MED439
KING SAUD UNIVERSITY

pediatric
439 Teamwork

Revised by The Academic Leader
Homoud Algadheb

Congenital heart disease

objectives:

- Understand the circulatory changes at birth.
- Know the classification of CHD.
- Understand the presentation of different cardiac diseases.
- Know acyanotic heart diseases including VSD, ASD, AVSD, PDA, coarctation of Aorta,aortic valve stenosis and pulmonary valve stenosis.
- Common cyanotic heart diseases including tetralogy of Fallot, transposition of the great arteries, total anomalous pulmonary venous return, hypoplastic left heart syndrome, truncus art Kawasaki disease,infective endocarditis, myocarditis, cardiomyopathy.

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Special thanks to team 437 & Faisal alsaif

Notes

Important

Book

INTRODUCTION

- Congenital heart disease prevalence is 1% of live births
- Etiology mostly unknown
- Chromosomal abnormalities 10 %

Congenital heart diseases (IMPORTANT TABLE)	
Specific CHD (major types)	**Incidence**
Ventricular Septal Defect (VSD)	30%
Patent Ductus Arteriosus (PDA) most common in premature	12 %
Atrial Septal Defect (ASD)	7%
Pulmonary valve stenosis (PS)	7%
Coarctation of Aorta (CoA)	5%
Tetralogy of Fallot (TOF) Most common cyanotic CHD	5%
Aortic valve stenosis (AS)	5%
D-Transposition of great arteries (D-TGA) Most common cause of severe cyanosis after birth	5%
Atrioventricular septal defect (AVSD)	2%

Chromosomal Abnormality Associated with CHD		
Trisomy 21 (Down syndrome) **(MCQ)**	AVSD	VSD
Trisomy 18 (Edwards syndrome)	VSD	PDA, CPVD
Trisomy 13 (Turners syndrome)	PDA,VSD,ASD	TAPVR
Digeorge syndrome	Interrupted aortic arch	Cono-truncal abnormality
Williams syndrome	Supra-aortic stenosis	Pulmonary Branches Stenosis
Noonan syndrome	Pulmonary valve stenosis	Hypertrophic Cardiomyopathy
Turner syndrome	Coarctation of aorta	Bicuspid aortic valve

Beyond your level

key concepts

Blood flow:

blood flow depends on:

- **Resistance:**

Blood flow easier to low resistant organs

- **Pressure Gradient:**

Blood flow from high pressure to low pressure chambers

- **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

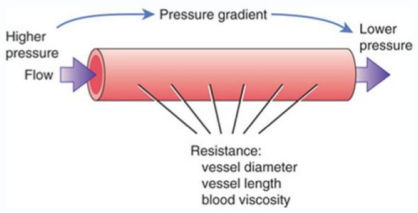
- **Systemic duct dependent circulation**
  example: critical aortic valve stenosis, critical coarctation of aorta, Interrupted aortic arch, HLHS

CVS Flow and Resistance

★ (MCQ)
Which one is the most determinant of flow? **Radius**

high viscosity increases resistance
e.g. in polycythemia the resistance will be high but in anemia it will be low

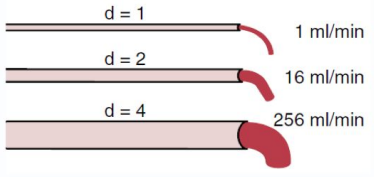
Flow



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

Q = Flow
ΔP = Pressure Gradient
R = Resistance

Resistance



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

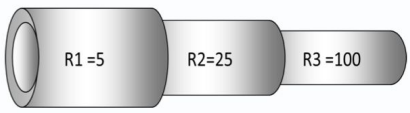
R: Resistance
η : Viscosity
L: Length
r: Radius

CVS Resistance

In **serial circuit** the total resistance will be equal to the sum of the three arteries

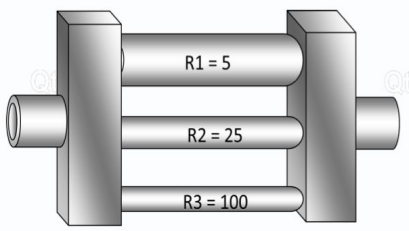
In the **parallel circuit** the total resistance will be always less than the artery with lowest resistance
E.g. coarctation of aorta , creates an additive resistance

Serial Circuit



R total = R1 + R2 + R3
R total = 5 + 25 + 100
R total = 130

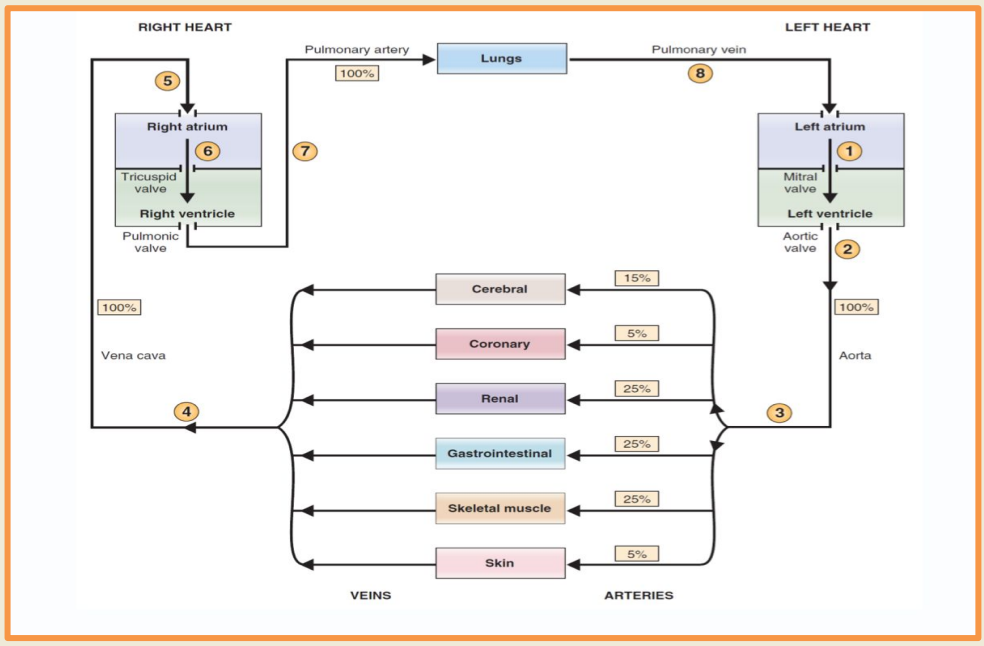
Parallel Circuit



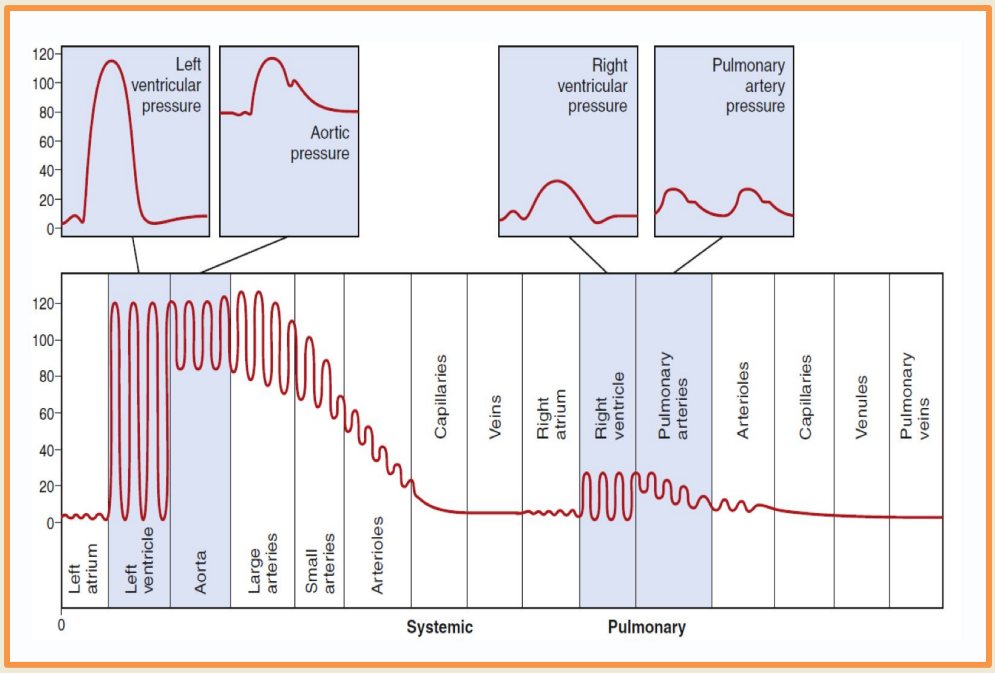
1/R total = 1/R1 + 1/R2 + 1/R3
= 1/5 + 1/25 + 1/100
= 20/100 +4/100+1/100 = 25/100 = 1/4
R total = 4

