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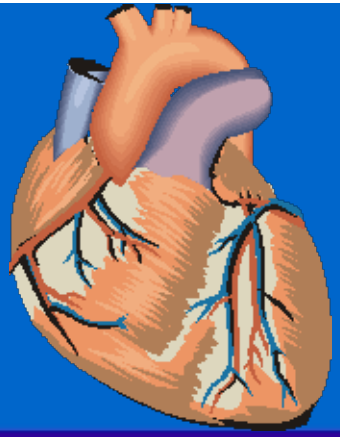
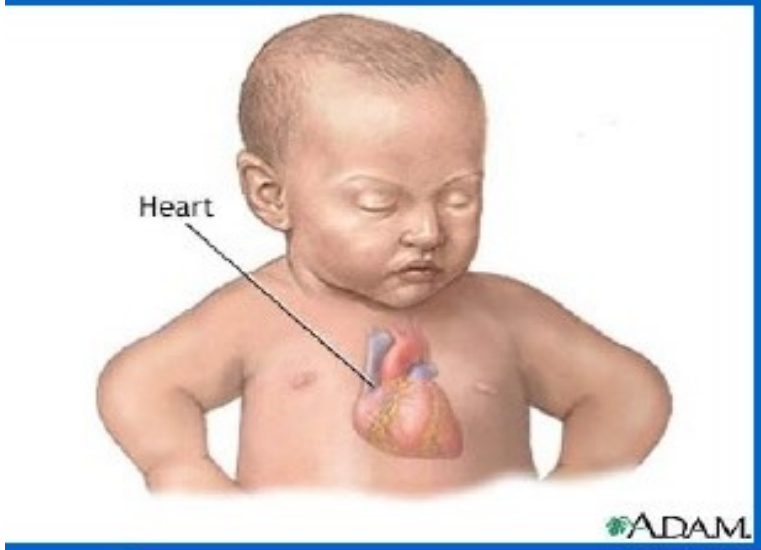
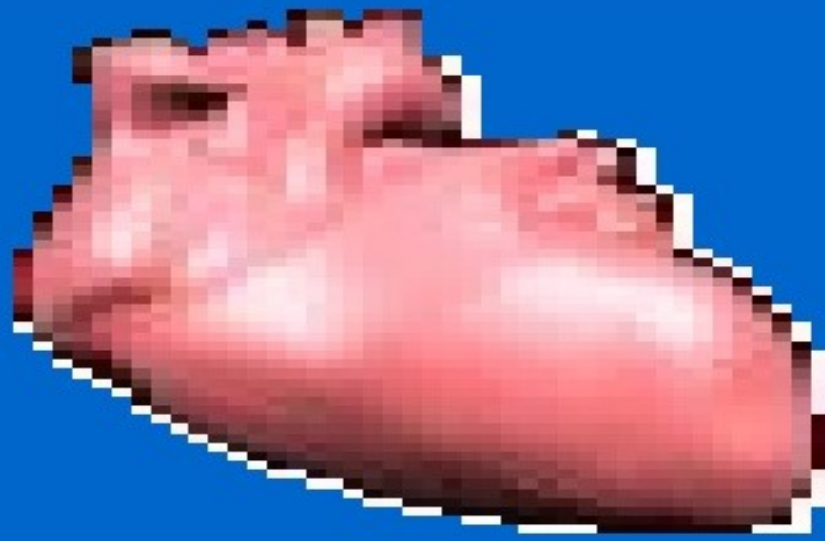
FACULTY OF NURSING SCIENCES

By- SUDHA BENJAMINI
Associate Professor
Faculty of Nursing



Congenital Heart Disease





Congenital Heart Disease

- Congenital heart disease (CHD) occurs in 1/125 live births.
- most common birth defect
- occur during the 1st 8 wks. of fetal development
- majority have no known cause

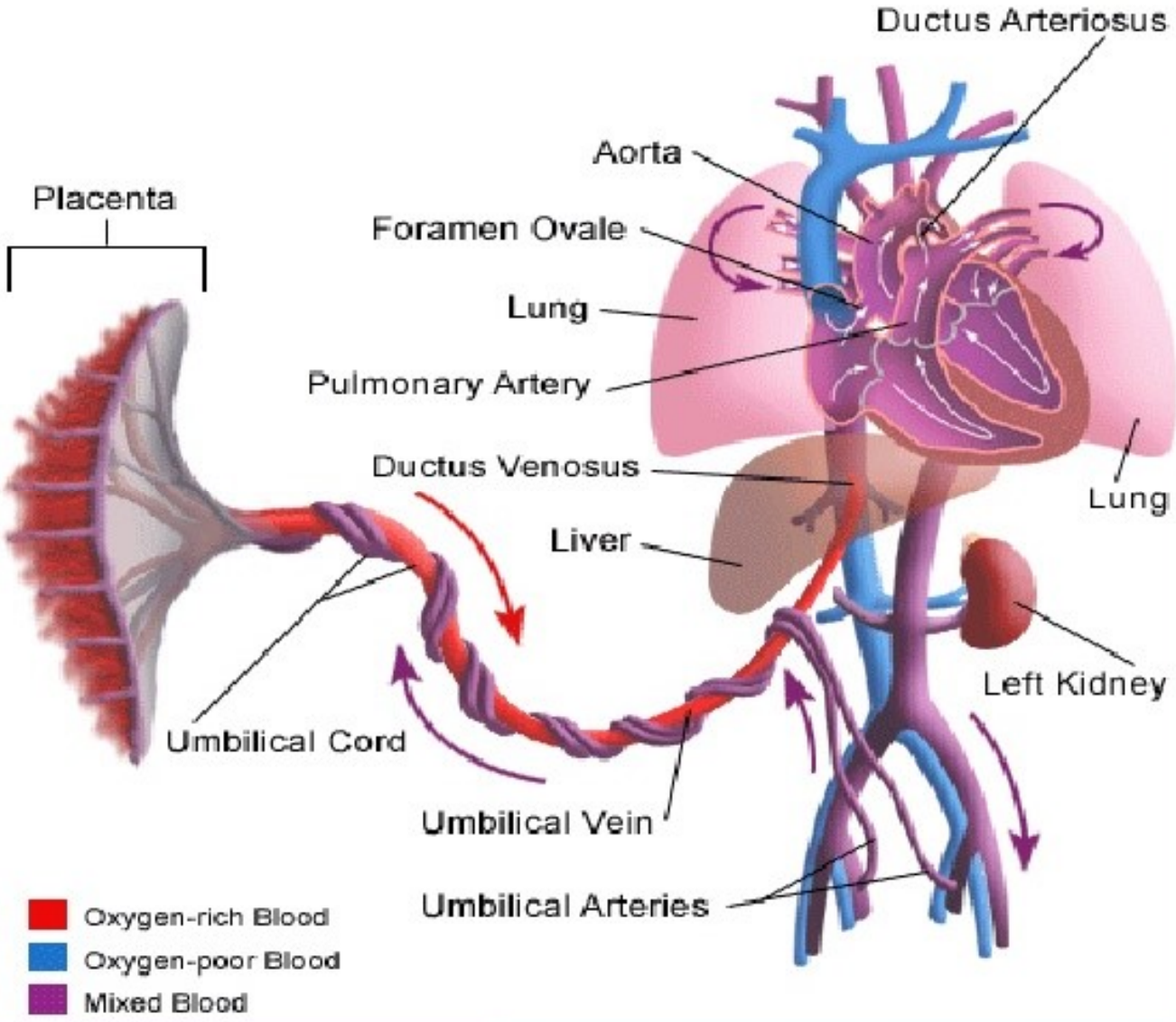
Factors Contributing to CHD

- 85 to 90 % of cases, there is no identifiable cause for the heart defect → generally considered to be caused by multifactorial inheritance.
- factors are usually both genetic and environmental, where a combination of genes from both parents, in addition to unknown environmental factors, produce the trait or condition.
- Maternal Factors:
  - seizure disorders w/ intake of anti-seizure medications
  - intake of lithium for depression
  - uncontrolled IDDM
  - lupus
  - german measles (rubella) – 1st trimester of pregnancy

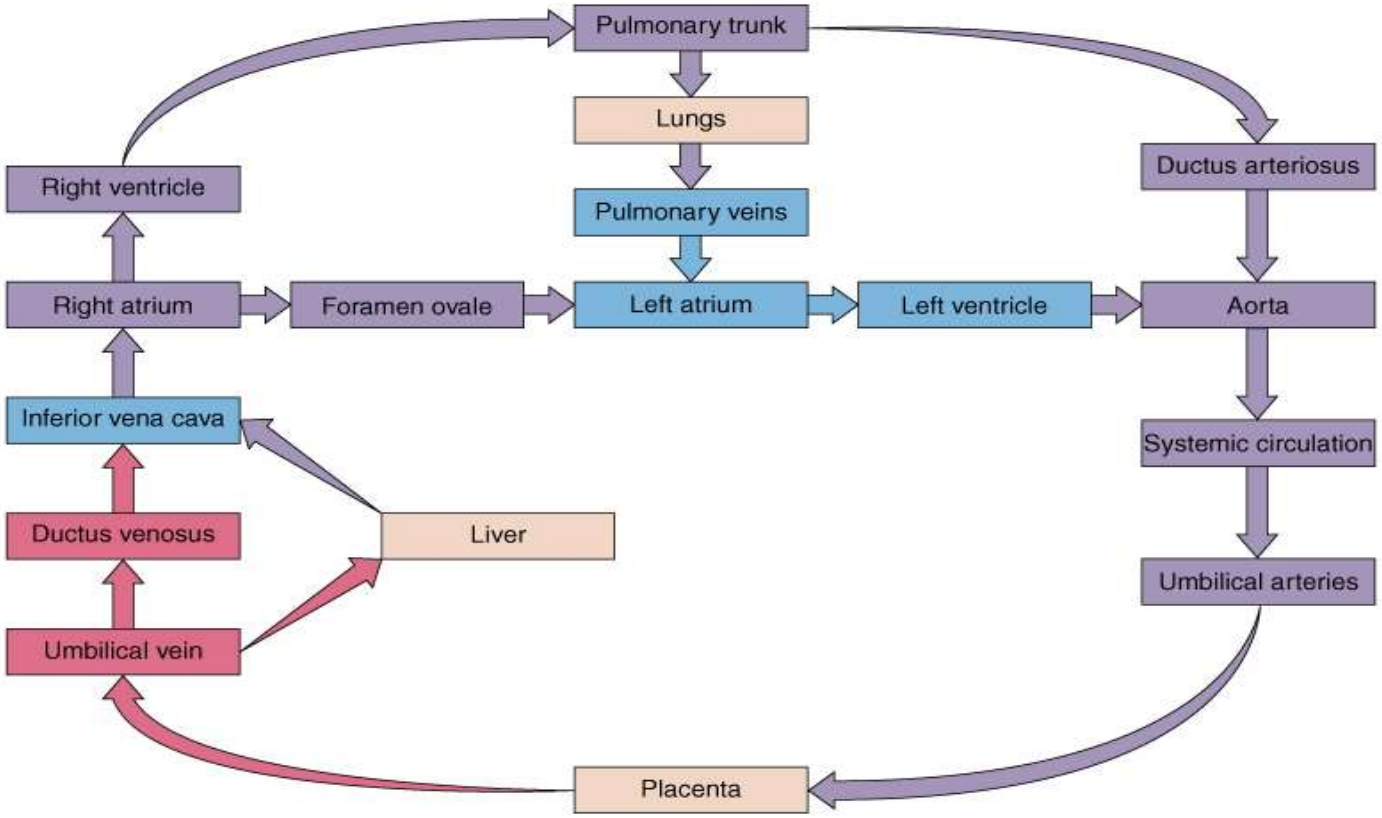
Factors Contributing to CHD

- Family History:
  - risk increases when either parent has CHD, or when another sibling was born w/ CHD
  - If you have had one child with CHD, the chance that another child will be born with CHD ranges from 1.5 to 5 %, depending on the type of CHD in the first child.
  - If you have had two children with CHD, then the risk ↑ to 5 to 10 %, to have another child with CHD.
  - If the mother has CHD, the risk for a child to be born with CHD ranges from 2.5 to 18 percent, with an average risk of 6.7 percent.
- Chromosome abnormalities:
  - 5 to 8 % of all babies with CHD have a chromosome abnormality
  - includes Down syndrome, trisomy 18 and trisomy 13, Turner's syndrome, Cri-du-chat syndrome

