal blood flow to the body
- Causes high blood pressure to the head and low blood pressure to the abdomen and legs
- May have other associated defects

Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

Coarctation of the Aorta-Critical CHD

- Part of the spontaneously AORTA (Ao) is narrow



Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

Coarctation of the Aorta-**Critical CHD**

- The narrowed part must be widened
  - -Surgical repair
    - •Removal of the narrow segment
    - •Reconstruction/patching of the AORTA
  - -Catheterization and stretching the narrow part from the inside with a balloon devise (Balloon Angioplasty)
    - •A stent is put in place to keep the dilated segment open

Congenital Heart Disease
A Brief Review
Types of CHD

Acyanotic Heart Defects

Coarctation of the Aorta-**Critical CHD**

- Even with surgery, hypertension may persist
- Medication is needed for the hypertension and arrhythmia
- Cardiologist follow up is necessary long term

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Cyanosis means a blue color to the skin
- It means that the oxygen level in the blood is low
- There are several potential causes
  - -Congenital Heart Disease with abnormal blood flow to the lungs is our focus
  - -Other causes include poisoning and lung disease
- Giving 100% oxygen does not always correct the problem

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease
How Babies Present at Birth

- Problems breathing
- Ashen or bluish skin color
- Poor feeding
- Extreme sleepiness
- Pounding heart
- Weak pulse

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Transposition of the Great Arteries (TGA)

- The Great Arteries are the AORTA and the PULMONARY ARTERY
- In TGA, AORTA connected to the RIGHT VENTRICLE

  -Sends unoxygenated blood back to the body
- PULMONARY ARTERY is connected to the LEFT VENTRICLE

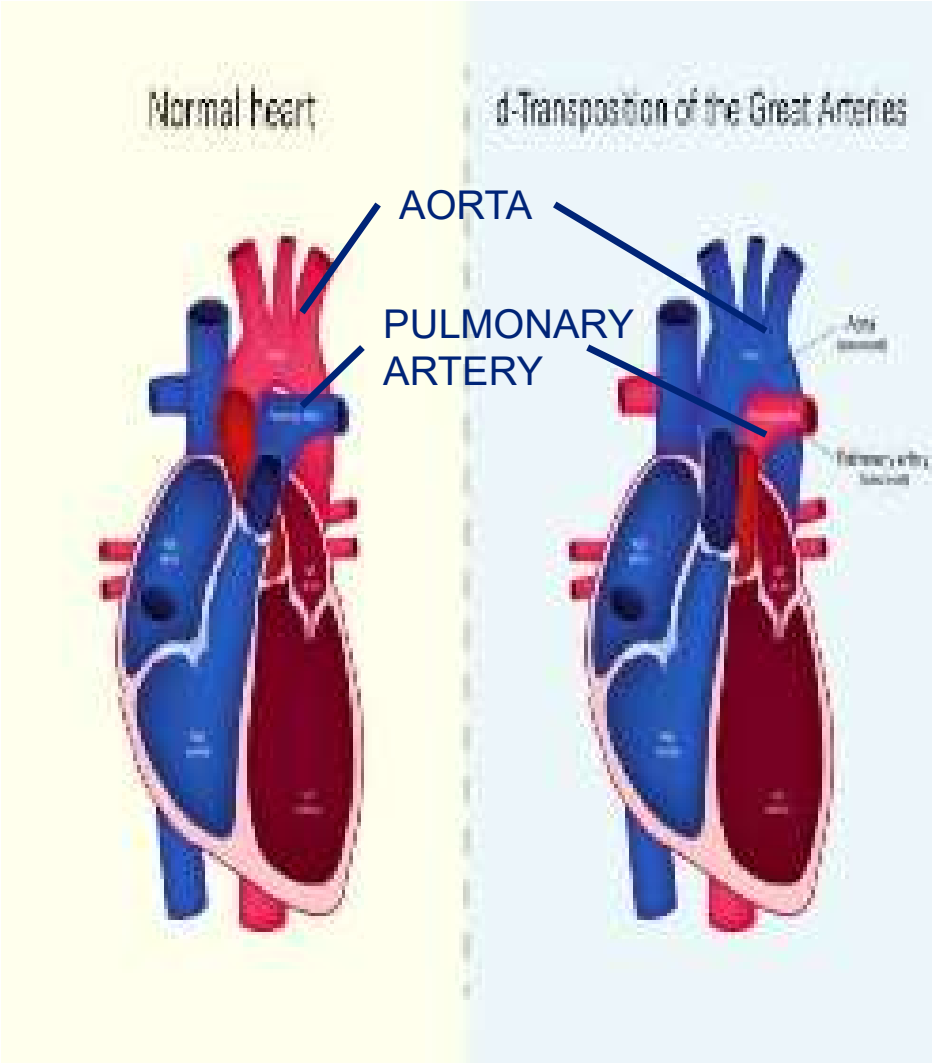
  -Sends oxygenated blood back to the lungs

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Transposition of the Great Arteries (TGA)

- Note in the TGA that the AORTA is Blue and the PULMONARY ARTERY is Red
- The AORTA comes from the RIGHT VENTRICLE
- The PULMONARY ARTERY comes from the LEFT VENTRICLE