cardiovascular physiology overview

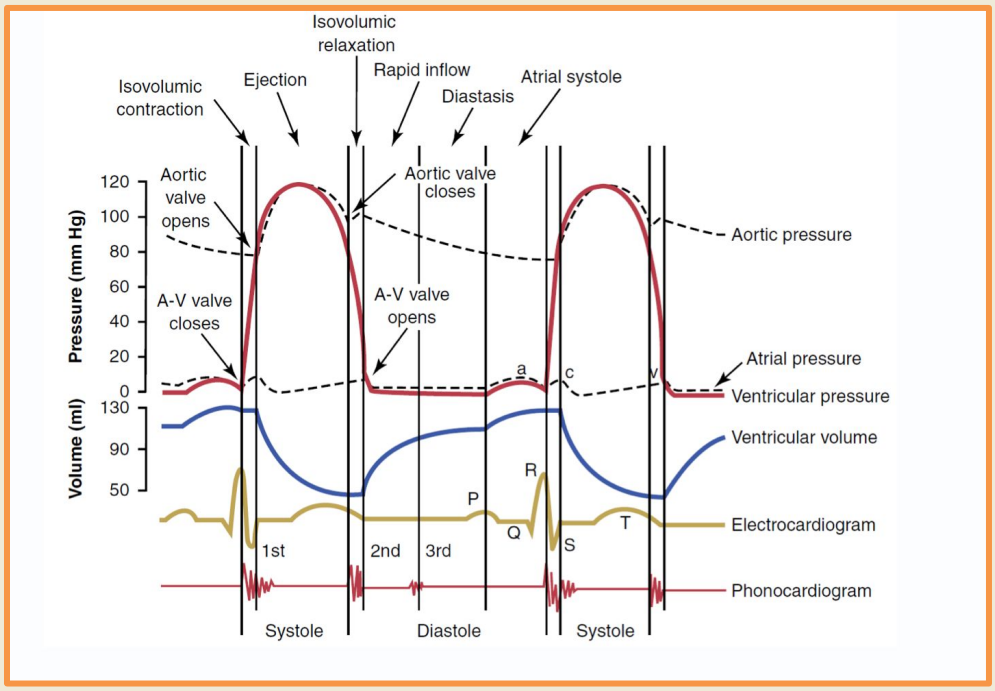
CVS Circulation



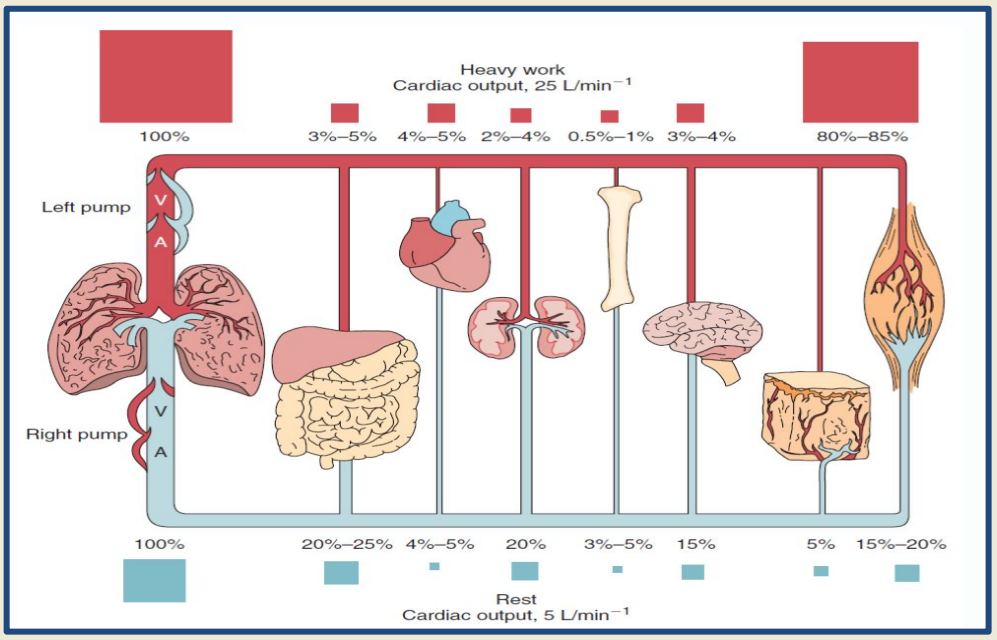
CVS Driving Pressure



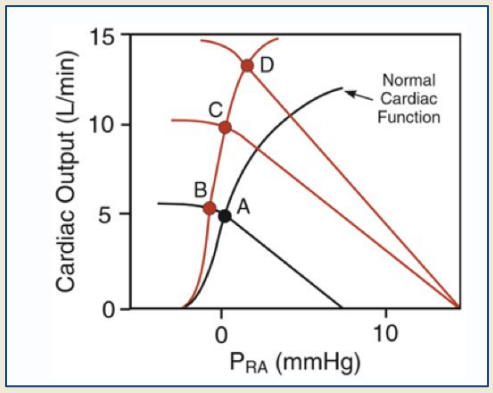
Cardiac Cycle



Cardiac Systemic Output



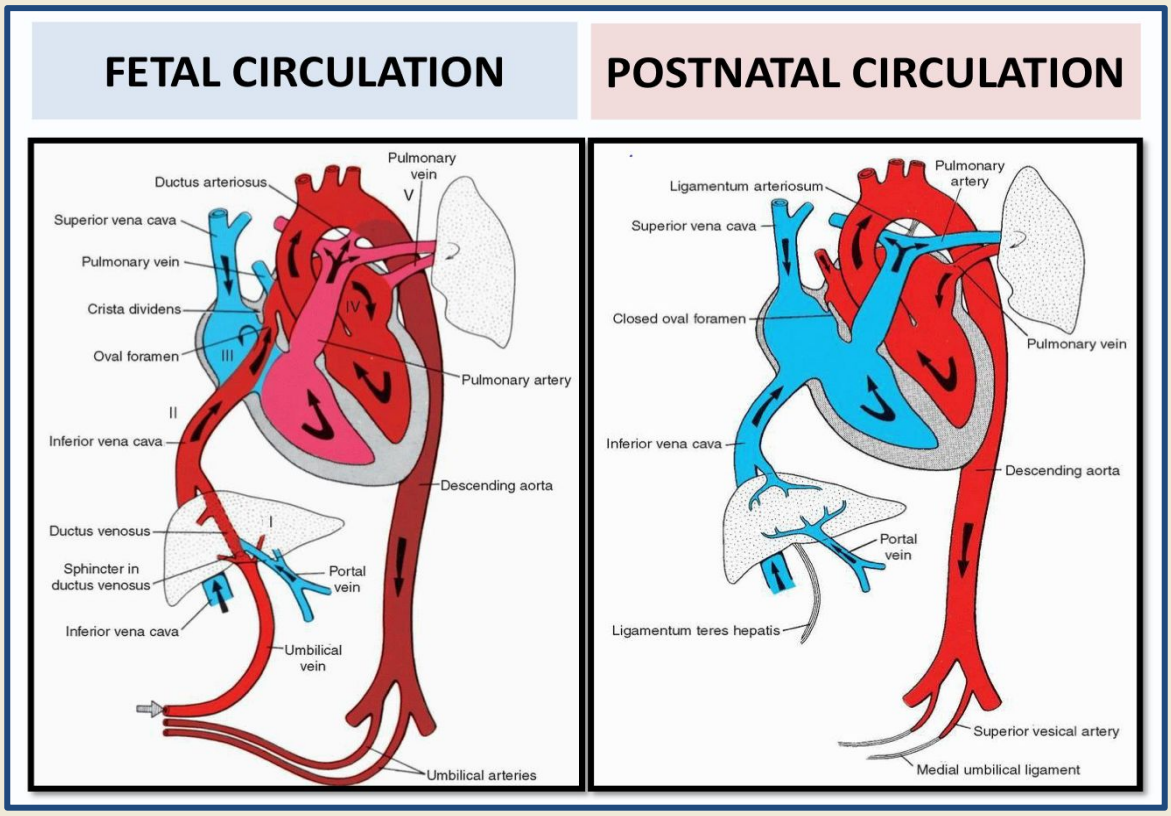
Cardiac and Venous Function Curve



Cardiac Output Increases during Exercise due to:

- **Increase contractility**
- **Increase preload**
- **Reduce afterload**
- **Increase heart rate**

Fetal vs Postnatal Circulation (MCQ)



- Lung is not utilized (placental instead), added in parallel so systemic vascular resistance is low.

- Fetal life = higher pulmonary vascular resistance (**special in fetal circulation**, opposite to postnatal), while the systemic vascular resistance is even lower than postnatal.
  lung takes 5-10% of blood flow in fetal life to ensure lung survival through the pulmonary artery. Since it only takes 5-10, pulmonary vein also ends up with the same resistance, so does the blood flow to left atrium , so left atrium has low pressure in fetal life.

- SVC, IVC, foramen ovale all open in the right atrium (the foramen ovale shunts right to left because it is pressure dependent, and the right atrium has more pressure than left atrium in fetal life. Ductus venosus decreases the flow to the heart (creates resistance) from umbilical vein, to protect the heart from the high low

CVS CHANGES AFTER BIRTHS

- Closure of ductus arteriosus[1]
- Closure of ductus venosus[2]
- Closure of foramen ovale[3]
- Increase of systemic vascular resistance after clamping the placenta
- Decrease of pulmonary vascular resistance[4] after the baby cries

- What is a sign that the lung is starting gas exchange when the baby is born? the child's first cry —> makes the lung work (that's why not hearing a cry is concerning).
- When you cut the umbilical cord, you:

1- Eliminate the placenta, which increase the systemic vascular resistance —> decrease the pulmonary vascular resistance (normalizes after 6-8 weeks)
2- Decrease the blood sources of right atrium, to only IVC and SVC
3- The ductus arteriosus closes (due to pressure gradient)—> constricts in 1-2 days (physiological closure)), and fibrose in 1-2 weeks due to decrease in prostaglandin (anatomical closure)

Classification of CHD

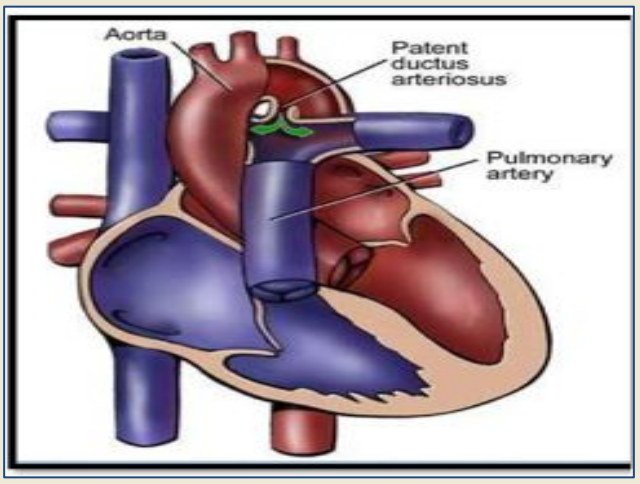
Cyanotic heart disease	Acyanotic Heart Disease
Decreased pulmonary flow: • Tetralogy of Fallot • Tricuspid atresia • Other univentricular heart with pulmonary stenosis	**Left – Right shunt lesions:** • Ventricular septal defect • Atrial Septal Defect • Atrio-ventricular Septal Defect • Patent Ductus Arteriosus
increase 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**"

1- Due to the increase in SVR and decrease in pulmonary vascular resistance
2- Due to cut of umbilical circulation—> no flow through the ductus venosus —> thromboses and closes
3- Due to the decrease in right atrial pressure after the elimination of umbilical vein flow, makes the left atrium higher in pressure compared to right atrium —> closes due to the pressure difference
4- Will not normalize until 6-8 weeks which is why some murmurs are not heard at birth, but rather weeks after, once the pulmonary vascular resistance fall (e.g. VSD and PDA)

