

Acyanotic CHD

Dr.Emamzadegan

**Pediatric & Congenital
Cardiologist**

Acyanotic CHD

- Epidemiology
- ETIOLOGY :

Most cases of congenital heart disease were thought to be **multifactorial** and result from a combination of **genetic** predisposition and **environmental** stimulus.

ETIOLOGY :

- **chromosomal abnormalities :**
trisomy 21, 13, and 18 and Turner syndrome;

heart disease is found in more than 90% of patients with trisomy 18, 50% of patients with trisomy 21., and 40% of those with Turner syndrome.

chromosomal abnormalities

- Deletion of a large region of chromosome 22q11.2, known as the **DiGeorge** critical region. (**CATCH 22**)

Environmental

- 1. Fever & Flu like illness during pregnancy**
- 2. Vitamin A**
- 3. Viral infections (Rubella)**
- 4. Drugs(Valproate ;lithium ; Accutane)**
- 5. Alcohol; smoking?**
- 6. Diabetic mother**
- 7. Lupus (Congenital CHB)**
- 8. Chemical materials**

ACYANOTIC CHD

- The most common lesions are those that produce a **volume load**, and the most common of these are left-to-right shunt lesions .(**ASD,VSD,CAVSD,PDA**)
Atrioventricular (AV) valve regurgitation and some of the cardiomyopathies are other causes of increased volume load.

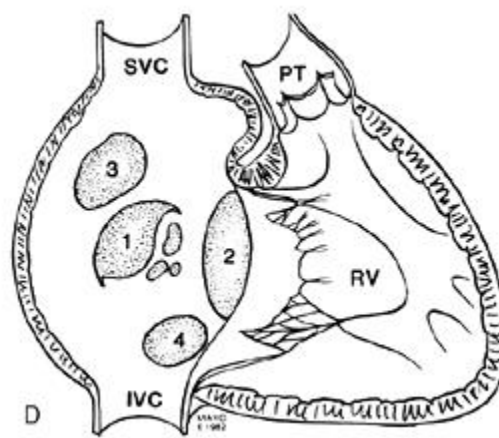
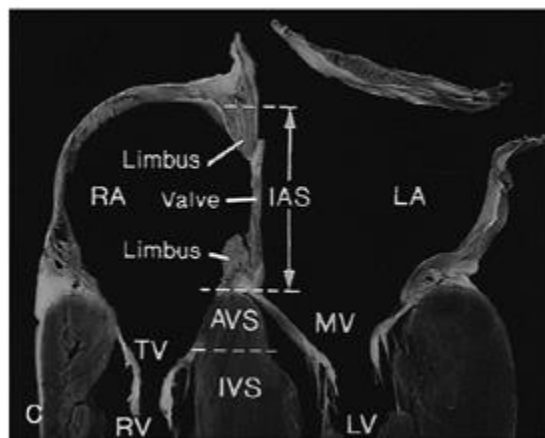
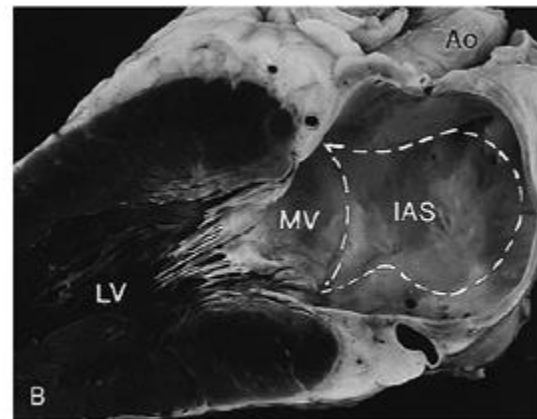
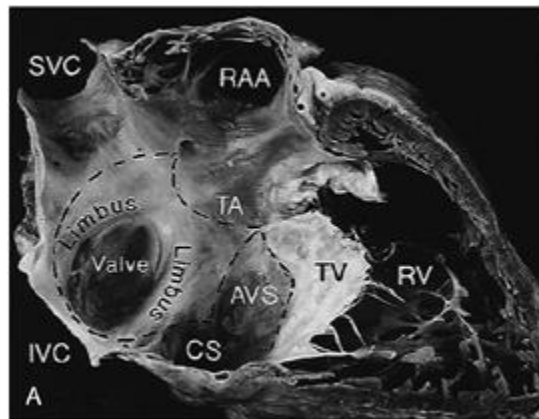
ACYANOTIC CHD

- The second major class of lesions causes an increase in **pressure load**, most commonly secondary to ventricular outflow obstruction (**pulmonic or aortic valve stenosis**) or narrowing of one of the great vessels (**coarctation** of the aorta).

