Heart defects in children

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Incidence of congenital heart defects (CHD)

- Average 8 in 1000 live births
- 10-15% of heart defects are complex
- In most cases it is possible to correct heart defects (non invasive methods or cardiac surgery), usually in the first year of life

Etiology

- multifactorial, usually unknown

- Genetic etiology in 10 % of congenital heart defects

  Higher risk of disease in children with relevant family history of cardiovascular abnormalities

- Environmental factors acting in utero: infections (e.g. rubella), diabetes mellitus, lupus erythematosus, alcoholism (FAS), some medications (e.g. warfarin)

Examples of heart defects in chromosomopathy:

- **Down Syndrome** (trisomy 21): VSD, defects of atrio-ventricular septum
- Edward's Syndrome (trisomy 18): complex heart defects
- Turner Syndrome (45 X0): aortic valve stenosis, coarctation of the aorta
- chromosome 22q11.2 deletion: defects of the aorta, tetralogy of Fallot, persistent truncus arteriosus
- William's syndrome (chromosome 7q11.23 microdeletion): stenosis of aorta, stenosis of pulmonary artery

Proper understanding of heart defect problems requires a reminder:

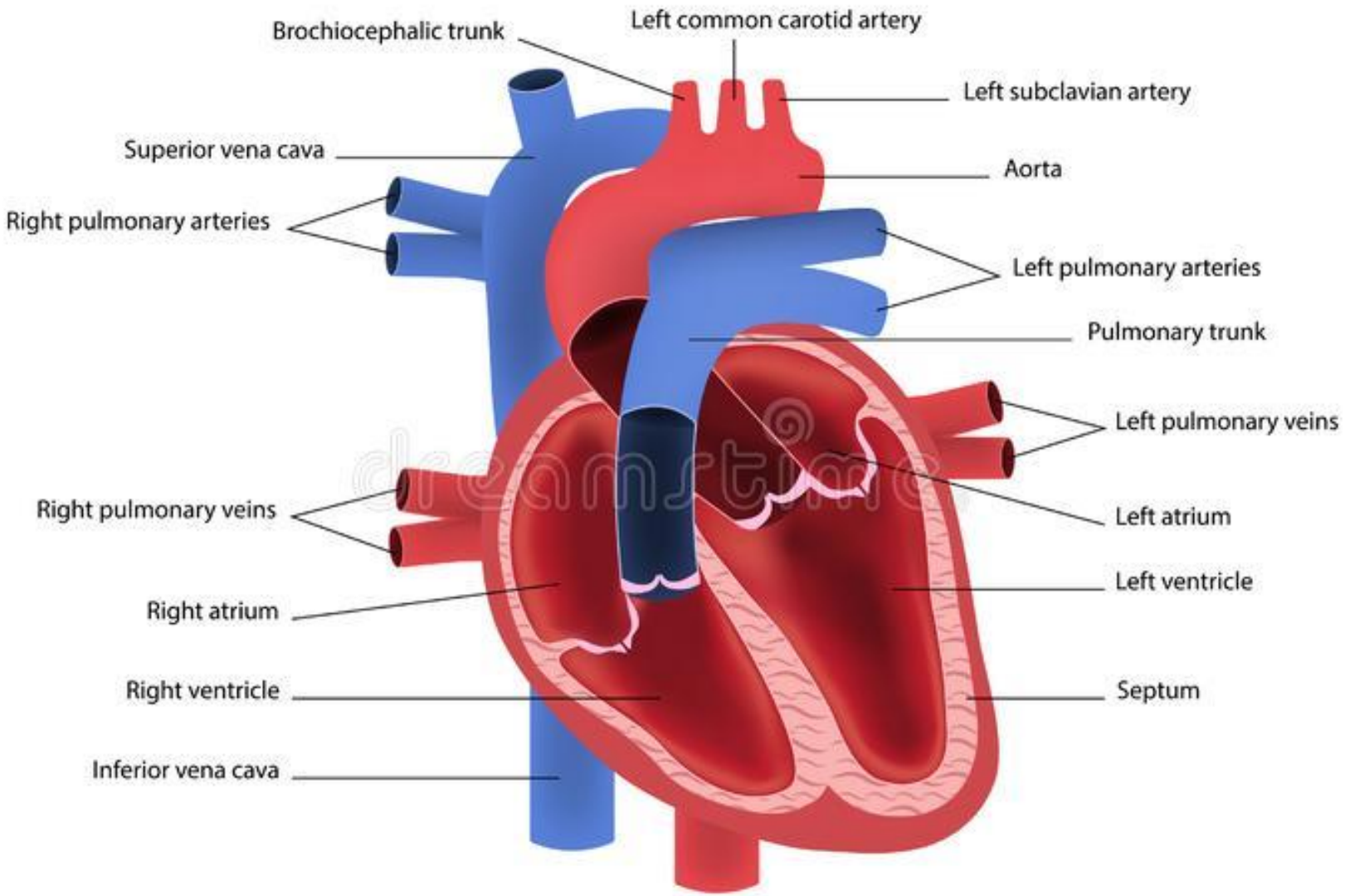
- NORMAL HEART ANATOMY

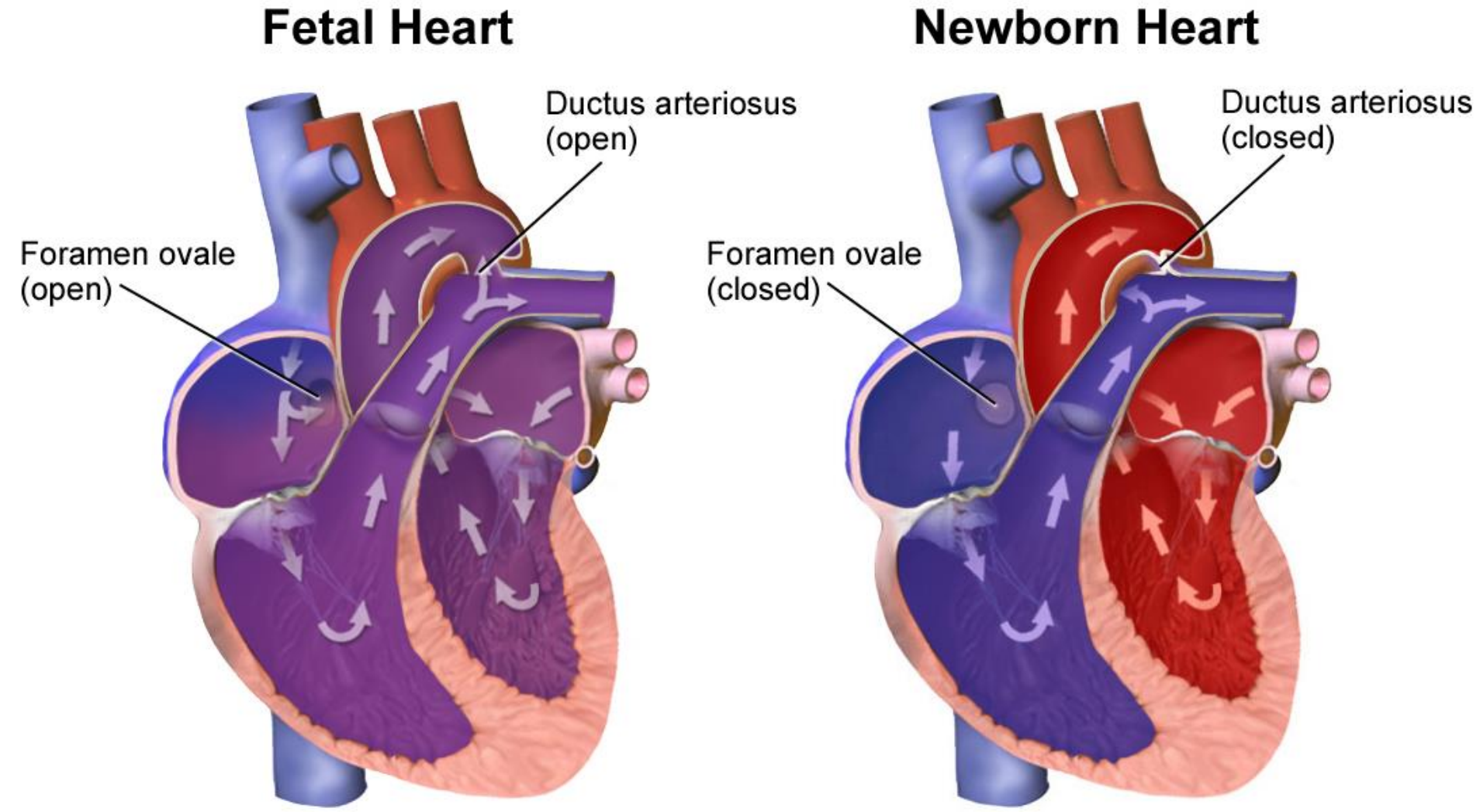
- FETAL CIRCULATION:

