SMLE essential topic!

Pediatrics TeamWork

437

Congenital Heart Diseases

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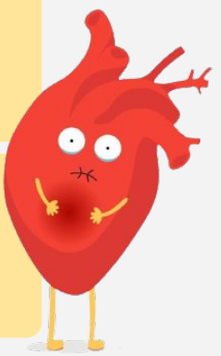
Notes Previous Notes Book Important!

Overview

Epidemiology

Congenital heart disease (CHD) ~ 1% of live births:

Ventricular septal defect (VSD)	30% Most common	Pulmonary valve stenosis (PS)	7%
Patent ductus arteriosus (PDA)	12%	Coarctation of aorta (CoA)	5%
Atrial septal defect (ASD)	7%	Tetralogy of fallot (TOF)	5%
Aortic valve stenosis (AS)	5%	Atrioventricular septal defect (AVSD)	2%
D-transposition of great arteries (D-TGA)	5%		



Etiology

Etiology: Mostly unknown

- **Chromosomal abnormalities (10%)**
- **Trisomy 21:** AVSD
- **Trisomy 18:** complex
- **Trisomy 13:** complex
- **DiGeorge Syndrome:** interrupted aortic arch , conotruncal abnormalities
- **Turner syndrome:** Coarctation of Aorta, Bicuspid aortic valve
- **Williams Syndrome:** Supra-aortic stenosis, Pulmonary branches stenosis
- **Noonan Syndrome:** Dysplastic pulmonary valve stenosis, hypertrophic cardiomyopathy

	Cardiac abnormalities	Frequency
Maternal disorders		
Rubella infection	Peripheral pulmonary stenosis, PDA	30%–35%
Systemic lupus erythematosus	Complete heart block (anti-Ro and anti-La antibody)	35%
Diabetes mellitus	Incidence increased overall	2%
Maternal drugs		
Warfarin therapy	Pulmonary valve stenosis, PDA	5%
Fetal alcohol syndrome	ASD, VSD, tetralogy of Fallot	25%
Chromosomal abnormality		
Down syndrome (trisomy 21)	Atrioventricular septal defect, VSD	30%
Edwards syndrome (trisomy 18)	Complex	60%–80%
Patau syndrome (trisomy 13)	Complex	70%
Turner syndrome (45XO)	Aortic valve stenosis, coarctation of the aorta	15%
Chromosome 22q11.2 deletion	Aortic arch anomalies, tetralogy of Fallot, common arterial trunk	80%
Williams syndrome (7q11.23 microdeletion)	Supravalvular aortic stenosis, peripheral pulmonary artery stenosis	85%
Noonan syndrome (PTPN11 mutation and others)	Hypertrophic cardiomyopathy, atrial septal defect, pulmonary valve stenosis	50%
Duchenne muscular dystrophy	Cardiomyopathy	78% by age 20

ASD, atrial septal defect; PDA, persistent ductus arteriosus; VSD, ventricular septal defect.

From the book!

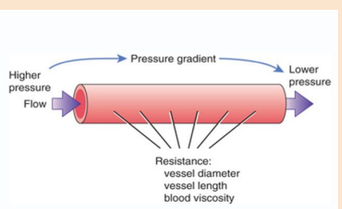
Overview

Key concepts

Blood flow	**Resistance**	Blood flow easier to low resistant organs	Pulmonary vascular resistance Systemic vascular resistance
	Pressure Gradient	Blood flow from high pressure to low pressure chambers systemic is high pressure pulmonary is low pressure	
	Flow Obstruction	Blood flow away from site of obstruction if alternative path exist	
Duct Dependent CHD	Congenital heart diseases that need patency of ductus arteriosus for survival	Pulmonary duct dependent circulation	Examples: critical pulmonary stenosis or pulmonary atresia
		Systemic duct dependent circulation	Example: critical aortic valve stenosis, critical coarctation of aorta, Interrupted aortic arch, HLHS

1.CVS Flow and Resistance

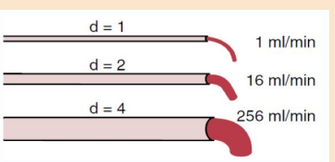
Flow



$$F = \frac{\Delta P}{R}$$

F = Flow
ΔP = Pressure Gradient
R = Resistance

Resistance



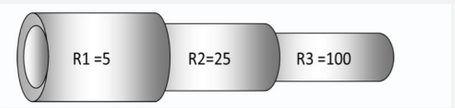
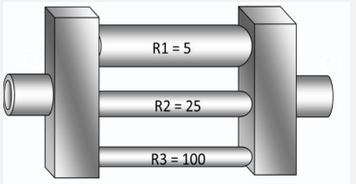
$$R \ \alpha \ \frac{\eta \times l}{r^4}$$

R: Resistance
η : Viscosity
L: Length
r: Radius

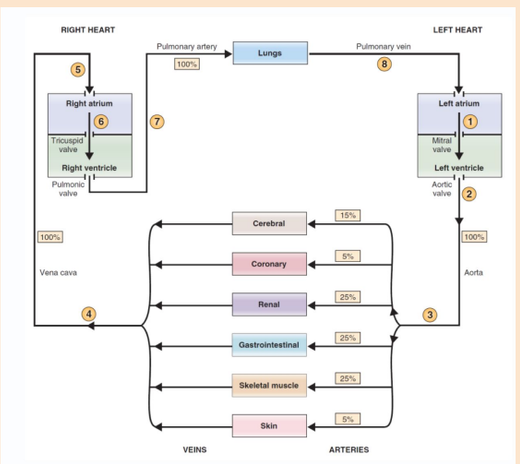
little change in diameter cause significant change in flow by power of 4

- we advise the mother to not take ibuprofen or NSAID in late pregnancy because it can close ducts arterioles in utrolife and end up with severe right side heart failure .
- prostaglandin is IV meds that keep ductus open