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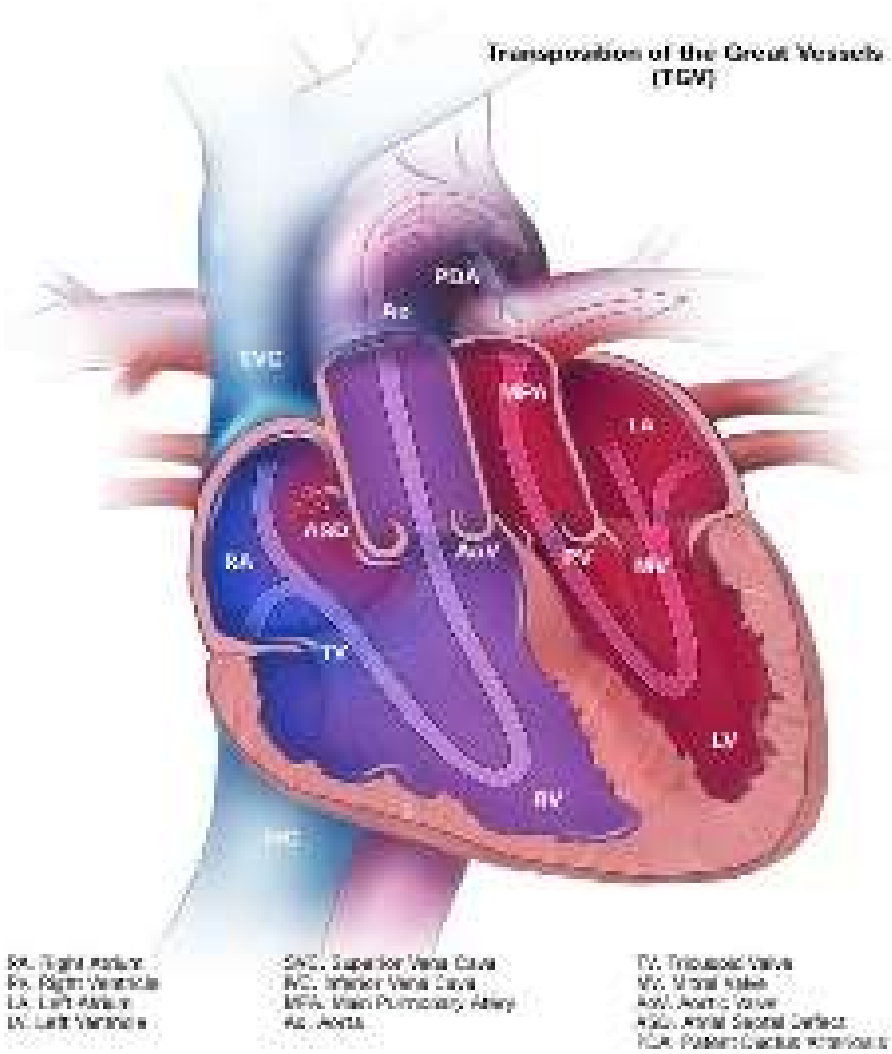
A Brief Review

Cyanotic Heart Disease

Transposition of the Great Arteries (TGA)

- The only way oxygenated blood can get to the Body is if there is mixing with the unoxygenated blood (Purple) pumped out by the RIGHT VENTRICLE to the Body



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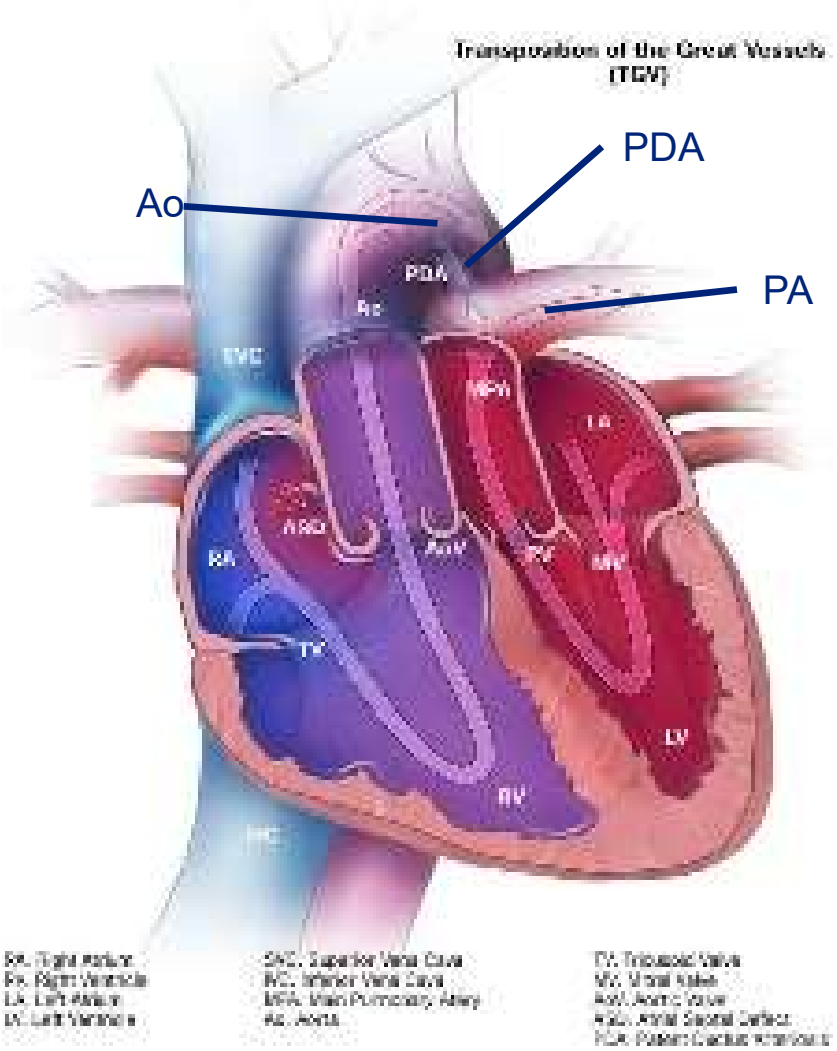
A Brief Review

Cyanotic Heart Disease

Transposition of the Great Arteries (TGA)

- The mixing only happens if there is
  - -A Patent Ductus Arteriosus (PDA)
    - Connection between the AORTA (Ao) and the PULMONARY ARTERY (PA)



