

# CONGENITAL HEART DISEASES

Congenital Heart Disease is considered to be the most common birth defect. According to the American Heart Association, approximately 35,000 babies are born each year with some type of congenital heart defect. Congenital heart disease is responsible for more deaths in the first year of life than any other birth defects.

6,16 / 1000 viable newborns in Czech Republic  
!!! Critical heart diseases 2,35/ 1000 viable newborns

## Sexual differences in CHD

boys ..... 51%

girls.....49%

Multifactorial etiology : a/ genetics

Down syndrome, trisomy 13, Turner's syndrome, Marfan syndrome,  
Noonan syndrome,

b/ risk factors

# PRENATAL DETECTION

- Before birth, an obstetric ultrasound scan may be used to screen pregnant women for signs of CHD in their unborn babies. This screening scan is often performed around 20 weeks of pregnancy when the fast moving structures of the fetal heart are large enough to be more easily imaged. If CHD is suspected, a mother will be referred for a fetal echocardiograph, which is a more detailed, diagnostic ultrasound scan by a specialist cardiologist. It is increasingly possible for specialists to screen for CHD as early as 14 weeks, if CHD is suspected from other factors, such as a family history.

# Physical examination and diagnostic tools in CHD

## I. Observation of pulses

weakened pulses - cardiac failure

comparison of the strength and timing of femoral and radial pulses  
/ CoA/

bounding peripheral pulsations – suggest systemic hypertension or lesions associated with an aortic run off / i.g. PDA, truncus arteriosus/

## II. Cyanosis

III. Murmurs and character of heart sounds /systolic, diastolic murmurs  
innocent murmurs= functional- varies in intensity with time, position and activity level/

## IV. Respiratory and heart rate

## V. Blood pressure

## V. Oxygen test

## VI. Echocardiography, ECG, Radiography

# Congenital heart diseases

**1. According anatomical structure**

**2. With right-left or left –right shunt  
/with cyanosis x without cyanosis/**

**3. Critical / incompatible with life/ x no critical**

# CRITICAL CONGENITAL HEART DISEASE

## A. HEART FAILURE

1. tachycardia over 150/min
2. tachypnoe over 60/min
3. fatigue
4. hepatomegaly
5. fluid retention

## B. HYPOXIA

1. cyanosis
2. metabolic acidosis

# Management in infant with critical congenital heart disease

**Emergency for transport to Special Surgical Centre  
/incubator, O2, drugs, prostaglandins/**

**Report with details from history of patient, administered  
drugs and liquids, last feeding.....**

**Results of examinations before transport**



# Congenital Heart Disease

## **Acyanotic Congenital Heart Disease**

Atrial Septal Defect

Ventricular Septal Defect

Patent Ductus Arteriosus

Atrio-Ventricular Canal Defect (Endocardial Cushion Defect)