1.CVS Flow and Resistance

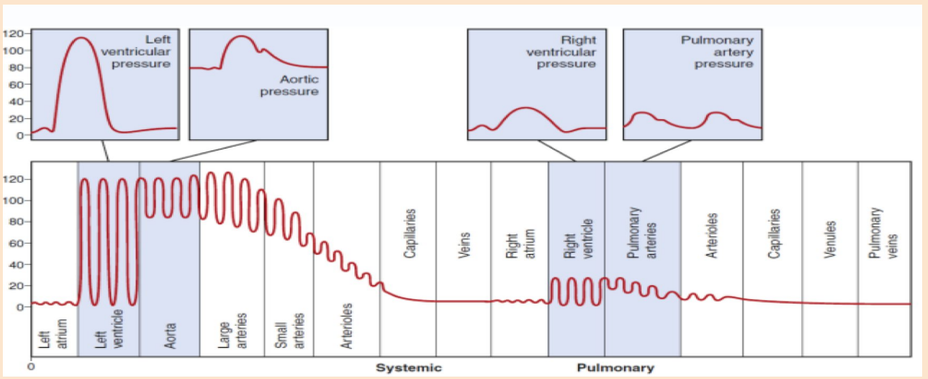
Serial Circuit	Parallel Circuit
	
R total = R1 + R2 + R3 R total = 5 + 25 + 100 R total = 130	1/R total = 1/R1 + 1/R2 + 1/R3 = 1/5 + 1/25 + 1/100 = 20/100 +4/100+1/100 = 25/100 = 1/4 R total = 4 - if you add more R the R total will be low - الناتج هنا بيكون اقل من اقل قيمة مقاومة موجودة في الدائرة

2.CVS Circulation



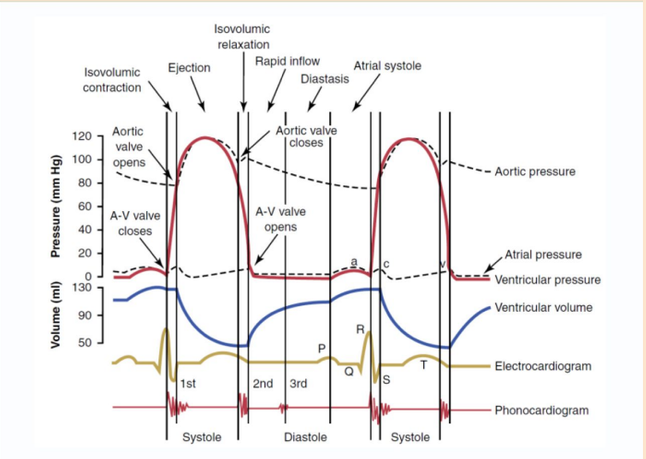
- any obstruction will cause increase in the pressure before obstruction.
- if the patient have pump problem the blood will accumulate in LV and gos backward to the LA than to lungs and it will cause right said heart failure .
- the resistance difference between the organs it determined according to its function and needs.

3.CVS driving pressure



- LV has capacity to go from (low pressure to accommodate LA flow) to (high pressure to overcome aortic pressure).

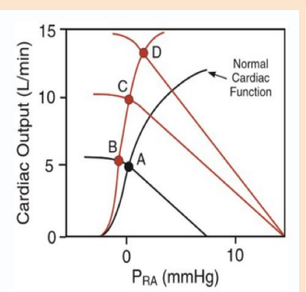
4.Cardiac Cycle

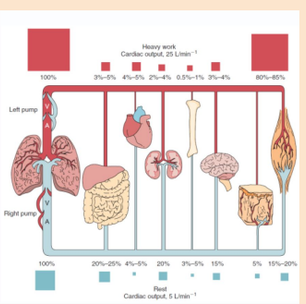


- **1st :** closer of mitral and tricuspid valve
- **2nd :** close of aortic valve
- **3rd:** raped filling of LV in the early diastole
- **4th:** atrial contraction
- 3rd + 4th are called galob rep and they are sign of heart failure.

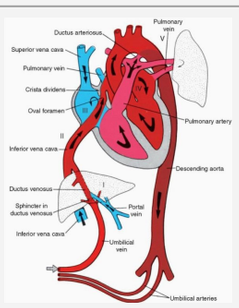
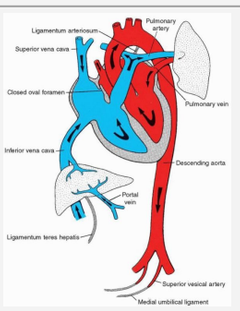
5.Cardiac Systemic Output

- Cardiac and Venous Function Curve
- Cardiac Output Increase during Exercise due to:
  - Increase contractility
  - Increase preload
  - Reduce afterload





6.Fetal vs Postnatal Circulation

FETAL CIRCULATION	POSTNATAL CIRCULATION
	
- foramen ovale is essential for life in fetus which is allow blood go from RA to LA directly . because RA in fetal life is higher pressure than postnatal and LA in fetal life is lower than postnatal - pulmonary resistance is high in fetal life so 10% of the blood will go to the pulmonary artery and 90% will go to systemic circulation through ductus arterioles	- once the baby cry after delivery the alveoli will open and the lung resistance will drop and by clamping the umbilical cord the systemic circulation resistant will go up so the blood in the RV will favor going to lungs. - After removing placenta the venous return to RA will depend on SVC and IVC so the RA pressure will go down.

CVS Changes after birth

- Closure of ductus arteriosus . placenta secret prostaglandin to keep it open after removing placenta and exposure to oxygen it will go on physiological constriction in 24h and anatomical closer in 2 weeks
- Closure of ductus venosus
- Closure of Foramen Ovale . because of the change in pressure that happened postnatally in RA and LA
- Increase of systemic vascular resistance
- Decrease of pulmonary vascular resistance

CHD

Classification of CHD

Cyanotic Heart Disease	Acyanotic Heart Disease
Decreased pulmonary flow: – Tetralogy of Fallot – Tricuspid atresia – Other univentricular heart with pulmonary stenosis	**Left – Right shunt lesions:** – Ventricular septal defect – Atrial Septal Defect – Atrioventricular Septal Defect – Patent Ductus Arteriosus
Increased pulmonary flow: – Transposition of great arteries – Total anomalous pulmonary venous return	**Obstructive lesions:** – Aortic stenosis – Pulmonary valve stenosis – Coarctation of Aorta

"Critical" Obstructive lesions: Present with "cyanosis"

