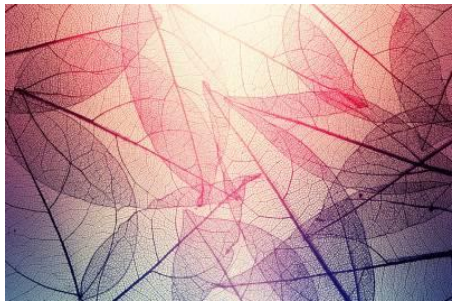
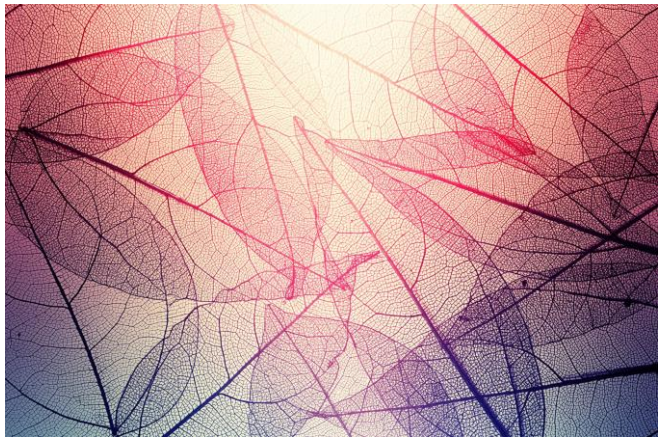


Pediatric cardiology

MUDr. Lenka Majerová

Department of Pediatrics and Inherited Metabolic Disorders, General University Hospital in Prague



The most common diagnoses

- **Congenital heart diseases (ventricular septal defect)**
- Cardiomyopathies (hypertrophic, dilated)
- Inflammatory diseases (endocarditis, myocarditis, pericarditis)
- Arrhythmias (supraventricular tachycardia)
- Systemic arterial hypertension (secondary)
- Pulmonary hypertension (persistent pulmonary hypertension in a newborn)
- Heart tumors

When setting the diagnosis...

- Medical history
- Physical examination - clinical signs and symptoms
- Imaging studies:
  - ECG
  - chest scan
  - **echocardiography**
  - CT
  - MRI
  - Cathetrisation
- Cardiac stress test (ergometry)

Medical history

- **Family** – genetics (syndromes, familiar inheritance of VCC)
- **Prenatal infections** (TORCH), **drugs** (ibuprofen), **X-rays**
- **Prematurity** – persistent ductus arteriosus, foramen ovale apertum
- **Problems with feeding (breaks), failure to thrive**
- **Frequent respiratory infections**
- **Fatigue** (at rest/on exercise)
- **Excessive sweating**
- **Syncope**
- **Chest pain** (often of musculoskeletal origin)
- **Palpitations**

Physical examination (aspection, palpation, auscultation)

The most common clinical signs and symptoms

- Dyspnea, tachypnea
- Cyanosis
- Murmurs
- Heart failure
- Pulmonary arterial hypertension (Eisenmenger syndrome)

Dyspnea and tachypnea

- Dyspnea – subjective feeling of shortness of breath
  - ***Objectification! Newborns & toddlers:***
    - Using of accessory breathing muscles resulting in to retraction of the belly, jugulum, intercostal spaces and/or nasal flaring

Sternal retractions - Labored breathing - Wikipedia

- Tachypnea
  - ***The age of a patient!***

A. Newborn to 2 months: 60 breaths per minute

B. Infant 2 months to 1 year: 50 breaths per minute

C. Preschool Child 1 to 5 years: 40 breaths per minute

D. School age Child: 20-30 breaths per minute

E. Adults: 20 breaths per minute

Cyanosis

- Bluish colouring of the skin and/or mucous membranes; >50g deoxygenated Hb

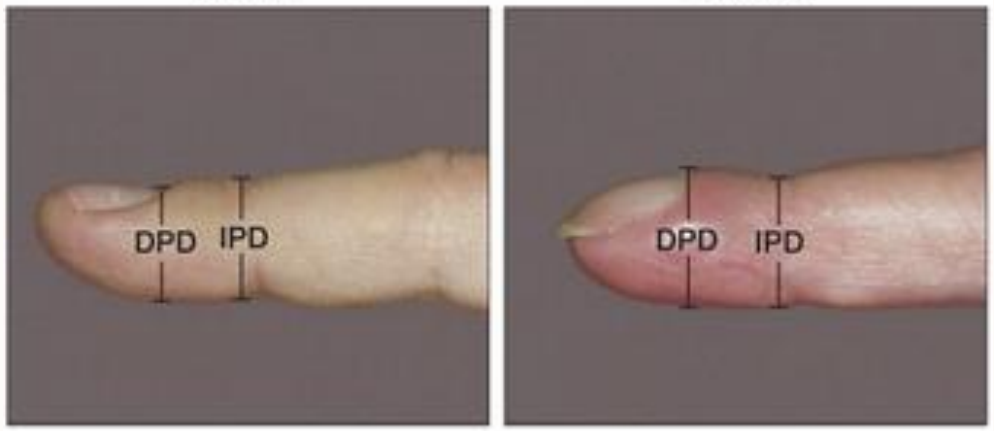
- Central vs. peripheral

Central cyanosis – „warm“

- decreased satO2 in arterial blood
- tongue and mucous membranes
- polyglobulia, clubbing („drumstick fingers“), thromboembolism

Normal

Clubbed

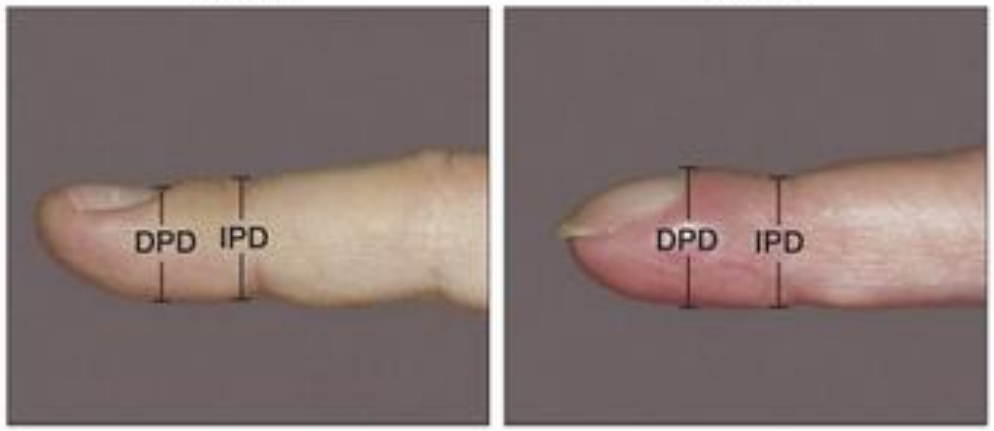


Peripheral cyanosis – „cold“

- low cardiac output and other shocks
- satO2 in arterial blood normal
- decreased venous satO2
- acrocyanosis

Normal

Clubbed



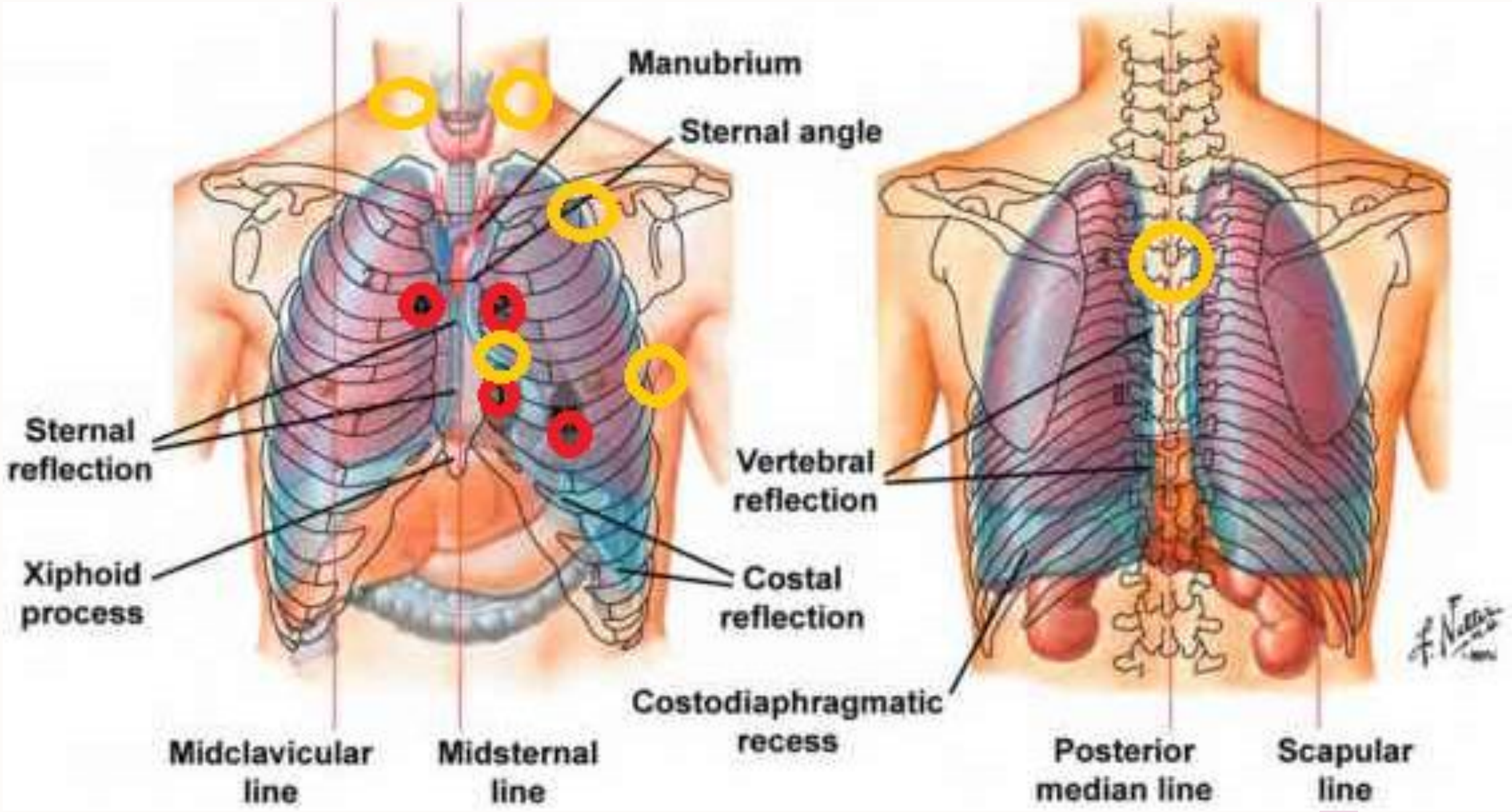
Asessment of the heart rate according to the age

Normal range of HEART RATE For children

AGE	RESTING HEART RATE (BPM)
Newborn (0-1 month)	70-190
Infant (1-11 months)	80-160
Toddler (1-3 years)	80-130
Preschooler (3-5 years)	80-120
School-aged child (6-12 years)	70-110
Adolescent (12-18 years)	60-100

Murmurs

- **Additional sound** to the normal heart sound caused by **turbulent flow** in the heart
- Organic vs. innocent (physiologic)
- Description of a murmur
    1. timing (systole/diastole/continuous)
    2. punctum maximum
    3. intensity – Levin‘s scale (1/6 – 6/6)
    4. propagation


Manubrium
Sternal angle
Sternal reflection
Xiphoid process
Midclavicular line
Midsternal line
Costodiaphragmatic recess
Costal reflection
Vertebral reflection
Posterior median line
Scapular line

Innocent murmur = physiologic

- 70 % of healthy children (toddlers, preschool age)
- Always systolic
- Max 3/6
- No thrilling
- Changing with body position
- No clinical symptoms
- No ECG changes

Heart failure

- Progressive condition in which the heart muscle is unable to pump enough blood to meet the body's needs for blood and oxygen
- Acute vs. chronic
- Forward vs. backward
- Right-side vs. left-side
- Compensated vs. decompensated

Forward failure

- Similar for right and left side, right side => lower pessure => better tolerance
- Tiredness, intolerance of excersice; left side – also syncope
- Paleness
- Peripheral cyanosis, cold and sweaty periphery
- Capillary refill longer than 3 sec
- Kidney failure, hyperkaleamia
- Necrotising enterocolitis (acute abdominal syndrome)

Right side backward

- Hepatosplenomegaly (liver >2cm below the last rib in the right midclavicular line)
- ↑ hepatojugular reflux when pushing below the liver
- Oedemas, ascites (small children – genitalia, bigger children - legs)
- Tachypnea, dyspnea
- Failure to thrive; pale, sweaty skin, cutis marmorata (bluish mottling of the skin)

Left side backward

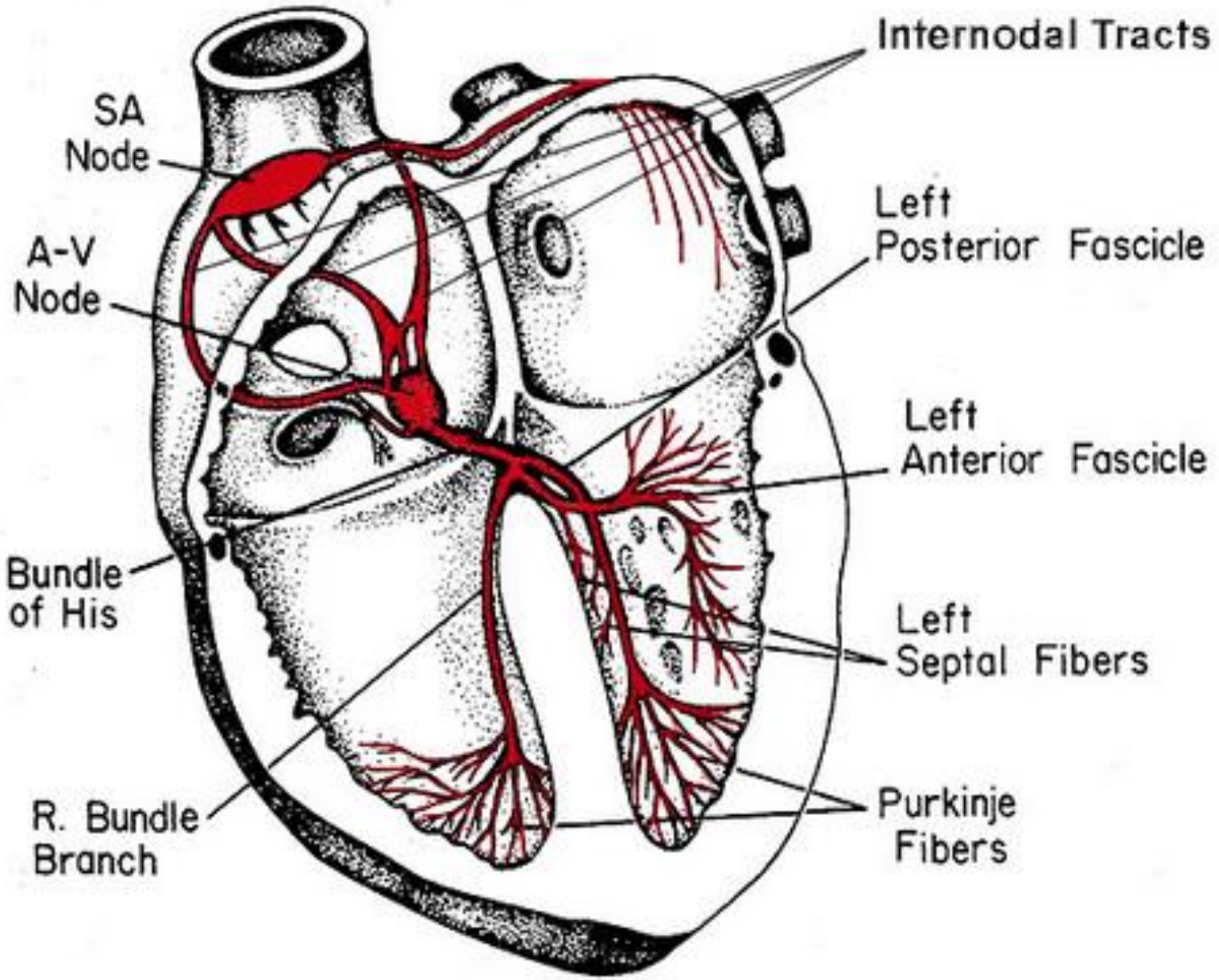
- Tachypnea, dyspnea
- Pulmonary oedema

Imaging studies

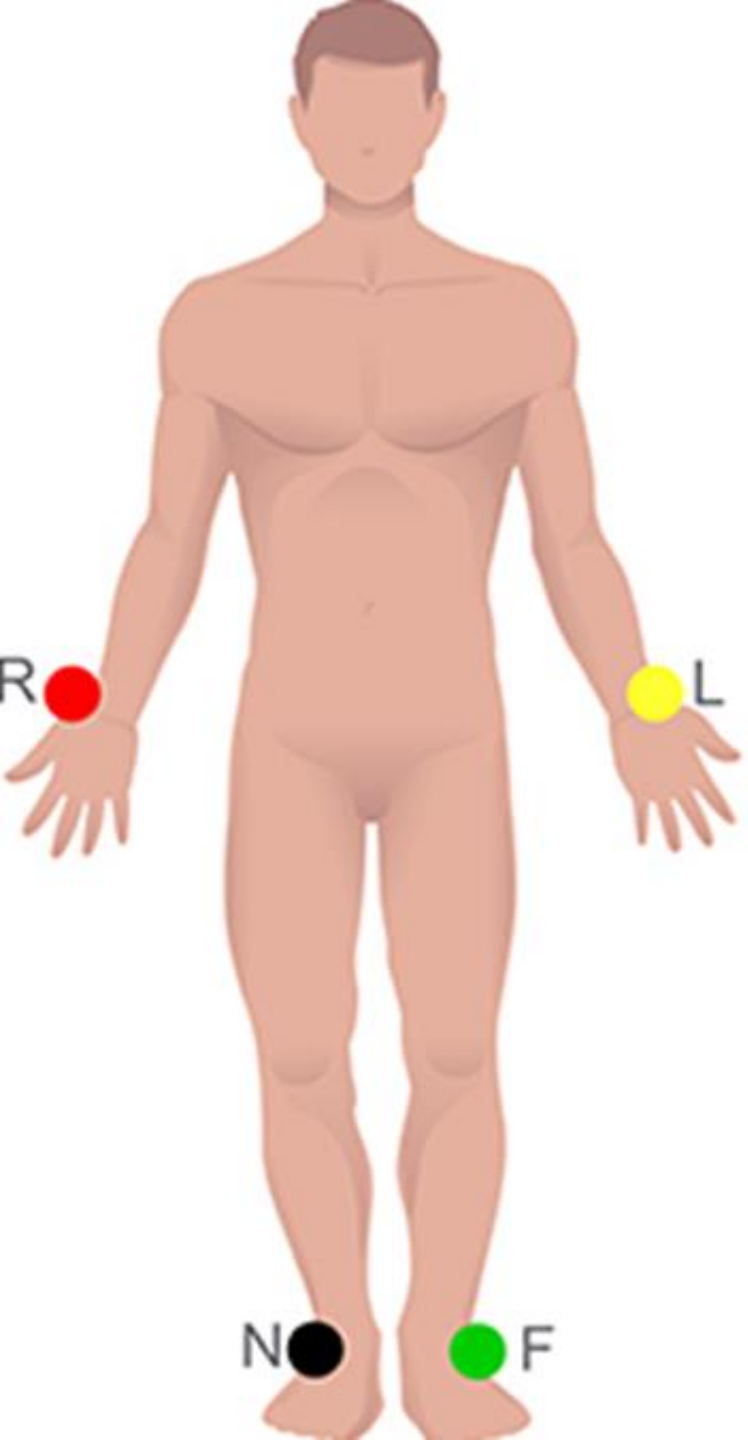
ECG

- Electric potentials detection

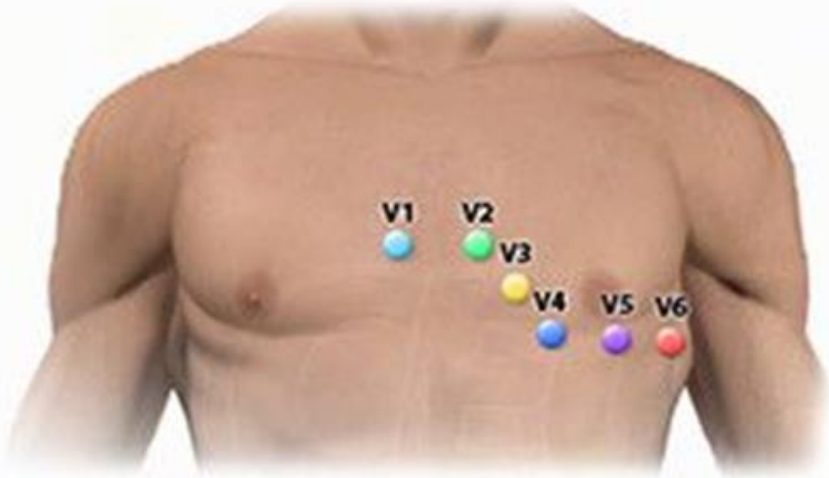
- Types of ECG detection:
  - 12-lead ECG
  - ECG Holter (usually 24 hours)
  - event recorder (implanted subcutaniously)

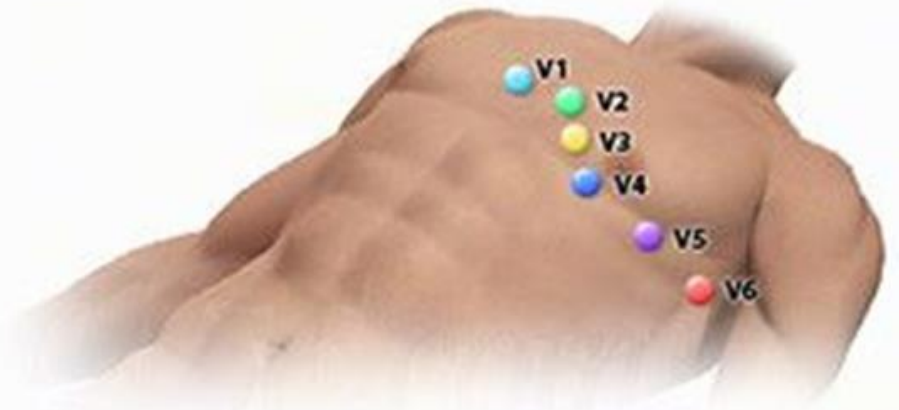


ECG electrodes placement

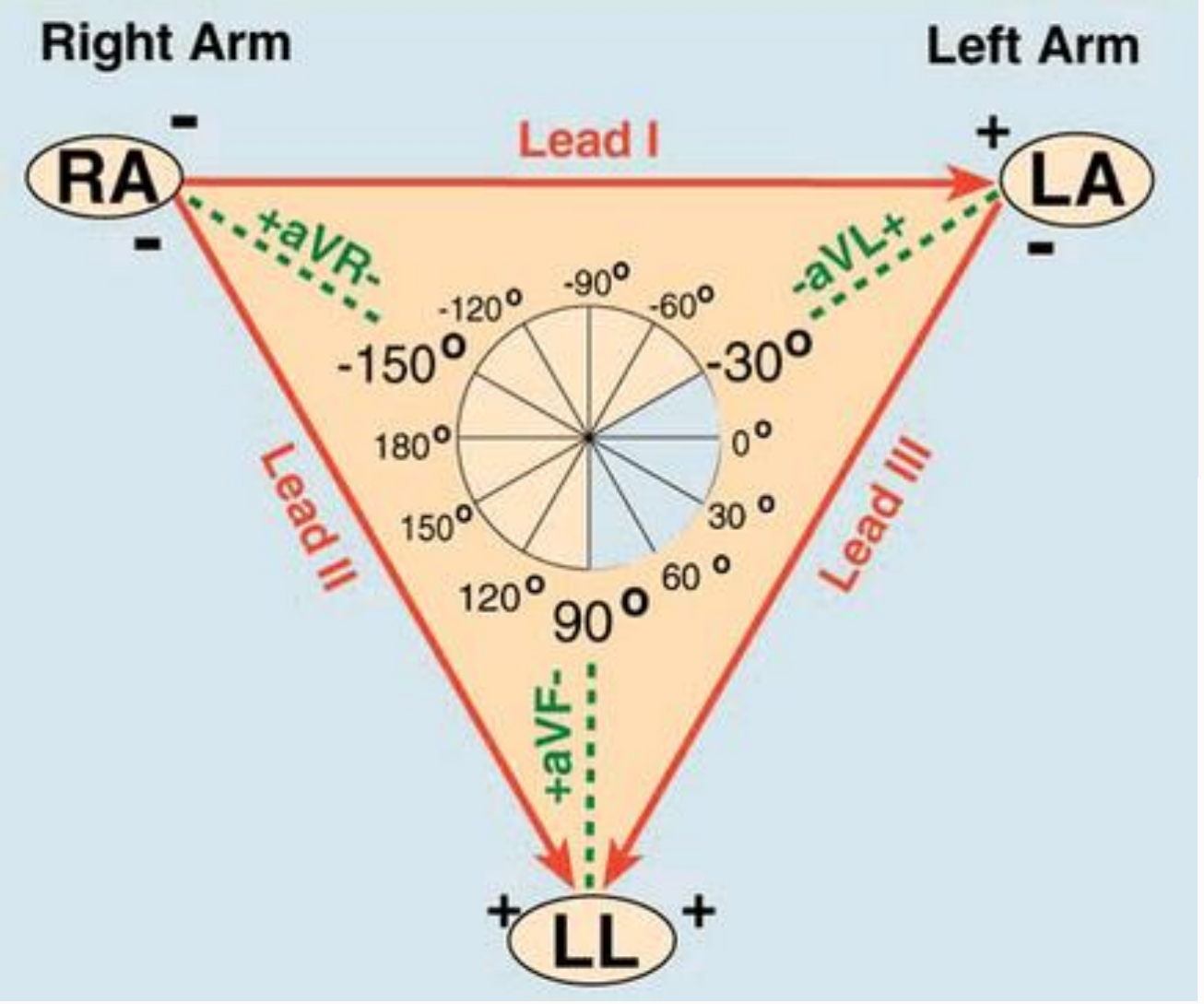


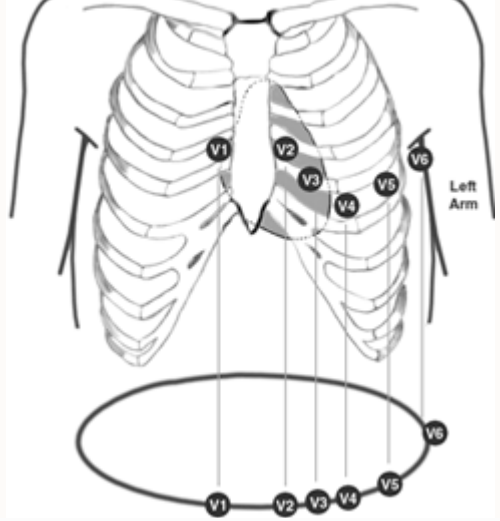
Precordial Leads (chest lead placement)



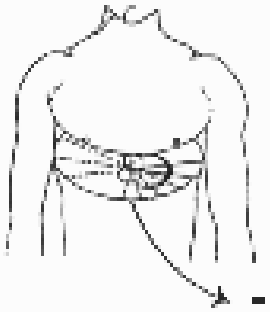


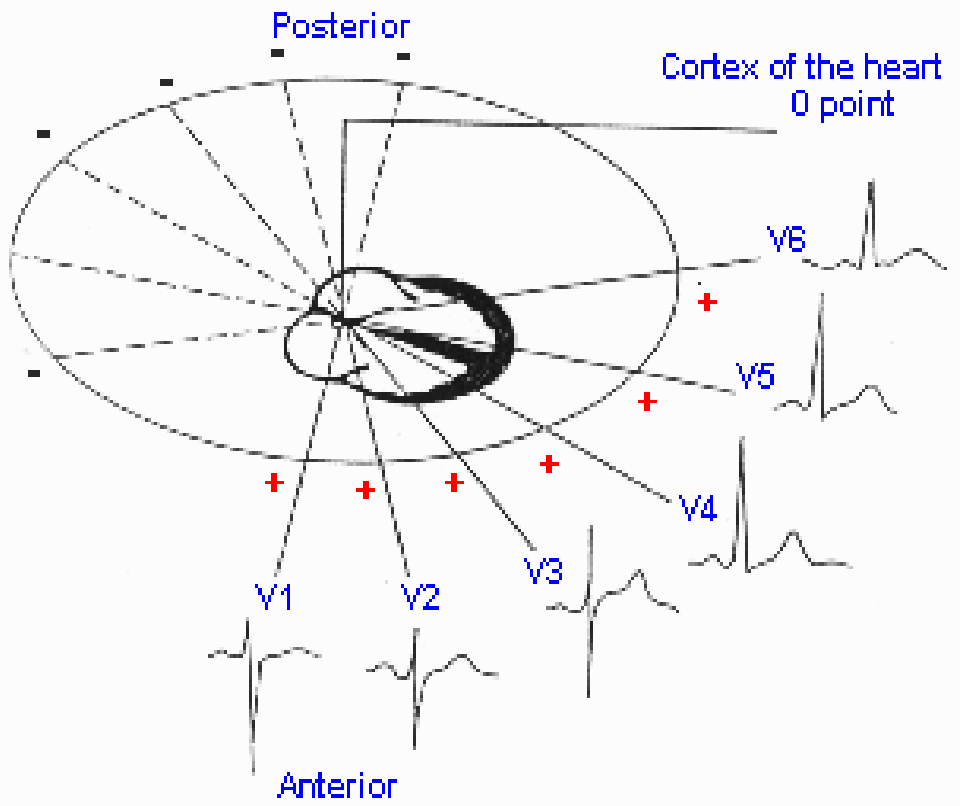
- V1 - 4th intercostal space to the right of the sternum
- V2 - 4th intercostal space to the left of the sternum
- V3 - Halfway between V2 and V4
- V4 - The left midclavicular line in the 5th intercostal space
- V5 - The left anterior axillary line at the same horizontal level as V4
- V6 - The left midaxillary line at the same horizontal level as V4 and V5

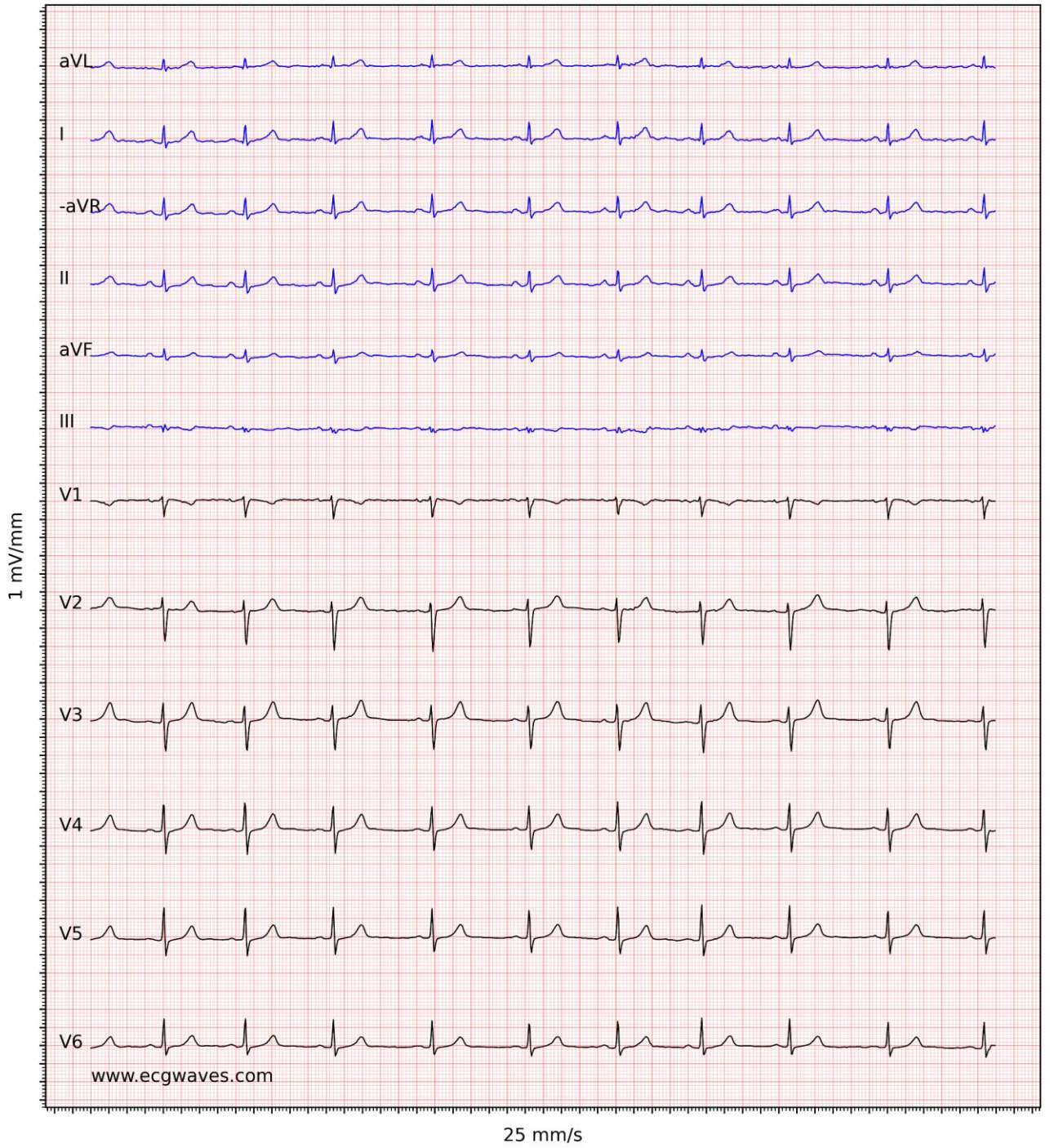


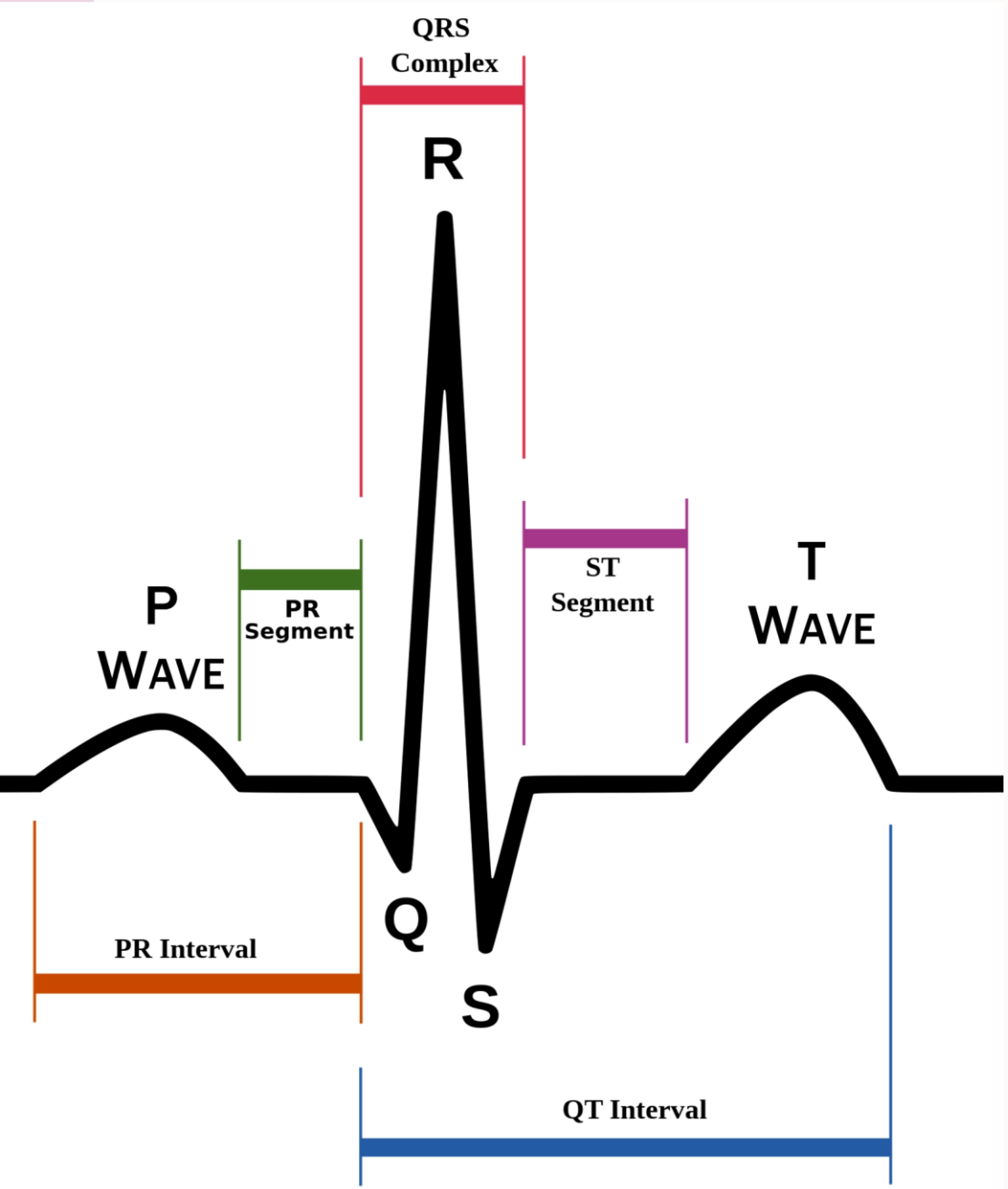


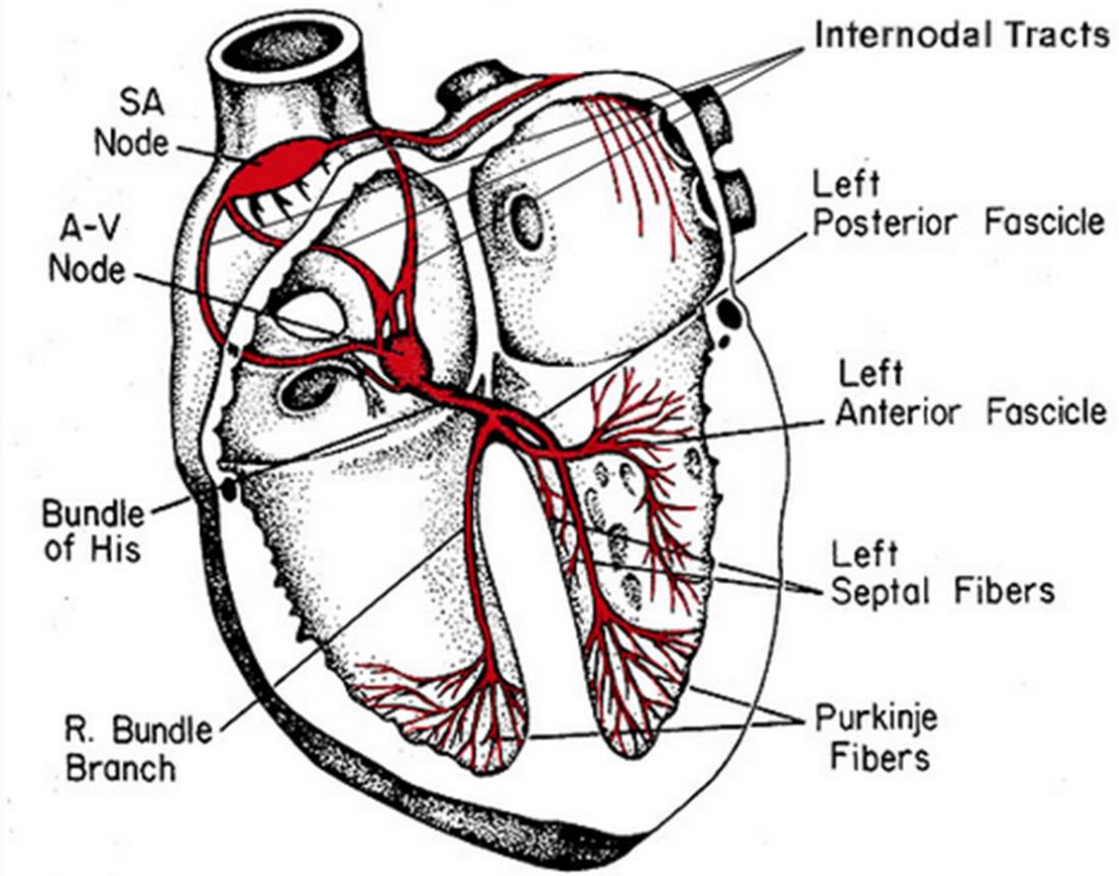
Horizontal Plane






aVL
I
-aVR
II
aVF
III
V1
V2
V3
V4
V5
V6
1 mV/mm
www.ecgwaves.com
25 mm/s





- **P wave** – depolarisation of atria
- **PQ (PR) interval** – the time between atrial depolarisation and ventricular depolarisation
- **QRS complex** – depolarisation of the ventricles
- **ST segment** (isoelectric line) - the region between the end of ventricular depolarisation and beginning of ventricular repolarisation