Pulmonary Stenosis

Left Ventricular Outflow Obstruction

Coarctation of the Aorta

## **Cyanotic Congenital Heart Disease**

Tetralogy of Fallot

Pulmonary Atresia with VSD

Pulmonary Atresia with Intact Ventricular Septum

Transposition of the Great Arteries

Double Outlet Right Ventricle

Ebstein's Anomalies

Hypoplastic Left Heart

# The most frequent critical congenital heart diseases in newborn child

**Transposition of the great arteries**

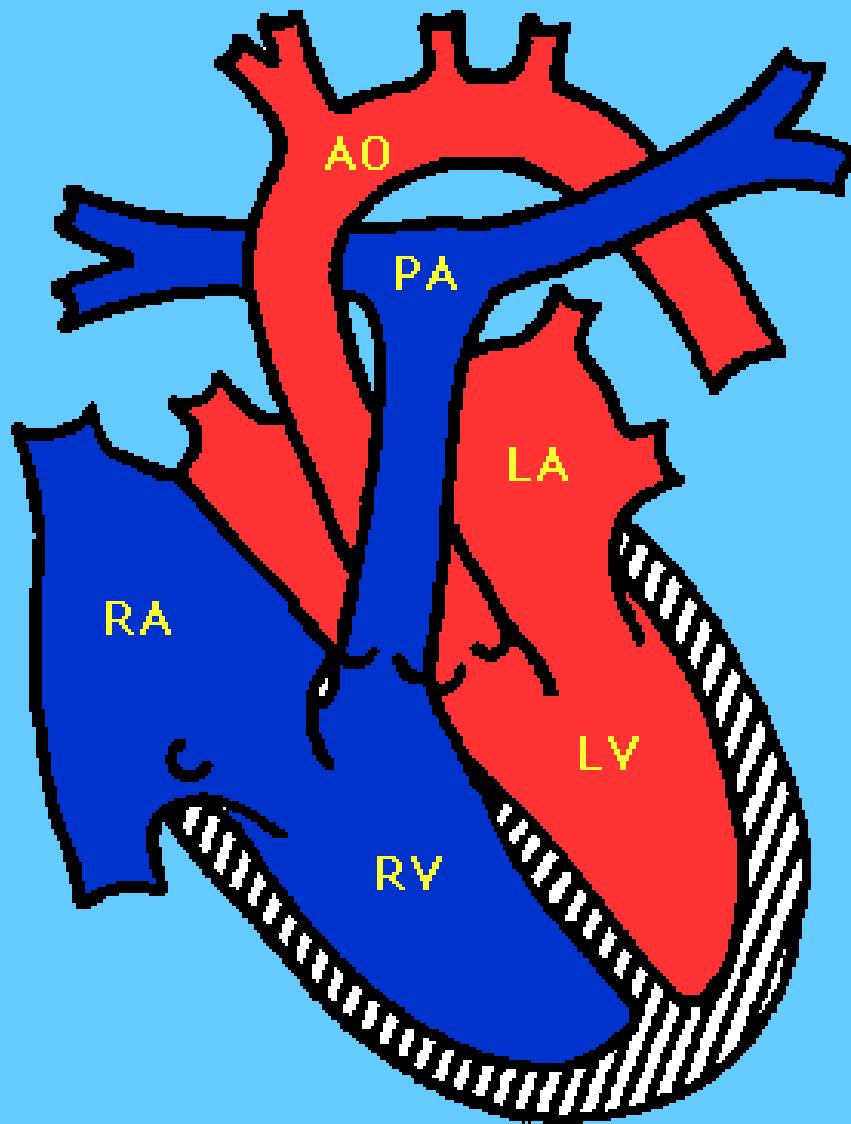
**Coarctation of the aorta**

**Pulmonary atresia**

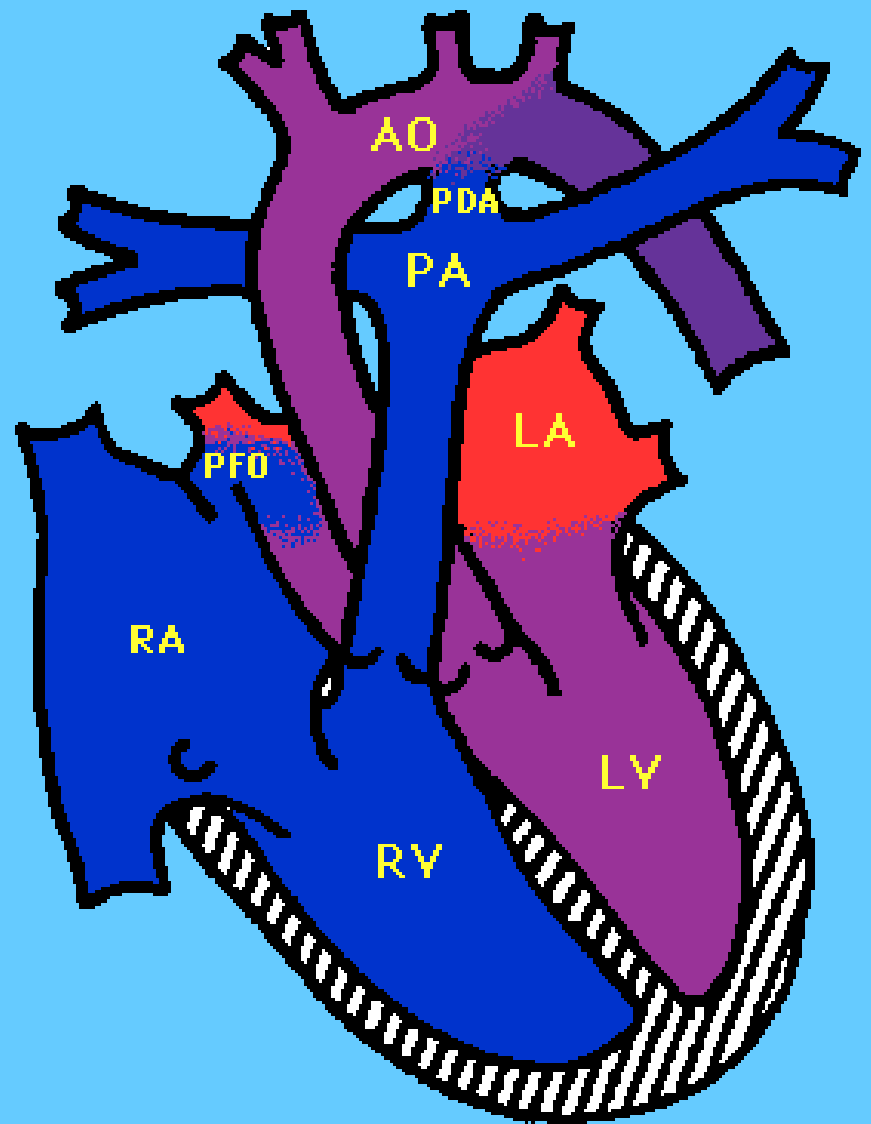
**Hypoplastic left ventricle**



# Persistence of the Fetal Circulation

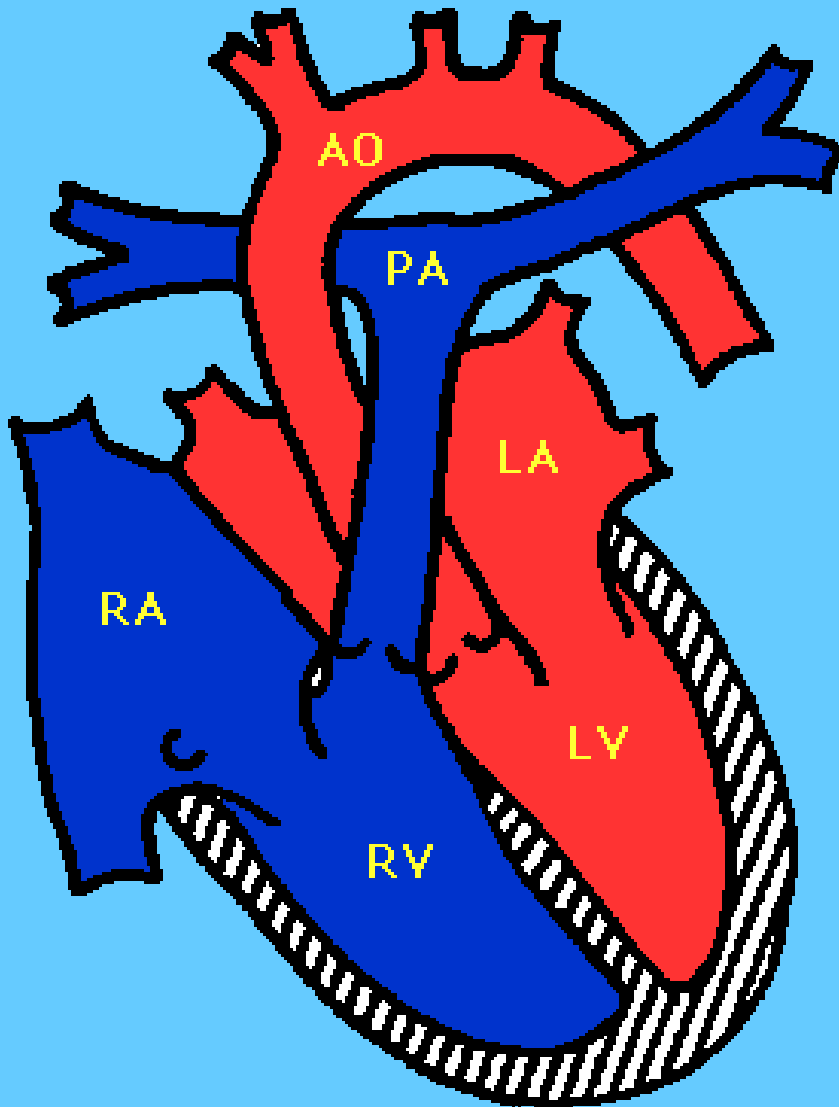


Normal