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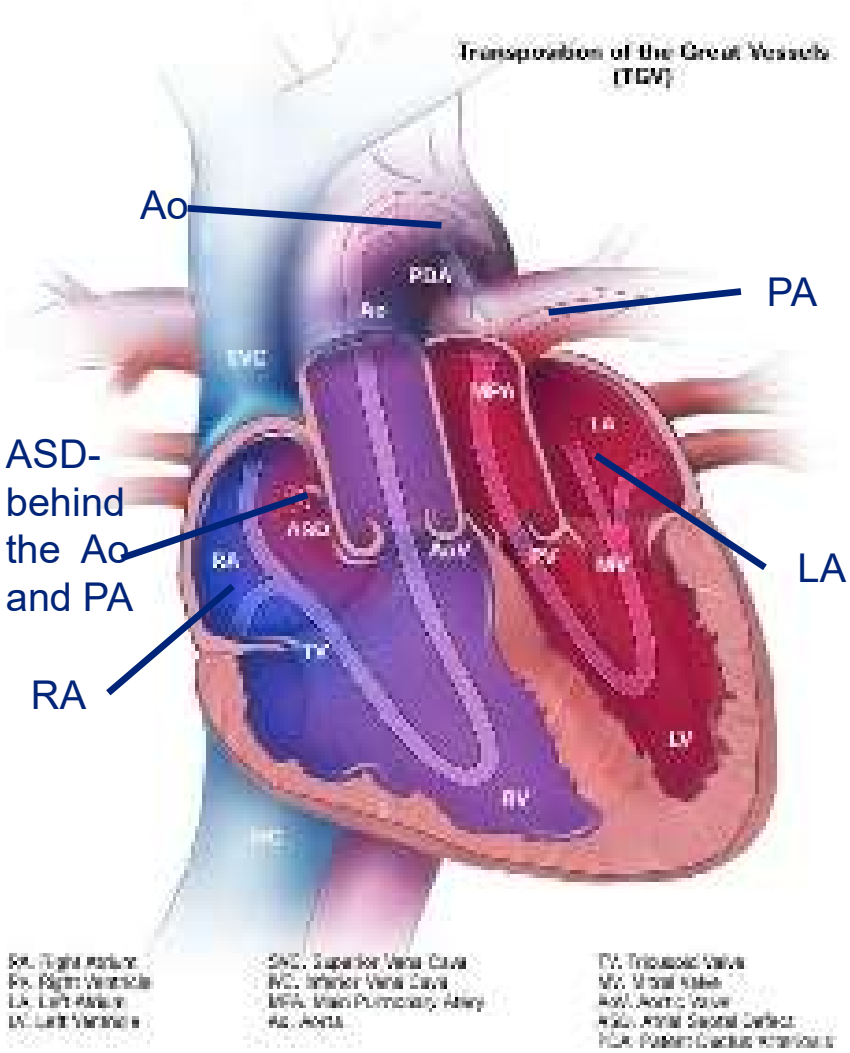
A Brief Review

Cyanotic Heart Disease

Transposition of the Great Arteries (TGA)

- The mixing only happens if there is
  - -An ATRIAL SEPTAL DEFECT (ASD)
    - Connection between the Right ATRIUM and Left ATRIUM



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Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Transposition of the Great Arteries

- Treatment is to surgically switch the great vessels to their correct position
- Initial treatment consists of keeping the PDA open
- Intravenous prostaglandin E1 (PgE1) infusion is used to keep PDA open to promote pulmonary blood flow

  -increased left atrial pressure,

  -causes left-to-right shunting and mixing at the atrial level

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Transposition of the Great Arteries

- More than 98 percent of surgically-treated infants survive their infancy.
- Most children who had TGA surgery recover and grow normally
- There are potential future risks for
  -arrhythmias,
  -leaky valves
  -other heart issues.

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Ebstein Anomaly
- Tricuspid Valve Defect
  - -rare heart problem
  - -the blood flows from the Right Ventricle backward into the right atrium
    - •heart enlargement
    - •fluid buildup in the body

Congenital Heart Disease
A Brief Review

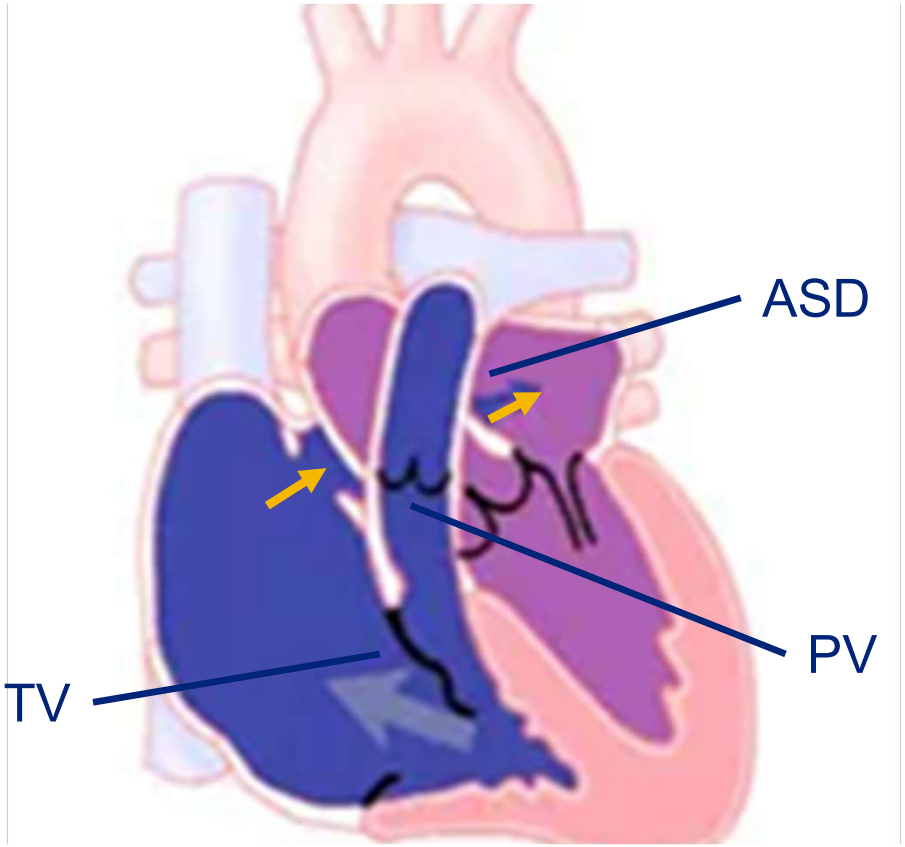
Cyanotic Heart Disease

- Ebstein Anomaly
  - -atrial septal defect
    - high pressure in the right atrium forces unoxygenated blood into the left atrium
    - Mixed blood goes to the body.