ECG evaluation

- Regularity – regular/irregular
  - In children often respiratory arrhythmia => higher frequency during inspiration, slower during exspiration - physiologic
- Rhythm – sinus/sinus-like/atrial/junctional
  - Sinus rhythm – positive P waves in I, II, III, aVF before every QRS complex
  - In children often sinus-like/atrial which is physiologic
- Frequency (evaluation of tachy- / bradycardia according to the age – charts)
  - 3000 / number of small squares in 50 mm/s – in children more comonlly used speed of paper 50 mm/s
  - 1500 / number of small squares in 25 mm/s
- Electric axis
- Intervals – PQ, QRS, QTc (according to the age, frequency respectively – charts)
- Morfology and QRS voltage
- ST segments changes
- T waves

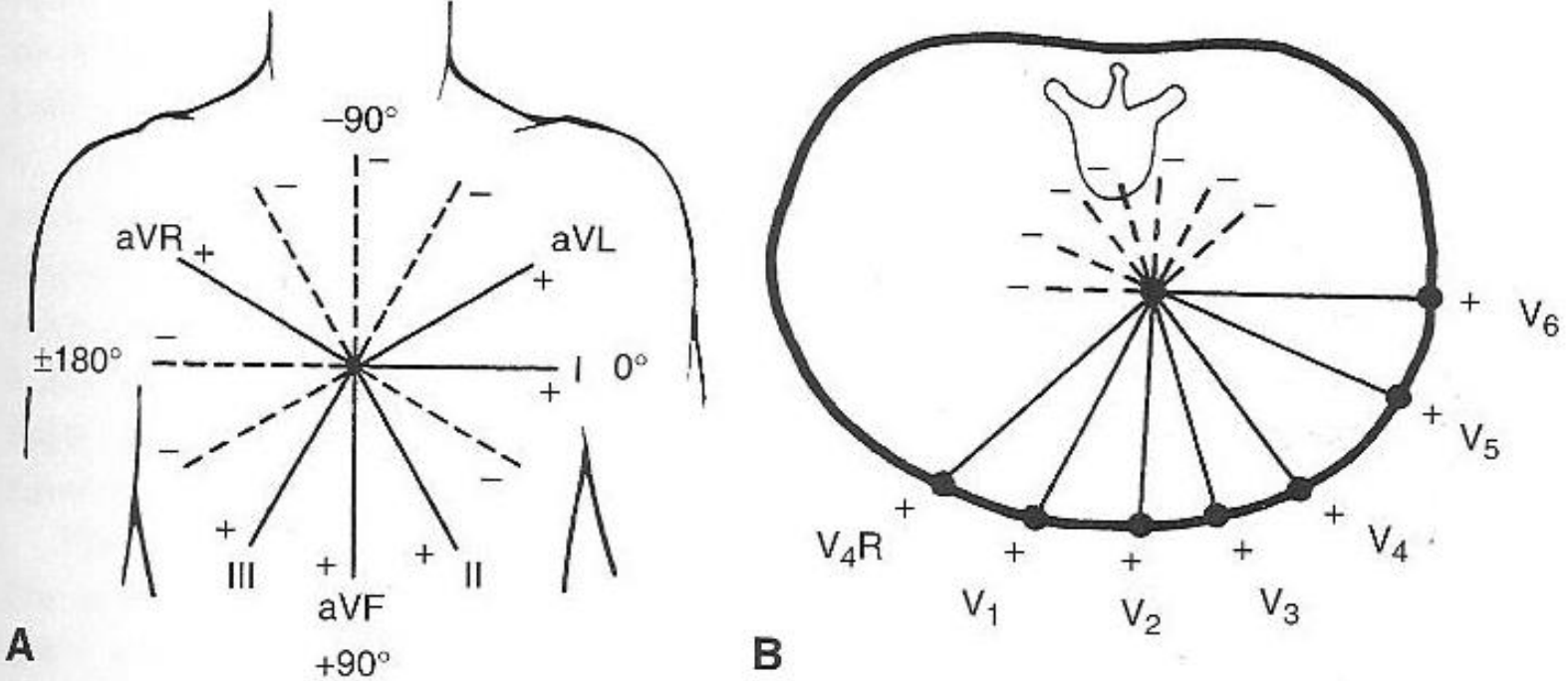
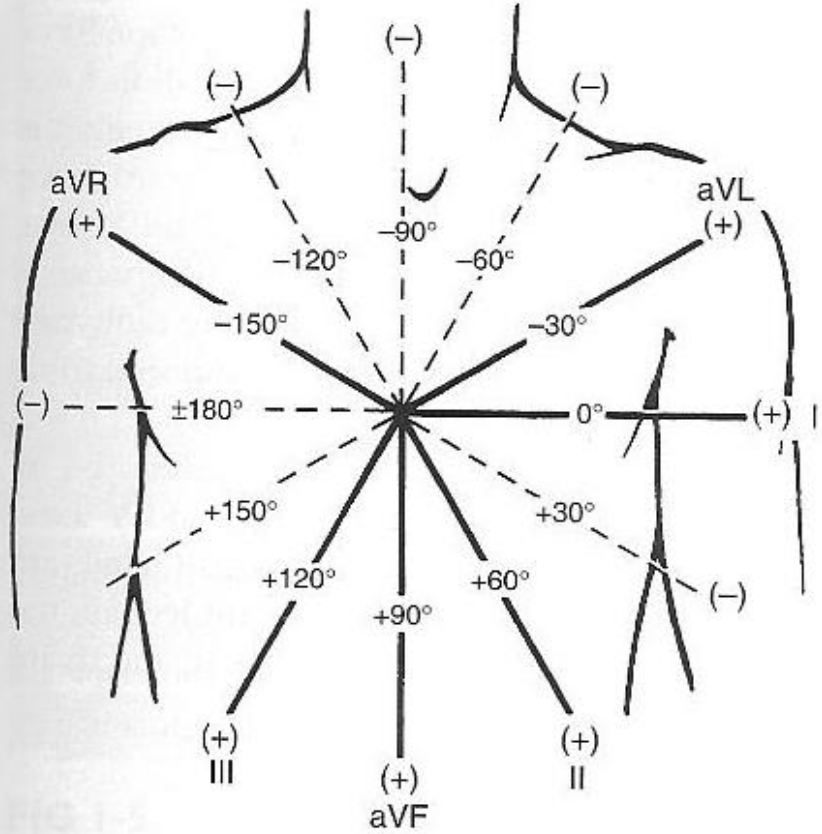


FIG 1-2.

Hexaxial **(A)** and horizontal **(B)** reference systems. The combination of **A** and **B** constitutes 12- (or 13-) lead ECG.



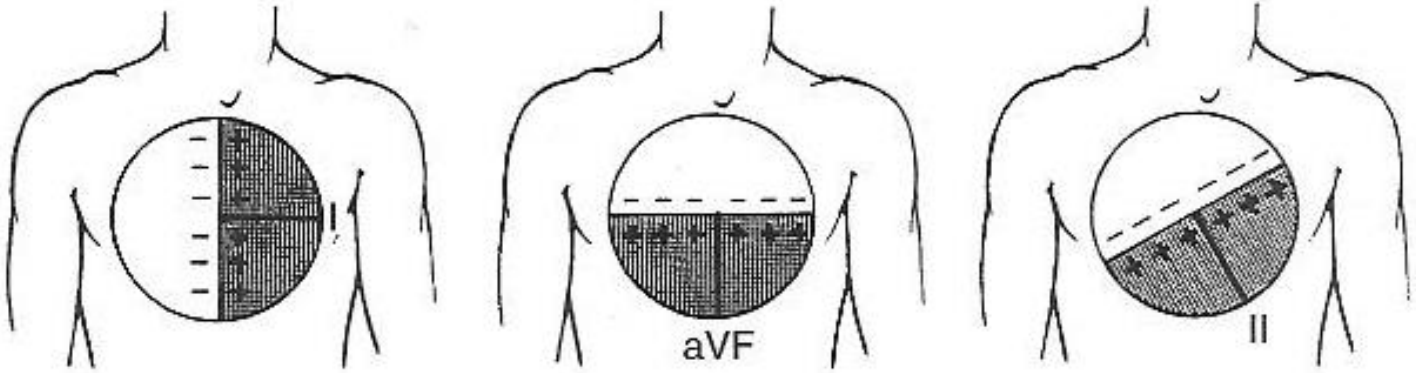


FIG 2-8.
Polarity of limb leads (viewed from the patient's front). Lines perpendicular to leads I, aVF, the areas into positive *(shaded)* and negative *(unshaded)* segments for polarity.

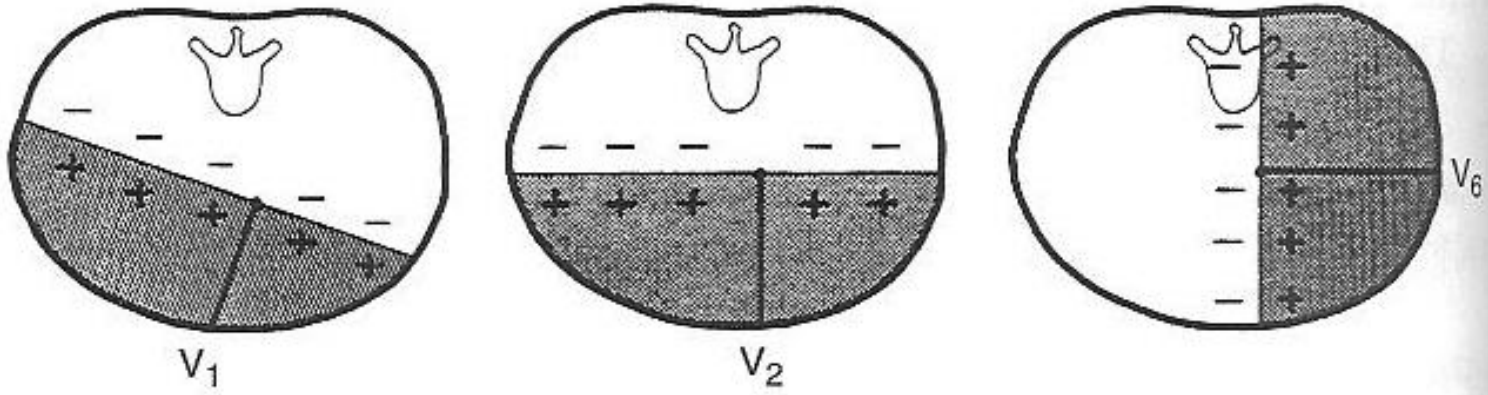
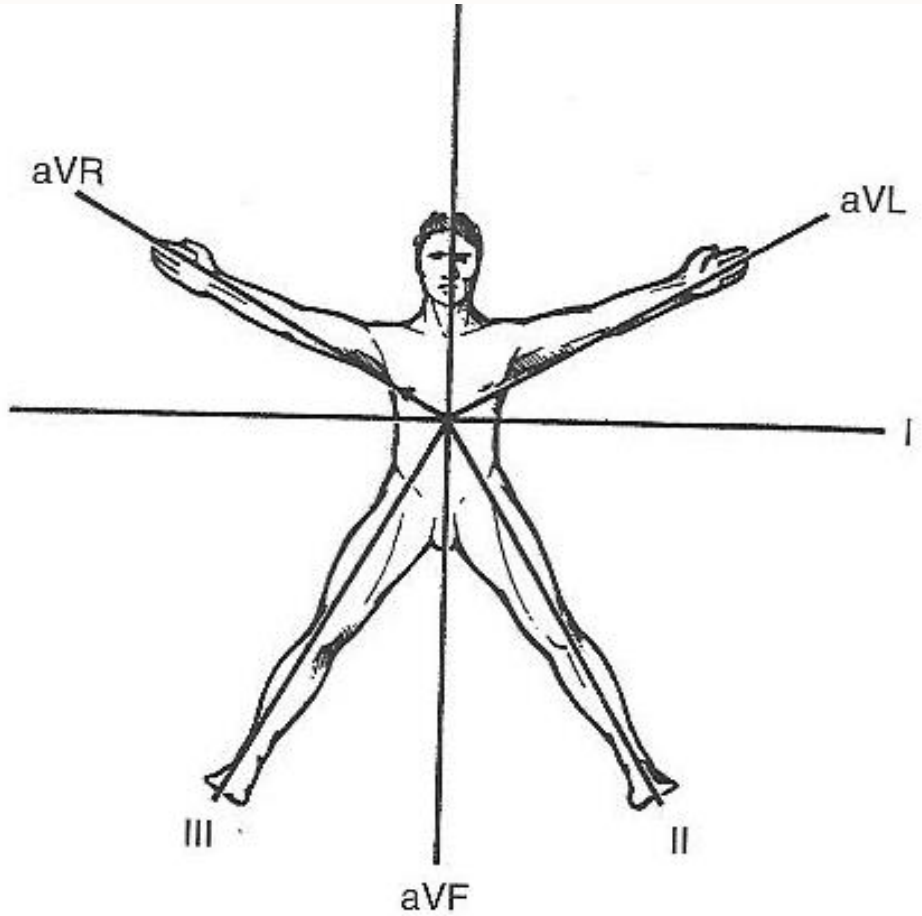
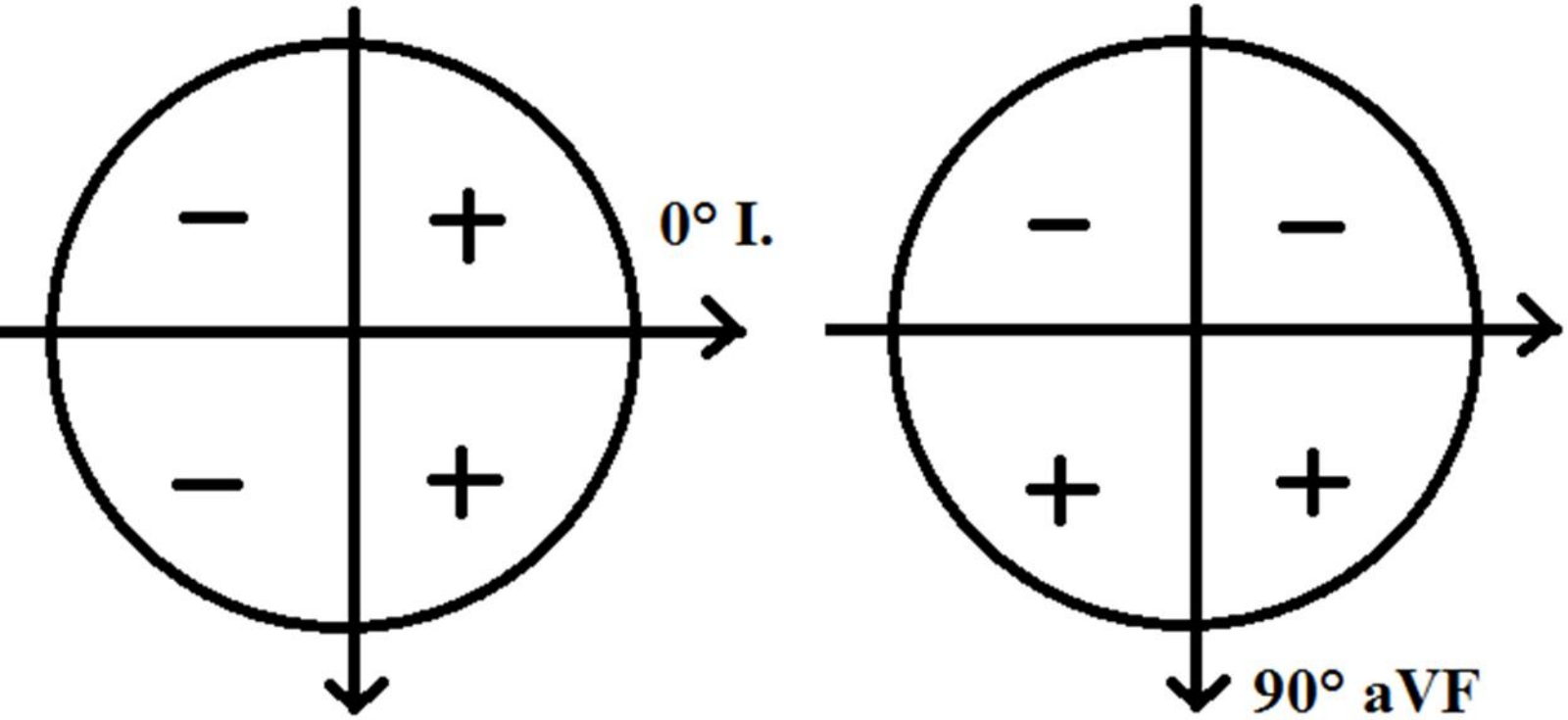
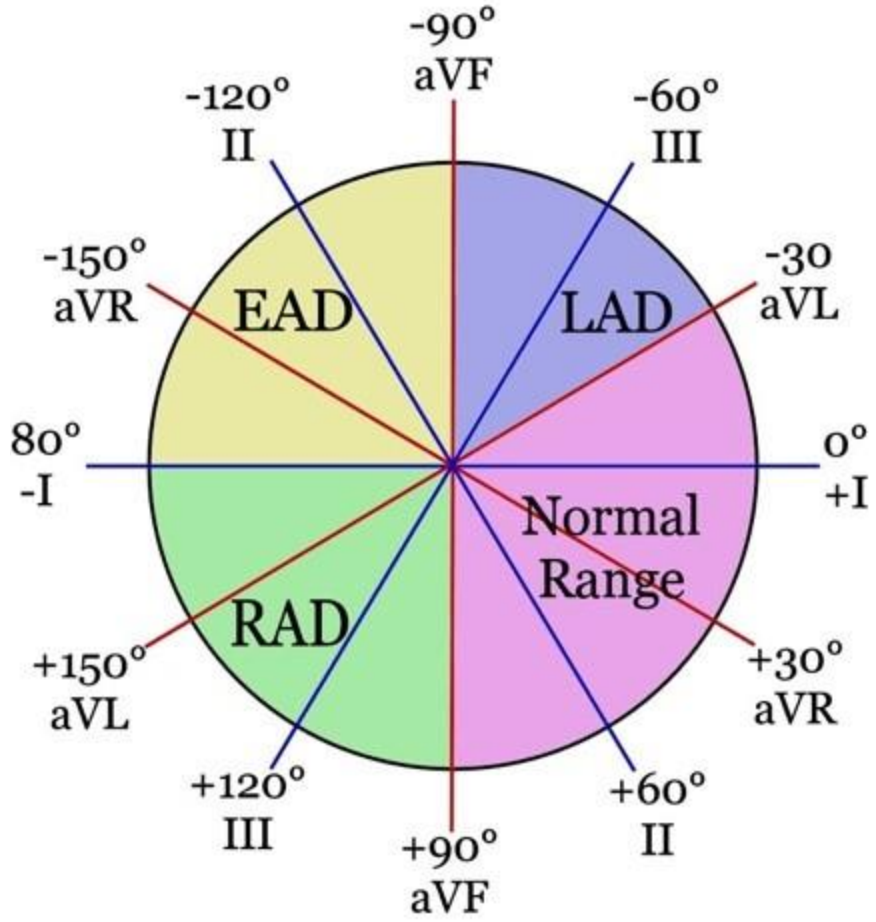


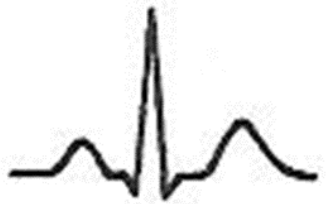
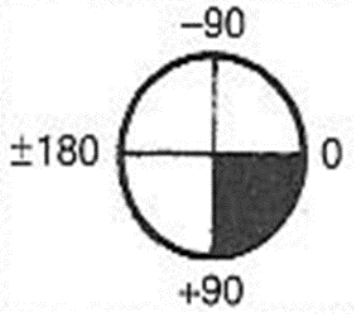
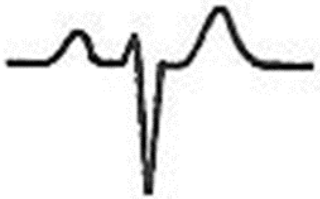
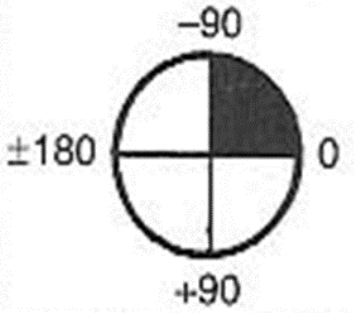
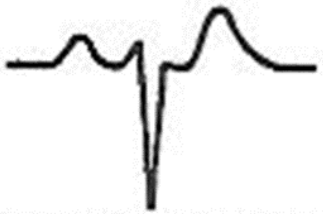
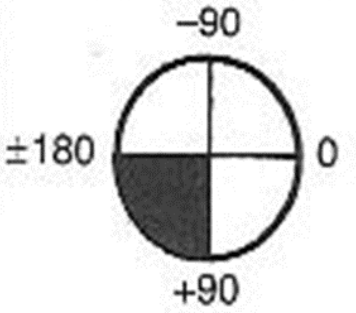
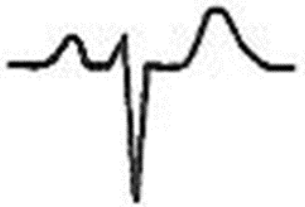
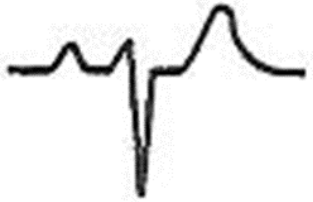
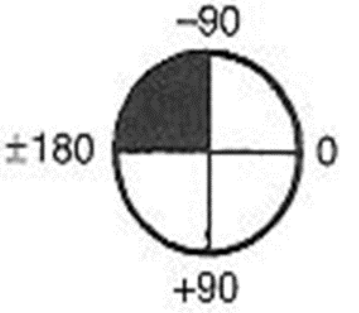
FIG 2-9.
Polarity of precordial leads. Lines drawn perpendicular to leads V_1, V_2, and V_6 divide t positive (*shaded*) and negative (*unshaded*) segments for polarity.

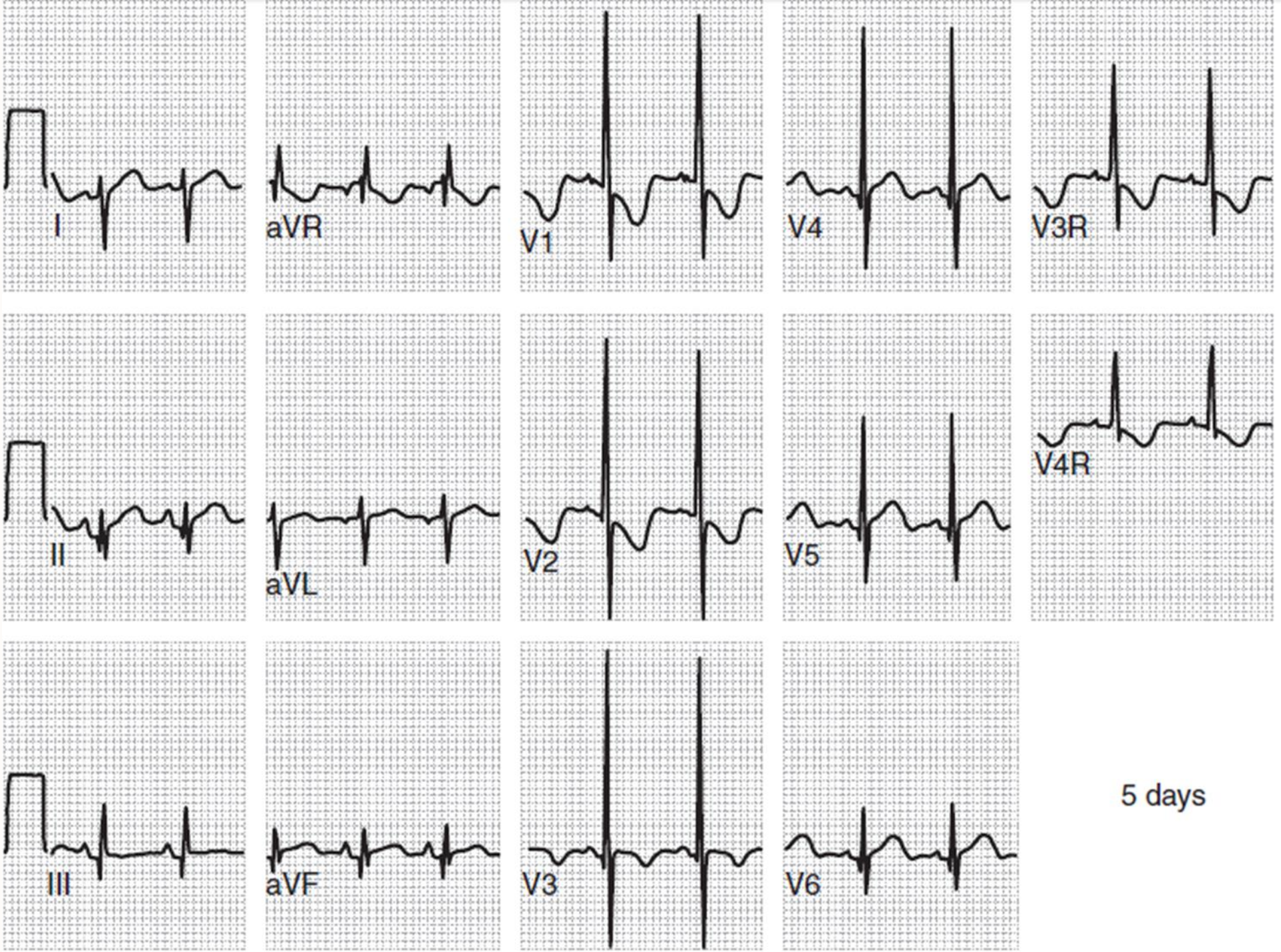


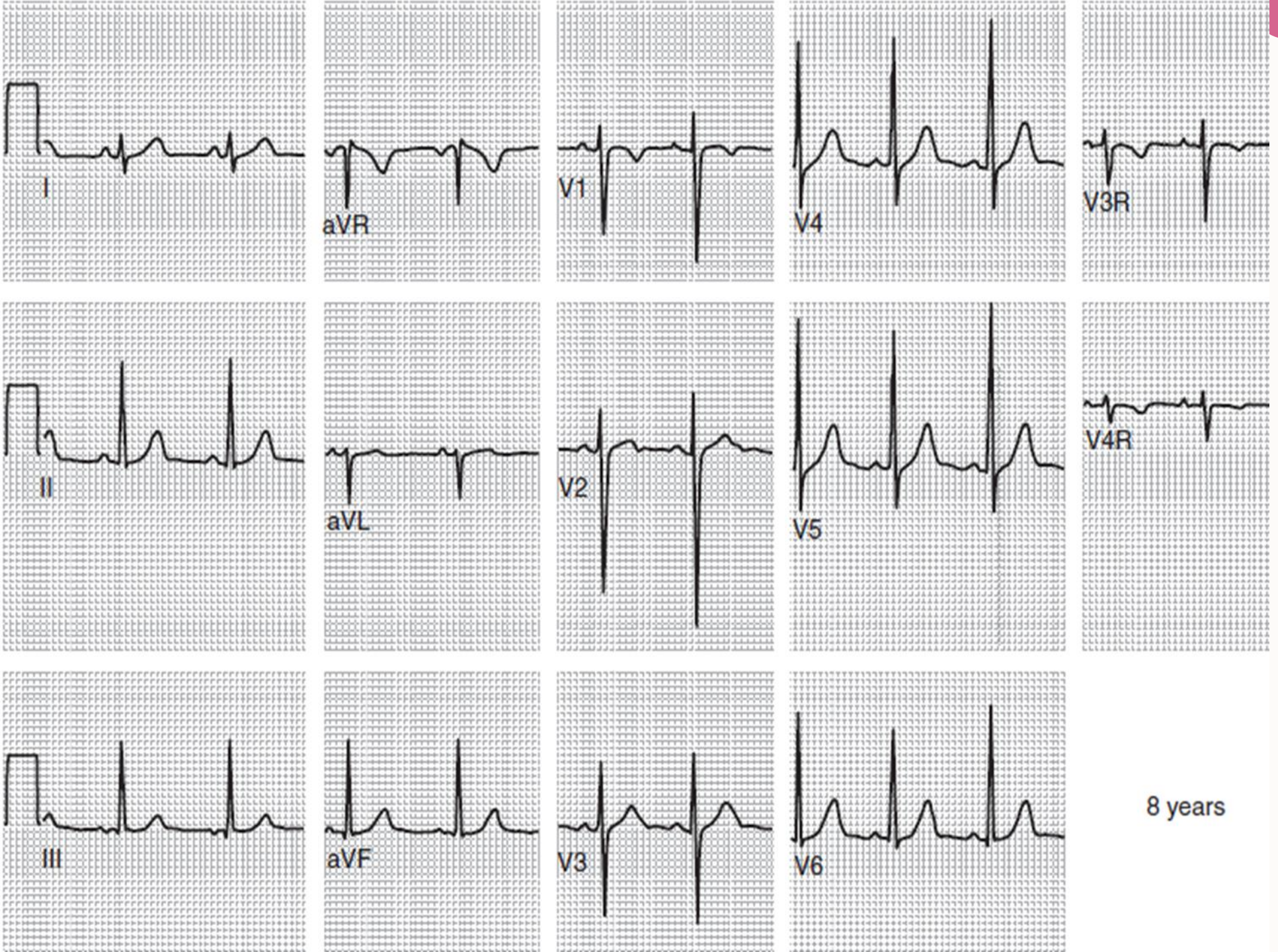
- Electric axis evaluation – the easiest way => to look at leads I and aVF (which are perpendicular to each other) and use final vectors to find the resultant according to this hexaxial reference system – difference between a newborn and an adult





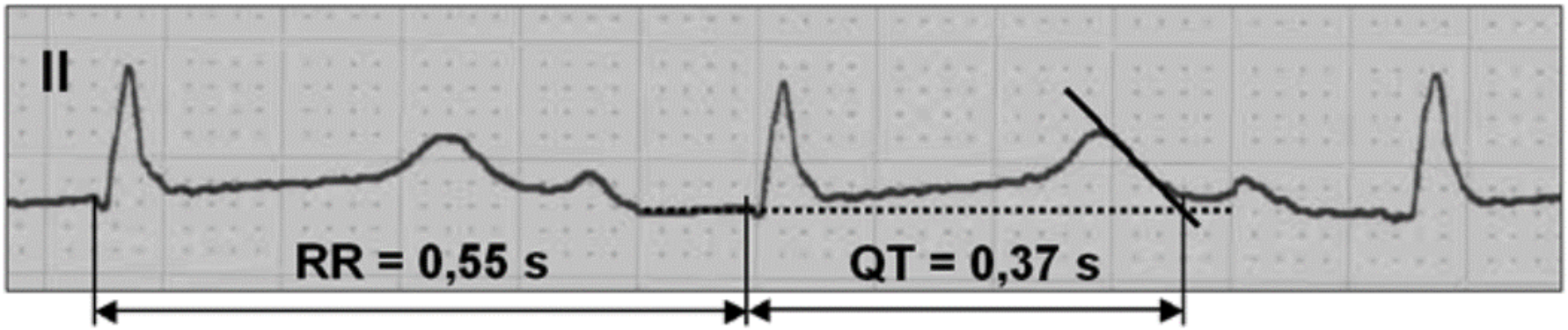
0° – +90°			
0° – –90°			
+90° – ±180°			
–90° – ±180°			