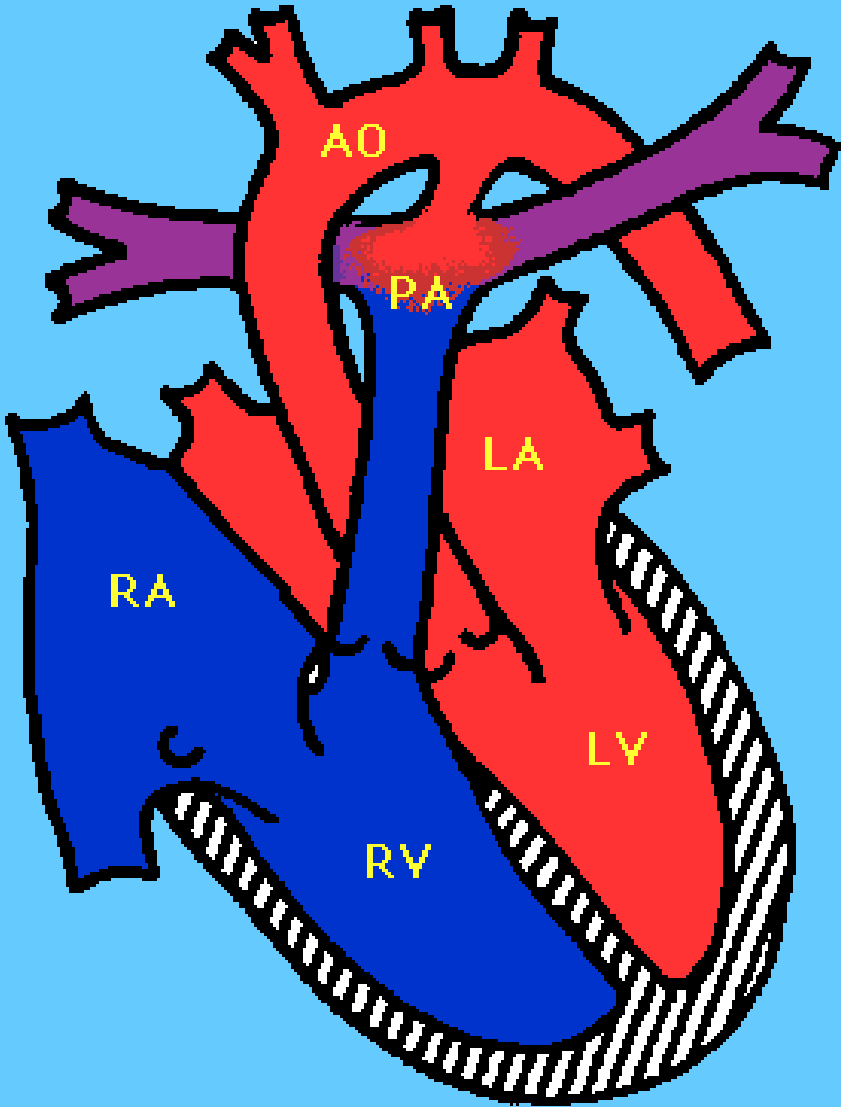


Persistence of the Fetal Circulation

# Patent Ductus Arteriosus



Normal

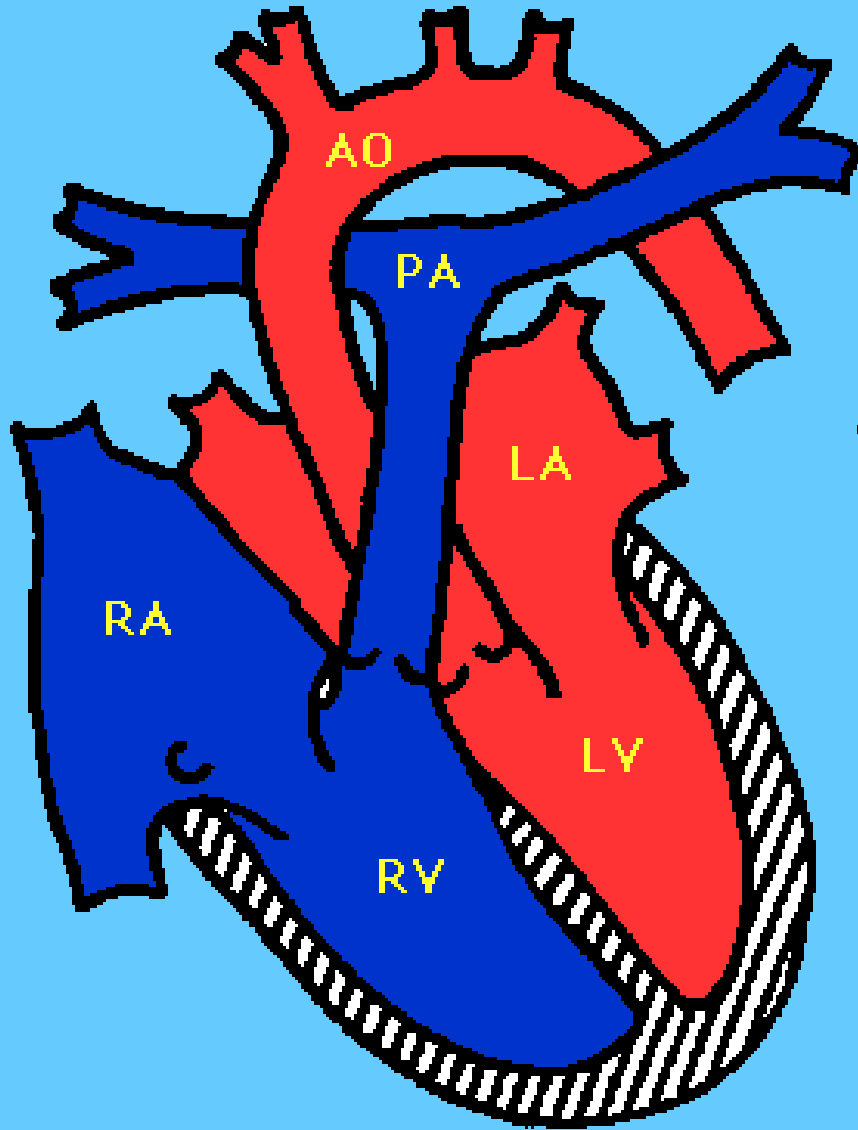


Patent Ductus Arteriosus

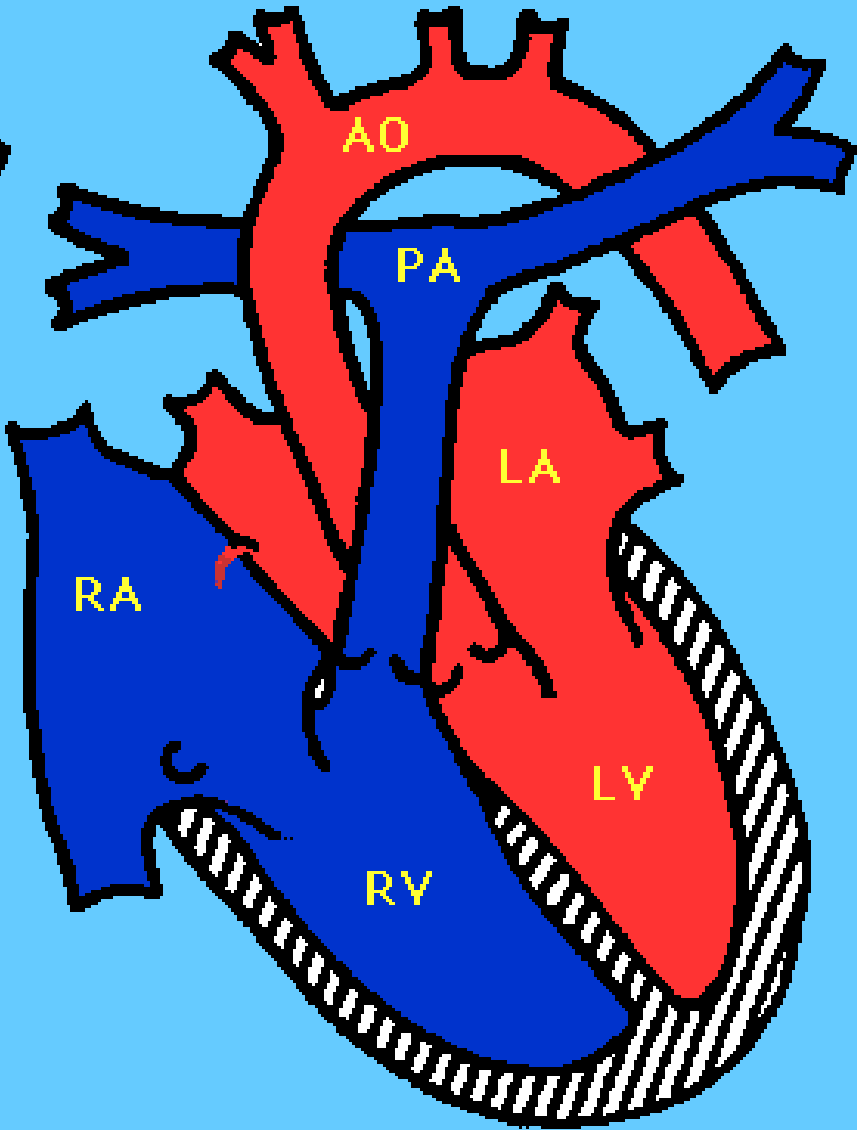
# Patent ductus arteriosus

Before birth, a large artery (ductus arteriosus) lets the blood bypass the lungs because the fetus gets its oxygen through the placenta. The ductus normally closes soon after birth so that blood can travel to the lungs and pick up oxygen. If it doesn't close, the baby may develop heart failure. This problem occurs most frequently in premature babies. Drug treatment /indomethacin/ often can close the ductus. If that doesn't work, surgery can close it.