Atrial septal defect

- **Etiology** : Failure of septal growth or excessive reabsorption of septum.
- **Frequency** : 10% of CHD
- **Male : Female** = 1 : 3
- **Typing**: Secundum ,Primum, Sinus venosus
- **The most common type** : ASD Secundum
- **Lt to Rt shunt** : Size of ASD & Compliance of RV & LV
- **ASD closure** : If significant shunt at around 3 years

ASD



Atrial septal defect

- **ASD closure device : Amplatzer**
for ASD secundum
- **Prophylaxis for Endocarditis:**
for non secundum types

Atrial septal defect

- An isolated valve-incompetent patent foramen ovale (**PFO**) is a common echocardiographic finding during infancy.
- **Ostium secundum defect:**

Partial anomalous pulmonary venous return, most commonly of the right upper pulmonary vein, may be an associated lesion in **Sinus venosus type**.

Atrial septal defect

- **CLINICAL MANIFESTATIONS:**
 1. often asymptomatic
 2. rarely produces clinically evident heart failure in childhood.
 3. In younger children, subtle failure to thrive may be present;
in older children, varying degrees of exercise intolerance may be noted.

Atrial septal defect

4. In most patients, the **2nd heart sound** is characteristically **widely split** and fixed in its splitting in all phases of respiration.
5. **Systolic ejection murmur**
6. **Mid diastolic rumble usually indicates a Qp/Qs ratio of at least 2 : 1**

Atrial septal defect

- **DIAGNOSIS:**

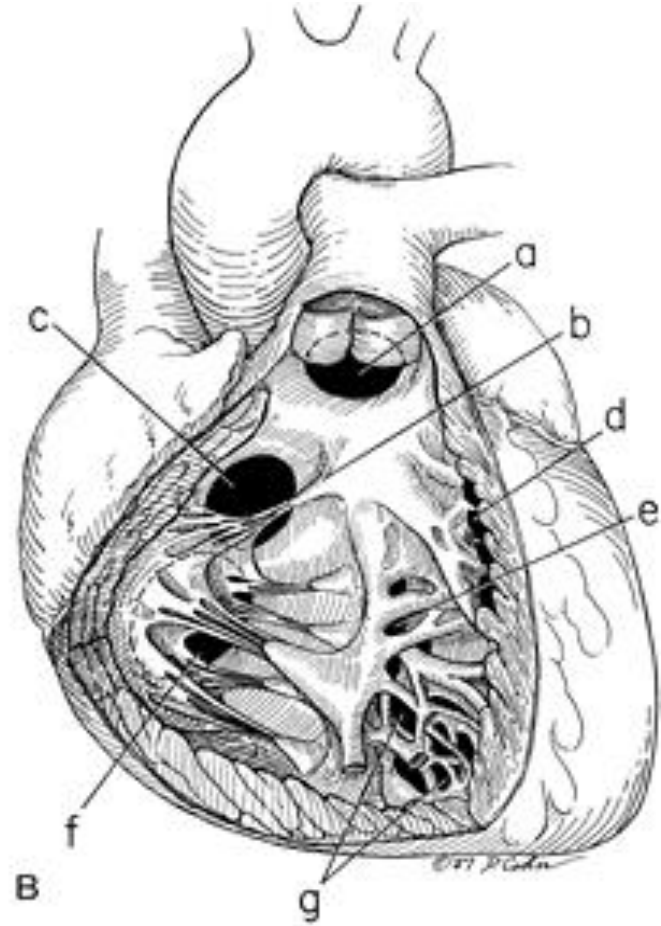
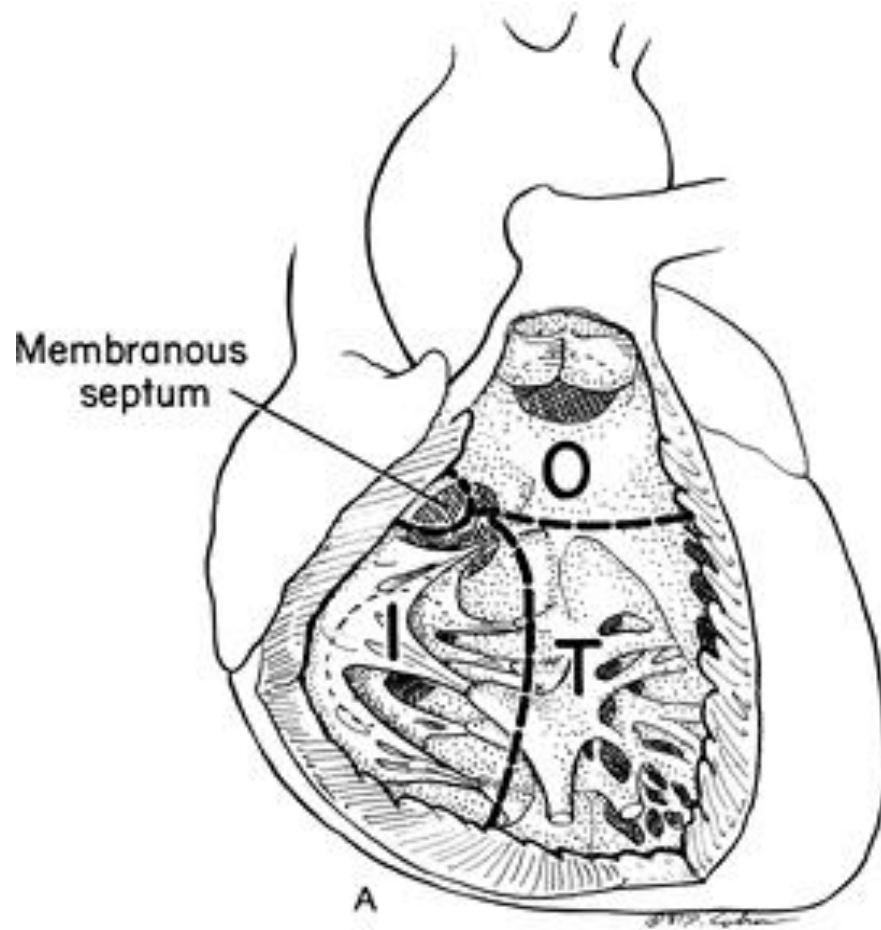
1.CXR :Cardiac enlargement is often best appreciated on the lateral view.