I
aVR
V1
V4
V3R
II
aVL
V2
V5
V4R
III
aVF
V3
V6
5 days

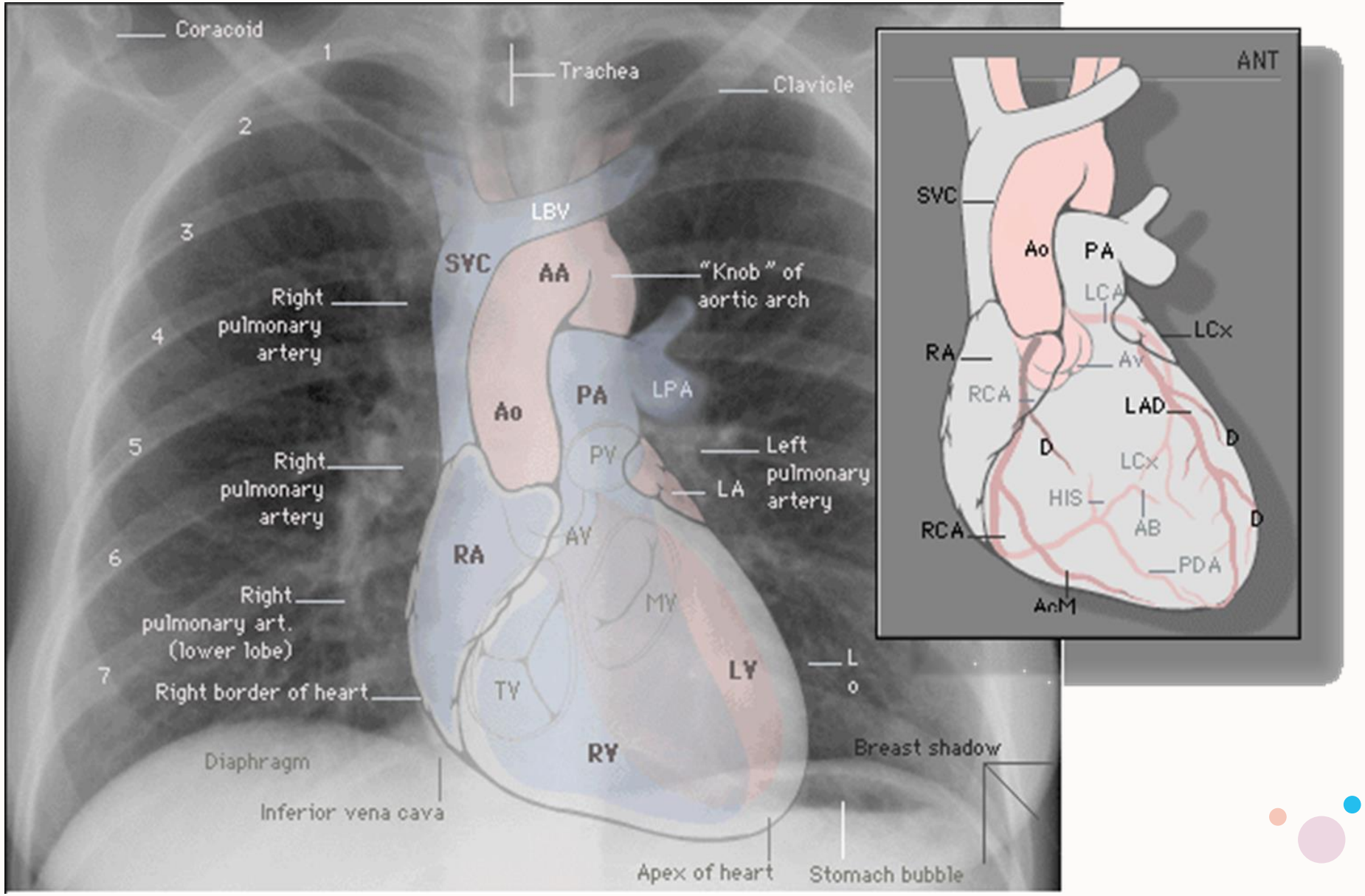

I
aVR
V1
V4
V3R
II
aVL
V2
V5
V4R
III
aVF
V3
V6
8 years

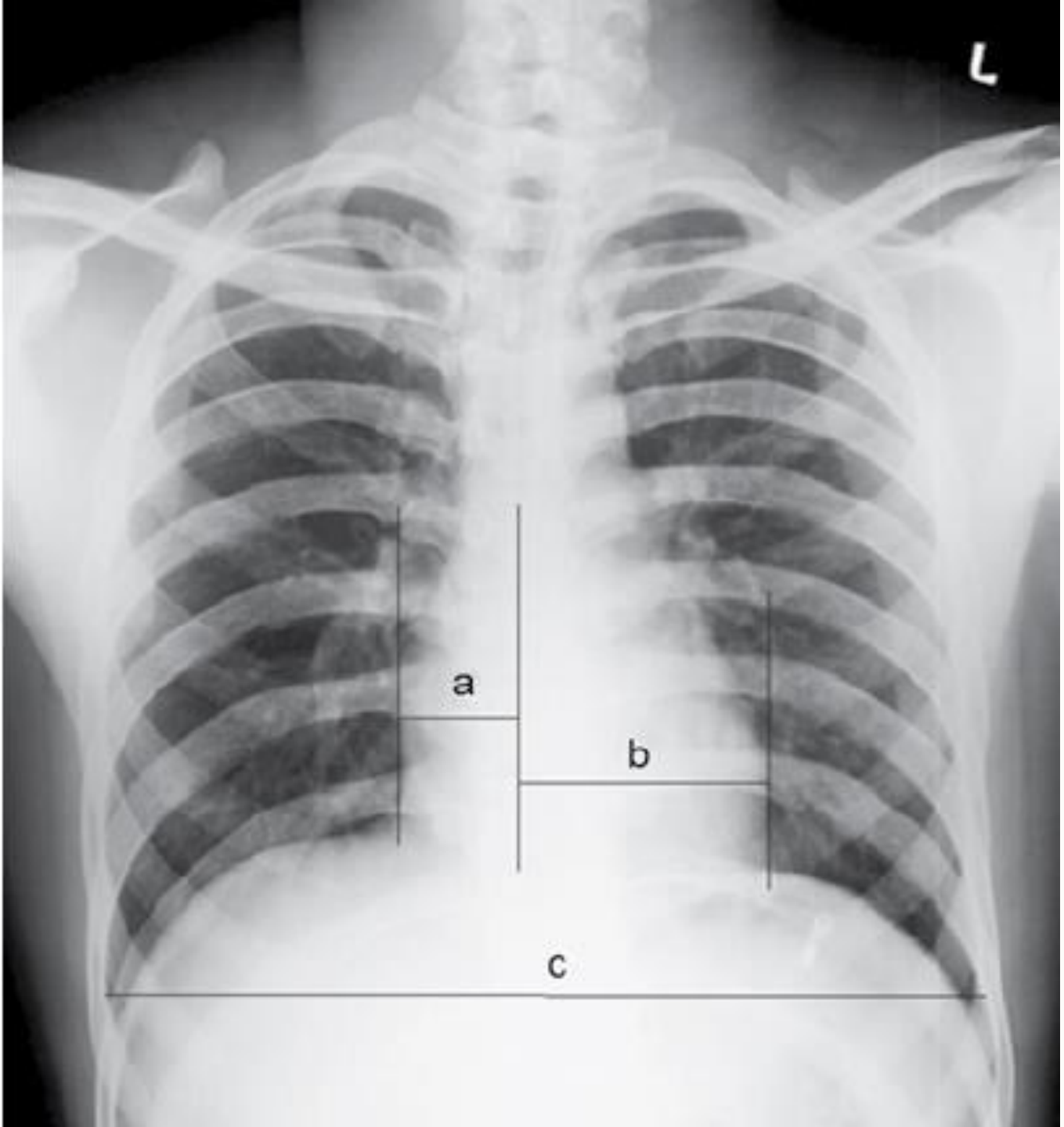
QTc – corrected QT interval (Bazett)

$$QTc\ [s] = \frac{QT\ [s]}{\sqrt{RR\ [s]}} = \frac{0,37}{\sqrt{0,55}} = 0,499\ [s]$$



Chest scan

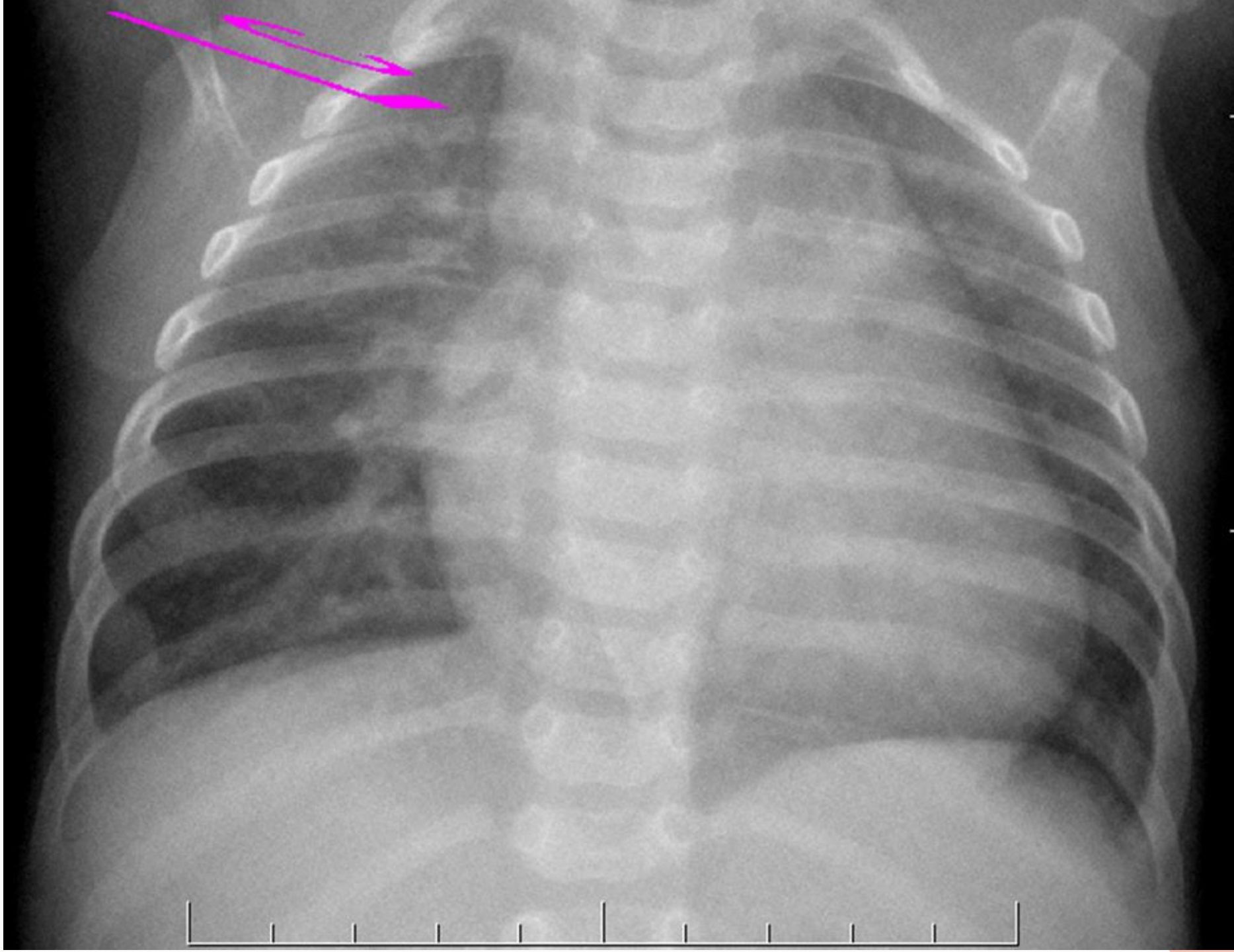

Coracoid
1
2
3
4
5
6
7
Trachea
Clavicle
LBV
SVC
AA
"Knob" of aortic arch
Right pulmonary artery
Ao
PA
LPA
Left pulmonary artery
PV
LA
Right pulmonary artery
AV
RA
Right pulmonary art. (lower lobe)
MV
Right border of heart
TV
LV
L o
RV
Diaphragm
Inferior vena cava
Breast shadow
Apex of heart
Stomach bubble
ANT
SVC
Ao
PA
LCA
LCx
RA
AV
RCA
LAD
D
D
LCx
HIS
AB
D
RCA
PDA
AcM



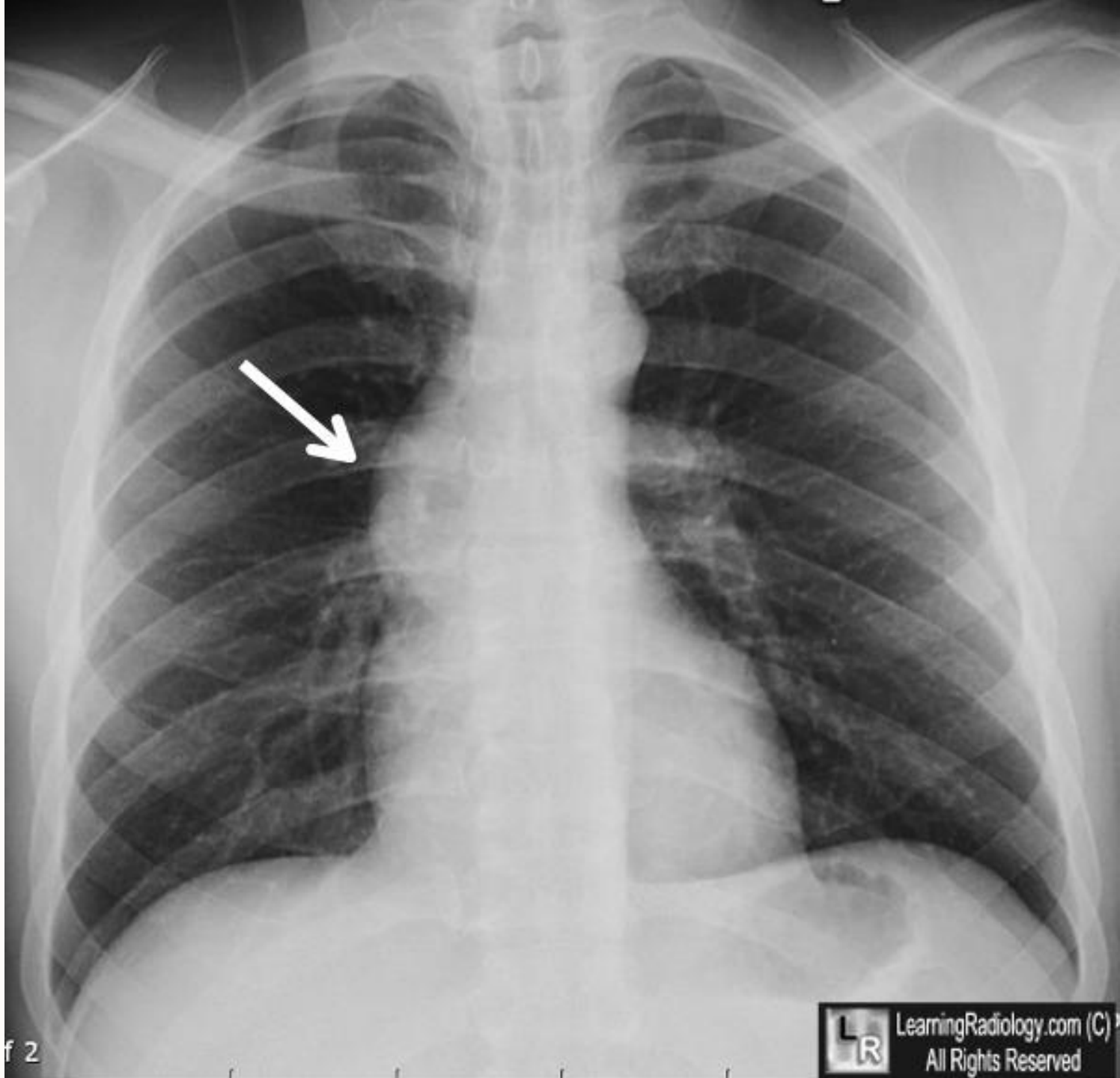
- Cardiothoracic ratio: a + b / c

- Normal range in children according to the age:
  - 1 year 0.61-0.45
  - 1-2 years 0.60-0.39
  - 2-6 year 0.52- 0.40
  - >7 years 0.50- 0.40

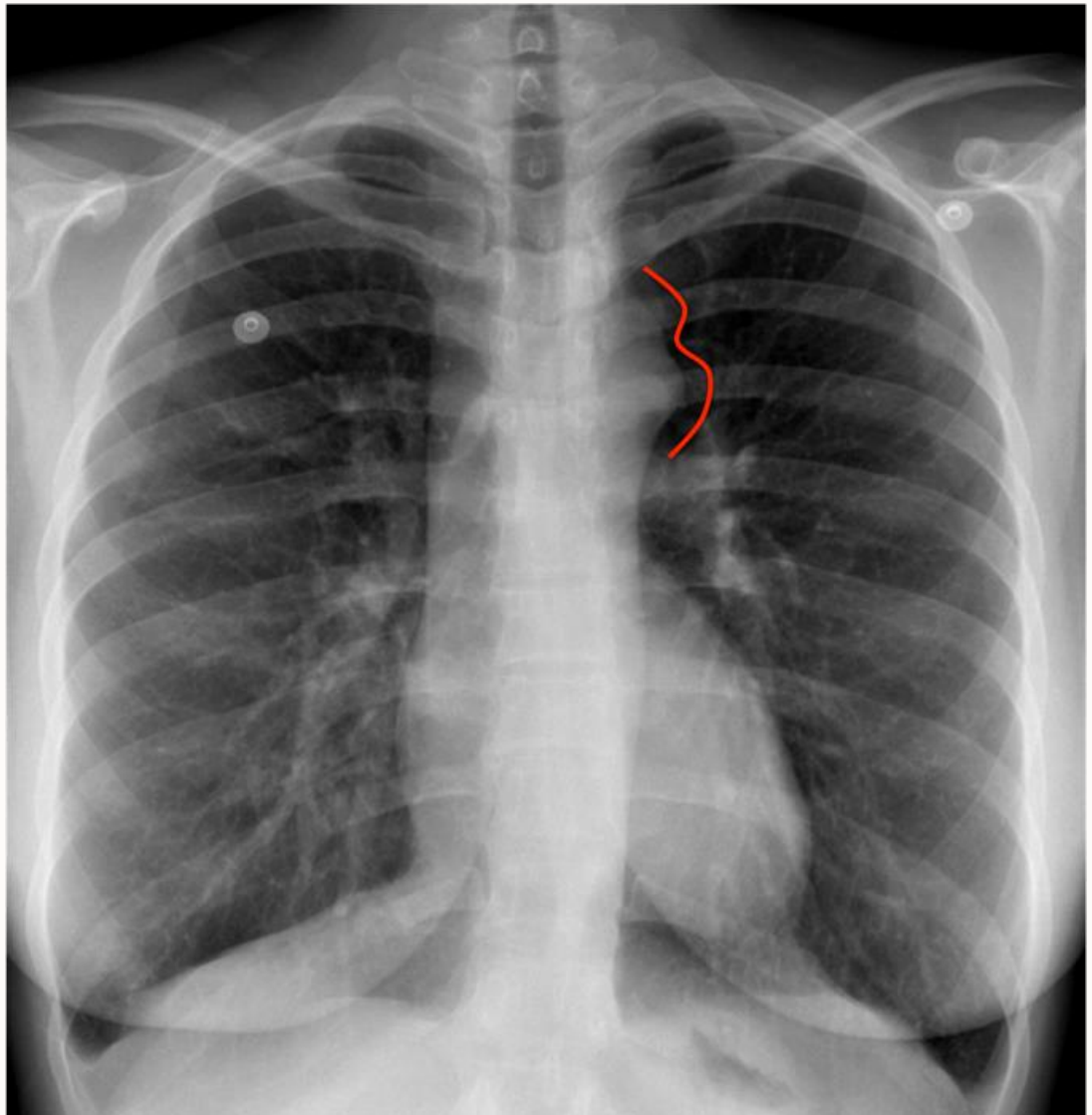
- L-R shunts (**VSD**)
- Cardiomegaly
- ↑ pulmonary vasculature

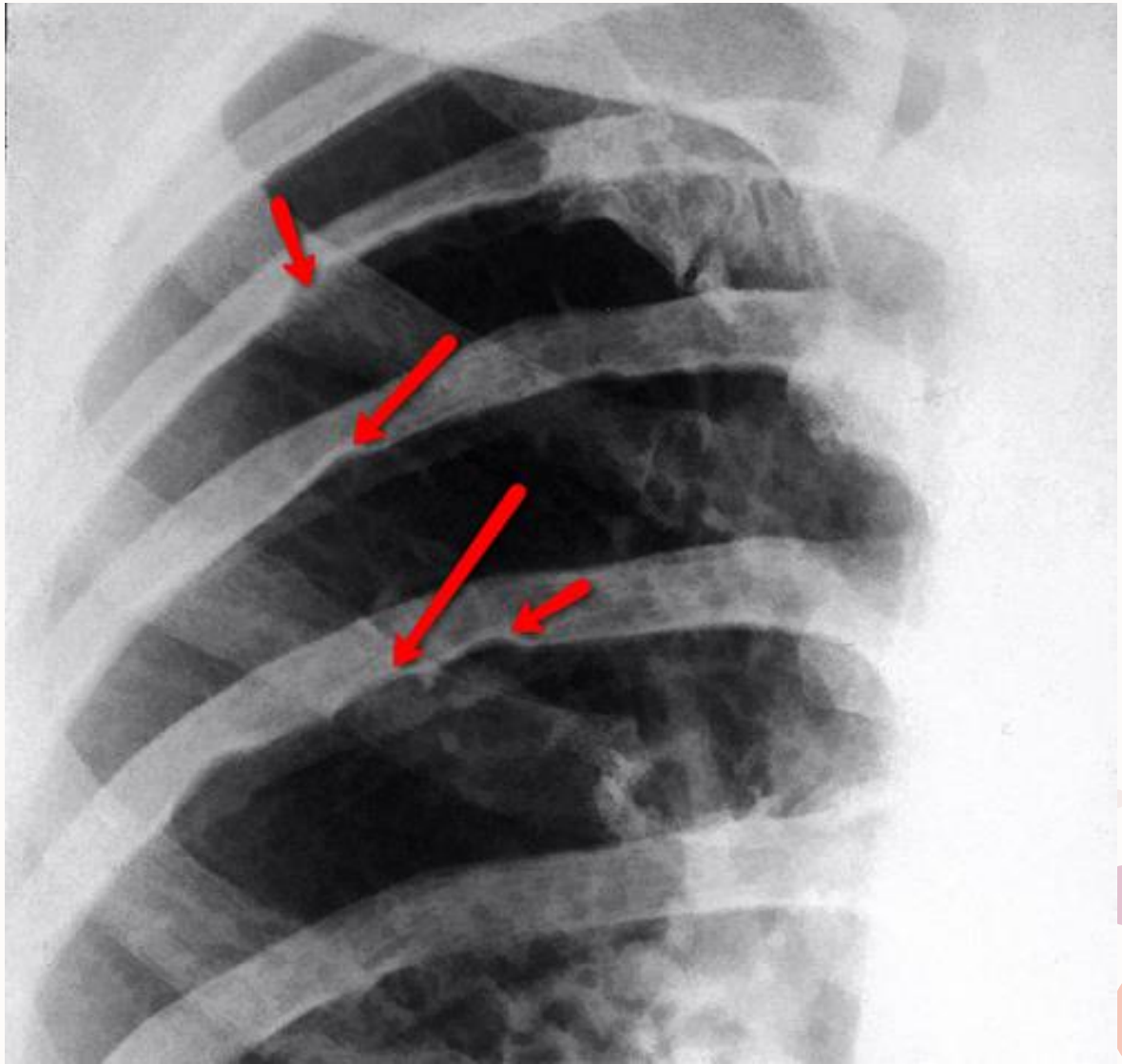


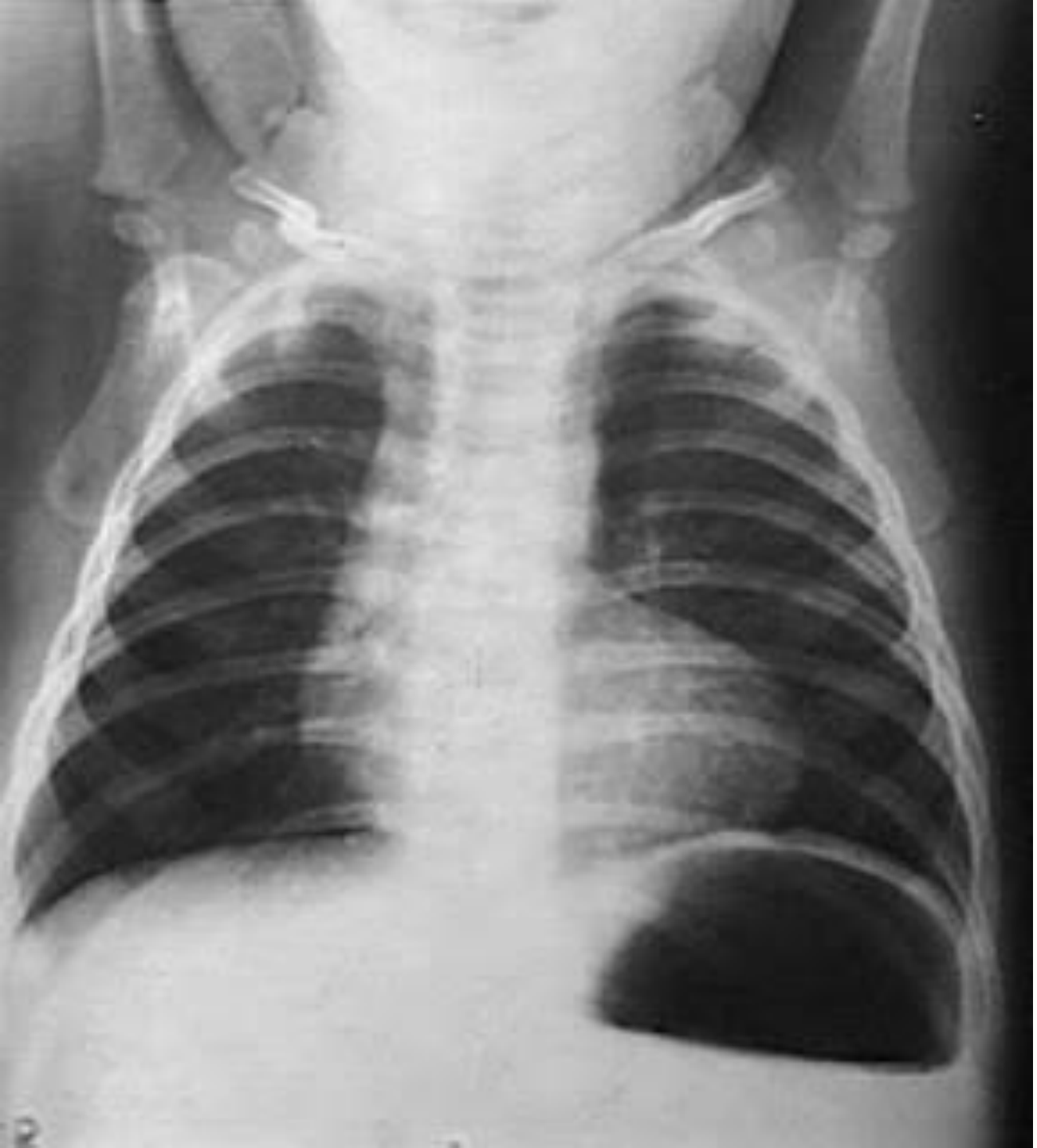
- **AS** – poststenotic dilatation of ascedent aorta



Sign of „3“ (dilated subclavian artery) & notching of the ribs (colaterals) => **CoAo**







- **TOF**
- Boot shape – like heart shadow
- Hypertrophy of RV
- Missing pulmonary knob
- ↓ pulmonary circulation

TAPVD – snowman´s sign

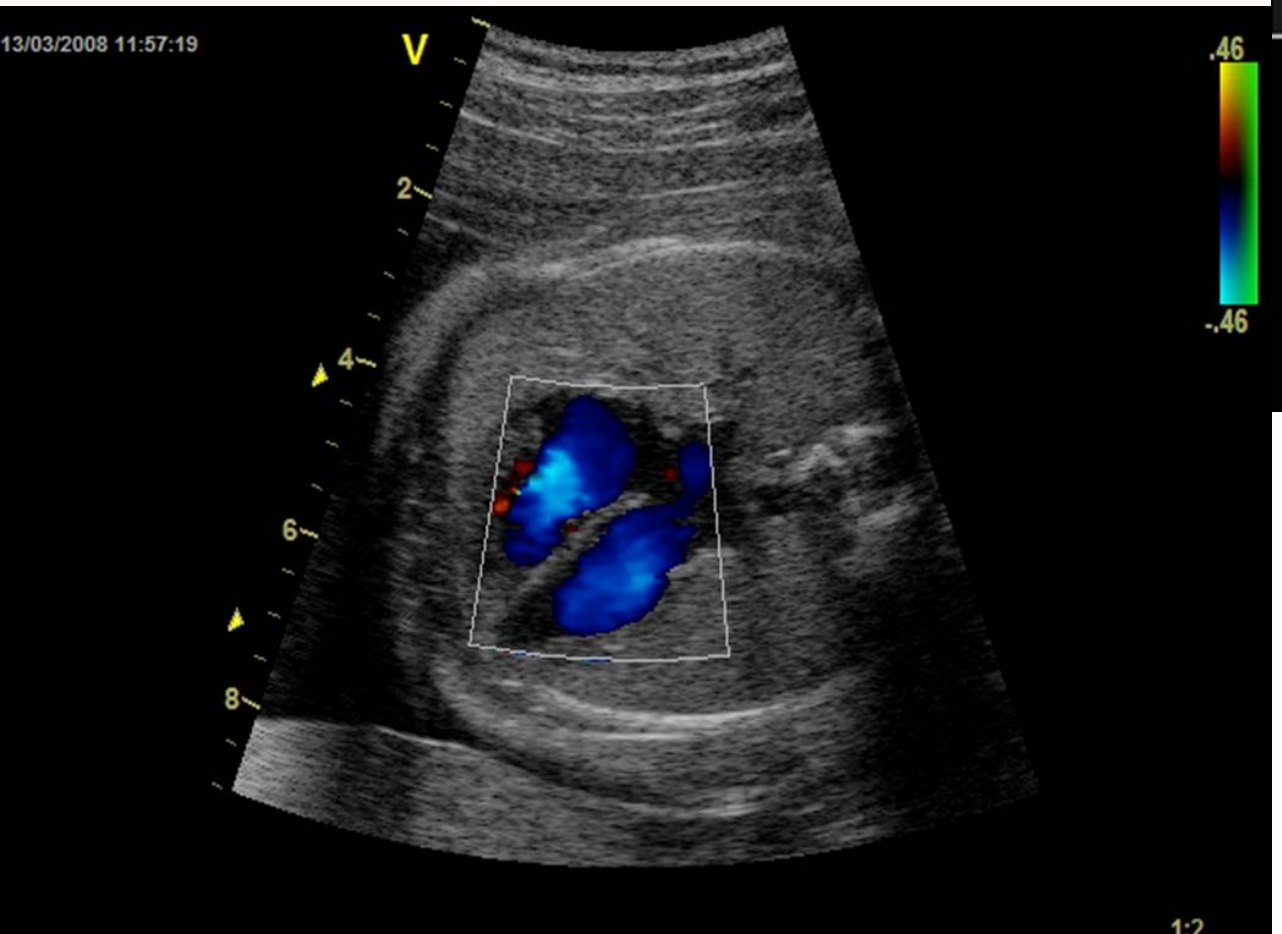
- **TGA**
- „egg-shaped heart“
- Narrow vascular peduncle

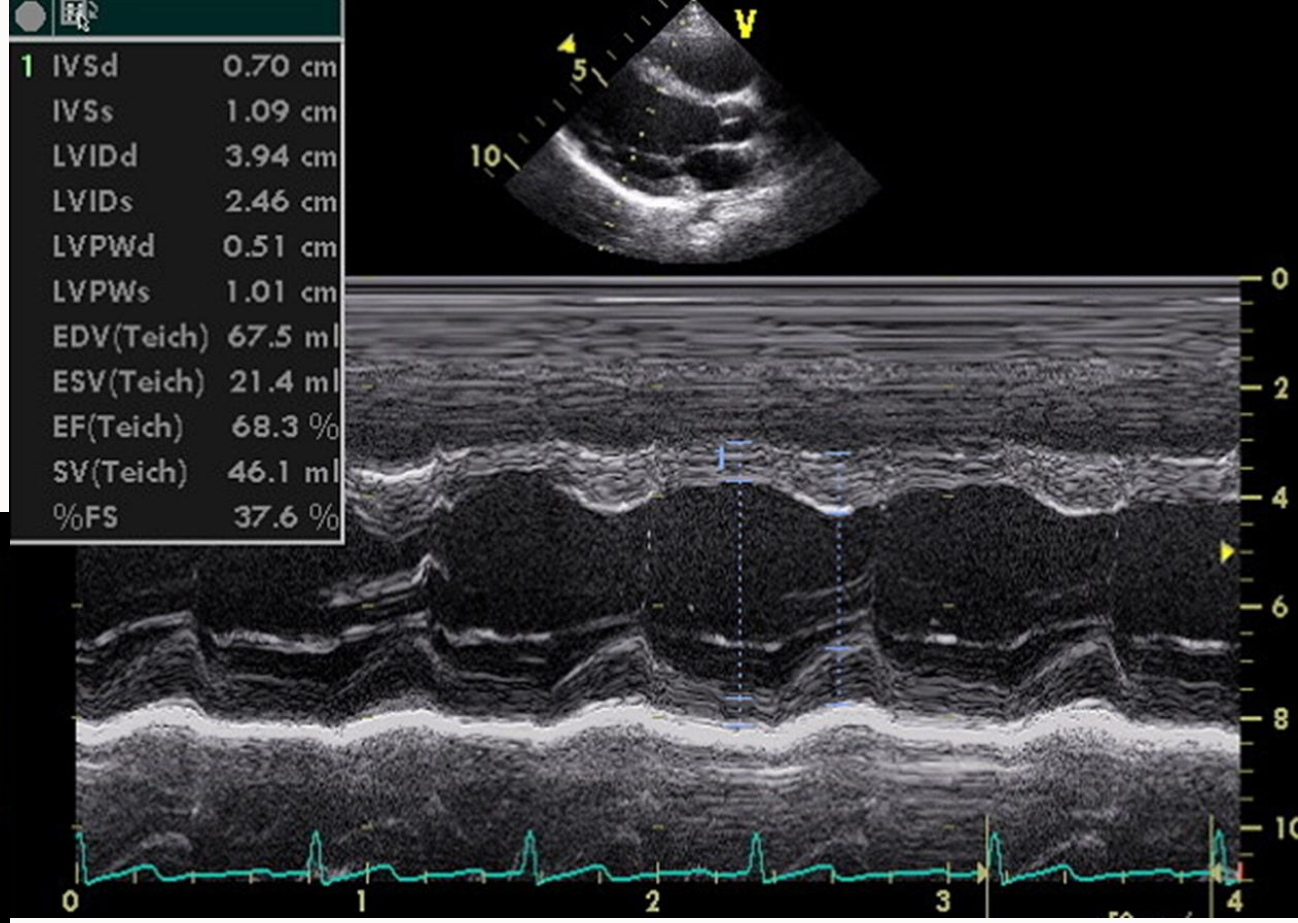
- Pulmonary oedema – **TAPVD obstructive type**
- Picture of „white lungs“

Echocardiography - the most important of all imaging methods in pediatric cardiology

- Different probes:
  - Frequency: 1-10 MHz, higher frequency = lower penetration
    - newborns 7-10 MHz
    - adolescents 3-4 MHz
  - According to the place of use
    - Transthoracic ECHO
    - Transesopfageal ECHO
    - Epicardial ECHO