# Patent Foramen Ovale

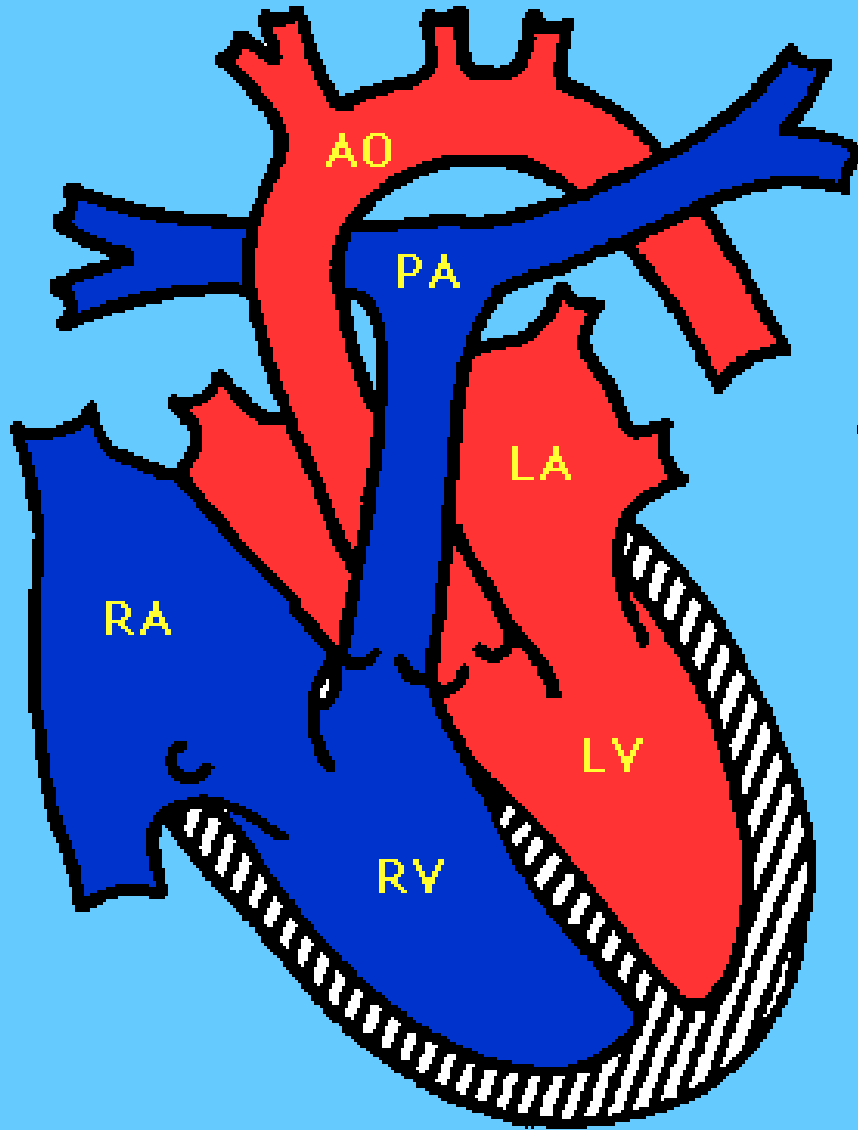


Normal

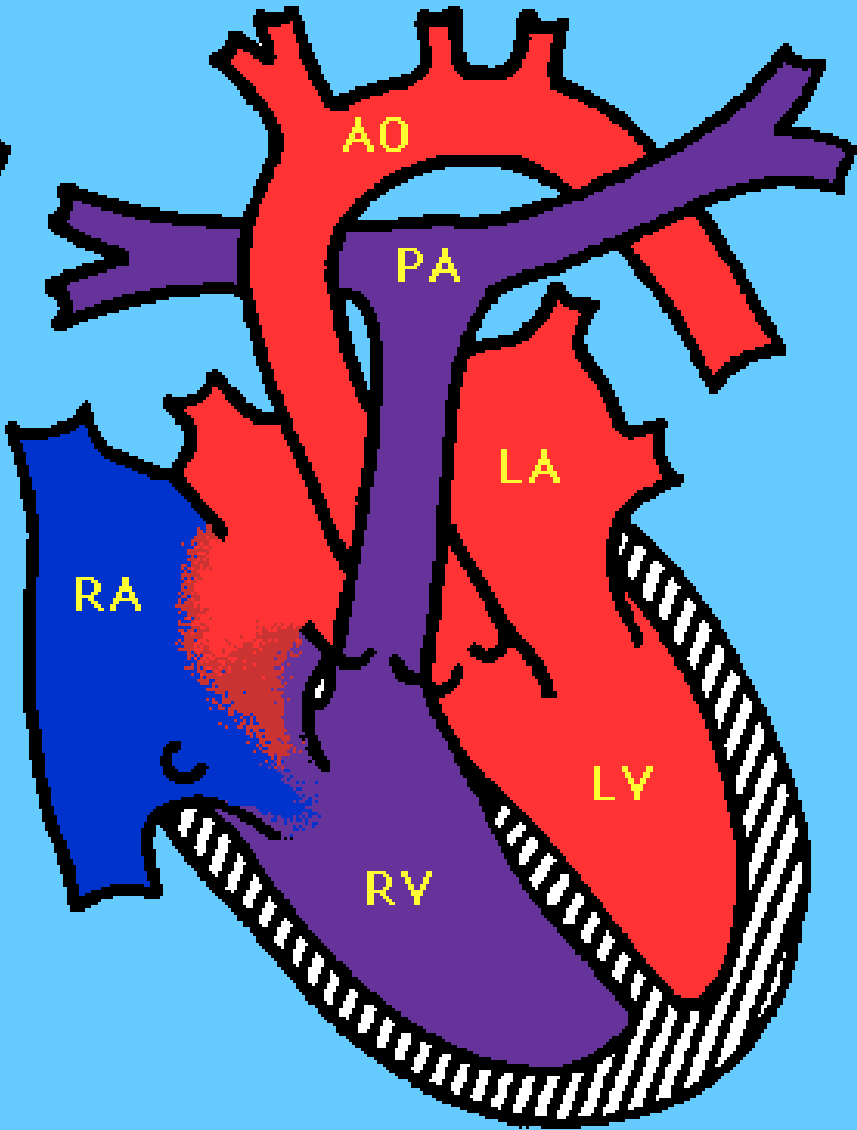


Patent Foramen Ovale

# Atrial Septal Defect

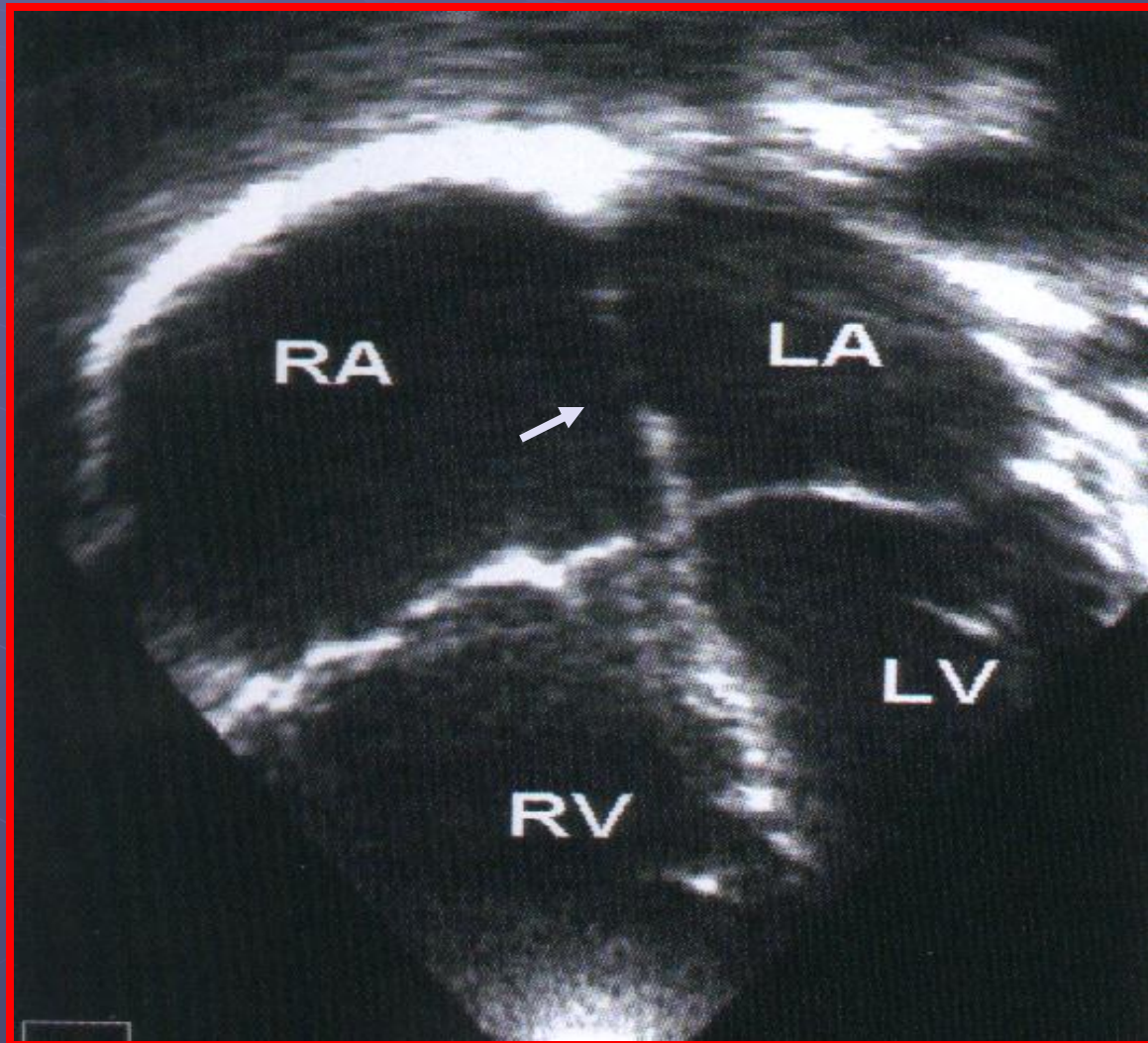


Normal



Atrial Septal Defect

# ASD



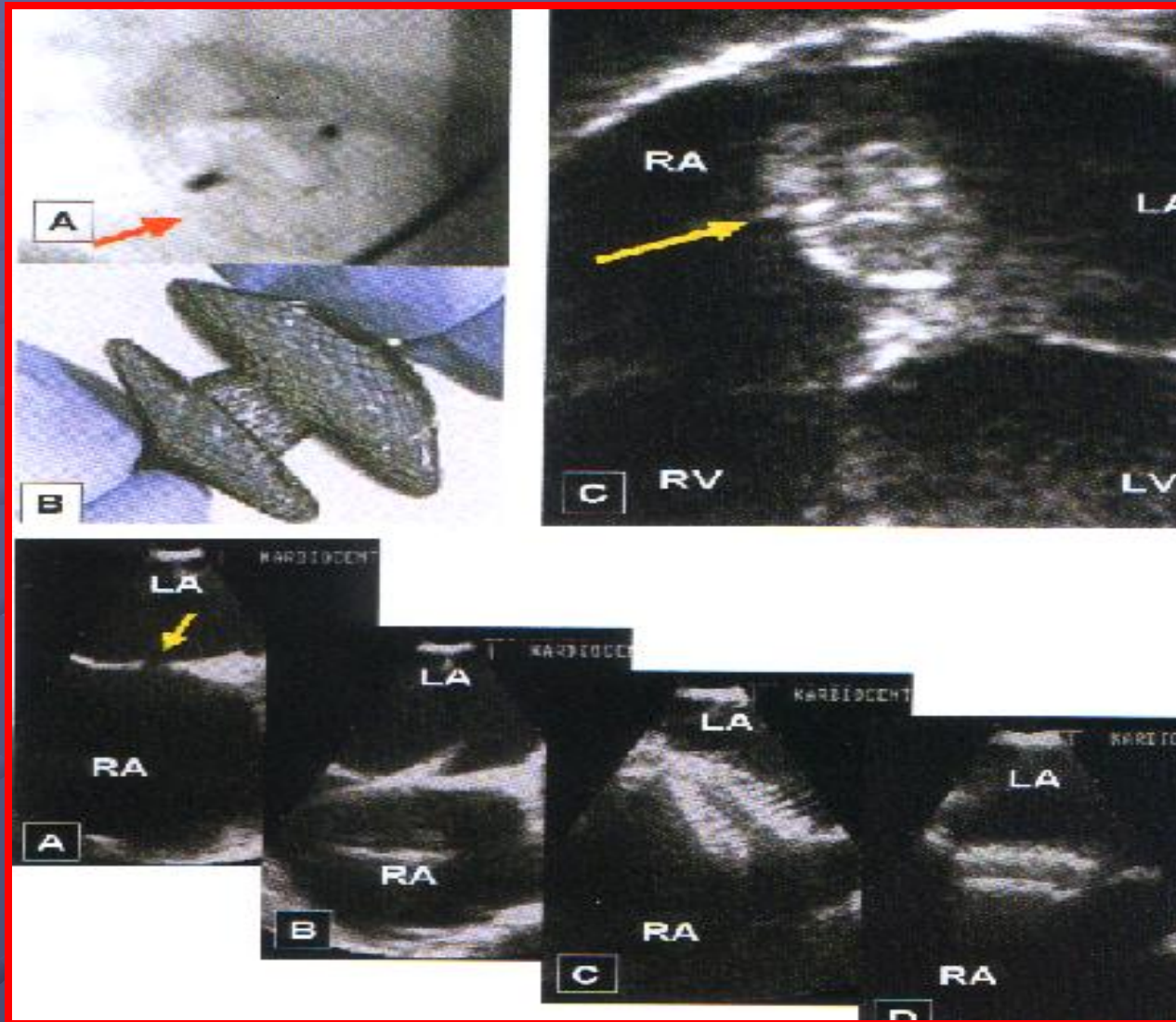


# Atrial septal defect (ASD)

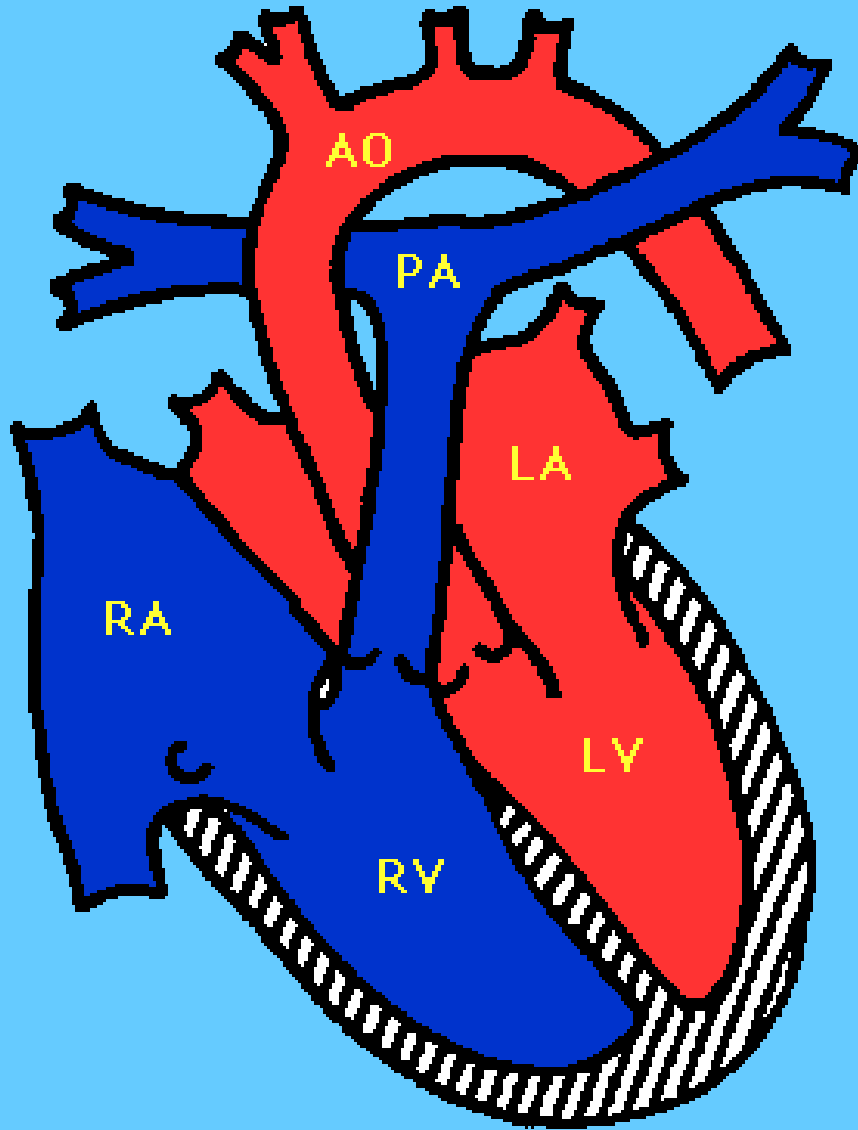
- In fetal circulation there is normally an opening between the two atria to allow blood to bypass the lungs. This opening usually closes about the time the baby is born. If the ASD is persistent, blood continues to flow from the left to the right atria.
- **Symptoms** /with small-to moderate-sized defects no symptoms /
- Frequent respiratory infections in children
- Difficulty breathing ([dyspnea](#))
- [Shortness of breath](#) with activity
- Sensation of feeling the heart beat ([palpitations](#))
- **Treatment** Surgical closure or a closure device (Amplatz device) into the heart through catheters.