Acyanotic Heart Diseases

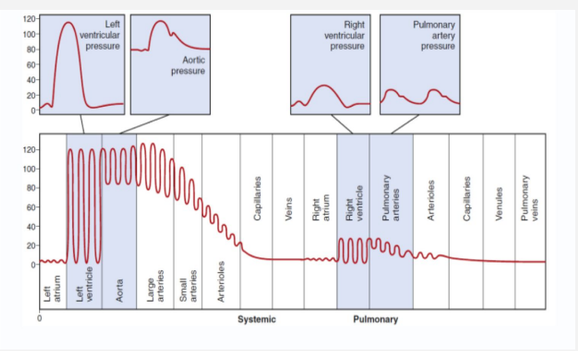
Those also known as left to right shunt lesions , and they include:

VSD | ASD | PDA | AVSD

Pathophysiology: L-R shunt

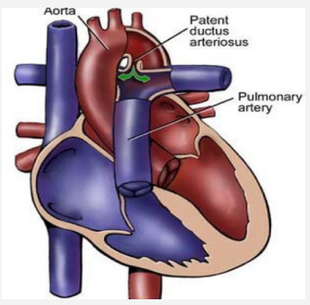
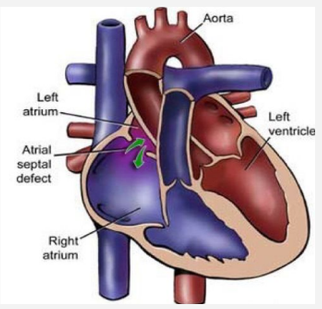
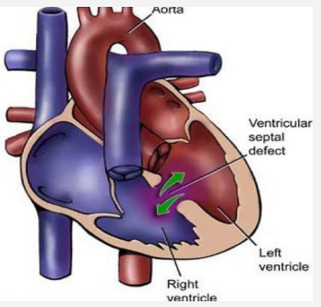
- L-R shunt toward pulmonary circulation.
- Increase cardiac output to the pulmonary circulation (Qp) CHF
- Reduce cardiac output to the systemic circulation (Qs)
- Increase Qp:Qs ratio normally it is 1:1 ratio

CVS driving pressure:



- LV pressure is higher than RV during systole but during diastole is almost the same
- aortic pressure is higher than pulmonary during both systole and diastole

Acyanotic Heart Diseases

Disease	PDA	ASD	VSD
			
PATHOPHYSIOLOGY	- L-R shunt at artery level - Dilated LA and LV - Enlarged pulmonary arteries	- L-R shunt at atrial level - Dilated RA and RV - Enlarged pulmonary arteries	- L-R shunt at ventricular level - Dilated LA and LV - Enlarged pulmonary arteries
SYMPTOM	- Small PDA: Asymptomatic - Moderate to large PDA: CHF	- Usually asymptomatic - Older children: activity related SOB or arrhythmia Rare: CHF	- Small VSD: Asymptomatic - Moderate to large VSD: CHF
EXAMINATION	- Small PDA: Silent - Large PDA: Continuous "machinery" murmur - Large PDA: Widened pulse pressure	- Fixed widely split second heart sound - Ejection systolic murmur	- Holosystolic murmur - Small muscular: ejection systolic murmur
INTERVENTION	- Mostly CATH closure - Surgery for premature and symptomatic babies	- Usually CATH closure around 3-6 years - Some types need surgical closure	-Usually surgical closure: 4-8 months - Older children when suitable: CATH closure
MEDICAL	**Anticongestive therapy:** - Diuretics often combined with captopril - Afterload reducing agents e.g. nitroglycerin - Nutritional support - ASD: usually no medication		

Complications if not treated:

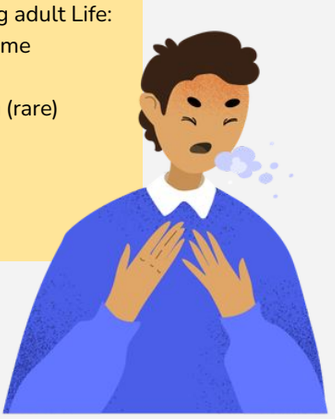
Untreated VSD and PDA beyond infancy

- Eisenmenger's syndrome irreversible and the only treatment is heart lung transplant
- Sign and symptom of CHF will disappear it could be VSD get smaller or it can be Eisenmenger's syndrome we can differentiate between them by cyanosis
- Patient will become cyanotic (R-L shunt)

Untreated ASD

Complications happened during adult Life:

- Eisenmenger's syndrome
- Atrial arrhythmias
- Paradoxical embolism (rare)



Next slide to talk briefly about AVSD

Acyanotic Heart Diseases

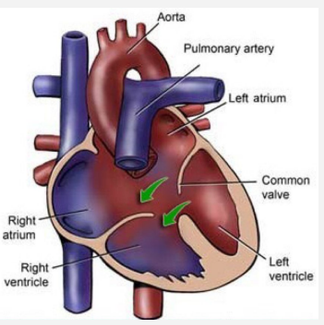
AVSD: Overview

- Incidence: 4 % of all CHD
  - Down Syndrome (50%)
- Divided into:
  - Complete AVSD
  - Partial AVSD
  - Transitional AVSD
- Balanced vs. Unbalanced

Down Syndrome

Pathophysiology:

- Similar to VSD and ASD
  - left to right shunt across the atrial level
  - Left to right shunt at and ventricular level
  - In addition: AV valve regurgitation
- Significant L-R shunting:
  - Pulmonary overcirculation
  - Increase Qp:Qs ratio.

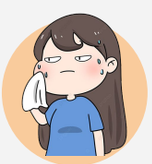


Clinical Features

Usually asymptomatic at neonatal period;

- Due to high pulmonary vascular resistance
- Baby may have slightly lower oxygen saturation

Symptoms of CHF started at few months of age:

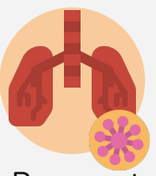


Diaphoresis

Poor feeding

Failure to thrive

Shortness of breath



Recurrent chest infection

Exercise intolerance

Physical examination:

- Feature of Down Syndrome
- Tachypnea
- Tachycardia
- Active precordium (watch this video to see this finding)
- Murmur: Pansystolic murmur
- Hepatomegaly

Acyanotic Heart Diseases

Diagnosis (AVSD)