Acyanotic Heart Disease (Left-to-Right Shunt lesions)

pathophysiology, symptoms and signs MCQ

PDA

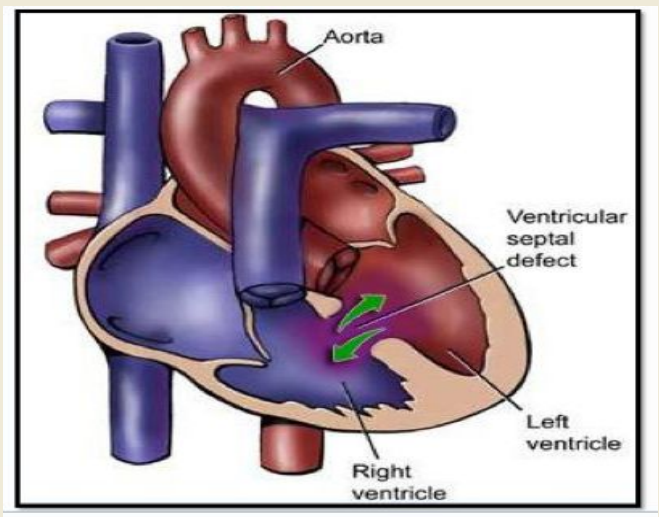


L-R shunt at artery level:
- Dilated LA and LV
-Enlarged pulmonary arteries

symptoms :
-Small PDA: Asymptomatic
-Moderate to large PDA: chronic heart failure

Examination:
- Small PDA: Silent
- Large PDA: Continuous "machinery" murmur
- Large PDA: Widened pulse pressure

VSD

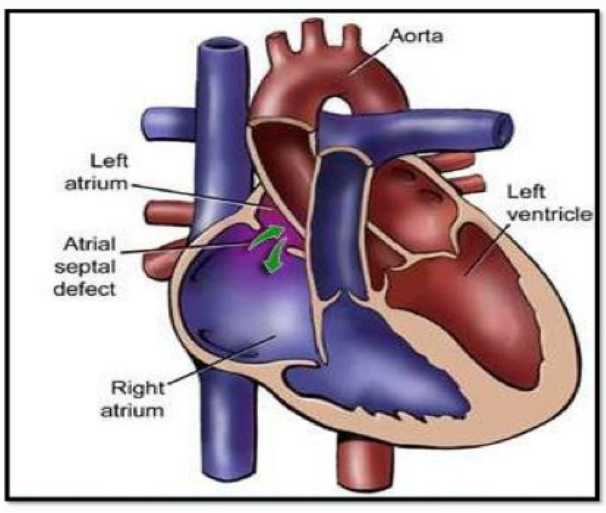


L-R shunt at ventricular level:
- Dilated LA and LV
- Enlarged pulmonary arteries

symptoms:
-Small VSD: Asymptomatic
-Moderate to large VSD: chronic heart failure

Examination:
- Holosystolic murmur
- Small muscular: ejection systolic murmur

ASD



L-R shunt at atrial level:
- Dilated RA and RV
-Enlarged pulmonary arteries

symptoms
-Usually asymptomatic
-Older children: activity related SOB or arrhythmia
-Rare: chronic heart failure

Examination
- Fixed widely splitted second heart sound
- Ejection systolic murmur

CONGESTIVE HEART FAILURE

exam scenario will include these S&S

SYMPTOMS:
- Diaphoresis
- Poor feeding
- Failure to thrive
- Shortness of breath
- Recurrent chest infection
- Exercise intolerance

SIGNS:
- Tachypnea
- Tachycardia
- Cardiomegaly
- Hepatomegaly
- Active precordium

No symptom during neonatal period in VSD due to high pulmonary vascular resistance Symptoms of CHF started ~ 2/12 of age

INVESTIGATION: L-R SHUNT

Diagnosis:

Chest X-ray:
Increased pulmonary vascular marking
Cardiomegaly

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

ECHO:
Confirm diagnosis

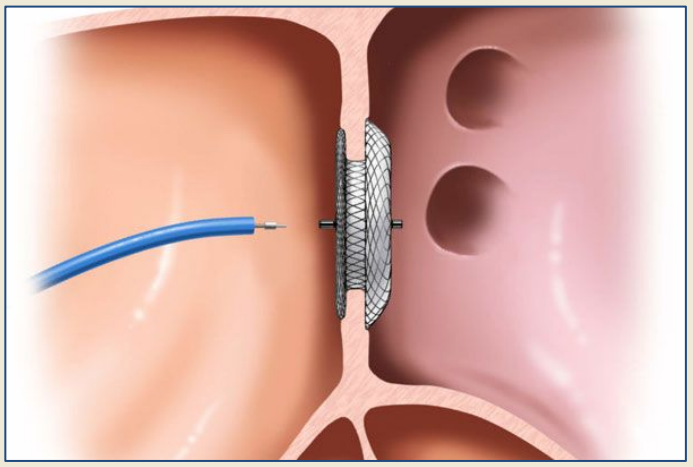
Cardiac Cath:
Not required for diagnosis
Might needed for more detailed hemodynamic assessment or when CATH intervention is planned

Medical treatment L-R shunt

- **Anticongestive therapy:**
  - Diuretics
  - Afterload reducing agents
- **Nutritional support**
- **ASD: usually no medication**

Intervention

VSD:
Usually surgical closure: 4-8 months
Older children when suitable: CATH closure



ASD:
Usually CATH closure around 3-6 years
Some types need surgical closure

PDA:
Mostly CATH closure
Surgery for premature and symptomatic babies

Prognosis

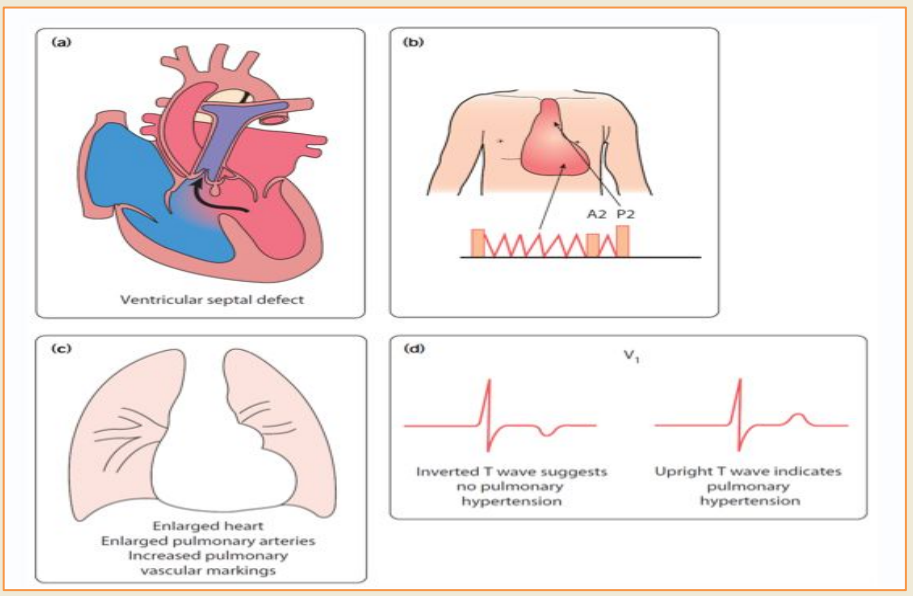
Untreated VSD and PDA beyond infancy:

- Eisenmenger's syndrome which manifest as:
  - Sign and symptom of CHF will disappear
  - Patient will become cyanotic
  - Leading to R-L shunt

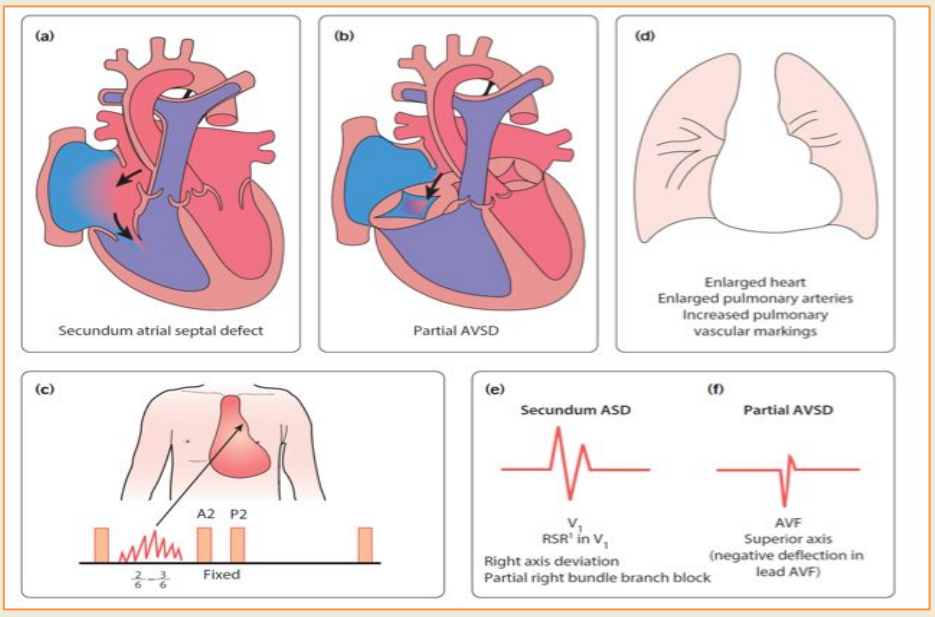
Untreated ASD:
Complications happened during **adult Life**:
- Eisenmenger's syndrome
- Atrial arrhythmias
- Paradoxical embolism (rare)

VSD

Determinant of flow is resistance and pressure: resistance is the most
important because in a very big VSD there will be equalization of flow despite
the pressure



ASD



Two types of ASD:

• **Secundum ASD** (80% of ASDs):
a defect in the centre of the atrial septum involving the foramen ovale.

• **Partial AVSD** (primum ASD): a defect in AVSD characterized:
an interatrial communication between the bottom end of the atrial septum and the atrioventricular valves (primum ASD) abnormal atrioventricular valves, with a left atrioventricular valve which has three leaflets and tends to leak (regurgitant valve).

Presentation:
• lung congestion (due to the left to right shunt increasing the blood volume in the right side of the heart)
• Commonly asymptomatic
• Recurrent chest infections/wheeze
• Arrhythmia from fourth decade onwards

Physical signs:
• ejection systolic murmur at the left sternal edge (due to the increase in pulmonary valve blood flow)
• Fixed, wide split in second heart sound (due to right ventricular stroke volume being equal in both inspiration and expiration.
• Partial AVSD = pansystolic murmur

Management:
only for significant ASD (large enough to cause right ventricle dilation)
• secundum ASD: Cardiac cath with insertion of occlusion device
• Partial AVSD: surgical (at 3-5 years of age)

you should take a break after this slide!
Do 10 jumping jacks and drink some water
it's an advice from ur leader!

