

# Perioperative Haemodynamic Goals in Paediatric Cardiac Surgery

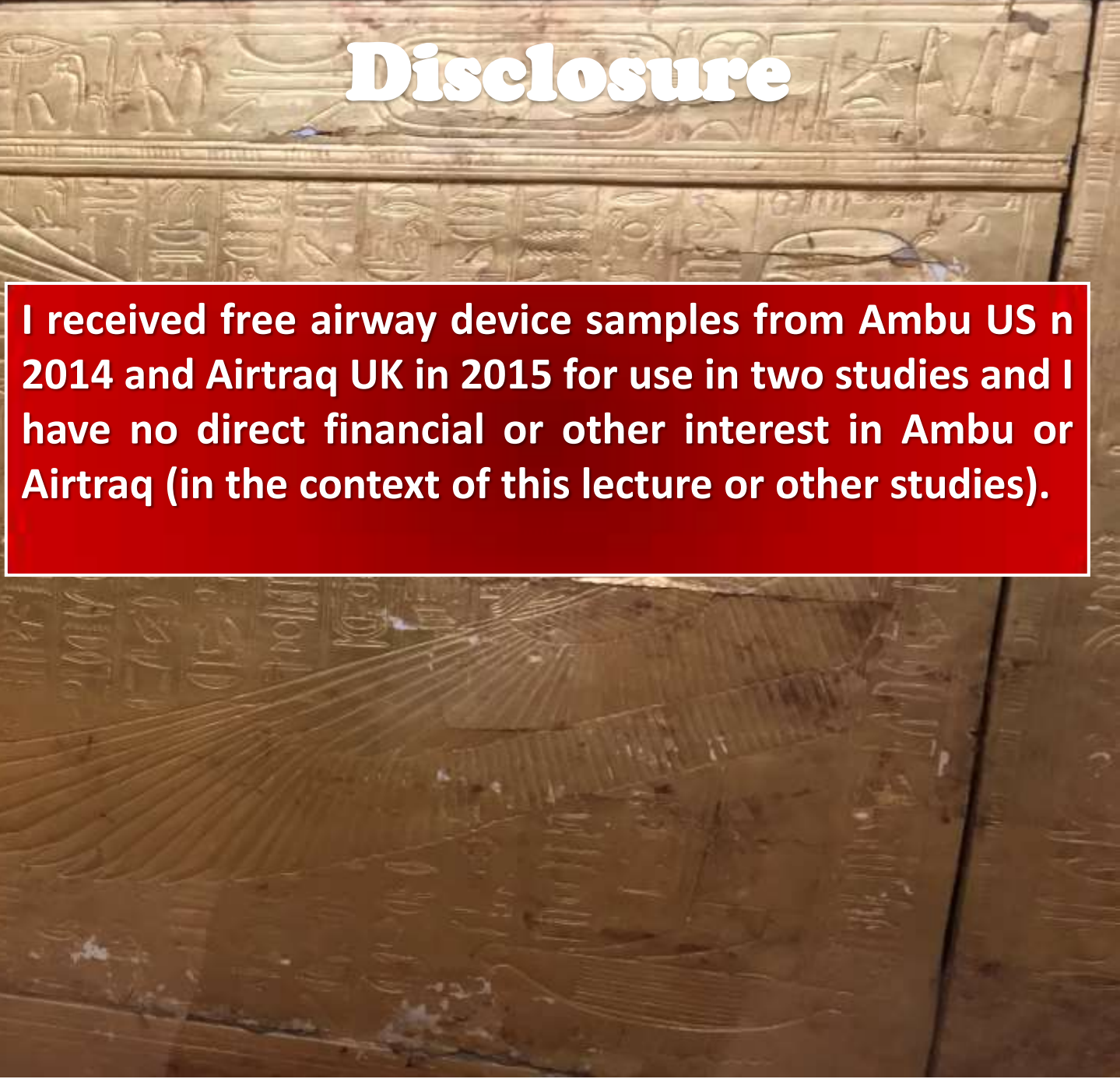
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Associate Professor, Anesthesiology Dept, Imam Abdulrahman Bin Faisal University, Dammam, Saudi Arabia,  
Association of Cardio-Thoracic Anaesthesia and Critical Care (ACTACC), UK.  
Association of Anaesthetists of Great Britain and Ireland (AABGI),  
Egyptian Cardiothoracic Anaesthesia Society (ECTAS), Thoracic Committee.



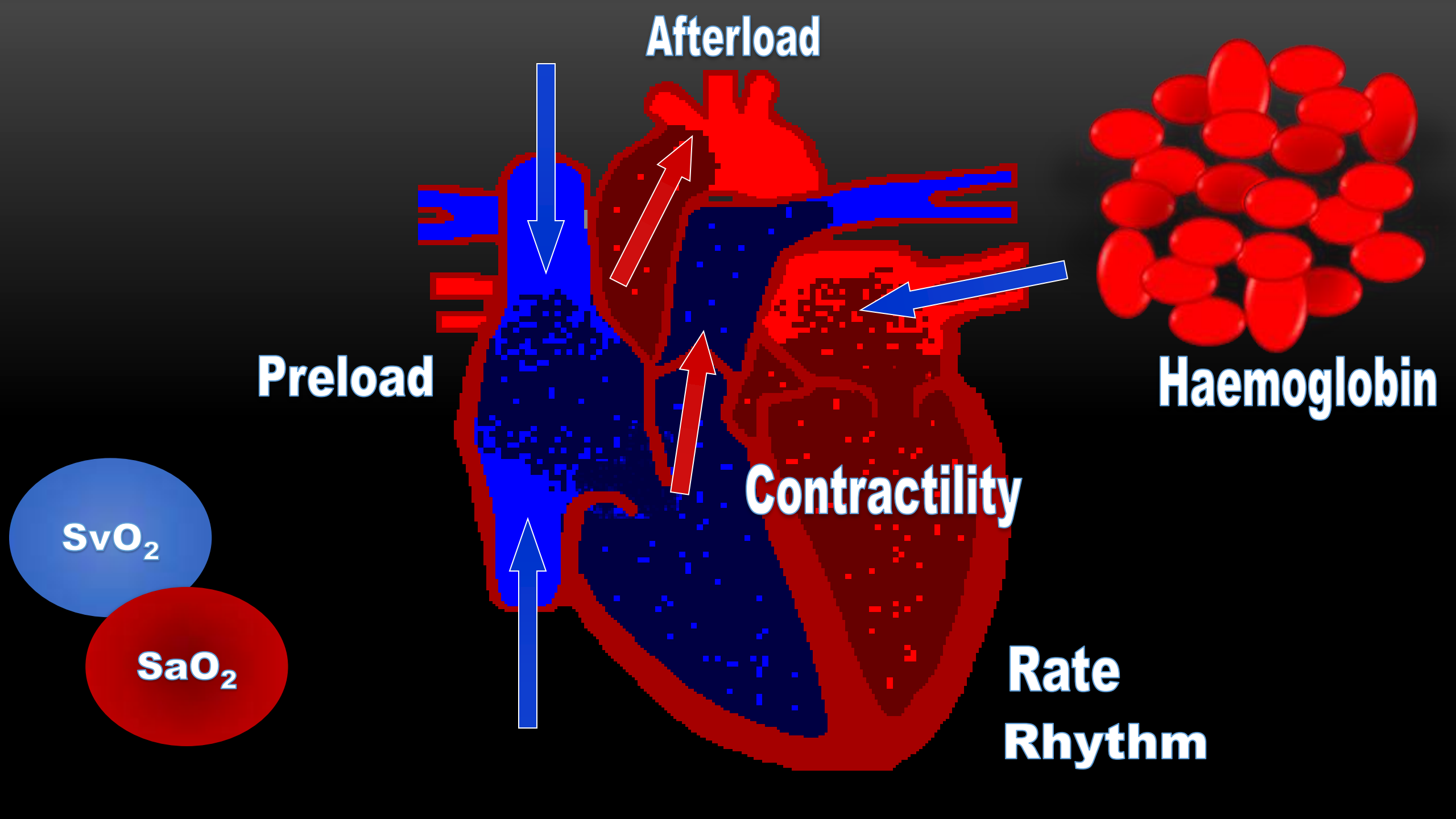


Mernptah statue, Western Bank, Luxor, Egypt, 2016



# Disclosure

I received free airway device samples from Ambu US in 2014 and Airtraq UK in 2015 for use in two studies and I have no direct financial or other interest in Ambu or Airtraq (in the context of this lecture or other studies).







1. Left-to-right shunt lesions.
2. Left-sided obstructive lesions.
3. Right-sided obstructive lesions.
4. Transposition of the great vessels.
5. Single ventricle.
4. Miscellaneous.

## Congenital Cardiac Diseases



**1. Left-to-right shunt lesions.**

**2. Left-sided obstructive lesions.**

**3. Right-sided obstructive lesions.**

**4. Transposition of the great vessels.**

**5. Single ventricle.**

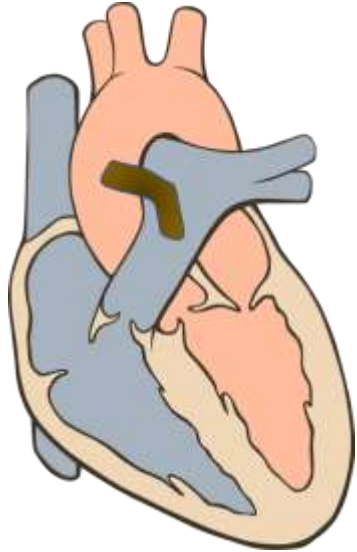
**4. Miscellaneous.**

- Patent ductus arteriosus.
- Atrial septal defects.
- Ventricular septal defects.
- Aortopulmonary window.
- Common atrioventricular canal.
- Double outlet right ventricle.
- Truncus arteriosus.
- Total anomalous pulmonary venous return.
- Partial anomalous pulmonary venous return.

