Pediatrics cardiology L1

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CONGENITAL HEART DISEASES (CHD)

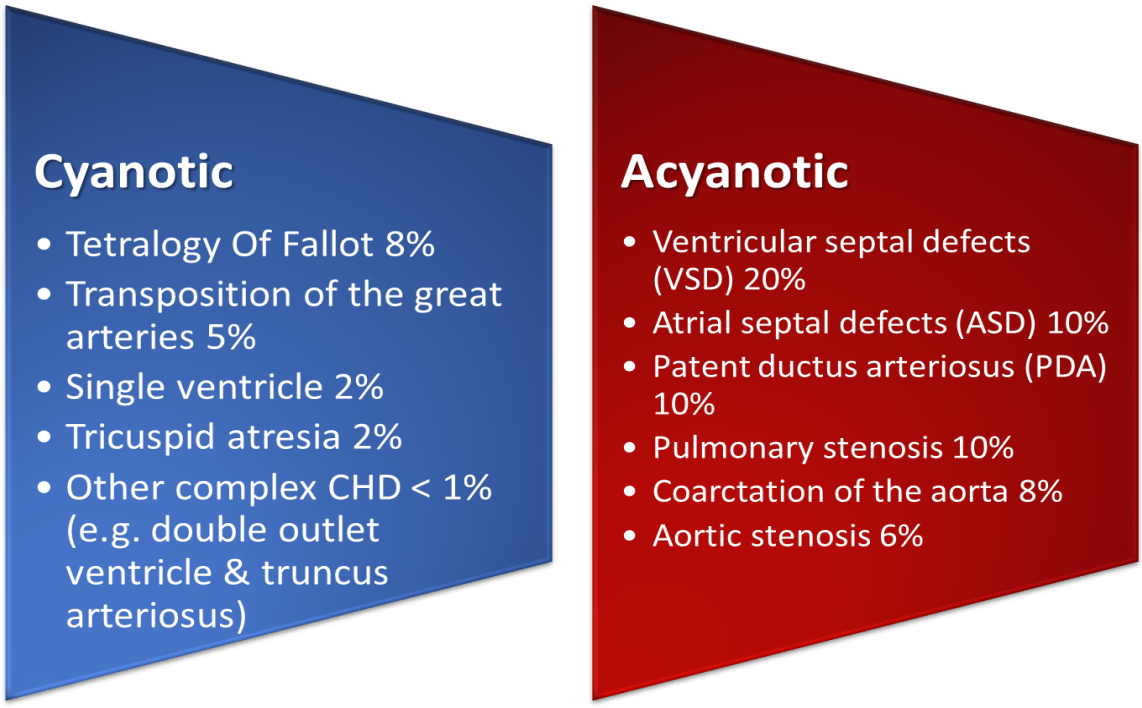
These are abnormalities in the cardio-circulatory structures or function that are presented at birth, even if it is discovered later.

etiology:

Mostly (80%) unknown: multifactorial

Interaction among many factors:Genetic,Environmental,Maternal diseases, etc.

About 20% of congenital heart disease (CHD) can be attributed to known causes such as genetic syndromes, chromosomal anomalies, and teratogens (Down syndrome, Turner syndrome, etc.)



Classification of congenital heart disease:

Conditions with ↑ PBF: (intar- or extracardiac Lt → Rt shunt "without" RV outflow obstruction): best examples VSD, PDA.

Conditions with ↓ PBF: (shunt "with" RV outflow obstruction): eg. TOF

Conditions with normal PBF: eg. Coarctation of aorta, Pulmonary stenosis (PS), Aortic stenosis (AS).

1. Conditions with ↑ PBF:

Examples:
Ventricular septal defect (VSD); Atrial septal defect (ASD); Patent ductus arteriosus (PDA); Total anomolus pulmonary venous drainage (TAPVD);

Clinical Features:

1 Signs & symptoms of heart failure
2 Recurrent chest infections
3 Failure to thrive

Complications:

Pulmonary hypertension +/- Eisenmenger's syndrome

Chest X-rays:

1 Cardiomegaly
2 Plethoric lungs.

Echocardiography :

Defining the defect with "no PS".