Congenital Heart Disease
A Brief Review
Cyanotic Heart Disease

Ebstein Anomaly

- Tricuspid Valve (TV)
  - In the wrong position
  - Valve's flaps (leaflets) are malformed
  - Narrow Pulmonary Valve (PV)
  - Decrease flow to the Lungs
  - Atrial Septal Defect (ASD)
  - Mixed blood (purple) goes to the body



Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Ebstein Anomaly
- Surgical repair or replacement of the tricuspid valve and closure of the atrial septal defect
- Medical therapy for heart failure or arrhythmias
- Children who have surgery do well.

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Tetralogy of Fallot

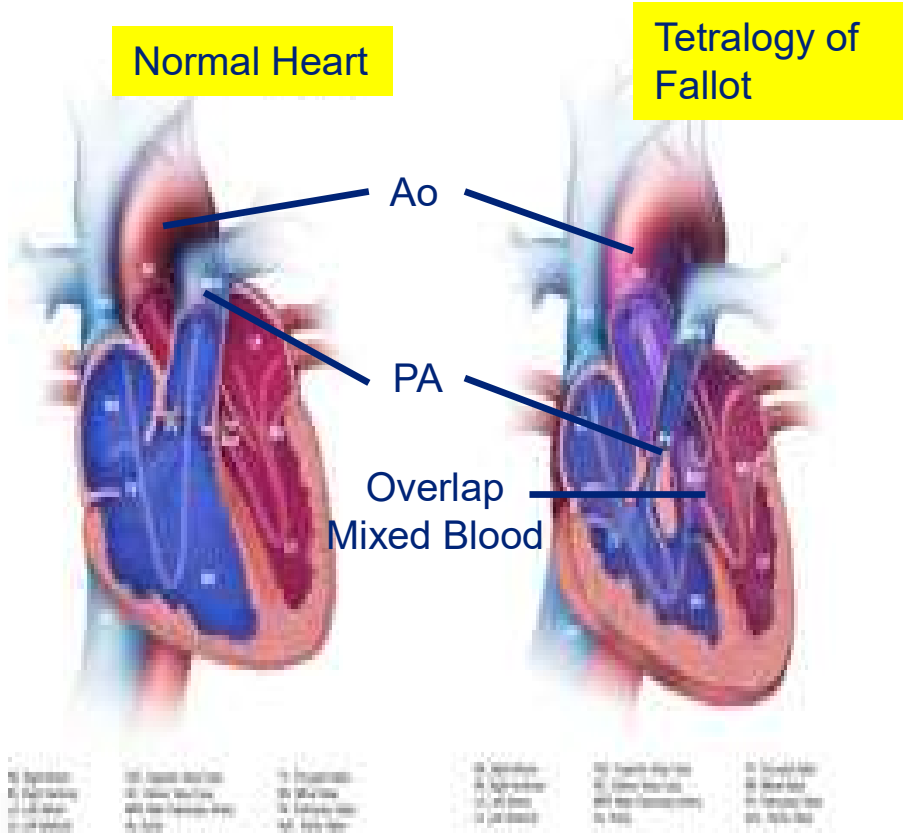
- There are four congenital heart defects found in tetralogy of Fallot
  - -Ventricular septal defect (VSD),
  - -Pulmonary stenosis,
  - -Over Riding Aorta (Misplaced aorta)
  - -Thickened right ventricular wall (right ventricular hypertrophy).

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Tetralogy of Fallot

- Mixing of oxygenated and unoxygenated blood (purple)
- The Aorta (Ao) overlaps; straddles both ventricles
- The Pulmonary Artery (PA) is small



CDC online

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Tetralogy of Fallot

- Three of the four congenital heart defects can be surgically repaired
  -Ventricular septal defect (VSD)
  -Pulmonary stenosis
  -Over riding Aorta (Misplaced aorta)
  The thickened right ventricular wall requires no surgical treatment

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Tetralogy of Fallot

- Sometimes the final surgery cannot be done right away
- In this case, a shunt is created that increases the blood flow to the lungs
- The shunt is later removed when the final repair is done
- Revision of the surgery is sometimes necessary in adolescence or early adult life

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Tetralogy of Fallot

- Most patients grow up to live normal adult lives
- The 10-, 20-, and 30-year survival rates were 95.8%, 92.7%, and 90.5%
- With the newer advances in cardiac surgery, the maximum age of survival is not known
- The predicted age currently is 50 years or older

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Three Related Defects are:

- Pulmonary Atresia
- Pulmonary Stenosis
- Tricuspid Atresia
- The common factor is that the blood flow to the lungs is impaired or prevented

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Three Related Defects are:

- Pulmonary Atresia
- Pulmonary Stenosis
- Tricuspid Atresia
- For a fetus to survive, there must be a way for the blood to get to the Lungs