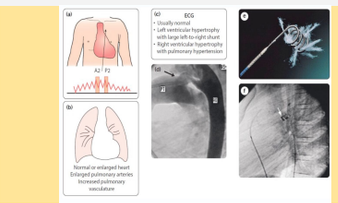
- Chest X-ray:
  - Increased pulmonary vascular marking
  - Cardiomegaly
- ECG:
  - Left Axis deviation with RVH is very suggestive of AVSD
- ECHO:
  - Confirm diagnosis
- Cardiac Cath:
  - Not required for diagnosis unless more detailed
  - hemodynamic assessment is needed

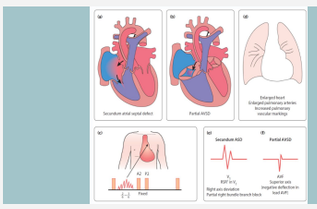
Treatment:

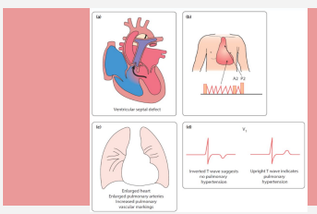
- Medical Rx:
  - Anticongestive therapy
  - Nutritional support
- Surgical closure for complete VSD:
  - Usually done before 6 months of age to avoid development of Eisenmenger's syndrome
    - Balanced AVSD: Biventricular repair
    - Unbalanced AVSD: Single ventricular repair

Acyanotic Heart Diseases

Investigations (L-R shunt)







Chest X-ray:

- Increased pulmonary vascular marking
- Cardiomegaly

ECG:

- Small lesion: Normal
- Moderate to large: chambers enlargement

ECHO:

- Confirm diagnosis

Cardiac Cath:

- Not required for diagnosis
- Might needed for more detailed hemodynamic assessment or
- when CATH intervention is planned

Congestive Heart Failure

SYMPTOMS	SIGNS
• Diaphoresis • Poor feeding • Failure to thrive • Shortness of breath • Recurrent chest infection • Exercise intolerance	• Tachycardia • Tachypnea • Cardiomegaly • Hepatomegaly • Active precordium

VSD

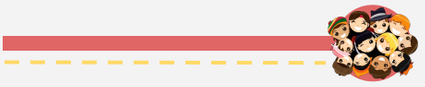
- No symptom during neonatal period
- due to high pulmonary vascular resistance

Symptoms of CHF started ~ 2/12 of age

– diaphoresis, poor feeding, and failure to thrive
– shortness of breath, recurrent chest infection
– exercise intolerance

CHF because of MI the lungs get congested from venous side however, in VSD lungs get congested from arterial side

Obstructive Lesions



Types

Aortic stenosis | Pulmonary valve stenosis | Coarctation of aorta

1.Coarctation of Aorta (CoA):

- Incidence: 5-7 % of all CHD
- Associated with Turner syndrome in female
- Arch interruption: seen in DiGeorge syndrome
- Can be: Discrete or Diffuse
- Can be mild to severe

Clinical features

➢ Physical Examination:
  - Differential cyanosis (severe CoA in newborn)
  - Signs of cardiac shock, severe metabolic acidosis
  - Reduced or absent femoral pulses
  - BP in lower limb lower than upper Limb BP
  - Radio- femoral delay

➢ Murmur:
  - Ejection systolic murmur at the back
  - Continuous murmur “due to collateral” at the back

Severity	Critical CoA	Mild CoA
Pathophysiology	• Spontaneous PDA closure - Lower body hypoperfusion - Hypotension - Acidosis - LV dysfunction - Cardiogenic Shock •“DUCT DEPENDENT CHD”	• Collateral vessels develop overtime • Flow maintained between proximal and distal aorta • Present later on life
Clinical Presentations	**Presented 2-3 weeks of life:** • Sign of CHF • Circulatory collapse • Shock • Death • “DUCT DEPENDENT CHD”	**Present later on life:** - Murmur - Chronic hypertension - Headache - Headache - Fatigue - Stroke » Rupture cerebral aneurysm

- One of signs of CoA in adults is radiofemoral delay however in pedia we see week flow
- Normally BP is higher in lower limb, if we have a patient with BP higher in the upper limb we should think of CoA.
- In CVS examination we should do 4 limb BP.

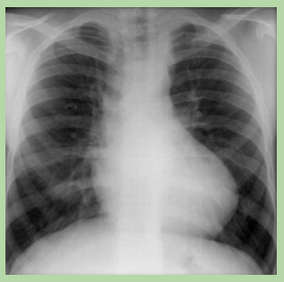
Obstructive Lesions

1.Coarctation of Aorta (CoA):

Diagnosis

Chest X-ray

- Cardiomegaly
- Prominent aortic knob
- Rib notching: Due to **intercostal collateral vessels**. Rarely seen before age of 10 years



ECG

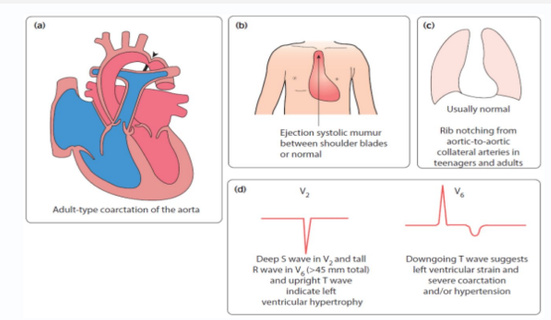
- Neonate: RV hypertrophy
- Older children: LV hypertrophy

ECHO

- Establish the diagnosis

Cardiac CT /MRI

- Might be needed to delineate the
- Arch anatomy



Treatment

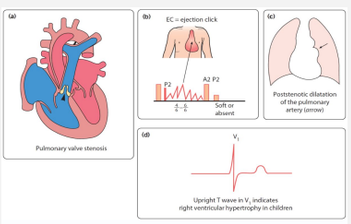
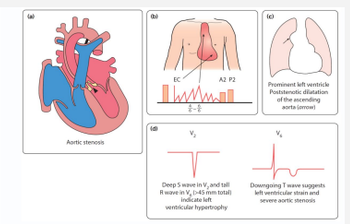
1.Critical CoA
- Duct Dependent CHD"
  - Prostaglandin E2 to keep PDA open

2.Surgery is the primary intervention

3.Trans-catheter balloon angioplasty +/- stent:
- Recurrent CoA
- Primary intervention: Discrete CoA in older children