Fetal Circulation



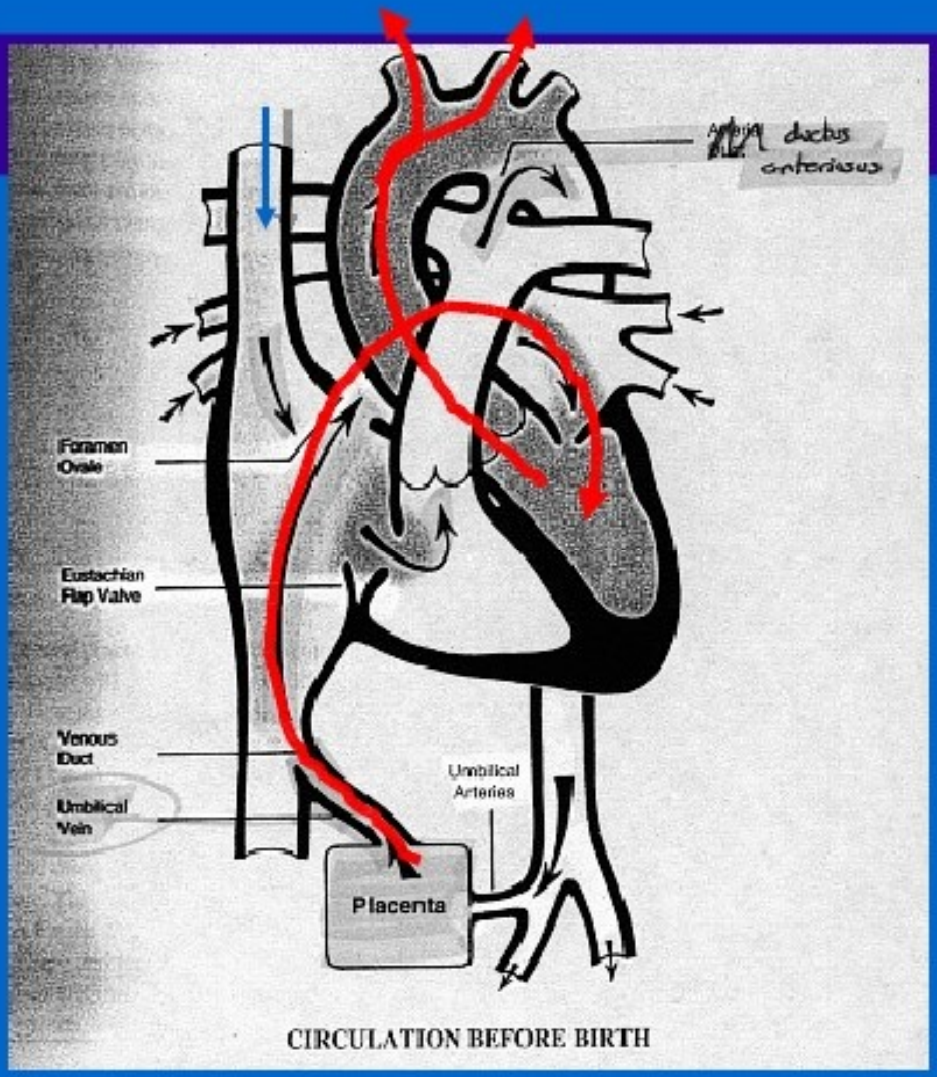
Flow Chart of Fetal Circulation



(c) Scheme of fetal circulation

Fetal circulation:

- Blood flows from the placenta
  - → IVC
  - → RA
  - → through the FO
  - → LA
  - → LV
  - → ascending aorta
  - → head & upper extremities
  - → returns via the SVC



CIRCULATION BEFORE BIRTH

Fetal circulation:

From the SVC

→ RA

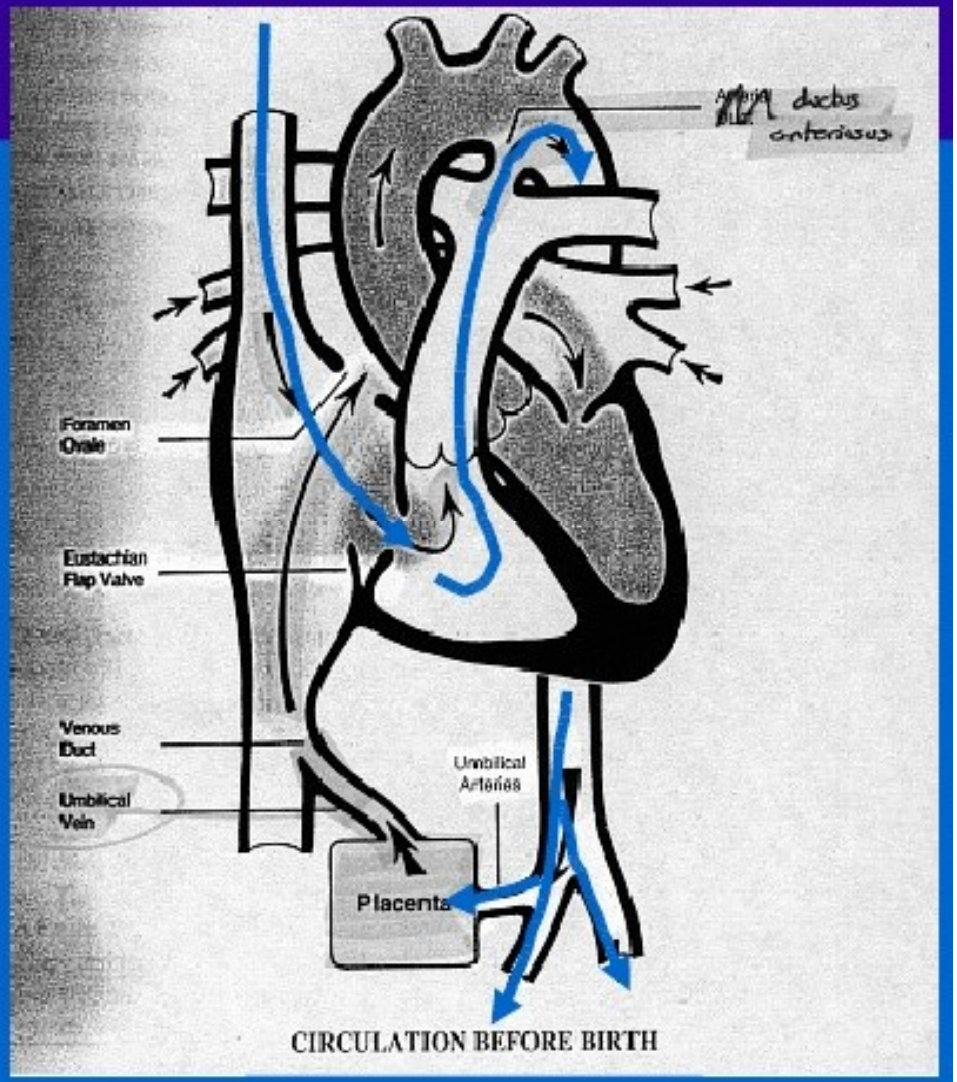
→ RV

→ pulm artery

→ through the PDA

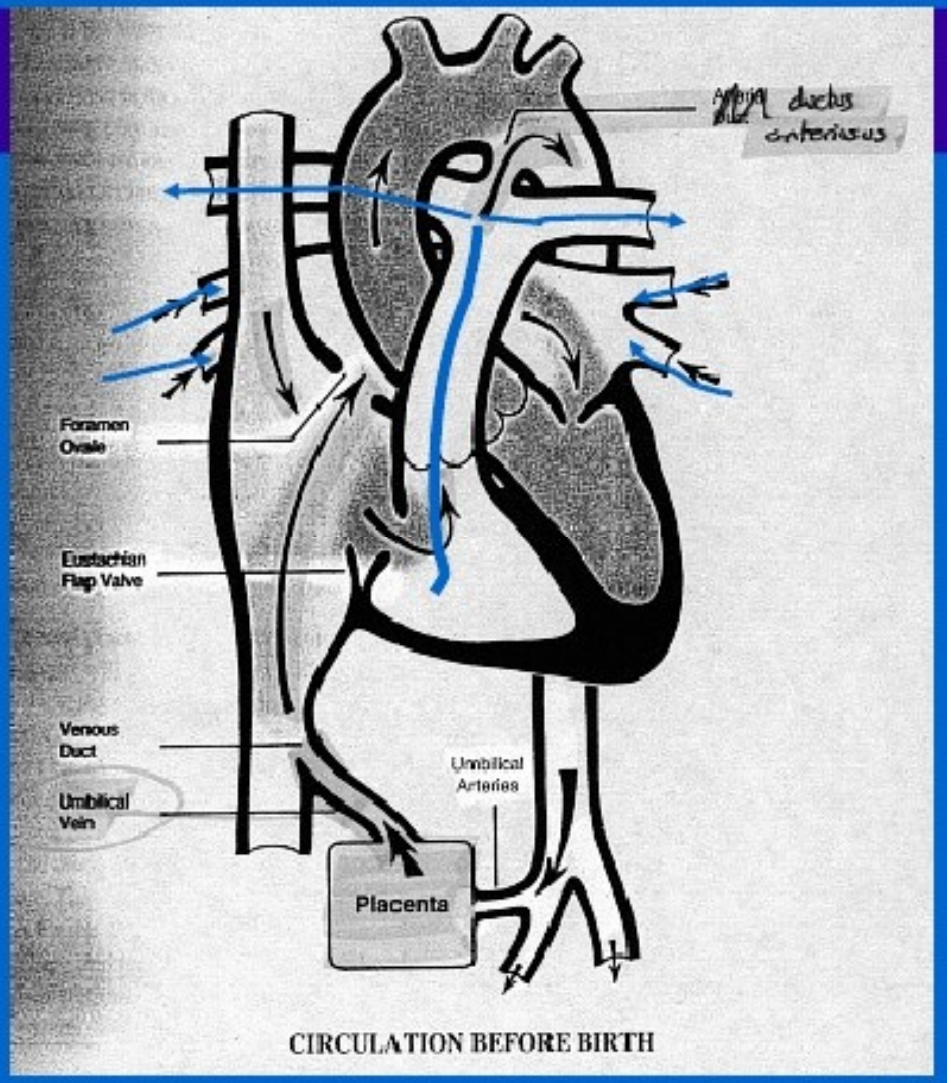
→ descending aorta

→ lower extremities and placenta



Fetal circulation:

Only a very small amount of blood is directed through the right and left pulmonary arteries to the lungs.



