

Congenital Heart Disease in Children

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➤ **CHD is the most commonest congenital malformation**

➤ **Incidence of congenital heart disease:**

– worldwide: 6-10 per 1,000*

– HK: 6.35 per 1,000**

*** at least 36% were considered as major cardiac malformations**

** Congenital Heart Disease in 56,109 Births - Incidence and Natural History*

Mitchell SC, et al

Circulation 1971; 43:323-332

*** Echocardiography as a tool for determining the incidence of congenital heart disease in newborn babies : a pilot study in Hong Kong*

Sung RY, et al

Int J Cardiol 1991; 30:43-47

Aetiology of Congenital Heart Disease

- Majority remains unknown and most cases are sporadic
- Chromosomal abnormalities
 - Down syndrome - atrioventricular septal defect
 - 22q11.2 deletion syndrome (DiGeorge syndrome)
- Familial
- Environmental
 - Congenital rubella infection - Patent ductus arteriosus
 - Maternal collagen disease - Complete congenital heart block

Presentation of heart disease in newborn

- Asymptomatic heart murmur
- Respiratory distress (heart failure)
- Cyanosis
- Shock

Differential diagnosis of asymptomatic murmur

- Ventricular septal defect
- Fallot's tetralogy
- Pulmonary stenosis
- Patent ductus arteriosus
- Aortic stenosis
- Atrial septal defect
- Innocent murmur

Differential diagnosis of respiratory distress

- Large left to right shunt (VSD, PDA, AP window)
- Hypoplastic left heart syndrome
- Aortic arch interruption
- Coarctation of aorta
- Complex defect

Differential diagnosis of cyanosis

- Fallot's tetralogy
- Transposition of great arteries
- Pulmonary atresia
- Tricuspid atresia
- TAPVD (obstructed)
- Complex heart lesion

Differential diagnosis of shock

- Hypoplastic left heart syndrome
- Aortic arch interruption
- Coarctation of aorta
- Complex defect

Heart failure

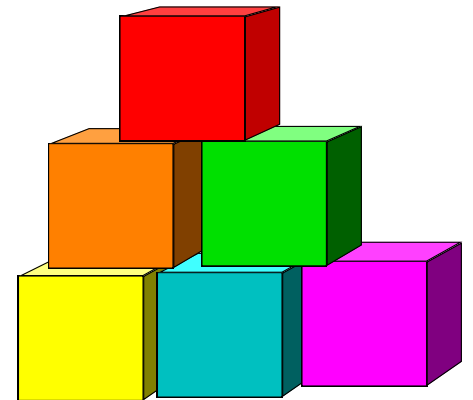
- Cardinal signs
 - Tachypnoea
 - Tachycardia
 - Hepatomegaly
- Feeding difficulty
- Slow weight gain
- Failure to thrive

Diagnosis of Congenital Heart Disease

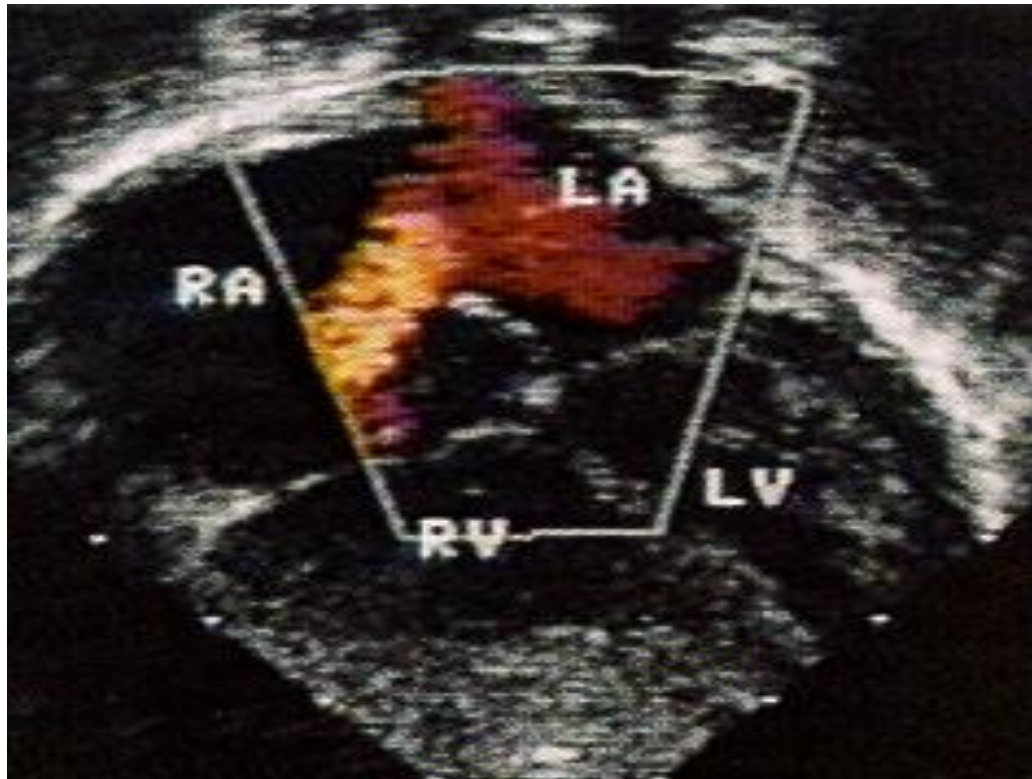
- ECG
- CXR
- **Echocardiography**
- CT scan
- MRI
- Cardiac catheterization