- low pulmonary flow in fetus
- foramen ovale in atrial septum
- ductus arteriosus - Botall's duct (connects pulmonary trunk with aorta descending)

- CHANGES IN CIRCULATION AT BIRTH

Normal heart anatomy





Fetal heart anatomy and changes after birth: https://www.youtube.com/watch?v=m3p5PsB6aZ4

https://upload.wikimedia.org/wikipedia/commons/7/76/Neonatal_Heart_Circulation.png

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Symptoms suggesting heart disease after birth

- Medical history: impairment of effort tolerance, fatique when feeding, sweating, loss of appetite (problems with weight gaining), reccurent respiratory system infections

- Examination: weight gain failure, dyspnoe, abnormal respiration (tachypnoe), tachycardia, heart murmurs, cardiomegaly, hepatomegaly, peripheral oedema, cold extremities, clubbed fingers, hypotonia, delayed development especially motor retardation

Heart murmur

Normally, there shouldn’t be any murmur

Murmurs are caused by abnormal blood flow through:

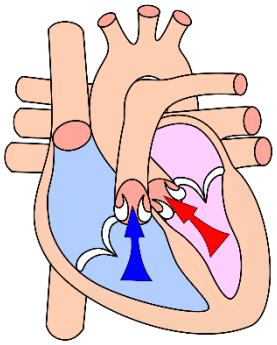
- too narrow valves (stenosis)
- or when there is leakage through the valve (regurgitation)
- or when there is an extra way of the blood flow e.g. the shunt between the left and right ventricle (ventricular septum defect)

Some murmurs may also occur during accelerated blood flow (hyperkinetic) e.g. in course of anemia or hyperthyroidism

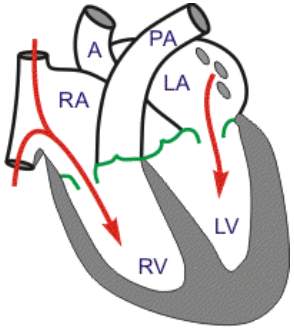
Heart murmur

During heart auscultation we hear 2 major tones:

- S1 – it is a sound of atrioventricular valves closure
- S2 – it is a sound of arterial valves closure



Time between S1 and S2 is a systolic phase (ventricular contraction)



Time between S2 and S1 is a diasystolic phase (blood flows from atrium to the ventricle)

The most important division of the heart murmurs is into:

- Systolic (heard during ventricular contraction)
- Diastolic

Heart sounds and murmurs explained

https://www.youtube.com/watch?v=wYZbMoWjLEg&t=20s

HEART MURMURS

- **Heart murmur :**
- Innocent heart murmur
- Pathological murmur

- Features of the innocent heart murmur
- Patient without pathological symptoms or heart defect
- Murmur is „soft”, only systolic
- Audible at the left edge of the sternum without radiation

Innocent heart murmur: fever, anemia

Levine Scale of heart murmurs

- 1/6 - very silent
- 2/6 - soft, audible but silent
- 3/6 - audible, but without the thoracic tremor
- 4/6 - audible, with thoracic tremor - „thrill”
- 5/6 - loud, audible with stetoscope slightly closed to the thorax
- 6/6 - very loud, audible even without the stetoscope