2.ECG : rsR'', RAD

Ventricular Septal Defect

- **25% of congenital heart disease** (p:1888)
- **The most common CHD**
- **Clinical findings of patients with a VSD vary according to the size of the defect and pulmonary blood flow and pressure.**
- In the immediate **neonatal period**, the left to-right shunt may be minimal because of higher right-sided pressure, and therefore the systolic **murmur may not be audible** during the 1st few days of life.

VSD



Ventricular Septal Defect

- **VSD Typing** : Inlet(endocardial cushion defect),membranous (the most common type= 67%),sub arterial (supracristal or sub pulmonic), muscular
- **Lt to Rt shunt** : size of defect & PVR(PAP/QP)
- **Mumur**: Holosystolic(pansystolic) at the lower LSB
- **Intensity of P2** : PAP
- **Early decrescendo diastolic MM at upper LSB= PI(PH)**

Ventricular Septal Defect

- **Large VSDs** with excessive pulmonary blood flow and pulmonary hypertension are responsible for **dyspnea**, feeding difficulties, **poor growth**, profuse **perspiration**, **recurrent pulmonary infections**, and **cardiac failure** in early infancy.

Ventricular Septal Defect

- The holosystolic murmur of a large VSD is generally less harsh than that of a small VSD.
- The presence of a **mid-diastolic**, low-pitched **rumble** at the apex is caused by increased blood flow across the mitral valve and indicates a **$Q_p : Q_s$ ratio of $>2 : 1$**

Ventricular Septal Defect

- **DIAGNOSIS :**

1. CXR, In patients with **small VSDs**, the chest radiograph is **usually normal**.

In **large VSDs cardiomegaly** with prominence of both ventricles, the left atrium, and the pulmonary artery . **Pulmonary vascular markings are increased**, and frank pulmonary edema' including pleural effusions, may be present.

DIAGNOSIS :

2.ECG : BVH, If pure RVH =PH

TREATMENT

- The natural course of a VSD depends to a large degree on the **size of the defect**. A significant number (30-50%) of small defects close spontaneously most frequently during the 1.st 2 yr of life.