Surgery is better than cath, BUT it has high risk of bleeding leading to death if we injured a major collateral vessel so we prefer cath more

2.Pulmonary and Aortic valves stenosis:

Pulmonary Valve Stenosis	Aortic Valve Stenosis
	
- Mild: no intervention - Moderat: - Sever : usually we baloon it and you may need surgery	- Usually we balloon it but it need replacement later

Cyanotic Heart Diseases

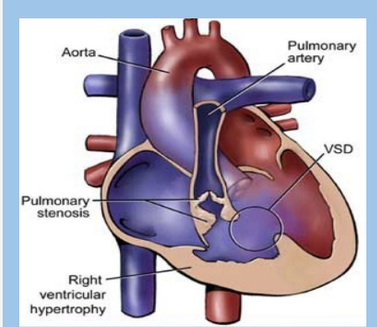
Types

Transposition of great arteries	Tricuspid atresia	Total anomalous pulmonary venous return	Truncus arteriosus
Tetralogy of fallot	Pulmonary atresia	Hypoplastic left heart syndrome	Ebstein's anomalies

-Transposition of GA is the most common cause of severe cyanosis after birth
-TOF is the most common CHD

Tetralogy of Fallot

- Most common cyanotic CHD
  - Incidence: 6 % of all CHD
- Can be associated with
  - DiGeorge Syndrome
  - Alagille syndrome
- Four basic components
  - Large VSD
  - Pulmonary stenosis
  - Overriding aorta
  - RV hypertrophy



الدكتور قال لنا نحط اليد اليمين على اليسار زي المومياء المصرية نسوي علامة اكس -

- Right hand = pulmonary and best heard in the left
- Left hand= aorta and best heard in the right
- 1st heart sound best heard in mitral area
- 2nd heart sound best heard in pulmonary area. **why?** Because in the aortic area i will hear aorta only but in pulmonary i will hear both aorta and pulmonary which is the 2nd heart sound
- In tetralogy the pulmonary will be small and aorta will dilate and override the pulmonary result in single 2nd heart sound

Clinical features:

Determined by severity of pulmonary stenosis:

- Most newborns are
  - Asymptomatic
  - Ejection systolic murmur
  - Initially: mild cyanosis which progresses over time
  - Might present with hypercyanotic spells “tet spell” if intervention delayed
- Newborn with critical PS or pulmonary atresia
  - Severe cyanosis when PDA close
  - “Duct dependent CHD”
  - Needs “Prostaglandin E2”

Cyanotic Heart Diseases

Tetralogy of Fallot

Clinical features of TET spell:

- Usually occur around 9-12 months of age
- Episodes of acute and severe cyanosis
- Might require emergency surgical intervention

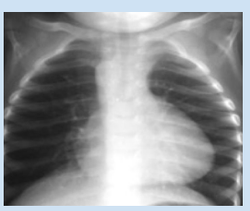
Management	Effect
Reduced anxiety	Reduce PVR
Oxygen	Reduce PVR
Sedation with morphine	Reduce PVR
Knee-to-chest position	Increase SVR
Phenylephrine	Increase SVR
IV fluid	Increase cardiac filling
Beta Blocker	Reduced heart rate

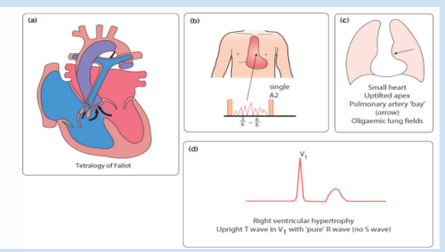
-**PVR:** pulmonary vascular resistance
-**SVR:** systemic vascular resistance
-Rarely we see CHD patient with CHF and cyanosis because they are physiology different :
1.CHF is caused by pulmonary congestion
2.Cyanosis caused by pulmonary reduced flow
-We can see patient with CHF and cyanosis in **Truncus Arteriosus**

Investigations:

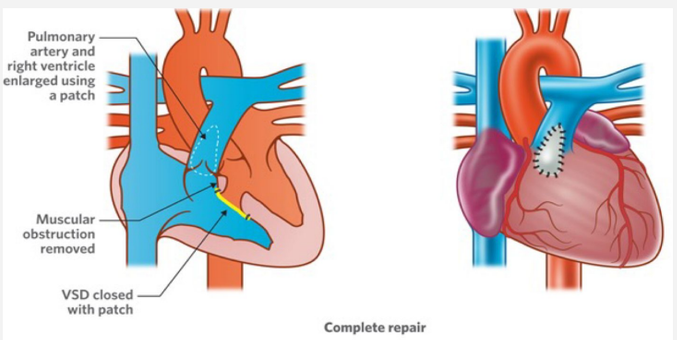
CHEST X-RAY:
- “boot-shaped heart”
- Oligemic lungs

ECG: RVH
ECHO: confirm diagnosis
CT/MRI: might be needed to delineate PA branches


Boot shaped heart



Treatment:

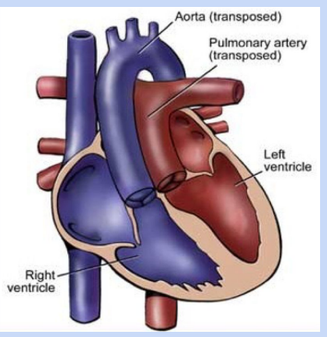


According to degree of pulmonary valve stenosis will determine the time of surgery. Some of the patient will need surgery after delivery direct and some patient we can do surgery at 12 months of age (at around 6 months, from book)

Cyanotic Heart Diseases

Transposition of Great Arteries

- Incidence: 4 % of all CHD
- Most common CHD presented with severe cyanosis at birth
- More common in male
- Higher incidence in infant of diabetic mother



Pathophysiology:

- ➢ In Normal heart:
  - Pulmonary and systemic circulations are in series
- ➢ In D-TGA:
  - Pulmonary and systemic circulations are in parallel
  - Deoxygenated blood circulates from and to the systemic circulation without proper mixing with oxygenated blood in the pulmonary circulation
  -
- ➢ Mixing of oxygenated and deoxygenated blood can occur at three levels:
  - Atrial level via ASD/PFO (most efficient) initial management until we do surgery
  - Great arteries level via PDA
  - Ventricular level via VSD (if present)

Clinical presentations:

