ACYANOTIC CONGENITAL HEART DISEASES

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INTRODUCTION

Incidence CHM $\approx 8 / 1000$ births

- In developed countries CHM $\approx 25\%$ of all malformations.

- Damage may occur through the influence of environmental factors during fetal development as:
  - teratogens
  - toxic agents
  - infections
  - alterations in blood flow.
INTRODUCTION

Cardiovascular abnormalities account for about 2% of heart diseases and malformations in 30% of all devices and systems.
I. Congenital heart disease with cyanosis (right-left shunt dominant)
- Tetralogy of Fallot
- Trilogy of Fallot
- Common arterial trunk
- Transposition of great arteries
- Ebstein disease
- Tricuspid atresia
- Hypoplastic left heart

II. Congenital without cyanosis or cyanosis minor (dominant right-left shunt)
- Atrial Septal Defect
- Ventricular septal defect
- Patent Ductus Arteriosus
- Atrioventricular canal
- Aorto-pulmonary window
- Anomalous pulmonary venous return

III. Cardiopaties without shunts
- Coarctation of the aorta
- Anomalies of the aortic arch

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Atrial Septal Defect

Ventricular Septal Defect

Patent Ductus Arteriosus

Coarctation of the aorta
Atrial septal defect

**Definition**

= Anatomic defect in the septum that causes communication between left atrium and right atrium.

ASD is 10% incidence of MCC.

CHM is a left right shunt, pulmonary flow increased necianogena.

**Intrauterine** "foramen ovale" - the blood of RA -> LA (lungs are shunt)

**Postpartum**: defects closes -> fibrous wall covering the oval fossa.

Lack closing ASD type = "fossa ovalis"
Atrial Septal Defect

Anatomical, depending on the location of the defect at parietal:

• ASD type foramen ovale
  - FO = persistent small flaw
  - Smaller than ASD secundum
  - Located in the same area fossa ovalis

• ASD secundum type
  - Lack septal higher
  - Situated in the middle portion of the interatrial septum
  - The most frequent, 70% of ASD
  - May emerge as a membrane multiperforata.
  - The combination of a ASD secundum type = + mitral Lutembacher syndrome.

Ostium primum atrial septal defect type •
- Parietal defect is located below,
- Associated with atrioventricular valvular defect of the product (cleft mitral valve).
Atrial Septal Defect

Anatomical, depending on the location of the defect at parietal:

• **ASD coronary sinus type**
  - Absence of interatrial wall at the site of shedding the coronary sinus vein.

• **sinus venosus atrial septal defect**
  - Superior location, at the estuary of SCV.
  - In some cases associated with pulmonary venous drainage in part, by this defect in RA.

• **unique atrium type ASD**
  - Totally missing septum
  - Appliance valve (mitral and tricuspid) normal.
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Hemodynamics

large ASD $\rightarrow$ L-R shunt

loading volume right cavities

overuse right cavities

Dilatation of the right cavities and pulmonary artery

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Atrial Septal Defect

Fiziopatologie

Parietal Defect \( \rightarrow \) \( L-R \) shunt

Blood pass LA \( \rightarrow \) RA \( \rightarrow \) charge pulmonary circulation \( \rightarrow \) HTAP

ASD large, nonoperated \( \rightarrow \) irreversible HTAP \( \rightarrow \) Eisemenger syndrome (shunt is inverse: cianosis and signs of cardiac failure)

Shunt volume depends on:
- Size of the defect,
- Compliance right ventricle
- Pulmonary vascular resistance

Inter-pulmonary stasis predispose them to infections.
Atrial Septal Defect

The clinical exam

**Asymptomatic:** discovered incidentally during medical examinations

**Symptomatic:** in the second or third decade of life.

On examination objective:
- systolic ejection grade II-III / VI in lung outbreak
- dedublation or strengthening second noise

Involving in children or young adults:
- fatigue, effort dyspnea,
- palpitations (extra beats / atrial fibrillation)
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Cardiotoracic X-ray

- It may be normal in case of small ASD

  OR

- Shadow heart is enlarged by widening RA and LA
- Pulmonary hipervascularisation

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Electrocardiogram -
- normal

OR

- signs of right ventricular hypertrophy
- incomplete right branch block

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Echocardiography
- It is the most simple and safe
- Identify atrial septal defect, location, size
- Highlights the shunt and the implications direction hemodynamic
- Exclude other associated abnormalities
Atrial Septal Defect

Cardiac catheterization

- Is not routine

- Indicated to surgical correction or interventional

- Cardiac catheterisation confirm ASD by passing the probe in LA.