Medical Treatment:

1 Antifailure medications.
2 Nutritional support
3 Treatment of infections
4 SBE prophylaxis

Surgical Treatment:

A Palliative: Pulmonary artery (PA) banding
B Definitive: closure of the defect

2 Conditions with ↓ PBF:

EXAMPLES:

1 Tetralogy of Fallot (TOF)

2 Double outlet right ventricle (DORV) with PS

3 Single ventricle with PS.

CLINICAL FEATURES:

1 Cyanosis

2 Hypercyanotic spells (cyanotic spells; Fallot spells; "tet" spells).

Complications:

1 Polycythemia

2 Cerebrovascular accident (CVA)

3 Brain abscess

Chest X-rays:

1 "Small" heart

2 Oligemic lung

Echocardiography :

Different cardiac anomalies "with PS".

Medical Treatment:

1 Beta-Blockers

2 Treatment of complication (eg. Hypercyanotic spells- see later)

Surgical Treatment:

A Palliative: shunt operation (e.g. BT shunt(1), Glenn operation(2))

B Definitive: according to the lesion.

Notes #"Pulmonary hypertension" is defined as a mean pulmonary artery pressure >25 mmHg at rest or >30 mmHg with exercise. It may be idiopathic, familial, or associated with multiple other diseases.

#"Shunt size": The extent of extra flow is assessed as the ratio of measured pulmonary blood flow (Qp) to systemic blood flow (Qs). In the normal case, where no connection exists, the ratio Qp:Qs is 1:1. Left-to-right shunting results in a Qp:Qs >1, while right-to-left shunting results in a Qp:Qs <1. For example, a Qp:Qs of 2:1 indicates that the pulmonary blood flow is twice that of systemic blood flow.

#The triad of systemic-to-pulmonary communication, pulmonary vascular disease and cyanosis is called

"Eisenmenger syndrome". The diagnosis of Eisenmenger syndrome implies that the development of

pulmonary vascular disease as a consequence of increased pulmonary blood flow, and requires exclusion of other causes of PAH.

VENTRICULAR SEPTAL DEFECT (VSD

The most common CHD

Anatomically: classified into:

1 Perimembranous VSD 80%

2 Muscular VSD 5%

3 Inlet VSD 5%

4 Outlet VSD.

Hemodynamically (functionally)classified into:

1 Small VSD (small Lt.-Rt. Shunt)

2 Moderate VSD

3 Large VSD

Pathophysiologiy:

The magnitude of the L-R shunt depends on: size of VSD & degree of pulmonary vascular resistance.

In large VSD, there is no resistance to the flow → large shunt ⟶ progressive increase in right ventricular (RV) & pulmonary artery pressure ⟶ 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 ⟶ small Lt.-Rt. Shunt pressure is normal in RV and pulmonary artery (PA).

Clinical features:

Small VSD: asymptomatic.

O/E: loud harsh, grade 4-6 pansystolic murmur at the lower left sternal border.

Large VSD: heart failure (between the age of 2-8 weeks); the patient will show tachypnoea, dyspnoea, recurrent pulmonary infections, feeding difficulty, poor growth and excessive sweating.

O/E: the murmur is soft with loud P2.

Diagnosis:

ECG: small VSD , normal ECG \ large VSD , biventricular hypertrophy

CXR;small VSD , normal \ large VSD , cardiomegaly, dilated pulmonary vessels (plethoric lung)

Echo: two-dimensional & Doppler echo must be done in all patients to determine the following:

Size

Anatomical location

Size & direction of intercardiac shunt

The degree of pulmonary hypertension

Presence of associated lesion (e.g. PS)

Natural history:

 # Small VSD: 50% close spontaneously (the majority during the 1st 2 years) ,If not closed remains asymptomatic.

 # Large VSD: Heart failure ,Recurrent chest infections ,Failure to thrive ,Infundibular pulmonary stenosis (protective!) ,Without surgical repair, most patients will develop pulmonary hypertension (PHT) and some reach to Eisenmenger syndrome (10%).