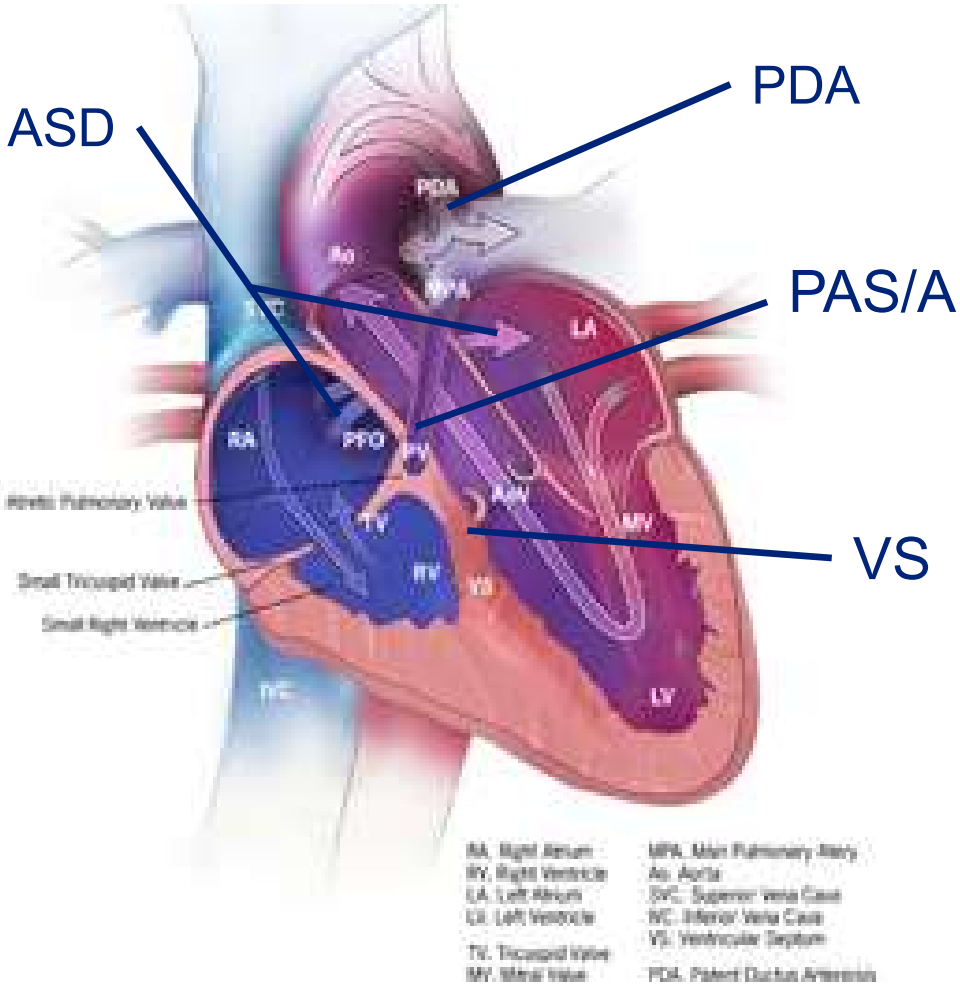
Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Pulmonary Atresia
- Pulmonary Stenosis
- Tricuspid Atresia
- Oxygenated blood and unoxygenated blood must mix
- This requires either
  - -Atrial Septal Defect
  - -Ventricular Septal Defect
  - -Patent Ductus Arteriosus

Congenital Heart Disease
A Brief Review
Cyanotic Heart Disease

- Pulmonary Atresia
- Pulmonary Artery Stenosis/Atresia (PAS/A) with INTACT Ventricular Septum (VS)
- Mixing of blood (purple) through the Ductus Arteriosus (PDA) or Atrial Septal Defect (ASD)



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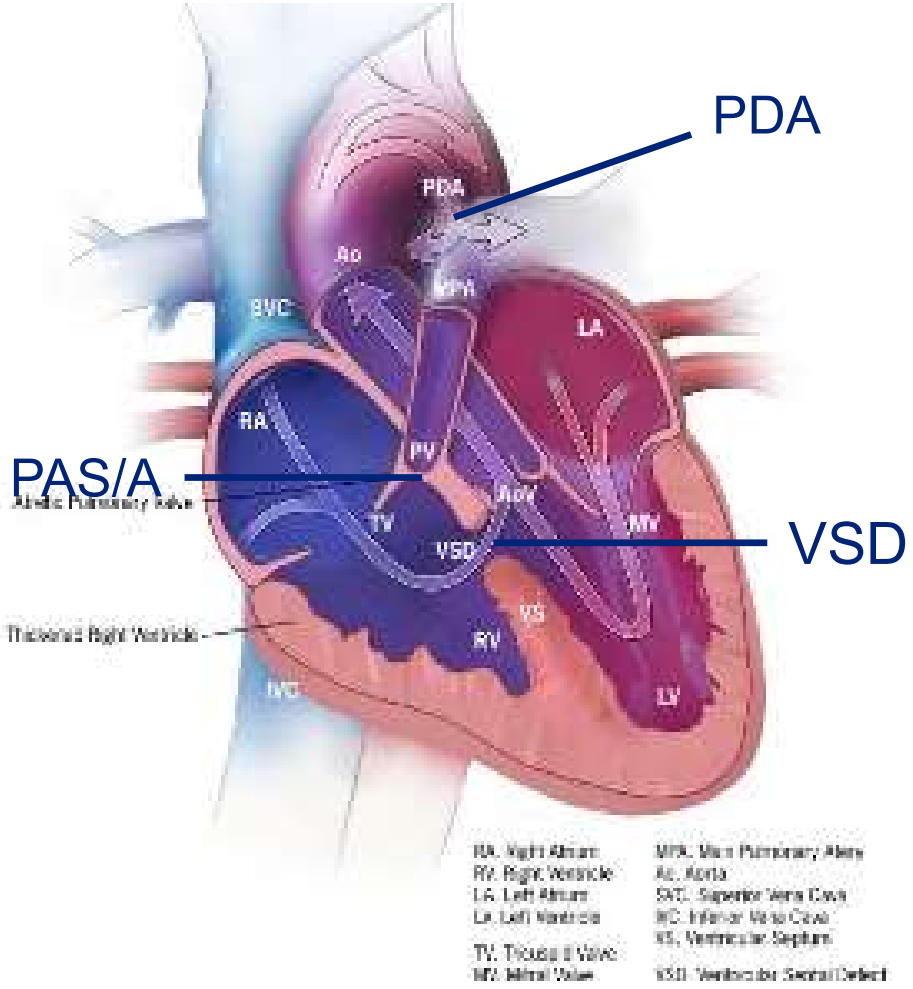
Congenital Heart Disease
A Brief Review
Cyanotic Heart Disease