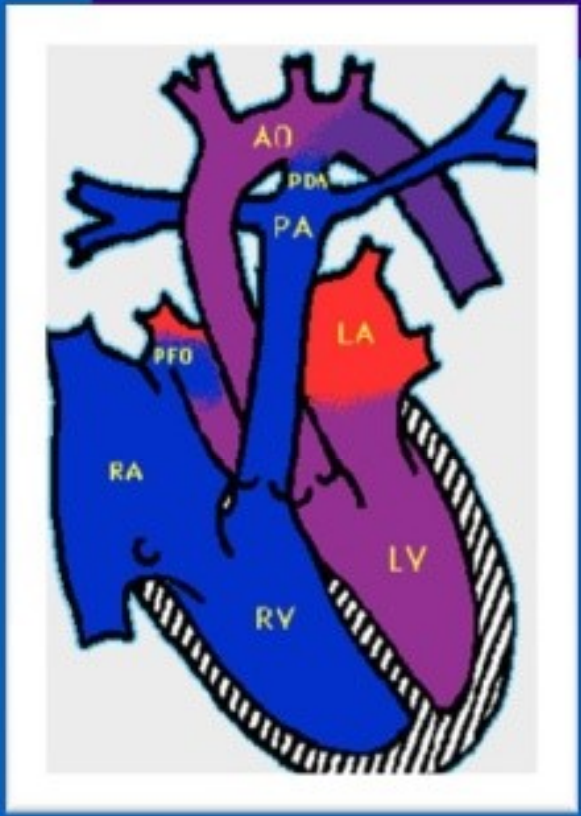
CIRCULATION BEFORE BIRTH

Fetal Structure	Adult Structure
Foramen Ovale	Fossa Ovalis
Umbilical Vein (intra-abdominal part)	Ligamentum teres
Ductus Venosus	Ligamentum venosum
Umbilical Arteries and abdominal ligaments	medial umbilical ligaments, superior vesicular artery (supplies bladder)
Ductus Arteriosum	Ligamentum arteriosum

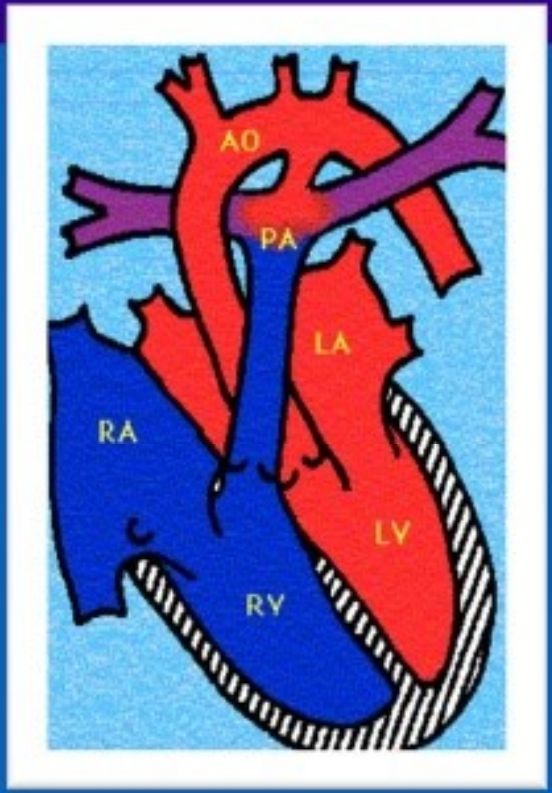
Blood circulation after birth:

The transformation from fetal to neonatal circulation involves two major changes:

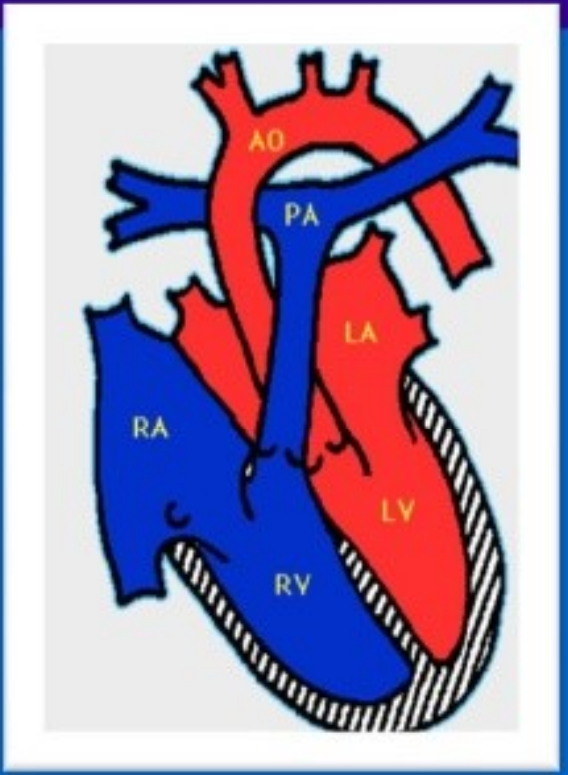
1. A marked increase in systemic resistance. caused by loss of the low-resistance placenta.
2. A marked decrease in pulmonary resistance. caused by pulmonary artery dilation with the neonate's first breaths.



Fetal Circulation



8 Hours old



24-72 hrs

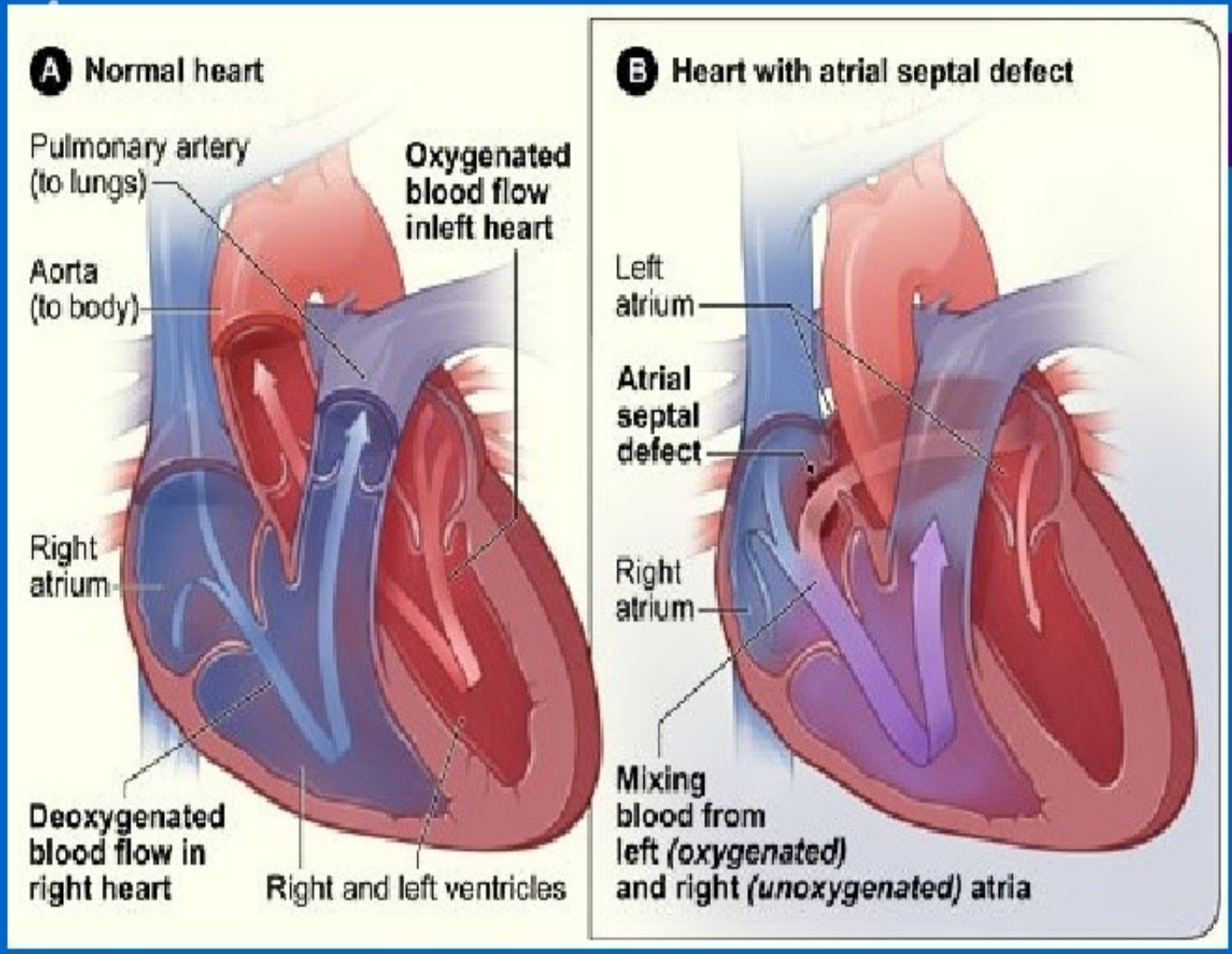
Blood circulation after birth:

- With the first breaths of air the baby takes at birth, the fetal circulation changes. A larger amount of blood is sent to the lungs to pick up oxygen.
- Because the ductus arteriosus (the normal connection between the aorta and the pulmonary valve) is no longer needed, it begins to wither and close off. (72 hrs.)
- The circulation in the lungs increases and more blood flows into the left atrium of the heart → ↑ pressure causes the foramen ovale to close and blood circulates normally

Acyanotic Congenital Heart Defects

Pink Baby (L → R shunt)

- L → R shunts cause CHF and pulmonary hypertension.
- This leads to RV enlargement, RV failure
- These babies present with CHF and respiratory distress.
- They are not typically cyanotic
- Examples: Patent Ductus Arteriosus (PDA)
  Ventricular Septal Defect (VSD)
  Atrial Septal Defect (ASD)
  Coarctation of the Aorta


A Normal heart
Pulmonary artery (to lungs)
Oxygenated blood flow inleft heart
Aorta (to body)
Right atrium
Deoxygenated blood flow in right heart
Right and left ventricles
B Heart with atrial septal defect
Left atrium
Atrial septal defect
Right atrium
Mixing blood from left (oxygenated) and right (unoxygenated) atria

Cyanotic Congenital Heart Defects

Blue Baby (R → L shunt)

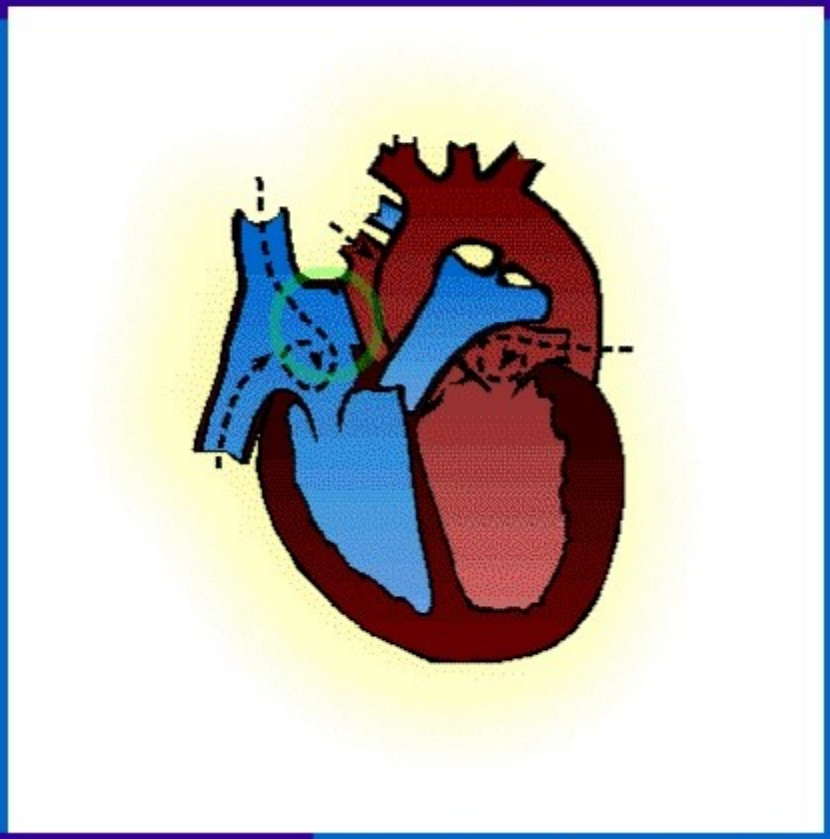
- R → L shunts cause hypoxia and central cyanosis.
- Venous blood is shunted from the R to the L side of the heart w/o passing through the lungs to be oxygenated
- Unoxygenated blood circulates in arteries → cyanosis
- Examples:
  - Tetralogy of Fallot (TOF)
  - Transposition of the Great Arteries (TGA)
  - Truncus Arteriosus (TA)

Atrial Septal Defect (ASD)