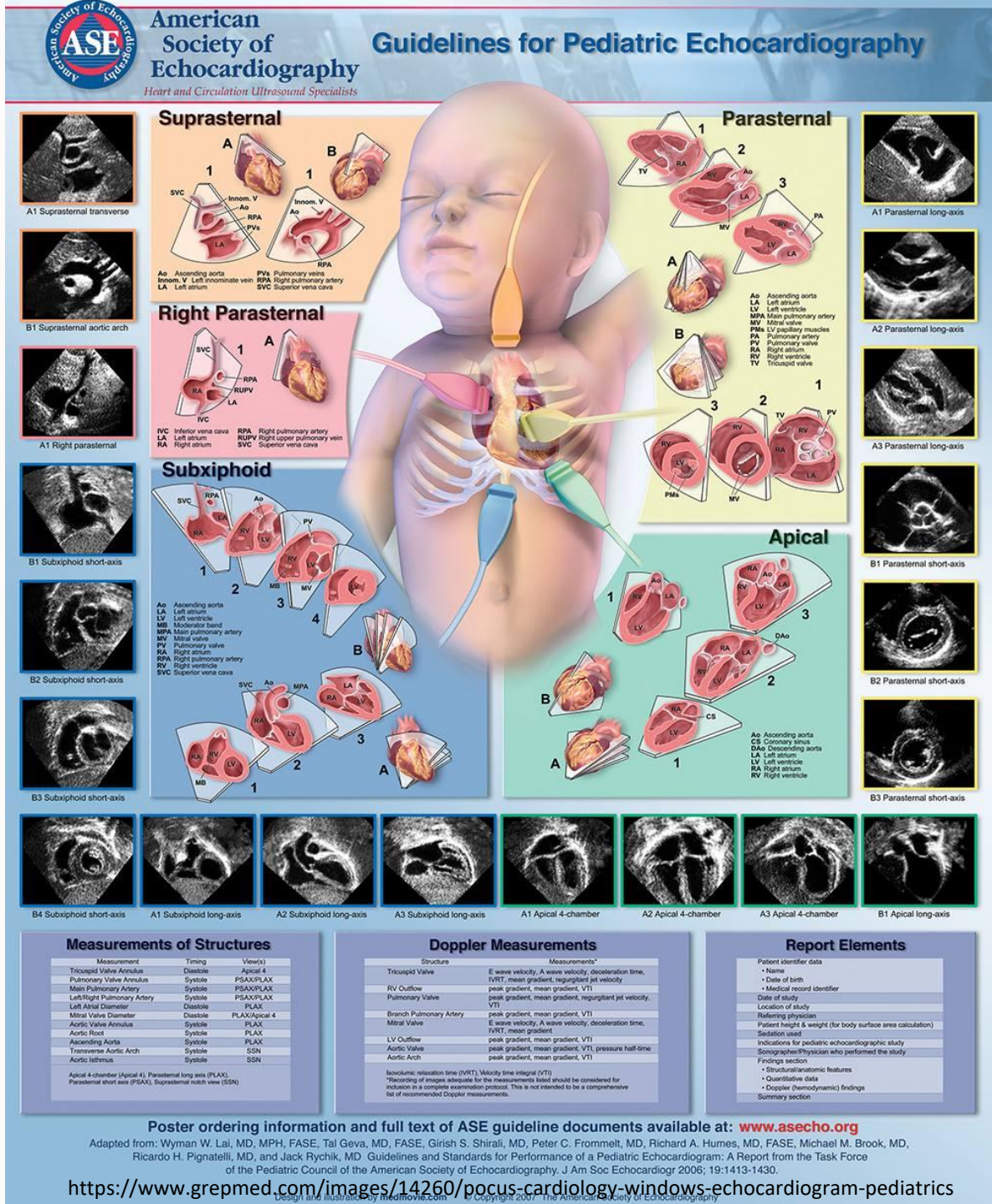
- Different types of imaging:
  - M-mode (1-D imaging)
  - 2-D imaging
  - 3-D imaging
  - Doppler imaging





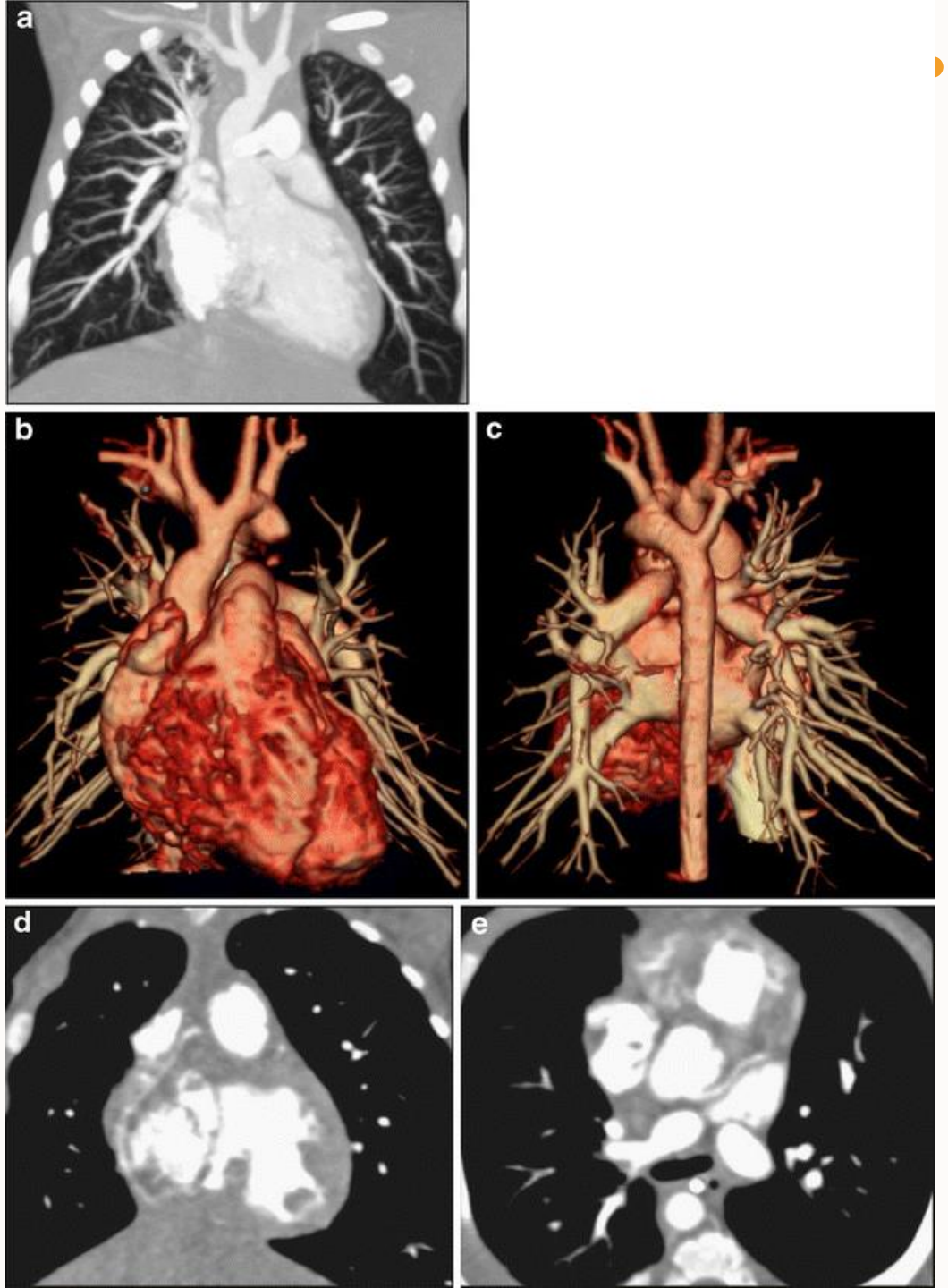
Fetal echocardiography!!

- **4 basic views/windows/:**
  - Subcostal /subxiphoid/
  - 4-chamber view (apical)
  - Parasternal
    - Long axis
    - Short axis
  - Suprasternal



https://www.grepmed.com/images/14260/pocus-cardiology-windows-echocardiogram-pediatrics

CT angiography



MRI

CT

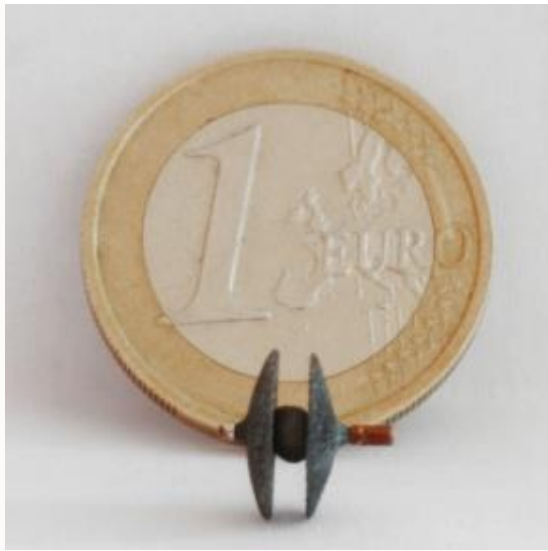
- Radiation
- Fast aquisition (seconds)
- Iodium contrast agent
- High spatial resolution
  - 0,5mm slice thickness
- Better available
- Cheaper
- Pacemakers/ICD: OK

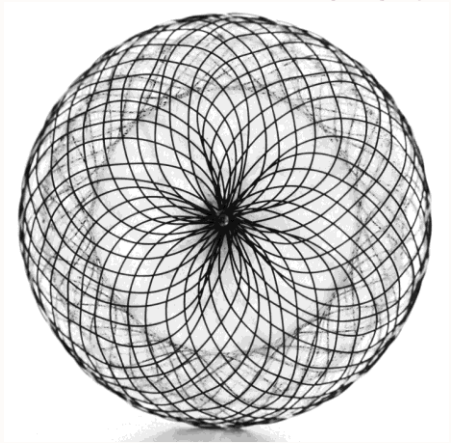
MRI

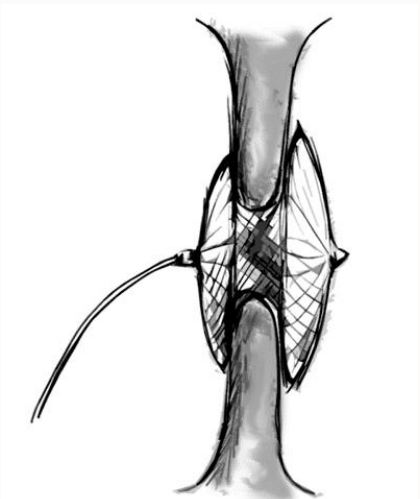
- No radiation
- Slow aquisition (min – 1 hour)
- Gad contrast agent +/-
- Lower spatial resolution
  - 3mm slice thickness
- Evaluation: chamber volumes, systolic function, valves, flows, tissue characterization
- More expensive
- Pacemakers/ICD: contraindicated (not all of them)

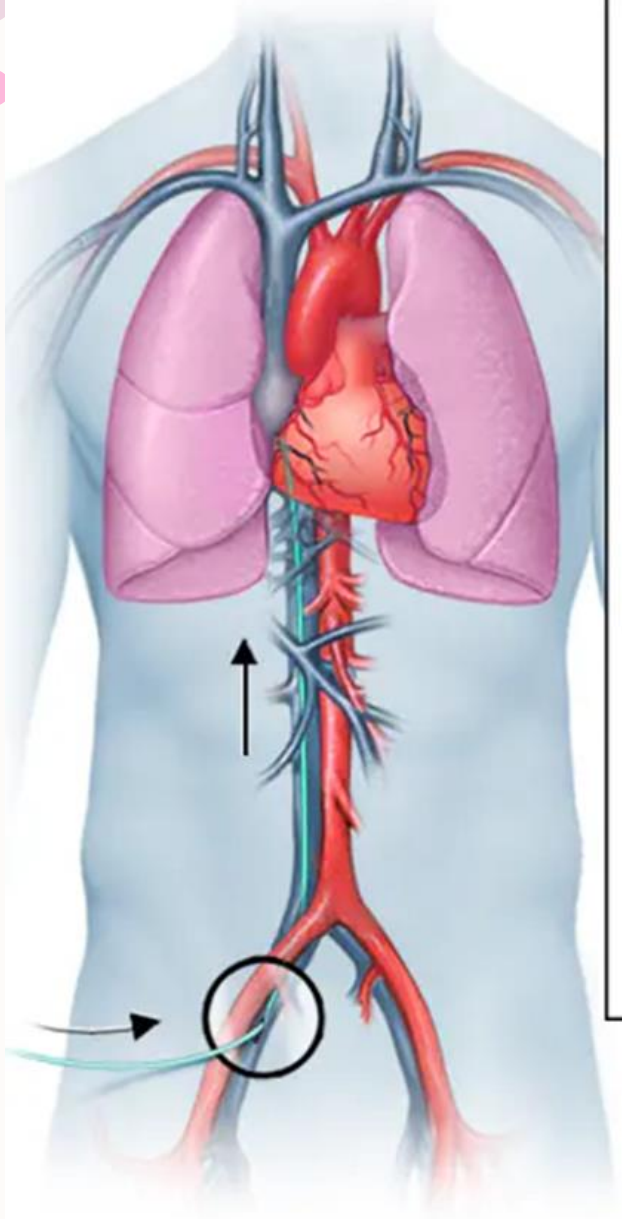
Catheterisation

- **Diagnostic** – hemodynamics, saturations of the oxygen, imaging of the stenosis which is not properly visible by echo, endomyocardial biopsy, electrophysiologic examination
- **Therapeutic**
  - Dilation of the stenosis/coarctation
  - Stent/valve placement
  - Baloon atrioseptostomy
  - Closure of ASD, PDA by occluders
  - Ablations

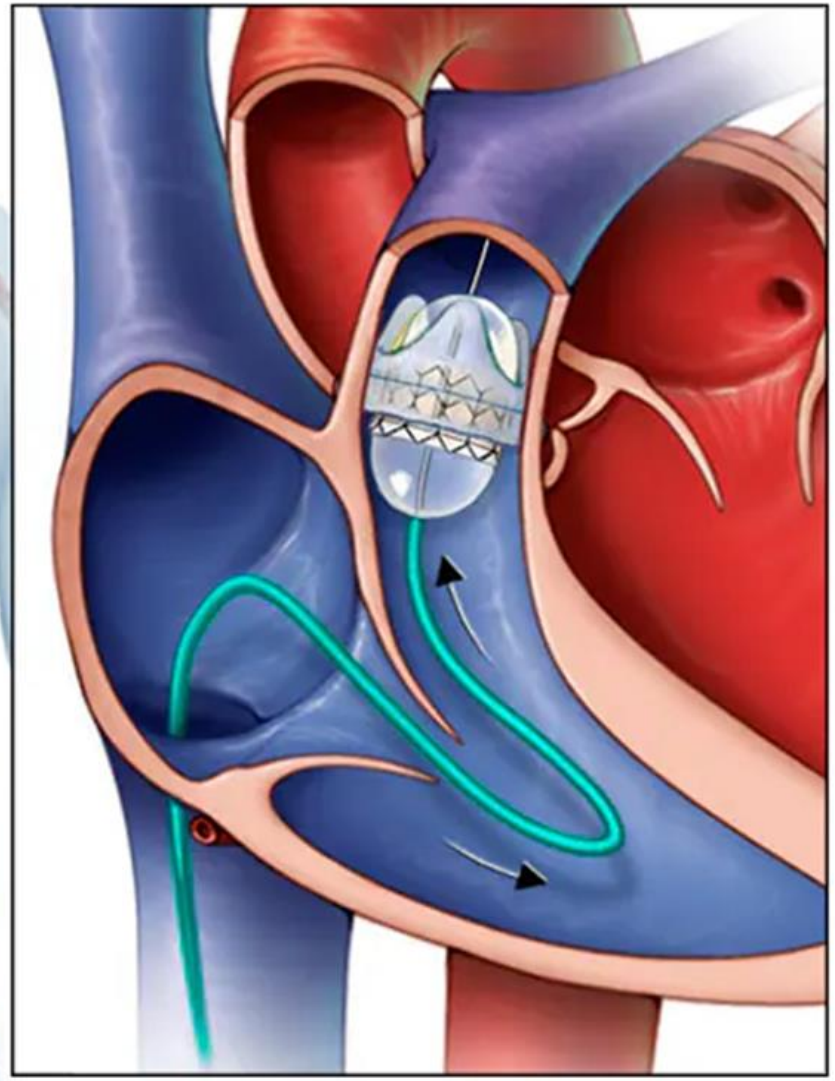




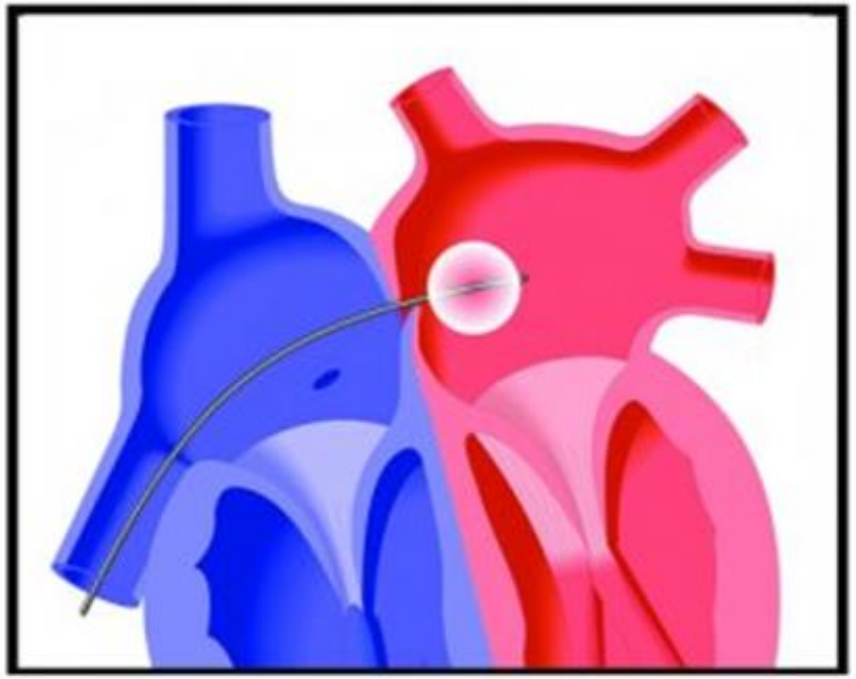


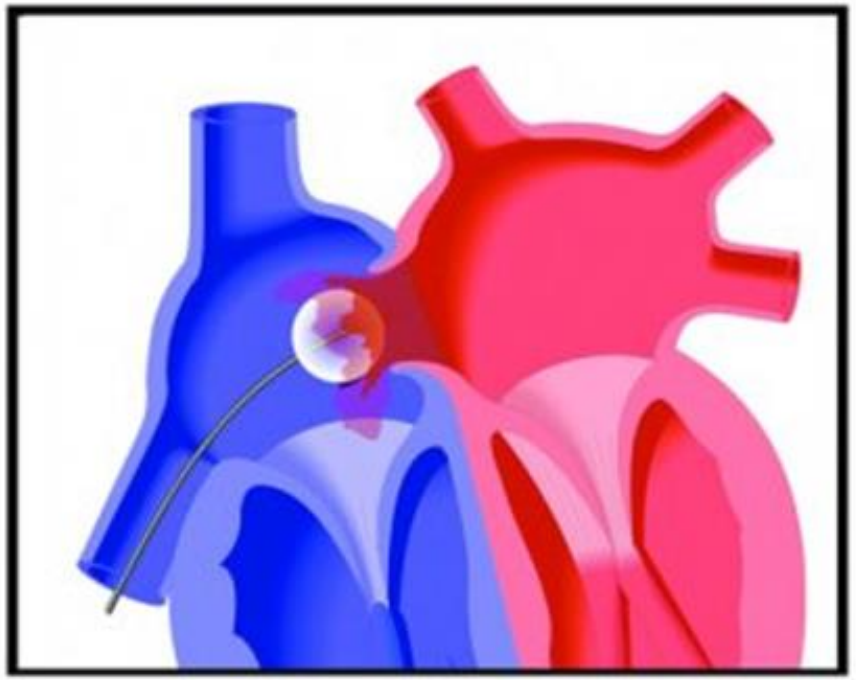


Catheter placed through femoral vein to heart



Balloon expands to place new valve inside existing valve





https://qcg.com.au/procedures/cardiac-catheter-ablation/

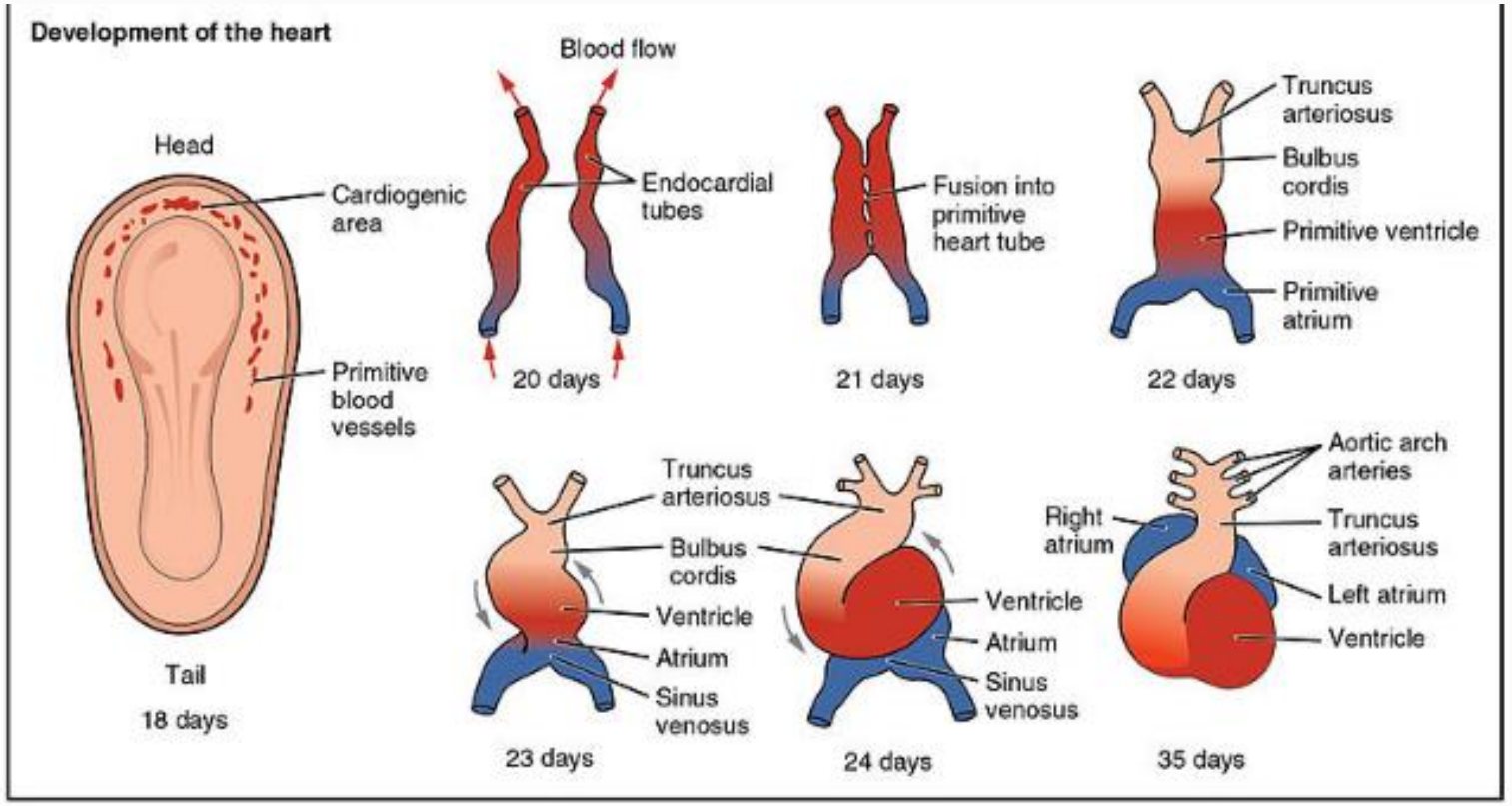
Cardiac stress test (ergometry)

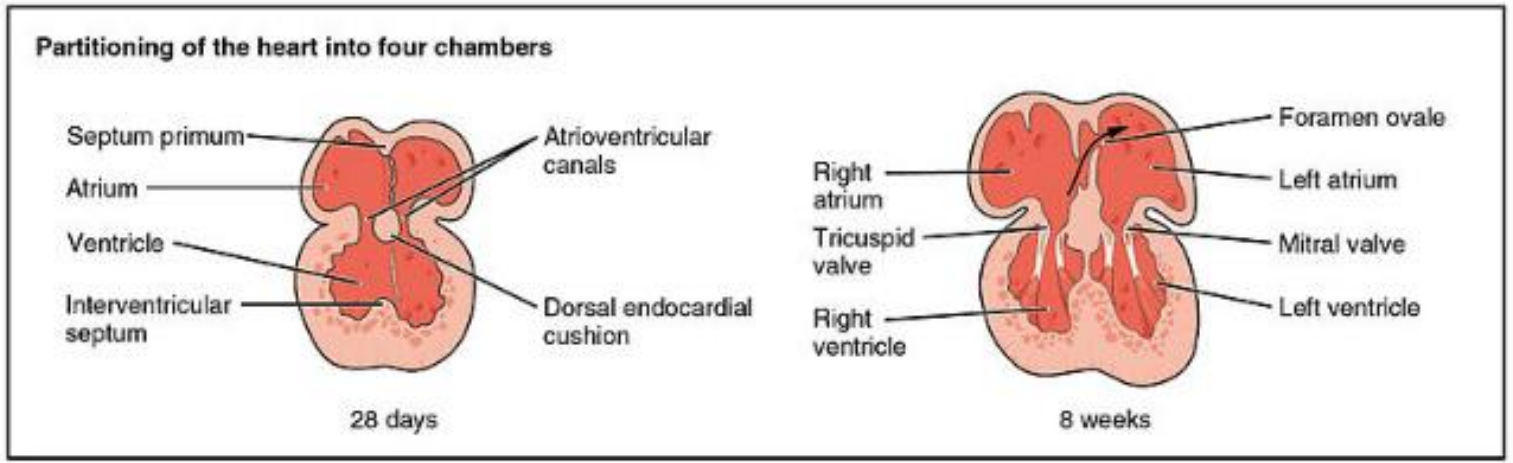
- Non-invasive examination => monitoring the work of the heart during exercise
- Bicycle ergometer => minimum height of 125-135cm or a treadmill (in smaller children) !!The key is the cooperation of a child!!
- Indications:
  - Asessement of physical capacity (after cardiosurgical operations, sportsmen)
  - Arrhythmias and syncope (during or soon after exercise)
  - When suspicion on lower coronary artery reserves (after cardiosurgery on coronary arteries, AS, in stenocardia, palpitations, dyspnea on exercise)
  - Asessement of the arterial blood pressure reaction (after cardiosurgery of CoA)
  - Preventively (positive family history – sudden cardiac death on exercise)
- Normal maximum heart rate = „220/min.“ minus „age of the patient“

Congenital heart diseases

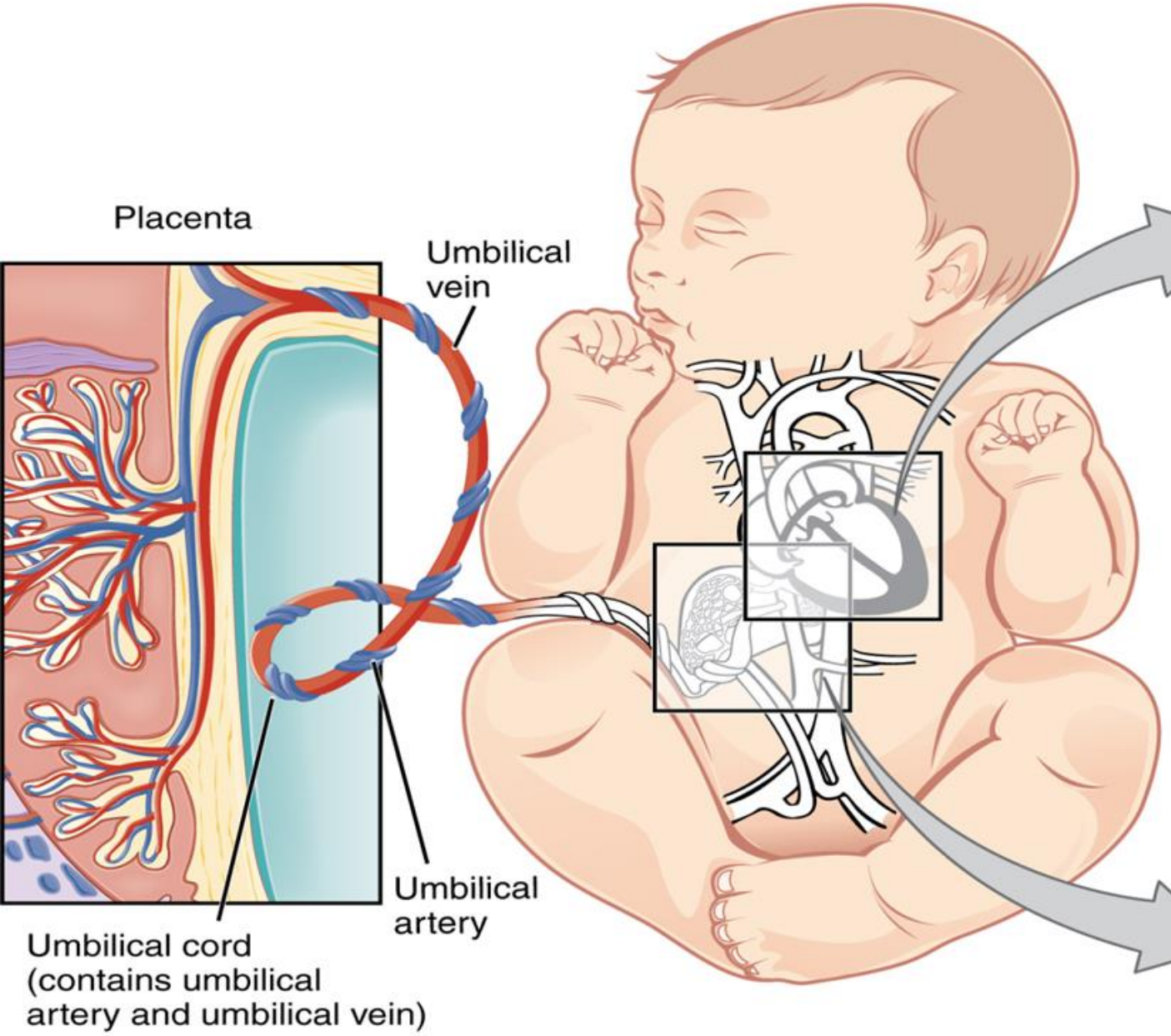
VCC – vitium cordis congenitum

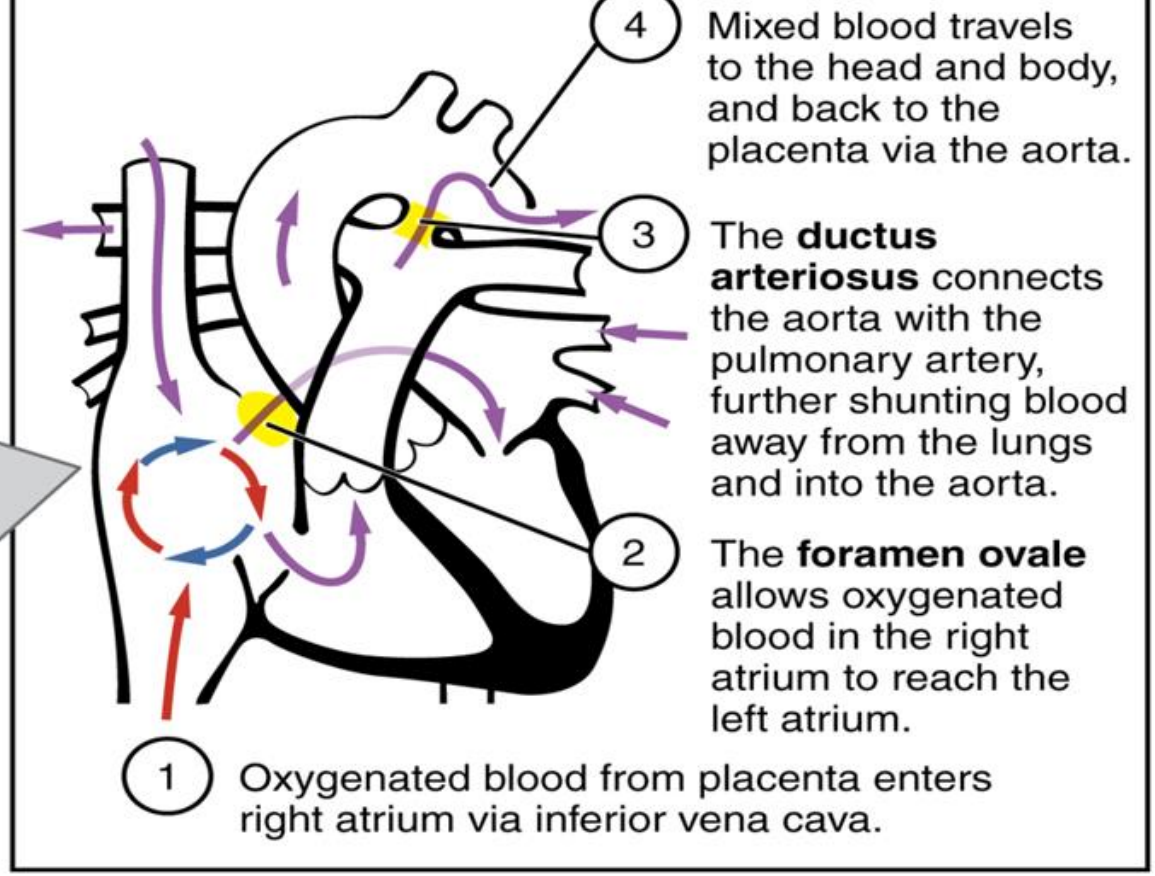
Embryology of the heart 3rd-10th gestational week