## Congenital Cardiac Diseases

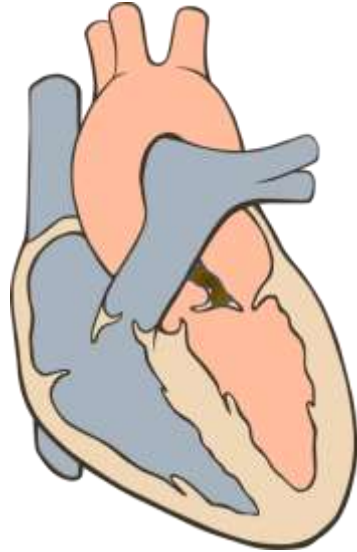
## Left-to-right shunt lesions

**50%** of all lesions



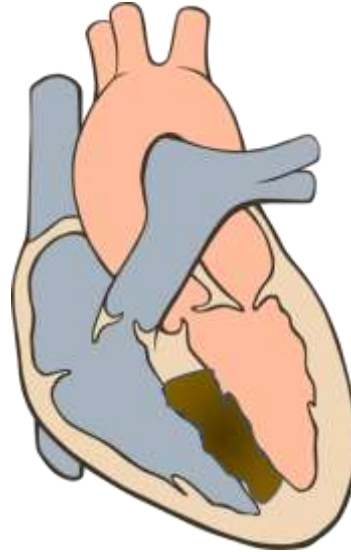
**PDA**

**1 : 2500** to **1 : 5000**  
live births



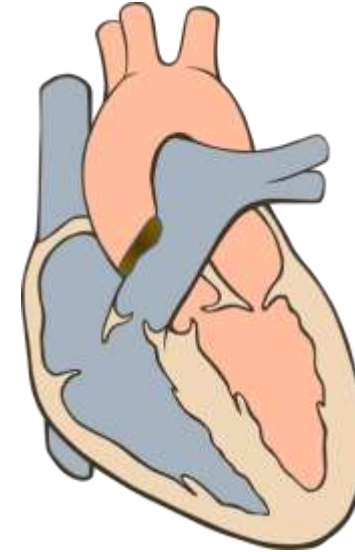
**ASD**

**5%** to **10%** of CHD



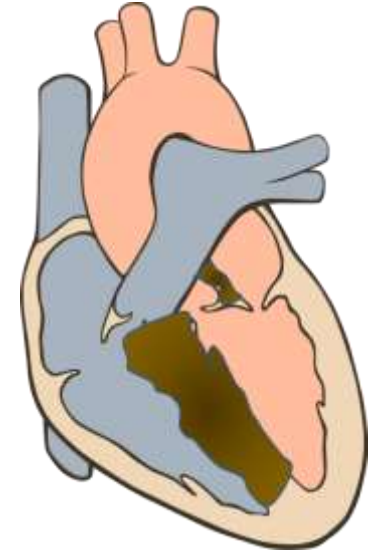
**VSD**

**20%** of CHD  
**2.6-5.7 : 1000** live births



**Aorto-pulmonary  
window**

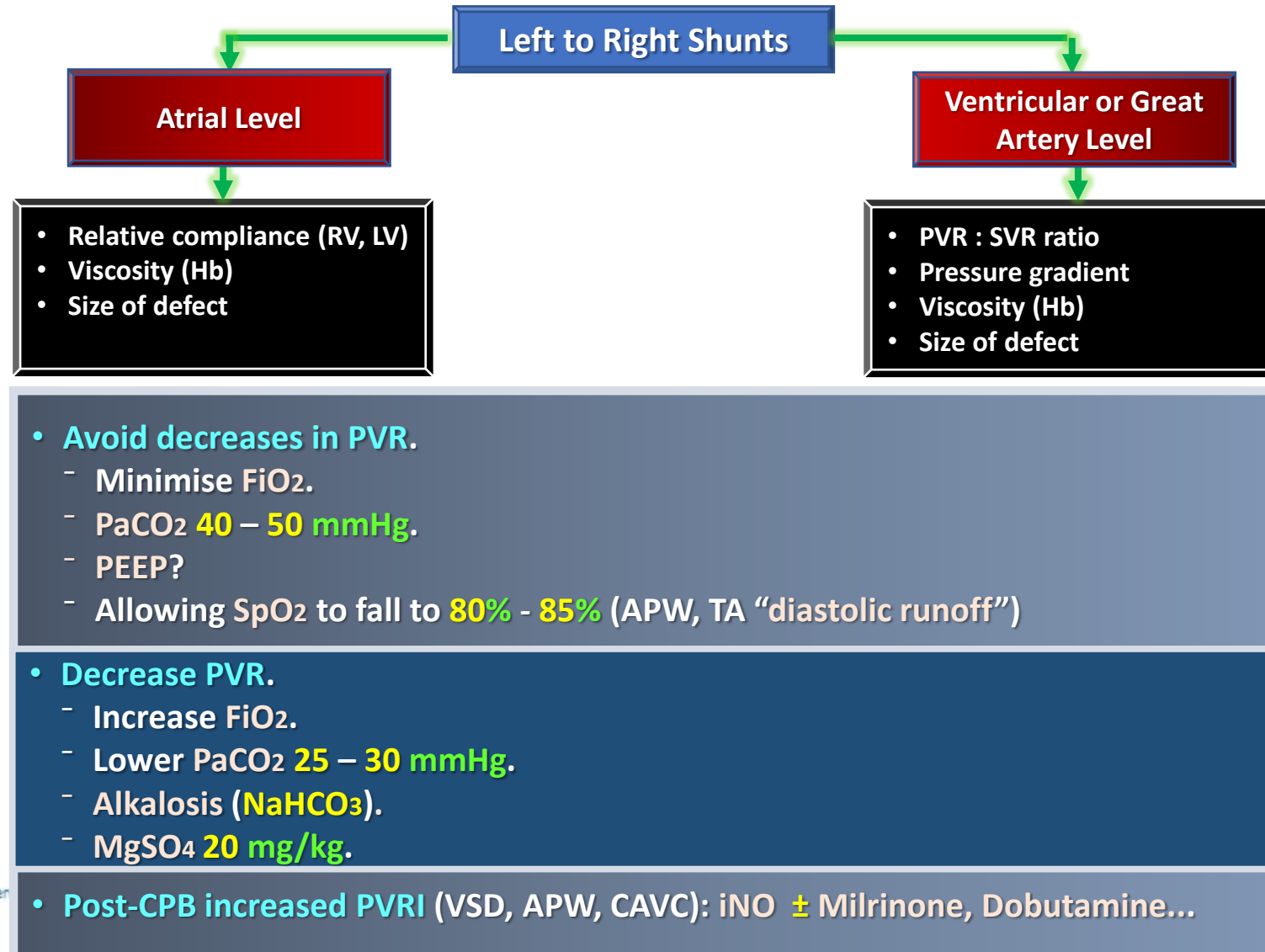
**0.1%** to **0.6%** of CHD



**Common AV  
canal**

**4%** to **5%** of CHD  
**0.19 : 1000** live births





## Double outlet right ventricle (**DORV**)

- Types of DORV are defined based on the relationship of the VSD to the great arteries:

TOF

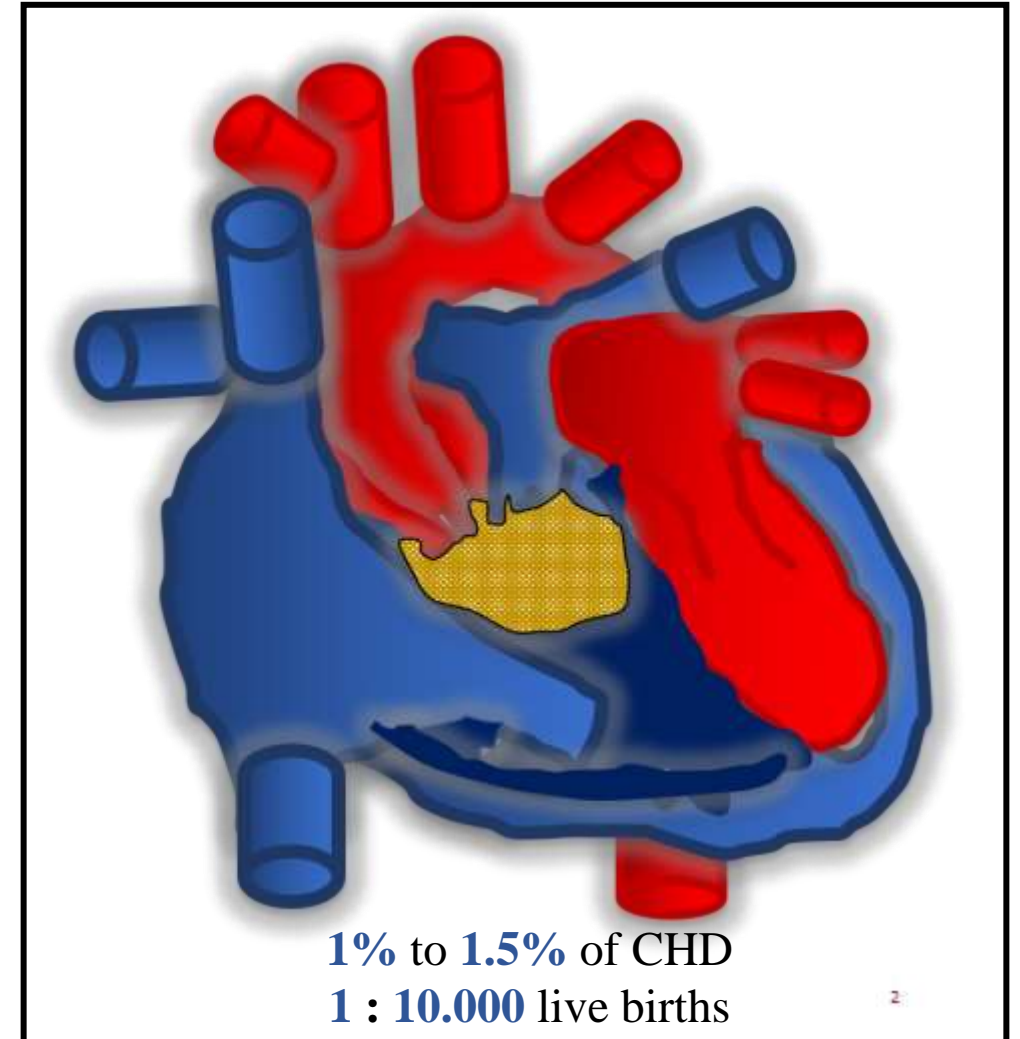
1. **Subaortic VSD** with or without pulmonary stenosis (51%-56%).

2. **Subpulmonary VSD** with or without subaortic stenosis and/or arch obstruction (30%).

TGA-side obstructive

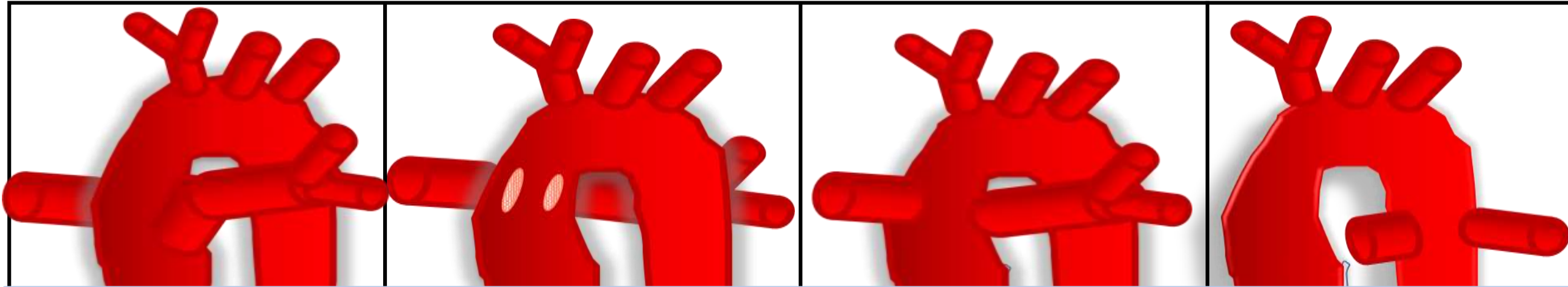
3. **Doubly committed VSD** (3%).

4. **Non-committed VSD** (12%-17%) L-R-Shunt



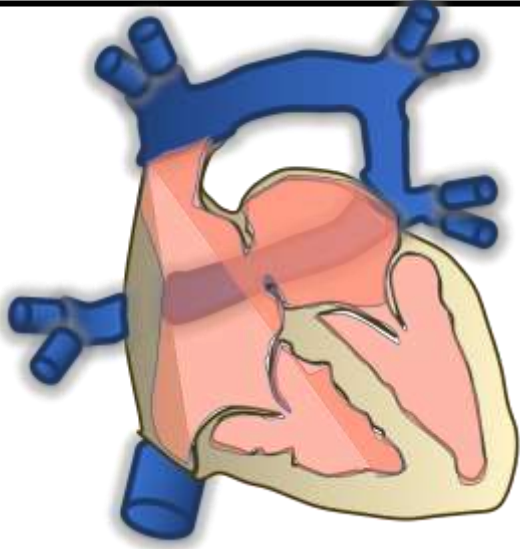
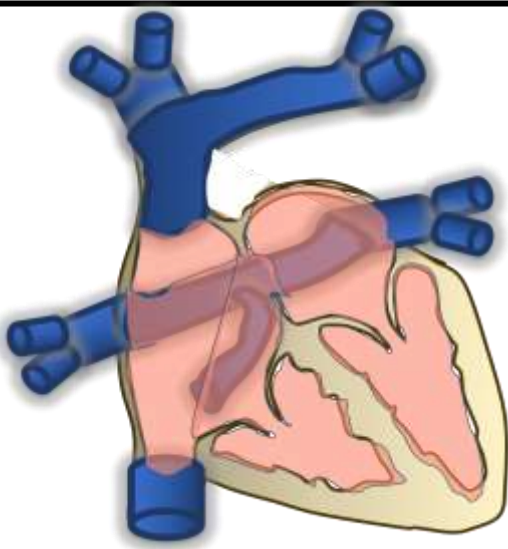
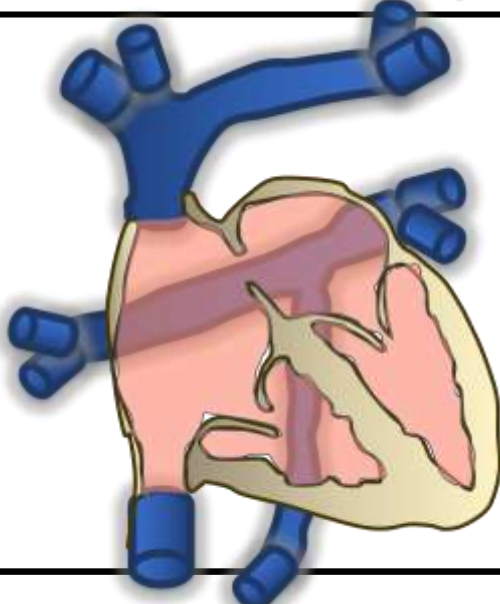
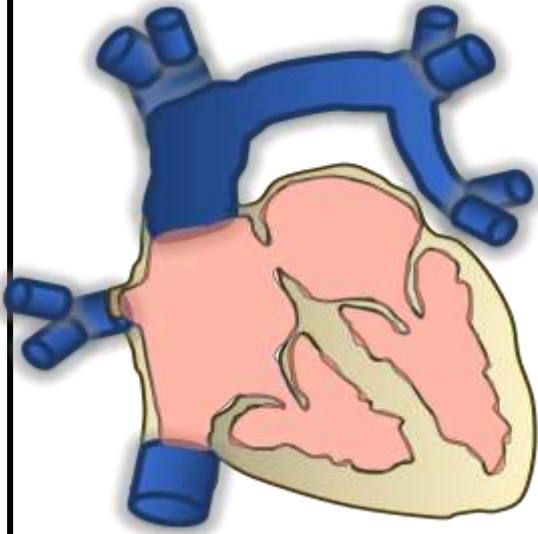