# ASD – AMPLATZ

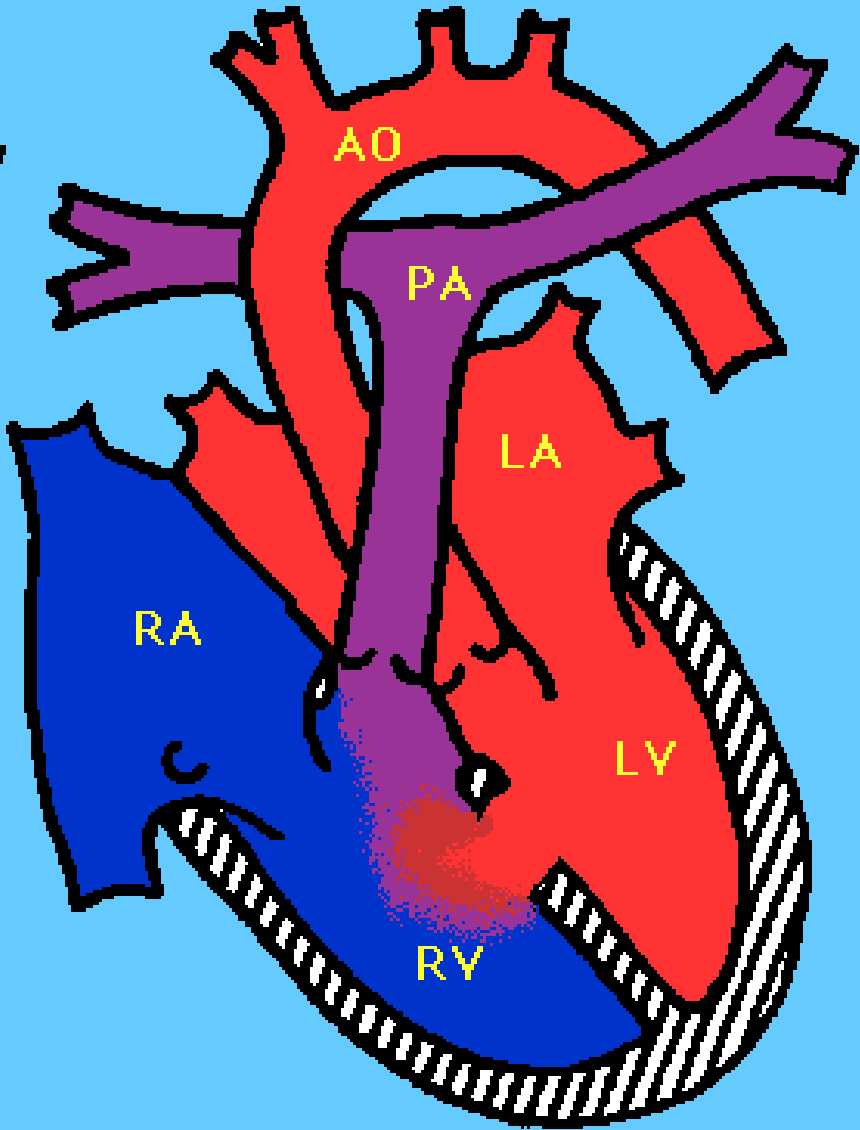
FROM LECTURE DOC. Urbanová



# Ventricular Septal Defect

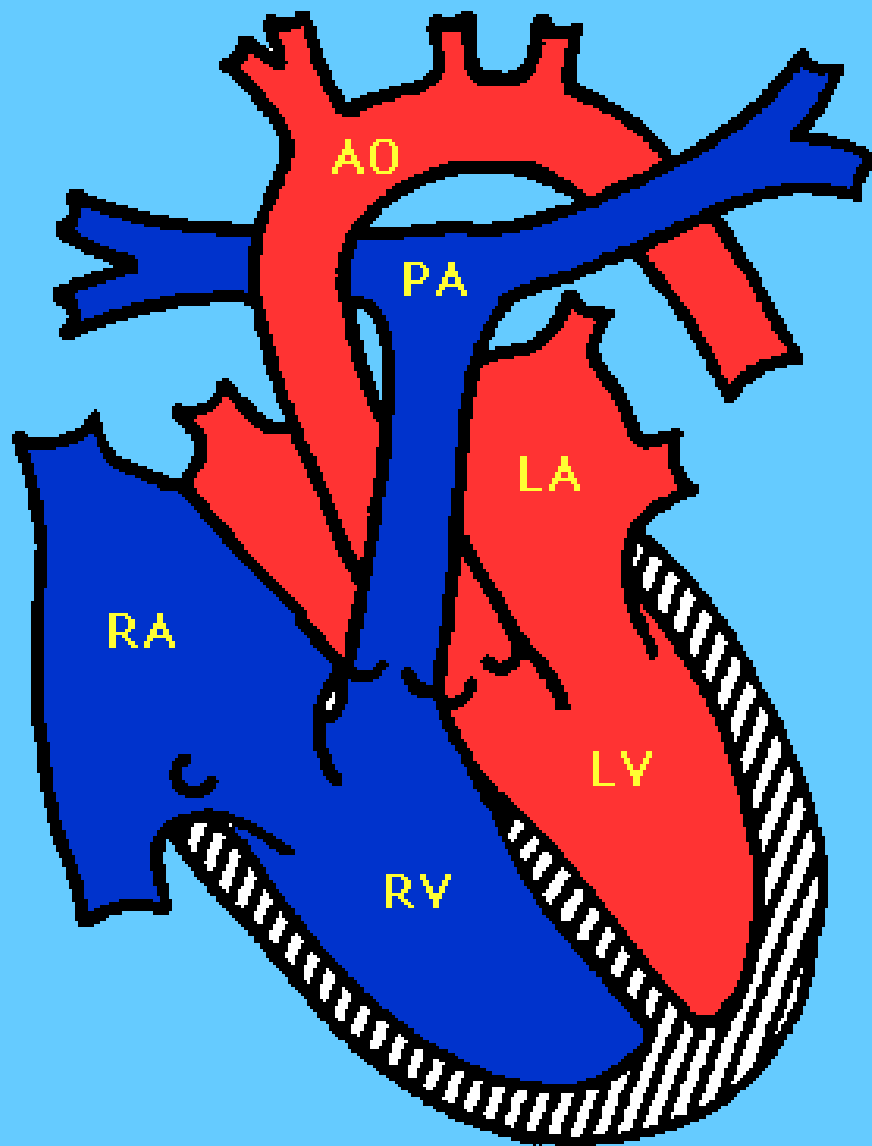


Normal

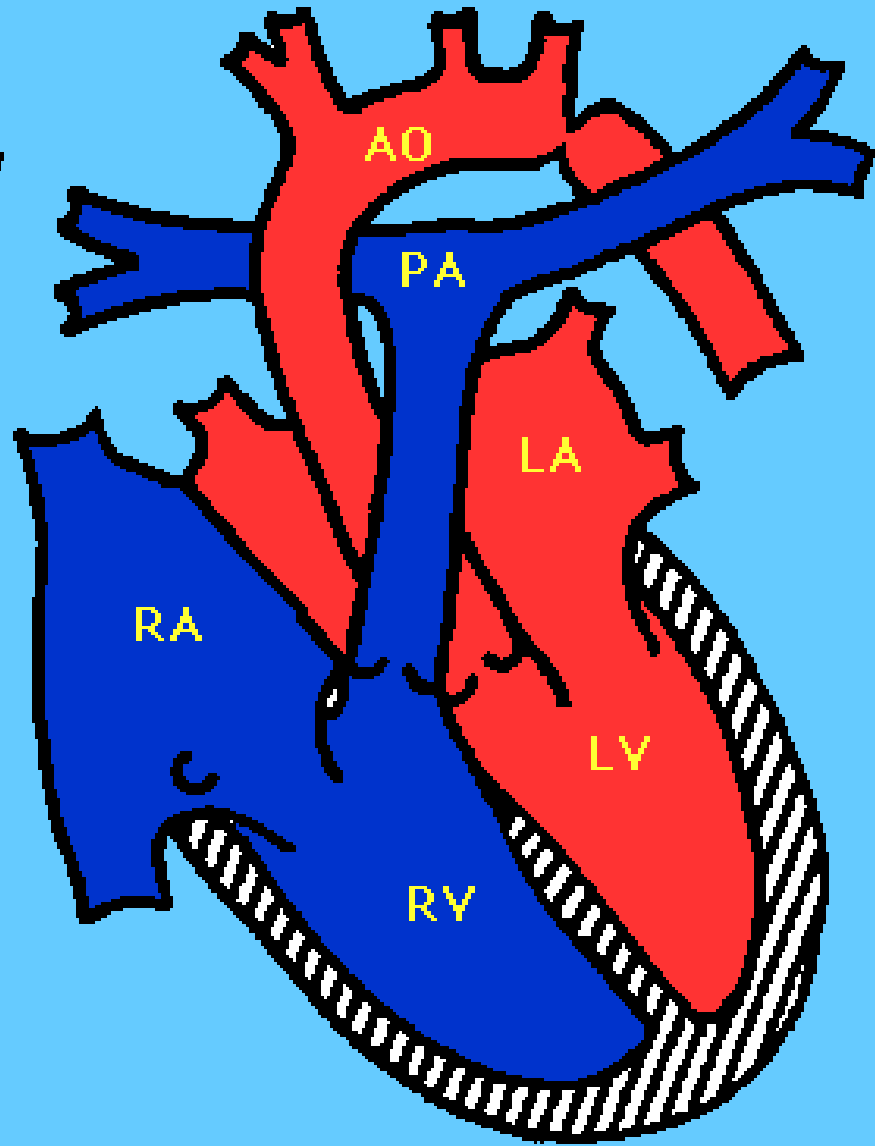


Ventricular Septal Defect

# Coarctation of the Aorta



Normal

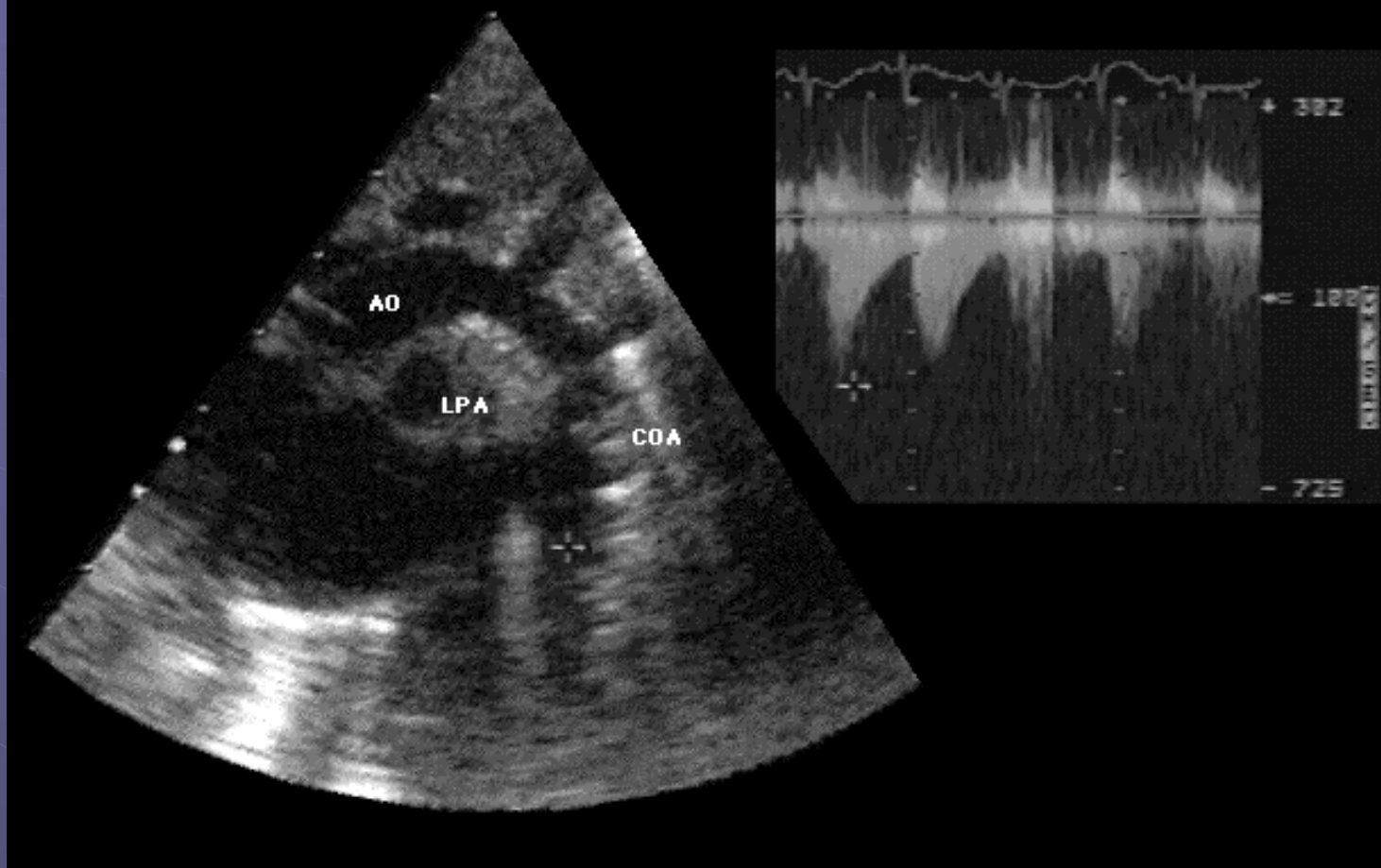


Coarctation of the Aorta

COA



## Coarctation of the Aorta, 2-D & CW Doppler, Suprasternal Notch





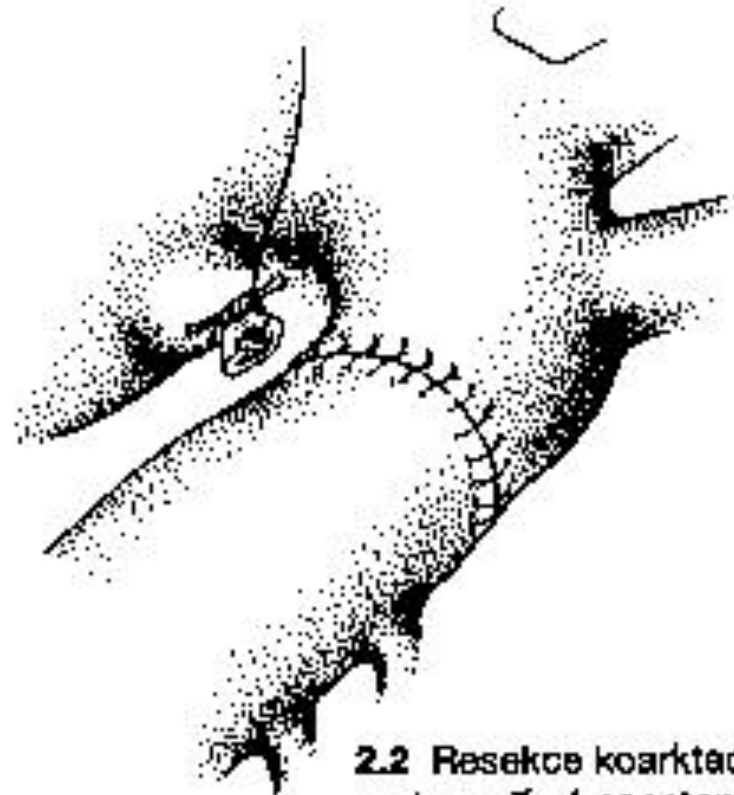
# Coarctation of the aorta

- Part of the aorta, the large artery that sends blood from the heart to the rest of the body, may be too narrow for the blood to flow evenly.
- **Symptoms: femoral pulse may not be felt at all or are weaker**
- :
- **Therapy:** A surgeon can cut away the narrow part and sew the open ends together, replace the constricted section with man-made material, or patch it with part of a blood vessel taken from elsewhere in the body. Sometimes, this narrowed area can be widened by inflating a balloon on the tip of a catheter (tube) inserted through an artery.