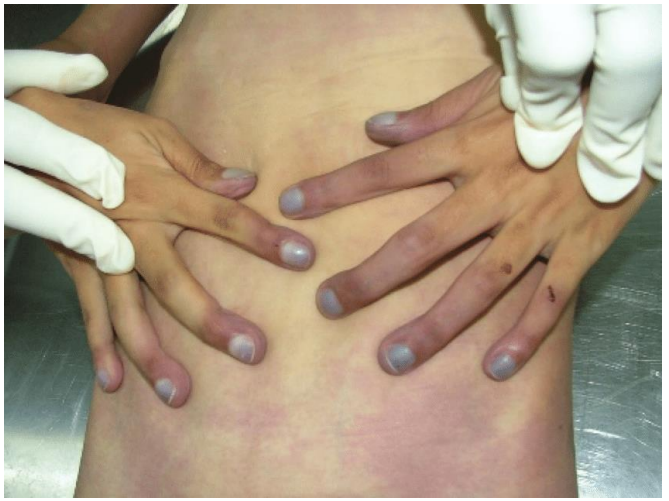
Cyanosis

- Peripheral cyanosis (hands and feet)
- Central cyanosis (tongue, mucous membranes)

- Occurs when concentration of deoxygenated haemoglobin is over 5 g%

- Evaluation of oxygenation of haemoglobin on skin colour is unreliable

- Saturation measured by oximeter (N ≥ 94%)



Diagnosis

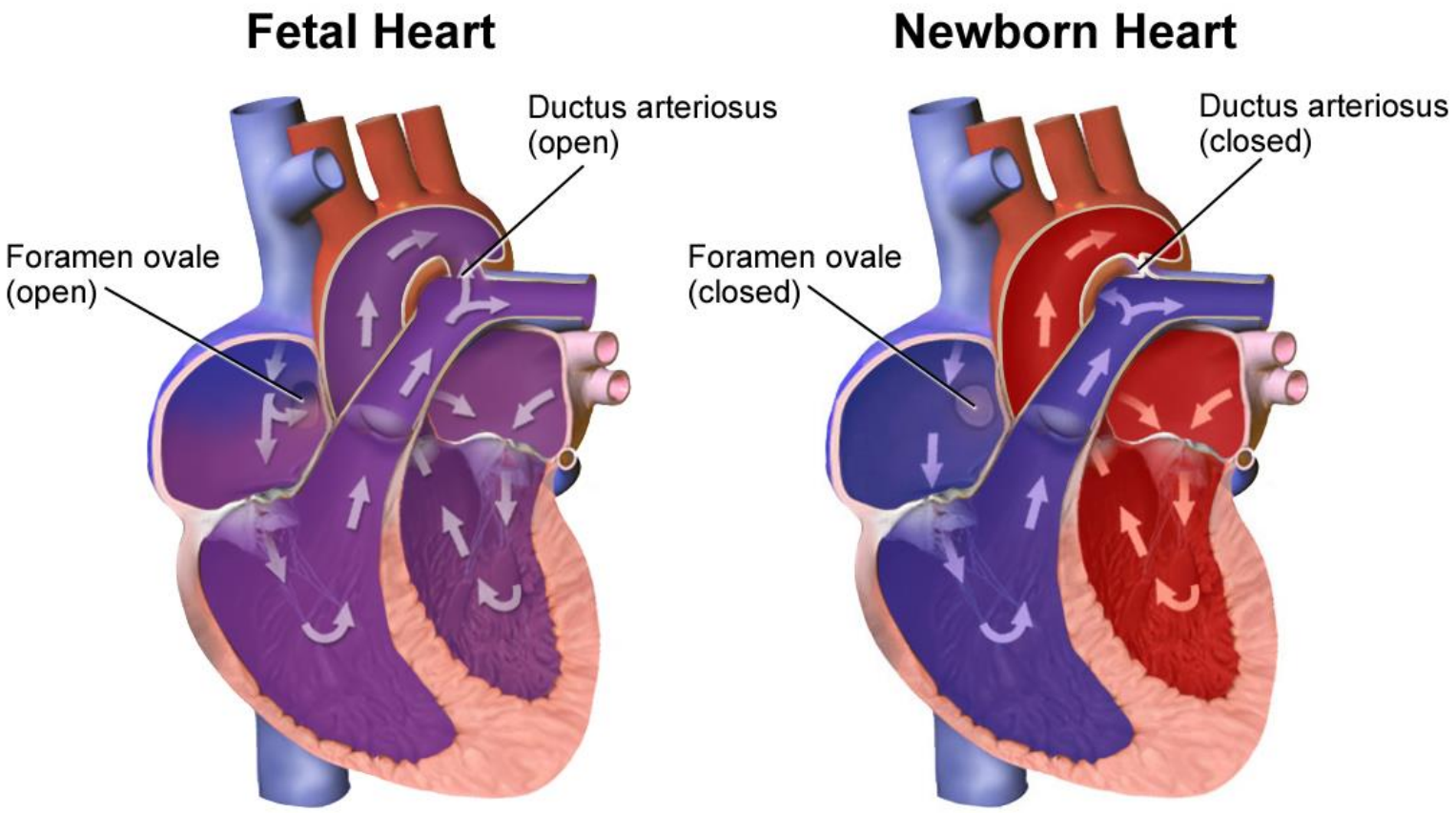
- Prenatal diagnosis : 18-20 hbd - usg of fetus to evaluate heart structure
- Mother with heart defect - echocardiography in fetus!
- **ECHO + doppler**
- History case, clinical features of heart failure, heart murmurs, cyanosis, cardiogenic shock
- Examination by: watching, palpating, percussion, auscultation
- Additional examinations: laboratory (NT- proBNP increased in heart failure), ECG, chest X-ray, magnetic resonanse imaging of the heart, cardiac catheterizationand angiography, isotopic angiography