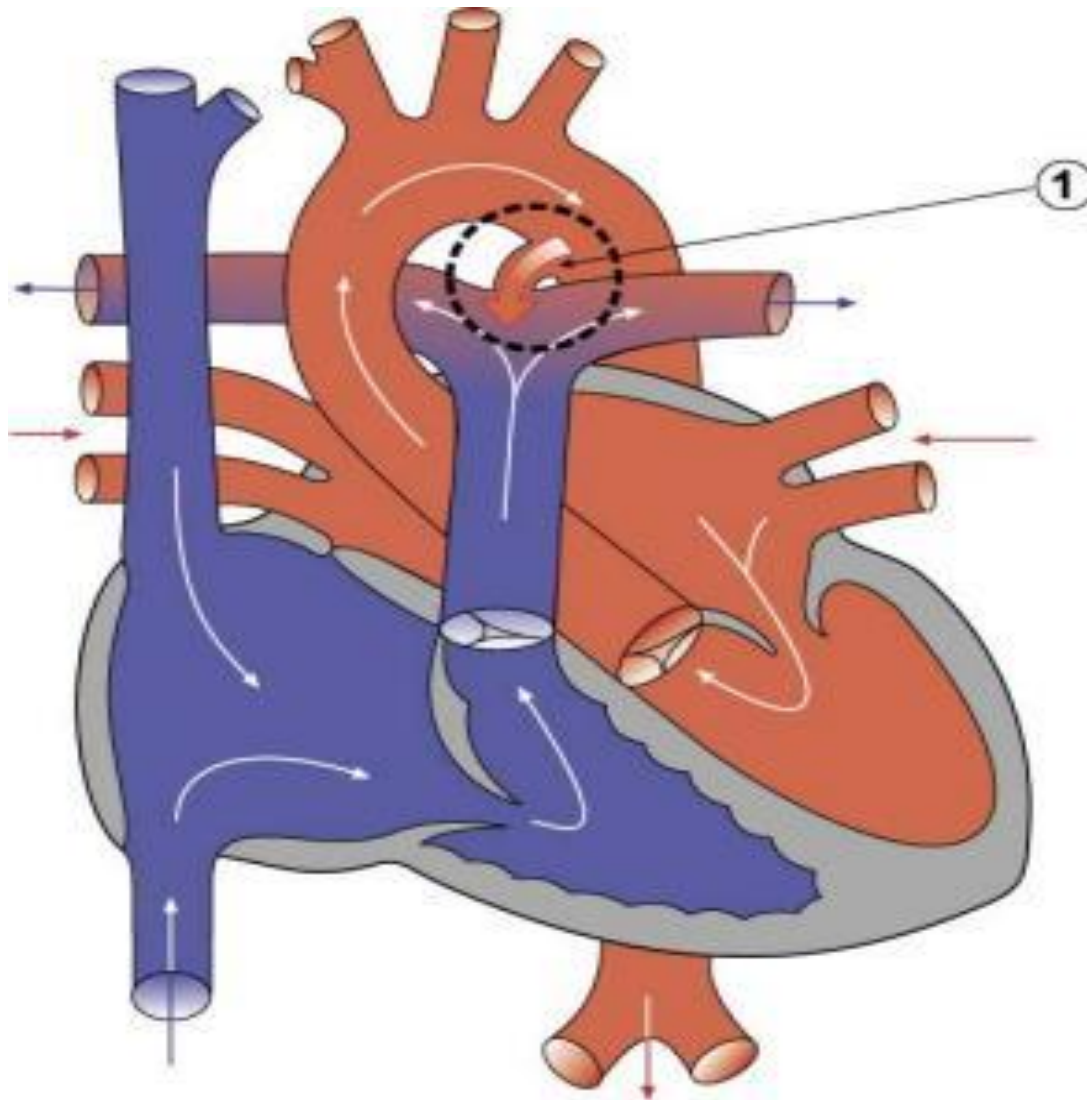
TREATMENT

- 35% of all VSDs close spontaneously.
- Small muscular VSDs are more likely to close (up to 80%) than membranous VSDs are (up to 35%).
- Supracristal VSD with AI = surgery
- **Small VSD closure** without symptom (no PH) **may not be required** but SBE prophylaxis is necessary.

Patent Ductus Arteriosus

- 5-10% of CHD
- **Clinical manifestations:**
 - a.** A small PDA does not usually have any **symptoms** associated with it.
 - b.** A large PDA will result in **heart failure** (after **6-8 w of life**) similar to that encountered in infants with a large VSD. **Retardation of physical growth** may be a major manifestation in infants with large shunts.