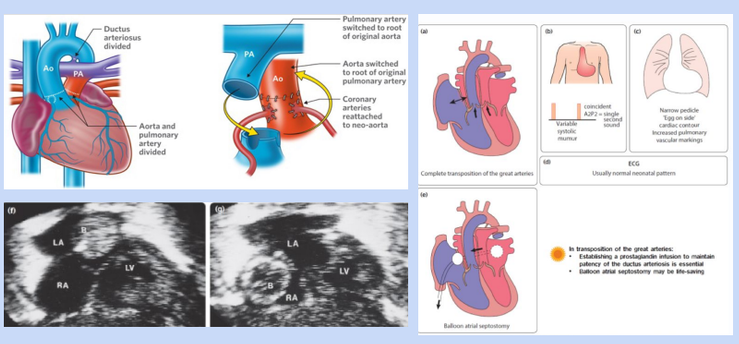
- Severely Cyanosis after birth
- Reverse differential cyanosis if pulmonary HTN
- No signs of respiratory distress
- Single second heart sound
- Typically: no murmur
- Hyperoxic test: FAIL

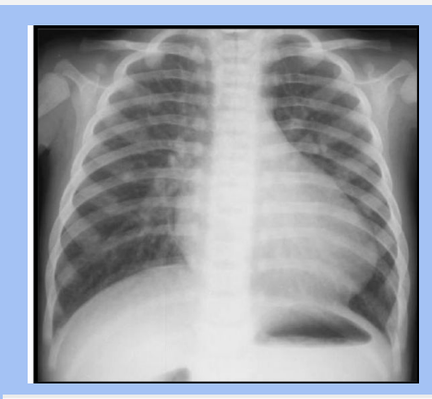
Investigations:

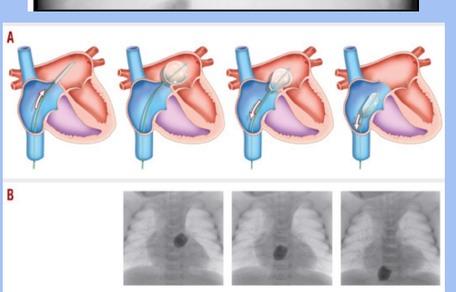
- ➢ Chest X-ray:
  - “egg on a string” appearance
- ➢ ECG: Typically normal
- ➢ ECHO: confirm diagnosis
- ➢ Cardiac Cath:
  - For septestomy
  - +/- coronary arteries anatomy

Management

- ➢ Supportive:
  - Prostaglandin E2
  - Balloon atrial septostomy (for better mixing)
- ➢ Definitive management:
  - Arterial Switch Operation (ASO) for simple D-TGA

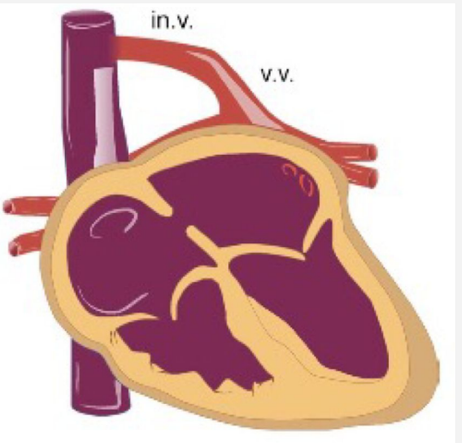






Cyanotic Heart Diseases

Total Anomalous pul,onsry venous return :TAPVD



- All 4 pulmonary veins returns to the right atrium
- Can be:
  - Supracardiac (50%)
  - Cardiac (25%)
  - Infracardiac (20%)
  - Mixed (5%)
- Can be:
  - Obstructed TAPVR
  - Non-obstructed TAPVR

Clinical features:

Cyanosis at birth due to mixing	Tachypnea due to lung congestion	+/- Hypotension if obstructed

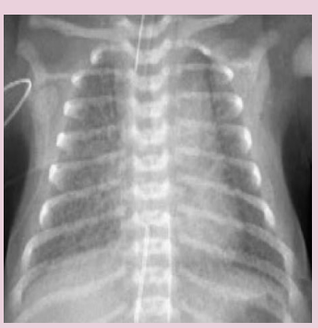
Diagnosis:

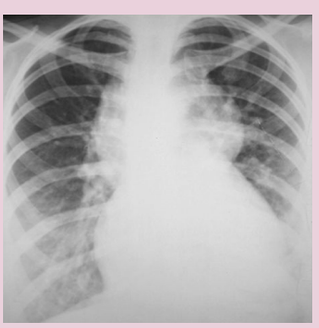
Chest X-ray:
- Figure of Eight
- Supracardiac “obstructed” worse type of TAPVD and they are sicker than TGA because they are cyanoced and hypotensive
- Small heart and RDS picture
- Infra-cardiac “obstructed”

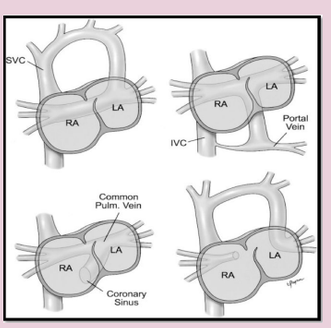
ECG: RVH

ECHO: Confirm diagnosis

Cardiac CT/MRI: Usually needed to further delineate the venous anatomy







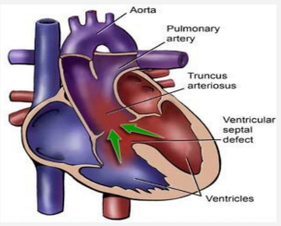
Treatment:

- **Surgery** and the outcome is excellent **(436 notes)**
- Stabilizing the patient prior to surgery as much as possible from a cardiovascular and metabolic standpoint is important. In a newborn with obstructive total anomalous pulmonary venous connection, stabilization often involves **mechanical ventilation**, **correction of acidosis**, **inotropic support**, and **administration of prostaglandin E1 for patency of patent ductus arteriosus** and, in patients with total anomalous pulmonary venous connection type III, for patency of the ductus venosus. **(Medscape)**
- **Nitric oxide** may be useful as a pulmonary dilator postoperatively in patients experiencing episodic pulmonary hypertension that is affecting cardiac output. **(Medscape)**