Diagnosis: Echocardiography (segmental approach)

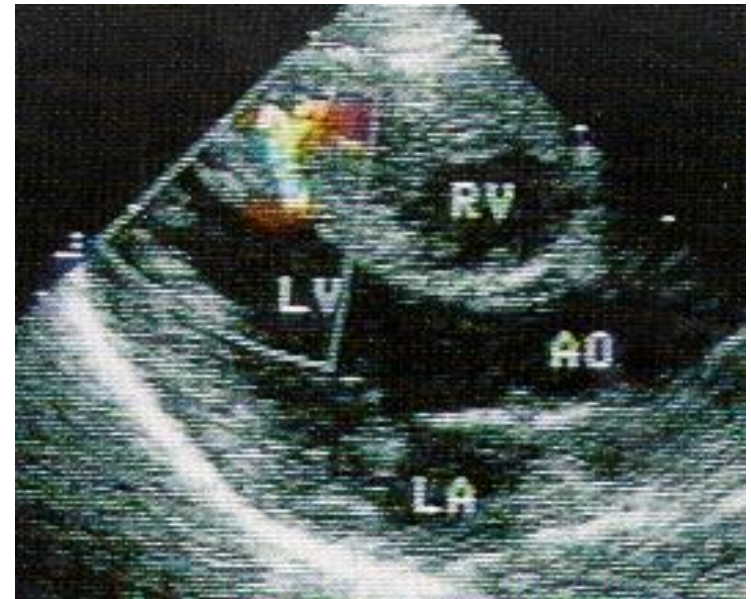
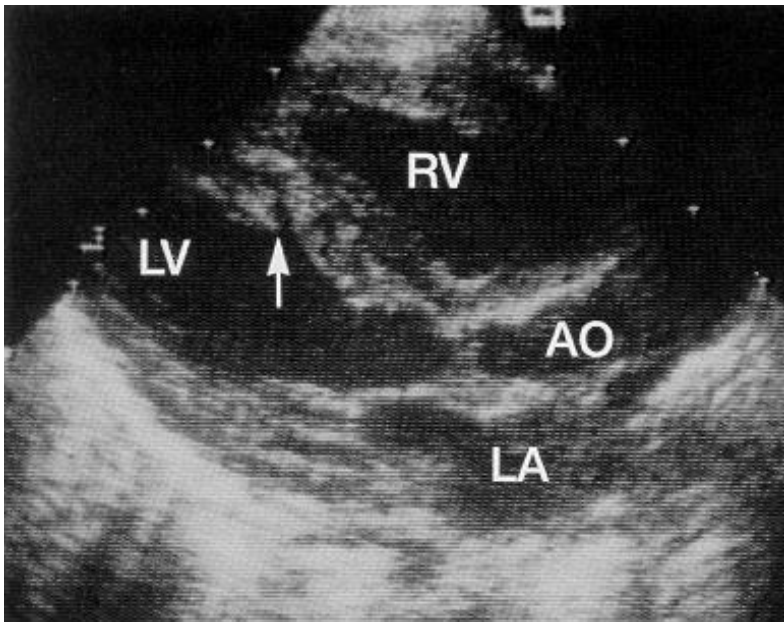
- The segmental approach divides the patient's cardiovascular system into individual segments and the connections between those segments.