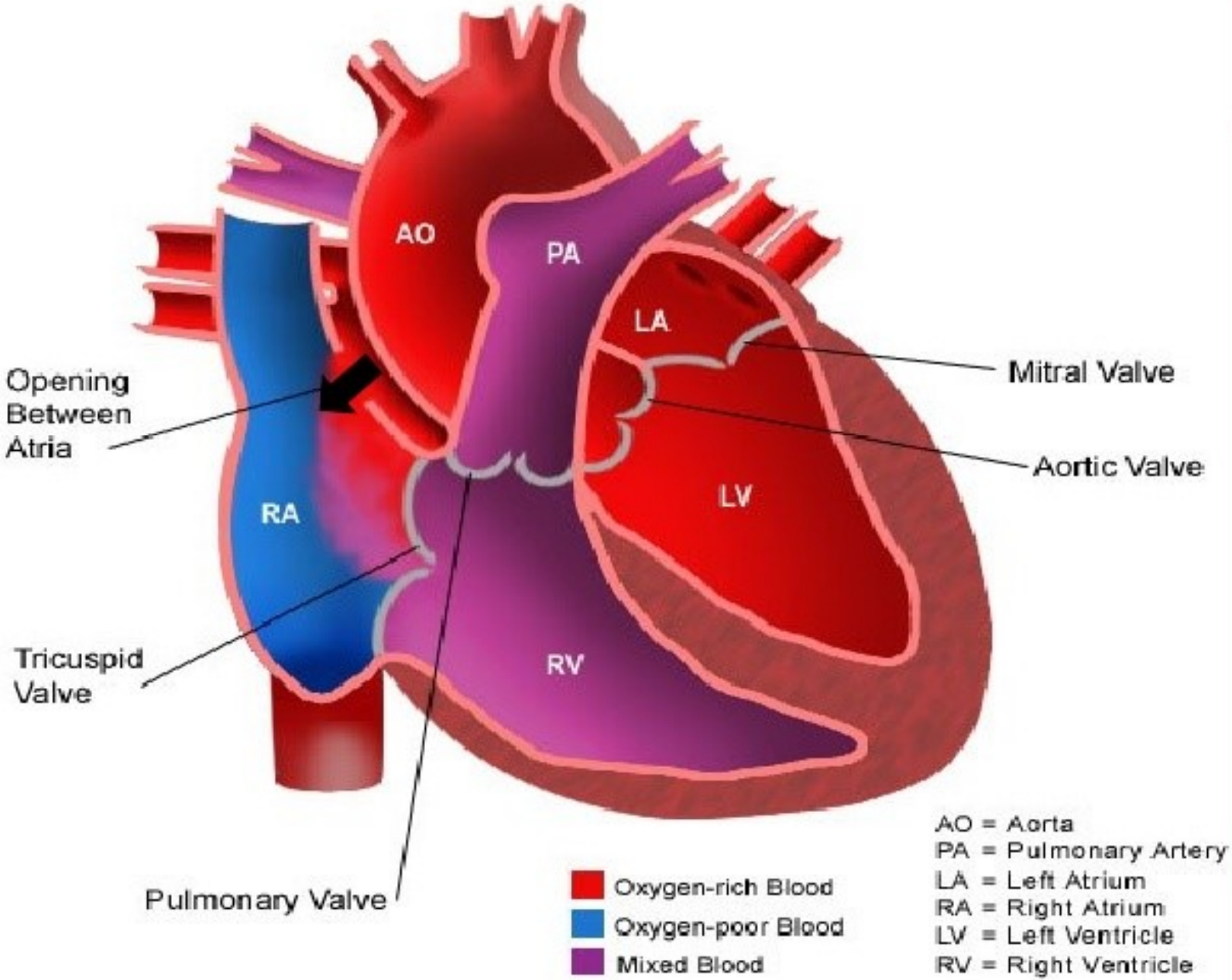
- an opening in the atrial septum
- An atrial septal defect allows oxygenated (red) blood to pass from the left atrium, through the opening in the septum, and then mix with unoxygenated (blue) blood in the right atrium
- During fetal heart devt. → the partitioning process does not occur completely, leaving an opening in the atrial septum
- occur in 4-10% of all infants w/ CHD

- EFFECTS: When blood passes through the ASD from the left atrium to the right atrium → a larger volume of blood than normal must be handled by the right side of the heart → extra blood then passes through the pulmonary artery into the lungs → pulmonary hypertension and pulmonary congestion

Atrial Septal Defect (ASD)



Atrial Septal Defect (ASD)



Atrial Septal Defect (ASD)

Signs and Symptoms

- child tires easily when playing
- infant tires easily when feeding
- fatigue
- sweating
- tachypnea, tachycardia
- shortness of breath, crackles
- poor growth
- murmur

Diagnostic tests:

- CXR – enlarged heart
- ECG
- 2D echo – show pattern of blood flow through the septal

Atrial Septal Defect (ASD)

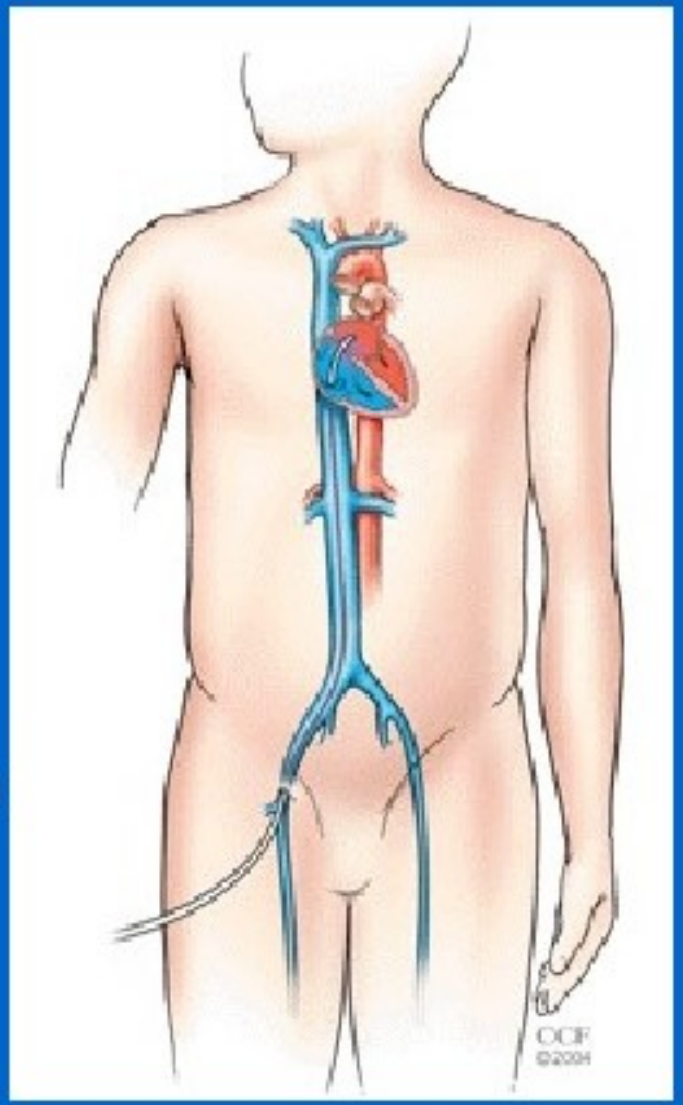
- 20% of atrial septal defects will close spontaneously in the first year of life.
- atrial septal defects may close spontaneously as a child grows
- Usually, an ASD will be repaired if it has not closed on its own by the time the child starts school - to prevent lung problems that will develop from long-time exposure to extra blood flow
- pulmonary arteries become thickened and obstructed due to increased flow, from left to right for many years (pulmonary vascular obstructive disease)

Treatment

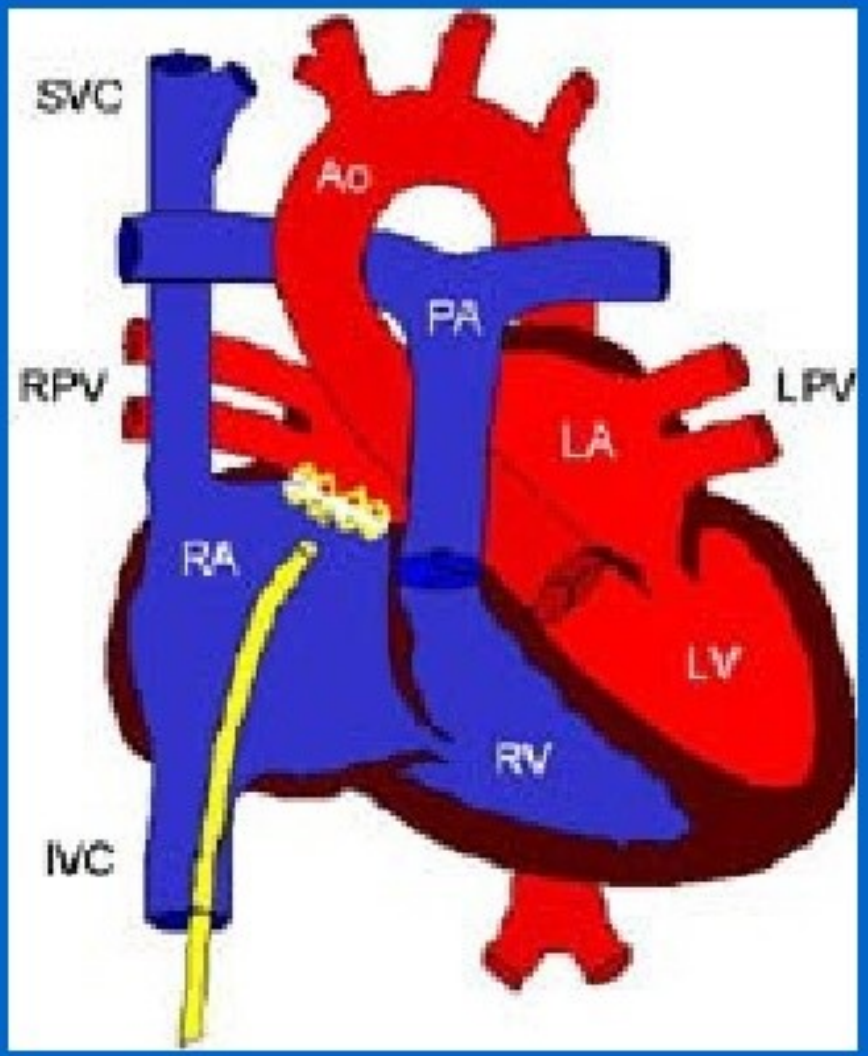
- medical management
  - digoxin - helps strengthen the heart muscle, enabling it to pump more efficiently.
  - diuretics – relieve pulmonary congestion
- infection control
  - prophylactic antibiotics to prevent bacterial endocarditis before dental procedures and other invasive procedures
- surgical repair
  - the patient is placed on cardiopulmonary bypass (the heart-lung machine), the right atrium is then opened to allow access to the atrial septum below
  - defect may be closed with stitches or a special patch.
  - the material utilized for patch closure of ASD's may be the

Treatment

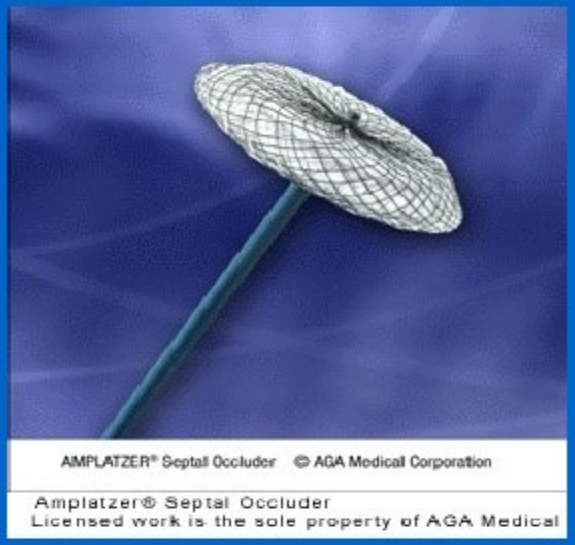
- Transcatheter management
- This technique involves implantation of one of several devices (basically single or double wire frames covered by fabric) using cardiac catheterization
- cardiac catheterization - involves slowly moving a catheter (a long, thin, flexible, hollow tube) into the heart. The catheter is initially inserted into a large vein through a small incision made usually in the inner thigh (groin area) and then is advanced into the heart



- An ASD closure device is moved through the catheter to the heart and specifically to the location of the heart wall defect
- Within a few days, the body's own tissue will begin to grow over the device. By 3 to 6 months, the device is completely covered by heart tissue and at that point becomes a part of the wall of the patient's heart.