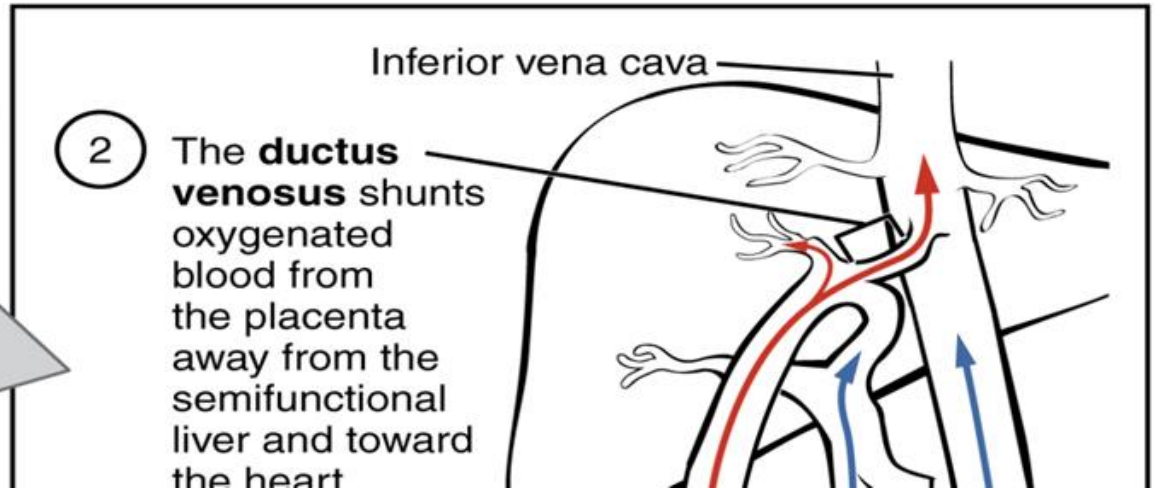




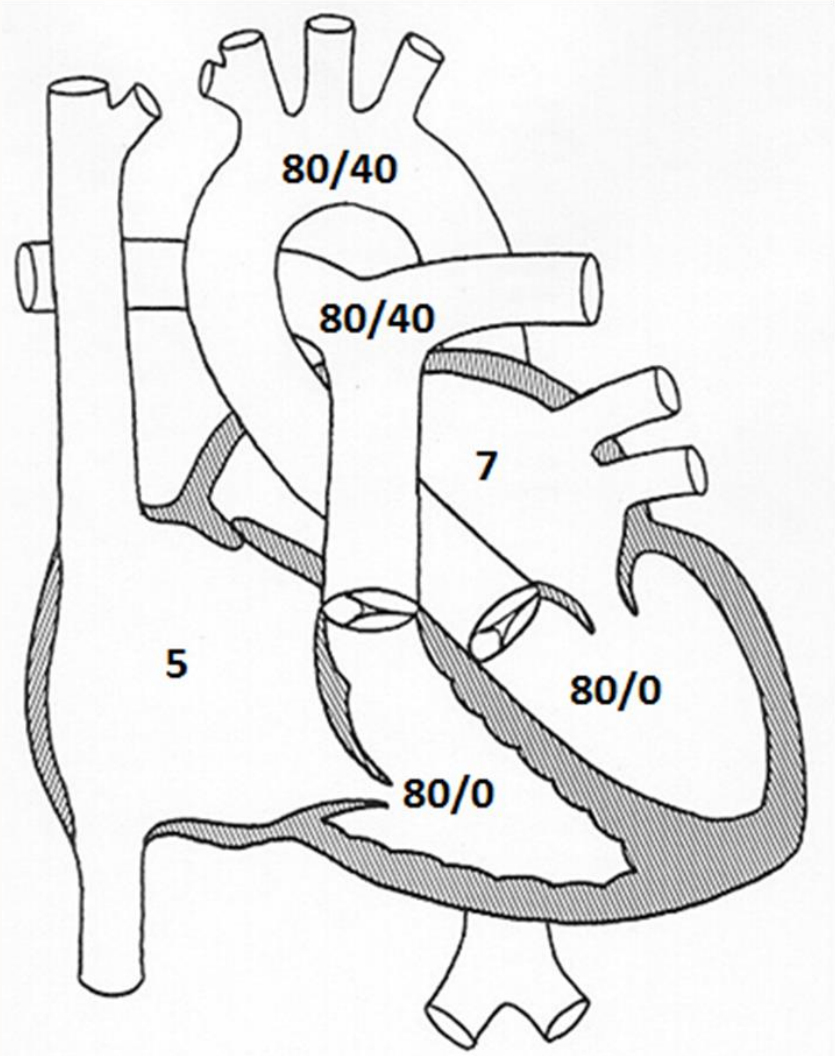
Fetal circulation

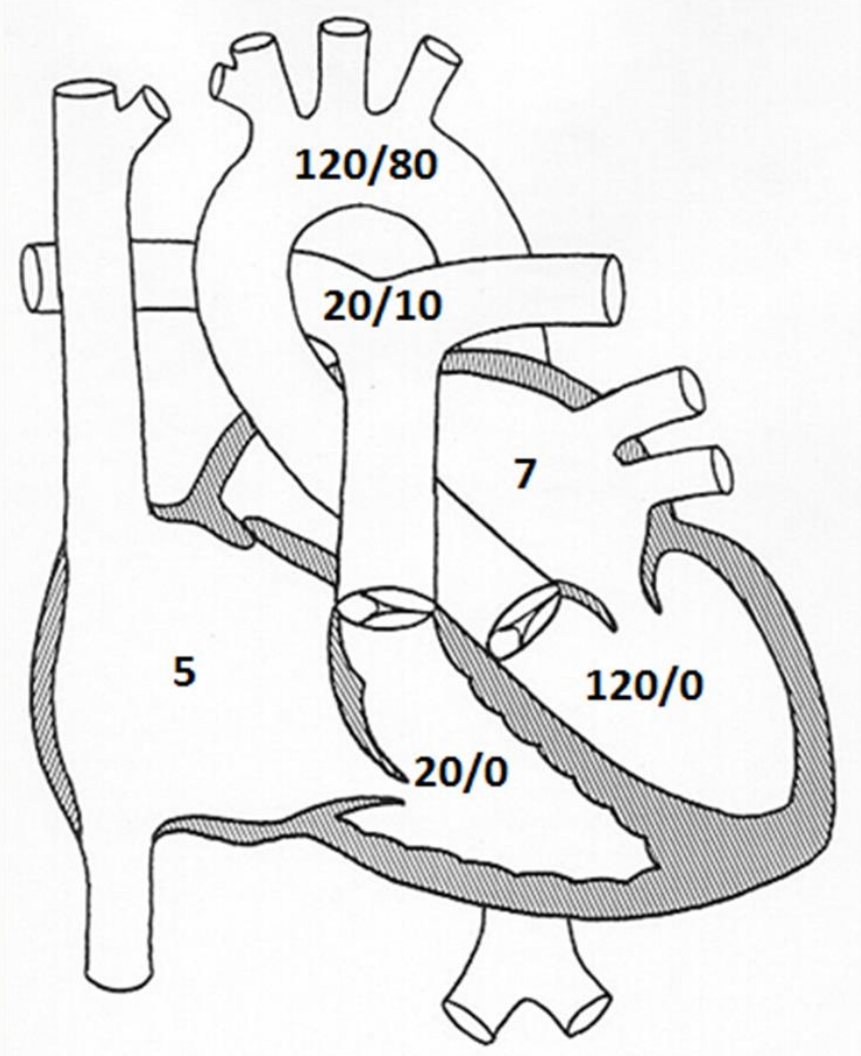






Hemodynamics - newborn vs. adult





Perinatal changes

- **1st breath** => **↑pO2** => **vasodilatation** in pulmonary arterial bed => **↓ pulmonary arterial resistence** => **↓ blood pressure in RV =>**
- Functional closure of **PDA** 10-18 hours after birth,
  - anatomically up to 3rd week of life
  - PDA in term newborns after 3 months of age => pathologic
- **FoA** – physiologic closure up to 6th month of age

Congenital heart diseases in numbers

- Most common of all congenital diseases – app. 30-40 %
- Incidence up to 1 % (6-8/1000 children)
- Up to 80 % require CS intervention
- 85-90 % simple VCCs
- Survival 80-85 % (also severe defects)

Prevalence of VCCs

- 6-8/1000 liveborn infants
- 41 % ventricular septal defect
- 9 % atrial septal defect
- 5 -6 % persistent ductus arteriosus, aortic stenosis, pulmonary stenosis, coarctation of the aorta
- 5 % transposition of the great arteries
- 4% tetralogy of Fallot
- 20% other VCCs

Prenatal cardiology

- A part of the 2nd trimestral screening
- Very important part of pediatric cardiology => very helpful for further managment of a child
- Take-off of the specialty in the 90´s of 20th century
- Centre of prenatal cardiology is in the Pediatric cardiac centre in Motol (Prague)
- Czech Republic prenatal detection with >80 % success in the latest years (USA around 40 %, SR around 20 %)
- The possibility of termination of the pregnancy (43 %)

Etiology - controllable causes

- Alcohol
- Warfarin, antivirotics, ATB, psychopharmaceutics
- Gestational DM
- X-ray, CT
- Infection: TORCH

Etiology - uncontrollable causes

- Genetics (chromosomal abnormalities, gene mutations)
- > 50 % of the cases are without any significant etiology!!!
- !!! consanguinity !!!

M. Down - A-V canal

Edwards, Patau

Turner - CoA

Marfan – Ao dilatation

DiGeorge - TOF

Classification of the VCCs

- **Central cyanosis**
  - cyanotic (35 %) D-TGA, TOF
  - noncyanotic (65 %) VSD, CoA

- **Severity**
  - critical (35 %)
    - Severe central cyanosis D-TGA
    - LCO + ↑pulmonary blood circulation + mild central cyanosis CoA, HLHS
  - noncritical (65 %) TOF, VSD

Therapy of the VCCs

- Depending on:
  - severity of VCC (critical vs. noncritical) => timing!,
  - complexity/type of VCC,
  - clinical state of a child, additional comorbidities,
  - weight/lenght,
  - possible advantages/risks from the intervention,
  - cardiosurgery (curative/palliation) vs. cathetrisation vs. watch & wait strategy (prevention of infective endocarditis!)

Cardiosurgery	Cathetrisational intervention
Invasivity ↑↑↑	Invasivity ↑
Open chest surgery : yes	Open chest surgery: no
ECC: yes	ECC: no
Arrhythmias: ↑↑↑	Arrhythmias: ↑
Infection: ↑↑↑	Infection: ↑
Pain: ↑↑↑	Pain: ↑
Hospitalisation : 7-10 d.	Hospitalisation: 3 days

Noncyanotic VCC- noncritical

- with L-R shunt

- **Ventricular septal defect (VSD) (40 %)**
- Atrial septal defect (ASD) (9 %)
- A-V septal defect (AVSDC)
- Ductus arteriosus persistent (PDA) (up to 6 %)

Symptomatology in L-R shunts

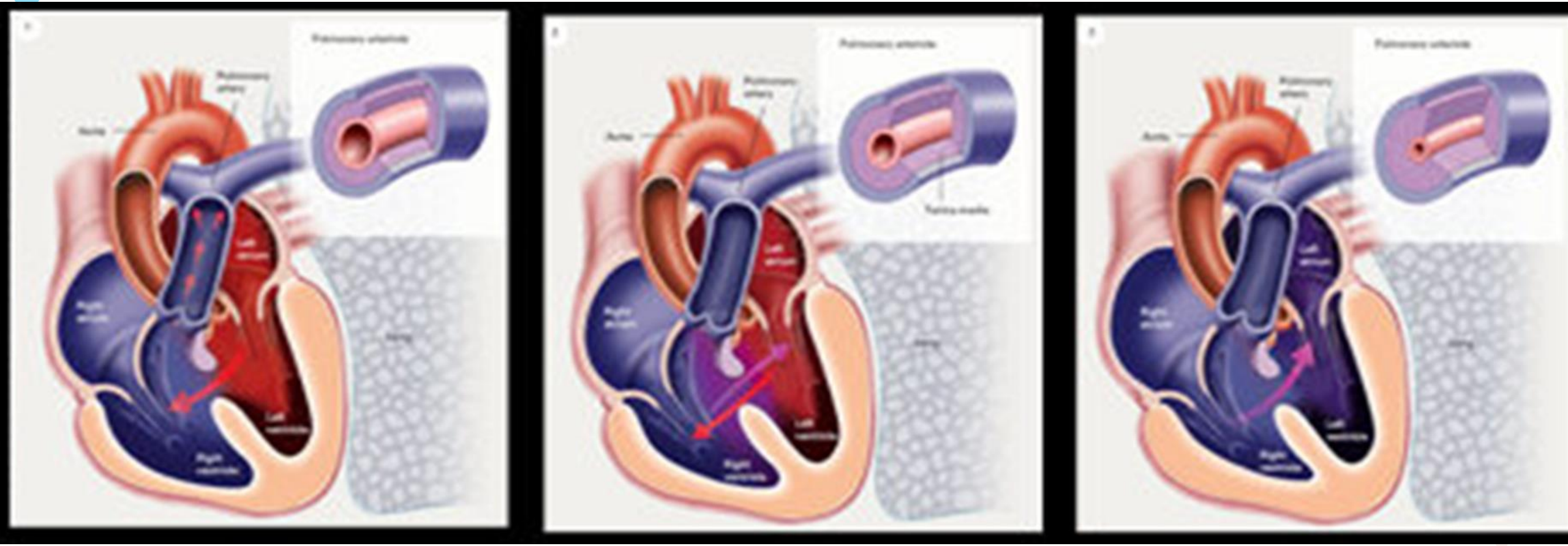
- Noncyanotic – „pink skin“
- Dyspnea
- Tired after feeding, failure to thrive
- Respiratory infections
- Murmur

Prognosis when treated

- Survival almost 100 %
- Quality of life is very good
- Even professional athletes

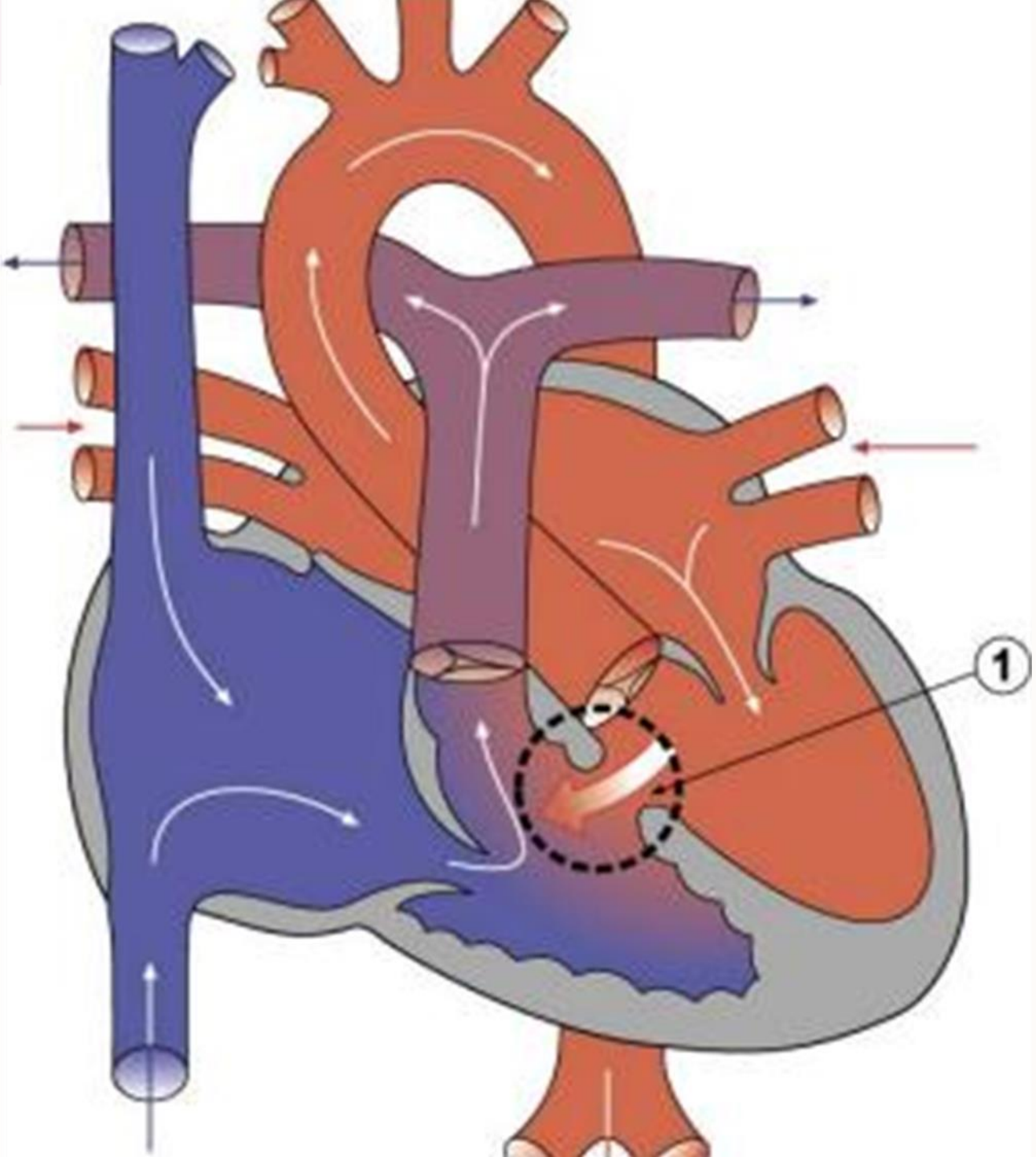
Prognosis when not treated properly or soon enough

- **Eisenmenger syndrome: 2. -10. year of life!**
- Long – therm damage of the pulmory arteries due to excessive blood flow
- The blood vessels in the lungs constrict = pulmonary resistence is rising !!!
- L-R shunt changes into R-L shunt
- Central cyanosis appear !!!
- Irreversible process = incurable; 10- year survival 50 %
- Only paliative treatement: Sildenafil, Bosentan, Prostacyklin
- Compensatory polyglobuly ... ↑ Viscosity ... Thromboembolic complications !!! (Th: hydratation !!!)



That‘s why..

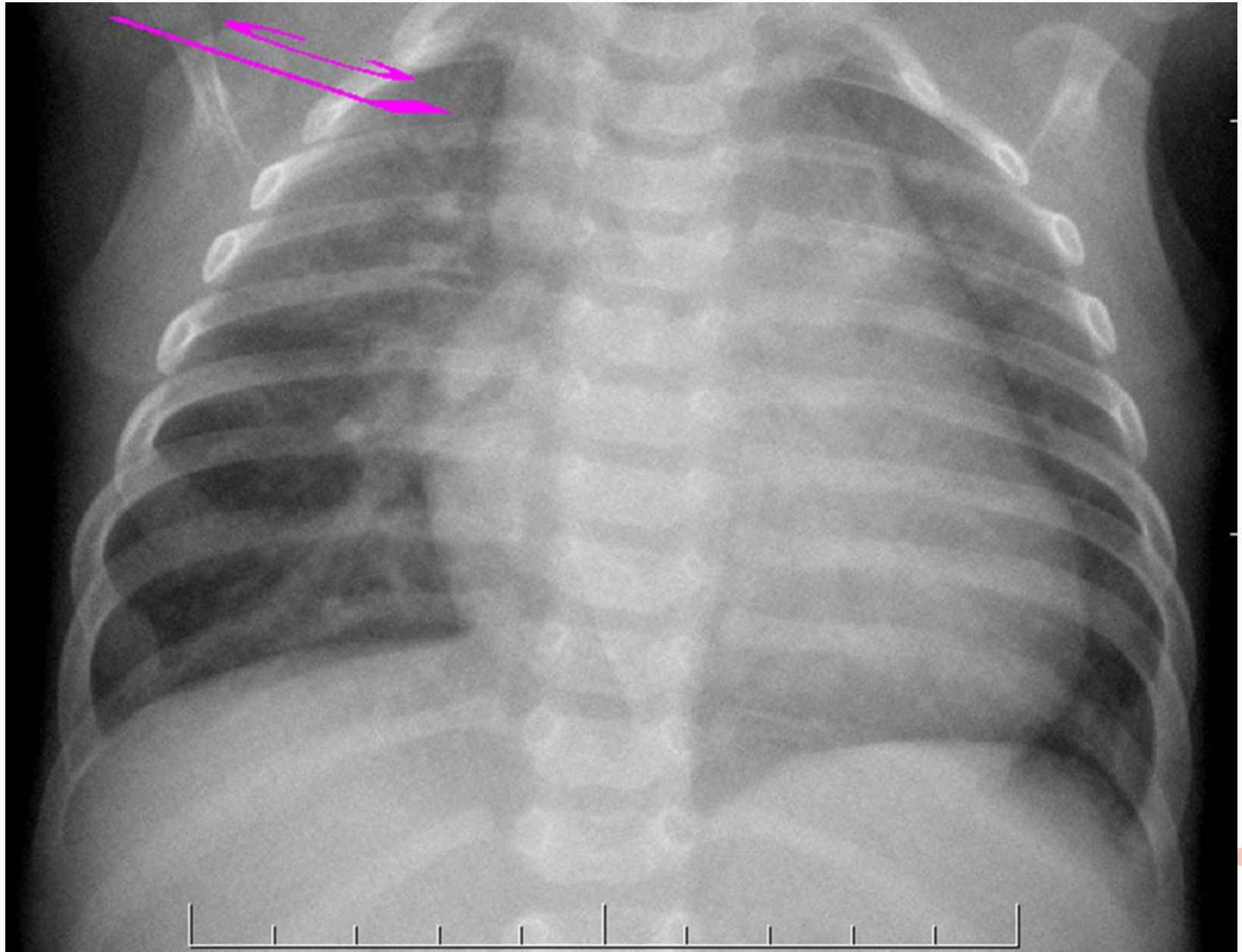
- Noncyanotic child …… BUT:
- Poor weight gain
- Recurrent respiratory infections
- ??? Murmur ???
- Rather send the patient to a pediatric cardiologist, than to treat Eisenmenger syndrome !!! !!!
- VCCs with L-R shunt

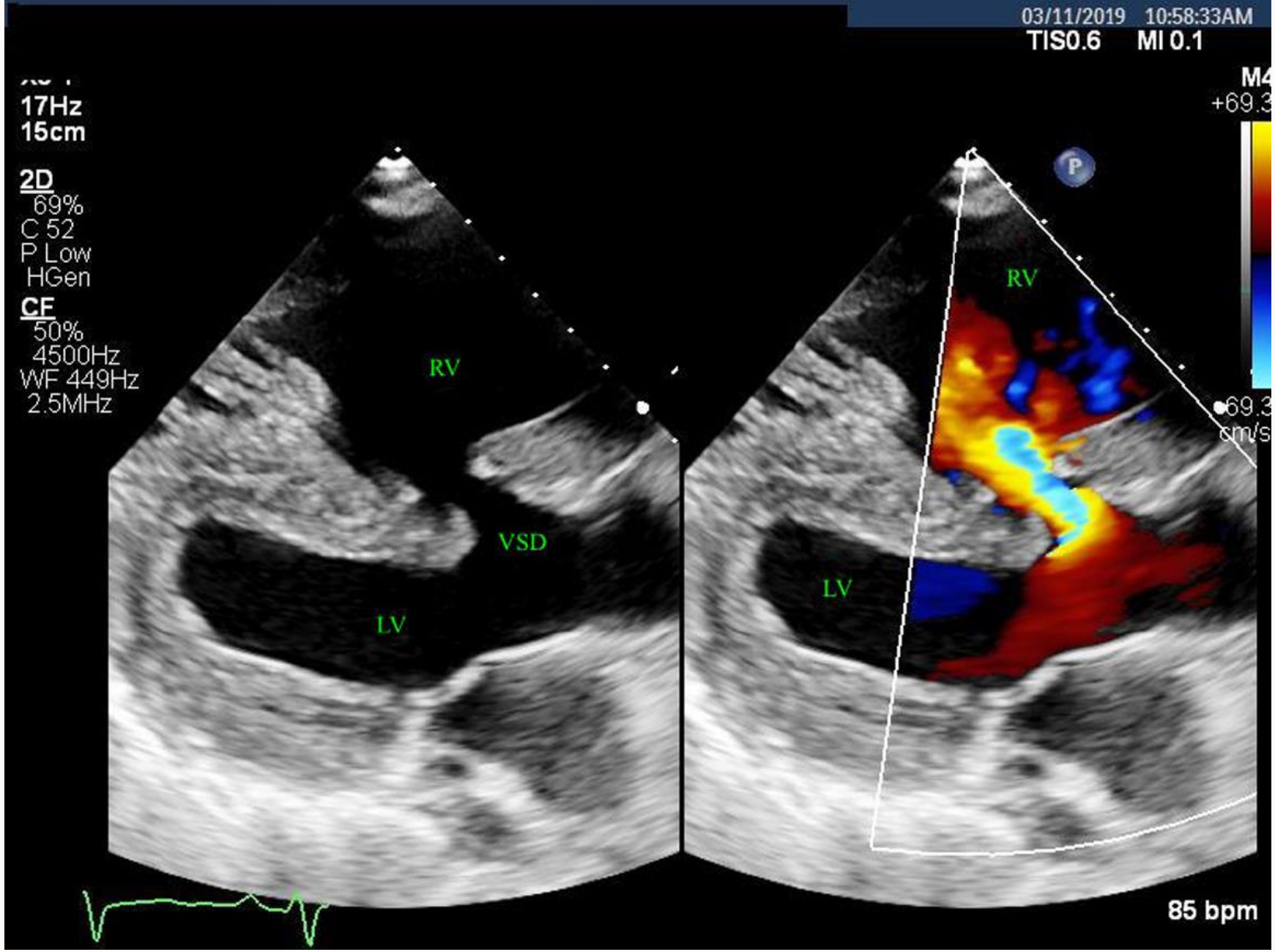


Ventricular septal defect

- The most common of all VCCs (41 %)
- **Systolic murmur** depends on the pressures inside of the heart (usually appears **after the drop of the pumonary vascular resistence**, individually **during first 2 months**) and **the size of the defect** (the bigger the „hole“ the less audible murmur and vice versa)
- **ECG:** normal or signs of hypertrophy of the LV
- **Chest X-ray:** prominent pulmonary vasculature, cardiomegaly (CTI > 0,5)
- **Echocardiography**
- Therapy: conservative (watch & wait) or surgical

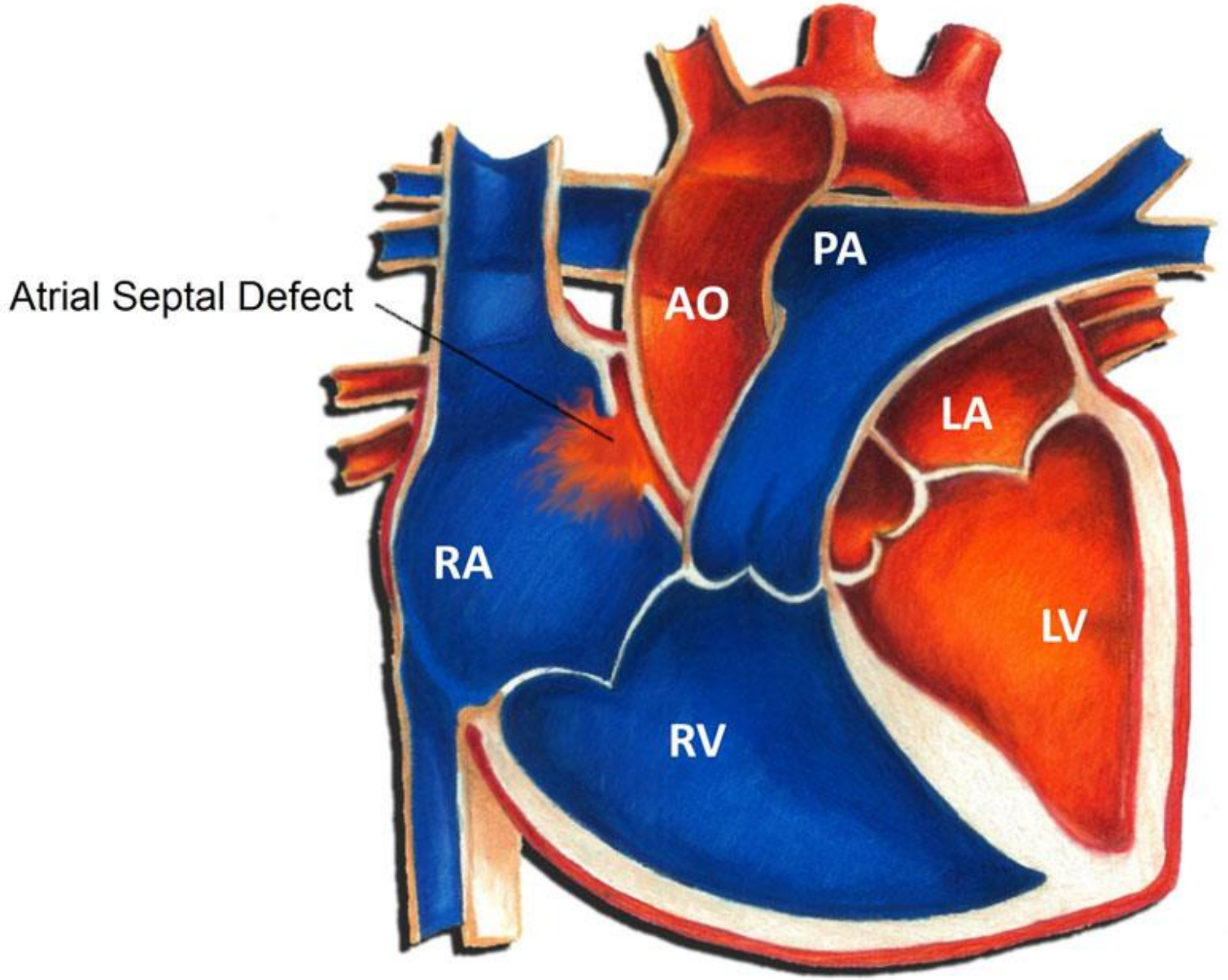
VSD - dilatation of the heart shadow, ↑ pulmonary circulation



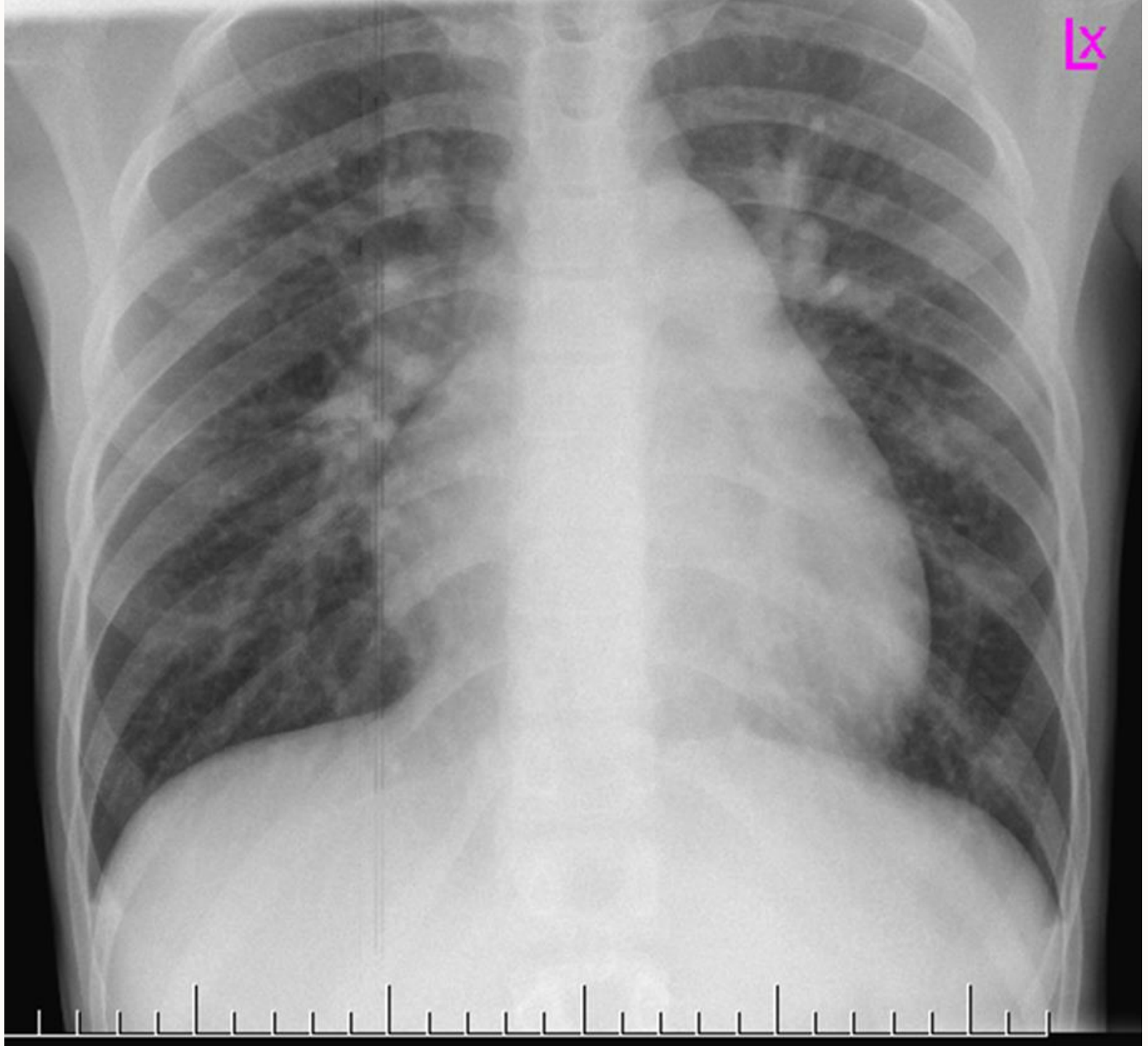


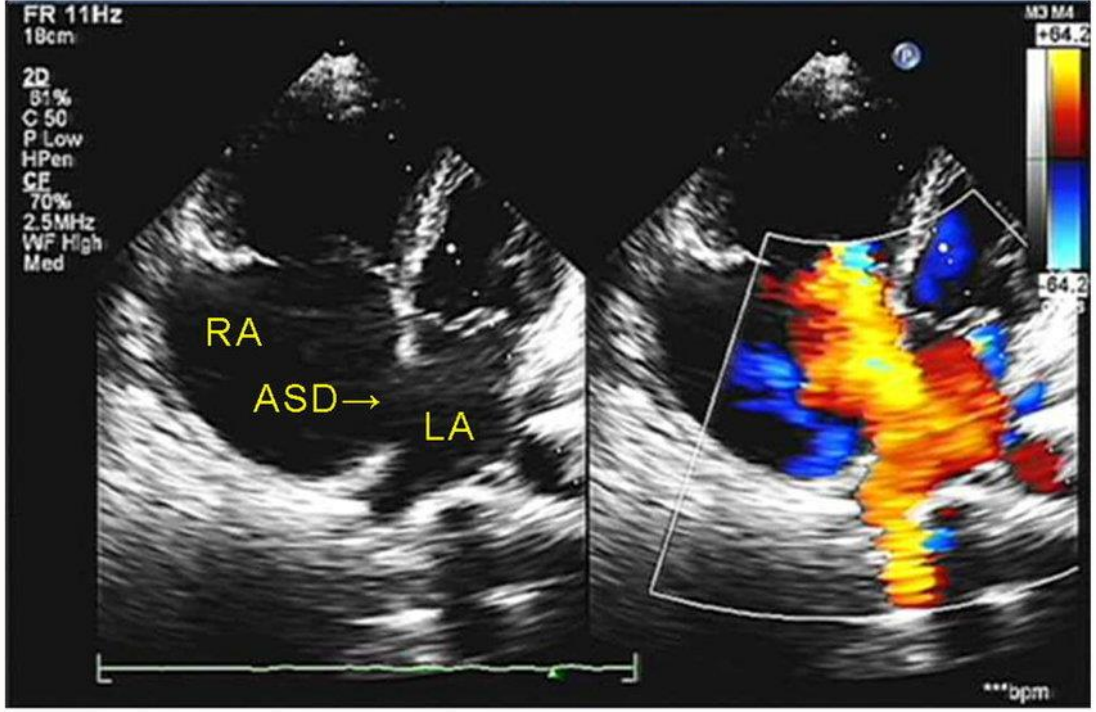
Echocardiographic evaluation of ventricular septal defects - Nayak - 2020 - Echocardiography - Wiley Online Library

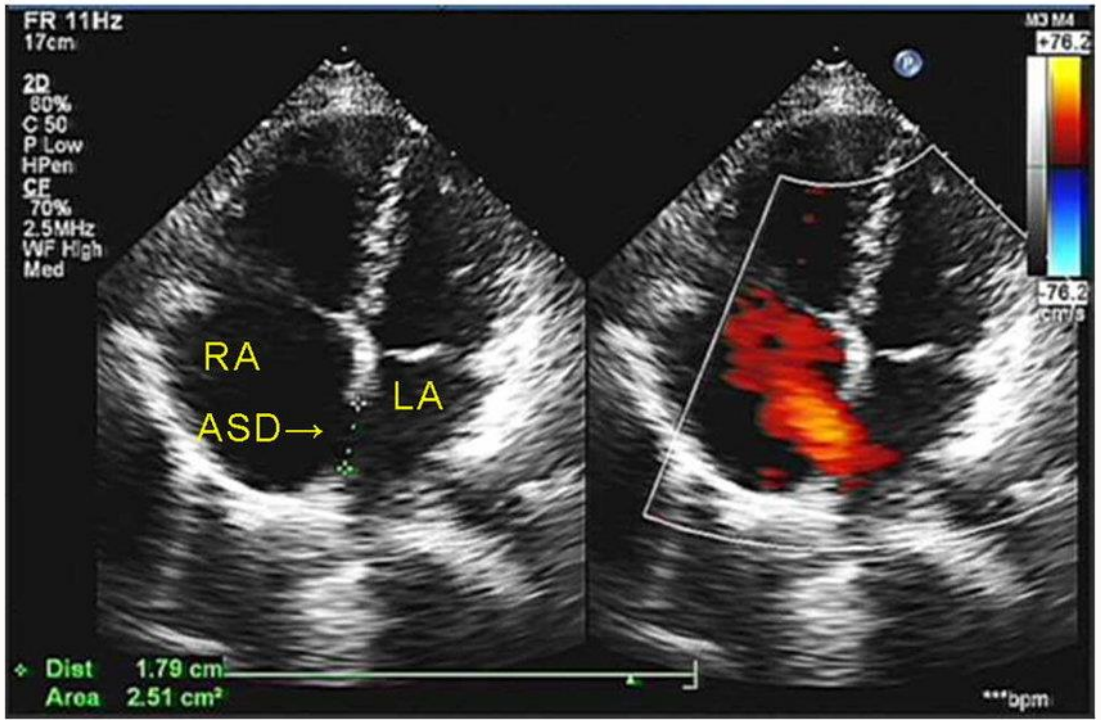
Atrial septal defect (9 %)