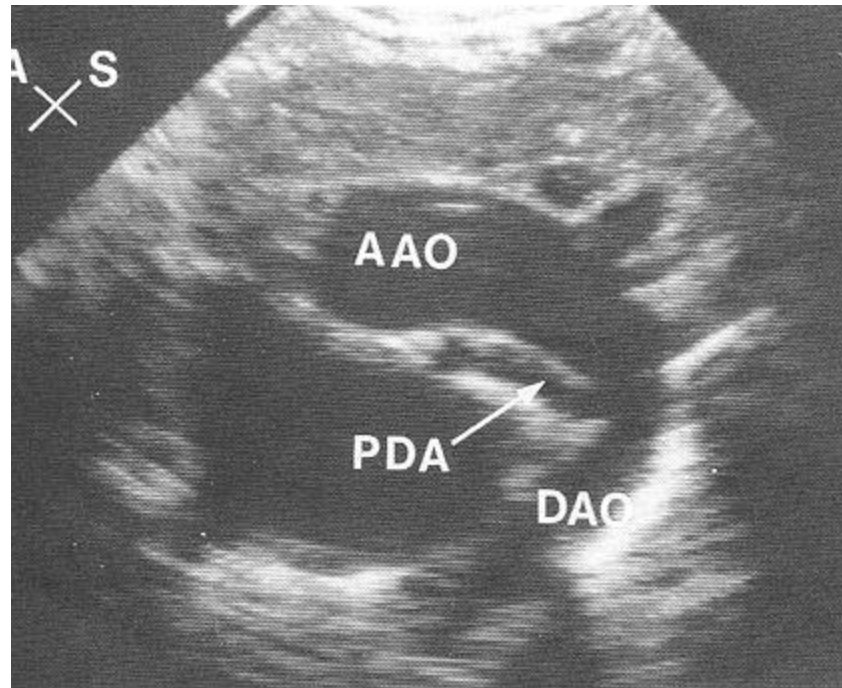
Atrial Septal Defect



Ventricular Septal Defect



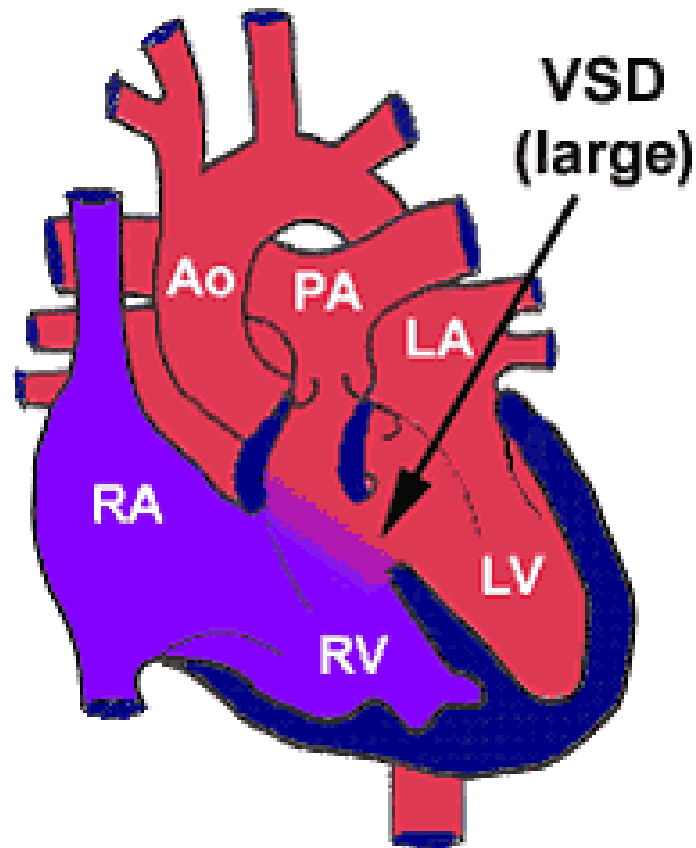
Patent Ductus Arteriosus



Treatment of CHD

- Surgery is the definitive treatment for most of the CHD
- Medical/palliative
 - Diuretic
 - Frusemide
 - Spironolactone
 - Digoxin
 - Vasodilators
 - ACEI (captopril)
 - Hydrallazine
- Interventional cardiac catheterization