Management:

Small VSD: The patient needs no treatment apart from follow-up.

Large VSD: we have medical, surgical and catheterization options.

Medical:

1 Treatment of heart failure.

2 Treatment of infection, especially respiratory.

3 Nutritional support.

ATRIAL SEPTAL DEFECT (ASD)

An opening in the inter-atrial septum other than patent foramen ovale

More common in females (F:M ratio is 3:1).

It has 4 types:

1 Primum ASD (in lower part) 10%

2 Secondum ASD (in the middle) 80%

3 Sinus venosus (near the vena cavae) 10%

4 Coronary sinus ASD: rare

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.

Clinical Features: (mostly secondum type)

Most of them are asymptomatic.

Normal growth

Dx accidentally: Wide, fixed splitting of 2nd heart sound, soft ejection systolic murmur at the 2nd left intercostal space, no thrill (grade 3) & parasternal RV heave

Dx: CXR:1. Cardiomegaly of RV configuration. 2. Increase pulmonary marking

 # ECG: 1 Right axis deviation (RAD) .2 Incomplete RBBB .3 Peaked P-wave (RA enlargement)

 # Echo

 # Cardiac Catheterization: Not essential for Dx, indicated in:1 Therapeutic aim. 2 Exclude associated cardiac anomaly as patent anomalies

Natural history:

1 Spontaneous closure occurs more than 80% of the time in patients with defects between 3 and 8 mm before 1½ years of age. An ASD with a diameter greater than 8 mm rarely closes spontaneously.

2 Most children with an ASD remain active and asymptomatic.

3 If a large defect is untreated, CHF and pulmonary hypertension develop in adults who are in their 20s and 30s.

4 With or without surgery, atrial arrhythmias (flutter or fibrillation) may occur in adults.

5 Infective endocarditis does not occur in patients with isolated ASDs.

6 Cerebrovascular accident, resulting from paradoxical embolization through an ASD, is a rare complication.

Treatment:

Closure of ASD is indicated after the age of 3 years when:

1 Symptomatic with CHF

2 Volume overload (Qp:Qs >1.5:1)

3 Pulmonary Hypertension

Surgical closure is indicated only in cases where catheter closure is not possible (primum) or failed

ATRIOVANTRICULAR 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 Down's syndrome, so all patients with Down's syndrome (children) should have cardiac evaluation if symptomatic or before 6 months .