PDA

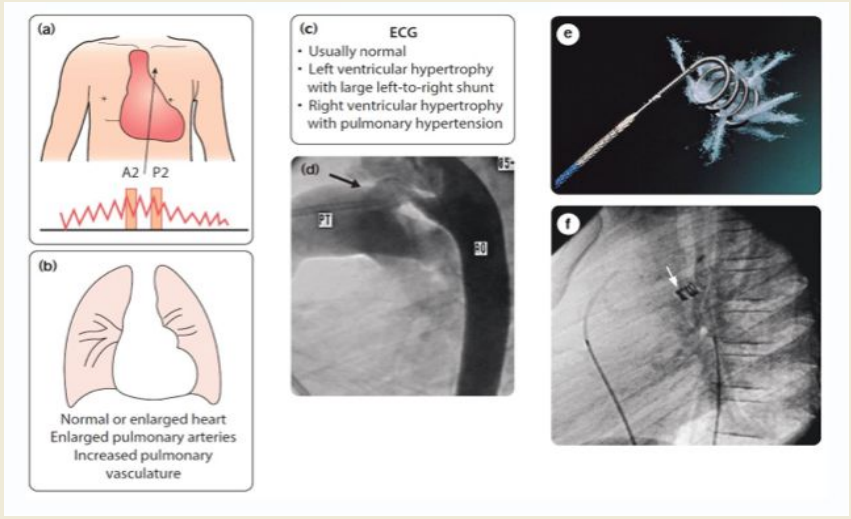
clinical features:

• continuous murmur beneath the left ventricle

• Increased pulse pressure (—> collapsing pulse)

Management:

Closure is recommended to avoid risk of bacterial endocarditis (via cath, at 1 year of age)



Atrioventricular septal defect (AVSD)

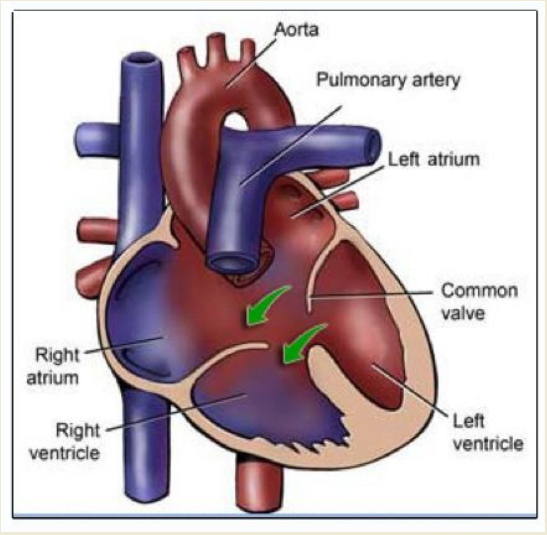
Incidence:

- 4 % of all CHD associated with Down Syndrome (50%)

Divided into:

- Complete AVSD, Partial AVSD, Transitional AVSD

Balanced vs Unbalanced



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.

Atrioventricular septal defect (AVSD)

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, which manifest as:**
  – Diaphoresis
  – Poor feeding
  – Failure to thrive
  – Shortness of breath
  – Recurrent chest infection
  – Exercise intolerance

Physical Examination:

- Feature of Down Syndrome
- Tachypnea
- Tachycardia
- Active precordium
- Murmur: Pansystolic murmur
- Hepatomegaly

Diagnosis:

Chest X-ray:
Increased pulmonary vascular marking
Cardiomegaly

ECG (MCQ)
Left Axis deviation with right ventricular hypertrophy is very suggestive of AVSD

ECHO
Confirm diagnosis

Cardiac Cath
Not required for diagnosis unless more detailed hemodynamic assessment is needed

Atrioventricular septal defect (AVSD)

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

Coarctation of Aorta (CoA)

Acyanotic Heart Disease obstructive lesion

- 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

Pathophysiology:

CRITICAL CoA

- **Spontaneous PDA closure:**

– Lower body hypo-perfusion
–Hypotension
–Acidosis
– LV dysfunction
–Cardiogenic Shock

- **Presented 2-3 weeks of life with:**

Sign of CHF

Circulatory collapse

Shock

Death

"DUCT DEPENDENT CHD"

MILD CoA

- Collateral vessels develop over time
- Flow maintained between proximal and distal aorta
- **Present later on life with:**

-Murmur

-Chronic hypertension

-Headache

-Fatigue

-Stroke

- ❖ Rupture cerebral aneurysm

Coarctation of Aorta (CoA)

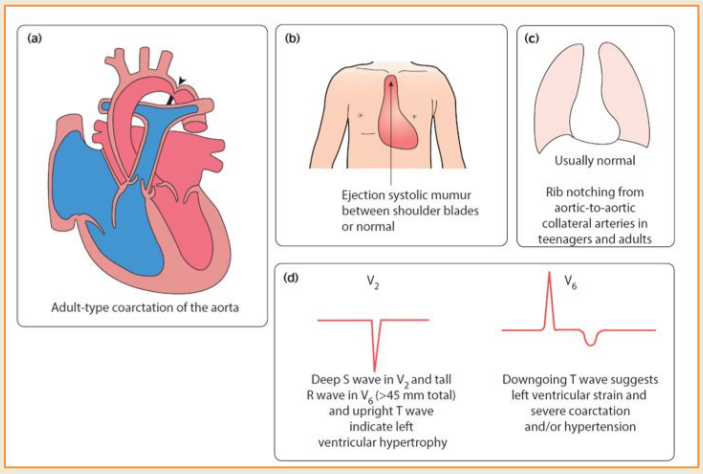
Physical Examination:

Differential cyanosis (severe CoA in newborn)
– Signs of cardiac shock
– 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

Diagnosis



Chest X-ray:

- Cardiomegaly
- Prominent aortic knob
- Rib notching

Due to intercostal collateral vessels, Rarely seen before age of 10 years

ECG:

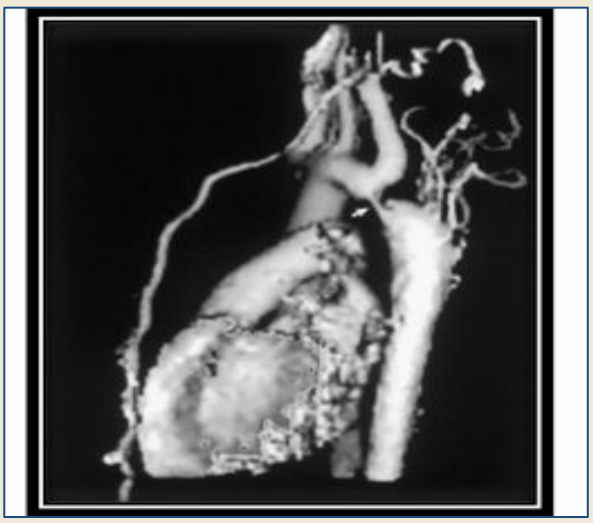
- Neonate: RV hypertrophy
- Older children: LV hypertrophy

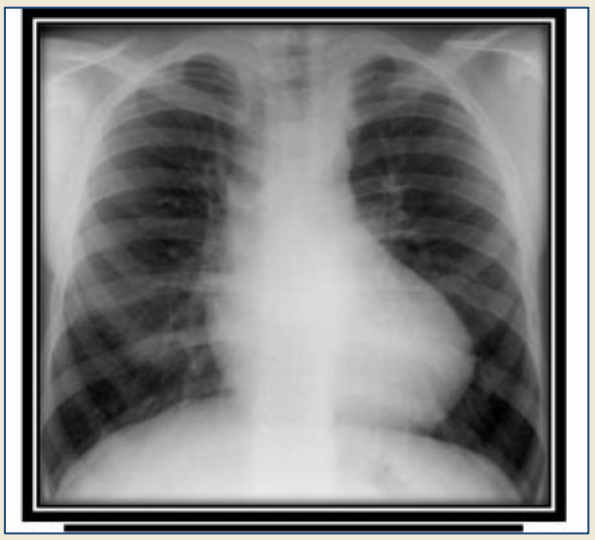
ECHO:

- Establish the diagnosis

Cardiac CT /MRI:

- Might be needed to delineate the arch anatomy



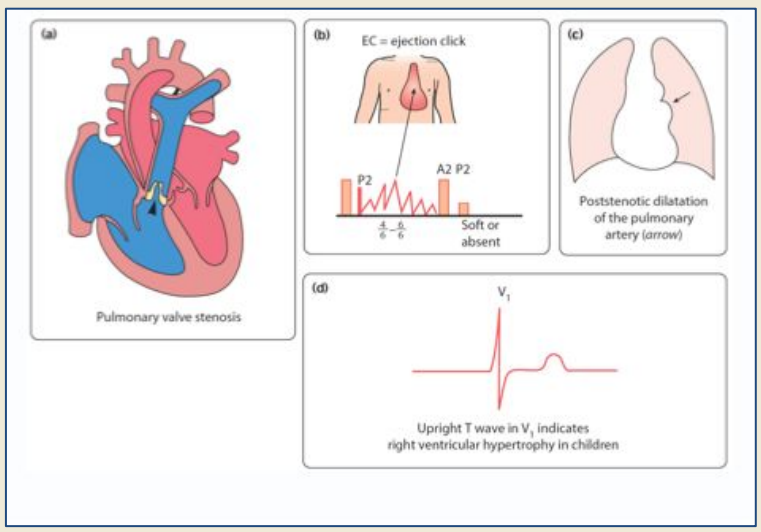


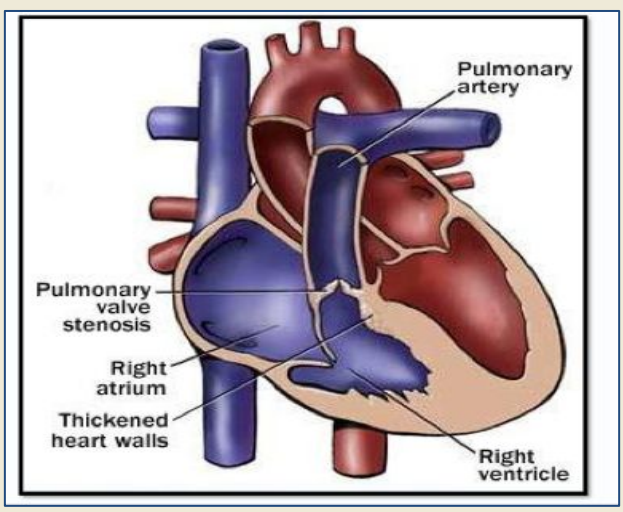
Coarctation of Aorta (CoA)

Treatment:

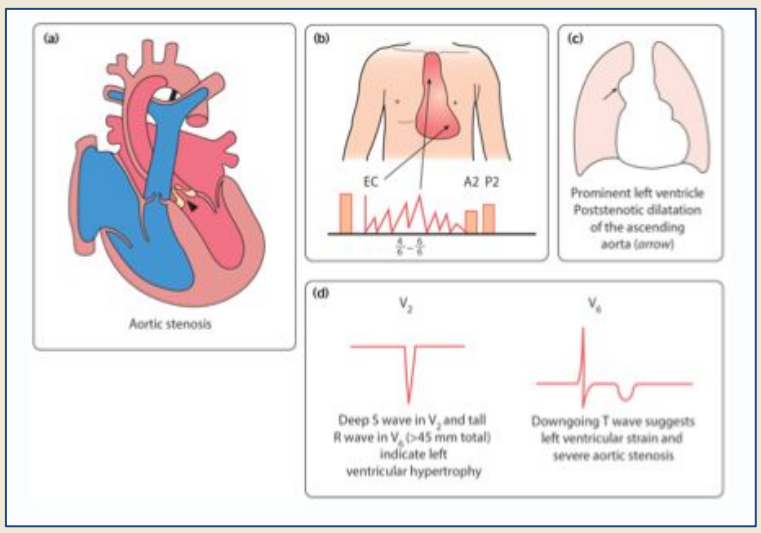
- **Critical CoA** “Duct Dependent CHD”
  Prostaglandin E2 to keep PDA open
- Surgery is the primary intervention
- Trans-catheter balloon angioplasty +/- stent:
  - Recurrent CoA
  - Primary intervention: Discrete CoA in older children

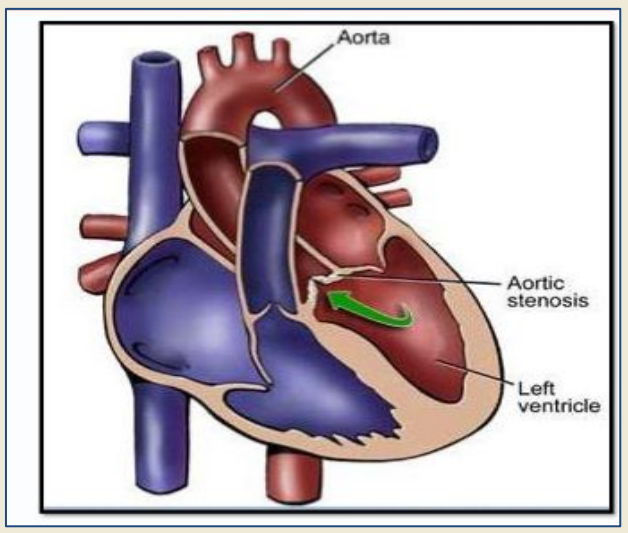
Pulmonary Valve Stenosis





Aortic valve stenosis



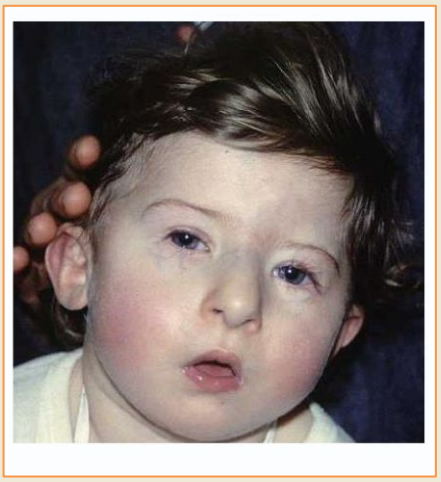


Cyanotic Heart Disease

(MCQ) Dr:if I ask you which one of the following choices is acyanotic heart disease you should know it

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

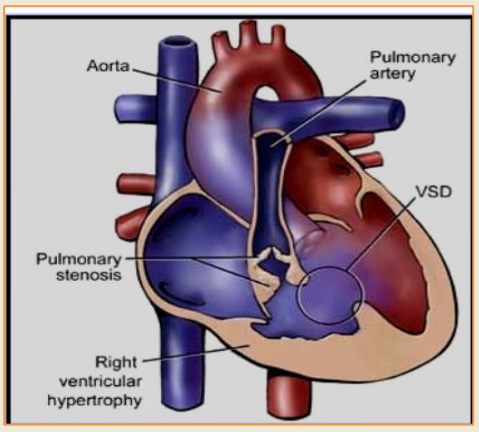
Tetralogy of fallot



- Most common cyanotic CHD the Incidence: 6 % of all CHD
- Can be associated with DiGeorge Syndrome and Alagille syndrome

★ Four basic components **important**

- Large VSD
- Pulmonary stenosis
- Overriding aorta
- RV hypertrophy



CLINICAL FEATURES: TOF

Determined by severity of pulmonary stenosis

Most newborns:

- Asymptomatic[1]
- Ejection systolic murmur
- Initially: mild cyanosis which progresses over time
- Might present with hypercyanotic spells[2] "tet spell" if intervention delayed
-Usually occur around 9-12 months of age – Episodes of acute and severe cyanosis

1- Diagnosed antenatally or following the identification of a murmur in the first 2 months of life.
2- The classical description of severe cyanosis, hypercyanotic spells and squatting on exercise developing in late infancy

Tetralogy of fallot

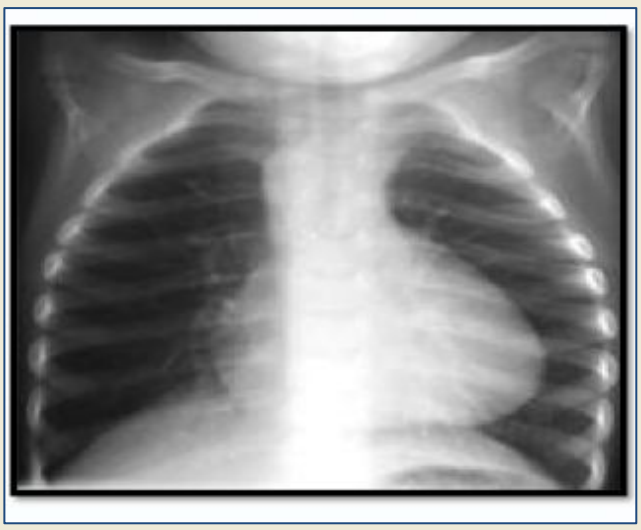
Management	Effect
Reduce anxiety give morphine	Reduce pulmonary vascular resistance
Oxygen most potent pulmonary dilator	Reduce pulmonary vascular resistance
Sedation with morphine	Reduce pulmonary vascular resistance
Knee to chest position	Increase systemic vascular resistance
Phenylephrine alpha agonist (vasoconstriction)	Increase systemic vascular resistance
IV fluid	Increase cardiac filling
Beta blocker	Reduce heart rate

CLINICAL FEATURES: TOF

- Newborn with critical PS or pulmonary atresia
  - Severe cyanosis when PDA close
  - "Duct dependent CHD"
  - Needs "Prostaglandin E2"

INVESTIGATION: TOF

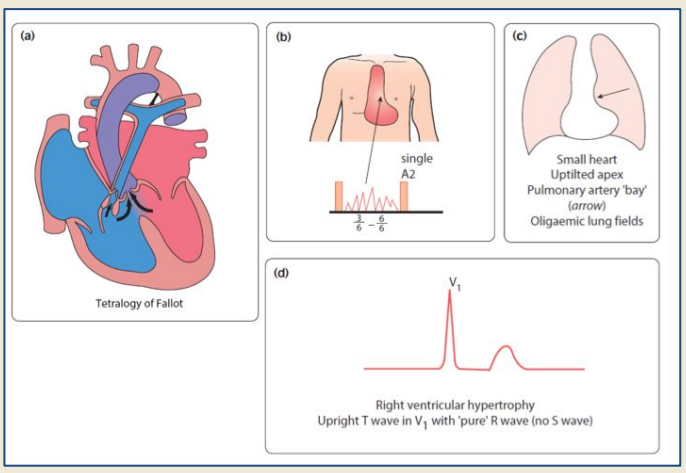
CHEST X-RAY:
"boot-shaped heart"
Oligemic lungs



ECG:
RVH

ECHO[1]:
confirm diagnosis

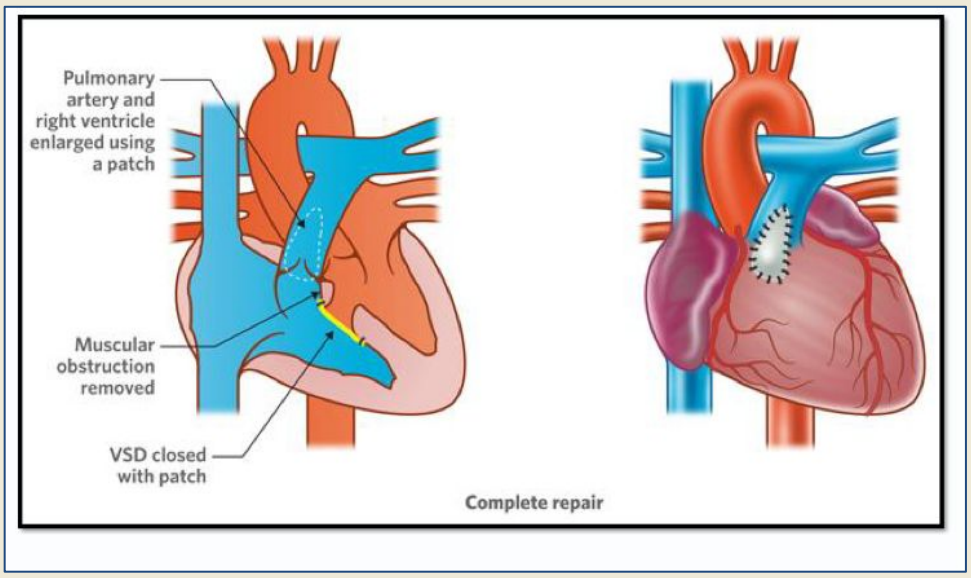
CT/MRI:
might be needed to delineate PA branches



Echo will demonstrate the cardinal features, but cardiac catheterization may be required to show the detailed anatomy of the coronary arteries.