Classification of heart defects

- The most common division: cyanotic and non-cyanotic
- With normal, increased or decreased pulmonary blood flow
- With or without blood leaking (left to right or right to left)

Cyanotic heart defects (Duct dependent)

- Tetralogy of Fallot– TOF
- Transposition of the Great Arteries – TGA
- Common Arterial Trunk (Truncus Arteriosus) - TA
- Total Anomalous Pulmonary Venous Drainage - TAPVD
- Tricuspid atresia - AT
- Hypoplastic Left Heart Syndrome - HLHS



After birth: increased pressure in aorta -> left – right shunt through ductus arteriosus
In cyanotic heart defects preservation of ductus arteriosus is sometimes the only way to provide blood flow to the lungs and sudsequent oxigenation

https://upload.wikimedia.org/wikipedia/commons/7/76/Neonatal_Heart_Circulation.png

Non-cyanotic heart defects

- Patent Ductus Arteriosus - PDA
- Atrial Septal Defect - ASD
- **Ventricular Septal Defect - VSD** (the most common heart defect in children)
- Atrioventricular Septal Defect - AVSD
- Coarctation of the Aorta - CoAo
- Critical Aortic Stenosis Ao –AS
- Critical Pulmonary Stenosis - PS

Congenital heart defects with symptoms occurrence in the first 3 days of life

- Transposition of the Great Arteries – TGA
- Hypoplastic Left Heart Syndrome - HLHS
- Critical Aortic Stenosis Ao –AS
- Critical Pulmonary Stenosis or Atresia - PS/PA
- Interruped Aortic Arch Ao - IAA

Congenital heart defects with symptoms occurrence between 4 and 14 days of life

- Tetralogy of Fallot – TOF
- PS/PA
- TGA+VSD+PS
- Coarctation of the Aorta (CoAo)

The most common congenital heart defects

Defects with left- to-right shunt (dyspnea is the main symptom)

Ventricular septal defect (VSD) (30% of defects)

Patent ductus arteriosus (PDA) (12%)

Atrium septal defect (ASD) (7%)