Equal no. of males and females are affected**.**

Classification

Partial ;ASD primum ,clefted mitral valve

Complete ;ASD primum ,large inlet VSD ,common single ario-ventricular valve

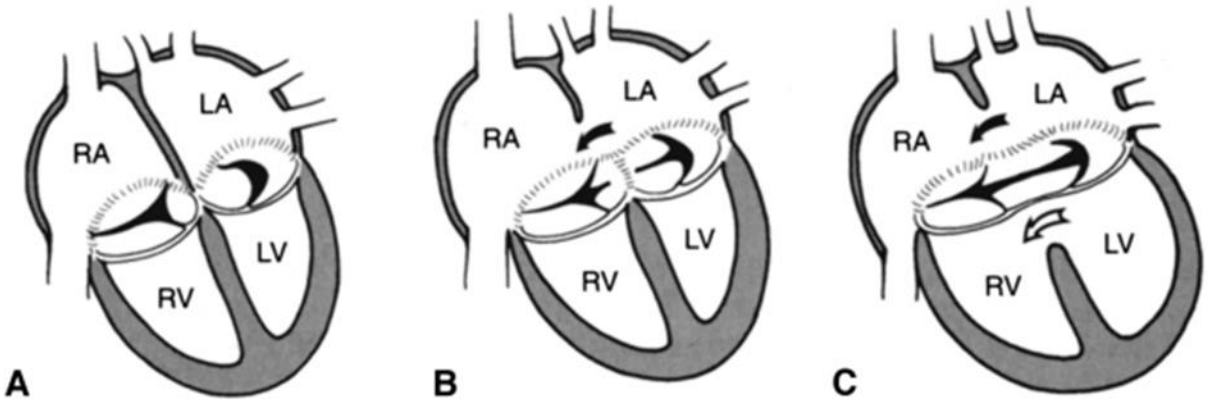


Figure 12-15 *Diagram of the atrioventricular (AV) valve and cardiac septa in partial and complete endocardial cushion defects (ECDs). **A**, Normal AV valve anatomy with no septal defect. **B**, Partial ECD with clefts in the mitral and tricuspid valves and an ostium primum atrial septal defect (ASD) (solid arrow). **C**, Complete ECD. There is a common AV valve with large anterior and posterior bridging leaflets. An ostium primum ASD (solid arrow) and an inlet ventricular septal defect (open arrow) are present. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.*

Clinical features:

Partial: either asymptomatic (mainly) or mild symptoms.

O/E: features of ASD + murmur of mitral regurgitation.

Complete: as the above with heart failure and/or pulmonary vascular disease.

CXR: Cardiomegaly is always present and involves all four cardiac chambers. Pulmonary vascular markings are increased

ECG: 1 “Superior” QRS axis with the QRS axis between -40 and -150 degrees is characteristic of the defect.

2 Most of the patients have a prolonged PR interval (first-degree AV block).

3 RVH or RBBB is present in all cases, and many patients have LVH, too.

Echo: is diagnostic.

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 or total septation), it will cause complete repair.

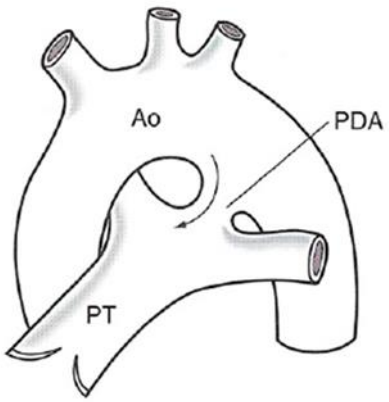
PATENT DUCTUS ARTERIOSUS (PDA)

It is a channel that connect the pulmonary artery with the descending aorta (isthmus part).

It results from the persistence of patency of the fetal ductus arteriosus after birth.

It is the most common lesion in infant of mothers with congenital rubella

PDA more common in females (like ASD).



Patent ductus

Clinical features:

Patients with small PDA usually are “asymptomatic” but large PDA will result in heart failure & pulmonary hypertension along with growth failure.

O/E: Collapsing pulse with pulse pressure

on auscultation classical continuous machinery murmur (systolic & diastolic) at the pulmonary area .

E C G ; In small PDA, it is normal, but large PDA left ventricular hypertrophy (LVH) or biventricular hypertrophy.

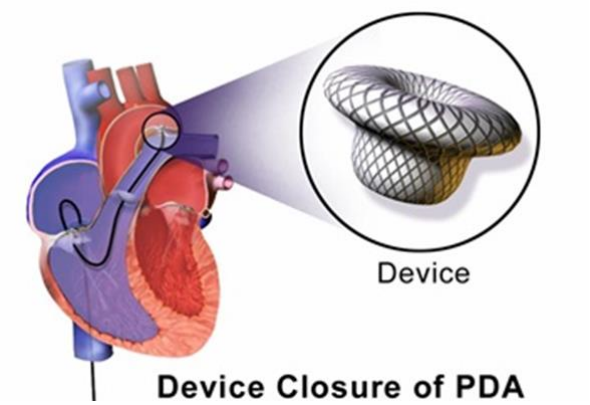
Chest X-ray: Cardiomegaly, plethoric lung, and prominent pulmonary conus .

Echo: is diagnostic

Rx:

“Closure should be done” (6 months – 1 year) whatever the size due to possible complications.

Most PDA closures can be done by transcatheter device or coil and some need surgical closure (when catheter closure is not possible or failed).



Device Closure of PDA