Acyanotic lesions



Ventricular septal defect

- Location: subarterial, perimembranous, muscular
- Size
- Associated lesions
 - eg. Coarctation of aorta

Natural history of VSD

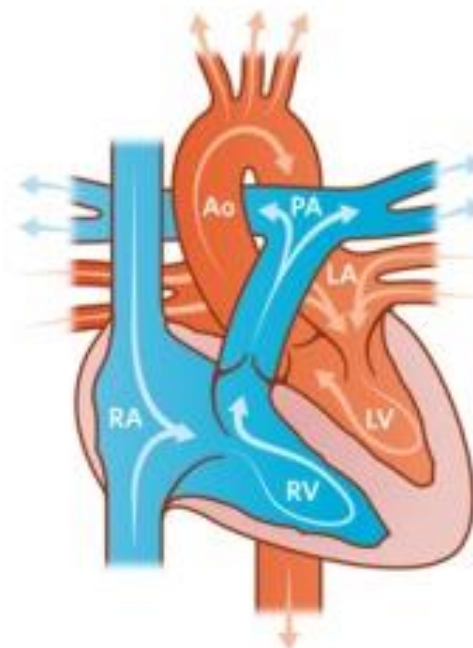
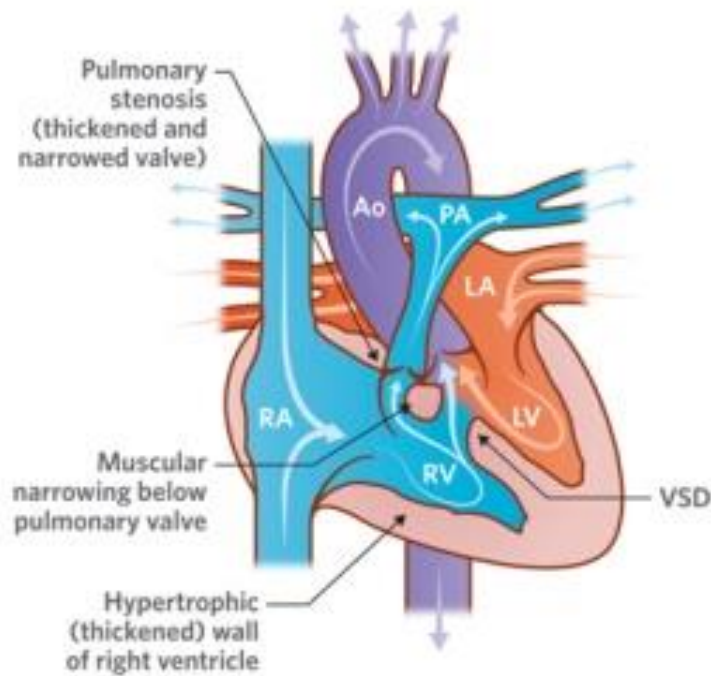
- Decrease in size
- Spontaneous closure
- Congestive heart failure
- Infective endocarditis
- Infundibular stenosis
- Eisenmenger syndrome

Management of VSD

- Leave it alone (? prophylaxis for infective endocarditis)
- Medical treatment to control heart failure
- Surgical repair
- Interventional catheterization to close the VSD with a device