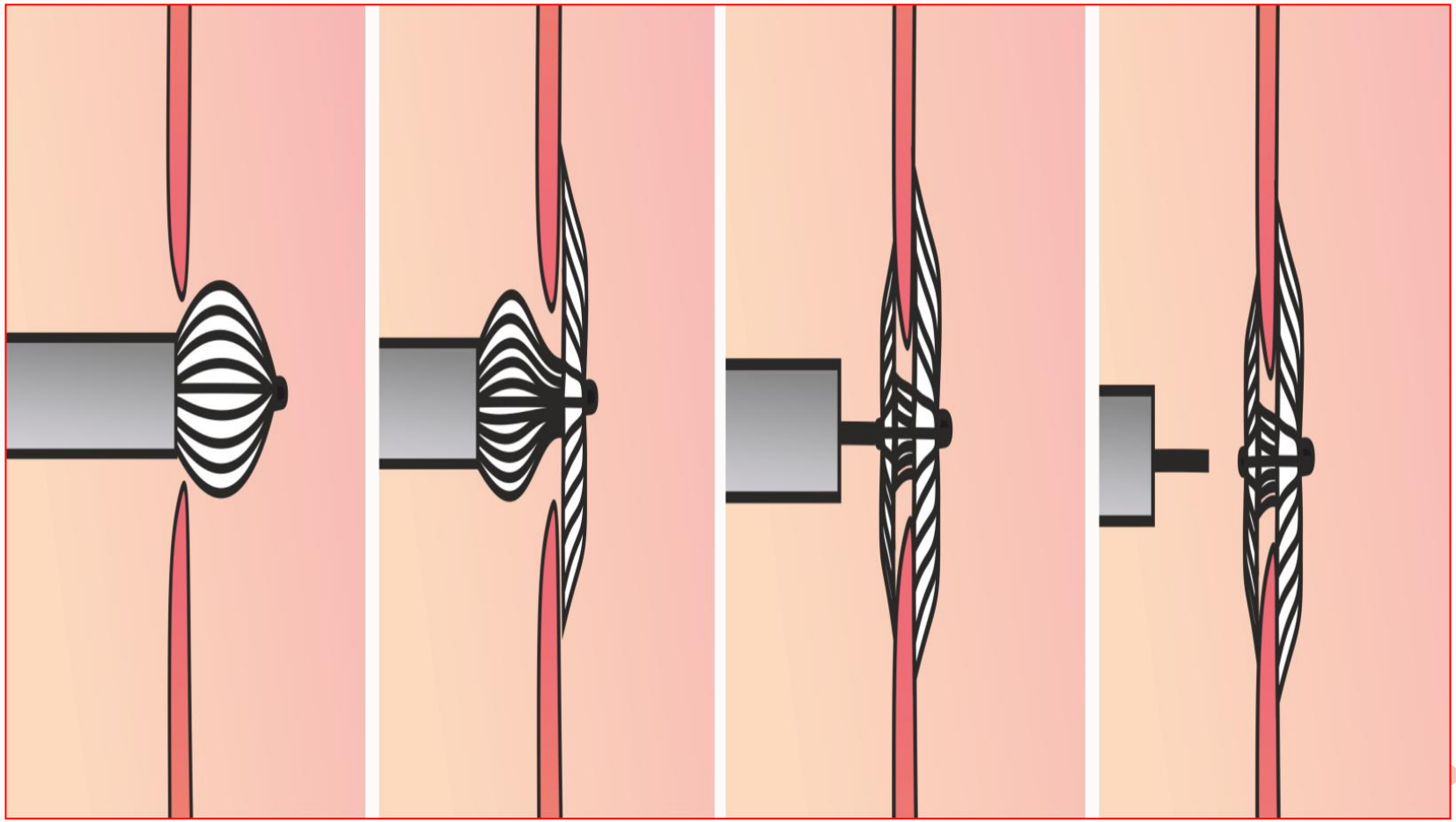
- **Systolic murmur** with punctum maximum above **pulmonary valve**, accentation/**splitting of the 2nd heart sound**
- **ECG:** hypertrophy of the right ventricule, IRBBB
- **Chest X-ray:** prominent pulmonary vasculature
- **Echocardiography**
- **Therapy:** cathetrisational or surgical

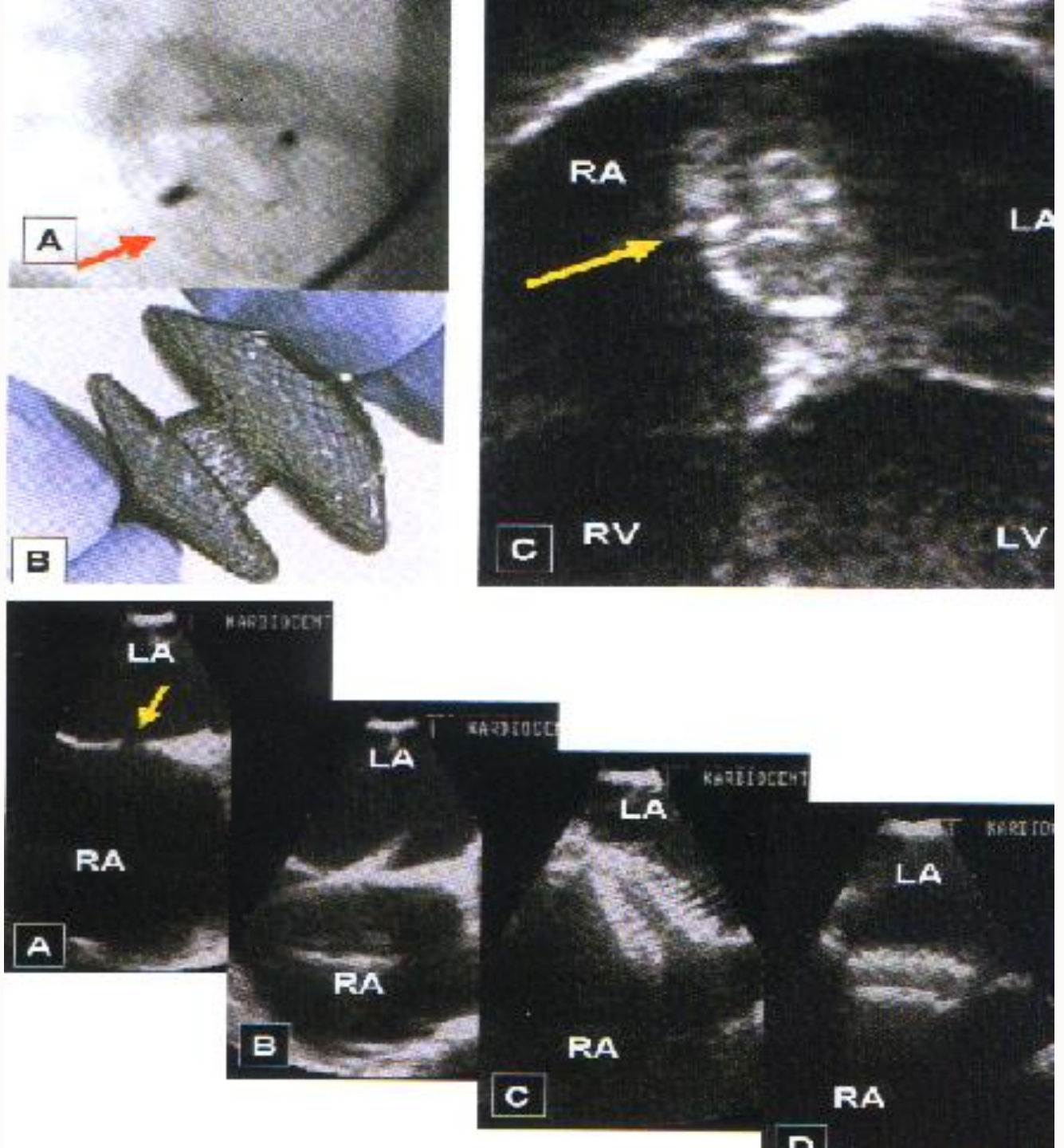



FR 11Hz
18cm
2D
81%
C 50
P Low
HPen
CE
70%
2.5MHz
WF High
Med
RA
ASD→
LA
M3 M4
+64.2
-64.2
***bpm

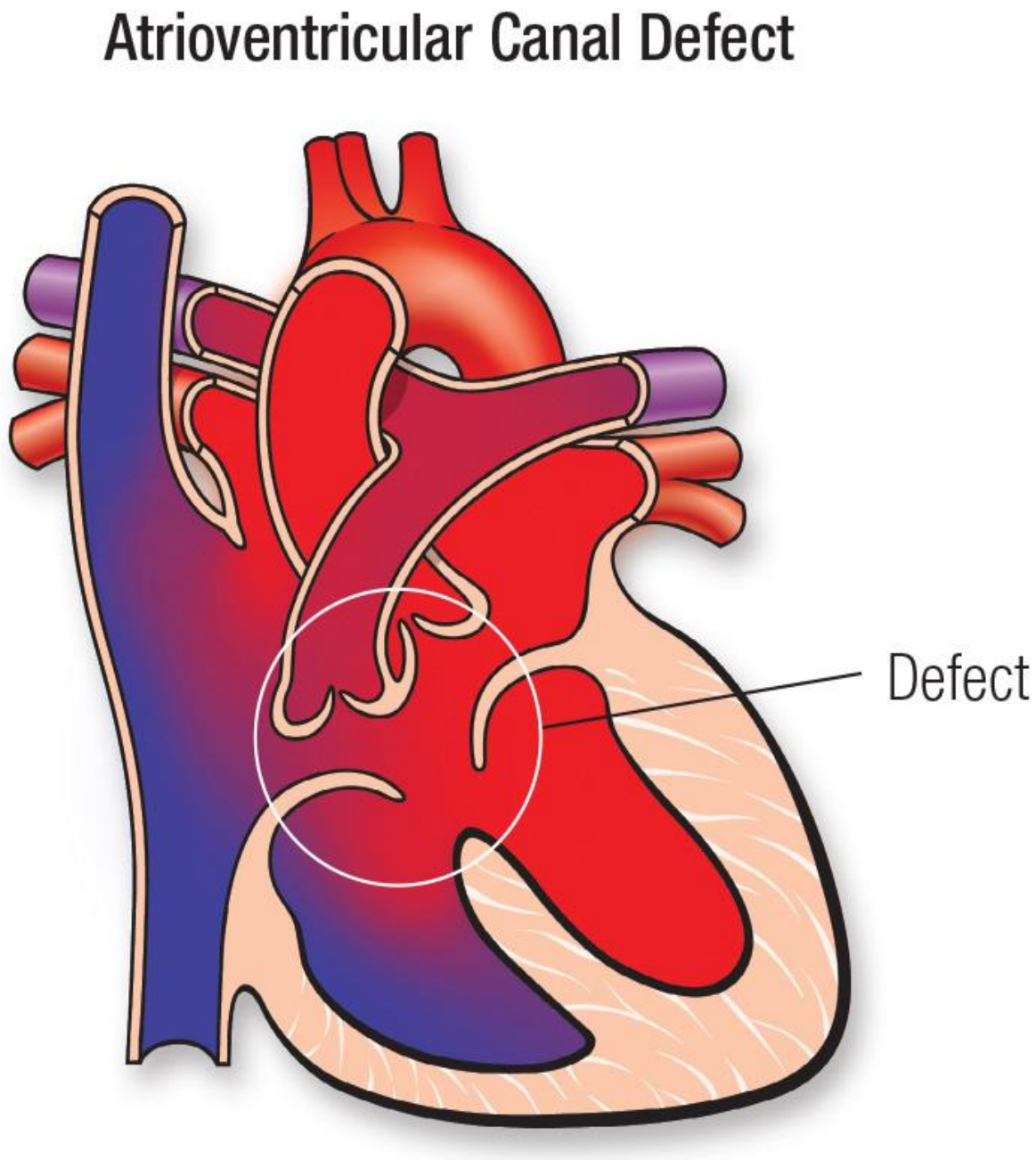

FR 11Hz
17cm
2D
80%
C 50
P Low
HPen
CE
70%
2.5MHz
WF High
Med
RA
LA
ASD→
M3 M4
+76.2
-76.2
Dist 1.79 cm
Area 2.51 cm²
***bpm

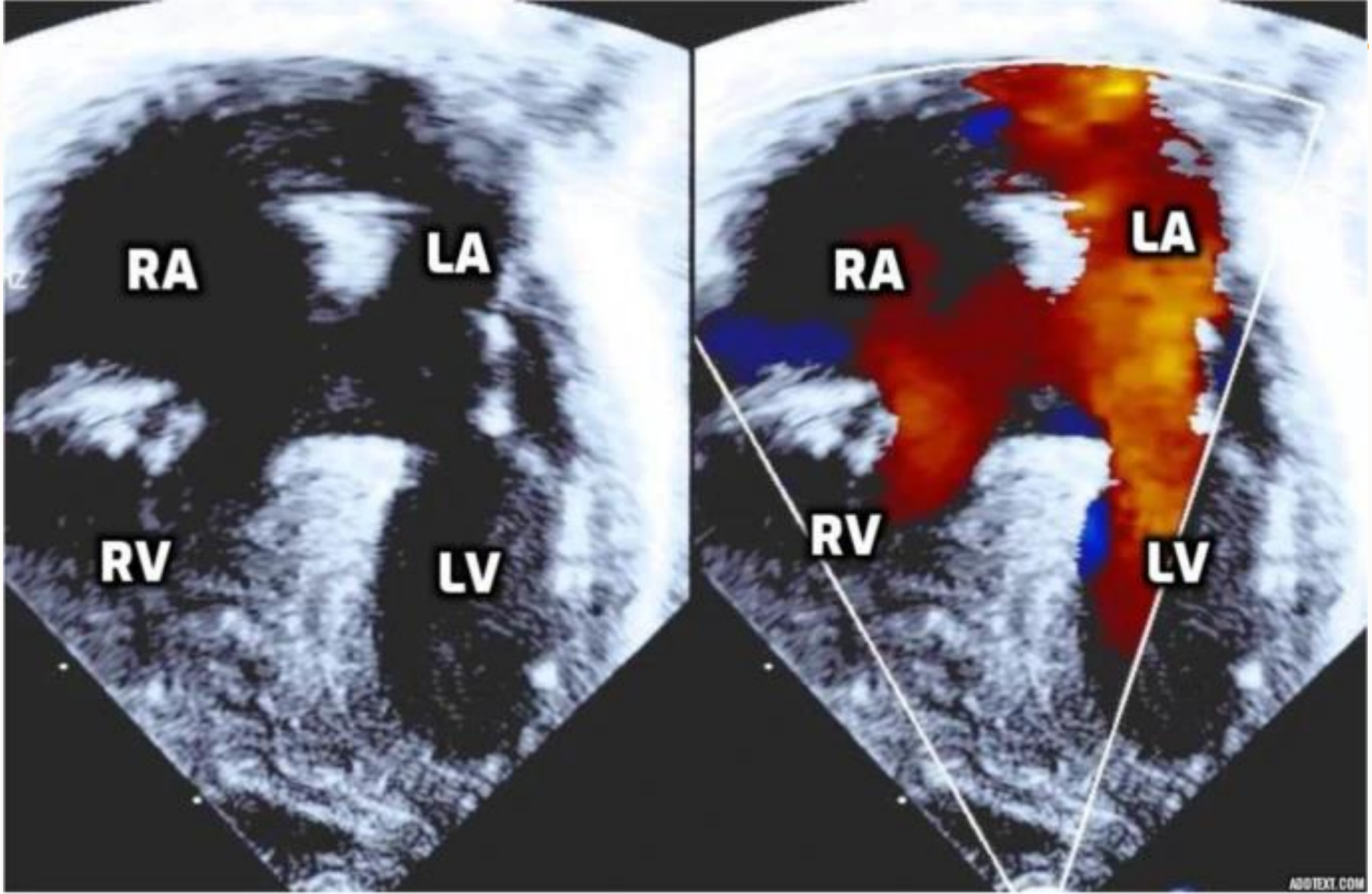
Cathetrisational closure of the ASD
=> Amplatz occluder



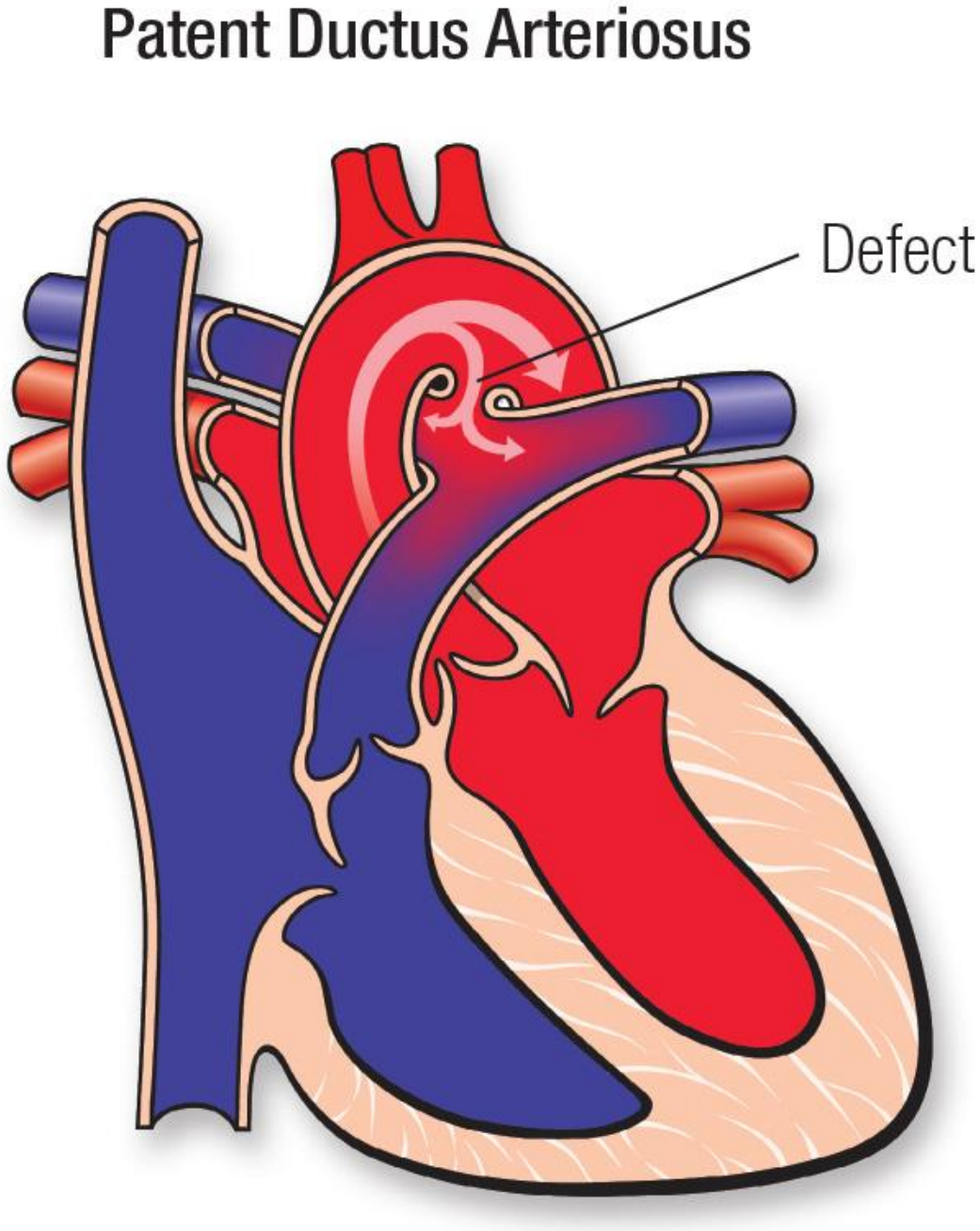

A
B
RA
LA
C
RV
LV
LA
RA
A
LA
RA
B
LA
RA
C
LA
RA

Atrioventricular septal defect – AV canal 4 % (m.Down 40 %)

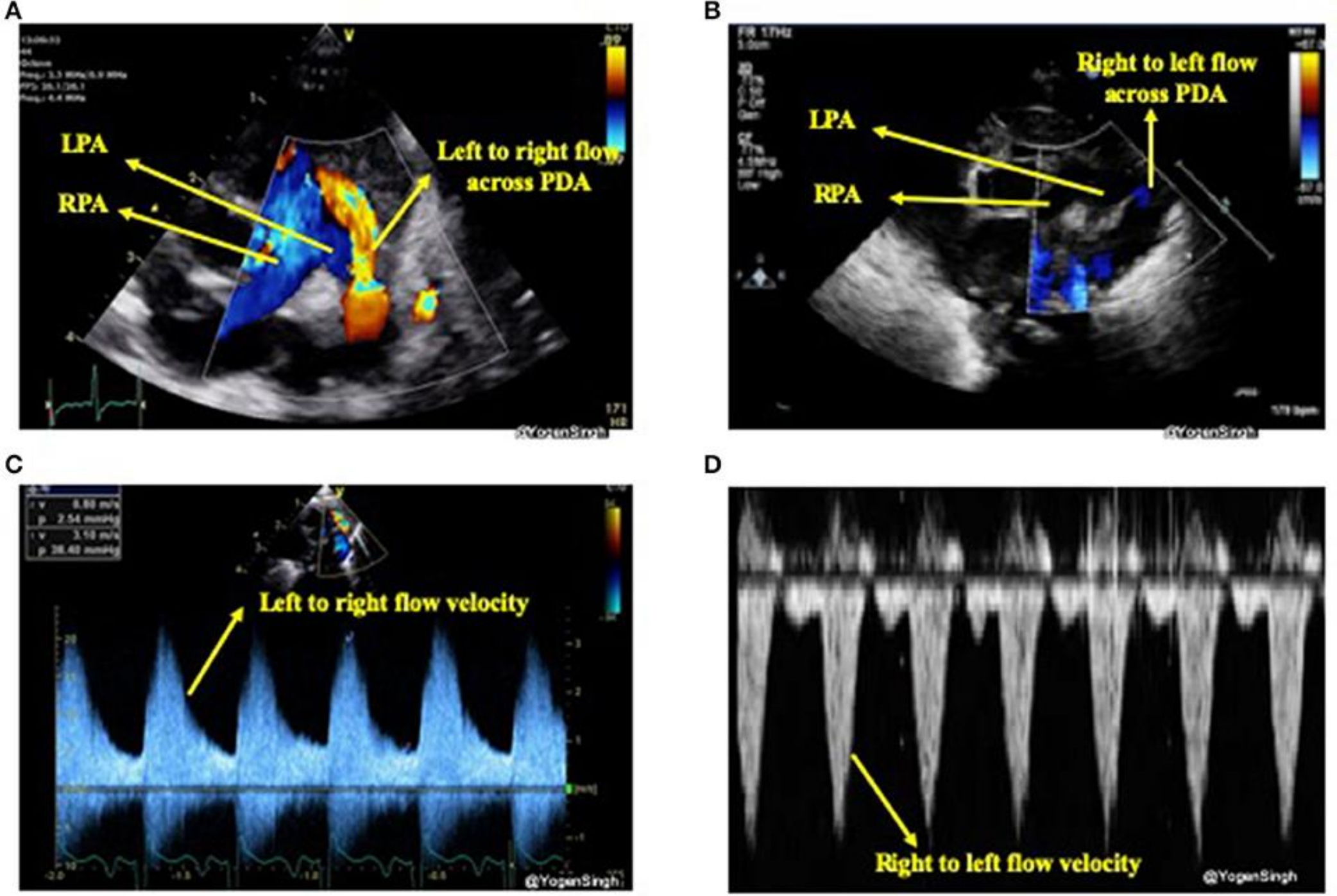



RA
LA
RV
LV
RA
LA
RV
LV
ADDTEXT.COM

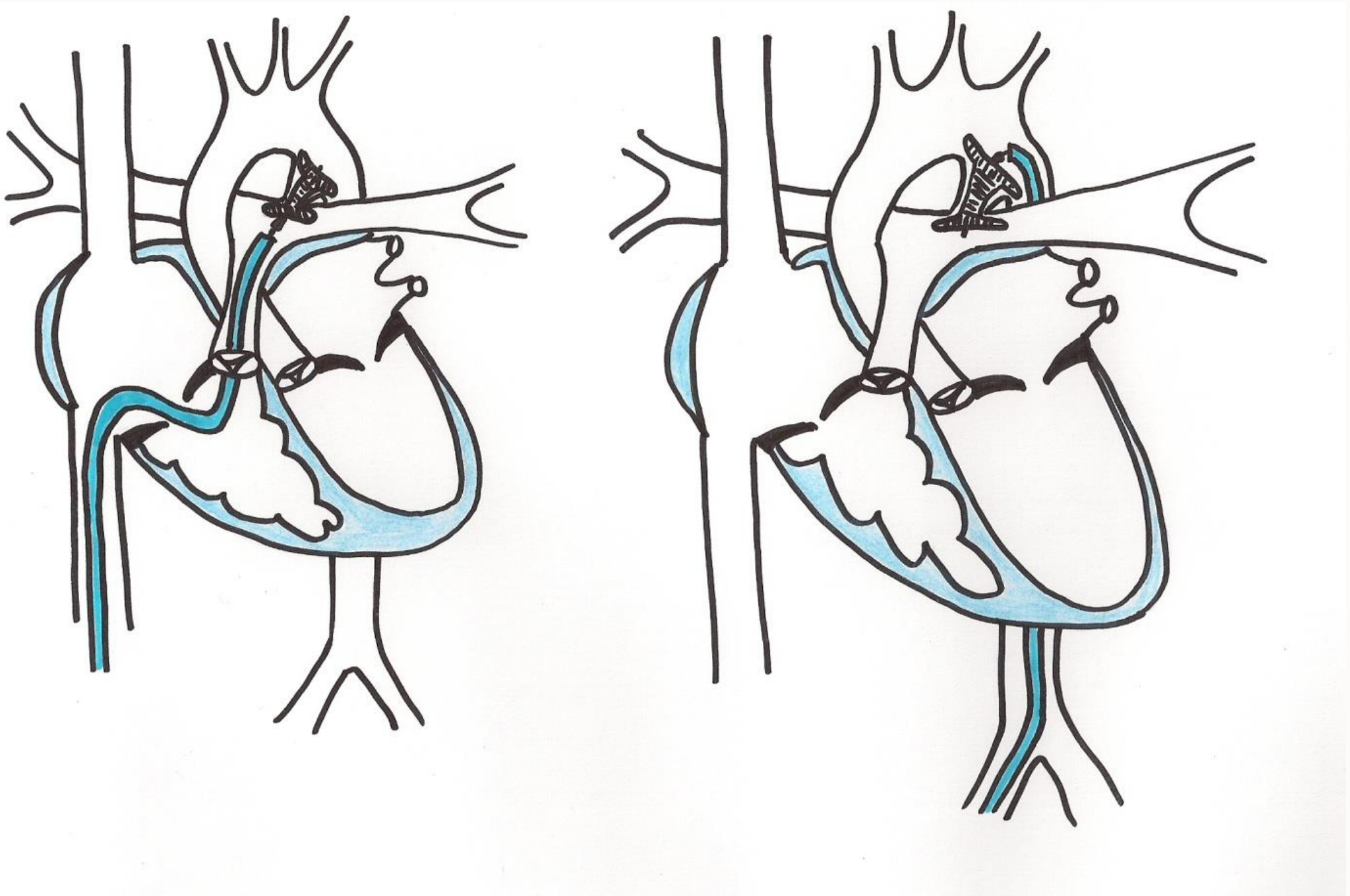
Persistent ductus arteriosus



- Incidence > 50 % in newborns with low birth weight (< 800 g)
- **Continuous/systolic murmur** typically **below left clavicule**
- **ECG:** hypertrophy of the LV
- **Chest X-ray:** prominent pulmonary vasculature, cardiomegaly
- **Echocardiography**
- **Therapy:**
  - in newborns (especially preterm) starting with medicamentous closure (Ibuprofen, Paracetamol i.v. repeatedly) or surgical ligation if non-effect,
  - in bigger children (at least > 3 kg but the more the better) cathetrisational closure or surgical ligation



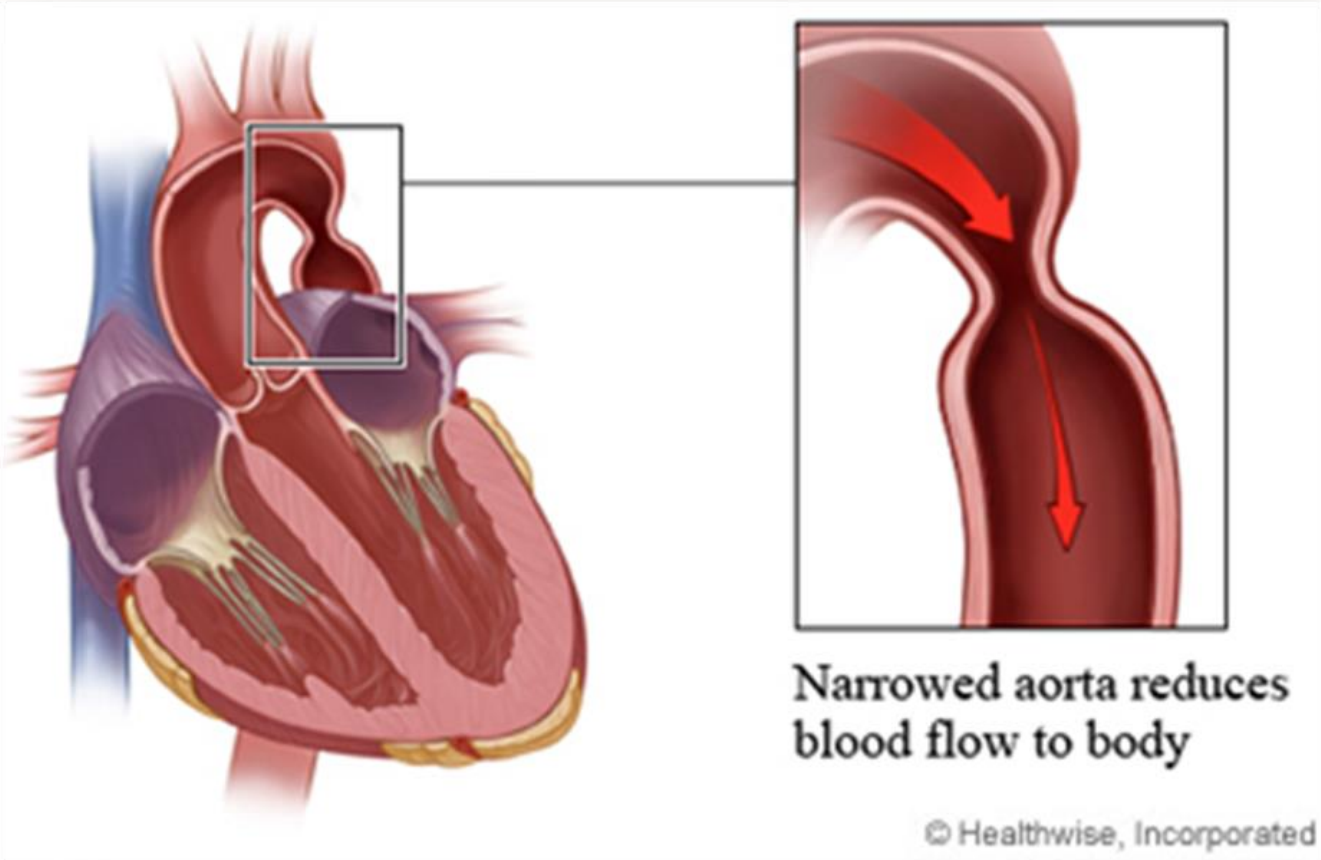
https://www.frontiersin.org/files/Articles/573627/fped-08-573627-HTML/image_m/fped-08-573627-g004.jpg



Noncyanotic VCC – without shunt: CoA (5 %)

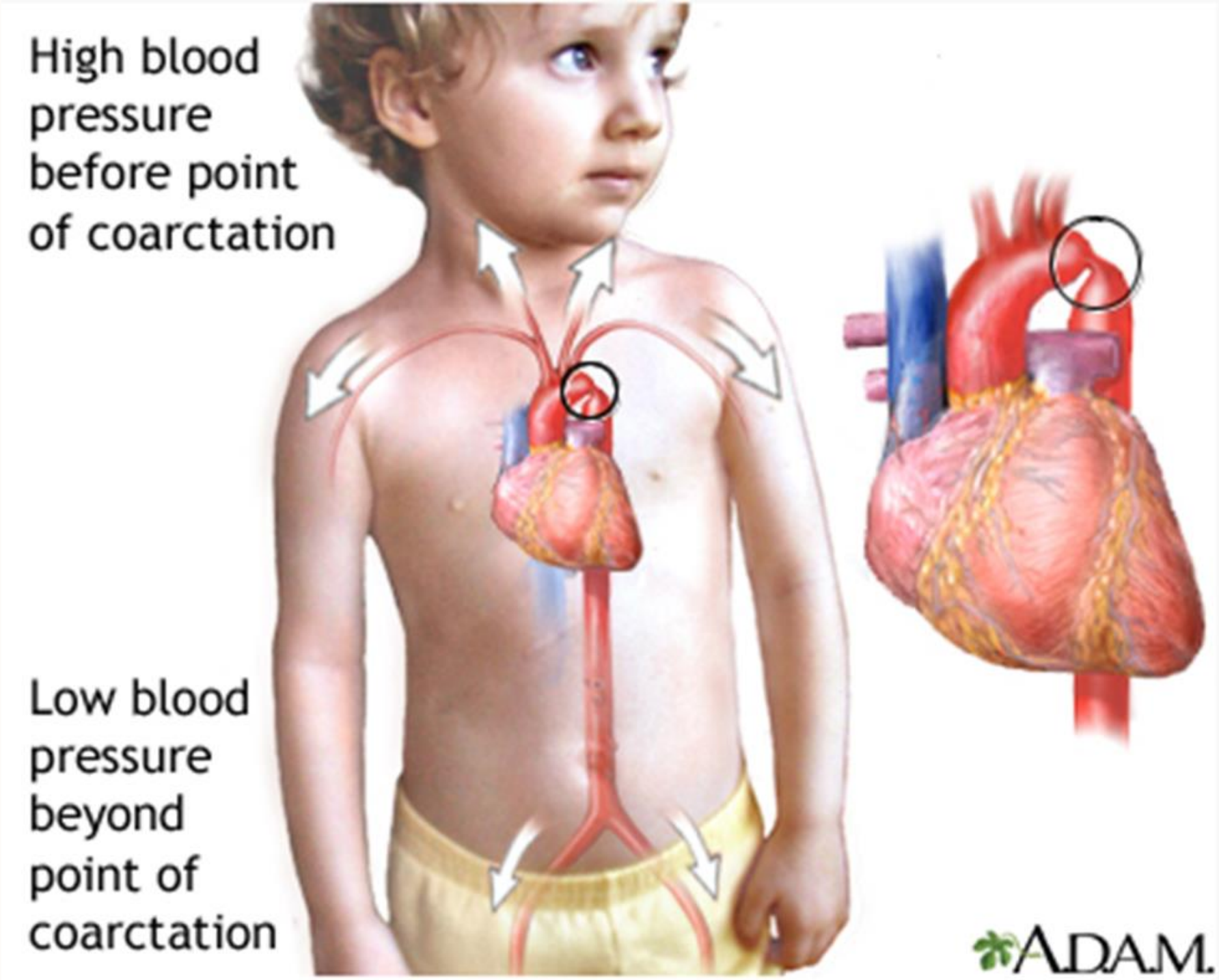
- Narrowing of the aortic isthmus => overgrowth of PDA tissue to Ao => when closing („contraction“) pulling Ao and causing its narrowing,
  - Rarely narrowing of thoracic or abdominal aorta
- More often boys
- Most common VCC in girls with