- ASD with long evolution, with severe PAH is catheterization to assess reversibility or surgical contraindication.
Atrial Septal Defect

Treatment

At children with little defect and shunt surgery is not indicated and are checked periodically.
Atrial Septal Defect

Treatment

ASD closure can be made:

1. Interventional - by placing a device ("umbrella") by cardiac catheterization, which will close the defect, avoiding surgery.
2. Surgical:

- By classical method - median sternotomy or thoracotomy and using CEC

Parietal defect is closed by:
- When the defect is small - direct suture
- Suturing a pericardial patch or textile
Atrial Septal Defect

Evolution

Favorable (in most cases), ASD is well tolerated. In the presence of heart failure or cardiomegaly major surgery is required in the first years of life.

- Depending on the size -in spontaneous closure of ostium secundum defect:
  - < 3 mm - 100%;
  - 3-8 mm approx 80%;
  - >8 mm - < 50%

- Usually sinus venosus ASD, coronary sinus, ostium primun not close spontaneously.

ASD medium - large undiagnosed childhood -> symptomatic decade 3rd or 4th Life (installation PAH)
Atrial septal defect

Ventricular Septal Defect

Patent Ductus Arteriosus

Coarctation of the aorta

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Ventricular Septal Defect

Definition

= Anatomic defect in the septum that causes communication between RV and LV

The incidence = 22% of congenital heart malformations

more often localized in the membranous portion of the septum, below the aortic orifice.
Ventricular Septal Defect

Hemodinamics

VDS = communication between left – right shunt.

Small VDS : do not allow high flow => the shunt is not inverse (stay left - right). It tends to close itself off.

Large VSD :
  • RV will receive a larger amount of blood RS
  • => increase the flow RV => increase the flow PA and increase the venous return
  • The pressures of the two ventricles equalizes => shunt It becomes optional or even reverses

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Ventricular Septal Defect

By size of the shunt:

- with small shunt
- with big shunt
- accented with pulmonary hypertension
- with moderate pulmonary stenosis
Ventricular Septal Defect

Depending on the anatomical location:

**VSD perimembranous:**
- The most common, 70-80% of VSD
- Close the septal tricuspid valve leaflets

**Muscular VSD:**
- Single or multiple ("swiss cheese")
- About 15% of VSD

**Infundibular VSD:**
- = Defect supracristal or subarterial
- 5-7% of VSD
- Impairment anatomic aortic valve through their neighborhood

**Atrioventricular VSD:**
- Is located in atrioventricular area

Outlet/Supracristal/Subpulmonar/Infundibular/Juxta-arterial

6% VSD / 30% Asians
Spontaneous closure uncommon

Perimembranous, paramembranous conoventricular

80% VSD
Aneurysm formation / Aortic regurgitation

Muscular
20% VSD
Spontaneous closure common

Inlet/AV canal type
Down syndrome

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The clinical exam:

**VSD asymptomatic** when small, trivial shunt

**VSD with moderately symptomatic shunt** - high -> phenomena of heart failure in early infancy:
- hepatomegaly
- tachypnea
- tachycardia
- dyspnea
- Difficulty with fatigue and sweating sucking
- subpondere
- Frequent lung infections

- **DSV large** unrestricted neglected evolves into Eisenmenger syndrome.
Clinical examination

Small VSD:
- Systolic murmur III / 6 degre, fourth and fifth intercostal space, right parasternal, "in a steam" with posterior irradiation

Large VSD:
- Systolic murmur (pansistolic), intense, rough in the spaces left intercostal III, IV and V with wide irradiation 'in hospital Wheel "by-humeral scapular region.
- Dedublation Zg II

In large VSD systolic murmur may be less expressive.
Ventricular Septal Defect

Cardio-pulmonary X-Ray

• Small VSD
  – heart appearance is normal
  – slightly decreased pulmonary transparency

• Large VDS
  – Cardiomegaly with the middle arch convex
  – Pulmonary vascular picture accentuated.
Ventricular Septal Defect

ECG

• **Small VSD** = normal

• **Large VSD**: left ventricular hypertrophy or biventricular hypertrophy
Ventricular Septal Defect

Echocardiography

Two-dimensional and Color Doppler:
- Highlights and locates VSD
- Direction shunt
- RV and the hemodynamic implications LS
- Calculated pressure gradient interventricular

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Ventricular Septal Defect

Cardiac catheterization

- When there are discrepancies between echocardiography and clinical manifestations

- Can detect small muscular VSD

- Measuring pressures in the various cavities and blood gas saturation

- Testing the reversibility or pulmonary vascular disease

- Indicated for surgical or interventional correction
Ventricular Septal Defect

Treatment

Small VSD:
- inter-fighting infection
- prophylactically - to prevent infective endocarditis
- periodic follow

Large VSD:

Medical:
inter-fighting infection
prophylactically - to prevent infective endocarditis

Treatment of Heart Failure

Interventional / Surgical - ventricular septal defect correction

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Interventional repair

- VSD muscle, small, single or multiple
- VSD closure using special sized devices without affecting neighboring structures (tricuspid valve).
Surgery treatment

Indication for surgery depends on several factors:
- Age, defect size, location, direction and magnitude of the shunt, pulmonary vascular resistances.