## Truncus Arteriosus 3% of CHD



- Efforts to **balance PVR** and **SVR** to make the ratio of  $Q_p : Q_s$  approach unity are essential.
- Use drugs that **maintain SVR** and **preserve myocardial function** (e.g. fentanyl, midazolam or etomidate).
- **Avoid lowering PVR**, it may lead to **pulmonary overcirculation**, lower DBP, risks for myocardial ischaemia.
- Patients with pulmonary hypertension may require increased  $FiO_2$  to maintain  $SaO_2$  between **80%** and **90%**.
- **DiGeorge syndrome** may require **perioperative calcium infusions** and use of **irradiated blood products**.
- **Inotropic support** is frequently required perioperatively.

## Total Anomalous Pulmonary Venous Return (**TAPVR**) 5% of CHD

			
<b>Supracardiac</b> 45%	<b>Cardiac</b> 25%	<b>Infra-cardiac</b> 25%	<b>Mixed</b> 5%

- TAPVR might be **not obstructed (L-R shunt)** or **obstructed pulmonary venous return (Pulmonary HT)**.



## Partial Anomalous Pulmonary Venous Return (**PAPVR**) 0.6% of CHD

- Avoid use of TEE, which may worsen obstructed pulmonary veins and obstruct non-obstructive veins.
- In obstructed pulmonary venous return and pulmonary hypertension (**Lower PVR**):
  - Avoid **overfilling** (only few millilitres might be disastrous).
  - **FiO2 = 1.0**.
  - **Hyperventilation**.
  - Systemic **alkalinisation**.
  - Deep **sedation** and **paralysis**.
  - Inotropic support  $\pm$  iNO for cyanotic patients.
  - Optimize **heart rate** and **rhythm** (pacing,..)
- In **unobstructed TAPVR** and **PAPVR**, there is a left-to-right shunt of pulmonary venous blood (**like as ASD..**)



## Shunt Lesions

L – R

### Consequences

- Systemic hypoperfusion
- Low *CO*, Hypotension
- LV volume load
- LV dysfunction

### Goals

- Avoid increase in *SVR*
- Avoid decrease in *PVR*
  - Low  $\text{FiO}_2$
  - Avoid hyperventilation



1. Left-to-right shunt lesions.
2. Left-sided obstructive lesions.
3. Right-sided obstructive lesions.
4. Transposition of the great vessels.
5. Single ventricle.
4. Miscellaneous.

## Congenital Cardiac Diseases



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**5. Single ventricle.**

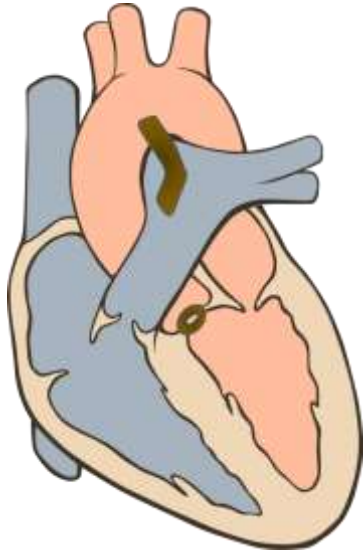
**4. Miscellaneous.**

- Aortic valve stenosis.
- Subvalvar aortic stenosis.
- Supravalvar aortic stenosis.
- Hypertrophic cardiomyopathy.
- Coarctation of the aorta.
- Interrupted aortic arch.
- Shone's anomaly.

**Congenital Cardiac Diseases**



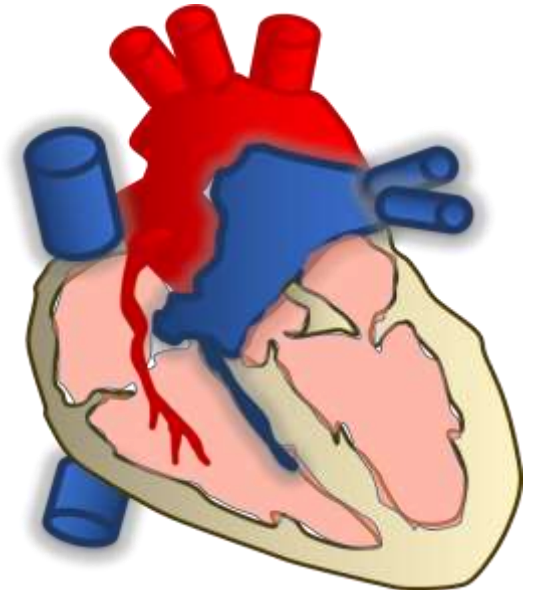
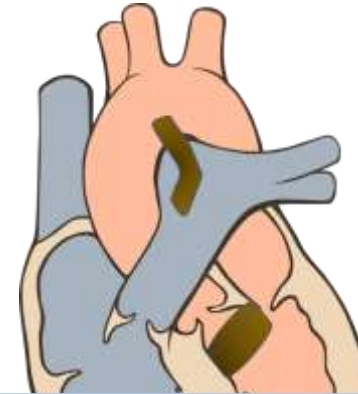
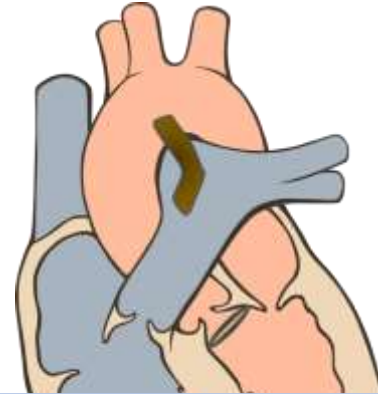
## Left-sided obstructive lesions (**LVOT**)



**AoV stenosis**

6% of CHD

**Endomyocardial fibroelastosis**



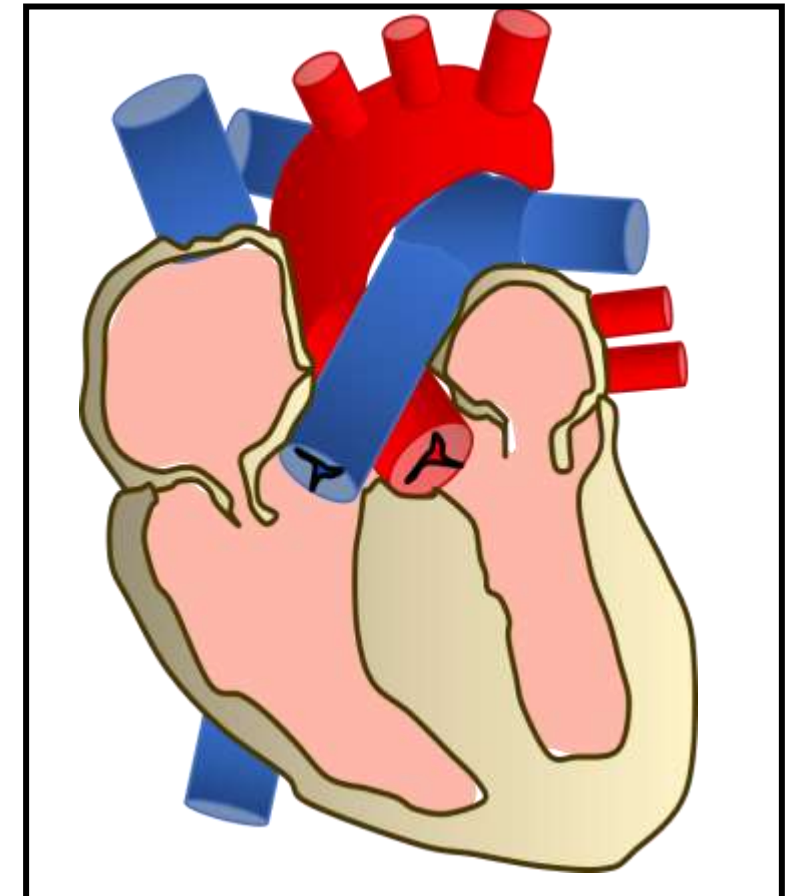
**Supravalvar AS**

Williams' syndrome  
± Coronary ostia

- Adequate *myocardial oxygen supply*.
- Avoid *tachycardia* (diastolic filling).
- Treat *tachyarrhythmia* (atrial contraction).
- Maintain or avoid decreases in *SVR*.
- Maintain an *adequate LV filling*.

## Hypertrophic obstructive cardiomyopathy (**HOCM**)

- MVR.
  - Impaired LV diastolic relaxation.
  - Thick walled, narrowed and tortuous **intramural coronary arteries**.
- Reduce the *LVOT gradient*.
    - Decrease *myocardial contractility*.
    - Maintain *SVR*.
    - Avoid *tachycardia / arrhythmias*.
    - Maintain an *adequate LV filling*.



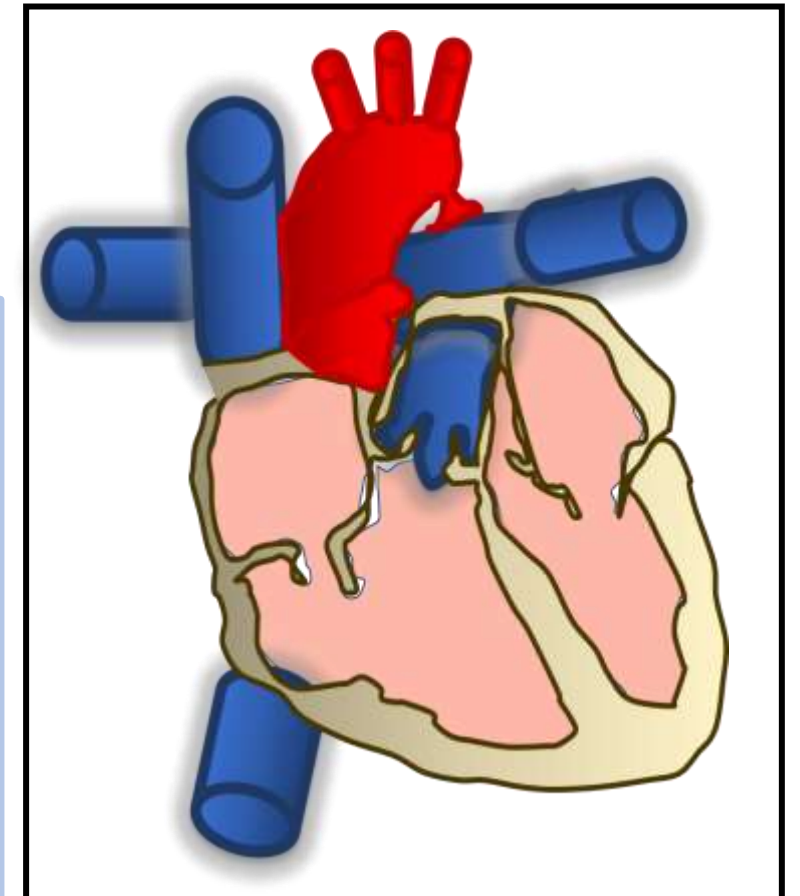
**HOCM**

**1 : 500** live births <sup>2</sup>

## Coarctation of the aorta

- Blood flow to the distal aorta is dependent on a **ductus arteriosus** and on **collateral circulation** later on.