Defects with right-to-left shunt (cyanosis is the main symptom)

Tetralogy of Fallot (5%)

Transposition of the great arteries (TGA) (5%)

Defects with blood mixing (dyspnea and cyanosis)

Atrioventricular septal defect (AVSD) (2%)

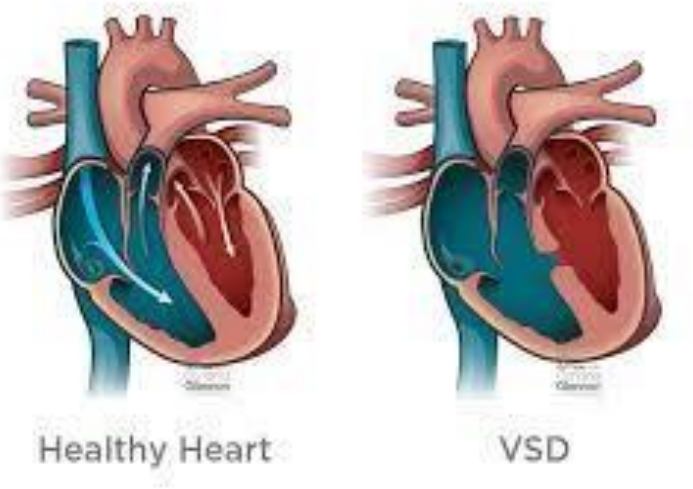
Defects of the outflow in the patient in good general condition (heart murmur):

- Pulmonary stenosis (PS) (7%)
- Aortic stenosis (AoS) (5%)

Defect of the outflow in the patient with shock

- Coarctation of the Aorta (CoA) (5%)

VSD - ventricular septal defects



- VSD: perimembranous or muscular

Small VSD (3 mm) (80-90% cases)

- Can be without symptoms
- Murmur – loud, systolic, at the left edge of the sternum
- Usually is spontaneously recovered

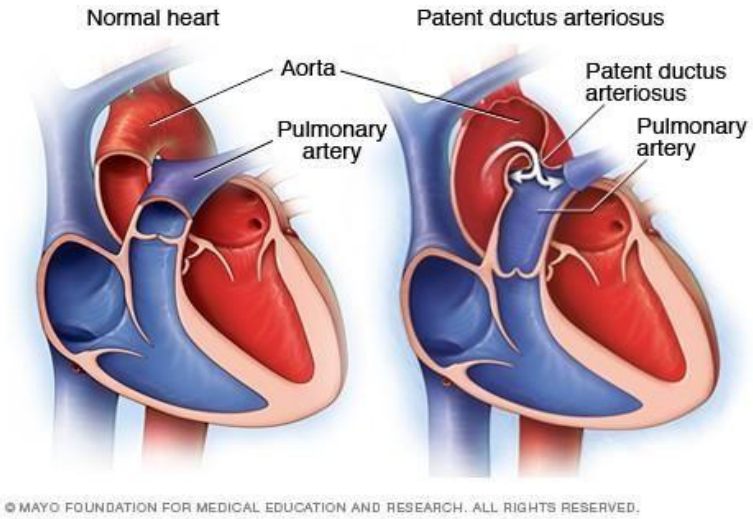
Large VSD (10-20% cases) (left to right shunt)

- Features of heart failure, silent murmur, tachypnoe, hepatomegaly,
- Loud second heart sound
- Chest X-ray : cardiomegaly, enlarged of pulmonary artery, intensified lung markings