- Pulmonary Artery Stenosis/Atresia (PAS/A) with VENTRICULAR SEPTAL DEFECT (VSD)
- Mixing of blood (purple) through the Ductus Arteriosus (PDA) and Ventricular Septal Defect (VSD)



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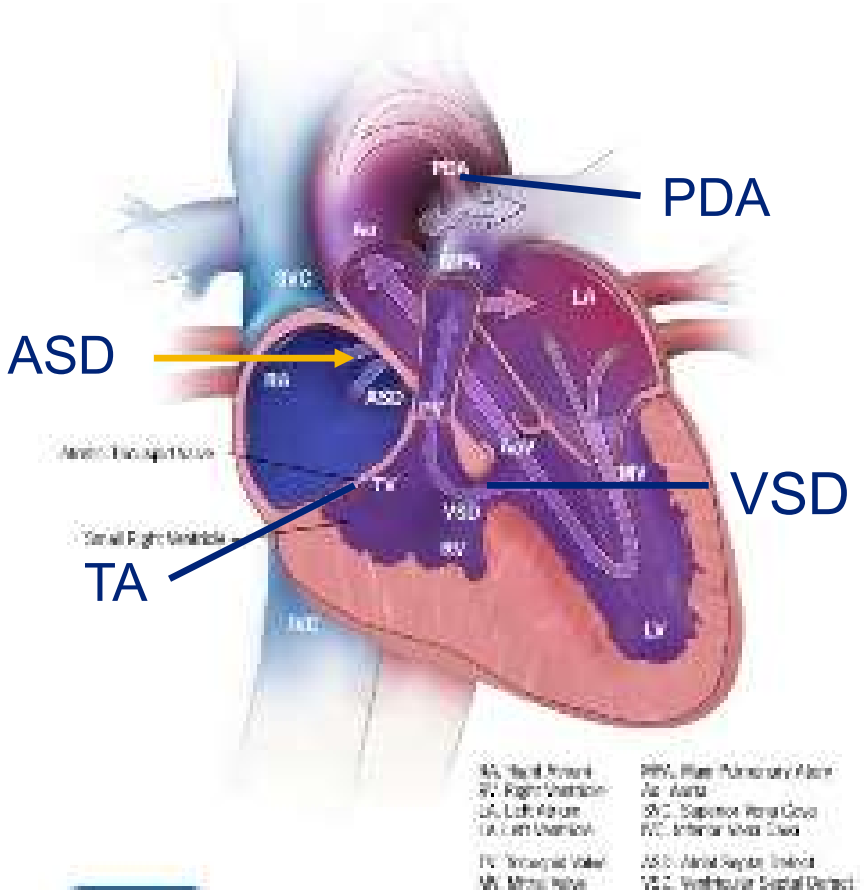
Congenital Heart Disease
A Brief Review
Cyanotic Heart Disease

- Tricuspid Atresia (TA)
- Mixing of blood (purple) through the Ductus Arteriosus (PDA), Atrial Septal Defect (ASD), and Ventricular Septal Defect (VSD)



CDC

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Tetralogy of Fallot, Pulmonary Atresia, Pulmonary Stenosis, and Tricuspid Atresia

- Similar problems with blood flow in the heart
  - Getting the unoxygenated blood to the Lungs
- The surgical treatments will be tailored to the specific combination of defects
- Tricuspid Atresia is the most difficult to treat

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Tetralogy of Fallot, Pulmonary Atresia, Pulmonary Stenosis, and Tricuspid Atresia

- To achieve a surgical correction different types of interventions are used

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Tetralogy of Fallot, Pulmonary Atresia, Pulmonary Stenosis, and Tricuspid Atresia

- Interventions:
  - Septostomy: making a hole between 2 heart chambers to allow blood to get to the lungs
  - Banding: putting a restrictive ring around a blood vessel; prevents excess blood flow from doing damage to an organ; e.g., the lungs

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Tetralogy of Fallot, Pulmonary Atresia, Pulmonary Stenosis, and Tricuspid Atresia

- Interventions:
  - Shunt procedures: making a connection in the heart so blood can get to the desired location
    - Examples of shunt procedures are:
      - Glen Procedure
      - Fontan Procedure

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Tetralogy of Fallot, Pulmonary Atresia, Pulmonary Stenosis, and Tricuspid Atresia

- Interventions:
  - Infants who have these surgeries are not cured
  - They are much improved
  - Require continued medical supervision
  - May require additional surgery as they mature
  - Some may require a heart transplant