  Turner´s syndrome
- Often present bicuspid AoV (85 %)
- 2 types:
  - **In newborns => critical !!!**
  - **In bigger children => non-critical**



Common clinical symptoms

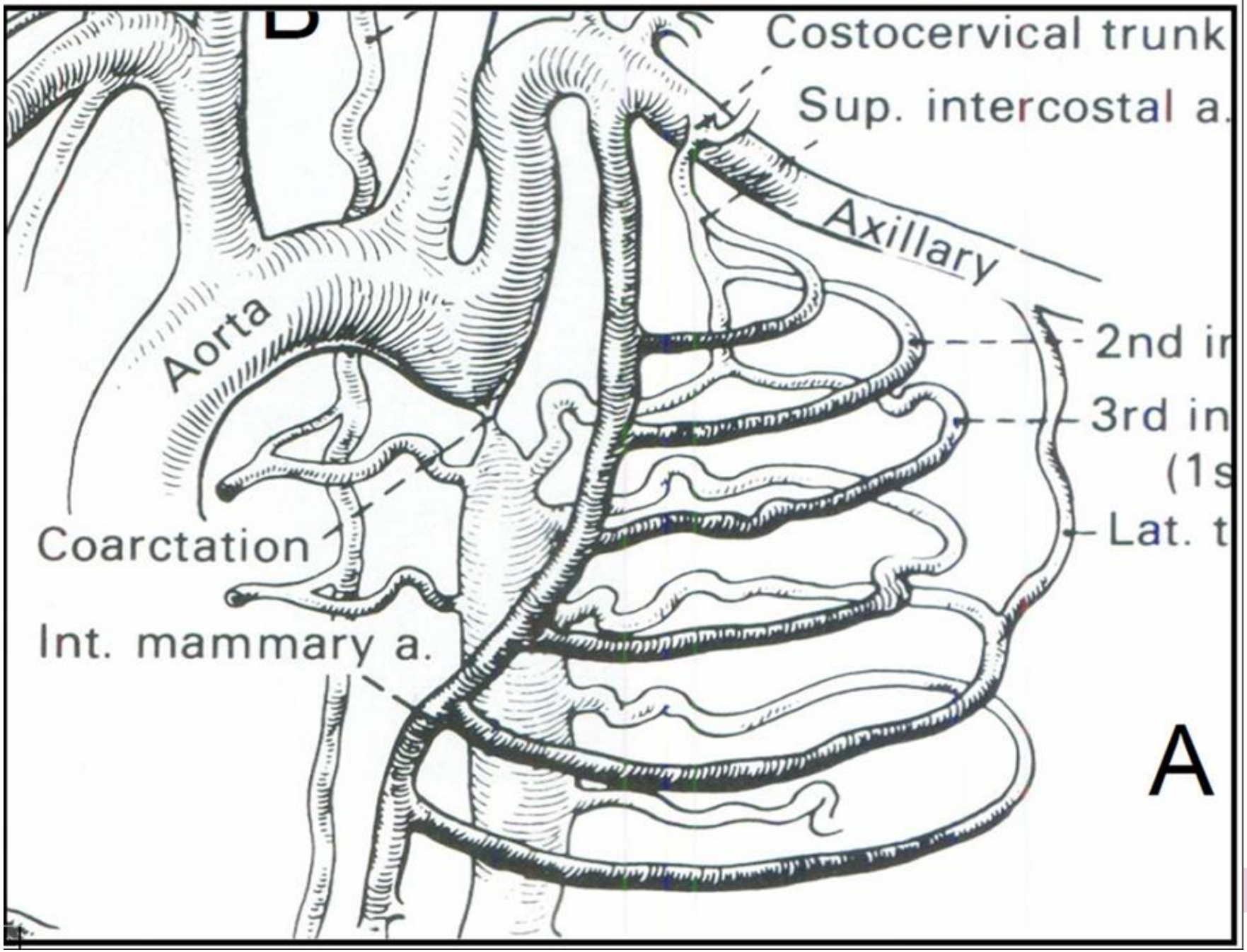
- Obstruction of the blood flow to lower extremities => hypotension + weak/no palpable pulses on the aa. femorales & hypertension + strong pulses on the arms
- **Clinical blood pressure gradient** => the difference of the blood pressure in the upper and lower extremities **20 mmHg and more** is significant !!
- **Systolic murmur**, often audible the best (**punctum maximum**) in the **interscapular area**
- **Echocardiography – abnormal pulsatility in the abdominal aorta!**

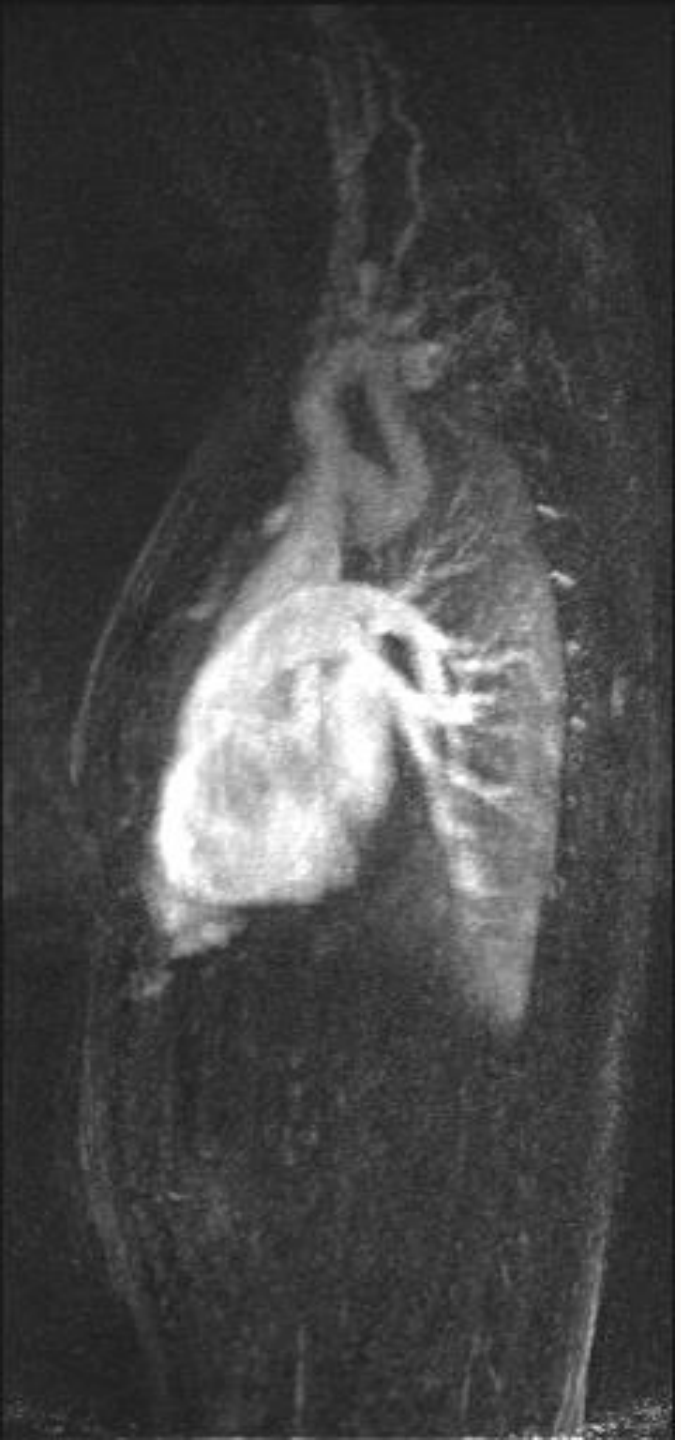

High blood pressure before point of coarctation
Low blood pressure beyond point of coarctation
A.D.A.M.

Clinical symptoms according to the age

- **Newborns - critical CoA** => symptoms of LCO + ↑pulmonary blood circulation + mild central cyanosis
- **Bigger children - non-critical CoA:**
  - Hypertension !!!
  - Possibly palpable pulsations on aa.femorales thanks to the system of collaterals
  - Epistaxis, impaired vision
  - Headache, migrenes, stroke

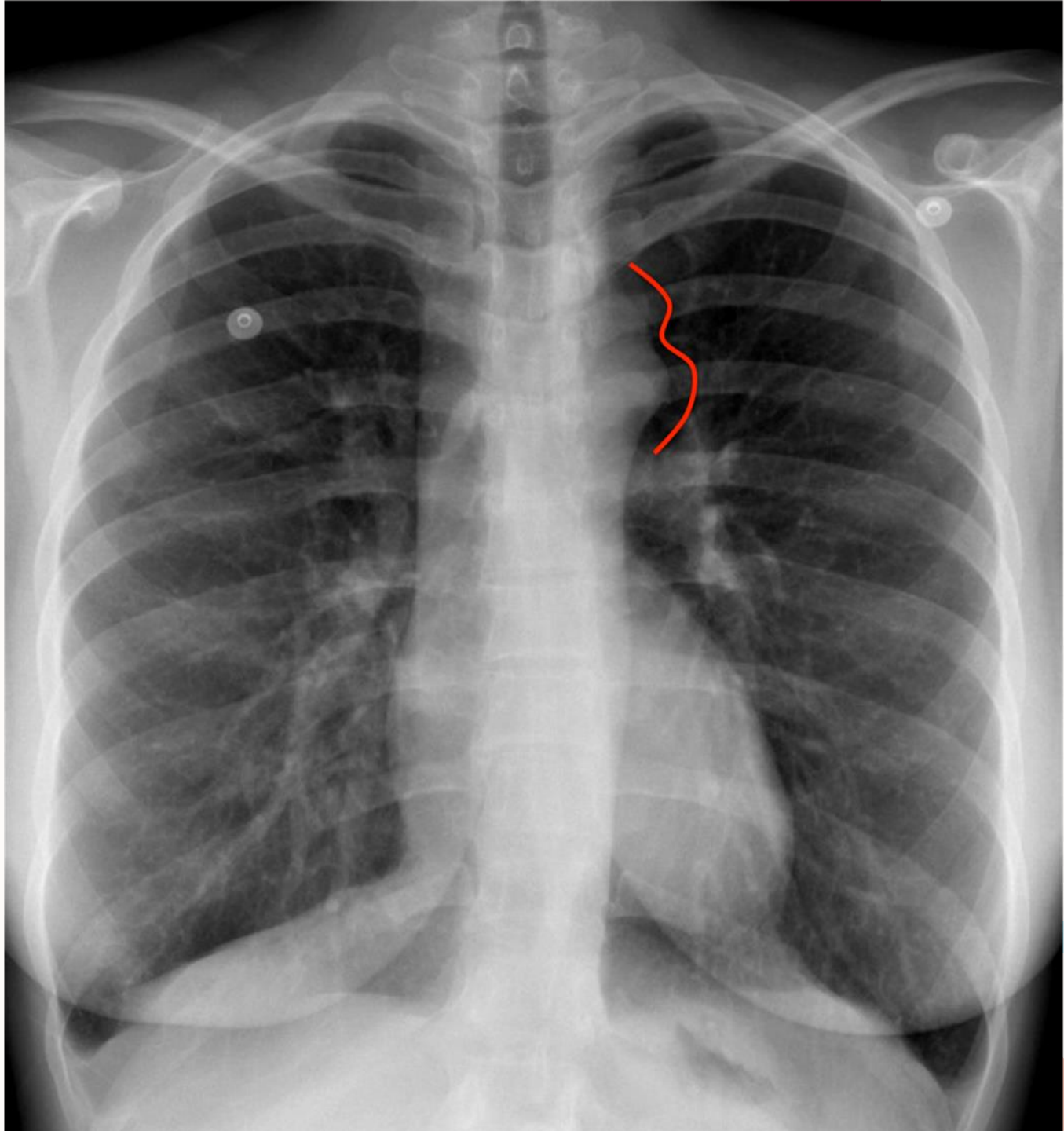
View of intercostal collaterals in coarctation of Aorta

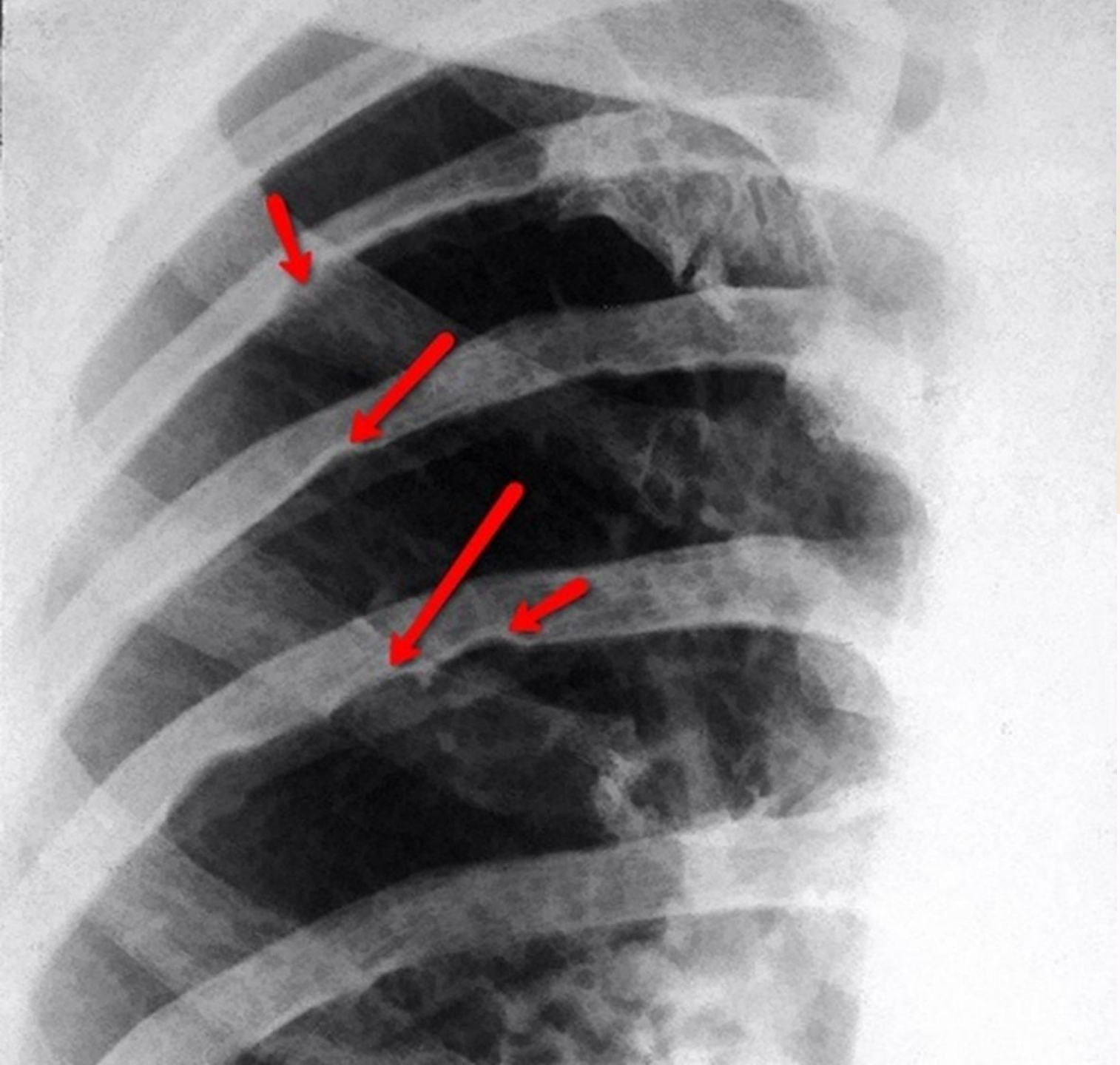


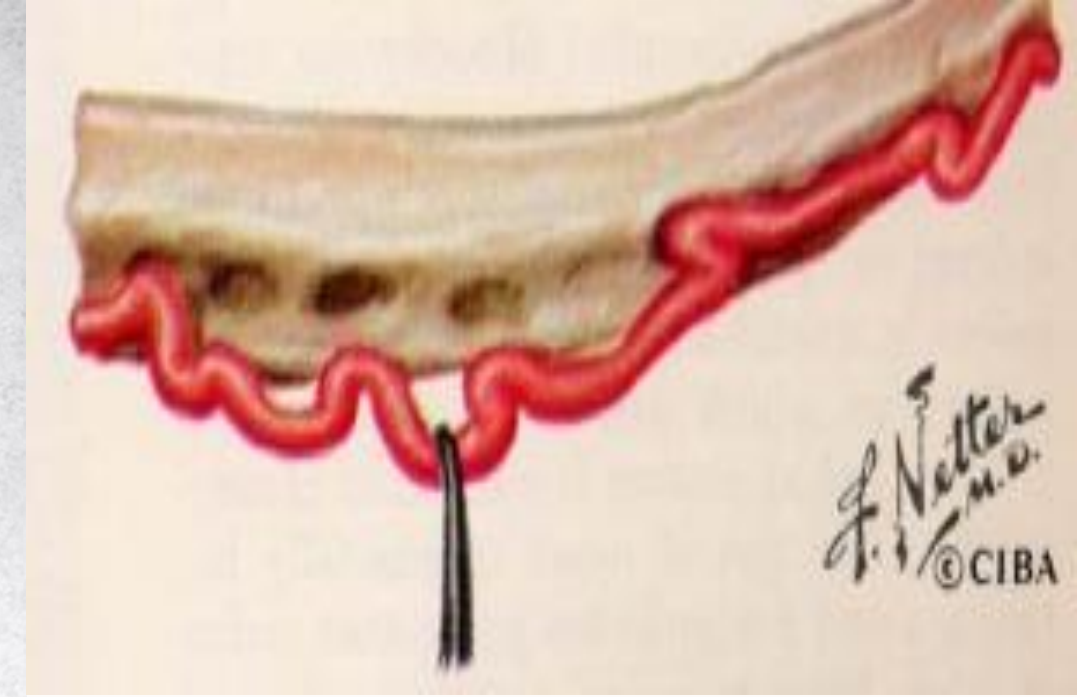


System of collaterals to the aorta usually from left subclavian artery
(MRI on the left side)

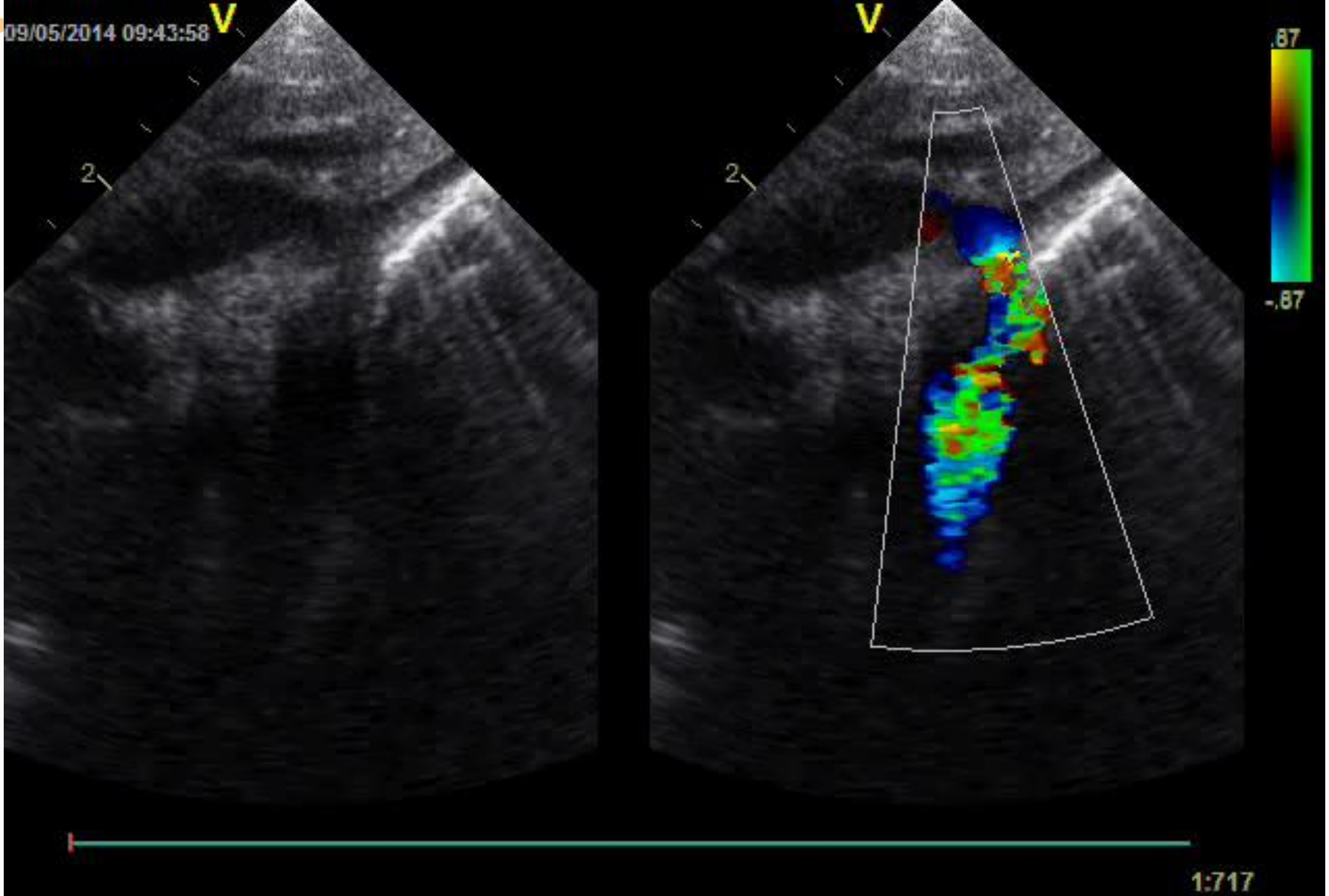
Sign of „3“ on the X-ray (on the right side)







Rib notching – caused by dilated intercostal arteries, collaterals respectively that are imprinting into the inferior margin of the ribs – „small bites“ look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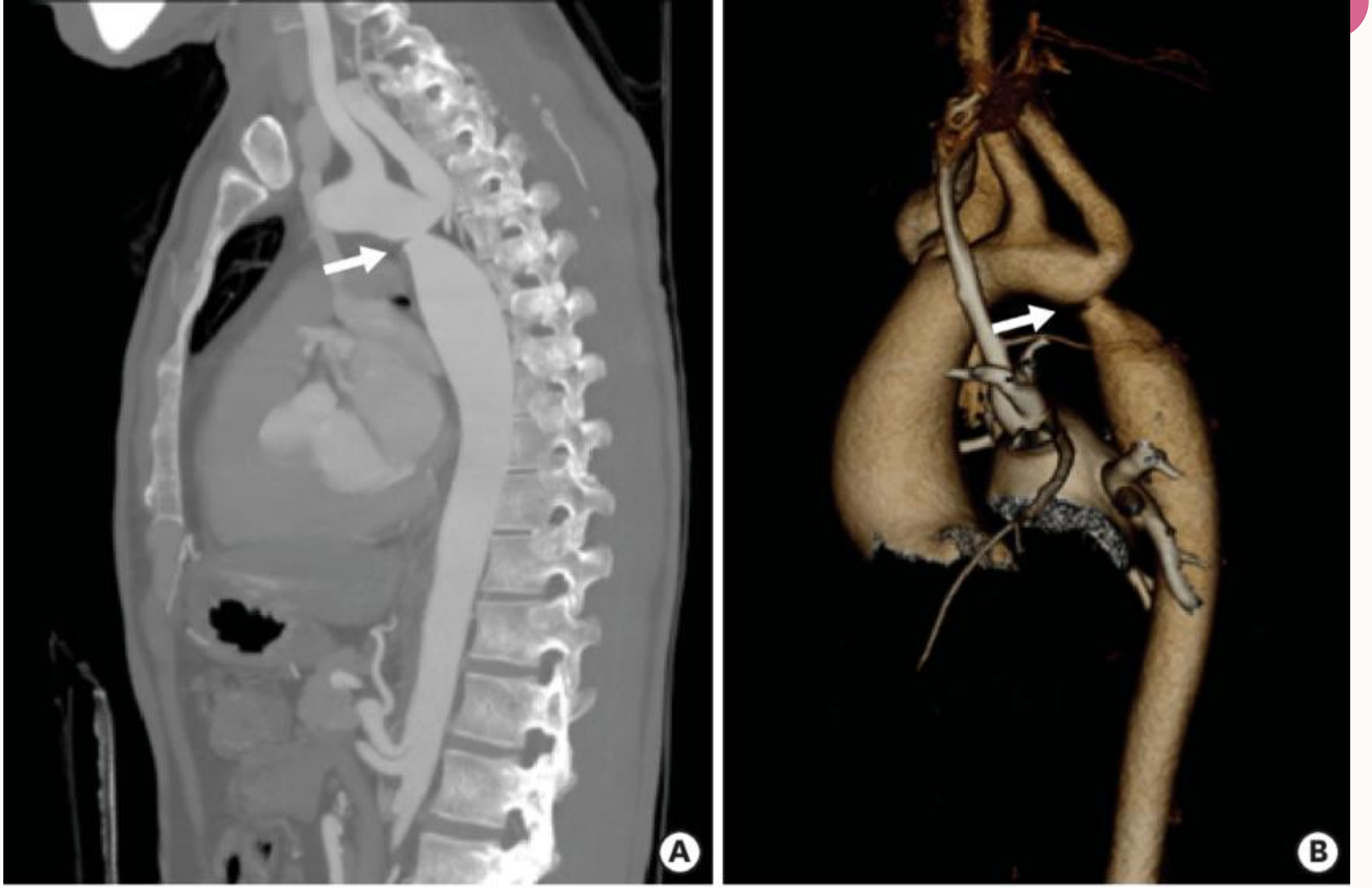
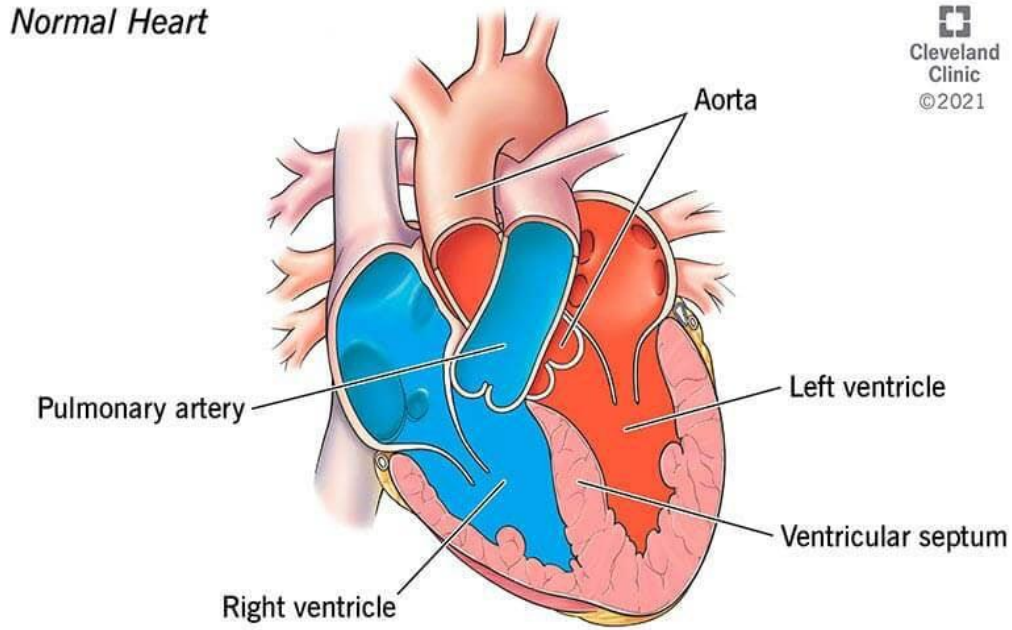


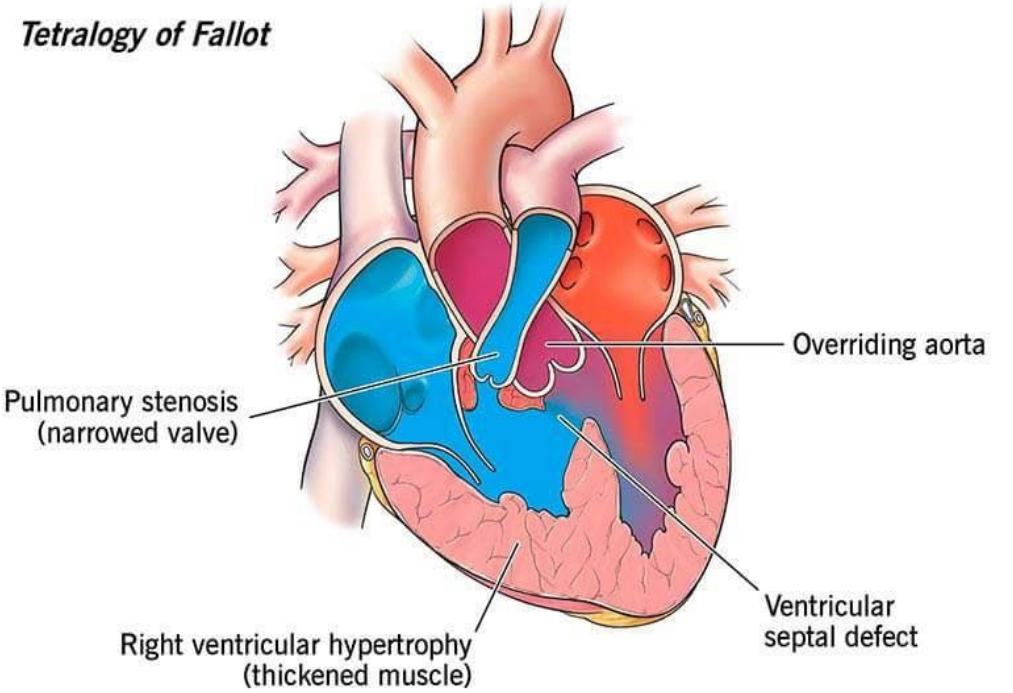
https://www.researchgate.net/figure/A-Computed-tomographic-angiogram-is-revealing-coarctation-of-the-aorta-just-at-the_fig2_331252305

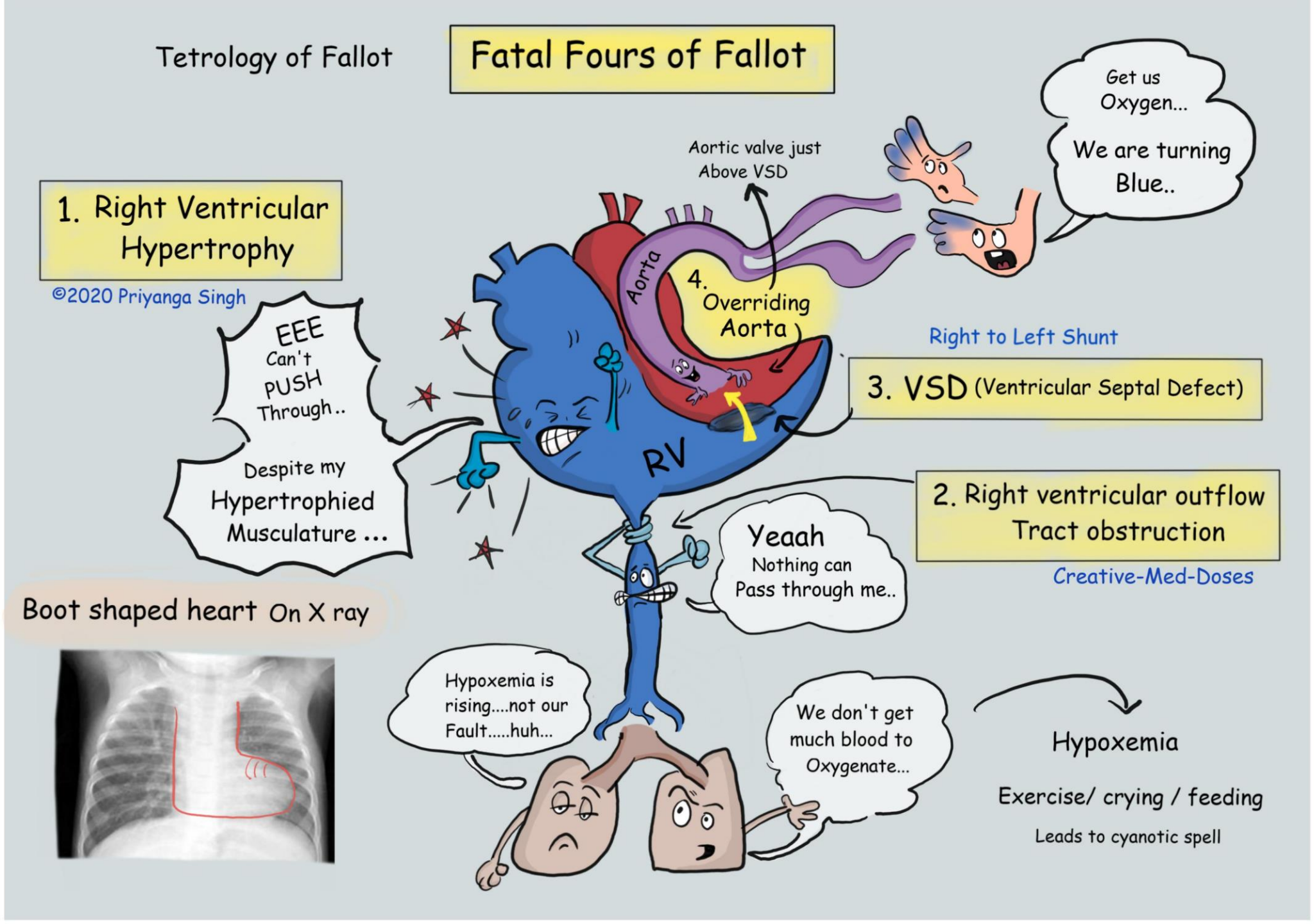
- **Therapy:** according to the age and technical possibilities surgical or cathetrisational
- **Possible long-term complications** even after correction:
  - 1. Recoarctation (clinical gradient > 20 torr!!!)
  - 2. Aneurysms – incidence 9 %, risk of the rupture, dissection of the aorta
  - 3. BAo – 2/3 of all children with CoA, risk of the dilation of the aortic root, valvular stenosis/regurgitation, IE
  - 4. Hypertension – atherosclerosis, hypertrophy LV (risk factor – delayed time of correction)
  - 5. Endothelial dysfunction – in delayed time of correction

Cyanotic – noncritical: TOF (4 %)