- Treatment: diuretics, kaptopryl, cardiac surgery in 3-6 months of life

Patent Ductus Arteriosus – PDA

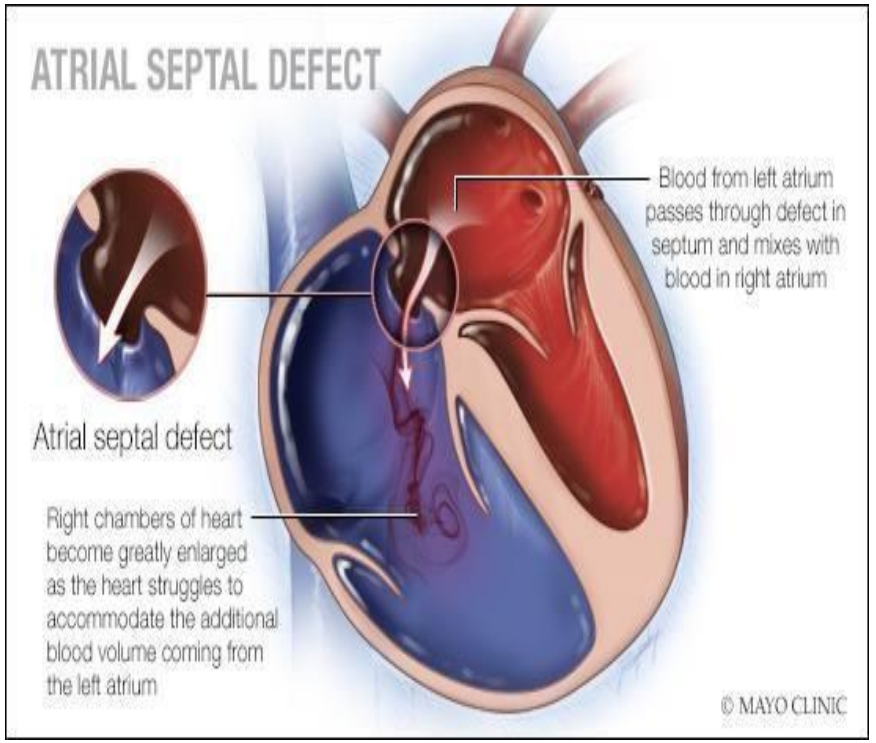
- Ductus arteriosus - Botall's duct – in fetus connects pulmonary trunk with aorta descending
- Normally it is closed within a few hours/days after birth
- 10% of heart defects in mature newborns
- 40% of heart defects in premature newborns born in 25 –27 week pregnancy
- Occurs 3 times more frequent in girls



- systolic-diastolic murmur in the second intercostal space in the left
- Chest X-ray and ECG can be normal; when PDA is significant the signs as in VSD
- Treatment: non-invasive (coil) or cardiac surgery

ASD- Atrial septal defect:

- ASD type II (80% ASD) defect in the middle part of the septum (foramen ovale)
- ASD type I (loss of the part of atrial septum)



Sings and symptoms of ASD :

- often asymptomatic
- recurrent respiratory system infections
- arhythmia
- systolic murmur at the upper left edge of the sternum or silent apex (ASD I)
- Split second pulmonary sound (ASD I)

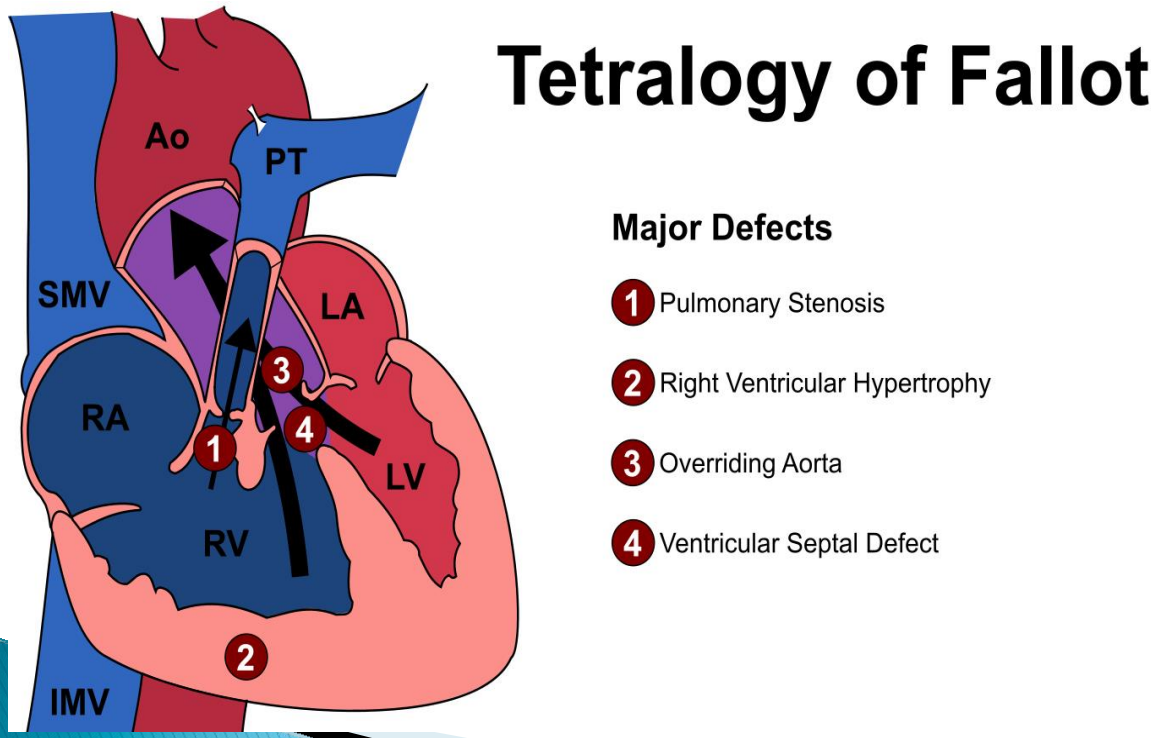
Treatment:

ASD type II – implant

ASD type I – cardiac surgery

Tetralogy of Fallot (ToF)

- **The most frequent cyanotic heart defect**
- **TETRALOGY - 4 components:**



Signs and symptoms:

- cyanosis,
- dyspnea,
- anoksemic attacks,
- fatigability,
- heart murmur,
- clubbed fingers

Treatment of heart defects

Duct-dependent CHD:

- Infusion of prostaglandin PGE1 immediately after labour – maintain the flow in patent ductus arteriosus

} Cardiac surgery

} In case of suspicion of congenital heart defect, patient should be referral and treated at the specialist centres

PROPHYLAXIS OF INFECTIVE ENDOCARDITIS

„2015 ESC Guidelines for the management of infective endocarditis „

The Task Force for the Management of Infective Endocarditis of the European Society of Cardiology (ESC)

Published: European Heart Journal (2015) 36, 3075–3123

- Antibiotic prophylaxis should only be considered for dental procedures requiring manipulation of the gingival or periapical region of the teeth or perforation of the oral mucosa

Class of recommendation: II a - Weight of evidence/opinion is in favour of usefulness/efficacy

Level of evidence: C - Consensus of opinion of the experts and/ or small studies, retrospective studies, registries

PROPHYLAXIS OF INFECTIVE ENDOCARDITIS

Cardiac conditions at highest risk of infective endocarditis for which prophylaxis should be considered when a high-risk procedure is performed:

1. Patients with any prosthetic valve, including a transcatheter valve, or those in whom any prosthetic material was used for cardiac valve repair.
2. Patients with a previous episode of IE.
3. Patients with Congenital Heart Disease (CHD):

(a) Any type of cyanotic CHD.

(b) Any type of CHD repaired with a prosthetic material, whether placed surgically or by percutaneous techniques, up to 6 months after the procedure or lifelong if residual shunt or valvular regurgitation remains.

PROPHYLAXIS OF INFECTIVE ENDOCARDITIS

- Single-dose 30–60 minutes before procedure

- No allergy to penicillin or ampicillin:

Amoxicillin or ampicyllin 50 mg/kg orally or i.v. (2 g for adults)

- Allergy to penicillin or ampicillin::

clindamycin 20 mg/kg orally or i.v. (600mg for adults)

*Alternatively, cephalexin 50 mg/kg i.v. for children (2 g for adults), cefazolin or ceftriaxone 50 mg/kg i.v. for children (1 g for adults). Cephalosporins should not be used in patients with anaphylaxis, angio-oedema, or urticaria after intake of penicillin or ampicillin due to cross-sensitivity.

Thank you for your attention