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

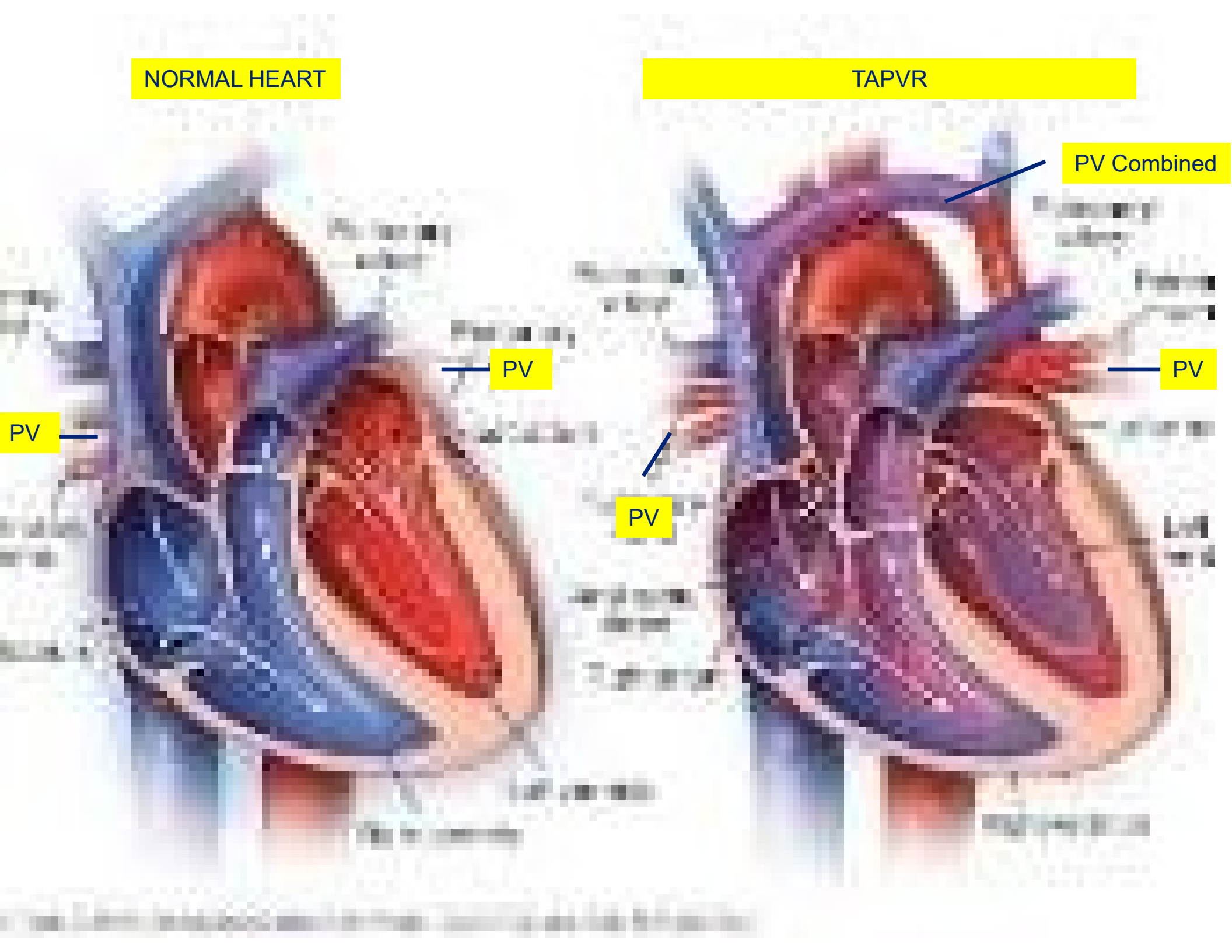
- Three heart defects
  - -Total Anomalous Pulmonary Venous Return
  - -Truncus Arteriosus
  - -Heterotaxia Syndromes

  While none of the heart defects in this presentation occur with high frequency, these are among the ones that are less rare

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Total Anomalous Pulmonary Venous Return (TAPVR)
  - -oxygen-rich blood does not return from the lungs to the left atrium
  - -the oxygen-rich blood returns to the right side of the heart.
  - -oxygen-rich blood mixes with oxygen-poor blood


NORMAL HEART
TAPVR
PV Combined
PV
PV
PV
PV

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Total Anomalous Pulmonary Venous Return (TAPVR)
  - -Babies with TAPVR usually have a hole between the right atrium and the left atrium (an Atrial Septal Defect-ASD)
  - -Allows the mixed blood to get to the left side of the heart to be pumped out the body

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Total Anomalous Pulmonary Venous Return (TAPVR)
  - -Some children can have other heart defects along with TAPVR
  - -Requires a difficult surgical intervention
  - -TAPVR surgery does not always have a good outcome

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Truncus Arteriosus
  - The Aorta and the Pulmonary Artery coming out of the heart remain connected as a single blood vessel
  - There is usually a hole between the bottom two chambers of the heart (ventricles)
    - Ventricular Septal Defect (VSD)

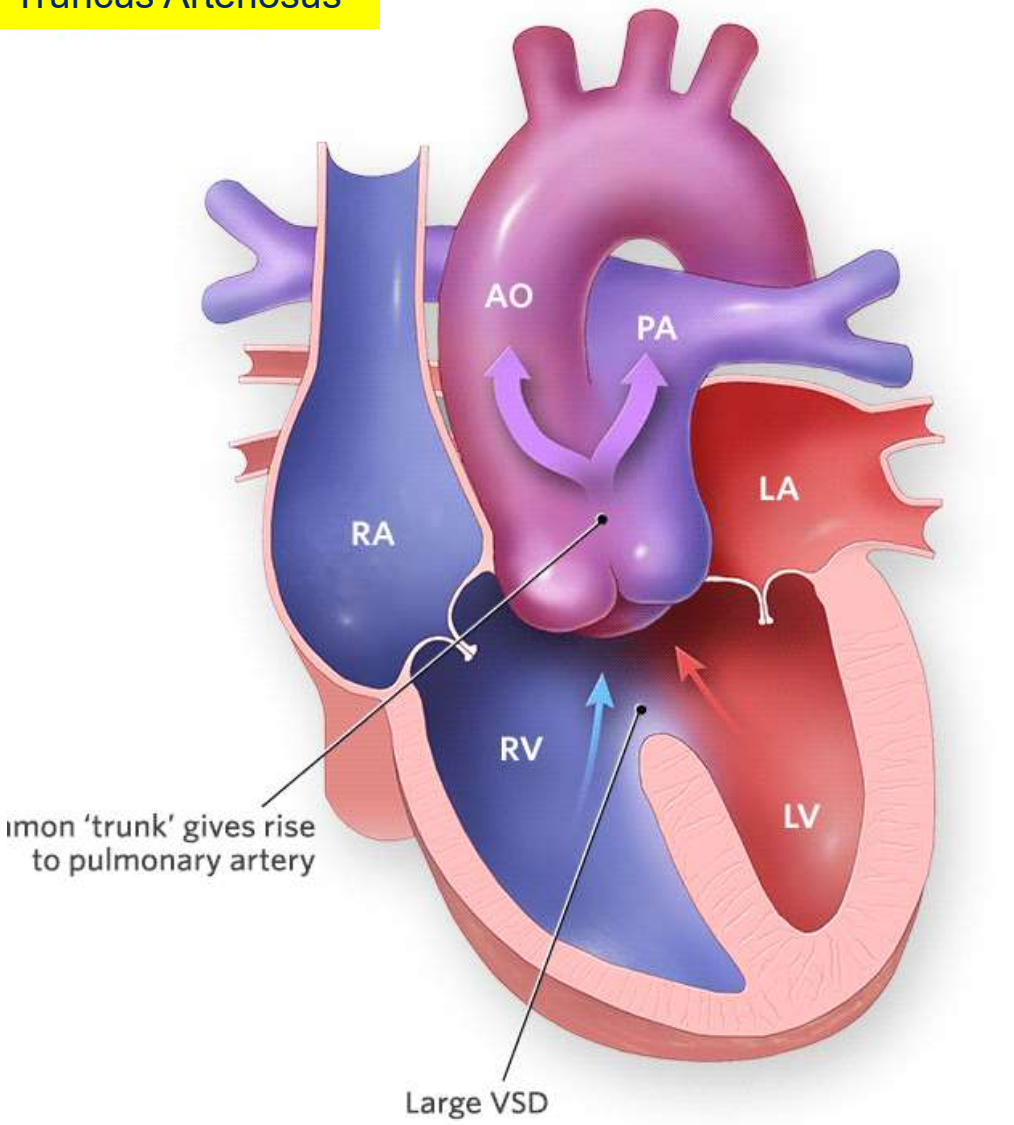
Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