Cyanotic lesions

Tetralogy of Fallot



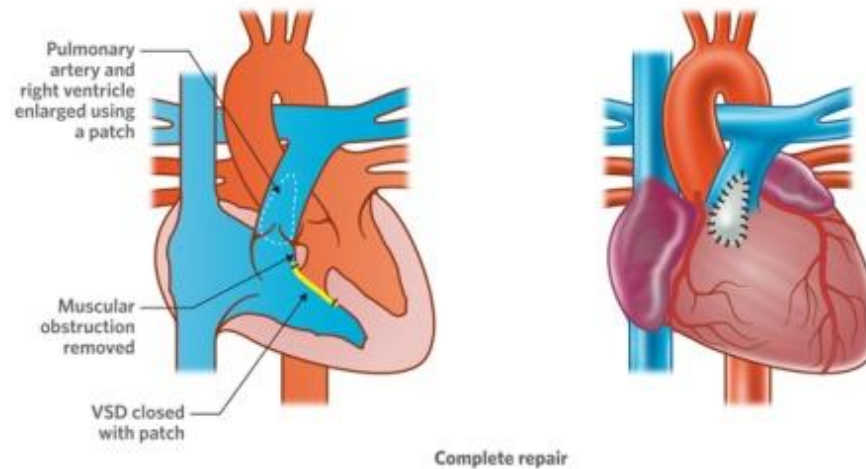
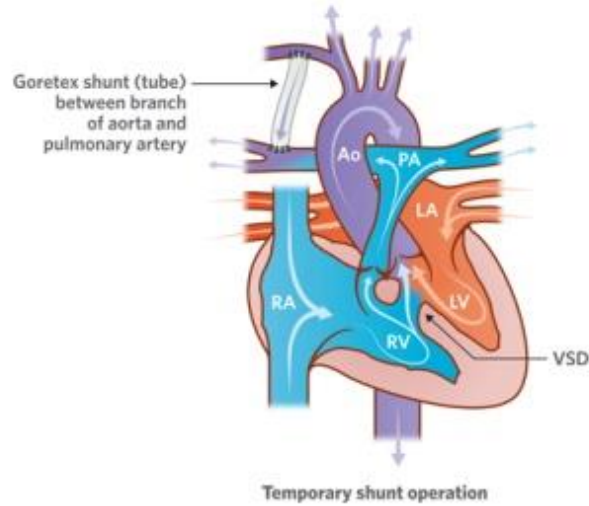
Normal heart and circulation

Natural course of TOF

- Depend on the degree and progression of RV outflow tract obstruction
- Severe: present in neonatal period, require urgent palliative surgery
- Moderate: present in infancy with cyanosis +/- cyanotic spells
- Mild: mild cyanosis with complications of polycythaemia (cerebral thrombosis etc)