- A **right-sided Art line** in a preductal artery.
- Avoid **hypertension**.
- Aortic **cross-clamping** (vasodilators, inotropes, UOP).
- Risk for spinal cord ischaemia (allow cooling to **35°C**).
- **Vasodilators**, **CCBs** and  **$\beta$ -blocker therapy** are used to treat post-correction hypertension (it might last for **2 w**, **mesenteric arteritis**?)

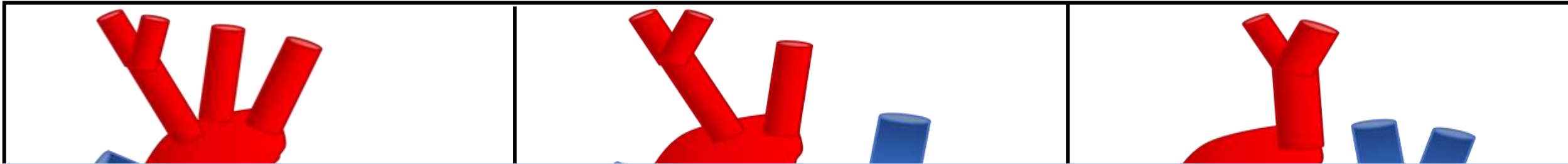


**Aortic Coarct**

**8%** of all CHD



## Interrupted aortic arch (IAA) 1% of CHD



- The use of prostaglandins, inotropic support (dopamine,..), and diuretics is standard.
- A femoral or umbilical Art line is usually placed.
- *Balanced anaesthesia* with *haemodynamic stability*.
- Intraoperative *DHCA* can potentially improve postoperative outcomes (bleeding? Aprotinin, washed RBCs).
- Use of irradiated blood will avoid graft-vs.-host reactions in infants with DiGeorge syndrome.
- Avoid *hypertension*.
- *Inotropes, fluid management, ventilatory adjustment, leaving chest open* should be considered after CPB.



## Shone's Syndrome

1. Levels of stenosis.

2. Location of the dominant lesion.

- Patients with a parachute MV / supra-avalvar mitral ring may show signs of *increased pulmonary congestion*.
- Patients with a high degree of sub aortic stenosis will exhibit *LV hypertrophy*.
- Prostaglandins to maintain the patency of the ductus arteriosus.
- $\beta$ -blockers to improve intracavitary laminar blood flow in patients with *dynamic LVOT*.
- Inotropes and diuretics for *CHF*.
- Milrinone and iNO are used to treat postoperative *pulmonary hypertension*.

Shone's Anomaly

## Obstructive Lesions

Left sided

Consequences

- ↓ Systemic perfusion
- Low CO, hypotension
- LV dysfunction
- Coronary hypoperfusion

Goals

- Avoid decrease in *SVR*
- Avoid decrease in *PVR*
- Maintain *Preload*
- Maintain ductal patency





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5. Single ventricle.
4. Miscellaneous.

## Congenital Cardiac Diseases



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**4. Miscellaneous.**

- Ebstein's anomaly.
- Tetralogy of Fallot.
- Pulmonary stenosis with intact VS.
- Pulmonary atresia with intact VS.
- Pulmonary atresia/VSD/multiple AP collateral arteries.

**Congenital Cardiac Diseases**



## Ebstein's Anomaly

1. Morphology of the TV.

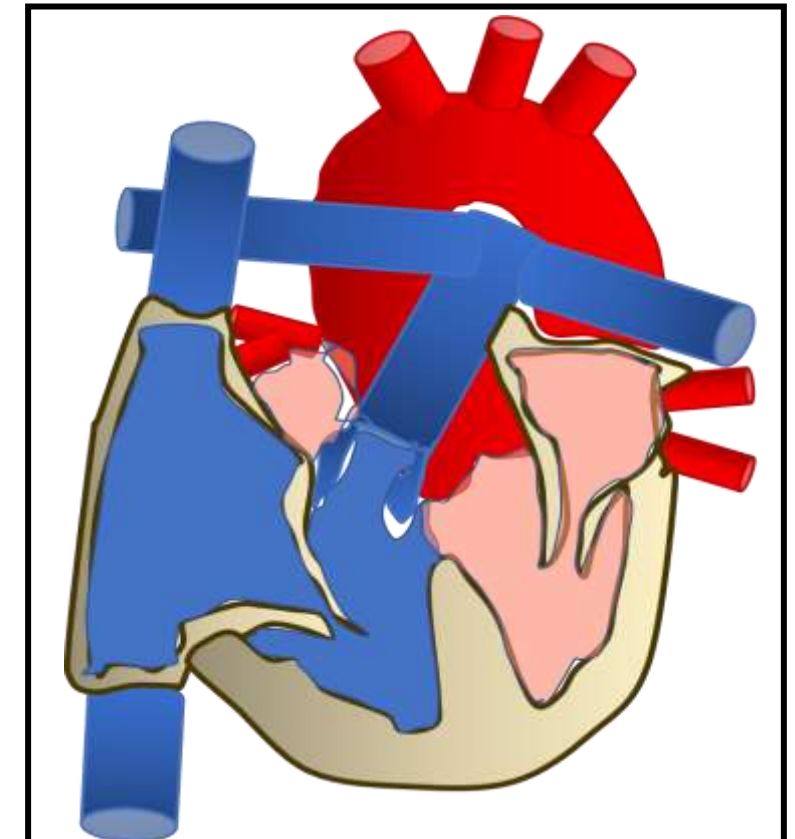
2. Size of the pumping chamber of the pulmonary ventricle.

- rSO<sub>2</sub> and TCD monitoring.
- Antifibrinolytic drugs to minimise pre-and-post CPB bleeding.
- **Right-to-left intracardiac shunt with low CO:**
  - Slow gas induction.
  - Intravenous ketamine + glycopyrrolate or etomidate.
  - Maintain *generous preload*.
  - Milrinone or dobutamine for *RV dysfunction*.
  - Lower *PVR*.
- **Prophylactic antiarrhythmic treatment** (RA and RV pacing, overdrive pacing, lidocaine, amiodarone).



## Tetralogy of Fallot

- A large unrestrictive VSD.
  - RVOT obstruction (dynamic and fixed).
  - An overriding of the aorta above the RVOT.
  - RV hypertrophy.
- VACTERL, DiGeorge syndrome, velocardiofacial syndrome, CHARGE.
  - Abnormal anatomy:
    - left subclavian artery might originate from the PA (shunt?).
    - LAD originates from RCA and crosses the RVOT inferiorly.
    - Left SVC drains into the coronary sinus in the RA (CVC?).
    - Absent PV syndrome (combined PS and incompetence) (↑ PBF, airway).



**Tetralogy of Fallot**

10% of all CHD



## 1. Degree of RVOT obstruction (cyanotic spells (R-L) to pink tets (L-R)).

- **Avoid RVOT obstruction:**
  - Reduce *contractility* ( $\beta$ -blocker).
  - Avoid decreases in *SVR* (ketamine/fentanyl, low MAC of sevoflurane, phenylephrine, norepinephrine).
  - Avoid *Hypovolaemia*.
  - Lower *PVR*.
- **Peripheral shunts** (contralateral Art line, thoracotomy, heparin, ventilate for 12-24 hrs).
- **Central shunts** (FiO2 mimic spontaneous breathing, vasopressors/fluid for  $\downarrow$ DBP and CoPP, heparin).
- **Surgical repair** (RV dysfunction (dopamine + NTG, dobutamine, milrinone), CHB, bleeding).
- **RV : LV** pressure ratio < 0.75 indicate adequate repair, TOE.

10% of all CHD

## Shunt Lesions

R – L

Consequences

- ↓ PBF
- Hypoxaemia
- LV volume load
- LV dysfunction

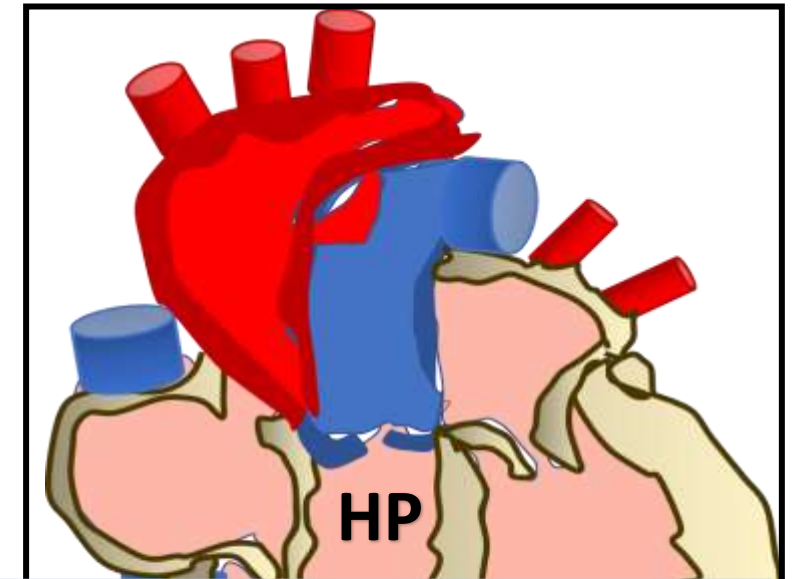
Goals

- Avoid decrease in *SVR*
- Lower *PVR*
  - High  $\text{FiO}_2$
  - Hyperventilation



## Pulmonary stenosis/atresia with intact ventricular septum

- It may be **valvular, subvalvular, or supra**valvular.
- **RV infundibular hypertrophy.**
- **Double chambered RV (HP, LP).**
- Severity depends on the size of **PDA** or **PFO**.
- Frequently associated with **Noonan's syndrome**.



- **Postoperative care:**
  - Optimize **RV filling pressures**.
  - Lower **PVR** (early postoperative use of pulmonary vasodilators)
  - Treat transient post-dilatation **RV dysfunction** secondary to PV insufficiency (**inotropes**). Caution should be exerted in patients with **infundibular hypertrophy**.



## Pulmonary atresia/VSD/ multiple aorto-pulmonary collateral arteries

- Similar to TOF.
- Unifocalization via a thoracotomy (*OLV (BB/DLT), hypoxaemia, haemodynamic instability, airway bleeding*).
- One-stage Unifocalization ( $\pm$  definitive repair) via a median sternotomy or clamshell incision
  - CPB on standby for life-threatening hypoxaemia.
  - Control as many of MAPCAs as possible prior to CPB to prevent cerebral injury (runoff into pulmonary circulation. PA valve conduit).
  - Close VSD? (pulmonary flow study “Lungs are perfused with one CO and mPAP < 30 mm Hg”).
- Post-CPB:
  - Treat RV failure (inotropes, pulmonary vasodilators).
  - Treat Intrapulmonary-bleeding (multiple suture lines, CPB, coagulopathy).
  - Treat lung reperfusion injury (PEEP, ARS, BAL, suctioning).

## Obstructive Lesions

### Right sided

#### Consequences

- ↓ PBF
- Hypoxaemia
- RV Hypertrophy/ dysfunction
- Tricuspid Regurgitation

#### Goals

- Avoid decrease in **SVR**
- Lower/ avoid increase in **PVR**
  - Hyperoxia
  - Mild hyperventilation
- Maintain **Preload**
- Maintain ductal patency





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4. Transposition of the great vessels.
5. Single ventricle.
4. Miscellaneous.