Tetralogy of fallot

TREATMENT

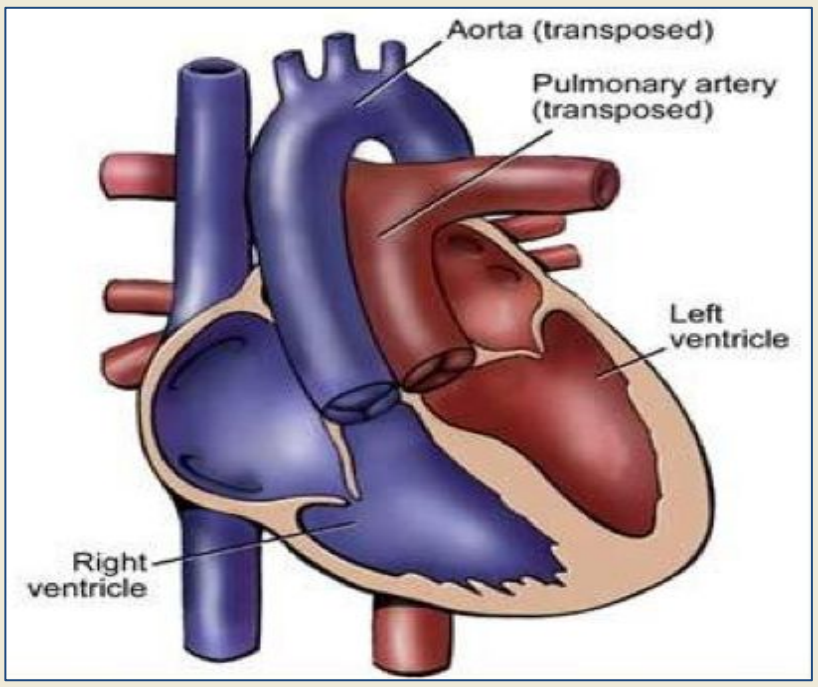


Management of TOF:

- Initial management is medical, with definitive surgery at around 6 months of age
- Infants who are very cyanosed in the neonatal period require a shunt to increase pulmonary blood flow (a modified Blalock–Taussig shunt)
- Management of hypercyanotic spells (if prolonged more than 15mins): treated according to need:
  - Sedation
  - Pain relief (morphine)
  - IV propranolol (or alpha agonist)
  - IV fluid
  - Bicarbonate (to correct acidosis)
  - Ventilation

Transposition of the 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



Transposition of the great arteries

PATHOPHYSIOLOGY: D-TGA

- **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 within oxygenated blood in the pulmonary circulation

★ Mixing of oxygenated and deoxygenated blood can occur at three levels:
– Atrial level via ASD/PFO (most efficient) MCQ so normally there is no ASD but with catheter we can go and open a hole between the two atriums so the blood can mix
– Great arteries level via PDA
– Ventricular level via VSD (if present)

PRESENTATION: D-TGA

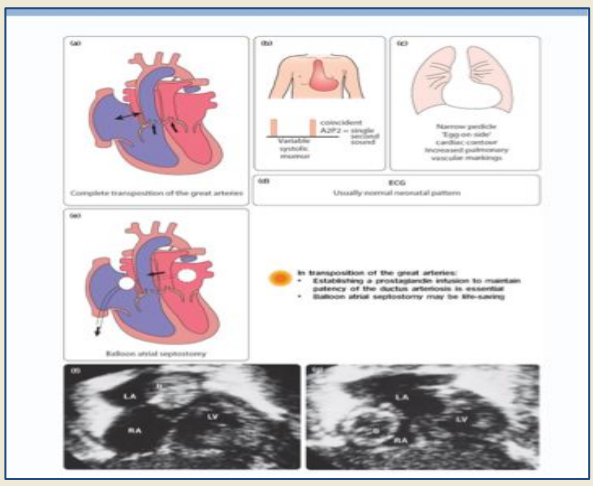
Cyanosis is always present, it can be
less severe in the presence of ASD

• Severely Cyanosis after birth. Cyanosis is always present, it can be less severe in the presence of ASD
• **"Reverse differential cyanosis" if pulm HTN**
• No signs of respiratory distress
• Single second heart sound
• Typically: no murmur
• Hyperoxic test: FAIL

INVESTIGATION: D-TGA

Chest X-ray: Increased pulmonary vascular marking due to increased pulmonary flow

• "egg on a string" appearance



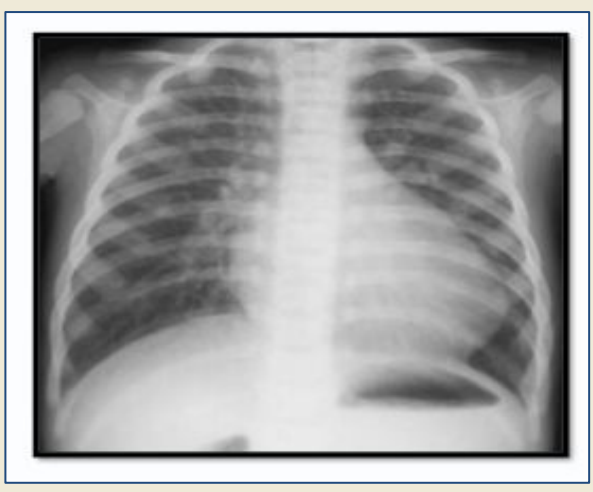
ECG:

• Typically normal

ECHO:
confirm diagnosis

Cardiac Cath:

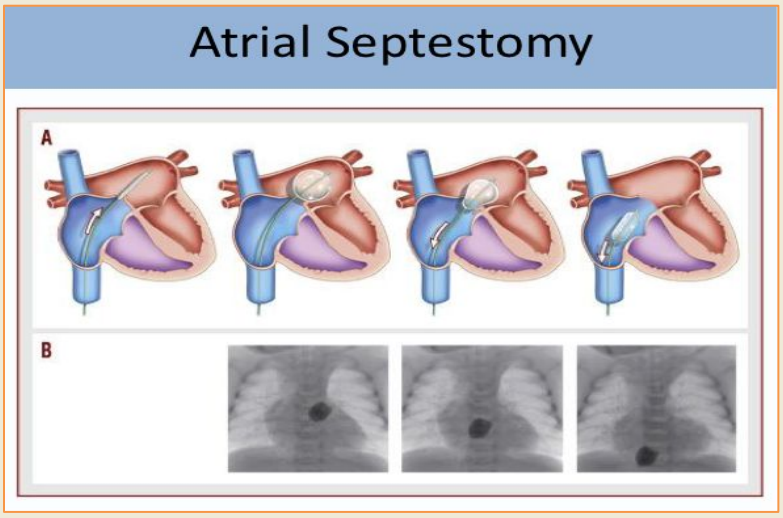
• For septostomy
• +/- coronary arteries anatomy

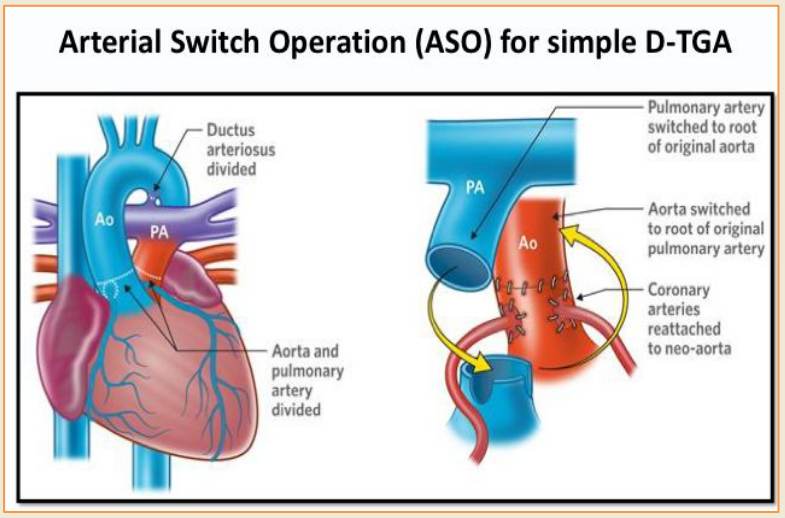


Transposition of the great arteries

MANAGEMENT: D-TGA

Supportive:
- Prostaglandin E2
- Balloon atrial septostomy (for better mixing)





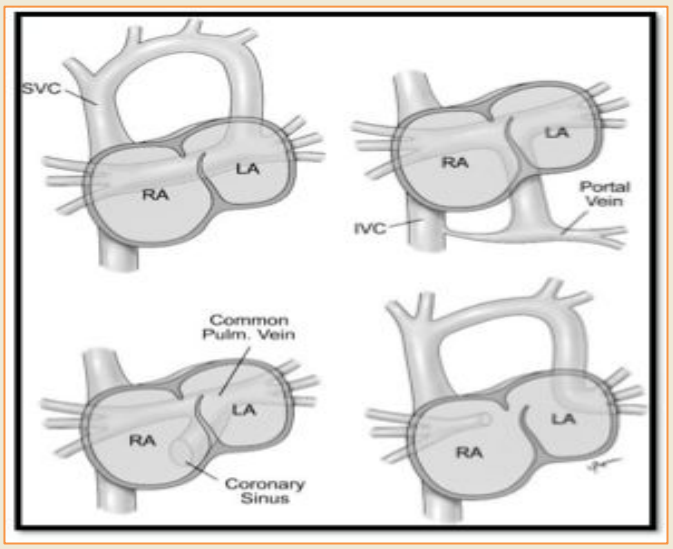
Total Anomalous Pulmonary Venous Return : TAPVD

- **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

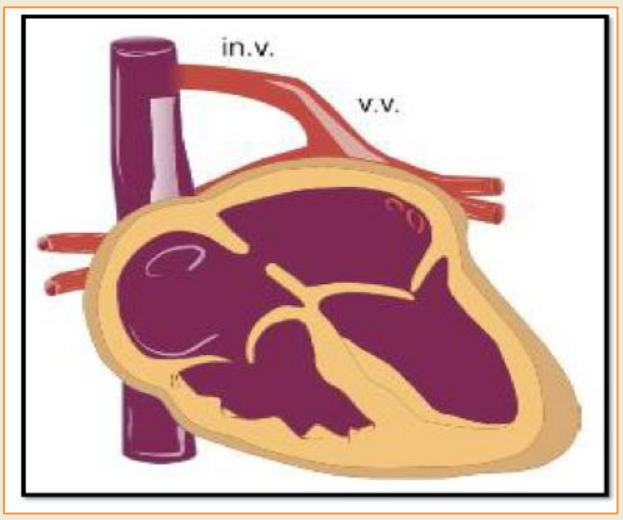


Equal saturation in all chambers (**unique feature to TAPVD**, the only congenital heart disease with this presentation)

Blue Baby MCQ

- How to differentiate between respiratory and cardiac cyanosis at birth?
  - **Respiratory:** combined with signs of respiratory distress (e.g. grunting) + improves with oxygen supplementation