# COA

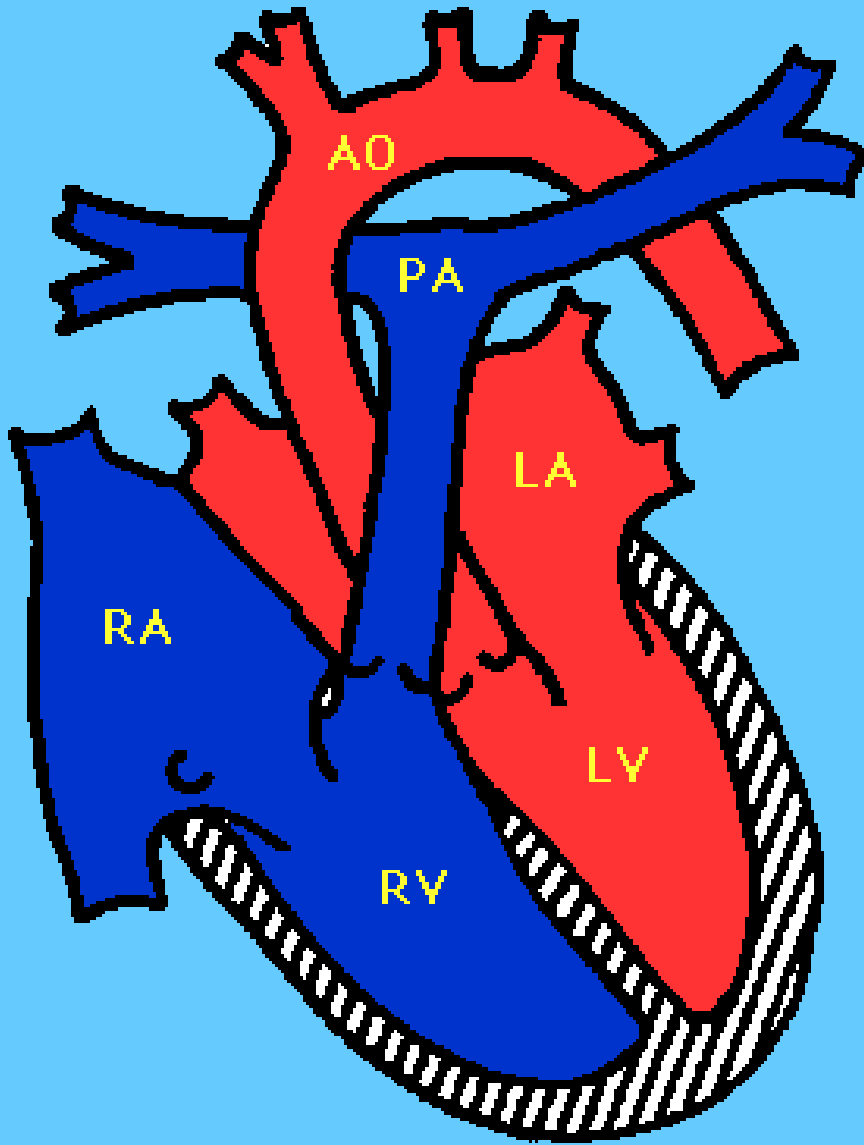


**2.1** Koarktace istimu aorty.  
Aorta je zúžena před  
odstupem tepenné dučeje.  
Naznačena jsou místa  
resekce koarktace.