PDA



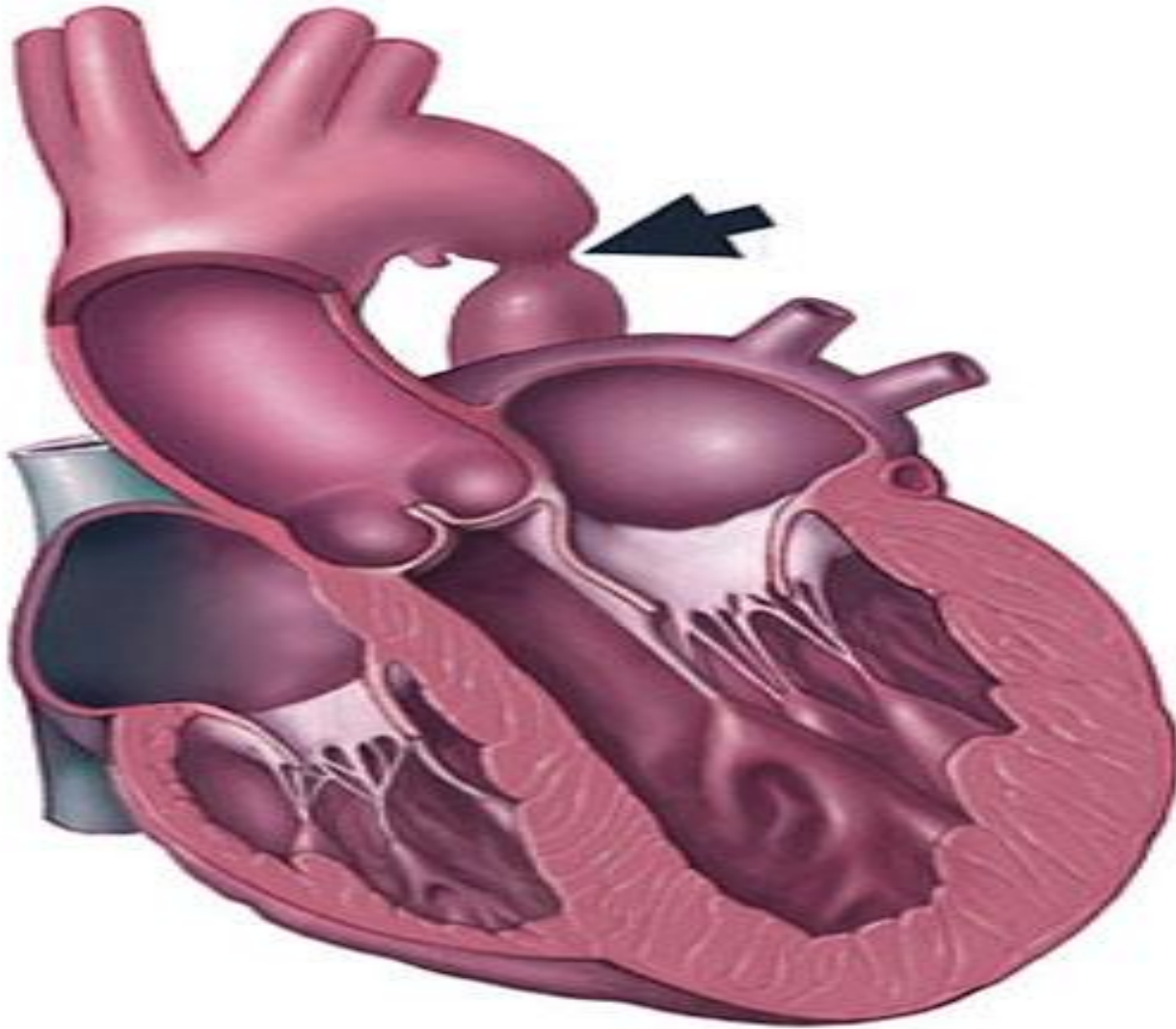
Patent Ductus Arteriosus

- bounding peripheral arterial pulses.
- A thrill, maximal in the 2nd left interspace, is often present.
- The **classic continuous murmur** is described as being like machinery in quality. It begins soon after onset of the 1st sound, reaches maximal intensity at the end of systole, and wanes in late diastole. (at Lt subclavicular)
- **P2 intensity = PAP**
- **Lt to Rt shunt: Size &PVR**

Patent Ductus Arteriosus

- **Diagnosis** : CXR ,ECG ,Echo
- **TREATMENT:** Irrespective of age, **all** patients with PDA require surgical or catheter closure (**Coil, Amplatzer**) ,because of the risk of Endocarditis.