Surgery
- Median approach, stenotomie using CEC approach as the defect transatrial
- Closes using pericardial patch or fabric, or continuous suture points separate the patch.

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Evolution

Small VSD: 80% may close spontaneously during the first months of life, after the age of 1 1/3.

Large VSD: congestive heart failure require surgical closure in the first year of life.

Children with large VSD and pulmonary vascular resistance Cres Eisenmenger syndrome can not be operated.

It may be complicated by:
- Infective endocarditis, recurrent lung infections
- Development of aortic insufficiency in VSD subarterial case.
Atrial septal defect

Ventricular septal defect

Patent ductus arteriosus

Coarctation of the aorta
Patent Ductus Arteriosus

Definition
= Persistent communication from the intrauterine between the pulmonary artery and the descending aorta distal to the origin of the left subclavian artery.

Incidence: 1/2000 term newborns

Prevalence among premature infants:
- 20% of premature infants
- 45% with G <1750g
- 80% G <1000g
**Patent Ductus Arteriosus**

**Pathophysiology:**

*Intrauterine* => *in the fetal circulation, CA has the role of maternal derived oxygenated blood into the systemic circulation, bypassing the lungs.*

*Blood from RA -> RV -> AP -> ductus arteriosus -> Ao downward.*

*After birth* => *functional closure under the action:*

- Elevated to O2 in the blood (during the first respiratory movements)
- Decrease in pulmonary vascular resistance (secondary pulmonary alveoli fill with air)
- Decrease in prostaglandins.
Arteriosus Ductus persistent postnatal => are S> D (A AP) Charging the volume and pressure of the pulmonary circulation.

Symptoms depend on the size PDA and pulmonary and peripheral vascular resistances.

Depending on blood flow through the channel describes:
- moderate rate
- high flow and pulmonary hypertension
- ductus arteriosus with pulmonary hypertension sea. (SDR Eisenmenger)
Clinical exam

PDA small or medium flow
- Patients are asymptomatic
- Systolic-diastolic murmur continuous covering second noise in the second intercostal space left and left subclavicular
- Pulse is extensive to all members
- Anterior fontanelle in infants often is throbbing!

PDA high rate:
- Clinical manifestations are present in the first year of life
  > congestive heart failure
  > respiratory infections
  > staturo deficit-weight
  > epistaxis.

When shunt reversal occurs, clubbing and cyanosis appears!
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X-Ray:

In small or moderate PDA:
- Normal heart

In large PDA:
- left cardiomegalie
- (Left Atrial Hypertrophy and Left Ventricular Hypertrophy)
- hyper-vascularized pulmonary aspects
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ECG

- Normal in case of failure or low volume environment

- Signs of overload of the heart left (Left Atrial Ventricular hypertrophy and Left ventricular hypertrophy)

- Right heart overload (severe PAH)
Echocardiography

- Highly accurate diagnosis
- Echocardiography is sufficient to establish the diagnosis
- Doppler: identify blood flow to the duct
**Patent Ductus Arteriosus**

**Management:**
- Fan support
- Fluid restriction

**PDA closure:**
- Medicine: prostaglandin synthesis inhibitors -> Indomethacin / Ibuprofen
- Non-surgical: pecutana closing the PCA (catheterization) = method of choice
- Surgical: thoracotomy at left intercostal space IV sectioning favorite duct ligation than to (safer and higher close rate)
**Patent Ductus Arteriosus**

Ducto-dependent CHM

PDA can be associated with CHM savior => = compensatory role maintaining open solution before surgery!

Infants with systemic blood flow ducto-dependent, may be in shock when the ductus closes → infusion of PG E1 can be life saving.

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Patent Ductus Arteriosus
Ducto-dependent CHM

Presentation:
Alprostadil "pint" 500 mg / ml; 1 ml ampoule;

Prostaglandin E1 dose of 0.05-0.1mcg / kg / min
- Vials of 1 ml to 500 mcg of active substance
- Is given intravenously, large caliber vein / umbilical using infusion pump

Patients diagnosed late will require high doses of prostaglandin E1 (0.15-0.20mcg / kg / min)

Adverse effects:
- Respiratory depression / apnea
- Hypocalcemia, hypoglycemia
- Hypotension, bradycardia or tachycardia
- Convulsions, cardiac arrest, sepsis, CID.
Patent Ductus Arteriosus

- **Evolution**
  - New Born - spontaneous closure occurs within the first 3 months of life
  - 50% are closed in the first 24 hours, 90% in the first 48 hours of life

- **Premature:**
  - spontaneous closure may last 1 year.
  - SDR Eisenmenger = final stage of PDA
  - Untreated => irreversible pulmonary vascular disease
Atrial septal defect
Ventricular septal defect
Patent Ductus Arteriosus
Coarctation of the aorta
Coarctation of the aorta

**Definition**
Congenital heart malformation characterized by narrowing and obstruction of the aortic lumen segment determined by intimal thickening media and tissue.