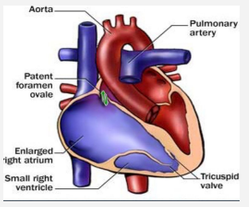
skipped

Others

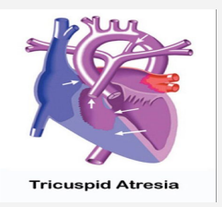
Truncus Arteriosus

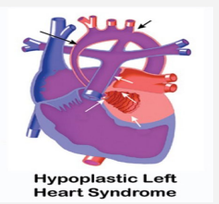


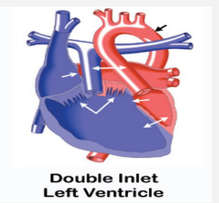
Ebstein's Anomaly



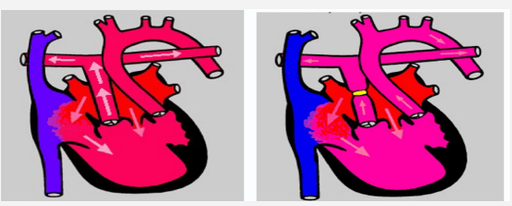
Single Ventricle
Complex CHD in which heart can not be separated into 4 chambers



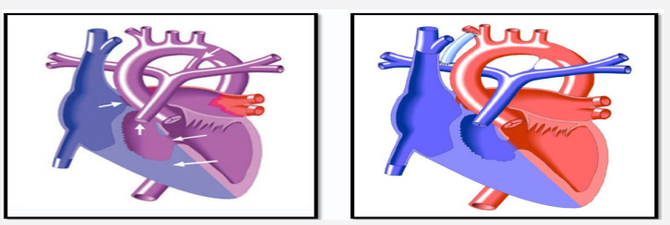




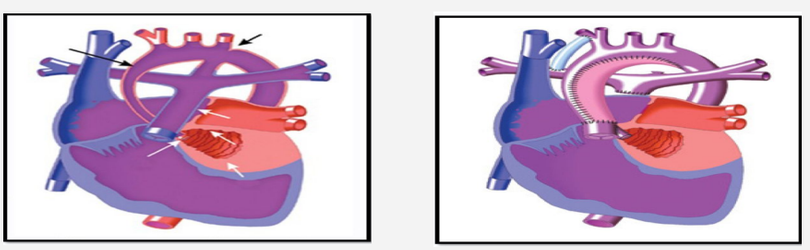
Double Inlet Left Ventricle
Stage 1: Pulmonary artery banding to restrict pulmonary flow



Tricuspid Atresia
Stage 1: BT shunt to augment pulmonary flow

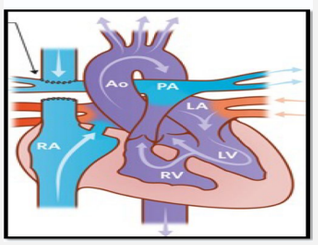


Hypoplastic Left Heart Syndrome
Stage 1: Major arch Construction (Norwood) with BT shunt

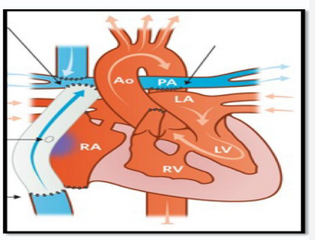


Common Stages Of Univentricular Heart Surgery

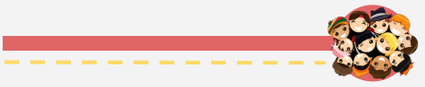
Stage 2: GLENN



Stage 3: FONTAN



Book!



- **What does cause the foramen ovale to close?** The change in the pressure difference between the atria; the pressure in left atrium becomes higher than the one right
- Duct dependent lesions will deteriorate at 1-2 days because usually it closes at this time!
- Innocent murmur can be heard normally in children (normal heart), hallmarks of an innocent ejection murmur are:

Asymptomatic	**Soft blowing murmur**	**Systolic murmur only, NOT diastolic**	**Left sternal edge**

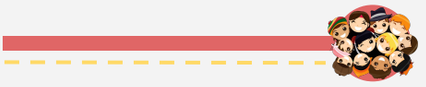
Also, normal heart sounds with no added sounds, no parasternal thrill and no radiation.

- Innocent (or flow) murmurs are often heard during a febrile illness or anemia.
- Causes of heart failure:

- Neonates – obstructed (duct-dependent) systemic circulation
  - Hypoplastic left heart syndrome
  - Critical aortic valve stenosis
  - Severe coarctation of the aorta
  - Interruption of the aortic arch
- Infants (high pulmonary blood flow)
  - Ventricular septal defect
  - Atrioventricular septal defect
  - Large persistent ductus arteriosus
- Older children and adolescents (right or left heart failure)
  - Eisenmenger syndrome (right heart failure only)
  - Rheumatic heart disease
  - Cardiomyopathy

- Symptoms of heart failure will increase up to the age of 3 months but may later subsequently improve as the pulmonary vascular resistance rises in response to the left-to-right shunt >> if left untreated, children will develop **Eisenmenger syndrome** (so improvement of symptoms of these patients is not good sign!!)
- Central cyanosis is less pronounced in **children with anemia**.
- Persistent cyanosis in an **otherwise well infant** is nearly always a sign of structural heart disease.
- Cyanosis with **respiratory distress** >> cyanotic heart disease, respiratory disorders, persistent pulmonary HTN of newborn, infections (GBS), inborn error of metabolism (metabolic acidosis and shock)
- Fetal anomaly scan (including scanning heart) performs between 18 weeks and 20 weeks gestation
- **Indications to do detailed fetal echocardiography:**
- Abnormal fetal anomaly scan
- Fetus at risk (Down syndrome, previous child with heart disease, either parents had congenital heart disease
- Cyanosis >> if **duct dependent**, prostaglandin is vital to survive
- Heart failure >> left to right shunt when **pulmonary vascular resistance falls**
- Shock >> when duct closes in severe left heart obstruction

Book!