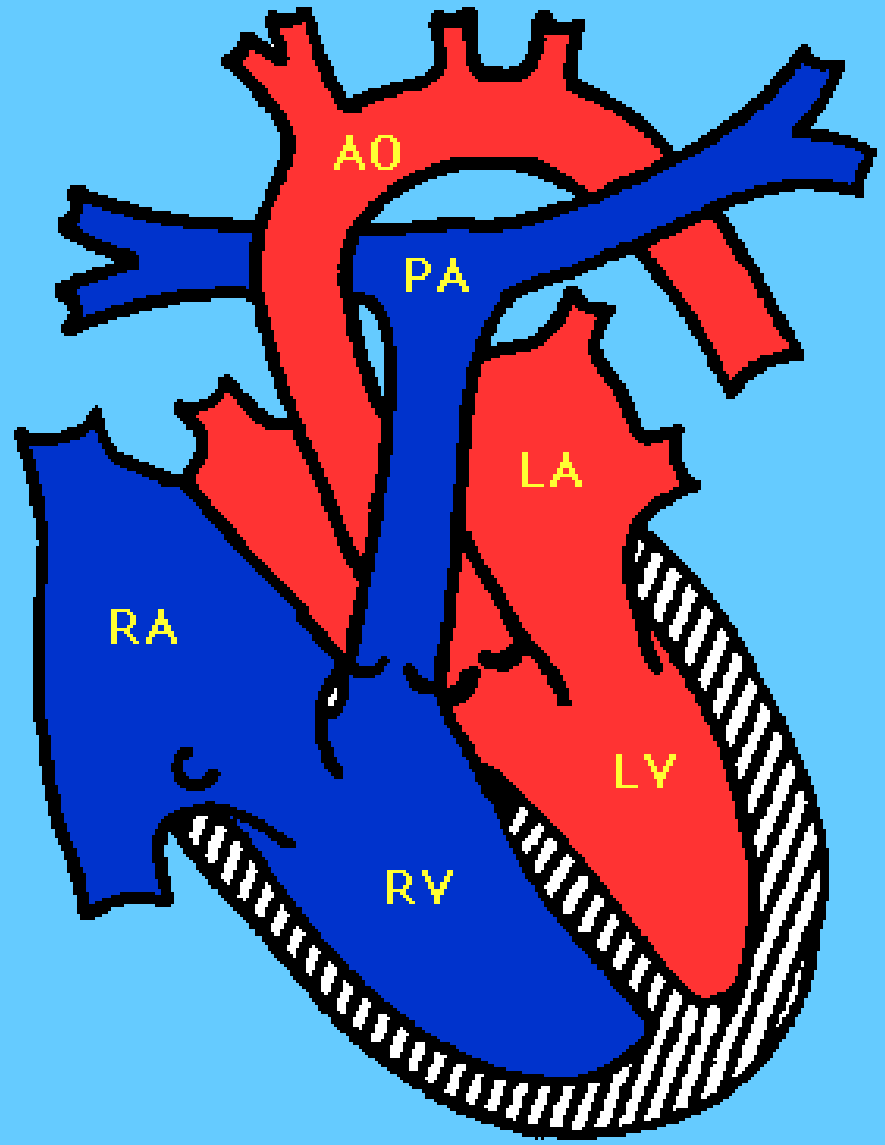


**2.2** Resekce koarktace  
aorty a přímá anastomóza  
koncem ke konci. Podvaz  
dučeje

# Aortic Valve Stenosis

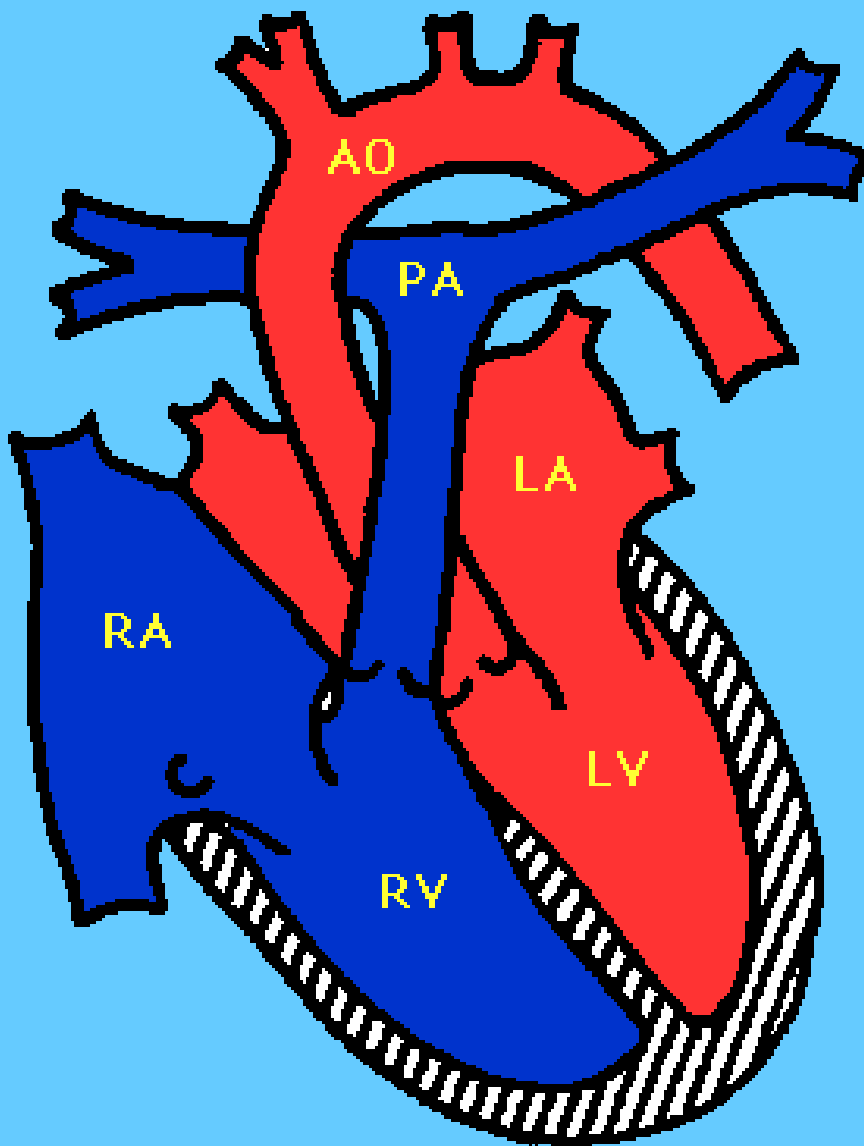


Normal

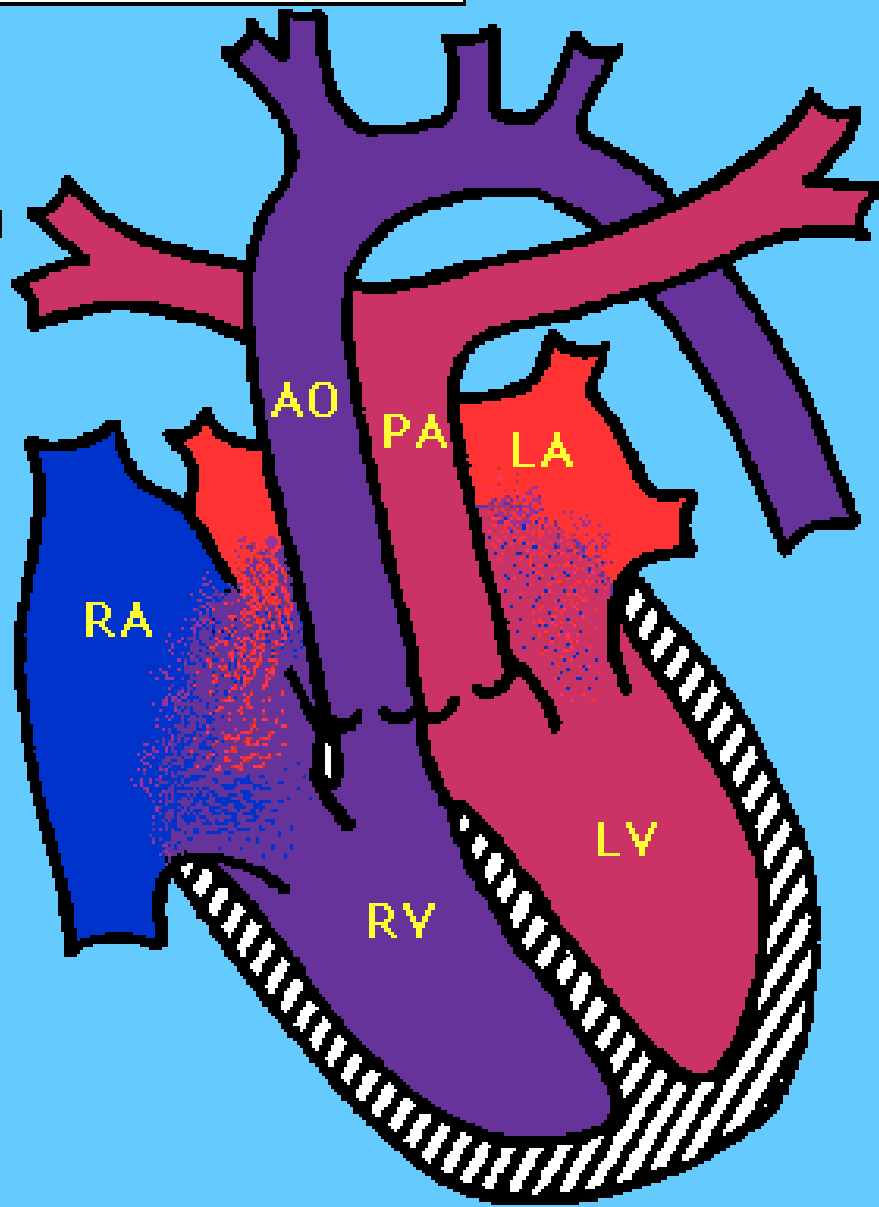


Aortic Valve Stenosis

# Transposition of the Great Vessels



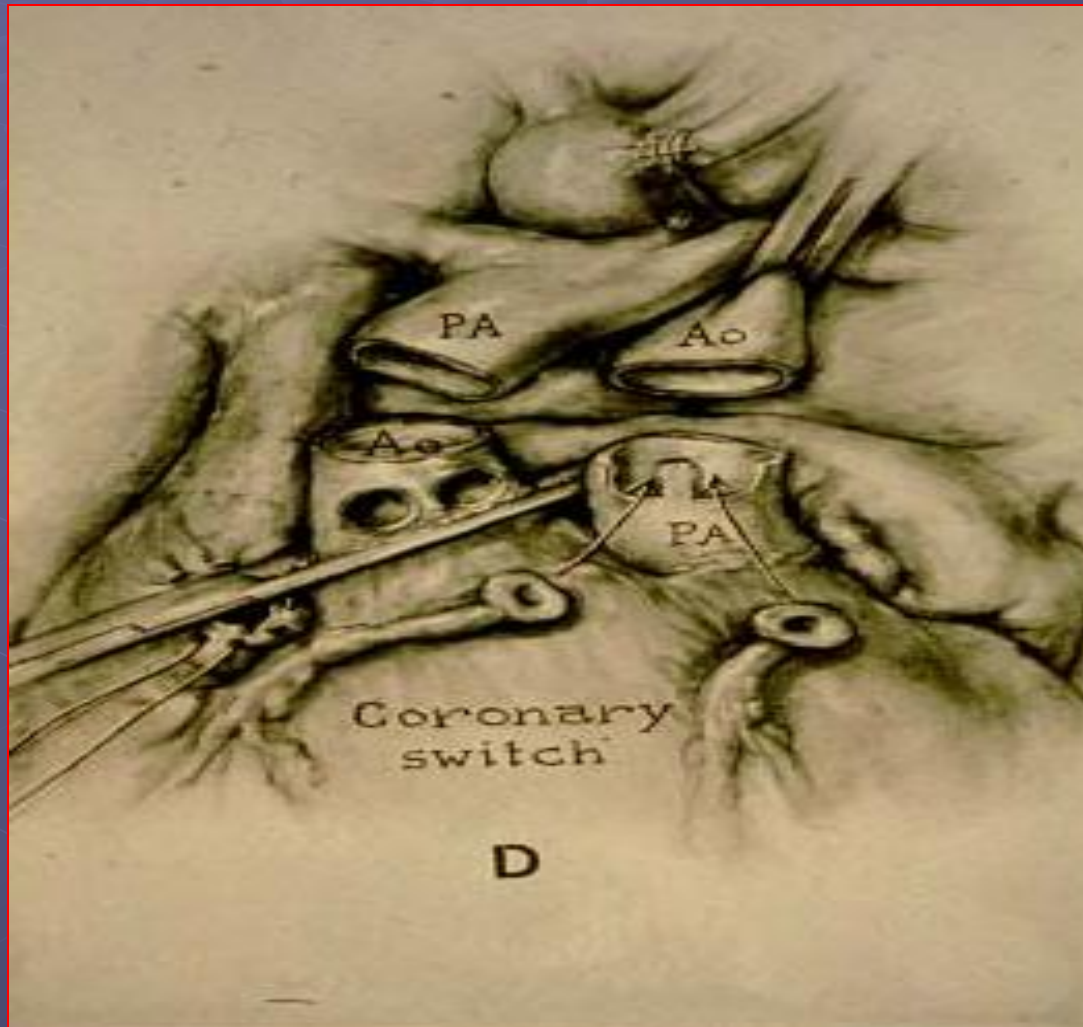
Normal



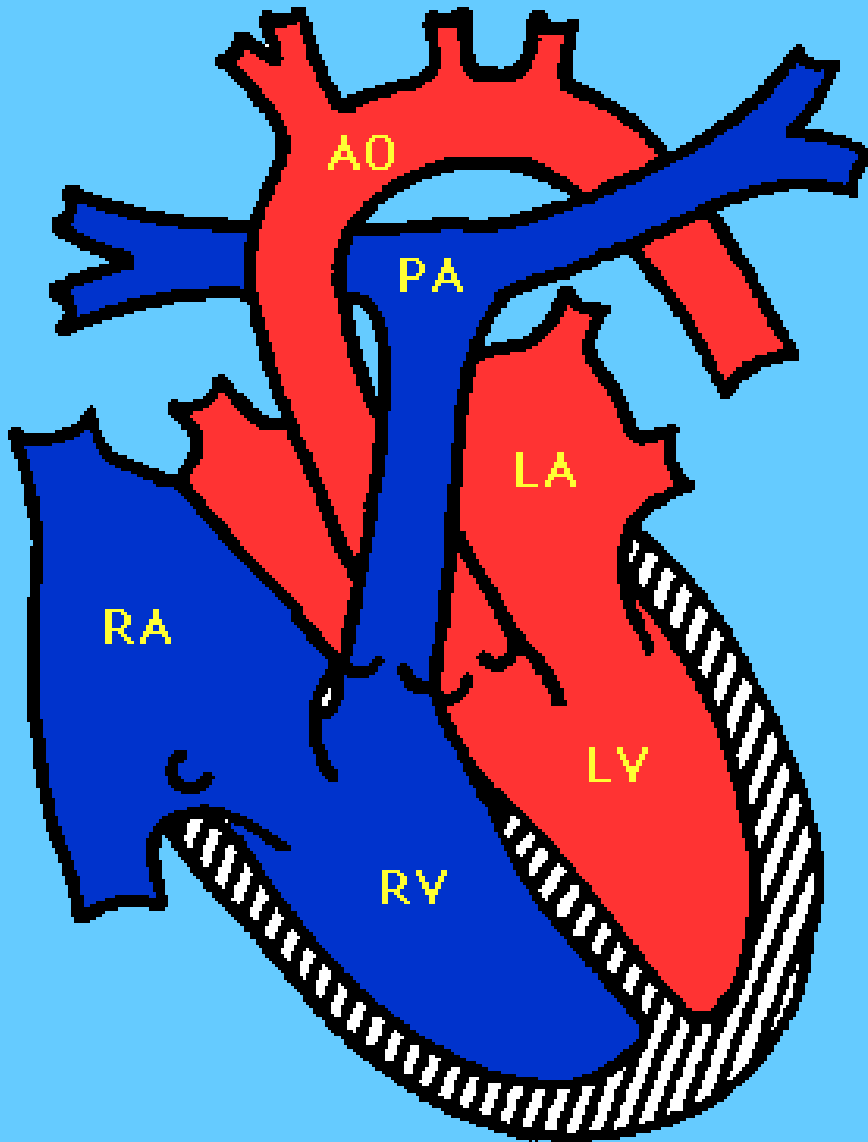
Transposition of the Great Vessels

# TGA - SWITCH

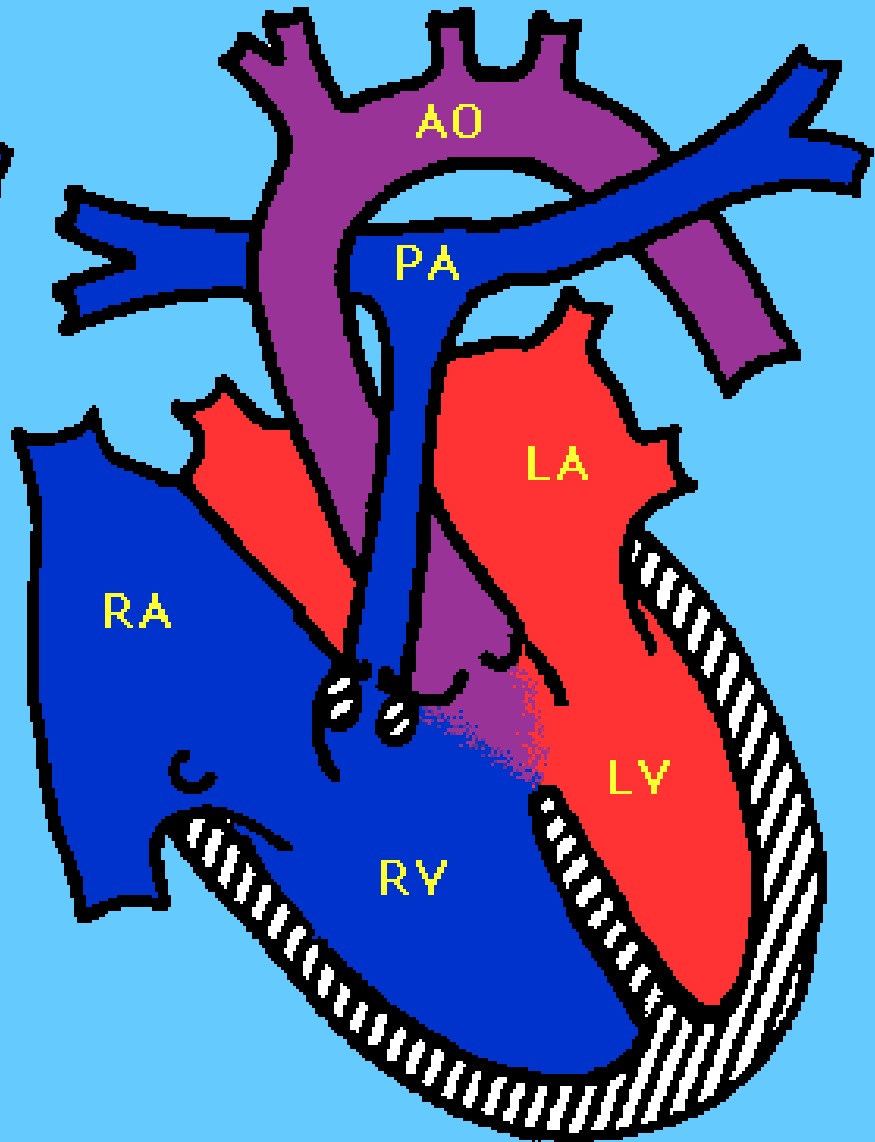
FROM LECTURE DOC. Urbanová



# Tetralogy of Fallot



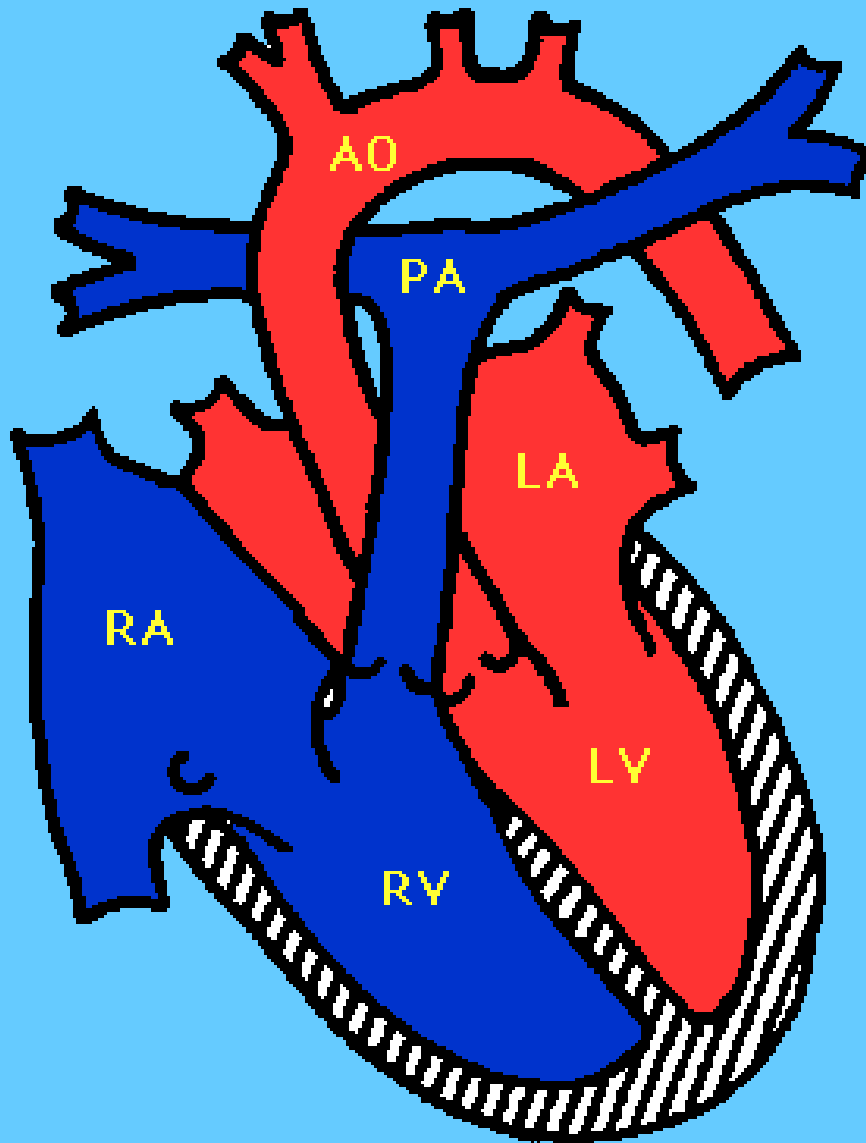
Normal



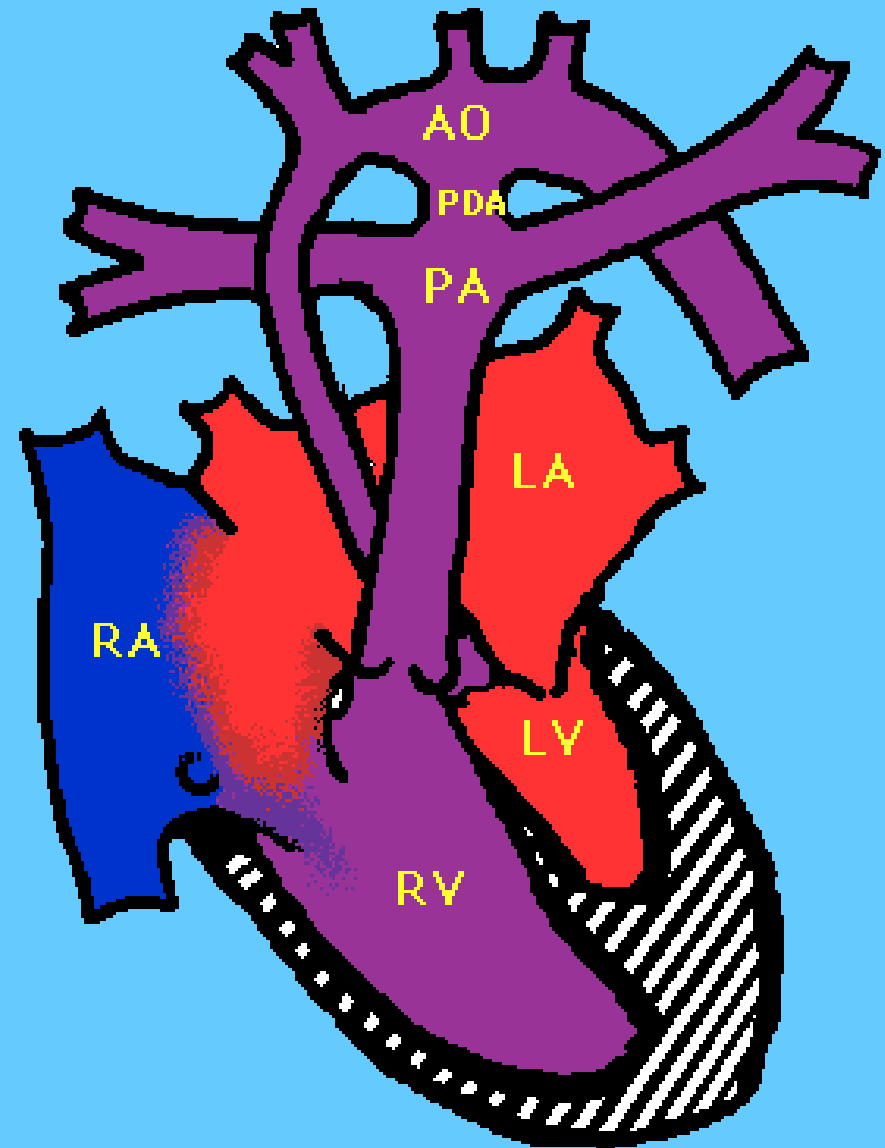
Tetralogy of Fallot



# Hypoplastic Left Heart Syndrome



Normal



Hypoplastic Left Heart

# HEART FAILURE

## Definition:

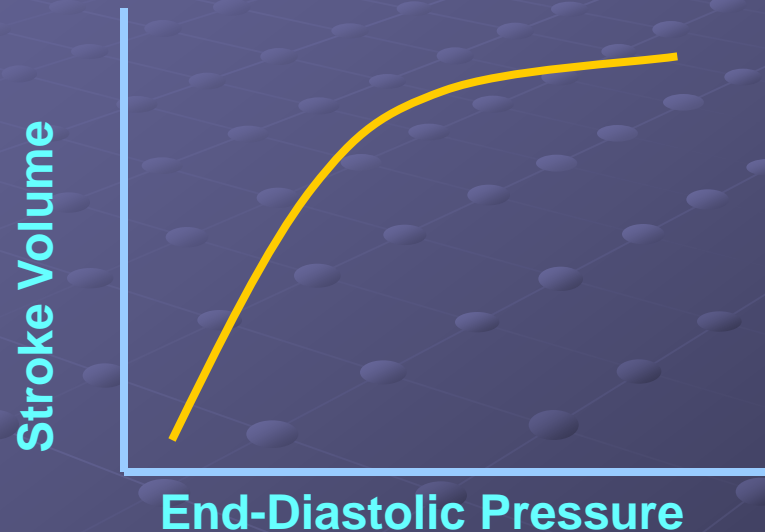
The heart cannot supply the blood flow demanded by the tissues, a clinical syndrome of symptoms and signs largely resulting from elevated atrial pressures can be recognized as congestive heart failure.

## STARLING LAW

The heart may fail if it is confronted with

- A. An abnormally high afterload
- B. An excessive preload
- C. Impaired myocardial contractility
- D. Inadequate diastolic filling / constrictive pericarditis, chronic tachyarrhythmia.../