**Location frequency** junction between distal aortic arch and descending aorta just below the origin of the left subclavian artery.

**Aortic coarctation isthmic** described as a spur produced by media and intima thickening of the aortic lumen protrudes from the rear and side walls of the aorta.

It represents 5-8% of all congenital heart malformations.

**Sex ratio male: female = 2/1**
Coarctation of the aorta

Anatomic lesion

The defect can be:
- **Preductal** (infantile form), or **postductal** (big child form)
- isolated or associated with other abnormalities (especially of the heart left)

Lumen of the aorta -> diameter <0.5 cm = **severe coarctation**

Poststenotic may occur dilations

In time => collateral circulation (left subclavian artery branches, intercostal arteries 3 and 4)
Coarctation of the aorta

Hemodynamics:

On both sides of coarctation area:
- Hypertension Preductal (cephalic extremity, upper limbs and pulmonary bed)
- Hypotension downstream (postductal).

In response to afterload => Left Ventricular Hipertrophy concentric.

Proper irrigation lower half of the body is done by:
- Increased systolic blood pressure over the area of coarctation
- Arteriolar vasoconstriction systemic
- Development of collateral circulation
Coarctation of the aorta

Tablou clinic

Severe forms of the disease _> 7-10 days of life:
Congestive heart failure brutal installed

- tachypnea
- cyanosis (especially lower limbs)
- lower extremities cold
  = weak pulse perceptible / absent femoral artery and Pedi
- acute renal failure (oliguria, anuria)
- shock
- oxygen saturation difference between the lowest and highest limbs.
Coarctation of the aorta

Tabloul clinic

La copilul scolar este evidentă circulația colaterală!

In most cases -> asymptomatic.

In older children or young adults: hypertension in the upper limbs, headache, epistaxis, dizziness.

Classical:
- development athletic top half of the body that contrasts with the development of the lower half of the body gracila
- pulse may well be knocked on the radial arteries and diminished / absent from the femoral arteries
- Hypertension

School children is evident collateral circulation!

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Coarctation of the aorta

Measuring blood pressure in 4 limbs:

- Increased at above the age-appropriate upper limbs
- Low levels of the lower limbs (over 10-15mmHg difference in blood pressure between the upper and lower limbs)

Crackles lung stasis hepatomegaly Systolic murmur II-III / 6 subclavicular left with irradiation Interscapular

The diagnosis is evoked:
- Blood pressure difference measured at the upper and lower limbs,
- Weak pulse in the femoral artery, in contrast to the radial pulse and carotid arteries mattress.
Coarctation of the aorta

Electrocardiogram:
- Left ventricular hypertrophy
- It can be normal
Coarctation of the aorta

Thoraco-pulmonary radiography:

Cord-size

LV hypertrophy variable

Poststenotica pre- and expansion of the aorta - the picture can be seen the number "3" on the upper left edge of the cardiac silhouette.
Coarctation of the aorta

Echocardiography

- The incidence suprasternal: coarctation area and the descending aorta coarctation post.
- Eco Doppler maximum flow rate will be measured at the aortic isthmus and appreciation pressure gradient
Coarctation of the aorta

Nuclear magnetic resonance

- is the gold standard in evaluating aortic coarctation

- noninvasive method

- determine the exact location and anatomy coarcatiei, the aorta and the collateral circulation.
Coarctation of the aorta

**Treatment**

**Medical**
- In infants with severe stenosis, congestive heart failure, insufficient kidney, metabolic acidosis.

**Urgent measures for resuscitation:**
- Intubation
- Prostaglandin infusion to maintain permeable ductus arteriosus
- Surgical or interventional (balloon dilatation).

**Blood pressure control**

**Interventional - Balloon Angioplasty**
- Non-invasive alternative to surgery
- The preferred method in case of recoarctation

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Coarctation of the aorta

Treatment

Surgical

It is appropriate at any age so urgently to save the lives of newborn and electively to prevent complications CoAo.

Surgical techniques:

- Resection technique end-to-end
- A flap technique. Left subclavian
- The technique of resection and interposition prosthesis

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Coarctation of the aorta

Natural evolution

- In severe forms or associated with other defects can present since birth congestive heart failure, severe acidosis, distal hypoperfusion -> medical and surgical treatment immediately.

- Asymptomatic forms can be detected in adulthood (elevated blood pressures, intense headache, epistaxis) or repeated hemorrhagic strokes

- CoAo untreated can reach age 35, 20% and even up to 50 years.

- Corrective surgery before the age of 14 -> 90% survival at 20 years