Example of Closure Device



Amplatzer Septal Occluder

Example of Closure Device

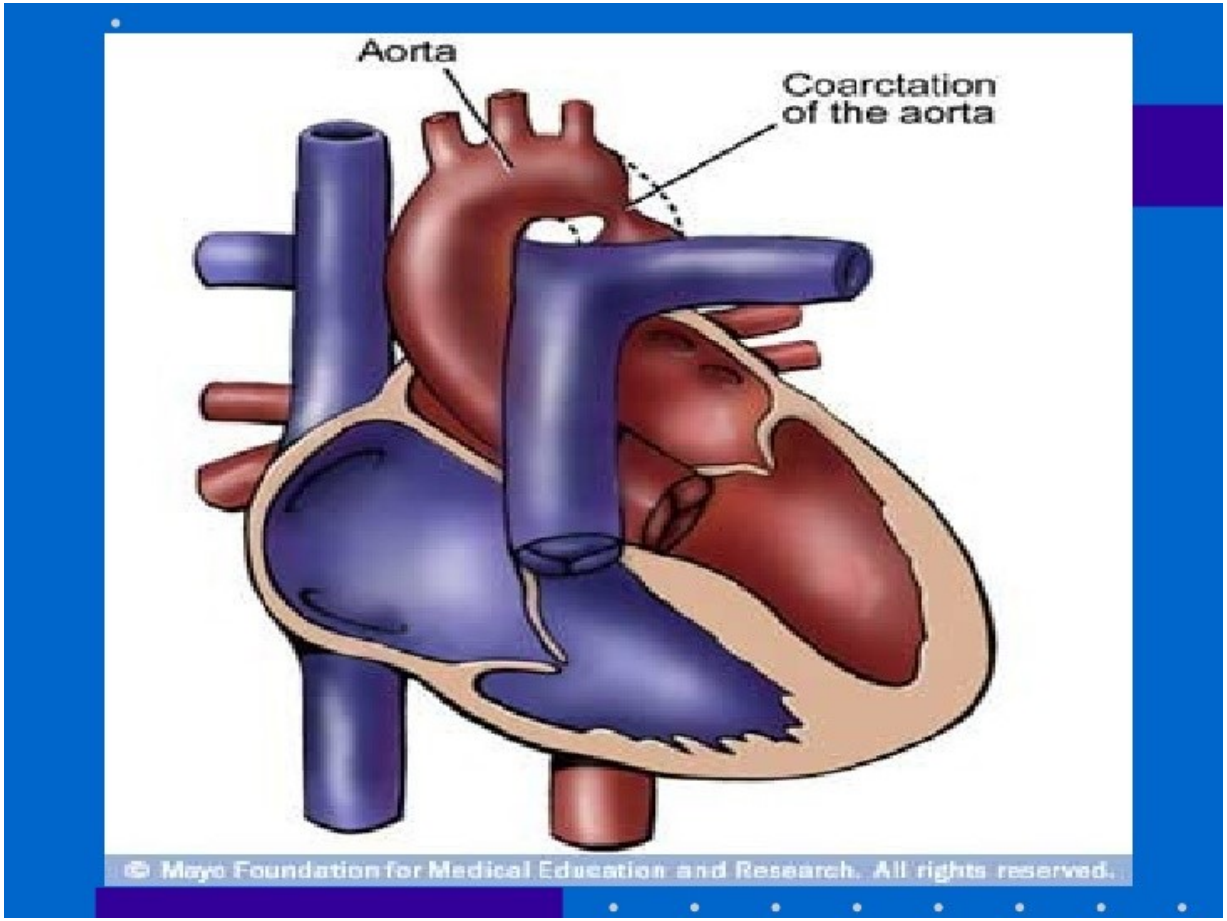
CardioSEAL Occluder

Coarctation of the Aorta

- Narrowing of the aorta
- can occur anywhere, but is most likely to happen in the segment just after the aortic arch. This narrowing restricts the amount of blood to the lower part of the body
- occurs in about 8-11 % of all children with CHD

EFFECTS:

- The left ventricle has to work harder to try to move blood through the narrowing in the aorta → left-sided heart failure
- BP is higher above the narrowing, and lower below the narrowing. Older children may have headaches from too much pressure in the vessels in the head, or cramps in the legs or abdomen from too little blood flow in that region.
- The walls of the arteries may become weakened by high pressure → spontaneous tears → cause a stroke or uncontrollable bleeding.


Aorta
Coarctation of the aorta
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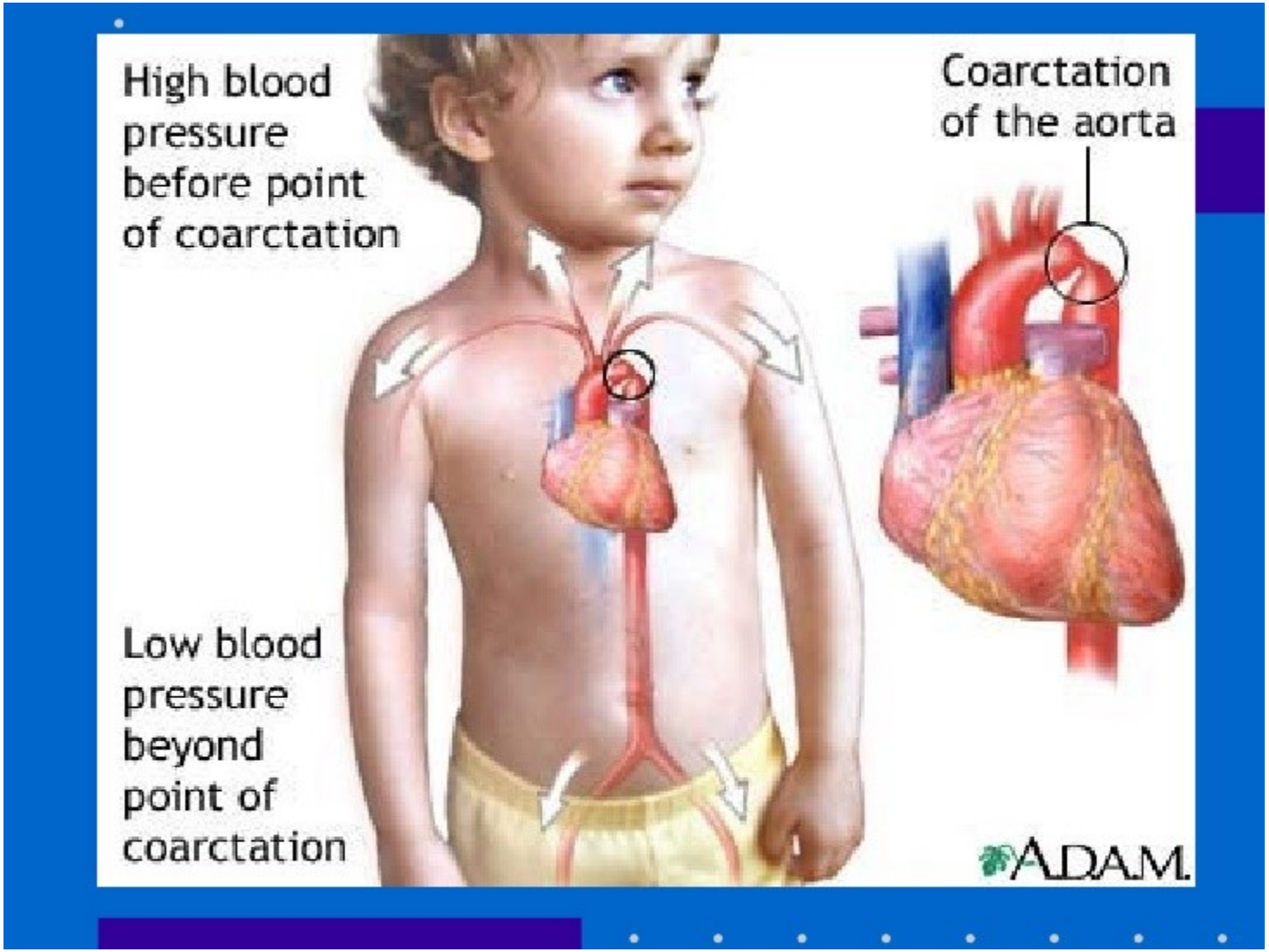

High blood pressure before point of coarctation
Coarctation of the aorta
Low blood pressure beyond point of coarctation
A.D.A.M.

Signs and Symptoms

- irritability
- pale skin
- sweating
- heavy and/or rapid breathing
- poor feeding
- poor weight gain
- cold feet and/or legs
- diminished or absent pulses in the feet
- BP in the arms significantly greater than the BP in the legs
- Mild narrowing may not cause symptoms at all. Often, a school-aged child or adolescent is simply noted to have high BP or a heart murmur on a physical examination. Some may complain of headaches or cramps in the lower sections of the body.

Treatment

- interventional cardiac catheterization

  - During the procedure, the child is sedated and a small, thin, flexible tube (catheter) is inserted into a blood vessel in the groin and guided to the inside of the heart

  - once the catheter is in the heart, the cardiologist will pass an inflated balloon through the narrowed section of the aorta to stretch the area open.

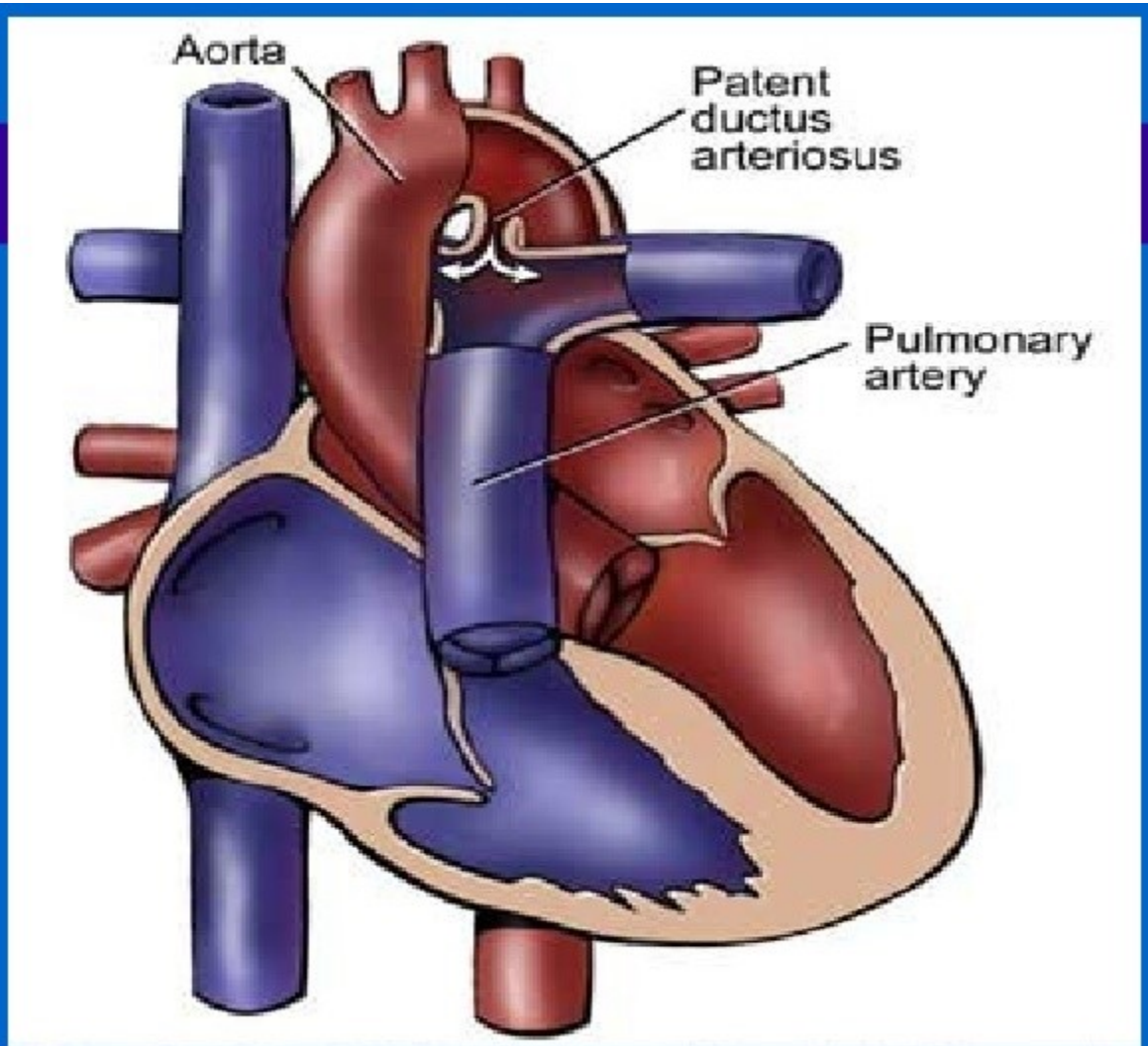
  - A small device, called a stent, may also be placed in the narrowed area after the balloon dilation to keep the aorta open.