- E. Changes in heart rate

# Frank-Starling Phenomenon



**“In the normal heart, the diastolic volume (preload) is the principal force that governs the strength of ventricular contraction.”**

*Otto Frank and Ernest Starling*

# C.O. = Stroke volume x Heart rate

- Stroke volume:
  - Preload
  - Myocardial contractility
  - Afterload:
    - systemic and pulmonary resistance
    - blood viscosity
- Heart Rate
  - Bradycardia
  - Sustained tachycardia

## SYMPTOMS OF HEART FAILURE

tachycardia

tachypnoe, dyspnoe

hepatomegaly

cough, fatigue, cold limbs, small diuresis

## BLOOD EXAMINATION IN HEART FAILURE

metabolic acidosis, hyperglycemia, increase of enzymes,  
osmolality of plasma, changes in ion composition of urine

# THERAPY of HEART FAILURE

## **1. Decrease of energetic requirement**

/ O<sub>2</sub>, temperature, nutrition, position, rest, artificial ventilation , therapy of infection, blood transfusion if present anaemia/

## **2. Increase of cardiac output /CO/**

**CO / per minute/ = pulse rate x pulse volume**

**a/ affecting of preload / blood amount comming to the heart/ - diuretics, infusion**

**b/ affecting of contractility- digoxin, catecholamines**

**c/ affecting of afterload - vasodilantia**

**d/ affecting of heart rate- beta blockers, atropin**