## Congenital Cardiac Diseases



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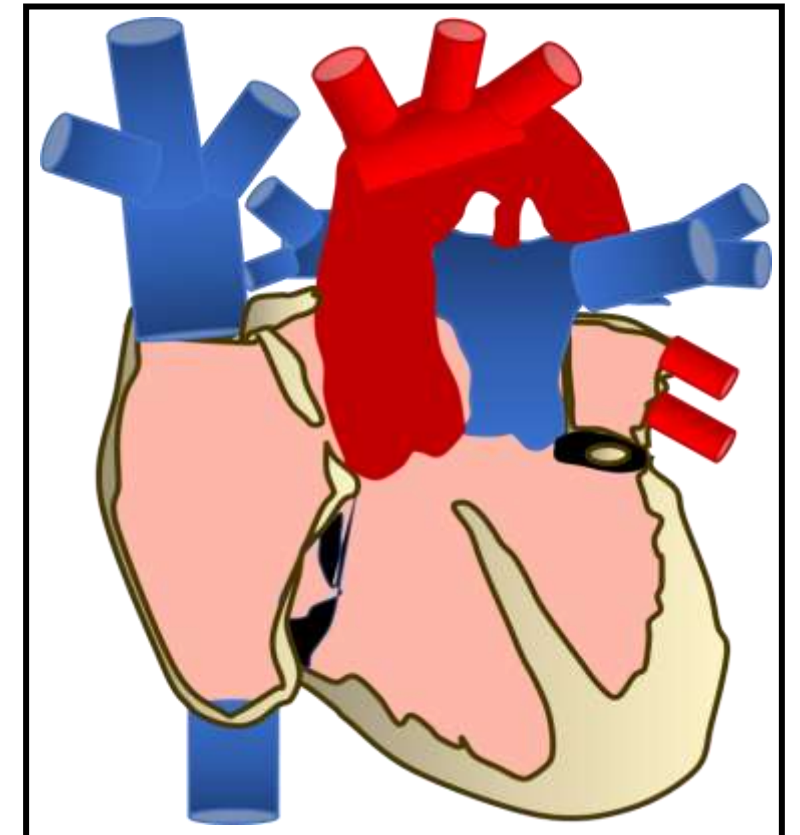
- TGA with intact VS.
- TGA with VSD.
- TGA with VSD + LV outflow tract obstruction.
- Congenitally corrected TGA.

**Congenital Cardiac Diseases**

## Transposition of the great arteries (**TGA**)

5% of CHD

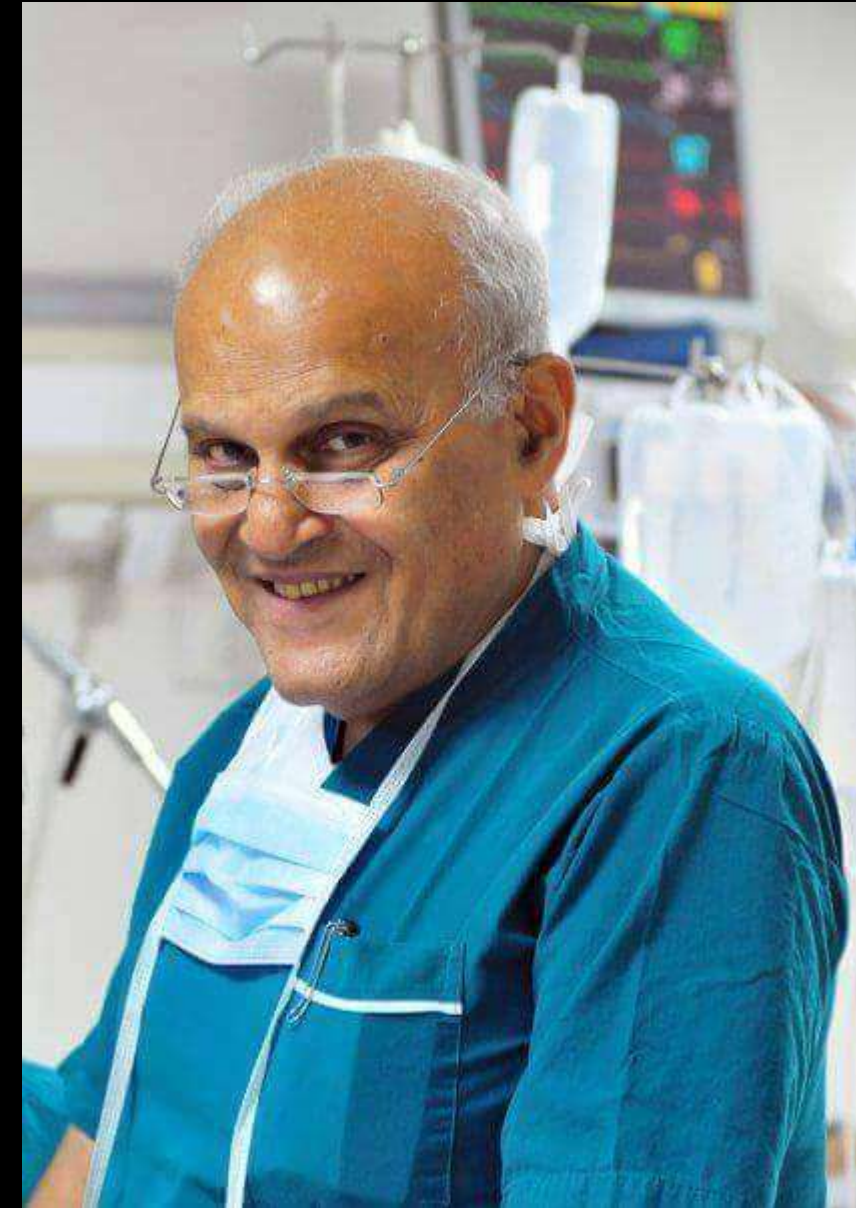
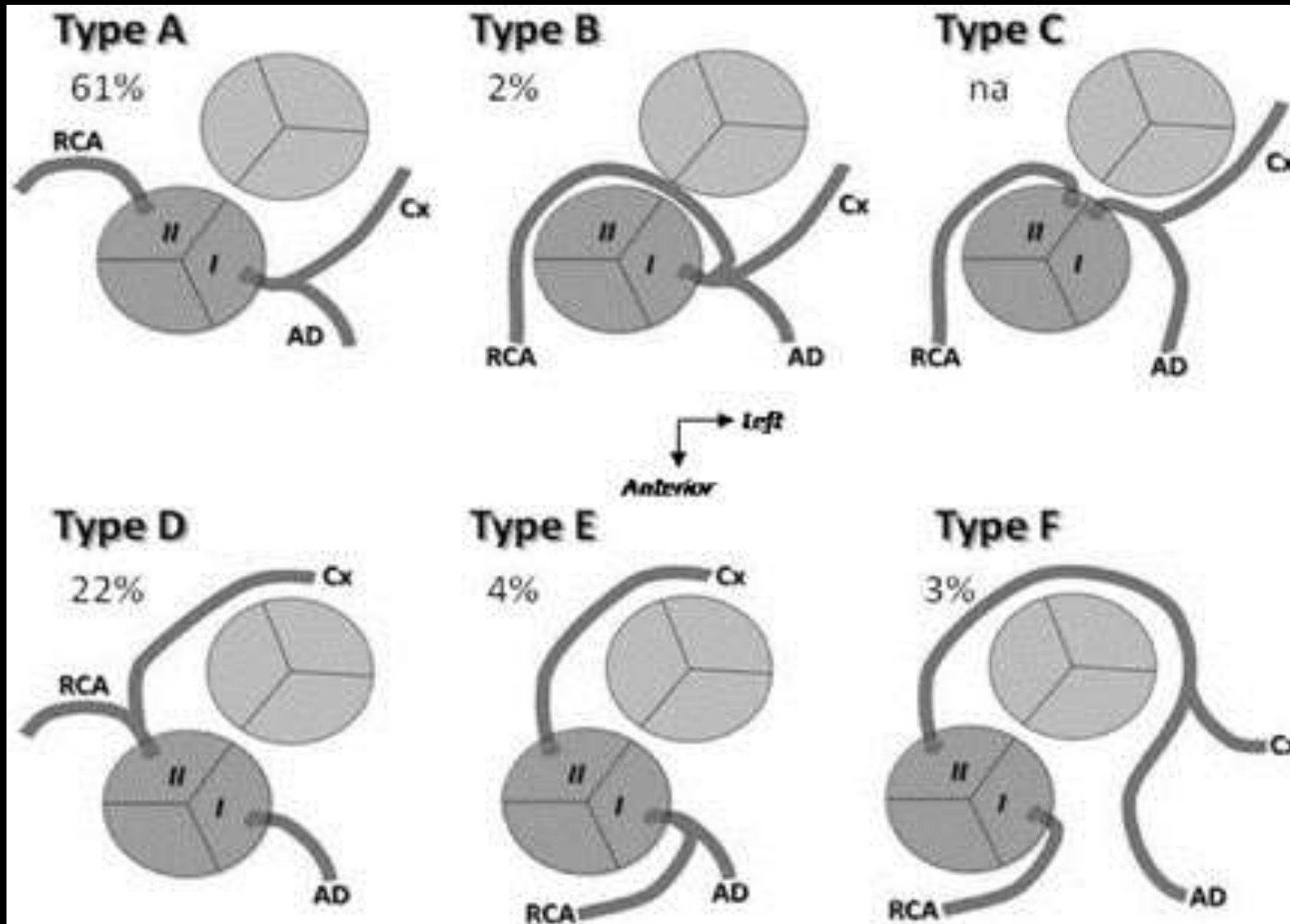
1. TGA with **intact ventricular septum (IVS)** (80%) [PFO, PDA, TV].
2. TGA with **VSD** (10-25%) [right arch, IAA, Ao Coarc].\*
3. TGA with **VSD** and **LVOTO** (30%) [subvalvular, PA stenosis].



**TGA**



# Coronary artery anatomy in T



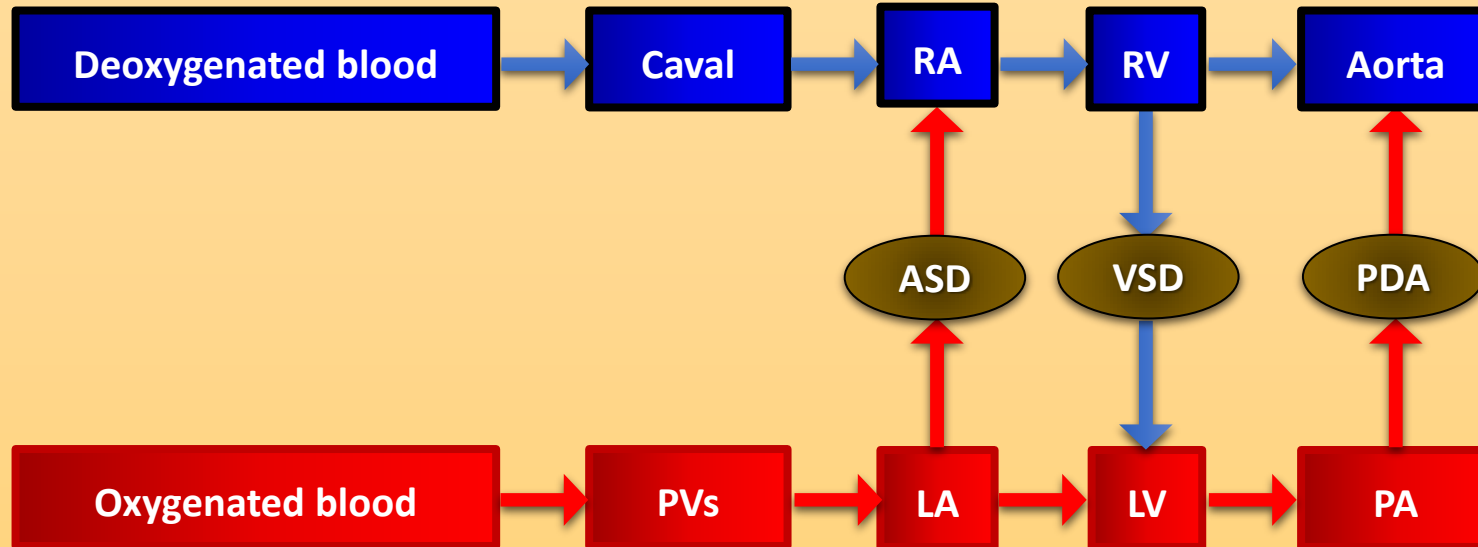
[Thorax](#). 1978 Aug;33(4):418-24.