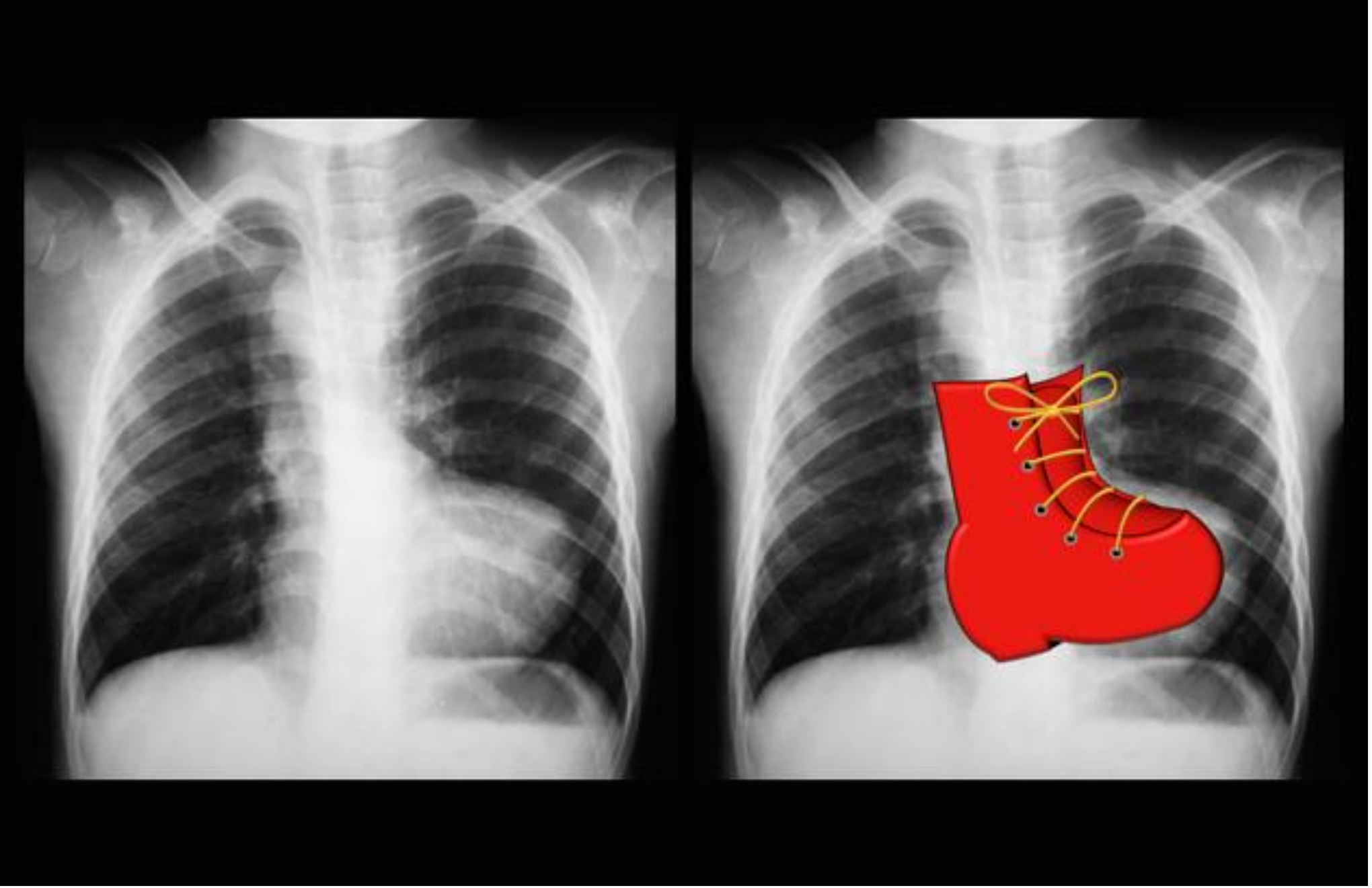
- Tetra = 4:
  - **Pulmonary stenosis** (valvular)
  - **Hypertrophy of RV** => pulmonary stenosis (infundibular/subvalvular)
  - **Ventricular septal defect** (VSD)
  - **Overriding of VSD by the aorta** (aorta shifted to the RV => 50 % arising from RV and 50 % from LV) => above VSD







RV outflow tract obstruction => high pressure in the RV (suprasyst.) => **R-L shunt => central cyanosis => sat.O2 75-90 %**



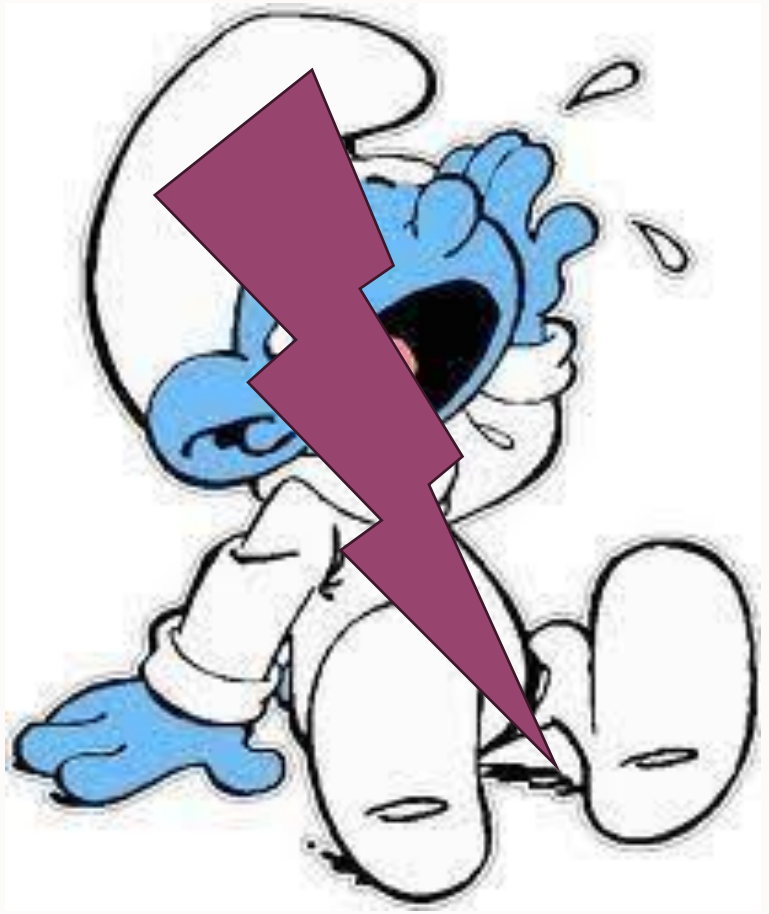
Boot-shaped heart

Hypertrophy of RV

Missing pulmonary knob

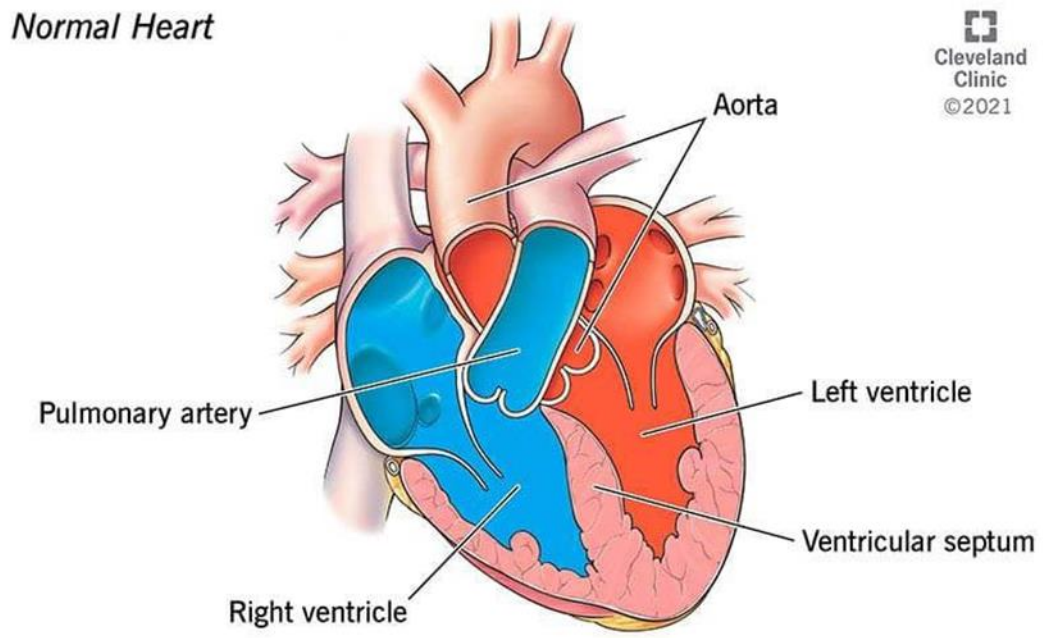
↓ pulmonary circulation

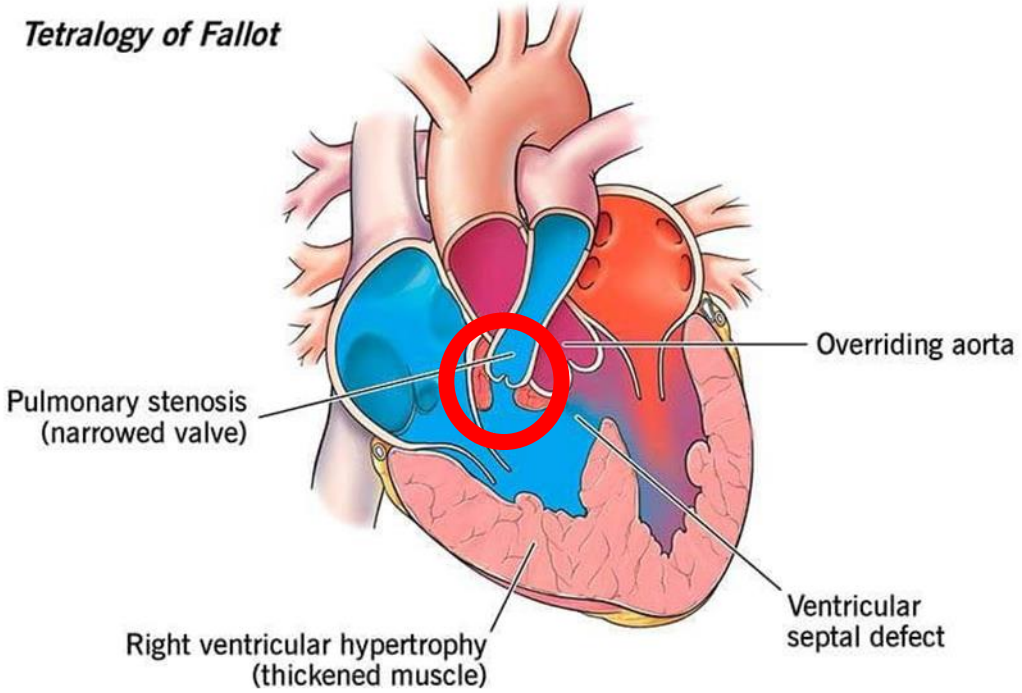
https://radiopaedia.org/cases/tetralogy-of-fallot-6



Urgent situation – hypoxic spell

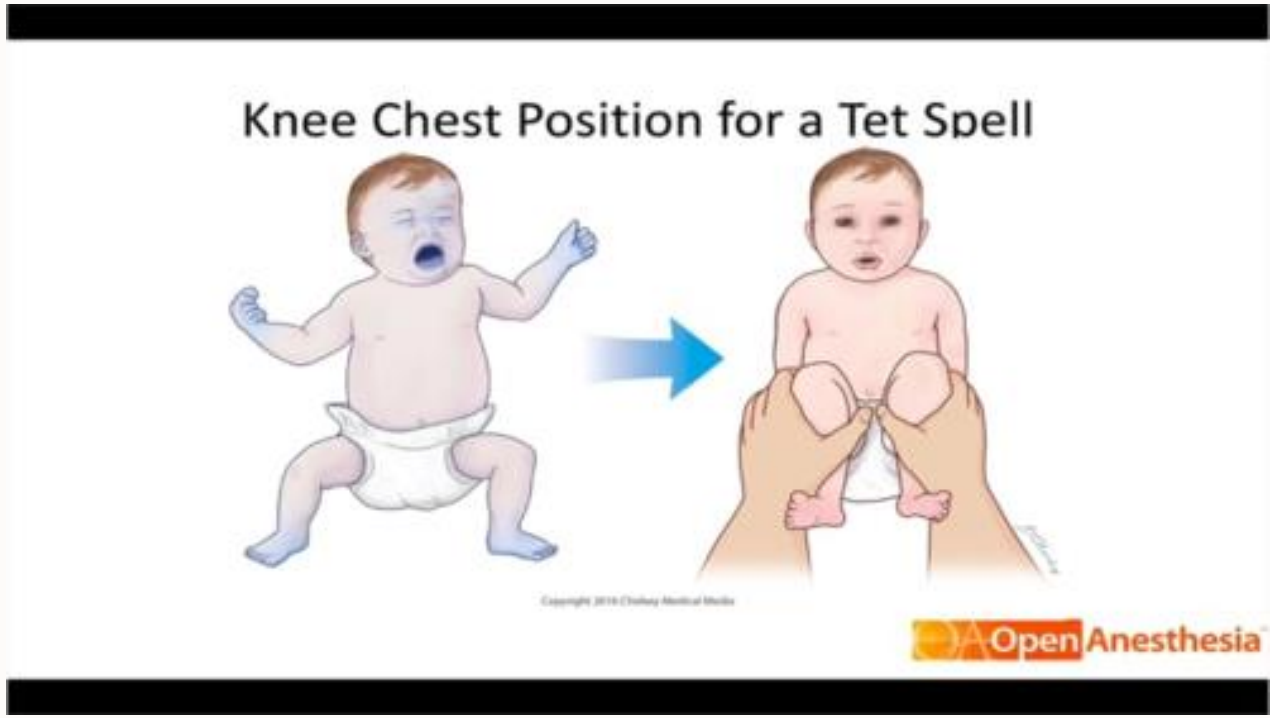
- Crying infants
- Activation of sympathetic NS
- ↑ contraction of subpulmonal heart muscle
- ↑ blood pressure in RV
- ↓ ↓ flow to the pulmonary artery
- ↑ R-L shunt
- satO2 z 85 % ↓↓ 60 % .. 40 % .. 0 %





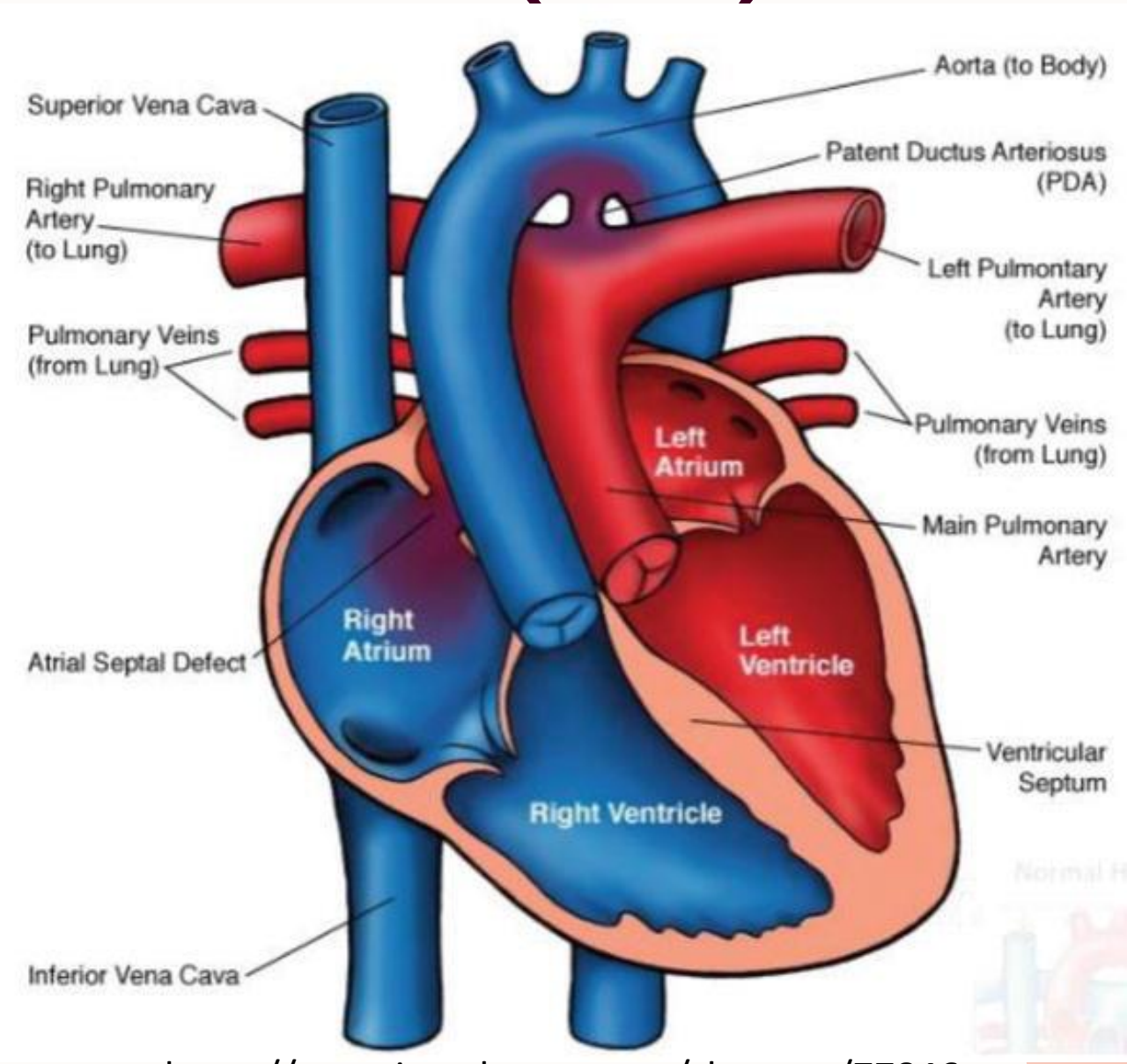
Treatment of hypoxic spell

- **!!! Restore the blood flow into PA!!!**
- Oxygen
- Calm down..sedate (Diazepam i.v.)
- Bending of the knees, squatting (increased venous return, increased syst. resistance)
- i.v. volume (albumin, 0,9% Sal. sol.)
- Ephedrine i.v.
- Norepinephrine i.v.
- Prevention – not letting babies cry; Betablockers !!!

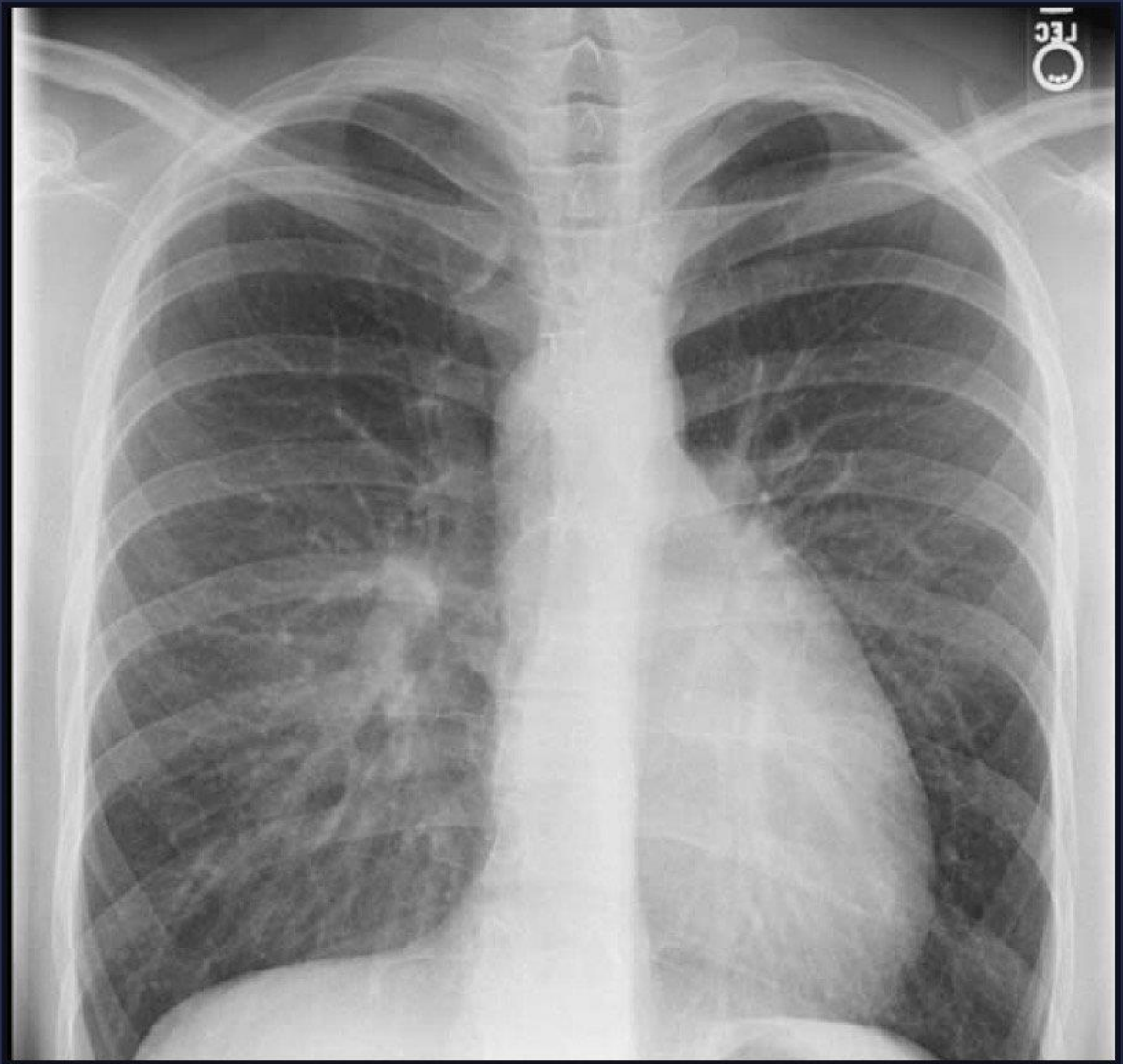


Cyanotic – critical: D-TGA (5 %)

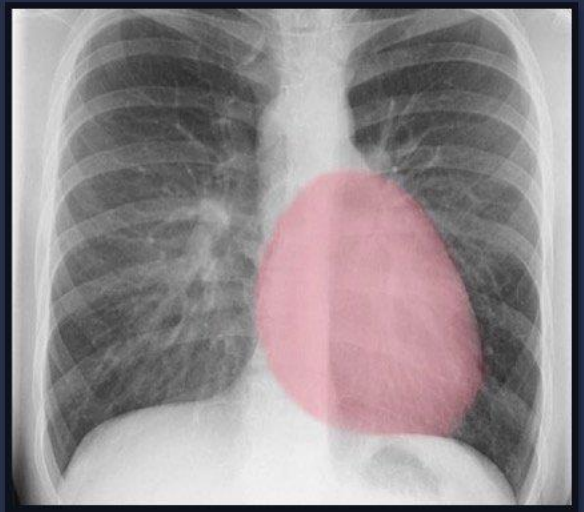
- The most common critical VCC
- 2-3x often in boys
- 50 % isolated, 50 % with other VCC
- Central cyanosis
- Sat. O2 <80 %
- 2 paralel circulations
- Mixing of the blood: FoA, PDA



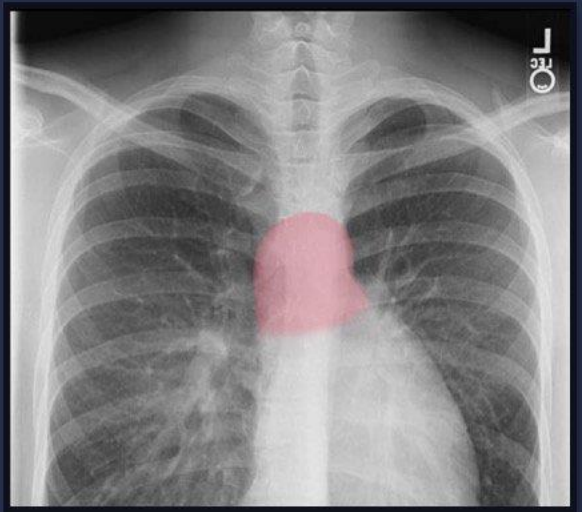
https://www.intechopen.com/chapters/77846



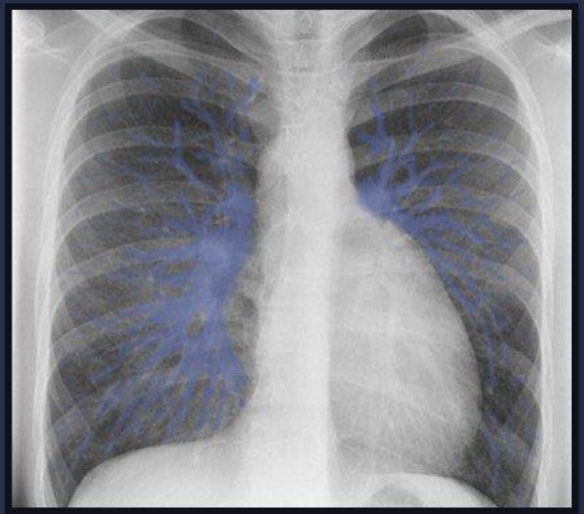
Original image by Madhero88 / CC BY-SA 3.0



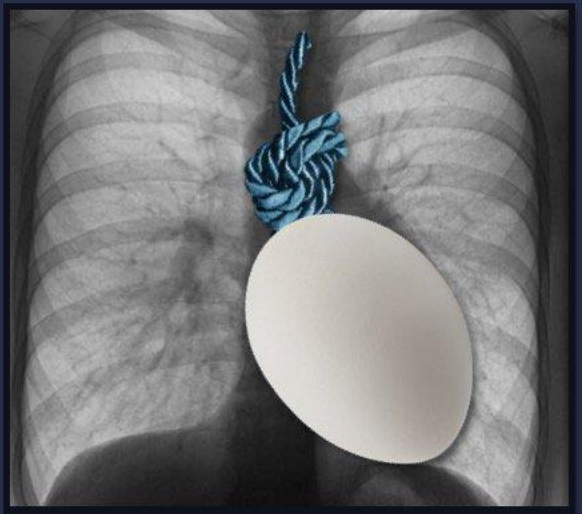
Oval Shaped Cardiac Silhouette



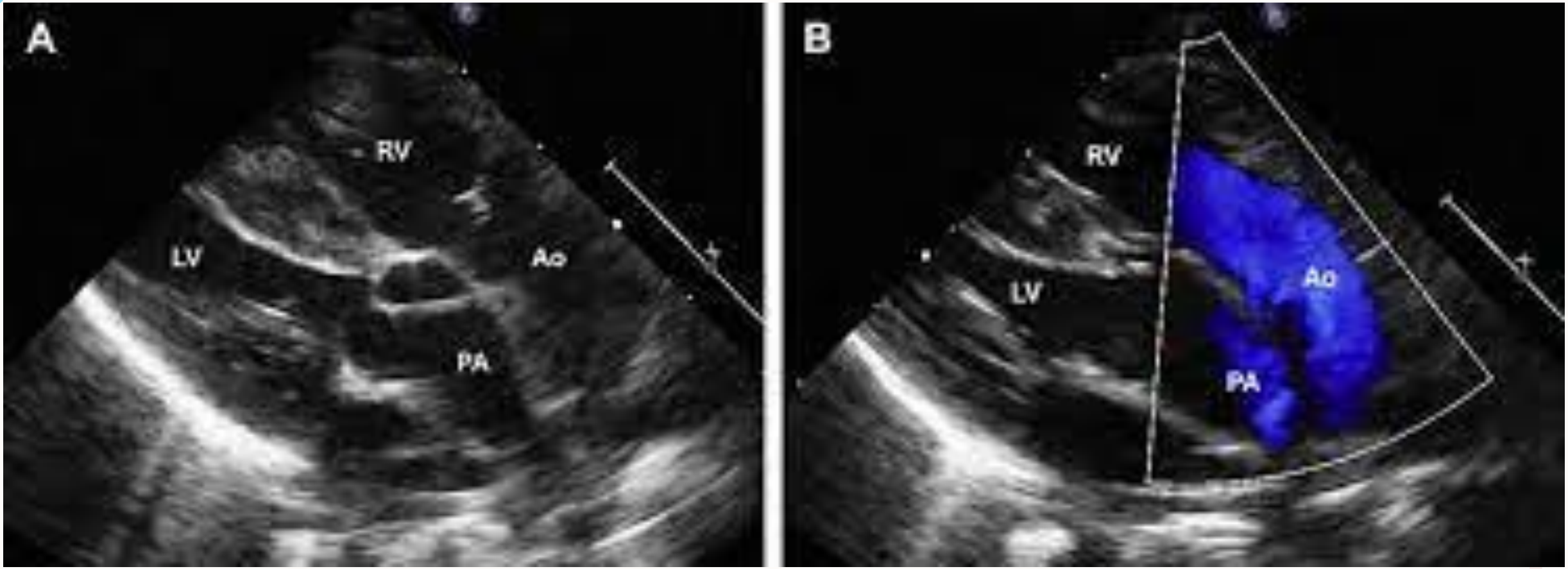
Narrow Vascular Pedicle



Increased pulmonary vasculature



Egg-on-a-string appearance



https://www.asecho.org/wp-content/uploads/2016/06/2016_Transposition-of-Great-Arteries.pdf

Therapy of TGA

- **Prostaglandin E i.v. continuously** (Alprostan-Léčiva) => keeps **PDA** open !!!
- **Side effects:** hypoventilation, apnoe, fever, diarrhea
- **Urgent baloon atrioseptostomy** in order to keep / make **FoA** bigger (if the mixing of the blood is not sufficient)
- **Anatomical (surgical) correction during the first 14 days**
- Excellent results => 10 years after cardiosurgery **survival of > 95 %** of the children

Hypertension in the childhood

Prevalence in the Czech republic

- 4 % of the children´s population
- 45 % of that obese children
- 15,4 % of obese children have arterial hypertension

Etiopathogenesis

- **Primary – essential hypertension 95 %**
- **Secondary hypertension 5 %** - predominantly up to the age of 10 years

Measurement of the blood pressure

- **A part of every preventive check-up since the age of 3 years**
- **Confirmation of hypertension** => repeated measurement at least during **2 more check-ups**, **unless it is a severe** hypertension
- **Width of the tourniquet** at least 40 % of the lenght of the arm (between the olecranon and acromion) + should cover app. 2/3 of an arm
- **After confirmation** of the hypertension the blood pressure should be **measured on each of 4 extremities**

Diagnostics – methods of measurement

- Auscultatory
- Oscillometric (automatic cuff devices)
- Doppler

Diagnostics

– ABPM (ambulatory blood pressure monitoring)

- Assessment during 24 hours long monitoring
- Excluding „white coat syndrome“
- Making the diagnosis in case of borderline hypertension
- Assessment of effectiveness of the therapy
- Problematic interpretation in toddlers and smaller children (missing charts)

Classification of the hypertension

- **Normal blood pressure < 90th percentile**
- **Elevated normal blood pressure (prehypertension) ≥ 90th - < 95th percentile or higher than 120/80 mmHg**
- **Stage 1 hypertension** (+ affected organs - hypertrophy LV, albuminuria) **95th – 99th percentile + 5 mm Hg**
- **Stage 2 hypertension** (+ bigger affection of the organs, failing of the organ function) **> 99th percentile + 5 mm Hg**

Primary hypertension

- Multifactorial
- Genetic factors (20-40 %)
- Obesity (BMI)
- Social background
- Lifestyle, nutrition
- Stress, smoking
- Unknown causes

Secondary hypertension

- Acute with sudden onset, transitory
- Chronic, longterm, persistent

Secondary hypertension

- Renal
- Renovascular
- Endocrine
- Farmacologically induced

Renal

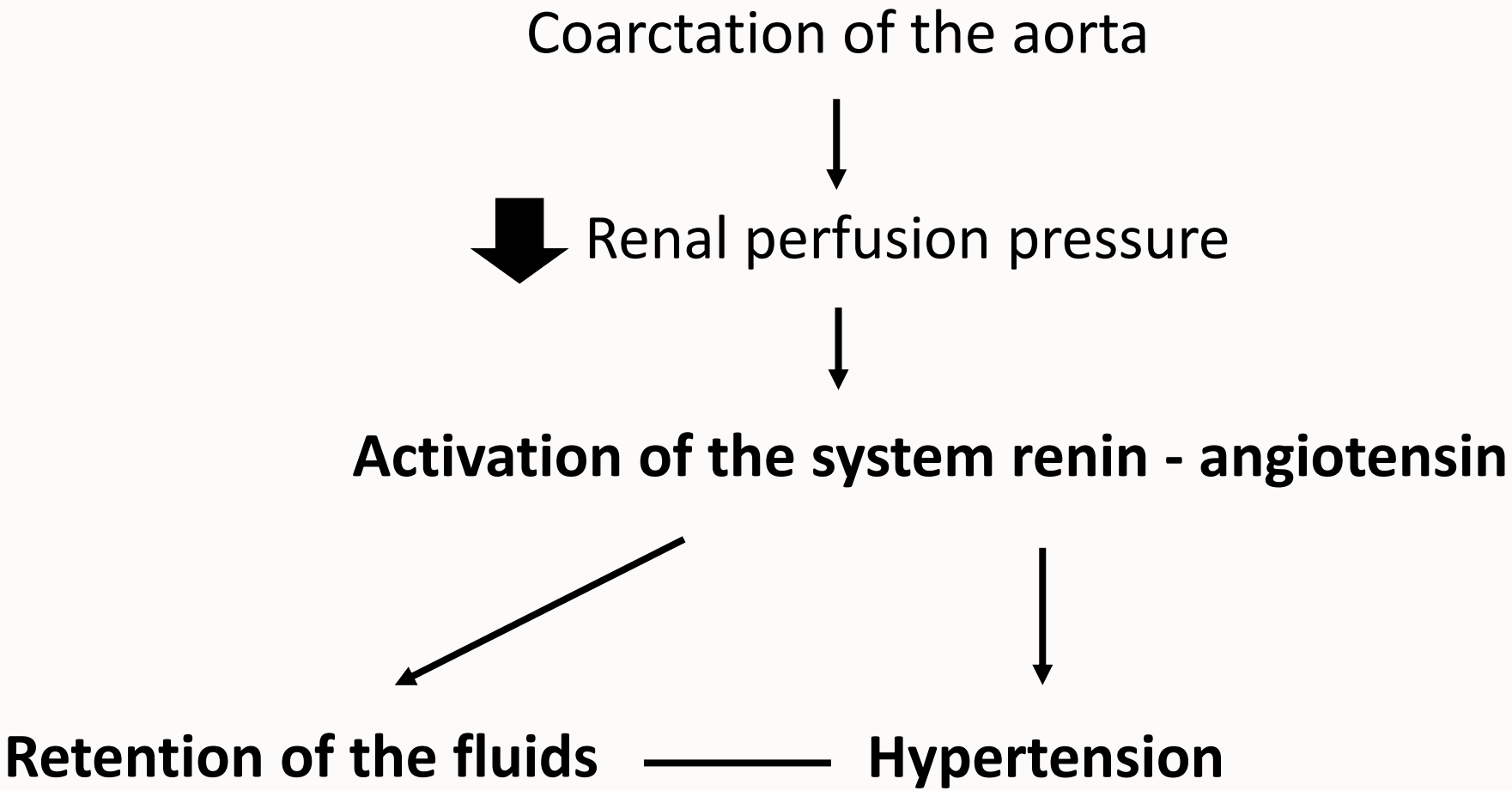
- 75-80 % cases of secondary hypertension
- In 25-50 % cases caused by pyelonephrititis
- Other possible causese:
  - Glomerulonephrititis
  - Trauma, tumors, congenital anomalies
  - Hemolytic uremic syndrome
  - Kidney surgeries and transplantation

Renovascular

- Renal artery (stenosis, fibromuscular dysplasia, thrombosis, aneurysm)
- Renal vene
- Umbilical artery
- Coarctation of the aorta
- Vasculitis
- Neurofibromatosis

Renovascular

- coarctation of the aorta => chronic, longterm, persistent



Endocrine

- Hyperthyreosis
- Hyperparathyreosis
- Congenital adrenal hyperplasia
- Cushing syndrome
- Primary hyperaldosteronism
- Feochromocytoma
- Neuroblastoma, ganglioneuroma

Farmacologically induced

- Corticosteroids and ACTH
- Contraception
- Sympatomimetics
- Stopping the antihypertensive treatment
- Amfetamin, cocain

Diagnostics

- Complete blood count
- Urea, creatinin, glykemia, Na, K, Ca, T4, TSH, plasmatic renin activity, catecholamins
- Urine chem + sed, microbiology, vanillylmandelic acid
- ECG, ECHO, ultrasound of the kidneys, Doppler of the renal arteries, angiography
- Eye (fundus) examination

Non-farmacologic treatment

- Reduction of the body weight
- Higher aerobic load
- Restriction of the salt in food
- Correction of the hyperlipidemia
- Adequate intake/supplementation (if needed) of the K, Ca, Mg

Farmacologic treatment

- If failure of lifestyle changes during 6 months or in case of moderate or severe hypertension
- Start of the treatment before the development of organic changes
- Helps to slow down the development of organic changes or to regress organic changes respectively

Farmacologic treatment

- The most commonly used medicaments in children:
  - ACE inhibitors
  - Blocators of Ca++ canals
  - ß-blockers
  - Sartans (angiotensin-II-receptor antagonists)

Thank you for your attention!