- **Types of ASD:** Secundum ASD (it involves **foramen ovale**), Partial AVSD (they have **regurgitant valve**)
- Partial AVSD requires surgical correction
- VSD: the smaller defect, the louder the murmur
- **Prevention of bacterial endocarditis in small VSD:** by maintaining good dental hygiene and avoiding body piercings or tattoos
- Ductus arteriosus connects the pulmonary artery to the descending aorta
- In preterm presence of PDA is not from congenital heart disease but due to prematurity

Summary

Left-to-right shunts

Lesion	Symptoms	Signs	Management
ASD			
Secundum	None	ESM at ULSE Fixed split S_2	Catheter device closure at 3–5 years of age
Partial AVSD	None	ESM at ULSE Fixed split S_2 Pansystolic murmur at apex	Surgery at 3 years of age
VSD			
Small (80%–90% of cases)	None	Pan-systolic murmur at LLSE	None
Large (10%–20% of cases)	Heart failure	Active precordium, loud P_2, soft murmur, tachypnoea, hepatomegaly	Diuretics, captopril, calories Surgery at 3–6 months of age
PDA	None	Continuous murmur at ULSE ±bounding pulses	Coil or device closure at cardiac catheter at 1 year of age, or ligation in preterm

ASD, atrial septal defect; AVSD, atrioventricular septal defect; ESM, ejection systolic murmur; LLSE, lower left sternal edge; PDA, persistent ductus arteriosus; ULSE, upper left sternal edge; VSD, ventricular septal defect.

- Clubbing of the fingers and toes will develop in older children with TOF
- If TET spells prolonged **beyond about 15 mins**, treatment should be give
- **Transposition of GA:** aorta is connected to the right ventricle and the pulmonary artery is connected to the left ventricle (discordant ventriculoarterial connection)

Summary

Cyanotic congenital heart disease

Lesion	Clinical features	Management
Tetralogy of Fallot	Loud murmur at upper left sternal edge Clubbing of fingers and toes (older) Hypercyanotic spells	Surgery at 6–9 months of age
Transposition of the great arteries	Neonatal cyanosis No murmur	Prostaglandin infusion Balloon atrial septostomy Arterial switch operation in neonatal period
Eisenmenger syndrome	No murmur Right heart failure (late)	Medication to delay transplantation

Summary

Common mixing

Lesion	Clinical features	Management
Atrioventricular septal defect (complete)	Down syndrome (often) Cyanosis at birth Breathless at 2–3 weeks of life	Treat heart failure medically Surgical repair at 3 months
Complex disorders (e.g. tricuspid atresia)	Cyanosis Breathless	Shunt (Blalock–Taussig) or pulmonary artery banding, then surgery (Glenn and later Fontan operation)

- **Carotid thrill** is always presenting in aortic stenosis. Ejection systolic murmur and apical ejection click
- Aortic valve replacement in neonates and children with significant aortic stenosis
- Pulmonary stenosis >> ejection systolic murmur
- Stent could be required for adult type CoA
- Interruption of the aortic arch is associated with other conditions such as: DiGeorge syndrome (absence of thymus, palatal defects, immunodeficiency and hypocalcemia, and chromosome 22q11.2 microdeletion)
- The sickest of all neonates with congenital heart disease are with **Hypoplastic left heart syndrome**; weakness or absence of **all peripheral pulses**, in contrast to weak **femoral pulses** in CoA

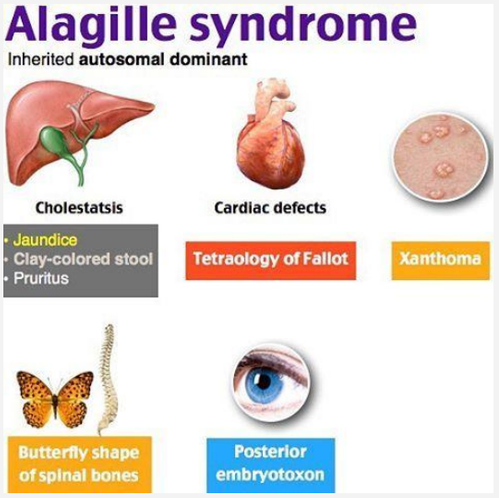
Summary

Left heart outflow obstruction in the sick infant – duct-dependent lesions

Lesion	Clinical features	Management
Coarctation of the aorta	Circulatory collapse Absent femoral pulses	Maintain ABC Prostaglandin infusion
Interruption of the aortic arch	Circulatory collapse Absent femoral pulses and absent left brachial pulse	Maintain ABC Prostaglandin infusion
Hypoplastic left heart syndrome	Circulatory collapse All peripheral pulses absent	Maintain ABC Prostaglandin infusion

Extra!

Alagille syndrome (Not from the book)



- In **reverse differential cyanosis**, the arms are more cyanotic than the legs. This occurs in children with transposition of the great arteries (TGA) when oxygenated blood from the pulmonary circulation enters the descending aorta through a patent ductus arteriosus.

Differential Cyanosis UL SaO_2 > LL SaO_2	Reversed Differential Cyanosis UL SaO_2 < LL SaO_2
- Severe pulmonary hypertension with PDA - Severe aortic coarctation H10(preductal) or interruption	- Transposition of great arteries (or Double-outlet right ventricle with subpulmonary ventricular septal defect) with severe pulmonary hypertension and PDA - TGA with severe aortic arch obstruction / interruption and PDA - Supracardiac TAPVD with PFO and PDA - Anomalous right subclavian artery connected by the ductus to the right pulmonary artery (Isolated RSA) (RUL SaO_2 lower than LUL and LL SaO_2)

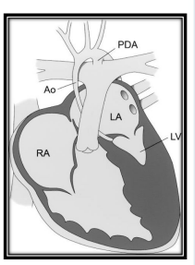
Hypoplastic left heart syndrome (436 slides)

HLHS

- HLHS: one of the most severe form of CHD
  - High morbidity and mortality
- Incidence: 1-2 % of all CHD
- multiple level of obstruction at left heart structures.
  - Mitral stenosis to mitral atresia
  - Variable degree of LV hypoplasia
  - Aortic stenosis to aortic atresia
  - Variable degree of ascending aorta hypoplasia

PATHOPHYSIOLOGY: HLHS

- No adequate flow across aortic valve to ascending aorta
- Relies on retrograde PDA flow to:
  - Brain
  - Coronary arteries
- Need ASD/PFO to shunt blood from LA to RA.



PRESENTATION: HLHS

- At birth: Cyanosis
- At 2-4 week of life:
  - Respiratory distress
  - Poor pulses/perfusion
  - Signs of cardiac shock