**Anatomy of the coronary arteries in transposition of the great arteries and methods for their transfer in anatomical correction.**

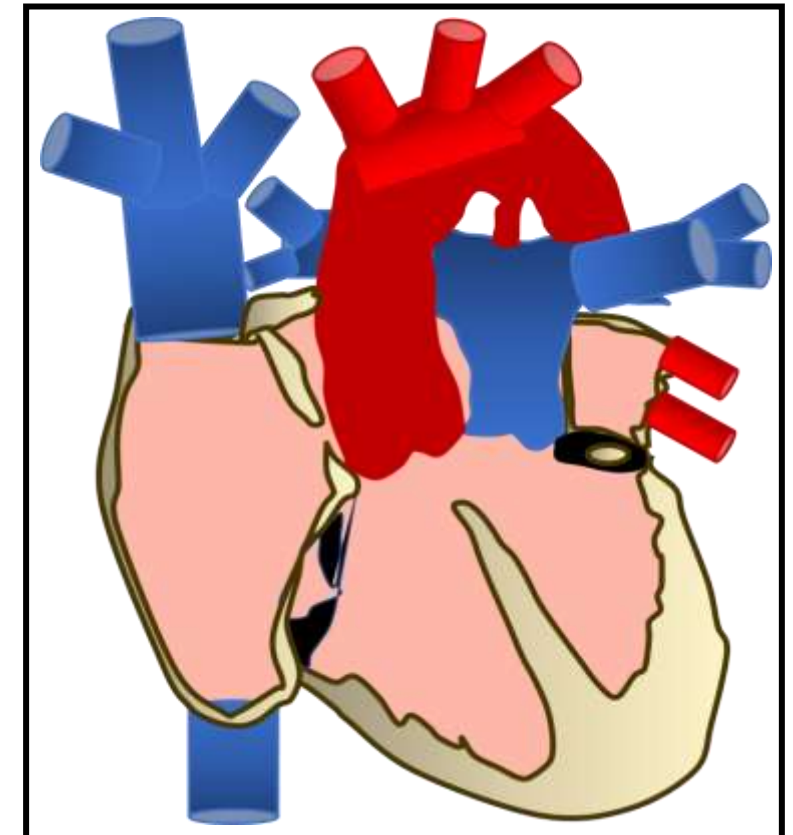
[Yacoub MH](#), [Radley-Smith R](#).

## Transposition of the great arteries (TGA)

5% of CHD



**Bidirectional (mixing) shunt**



**TGA**



## Transposition of the great arteries (**TGA**)

5% of CHD

- TGA with IVS undergoing deconditioning of LV:
  - Avoid *overdistention of LV* (diuretics, venesection (even 3-5 ml)).
  - Avoid increases in *afterload* (50–75 mmHg range).
  - Low LAP (4–6 mmHg).
- Post-CPB:
  - Treat *LV failure* (high LAP, hypoperfusion) (dopamine, NTG, CaCl<sub>2</sub>, pulmonary vasodilators).
  - Diagnose *obstructed translocated coronary arteries* (arrhythmia, ventricular failure, coagulopathy).
  - Sometimes, *leave chest open* (prolonged CBP (lung and myocardial oedema), transfusion, haemodynamic instability).



## Mixing Shunt Lesions

### Consequences

- $Q_p : Q_s$  correlates with **SVR : PVR**
- Varying degree of hypoxaemia
- ↑ Haematocrit
- ↑ Blood viscosity

### Goals

- Optimise  $DO_2$  (*Hct*, *CO*)
- Adjust **PVR / SVR** for optimum
  - $Q_s : Q_p$
  - $SaO_2$
  - $SvO_2$
- High  **$Q_p : Q_s$**  (Lower **PVR**)
  - Low  $FiO_2$
  - Avoid hyperventilation
- Low  **$Q_p : Q_s$**  (Lower **PVR**)
  - High  $FiO_2$
  - Hyperventilation

## Congenitally corrected transposition of the great arteries (**ccTGA**)

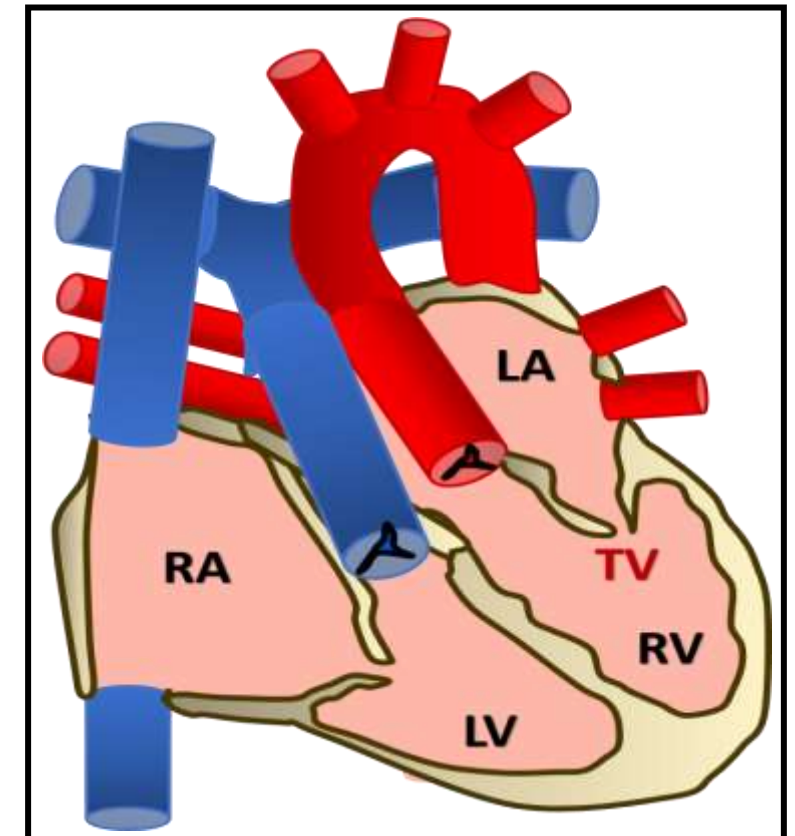


- Systemic RV dysfunction + systemic AV valve regurgitation:

- Monitor *LAP* and *RAP*.
- *Unload RV*.
- Avoid increases in *MAP*.
- *ECMO*.

- Post-CPB:

- Long CPB time (coagulopathy).



ccTGA



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4. Miscellaneous.

## Congenital Cardiac Diseases





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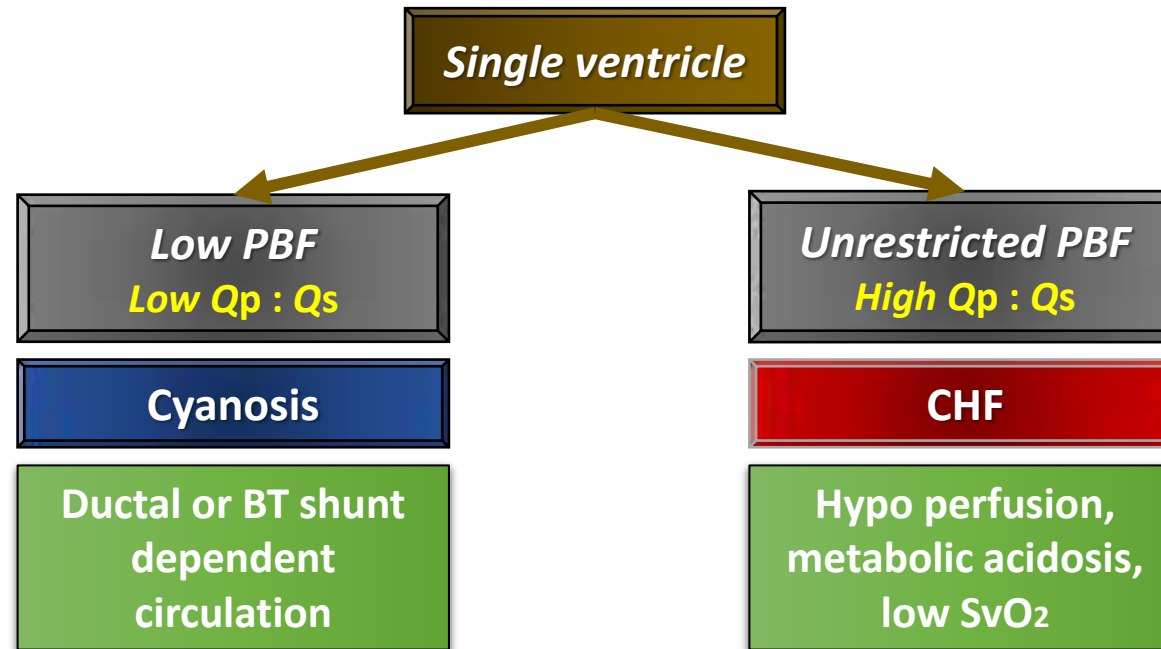
**5. Single ventricle.**

**4. Miscellaneous.**

- Hypoplastic left heart syndrome (**HLHS**).
- Tricuspid atresia.
- Staged approach to Fontan.
- Fontan circulation.
- Fontan circulation and non-cardiac surgery.

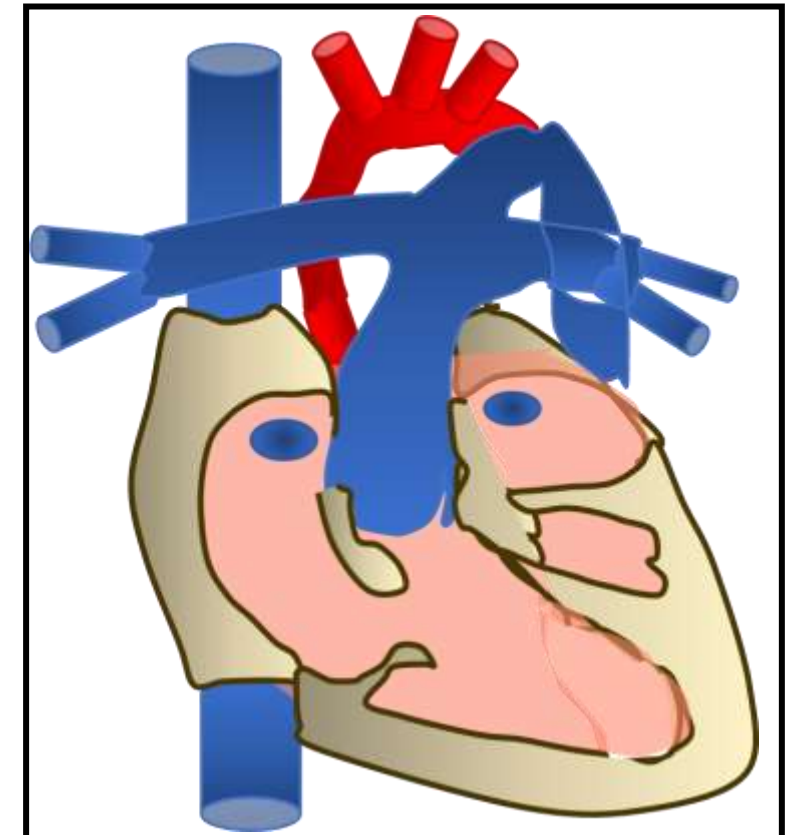
**Congenital Cardiac Diseases**

## Single-ventricle lesions



## Hypoplastic left heart syndrome (**HLHS**)

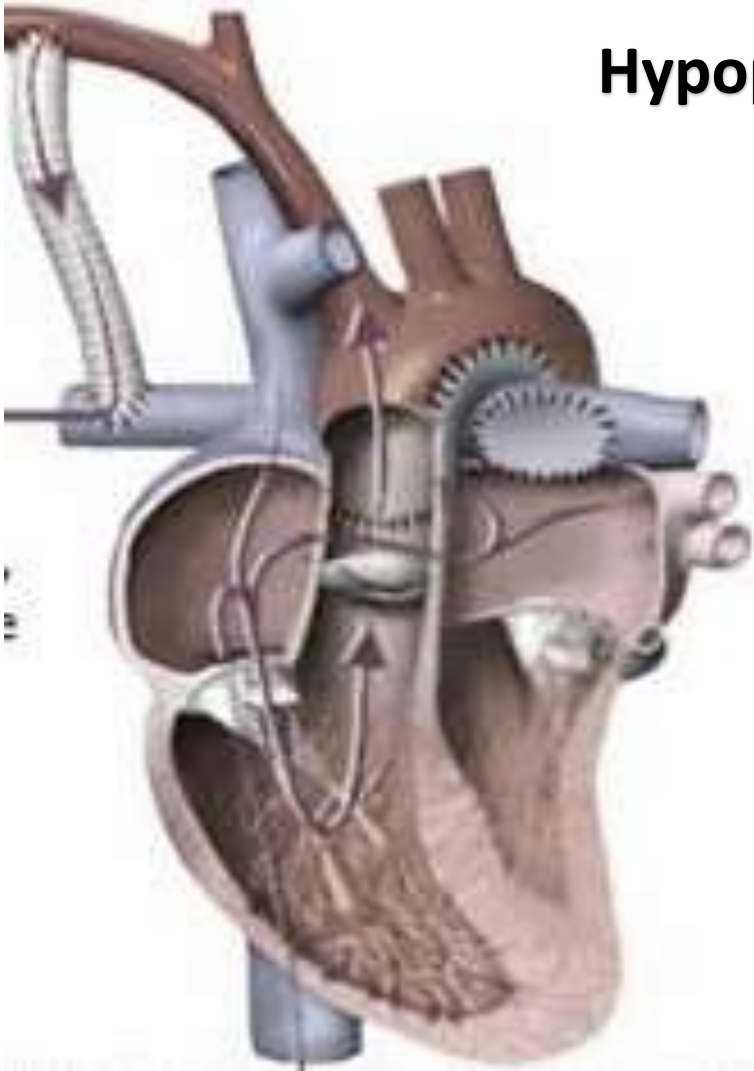
- Non-functional LV.
- **Pulmonary venous return (PFO, ASD, TAPVA).**
- **RA:** mixing of **systemic** and **pulmonary** circulation.
- **RV → PA → PA branches.**
- **RV → PA → PDA → Descending Aorta → Systemic circulation.**
- **RV → PDA → (Retrograde) three major arteries and Coronaries.**