- surgical repair

  Your child's coarctation of the aorta may be repaired surgically in an operating room. The surgical repair is performed under GA. The narrowed area is either surgically removed, or made larger with the help of surrounding structures or a patch.

Patent Ductus Arteriosus (PDA)

- characterized by a connection between the aorta and the pulmonary artery
- All babies are born with a ductus arteriosus.
- As the baby takes the first breath, the blood vessels in the lungs open up, and blood begins to flow → the ductus arteriosus is not needed to bypass the lungs
- Most babies have a closed ductus arteriosus by 72 hours after birth.
- In some babies, however, the ductus arteriosus remains open (patent) .
- The opening between the aorta and the pulmonary artery allows oxygenated blood to pass back through the blood vessels in the lungs.



Patent Ductus Arteriosus (PDA)

- In many children, there is no known reason for the ductus arteriosus remaining open. However, PDA is seen more often in the following:
- premature infants
- infants born to a mother who had rubella during the first trimester of pregnancy

EFFECTS:

- PDA → oxygenated blood passes from the aorta to the pulmonary artery & mixes w/ the unoxygenated blood w/c goes to the lungs → ↑ blood volume to the lungs → pulmonary hypertension & congestion
- Further, because blood is pumped at high pressure through the PDA, the lining of the pulmonary artery will become irritated and inflamed. Bacteria in the bloodstream can easily

Signs and Symptoms

- fatigue
- sweating
- tachypnea
- shortness of breath
- congested breathing
- disinterest in feeding, or tiring while feeding
- poor weight gain
- murmur
- increase systolic BP
- bounding pulse

Treatment

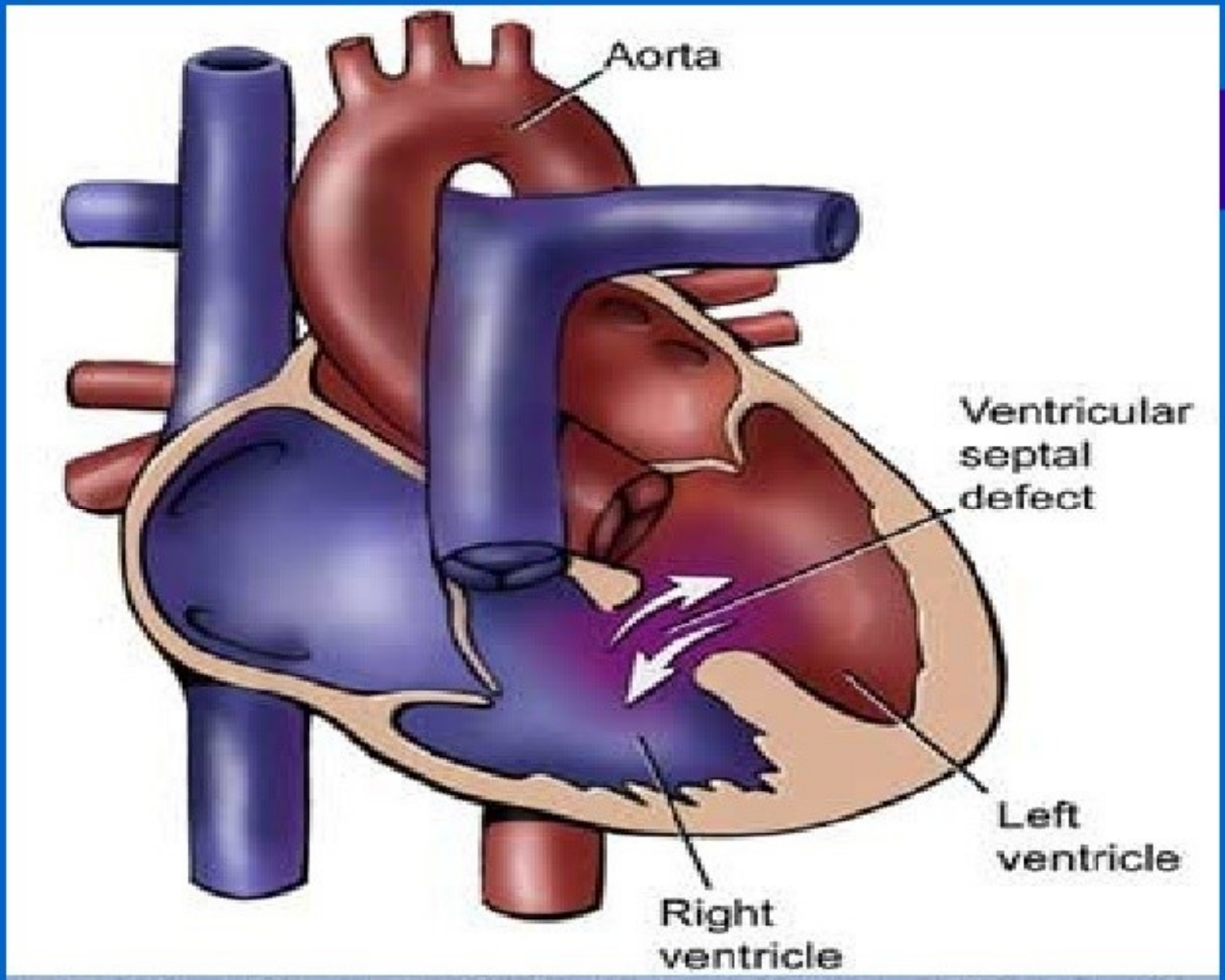
- Medical Management
  - Indomethacin IV (prostaglandin inhibitor) may help close a PDA.
    - works by stimulating the muscles inside the PDA to constrict, thereby closing the connection
  - Digoxin
  - Diuretics
- adequate nutrition
  (premature infants or those infants with a large PDA may become tired when feeding, and are not able to eat enough to gain weight)
  - high-calorie formula or breast milk
    Special nutritional supplements may be added to formula or pumped breast milk that increase the number of calories in each ounce, thereby allowing your baby to drink less and still consume

Treatment

- supplemental tube feedings
  - infants who can drink part of their bottle, but not all, may be fed the remainder through the feeding tube
  - infants who are too tired to bottle-feed may receive their formula or breast milk through the feeding tube alone.
- PDA surgical repair or closure
  - Repair is usually indicated in infants younger than 6 months of age who have large defects that are causing symptoms, such as poor weight gain and rapid breathing
  - Transcatheter coil closure of the PDA
  - PDA ligation
    - involves closing the open PDA with stitches or the

Ventricular Septal Defect (VSD)

- an opening in the ventricular septum
- allows oxygenated blood to pass from the left ventricle, through the opening in the septum, and then mix with unoxygenated blood in the right ventricle.
- VSDs are the most commonly occurring type of congenital heart defect, occurring in 14-17 % of babies born each year.
- occur when the partitioning process does not occur completely, leaving an opening in the ventricular septum.



Ventricular Septal Defect (VSD)

- EFFECTS:
- When blood passes through the VSD from the left ventricle to the right ventricle → a larger volume of blood than normal must be handled by the right side of the heart → extra blood then passes through the pulmonary artery into the lungs → pulmonary hypertension and pulmonary congestion → pulmonary arteries become thickened and obstructed due to increased pressure
- If VSD is not repaired, and lung disease begins to occur → pressure in the right side of the heart will eventually exceed pressure in the left → R to L shunt → cyanosis
- Due to high pressure --- tissue damage may eventually occur in the right ventricle → bacteria in the bloodstream can

Signs and Symptoms

- fatigue
- sweating
- tachypnea
- murmur
- heavy breathing
- congested breathing
- disinterest in feeding, or tiring while feeding
- poor weight gain
- The larger the opening, the greater the amount of blood that passes through and overloads the right ventricle and lungs.

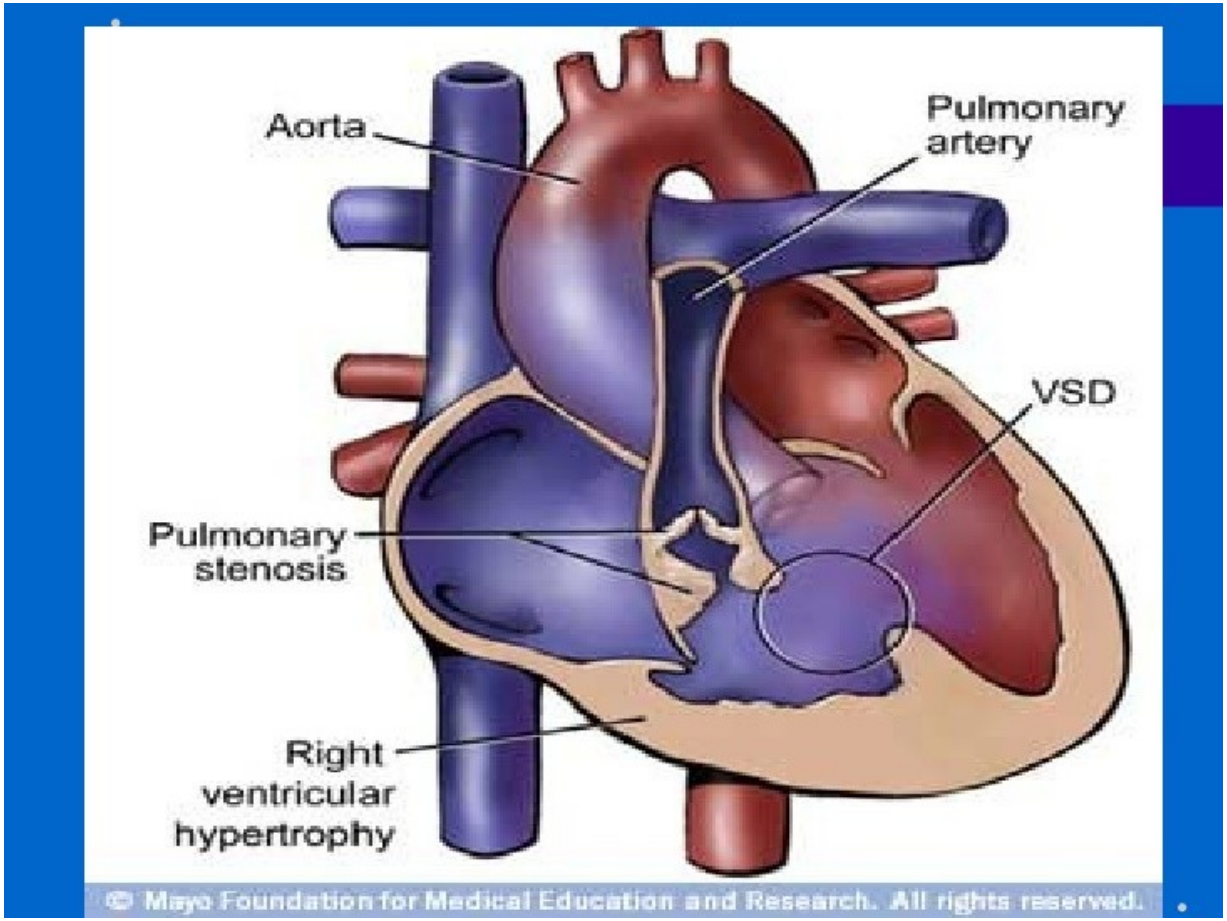
Treatment

- Medical management
  - digoxin
  - diuretics
- Adequate nutrition
  - high-calorie formula or breast milk
  - supplemental tube feedings
- Prophylactic antibiotics to prevent bacterial endocarditis
- Surgical repair – VSD will be closed w/ stitches or special patch
- Interventional cardiac catheterization – Septal occluder

Tetralogy of Fallot (TOF)

- a complex condition of several congenital defects that occur due to abnormal devt. of the fetal heart during the first 8 weeks of pregnancy. These problems include the following:

2. ventricular septal defect (VSD)
3. Pulmonary valve stenosis
4. overriding aorta - The aorta sits above both the left and right ventricles over the VSD, rather than just over the left ventricle. As a result, oxygen poor blood from the right ventricle can flow directly into the aorta instead of into the pulmonary artery to the lungs.
5. Right ventricular hypertrophy - The muscle of the right ventricle is thicker than usual because of having to work harder than normal.