  - **Cardiac:** breathing normally + does NOT improve with Oxygen



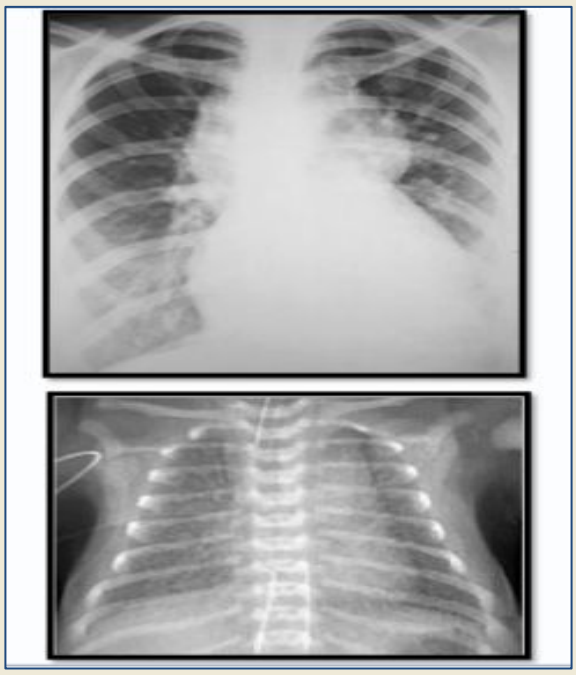
Total Anomalous Pulmonary Venous Return : TAPVD

PRESENTATION: TAPVD

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

Diagnosis:

Chest X-ray:
Figure of Eight
Supracardiac “obstructed”
Small heart and RDS picture
Infra-cardiac “obstructed”

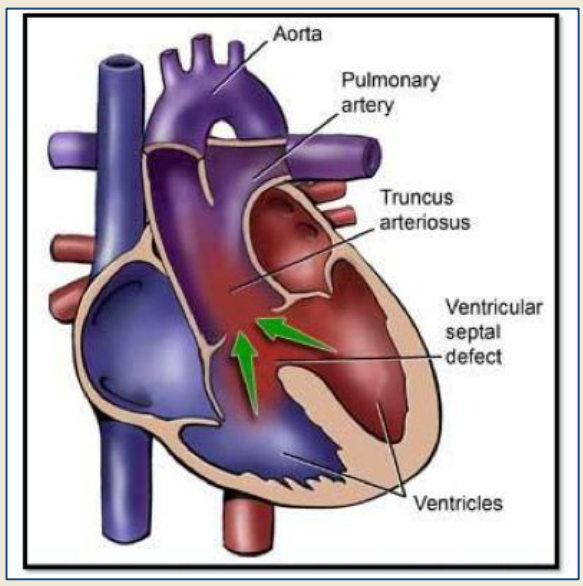


ECG:
RVH

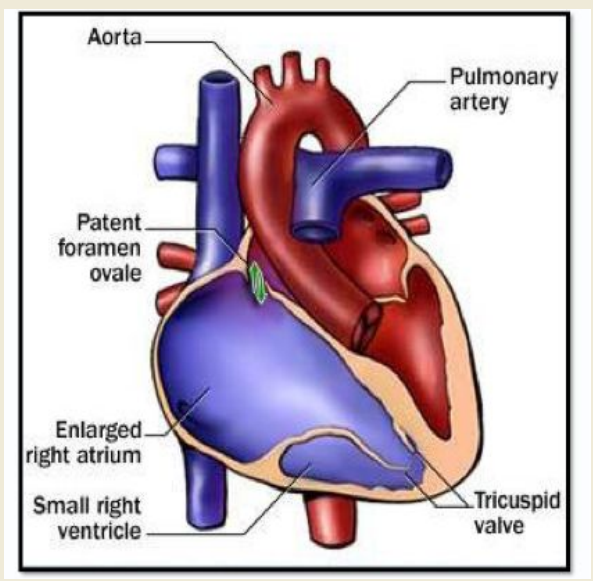
ECHO:
Confirm diagnosi

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

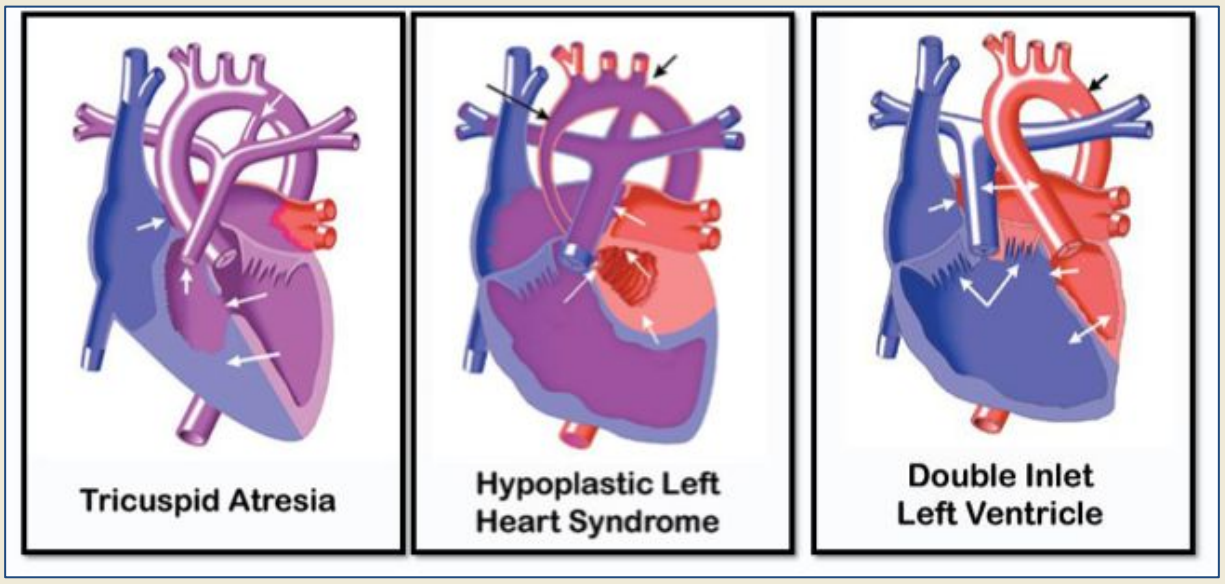
Truncus Arteriosus



Ebstein’s Anomaly

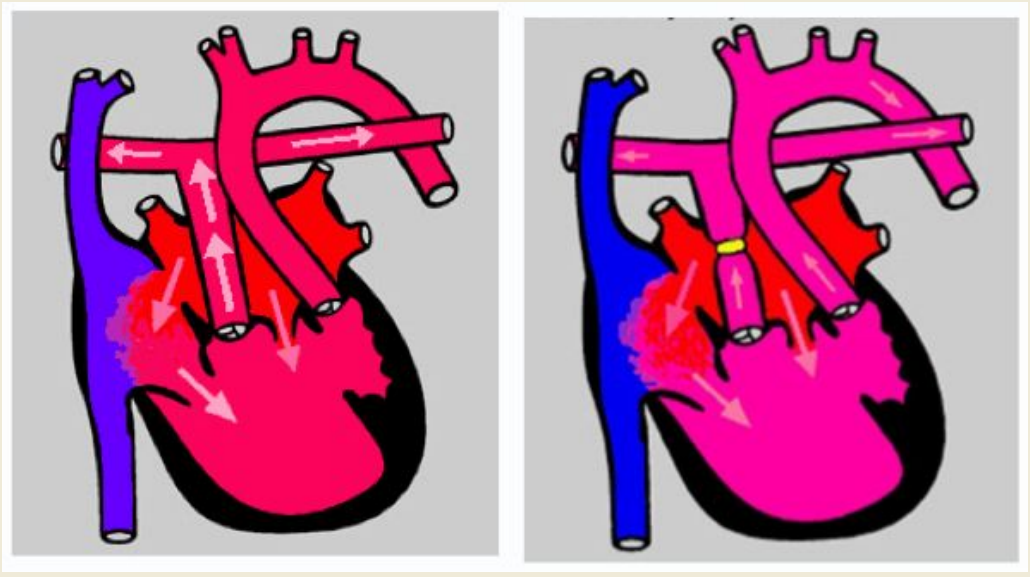


Single Ventricle



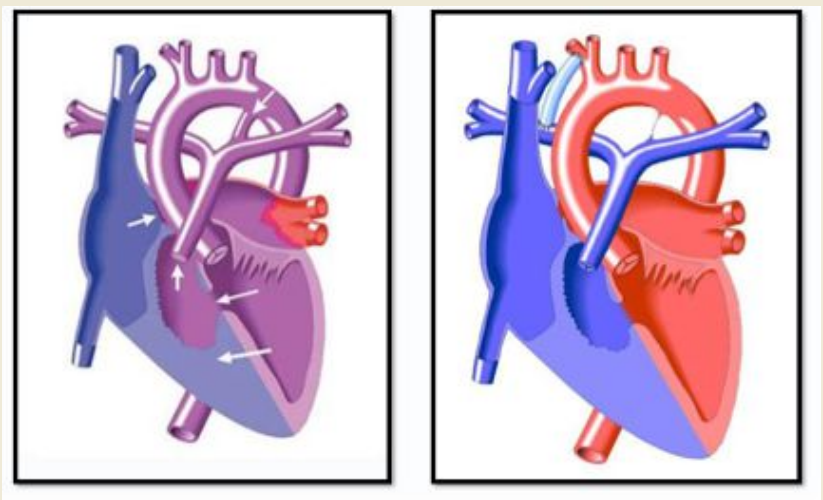
Complex CHD in which heart can not be separated into 4 chambers

Double Inlet Left Ventricle



Stage 1: Pulmonary artery banding to restrict pulmonary flow

Tricuspid Atresia



Stage 1: BT shunt to augment pulmonary flow

Only the left ventricle is effective, the right is small and nonfunctional

- Presentation:
  - If duct dependent: cyanosis at birth
  - If not: becomes cyanosed and breathless later
- Management: early palliation to maintain secure blood supply to the lung.

details are extra from abu wardah ;)

Hypoplastic Left Heart Syndrome [1]

In this condition there is underdevelopment of the entire left side of the heart). The mitral valve is small or atretic, the left ventricle is diminutive, and there is usually aortic valve atresia. The ascending aorta is very small, and there is almost invariably coarctation of the aorta.