Repair of TOF

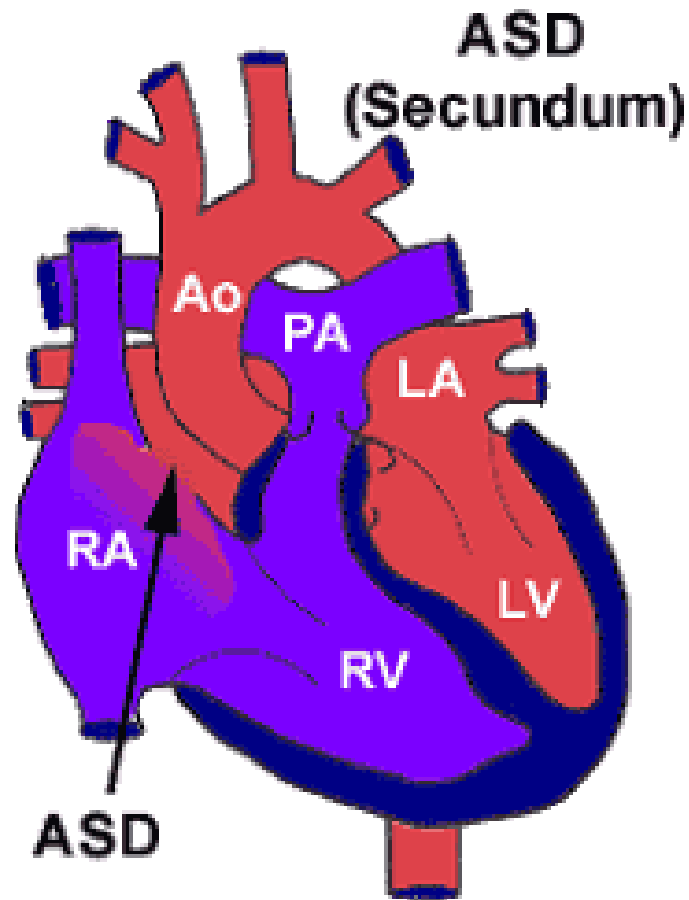
Repair of tetralogy of Fallot



Management of TOF

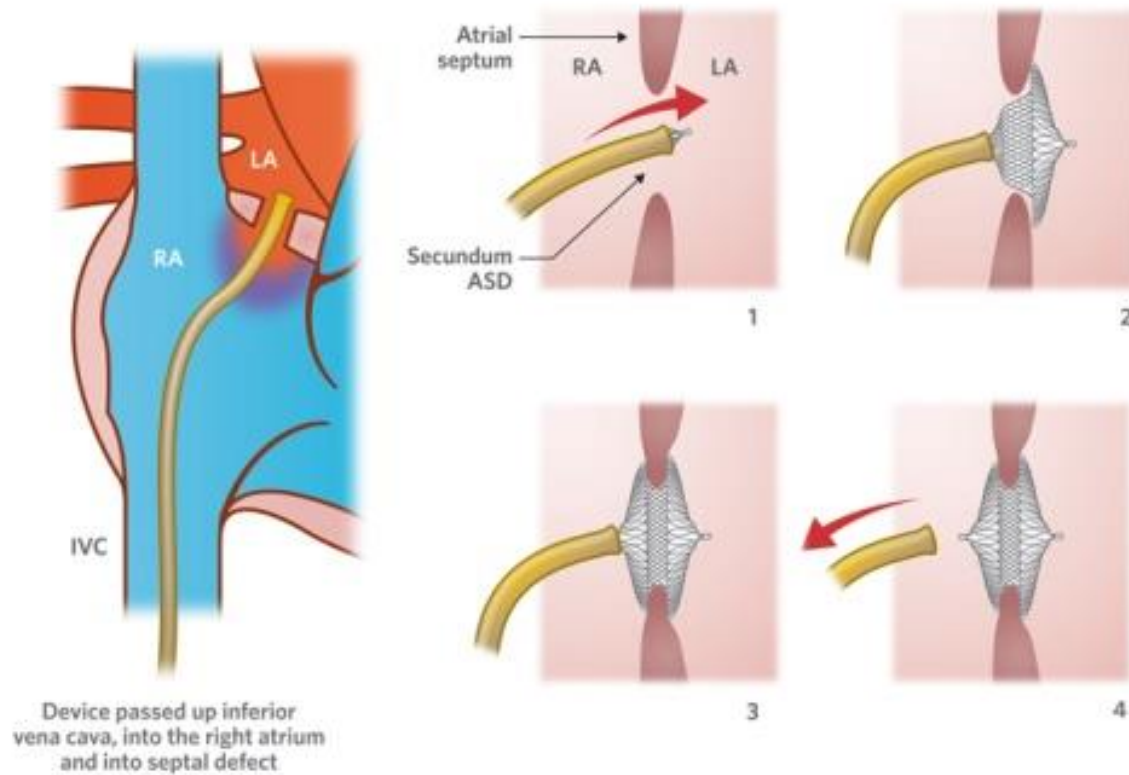
- Total correction by surgery
- Palliative surgery (Blalock Taussig shunt)
- Prophylaxis for infective endocarditis