Coarctation



Coarctation of Aorta

- **98% occur just below the origin of the left subclavian artery at the origin of the ductus arteriosus (Juxtaductal coarctation).**
- **Male : Female = 2 : 1**
- **Turner syndrome**
- CoA is associated with a **bicuspid aortic valve** in more than 70% of patients.

Coarctation of Aorta

- Mitral valve abnormalities and subaortic stenosis are potential associated lesions. When this group of left-sided obstructive lesions occurs together, they are referred to as the **Shone complex**.
- can occur as a **discrete juxtaductal obstruction** or as **tubular hypoplasia** of the transverse aorta.

Coarctation of Aorta

- **Blood pressure is elevated in the vessels that arise proximal to the coarctation.**

CLINICAL MANIFESTATION

- **Infancy = No symptom; Heart Failure**
- **After infancy :** is not usually associated with significant symptoms. weakness or pain (or both) in the legs after exercise, but in many instances, even patients with severe coarctation are **asymptomatic**.
- **Older children :** hypertensive on routine physical examination.

CLINICAL MANIFESTATION

- **The classic sign of coarctation of the aorta is a disparity in pulsation and blood pressure in the arms and legs.**
- **A radial-femoral delay**
- **In older than 1 yr, about 90% of whom have systolic hypertension in an upper extremity greater than the 95th percentile for age.**

CLINICAL MANIFESTATION

- **A systolic murmur is heard along the left sternal border with a loud 2nd heart sound.**
- **Differential cyanosis = PDA**

DIAGNOSIS

- **CXR** : Cardiac enlargement and pulmonary congestion are noted in infants with severe coarctation.

Notching of the inferior border of the ribs from pressure erosion by enlarged collateral vessels is common. (Late)

COA

- In the immediate postoperative course, "rebound" hypertension is common.
- **POST COARCTECTOMY SYNDROME :**
Postoperative mesenteric arteritis may be associated with acute hypertension and abdominal pain in the immediate postoperative period. (Nausea, Vomiting, leukocytosis, intestinal hemorrhage, bowel necrosis, and small bowel obstruction).

POST COARCTECTOMY SYNDROME

- **Relief** is usually obtained with antihypertensive drugs (nitroprusside, esmolol, captopril) and intestinal decompression; surgical exploration is rarely required for bowel obstruction or infarction.

Treatment

CoA with HF in infancy:

PGE1, inotropics, diuretics

1. **Coarctectomy**
2. **Balloon angioplasty**; Should recoarctation occur, is the procedure of choice.
3. **Intravascular stents**; are commonly used, especially in adolescents and young adults.

Complications

- **Subarachnoid or intracerebral hemorrhage** may result from rupture of congenital aneurysms in the circle of Willis.
- **Rupture of normal vessels; these accidents are secondary to hypertension.**
- Aneurysms of the descending aorta or the enlarged collateral vessels may develop.
- Infective endocarditis or endarteritis

CoA

- **PHACE syndrome :**

posterior brain fossa anomalies, facial hemangiomas, arterial anomalies, cardiac anomalies and CoA, eye anomalies; may have stroke.

Endocardial Cushion Defect

- A-V canal defect (AVSD)
- **Etiology:** failure of septum to fuse with endocardial cushion
- **Complete defect:** ASD primum, inlet VSD, cleft in Ant leaflet of MV & septal leaflet of TV
- **Partial defect:** ASD primum +MR (cleft)

Endocardial Cushion Defect

- **Pathophysiology** :ASD+VSD+ AV valve insufficiency
- **Clinical manifestation**: CHF over the 6-8 w of life, PH (**eisenmenger ,PVOD**)
- **Down syndrome**: near 50% with CAVSD
- Murmur may be not significant (Down=Echo)
- **CXR**: C/T & PVM increased
- **ECG**:LAD,BVH

Treatment

- Digoxin + Diuretic then surgery

AS

- Mild : 10-30 mmhg
- Moderate : 30-60 mmhg
- Severe : >60mmhg(sudden death)
- Systolic murmur in upper Rt sternal border
- Treatment : PTAC

PS

- Mild : 10-30 mmhg
- Moderate : 30-60 mmhg
- Severe >60 mmhg
- Systolic murmur in upper Lt sternal border
- Noonan syd
- PPS (Congenital Rubella)
- Treatment : PTPC