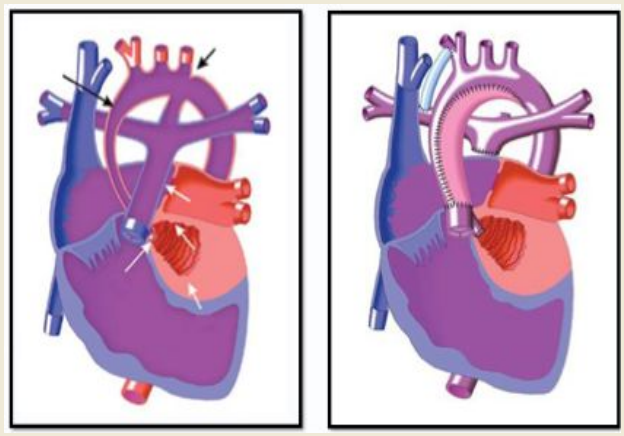
Clinical features:

These children may be detected antenatally at ultrasound screening.
This allows for effective counselling and prevents the child from becoming sick after birth. They may be identified on oxygen saturation screening. If they do present after birth, they are the sickest of all neonates presenting with a duct-dependent systemic circulation. There is no flow through the left side of the heart, so ductal constriction leads to profound acidosis and rapid cardiovascular collapse. There is weakness or absence of all peripheral pulses, in contrast to weak femoral pulses in coarctation of the aorta.

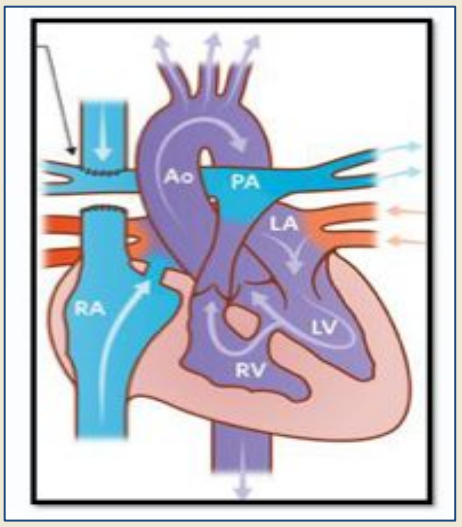
Management:

The management of this condition consists of a difficult neonatal operation called the Norwood procedure. Children who have complex lesions or are small for gestational age undergo hybrid procedures that are a combination of cardiac catheter and surgical operation. This is followed by a further operation (Glenn or hemi-Fontan) at about 6 months of age and again (Fontan) at about 3 years of age.

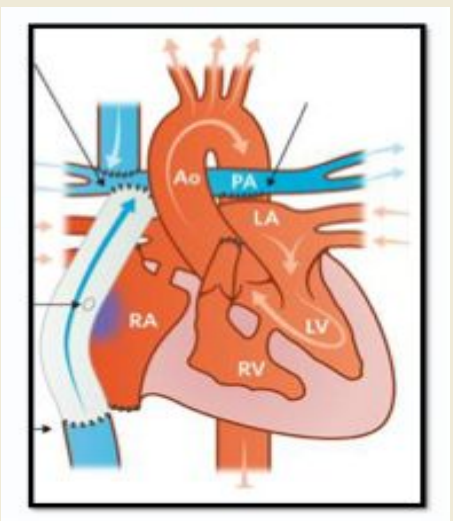
Common Stages Of Univentricular Heart Surgery



Stage 1: Major arch Construction (Norwood) with BT shunt



Stage 2: GLENN



Stage 3: FONTAN

1- Presentation: weak/absence of ALL peripheral pulses (vs only femoral in aortic coarctation)
Management: Norwood procedure

Extra

Eisenmenger syndrome:

If high pulmonary blood flow due to a large left-to-right shunt or common mixing is not treated at an early stage, the pulmonary arteries become thick walled and the resistance to flow increases.

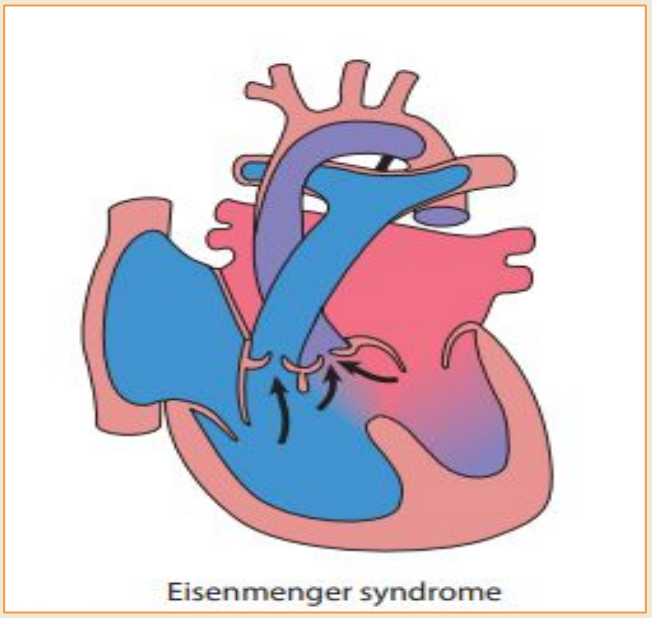
Gradually, those children that survive become less symptomatic as the shunt decreases.

Eventually, at about 10–15 years of age, the shunt reverses and the young person becomes blue, which is **Eisenmenger syndrome**.

This situation is progressive and the adult will die in right heart failure at a variable age, usually in the fourth or fifth decade of life.

Treatment is aimed at prevention of this condition, with early intervention for high pulmonary blood flow.

Transplantation is not readily available (cardiopulmonary transplantation) although medication to palliate pulmonary vascular disease is now available (see the



Eisenmenger syndrome

Tricuspid atresia:

In tricuspid atresia only the left ventricle is effective, the right being small and non-functional.

Clinical features:

There is ‘common mixing’ of systemic and pulmonary venous return in the left atrium.

Presentation is with cyanosis in the newborn period if duct dependent, or the child may be well at birth and become cyanosed or breathless.

Management:

Early palliation (as with all the common mixing complex diseases) is performed to maintain a secure supply of blood to the lungs at low pressure, by: • a Blalock–Taussig shunt insertion (between the subclavian and pulmonary arteries) in children who are severely cyanosed • pulmonary artery banding operation

Completely corrective surgery is not possible with most, as there is often only one effective functioning ventricle

Extra

Aortic stenosis

The aortic valve leaflets are partly fused together, giving a restrictive exit from the left ventricle
. There may be one to three aortic leaflets.
Bi-cuspid aortic valve is a common lesion seen in up to 1% of the population and may be inherited.
Aortic stenosis may not be an isolated lesion. It is often associated with mitral valve stenosis and coarctation of the aorta, and their presence should always be excluded.

Clinical features:
Most present with an asymptomatic murmur.
Those with severe stenosis may present with reduced exercise tolerance, chest pain on exertion, or syncope.
In the neonatal period, those with critical aortic stenosis and a duct-dependent systemic circulation may present with severe heart failure leading to shock.

Physical signs:

- Small volume
- slow rising pulses
- Carotid thrill (always)
- Ejection systolic murmur maximal at the upper right sternal edge radiating to the neck
- Delayed and soft aortic second sound
- Apical ejection click.

Management:
In children, regular clinical and echocardiographic assessment is required in order to assess when to intervene.
Children with symptoms on exercise or who have a high resting pressure gradient (>64 mmHg) across the aortic valve will undergo **balloon valvotomy**.

Balloon dilatation in older children is generally safe and uncomplicated, but in neonates this is much more difficult and dangerous.
Most neonates and children with significant aortic valve stenosis requiring treatment in the first few years of life will eventually require aortic valve replacement.
Early treatment is therefore palliative and directed towards delaying this for as long as possible

Extra

The pulmonary valve leaflets are partly fused together, giving a restrictive exit from the right ventricle.

Clinical features:
Most are asymptomatic.
It is diagnosed clinically. A small number of neonates with critical pulmonary stenosis have a duct-dependent pulmonary circulation and present in the first few days of life with cyanosis.

Physical signs:
- An ejection systolic murmur best heard at the upper left sternal edge; thrill may be present.
- An ejection click best heard at the upper left sternal edge.
- When severe, there is a prominent right ventricular impulse (heave)

Management:

Most children are asymptomatic and when the pressure gradient across the pulmonary valve on Doppler echocardiography becomes markedly increased (> about 64 mmHg), intervention will be required.
Transcatheter balloon dilatation is the treatment of choice in most children.

Questions

1- Alan, a 4-month-old boy, sees his general practitioner for an ear infection. On listening to his chest a heart murmur is heard.
Which one of the following features most suggests that it requires further investigation?
Select one answer only.
A. A thrill
B. Disappearance of murmur on lying flat
C. Murmur maximal at the left sternal edge
D. Sinus arrhythmia
E. Systolic murmur

2-Sunil, a 3-month-old infant, presents with breathlessness and sweating on feeding. He has had several chest infections. You suspect heart failure. Which of the following is most likely to be correct regarding his heart failure?
Select one answer only.
A. Hepatomegaly is not a common feature at this age
B. It is caused by Eisenmenger syndrome
C. It is due to left heart obstruction
D. It is due to a left-to-right shunt
E. It is due to an increase in right-to-left shunt

3- Tariq, who is 6 weeks old, is admitted directly from the cardiology clinic with heart failure. He has a large ventricular septal defect. The cardiologist has recommended treatment with furosemide and spironolactone. His mother wants to know why he has only now started to have problems.
Which of the following statements provides the best explanation?
Select one answer only.
A. At birth and for the first few weeks the ductus arteriosus remained patent and this balanced the flow across the septal defect
B. Pulmonary vascular resistance is increasing and blood is now flowing from right to left
C. The left ventricle is now failing due to its progressive dilatation
D. The pulmonary vascular resistance falls after birth and now flow from left to right across the septal defect is much greater
E. Volume overload results in decreased return to the left ventricle and a reduction in cardiac output related to a reduced end-diastolic filling pressure

4- Which of the following is the most common type of congenital heart disease
Select one answer only.
A. Atrial septal defect
B. Persistent arterial duct
C. Pulmonary stenosis
D. Tetralogy of Fallot
E. Ventricular septal defect

Answers

1- **A.** A thrill
Correct. A thrill is a palpable murmur, i.e. a loud murmur. It always requires further investigation.

2-**D.** It is due to a left-to-right shunt
Correct. After the 1st week of life, progressive heart failure is most likely due to a left-to-right shunt, most often from a ventricular septal defect.

3- **D.** The pulmonary vascular resistance falls after birth and now flow from left to right across the septal defect is much greater
Correct. The pulmonary vascular resistance falls over the first few weeks of life. This increases the flow across the septal defect and leads to progressively worsening heart failure.

4- **E.** Ventricular septal defect
Correct. This is the most common single group of structural congenital heart disease (30%).