Atrial septal defect



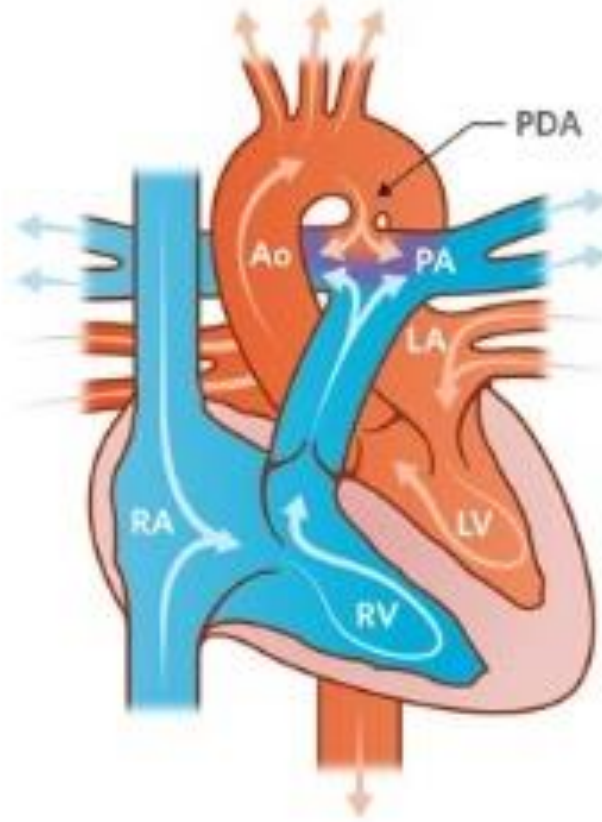
Interventional closure of ASD

Atrial septal defect closure using an expanding device



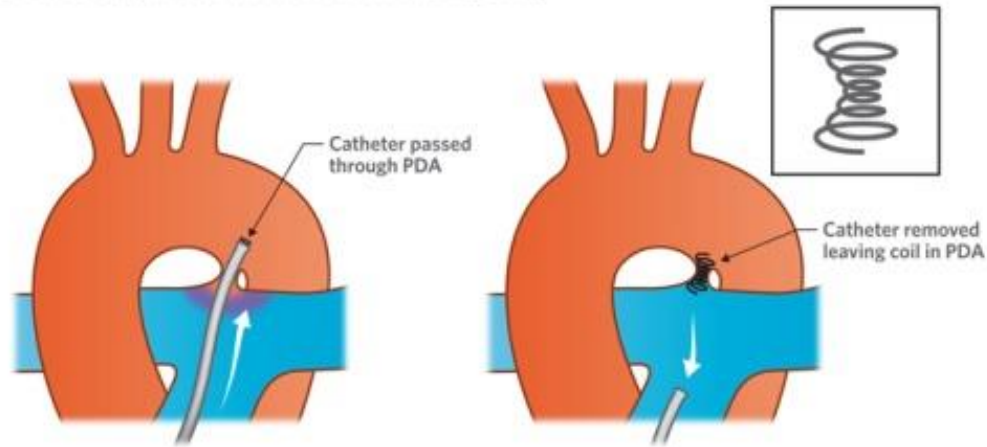
Patent ductus arteriosus (PDA)

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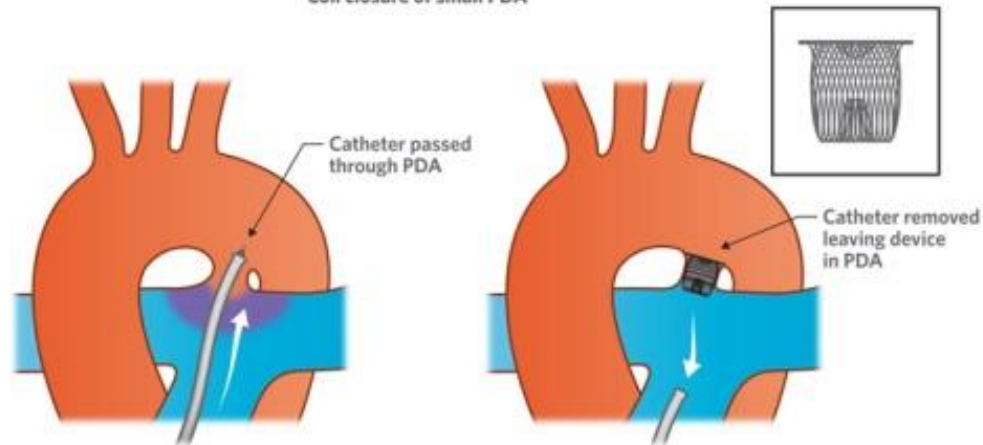


Interventional closure of PDA

Closure of patent ductus arteriosus (PDA)



Coil closure of small PDA



Device closure of larger PDA

Conclusion

- CHD is the most commonest congenital abnormalities in clinical practice
- Haemodynamics in various lesions determine its clinical presentations
- The main treatment is either surgical repair or interventional, medication are for symptomatic relief