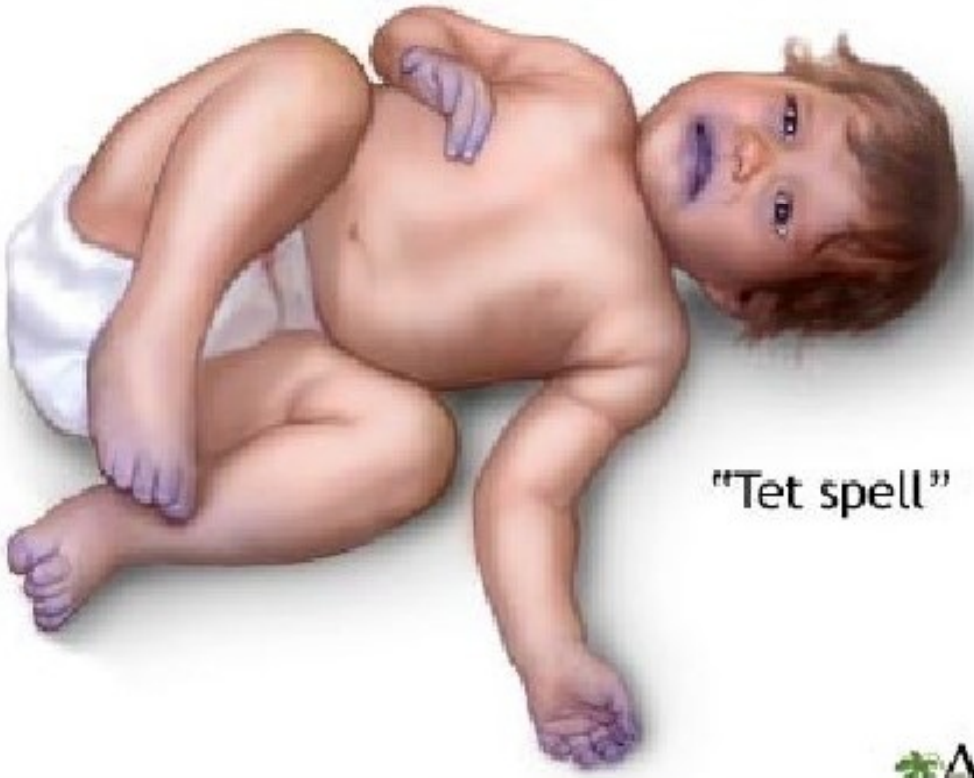
Tetralogy of Fallot (TOF)

- EFFECTS:
- If the right ventricle obstruction is severe, or if the pressure in the lungs is high → a large amount of oxygen-poor (blue) blood passes through the VSD, mixes with the oxygen-rich (red) blood in the left ventricle, and is pumped to the body → cyanosis
- The more blood that goes through the VSD, the less blood that goes through the pulmonary artery to the lungs → ↓ oxygenated blood to the left side of the heart.
- Soon, nearly all the blood in the left ventricle is oxygen-poor (blue). This is an emergency situation, as the body will not have enough oxygen to meet its needs.

Signs and Symptoms

- Cyanosis (blue color of the skin, lips, and nail beds) that occurs with such activity as crying or feeding
- Some babies do not have noticeable cyanosis, but may instead be very irritable or lethargic due to a decreasing amount of oxygen available in the bloodstream.
- Murmur
- Tachycardia
- Irritability
- Syncope
- Clubbing of fingers

Children with Tetralogy of Fallot exhibit bluish skin during episodes of crying or feeding.



"Tet spell"

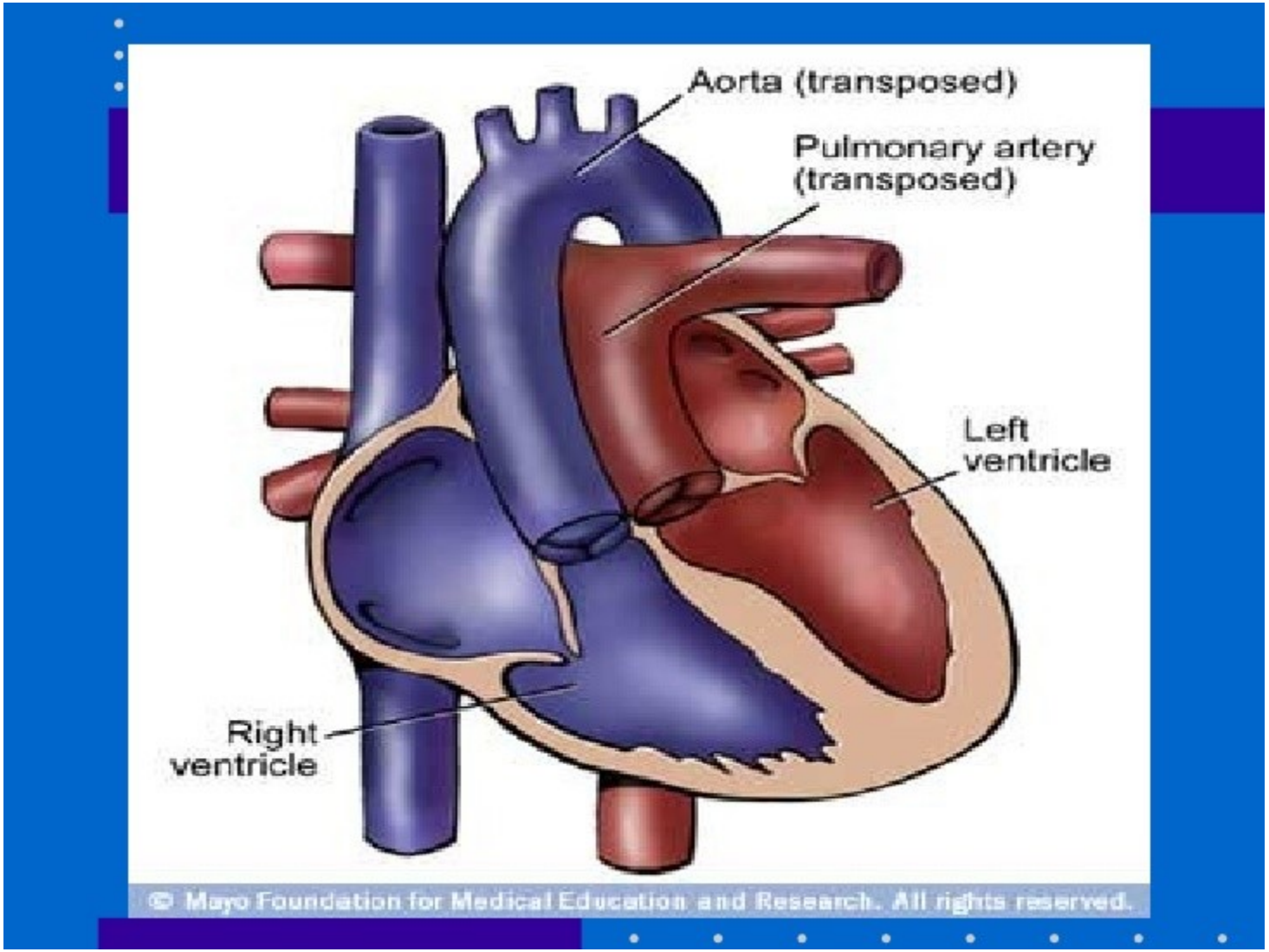
A.D.A.M.

Treatment

- Tetralogy of Fallot is treated by surgical repair of the defects. A team of cardiac surgeons performs the surgery, usually before an infant is 1 year old. In many cases, the repair is made at around 6 months of age, or even a little earlier. Repairing the heart defects will allow oxygen-poor (blue) blood to travel its normal route through the pulmonary artery to receive oxygen.
- The operation is performed under general anesthesia, and involves the following:
- The ventricular septal defect is closed with a patch.
- The obstructed pathway between the right ventricle and the pulmonary artery is opened and enlarged with a patch. If the pulmonary valve is small, it may be opened as well.

Transposition of the Great Arteries (TGA)

- the aorta is connected to the right ventricle, and the pulmonary artery is connected to the left ventricle
- Oxygen-poor (blue) blood returns to the right atrium from the body → passes through the right atrium and ventricle, → into the misconnected aorta back to the body.
- Oxygen-rich (red) blood returns to the left atrium from the lungs → passes through the left atrium and ventricle, → into the pulmonary artery and back to the lungs.
- Other heart defects are often associated with TGA
  - atrial or ventricular septal defect
  - may be necessary in order for the infant with TGA to survive
  - Allow mixing of blood – providing at least smaller amounts of oxygen to the body



Signs and Symptoms

- Cyanosis soon after delivery
- rapid breathing
- labored breathing
- rapid heart rate
- murmur
- cool, clammy skin

Treatment

- admitted to the NICU, placed on oxygen, and possibly even on a ventilator, IV medications to help the heart and lungs function more efficiently.
- a cardiac catheterization procedure will usually be performed to evaluate the defect(s) and the amount of blood that is mixing.
- as part of the cardiac catheterization, a balloon atrial septostomy may be performed to improve mixing of oxygen-rich (red) and oxygen-poor (blue) blood.
  - A catheter with a balloon in the tip is used to create an opening in the atrial septum
  - The catheter is guided through the foramen ovale (a small opening present in the atrial septum that closes shortly after birth) and into the left atrium.
  - The balloon is inflated.