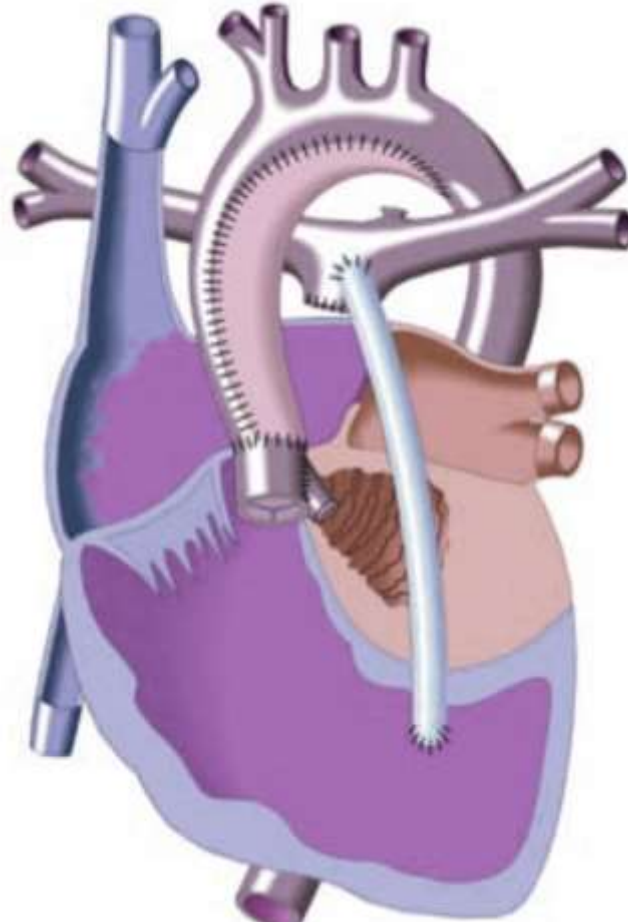
**HLHS**



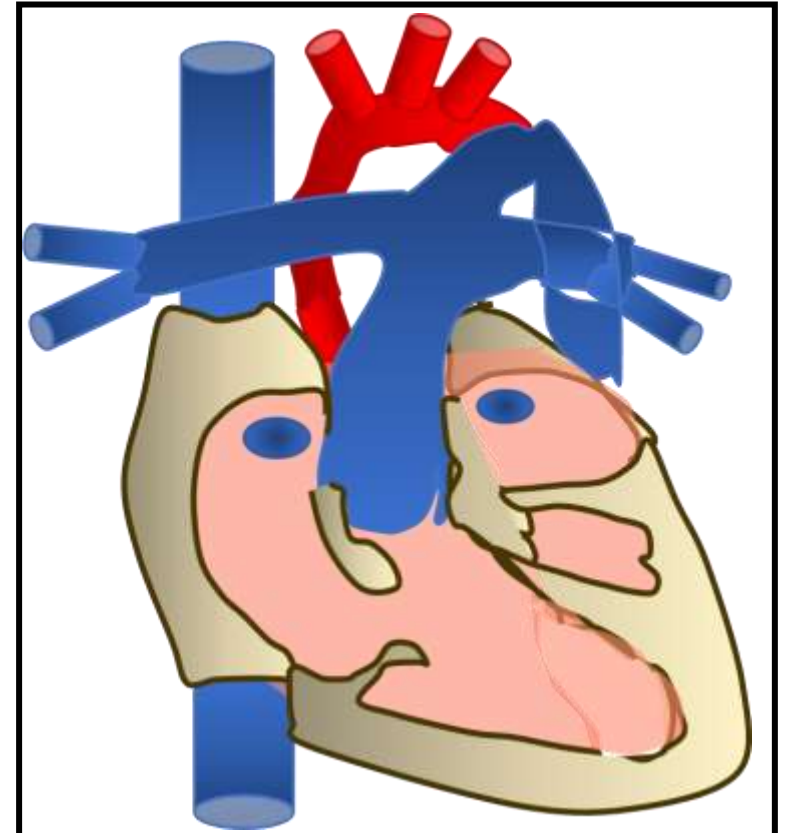
## Hypoplastic left heart syndrome (**HLHS**)



Norwood



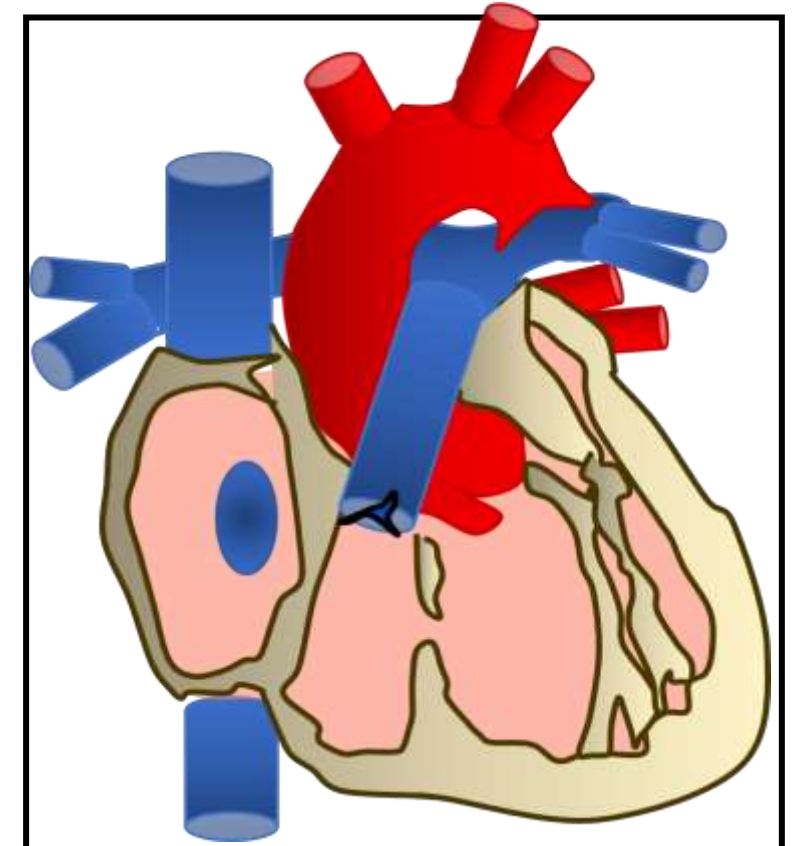
Sano



HLHS

## Tricuspid atresia (TA)

- Agenesis of tricuspid valve.
- Single RV.
- Ductal-dependent PBF (**cyanosis**)
- Those variant with **VSD** may have **adequate** or **excessive** PBF.



Tricuspid atresia

- Doppler Aortic flow
- SvO<sub>2</sub>
- SaO<sub>2</sub>

**Qp: Qs**

**High**  
**Low Qs, SvO<sub>2</sub>**

Commonest    Wide Pulse Pressure  
Low DBP (runoff)

- Adequate preload
- Hb 14-15 g/dl
- Increase **PVR**
  - Reduce alveolar PO<sub>2</sub>
  - Low FiO<sub>2</sub>
  - Increase alveolar PCO<sub>2</sub>
  - Inotropes, vasodilators

**Low**  
**Low Qp**

Narrow Pulse Pressure  
Normal to high DBP

- Adequate preload
- Hb 14-15 g/dl
- Technical (larger or proximal shunt)
- Increase **SVR** (hypoperfusion)
- Lower **PVR**
  - High FiO<sub>2</sub> (0.6-1.0)
  - Low to normal PaCO<sub>2</sub>
  - Normothermia, Alkali, iNO

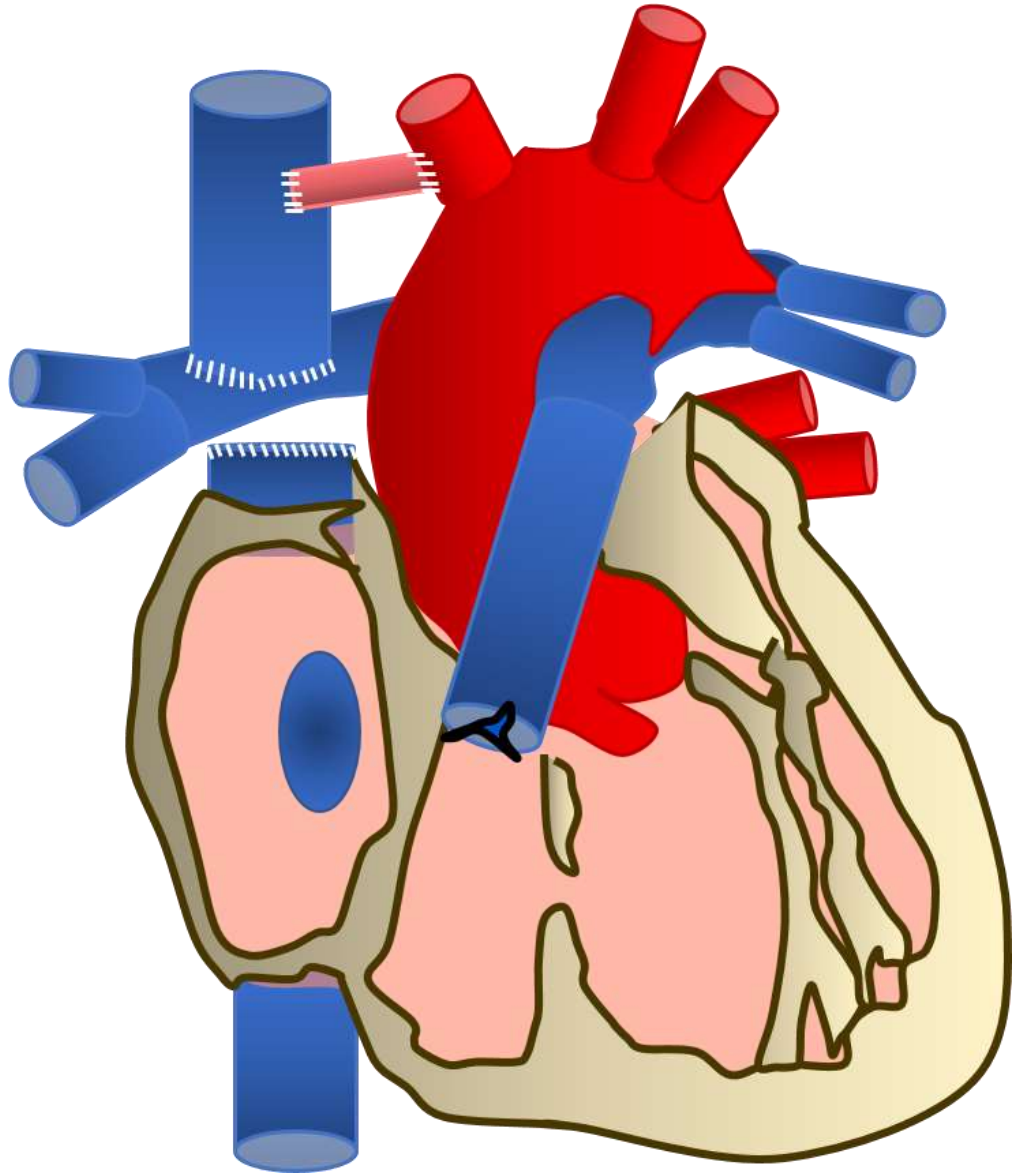
**Hypoxaemia**  
**Low SvpO<sub>2</sub> (30%)**

