CONGENITAL HEART DISEASES (CHD)
These are abnormalities in the cardio-circulatory structures or function that presented at birth, even if it is discovered later.

INCIDENCE: They occur in 8/1000 of live births (about 1% of this risk increase to 4% for 2 pregnancy following the birth of a child with CHD.

AETIOLOGY: Mostly unknown & 20% to siblings having CHD), but appears to result from interaction among many factors, genetic, environmental and maternal diseases, so it is a multifactorial aetiology.

CLASSIFICATION: I- Acyanotic CHD:
  a. Ventricular septal defects (VSD) 25%
  b. Atrial septal defects (ASD) 10%
  c. Patent ductus arteriosus (PDA) 10%
  d. Pulmonary stenosis 10%
  e. Coarctation of the aorta 8%
  f. Aortal stenosis 6%
II. Cyanotic CHD:

a. Tetralogy of Fallot (TOF) 8% (the most common)
b. Transposition of the great arteries (TGA) 5%
c. Single ventricle 2%
d. Tricuspid atresia 2%
e. Other complex CHD < 1% (e.g. double outlet ventricle & truncus arteriosus)

VENTRICULAR SEPTAL DEFECT (VSD)

The most common CHD

1. Anatomically: classified into:
   1. Perimembranous VSD 80%
   2. Muscular VSD 5%
   3. Inlet VSD 5%

   Hemodynamically (functionally): classified into:
   1. Small VSD (small Lt.-Rt. Shunt)
   2. Moderate VSD
   3. Large VSD
**PATHOPHYSIOLOGY:**

The functional disturbance caused by VSD magnitude of the L-R hunt depends on: size of VSD & degree of pulmonary vascular resistance not on the site of VSD.

In large VSD, there is no resistance to the flow, so there is large hunt and there will be progressive increase in right ventricular (RV) & pulmonary artery pressure, as a result of increase of the pulmonary vascular resistance which causes Rt-Lt. (Eisenmenger syndrome) which is irreversible.

In small VSD, there is high resistance to the flow through the VSD so there is small Lt.-Rt Shunt and the pressure is normal in RV and pulmonary artery (PA).

**CLINICAL FEATURES:**

Small VSD: the patients are asymptomatic. O/E ^ loud harsh, grade 4-6 pansystolic murmur at the lower left sternal border.

Large VSD ± heart failure occurs in infants between the age of 2-8 weeks; the patient will show tachypnoea, dyspnoea, recurrent pulmonary infections, feeding difficulty, poor growth and excessive sweating. Q/E the murmur is soft with loud P2 (the more pulmonary vascular resistance, the louder the P2 will be).
DIAGNOSIS:
1. ECG:
   a. small VSD -> normal ECG
   b. large VSD -> biventricular hypertrophy
2. CxR:
   a. small VSD -> normal
   b. large VSD -> cardiomegaly, dilated pulmonary vessels (plethoric lung)
3. Echo: Two-dimensional & Doppler echo must be done in all patients to determine the following:
   a. Size
   b. Anatomical location
   c. Size & direction of intercardiac shunt
   d. The degree of pulmonary hypertension
   e. Presence of associated lesion (as TOF)
4. Catheterization & angiography: A small number of patients with VSD requires cath. (because echo is more sensitive & more specific).
   a. Provide a clear anatomical picture of the location and no. of VSD in patients when surgery is required.
   b. Estimation of magnitude of the shunt & pulmonary vascular resistance (to know if the patient is still operable or not).
   c. Closing some muscular VSDs with catheter derived devices.
NATURAL HISTORY:

^ Small VSD: about 50% will close spontaneously; the majority during the 1st 2 years of life;
  the other will remain asymptomatic. ^ Large VSD: most patients need surgical repair, in the 1st year of life,
Without surgical repair, most patients will develop pulmonary hypertension
and some reach to Eisenmenger syndrome (10%), but 5% will develop infundibular & pulmonary stenosis. S

MANAGEMENT:

Small VSD: surgery is not indicated (because the mortality & morbidity of surgery > natural history of the VSD). The patient needs no treatment apart from follow-up & SBE prophylaxis.
S Large VSD: we have medical, surgical and catheterization options.
1.  Medical:
   a. Treatment of heart failure.
   b. Treatment of infection, especially respiratory.
   c. SBE prophylaxis.
   d. Nutritional support.
2.  Surgical:
   a. Either pulmonary artery banding
   b. Or total surgical repair,
Indications:
b. Repeated pulmonary infections.
c. Failure to thrive and growth failure.
d. Any degree of pulmonary hypertension (after 2 years of age).

3. Catheterization based treatment: Most muscular VSD and some membranous VSD can be closed by devices placed during cardiac catheterization.

ATRIAL SEPTAL DEFECT (ASP)
an opening in the inter-atrial septum other than patent foramen ovale; more common in females; F:M ratio is 3:1.

It has 3 types:
1. Primum ASD (in lower part) 10%
2. Secondum ASD (in the middle) 80%
3. Sinus venosus (in the upper part) 10%

HEMODYNAMIC EFFECT: There will be chronic Lt-Rt. Shunt which causes volume overload on the Rt. Sided cardiac structures & result in their dilatation & increase of pulmonary blood flow.
C/F(MQSTLY SECONDUM TYFE):
Most of them are asymptomatic. S Normal growth
Dx accidentally: fixed splittin of 2nd heart sound, soft ejection systolic murmur at the 2 left intercostal space, no thrill & parasternal heave. S DX:
A. CXR
Cardiomegaly of RV configuration
   Round apex peak
   Increase pulmonary marking
B. ECG:
   Right axis deviation (RAD)
   Incomplete RBBB
   Peaked P-wave (RA enlargement)
C- Echo: Trans-thoracic & trans-esophageal echocardiography is essential for Dx.
D. Cardiac Catheterization: Not essential for Dx, indicated in: Therapeutic
   aim & 'Exclude associated cardiac anomaly as patent anomalies.

**Natural history:** ASD is abenign lesion

Rarely causes heart failure or pulmonary hypertension in the 1st decade of life

Rarely complicated by infective endocarditis

Rarely closed spontaneously after infancy
Treatment;
Closure of ASD in asymptomatic patients is done at 4-5 y
Either by occluder, which if failed by surgical repair.

Atrioventricular septal defect
(endocardial cushion defect)

It is a group of anomalies sharing a defect at the site of atrioventricular septum
And abnormality in the atrioventricular valve.

Common in downs syndrome, so all patients with downs syndrome should
Have cardiac evaluation if symptomatic or before 6 months of age

Equal no. of male and females are affected
CLASSIFICATION:
1. Partial: ASD primum, clefted mitral valve.
2. Complete: ASD primum, large inlet VSD, common single atrioventricular valve.

C/F:
1- Partial: either asymptomatic (mainly) or mild symptoms. Q/E: ASD murmur of mitral regurgitation. 2. Complete: as the above with heart failure and/or pulmonary vascular disease.

DX:
VCXR;
ECG: (prolonged PR interval), S Echo:

NATURAL HISTORY:
It depends on the size of various atrial and ventricular defects and the amount of mitral regurgitation.
The ostium primum ASD with no mitral regurgitation has the same benign natural history of simple secondum ASD, but with complete AV-canal defect, heart failure and/or pulmonary vascular disease may occur.
RX: repair the septal effect (patching), it will cause complete repair.