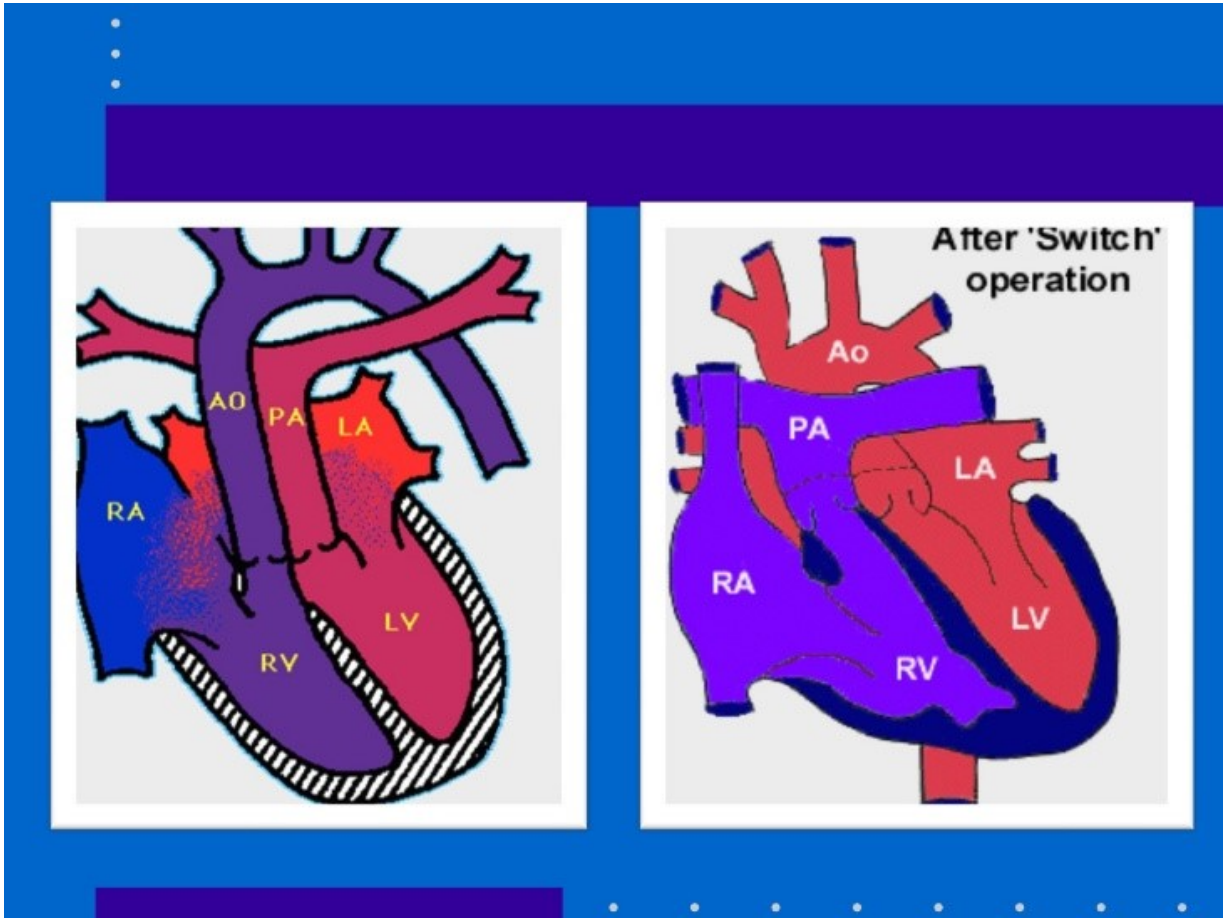
Treatment

- An IV prostaglandin E1 is given to keep the ductus arteriosus from closing.

Within the first 1 to 2 weeks of age, TGA is surgically repaired.

The "switch" operation is performed under GA, and involves the ff:

- The aorta is moved from the right ventricle to its normal position over the left ventricle.
- The pulmonary artery is moved from the left ventricle to its normal position over the right ventricle.
- The coronary arteries are moved so they will originate from the aorta and take oxygen-rich (red) blood to the heart muscle.
- Other defects, such as atrial or ventricular septal defects or a patent ductus arteriosus, are commonly closed.

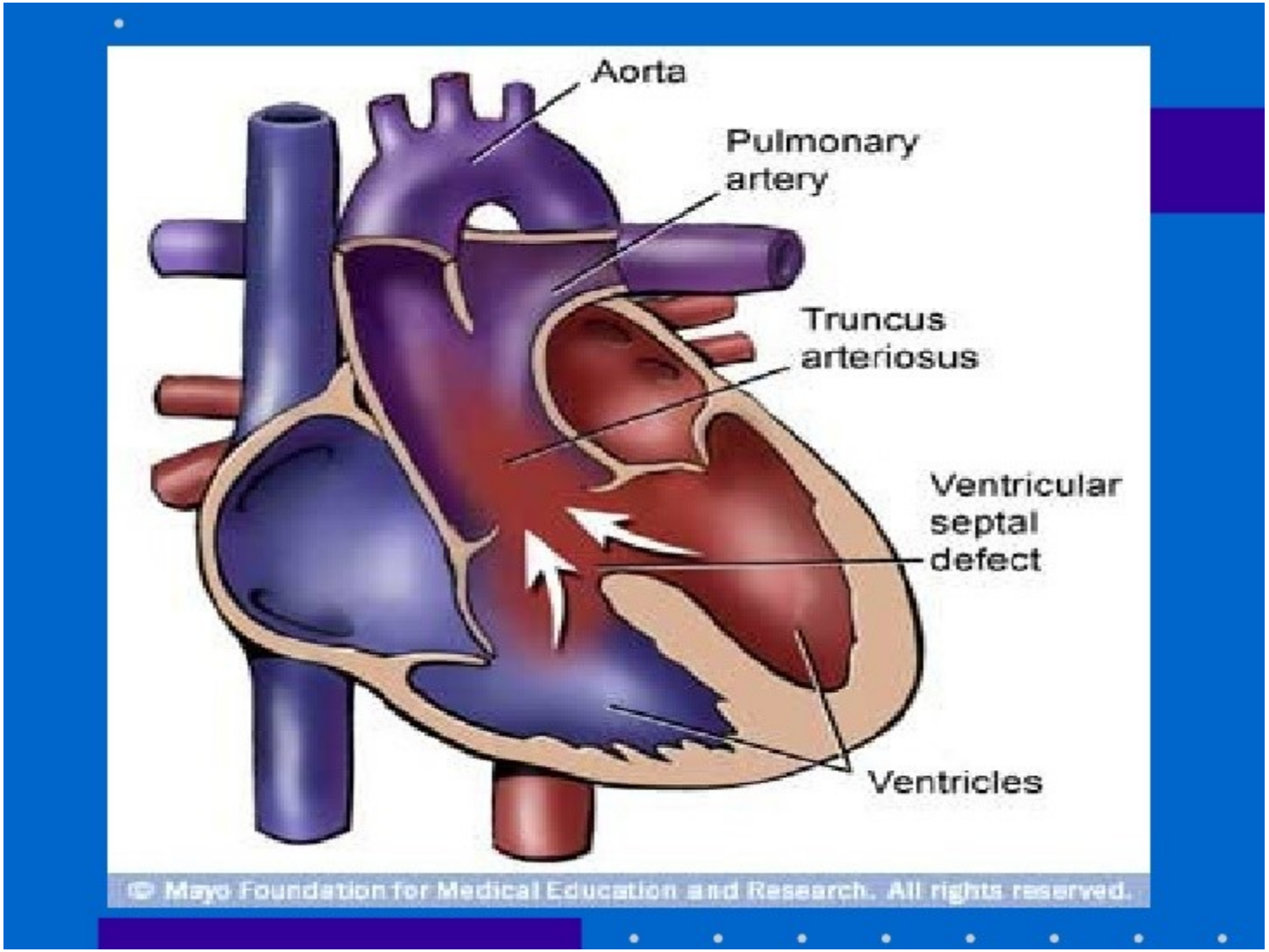

AO
PA
LA
RA
LV
RV
After 'Switch' operation
Ao
PA
LA
RA
LV
RV

Truncus Arteriosus

- The aorta and pulmonary artery start as a single blood vessel, which eventually divides and becomes two separate arteries.
- Truncus arteriosus occurs when the single great vessel fails to separate completely, leaving a connection between the aorta and pulmonary artery.
- Usually accompanied by a ventricular septal defect

EFFECTS:

- oxygen-poor (blue) and oxygen-rich (red) blood mix back and forth through the ventricular septal defect.
- This mixed blood then flows through the common truncal vessel. Some of it will flow to pulmonary artery and on to the lungs, and some of the mixed blood will go into the aortic branch and to the body.


Aorta
Pulmonary artery
Truncus arteriosus
Ventricular septal defect
Ventricles
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Signs and Symptoms

- cyanosis
- fatigue
- sweating
- pale skin
- cool skin
- rapid breathing
- heavy breathing
- rapid heart rate
- congested breathing
- disinterest in feeding, or tiring while feeding
- poor weight gain

Treatment

- Truncus arteriosus must be treated by surgical repair of the defects. However, medical support may be necessary until the best time for the operation to take place.
- medical management
  - Digoxin
  - Diuretics
  - ACE (angiotensin-converting enzyme) inhibitors - dilates the blood vessels, making it easier for the heart to pump blood forward into the body.
- adequate nutrition
  - high-calorie formula or breast milk
  - supplemental tube feedings

Treatment

- surgical repair

  Surgery is usually performed after the infant is 2 weeks old, but before the blood vessels in the lungs are overwhelmed by extra blood flow and become diseased.

  The operation is performed under general anesthesia, and involves the following:

  - The pulmonary arteries are detached from the common artery (truncus arteriosus) and connected to the right ventricle using a homograft (a section of pulmonary artery with its valves intact from a tissue donor).
  - The ventricular septal defect is closed with a patch.

Tricuspid Atresia

- In this condition, there is no tricuspid valve, therefore, no blood flows from the right atrium to the right ventricle.
- Blood in right atrium → foramen ovale → left atrium and left ventricle → aorta
- Tricuspid atresia defect is characterized by the following:
  - a small right ventricle
  - a large left ventricle
  - Small VSD and PDA
  - diminished pulmonary circulation
  - cyanosis - bluish color of the skin and mucous membranes caused from a lack of oxygen.
- A surgical shunting procedure is often necessary to increase

Nursing assessment

A. Become informed about the child's symptomatology and plan of medical care

- Obtain thorough nursing history to become familiar with the child and his family to recognize normal and abnormal patterns
- Discuss with the physician the plan for medical care

B. Make a baseline Nursing Assessment of the child's condition

- Observe and record information relevant to the child's growth and development
- Observe and record child's level of exercise tolerance
- Observe child's skin and mucous membranes for color and temperature changes

- Observe for clubbing of the fingers, especially the thumb nails, with thickening and shininess of the terminal phalanges-may occur in cyanotic children by 2-3 months of age.
- Observe for chest deformities
- Observe for respiratory pattern
- Palpate the child's pulses in all extremities
- Auscultate the child's heart
- Record vital signs

Nursing Diagnoses

- Impaired gas exchange related to altered pulmonary blood flow or oxygen deprivation
- Altered cardiac output related to specific anatomic defect
- Activity intolerance related to decreased oxygenation in blood and tissues
- Altered Nutrition: less than body requirements related to the excessive energy demands required by increased cardiac workload
- Increased potential for infection related to poor nutritional status
- Anxiety related to diagnostic procedures and hospitalization
- Developmental delay related to decreased energy, inadequate nutrition, physical limitations and social isolation
- Alteration in parenting related to parental perception of the child as vulnerable

Nursing Interventions

A. Provide adequate nutritional and fluid intake to maintain the growth and developmental needs of the child

- Feed in semi-erect position
- Provide small frequent feedings
- Provide foods with high nutritional value
- Determine child's likes and dislikes
- Strict input and output
- Daily weight

B. Prevent infection

- Prevent exposure to communicable diseases
- Immunizations should be up-to-date
- Handwashing should be observed
- Be certain that the child receives prophylactic medication for infective endocarditis

C. Reduce the workload of the heart since decreased activity and expenditure of energy will decrease oxygen requirements

- Uninterrupted rest
- Avoid unnecessary activities
- Prevent excessive crying
- Provide diversional activities
- Prevent constipation
- Relieve the respiratory distress associated with increased pulmonary blood flow or oxygen deprivation
  - Determine degree of respiratory distress
  - Include specific information in nursing record
  - Position child at 45 degree angle to decrease pressure of the viscera on the diaphragm and increase lung volume
  - Pin diapers loosely and provide loose-fitting pajamas for older children
  - Feed slowly
  - Tilt infant's head slightly
  - Suction the nose and throat if unable to cough out secretions
  - Provide oxygen therapy as needed
  - Improve oxygenation o that the body functions may be maintained

- Provide effective oxygen environment
- Observe response to oxygen therapy
- Observe response to oxygen weaning therapy
  - Relieve Hypoxic spells associated with cyanotic types of Congenital heart disease
  - Observe for “tet” spells
  - Encourage fluid intake
  - Obtain vital signs

- D. Observe child for symptoms of Congestive Heart Failure that occur frequently as a complication of Congenital Heart Disease
- E. Observe for the development of symptoms of infective endocarditis that may occur as a complication of congenital heart disease
- F. Observe for the development of thrombosis that may occur as a complication of congenital heart disease
- G. Prepare the child for diagnostic and treatment procedures
- H. Explain cardiac problems to child and parents


Thank you