- Hb 14-15 g/dl
- **ARS**
- **PEEP**

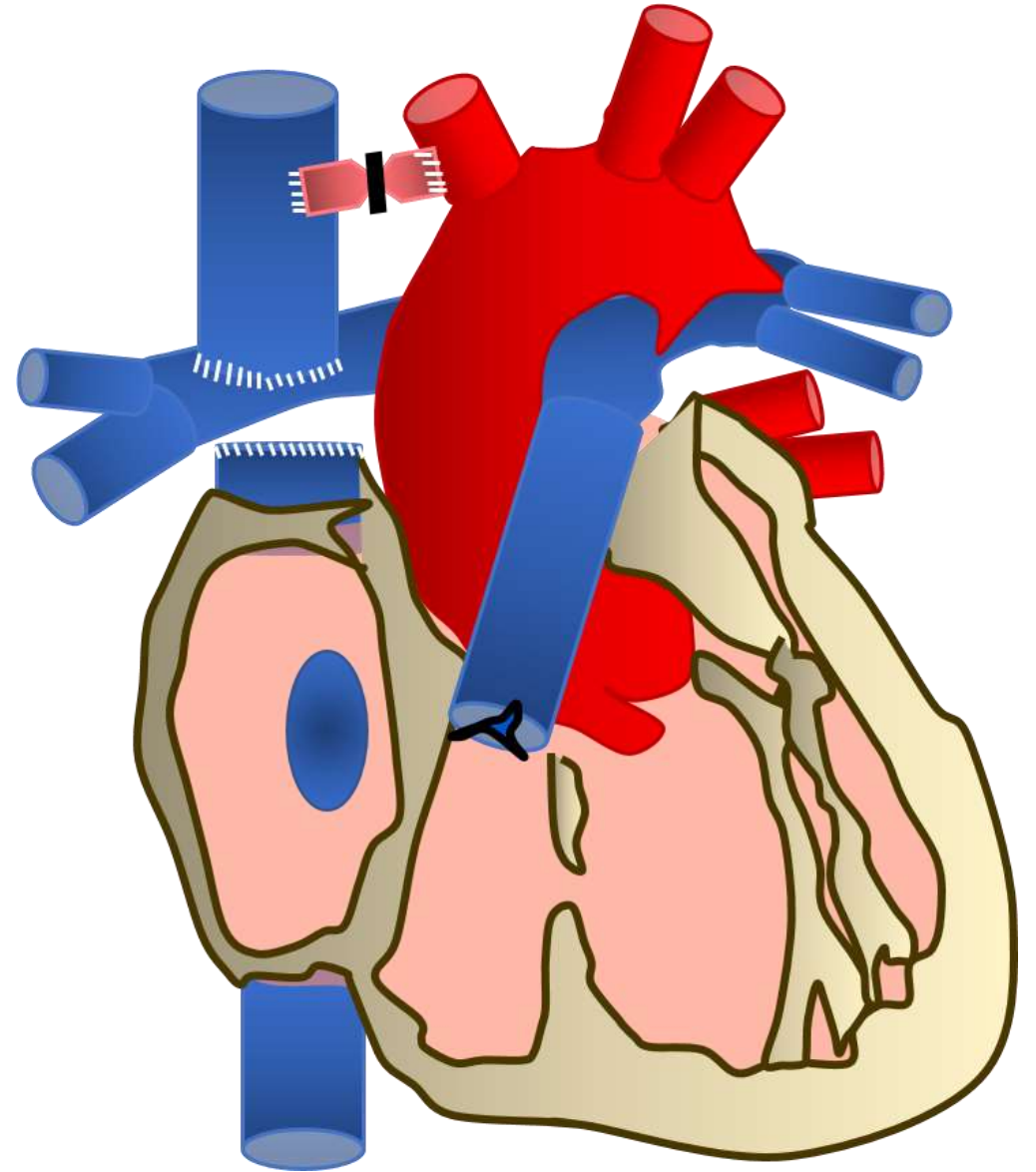
**• Post-CPB:**

- ARS, correct acidosis, CaCl<sub>2</sub>.
- Dopamine, milrinone (for high SVR) (rule out jeopardising coronary flow)
- Think about (residual aortic arch obstruction, valve obstruction / regurgitation)
- Haemostasis (fresh whole blood, platelets, fibrinogen, antifibrinolytic).





Glenn central shunt



Hemi-Fontan shunt



## Fontan Completion

Thorax. 1971 May; 26(3): 240–248.

### **Surgical repair of tricuspid atresia**

F. Fontan and E. Baudet

- **Preoperative:**

- Timing: **9-12 months**. shorter (restoration of optimum ventricular compliance), longer (VAE, hypoxaemia)
- Function of the *vital organ systems* and *coagulation*.

- **Intraoperative:**

- *Balanced anaesthesia, high dose fentanyl* (AV valve or ventricular dysfunction)
- *Art line* (location), *NIBP* (four extremities), *CVC* (thrombosis).
- *CPB*: DHCA? MUF

- **Post-CPB: Fontan:** improves  $\text{SaO}_2$ , Low CO.

- Maintain adequate *intravascular volume*.
- *Myocardial dysfunction*: Low dose of dopamine, milrinone (*diastolic dysfunction* or *valve regurgitation*).
- *Minimize the impediments to PBF* (suctioning, ARS, PVR, low-normal  $\text{PaCO}_2$ ,  $\pm$  PEEP < **6 cmH<sub>2</sub>O**).
- *Bleeding*: (fresh whole blood).
- *Pacing* (junctional rhythm).
- *Hypoxaemia* indicates some communication from the systemic venous system to RA (*fenestration*).



- **Points to be considered:**

- Single ventricle is no longer working with  $Q_p + Q_s$ .
- CO is not dependent on  $PBF$ .
- $\uparrow$  Venous pressure (risk of *surgical bleeding*).
- A well-functioning Fontan warm, well perfused, and acyanotic.
- All air must be evacuated meticulously.

- **Intraoperative:**

- Monitoring: Non-invasive (*superficial*), Invasive (*major*) (*location*), TEE, CVC ( $\sim$ PAP) (*risks*),  $rSO_2$ .
- Transcutaneous defibrillator/pacing pads.
- Careful titration of anxiolytics.
- Low CO (*hypotension, high CVP, lactic acidosis*).
  - ✓ Maintain  $CVP \leq 16$  mmHg.
  - ✓ Lower PVR ( $\uparrow$   $FiO_2$ ,  $PaCO_2 = 30$  mmHg, correct atelectasis, limit Ppk, normothermia, Milrinone, iNO).
  - ✓ Treat *ventricular dysfunction* and *arrhythmia*.
- Balanced anaesthesia (*etomidate*) high dose fentanyl (AV valve or ventricular dysfunction)
- Epidural analgesia/anaesthesia (*titrate LA, dural puncture* or SA).



1. Left-to-right shunt lesions.
2. Left-sided obstructive lesions.
3. Right-sided obstructive lesions.
4. Transposition of the great vessels.
5. Single ventricle.
4. Miscellaneous.

## Congenital Cardiac Diseases



**1. Left-to-right shunt lesions.**

**2. Left-sided obstructive lesions.**

**3. Right-sided obstructive lesions.**

**4. Transposition of the great vessels.**

**5. Single ventricle.**

**4. Miscellaneous.**

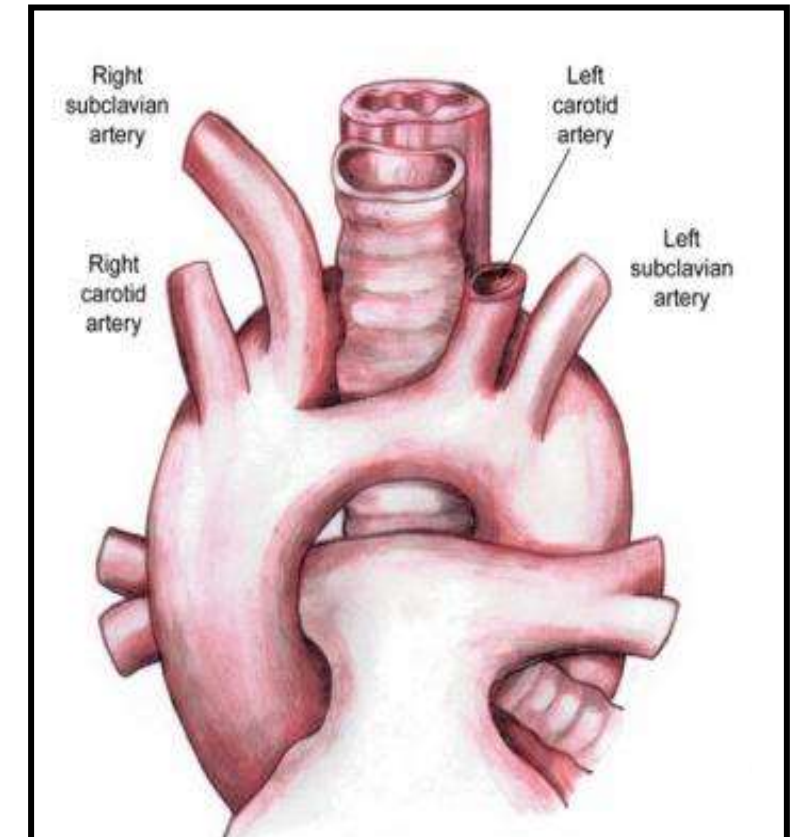
- **Vascular rings.**
- **Anomalies of the coronary arteries.**
- **Mitral regurgitation.**
- **Pericardial effusion and tamponade.**

**Congenital Cardiac Diseases**



## Vascular rings

- Large **vascular access**, **Art line**.
- **Inhalational induction** with maintenance of **spontaneous ventilation**.
- Left thoracotomy/VATS, **Smaller ETT or OLV**.



### Vascular rings

1% of all CHD



## Anomalies of the coronary arteries

- Large Infants with **ALCAPA** are often critically ill with little reserve.
- Minimise **coronary stealing** (**mild** ↑**PVR** (normocapnia, low  $\text{FiO}_2$ )).
- Smooth and gradual **induction**, fast **airway securing**, **volatile**?
- Titrate **fluids / inotropes**.
- **Post-CPB: inotropic, inodilator, coronary/systemic vasodilators, LVAD.**

- **LCA** from **right aortic sinus**
- **RCA** and **LCA** from **PA** (fatal)
- **RCA** from **PA** (ARCAPA)
- **LCA** from **PA** (ALCAPA)

### Anomalies of thr CA



## Mitral regurgitation

- Afterload reduction.
- Adequate preload and contractility.
- High–normal HR.
- Volatile anaesthetics (but not halothane).





## Pericardial effusion and tamponade

- **Maintain afterload. Etomidate or ketamine.**
- **High–normal preload**
- **Maintain contractility.**
- **High–normal HR.**
- **Pre-induction drainage.**
- **Keep the patient breathing spontaneously or gently assisted, if possible**



1. Left-to-right shunt lesions.
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4. Miscellaneous.

## Congenital Cardiac Diseases





The image shows a close-up of an ancient Egyptian stone relief. The central figure is a winged deity, likely Ptahhotep, shown in profile facing right. He has large, feathered wings and is wearing a kilt. The background is filled with hieroglyphs. A white text overlay is positioned across the center of the relief.

Thank you Teşekkür ederim.