Truncus Arteriosus

- Aorta and the Pulmonary Artery coming out of the heart remain connected as a single vessel
- Ventricular Septal Defect (VSD)

Truncus Arteriosus



Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Truncus Arteriosus
  - -Oxygen-poor blood and oxygen-rich blood are mixed
  - -Both flow into the Body and the Lungs
  - -Because of pressure differences between the lungs (low pressure) and the body (higher pressure), the lungs get more blood than normal

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Truncus Arteriosus
  - -The surgical repair is complex
  - -Artificial conduits may be necessary
  - -Artificial valves may be needed
  - -Additional surgeries may be needed as the child grows
  - -Other health problems may occur such as chronic lung disease

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Heterotaxia Syndrome (Isomerism)
  - Heart and other organs are in the wrong place in the chest and abdomen
    - The heart is rotated 180 deg. side to side
  - Commonly affects the lungs, liver, spleen, stomach and intestines
  - The spleen may be missing or exist as several mini spleens
  - The displacements can be a mirror image to the left or to the right
  - This is a rare condition

Congenital Heart Disease
A Brief Review

Cyanotic Heart Disease

- Heterotaxia Syndrome (Isomerism)
  - The heart surgery is tailored to the specific defects
  - Other organs may need surgery also
  - Conduction abnormalities of the heart may occur leading to arrhythmias
  - Neurodevelopmental problems may occur

Congenital Heart Disease
A Brief Review

Mortality and Survival

Most Recent Study:

Lesion Specific CHD Mortality Trends

Pediatrics, October 2022

Review of the time period 1999 to 2017 in the United States

Congenital Heart Disease
A Brief Review

Mortality and Survival

Infant Mortality

- 5.6% of all deaths had CHD as the cause
- In an additional 3%, CHD were contributing
  - Other diagnoses include prematurity, genetic, non-cardiac birth defects
- 23% deaths were from Single Ventricle Syndromes including Hypoplastic Left Heart and Truncus Arteriosus
- Trends were generally improving except for Total Anomalous Pulmonary Venous Return

Congenital Heart Disease
A Brief Review

Mortality and Survival

Child Mortality 1 to 4 years of age

- 4.3% of all deaths had CHD as the cause
- In an additional 1.5%, CHD were contributing
  - Other diagnoses include genetic and non-cardiac birth defects
- Single ventricle lesions remained the leading cause of death
  - This group had increases in the mortality rates

Congenital Heart Disease
A Brief Review

Mortality and Survival

Child Mortality 5 to 17 years of age

- 1.6% of all deaths had CHD as the cause
- In an additional 0.4%, CHD were contributing
  - Other diagnoses include genetic and non-cardiac birth defects
- Single ventricle lesions again were the leading causes of death

Congenital Heart Disease
A Brief Review

Mortality and Survival

- CHD mortality rates continue to decrease across infancy, childhood, and adolescence
- This is for most CHDs
- Single Ventricle defects have the highest mortality rates
- Based upon these statistics, most infants and children with CHDs will survive into adult life

Congenital Heart Disease
A Brief Review

Congenital Heart Disease in Adults

- Advances in diagnostic methods and in the surgical and medical care of infants and children with congenital heart disease have resulted in very high success rates for healthy survival
- In the United States, there are presently between 500,000 and 600,000 adults with congenital heart disease.
- Over 20,000 heart operations for congenital heart disease are performed in the United States each year

Congenital Heart Disease
A Brief Review

Congenital Heart Disease in Adults

- 25,000 infants currently born with congenital malformations of the heart and circulation
- Over 85% will reach adulthood
- Congenital heart disease in adults is still a largely unrecognized subspecialty but is emerging as a discipline that requires special expertise

Congenital Heart Disease
A Brief Review

Congenital Heart Disease in Adults

- Uninterrupted, long-term care is essential if this population is to be properly managed
- As managed care professionals
  - We must be aware of the infants and children who need our assistance
  - We must seize the opportunity to assist those who have transitioned to adult life

Congenital Heart Disease
A Brief Review

Conclusion

- You have now been exposed to some of the information you will need to assist your families
- Understanding some of the physiology related to Congenital Heart Defects enables you to better anticipate patient needs

Congenital Heart Disease
A Brief Review

Conclusion

- Knowing the terminology and some of the pathology will enable better communication with the patients, caregivers, and providers.
- Familiarity with Congenital Heart Disease enables you to consider appropriate referrals and support

You are not now Cardiologists, but you can now speak their language.

THANK YOU FOR YOUR ATTENTION

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QUESTIONS?