P.T. Management in Congenital Heart Diseases-

Congenital Heart Diseases-

Etiology:- results of aberrant embryonic development of a normal structure. / failure of such a structure to progress beyond an early stage of embryonic / fetal development.

Congenital Heart Diseases:-

- **Acyanotic**
  - **↓ L-R shunt**
    1) ASD
    2) VSD
    3) PDA / Aortopulm. window
    4) Aortic root to Rt heart shunt
    5) Multiple level shunt.
  - **Cont L-R shunt**
    1) Left heart malformations.
    2) Rt heart malformations.
- **Cyanotic**
  - **↑ Pulm. bd. flow**
    1) Complete transposition of great vessels.
    2) Single ventricle
    3) Common atrium
    4) TOF ± pulmonary atresia & red collateral arterial flow.
    5) Tricuspid atresia ± large VSD.
  - **↓ (N) OR ↓↓ pulm. bd. flow**
    1) tricuspid atresia
    2) Ebstein's anomaly.
    3) pulm. atresia ± intact ventricular septum
    4) Pulm. stenosis ± R→L atrial shunt.
    5) Single ventricle ± pulmonic stenosis.
    6) Pulm. A-V fistula.

Acyanotic CHD c L→R shunt:-

1) ASD

a) sinus venosus type occurs high in atrial septum near the entry of SVC

b) ostium primum:- form of AV septal defect that lie immediately adjacent to A-V valves. (seen commonly in Down's syndrome).

c) ostium secundum type:- involves fossa ovalis (mid-septal in location)

- L→R shunt magnitude depends on
  - defect size
  - diastolic properties of both ventricles
  - relative impedence in pulm. & systemic circulations.

→ L-R shunt causes diastolic overloading of RV & ↑ed pulm. blood flow.

→ in pts c ASD:-
  - physical underdevelopment.
  - ↑ed tendency for respi. infections.
  - after 4th decade - atrial arrhythmias.
    - pulm. art. hypertension.
    - bidirectional & even R→L shunting of blood.
    - cardiac failure.

Tt:-

- operative repair ideally in children aged 3-6 yrs.
- Defect is closed usually ε a patch of pericardium or of pp prosthetic material. ε g
- No surgery for small defects/ trivial L→R shunt
- Medical Management:- t/t of
  - Respi. infections.
  - antiarrhythmic medications.
  - usual t/t for HTN, coronary dis. or heart failure.

② VSD

- opening usually single & situated in membranous portion of septum.
- functional disturbance depends on size of defect & status of pulm. vascular bed.
- only small or moderate sized defects seen in adulthood.
- there could could be development of
  - pulm. vascular obstruction
  - RV outflow tract obstruction.
  - aortic regurgitation.
  - infective endocarditis.
- pts ε large VSD & pulm. HTN are at greatest risk for developing pulm. vascular obstruction.

- In pts ē severe pulm. vascular obstruction (Eisenmenger's syndrome) symptoms in adult life -
  - exertional dyspnoea
  - chest pain
  - syncope
  - hemoptysis.
  - R→L shunt leads to cynosis, clubbing & erythrocytosis.

T/t:-

- surgery not recommended for pts ē (N) Pulm. artery pressure ē small shunts (pulm-to-systemic flow ratio of less than 1.5 to 2.0:1.0)
- surgery indicated when there is a moderate to large L→R shunt ē a pulm.-to-systemic flow ratio > 1.5:1.00 & 2.0:1.0.

(3) PDA

- ductus arteriosus leads from bifurcation of pulm. artery to the aorta just distal to the left subclavian artery.
- in this anomaly → pulm. pressures are (N) & a gradient & shunt from aorta to pulm. artery persists throughout cardiac cycle. → leads to

- In adults who were born ⊂ a large L→R shunt thru the ductus, pulm. vascular obstruct° (Eisenmenger syndrome) ⊂ pulm. HTN, R→L shunting & cynosis have usually developed.

- Severe pulm vascular dis. results in reversal of flow thru the ductus, unoxygenated blood is shunted to the descending aorta, & the toes, but not fingers, toes become cynotic & clubbed, a finding termed differential cynosis.

- causes of death:- cardiac failure
  - infective endocarditis.
  - severe pulm. vascular obstruct°

- Severe pulm. vascular obstruction may cause
  - aneurysmal dilatation
  - calcification & rupture of ductus.

H/E:-

In absence of severe pulm vascular dis & predominant L→R shunting of blood, the ductus should be surgically ligated / divided.

④ Aortic root to Rt heart shunt:- Common causes:-
- coronary A-V fistula (between coronary artery & coronary sinus / RA/RV)
- anomalous origin of Left coronary artery from the pulm. trunk.

- Coronary A-V fistula :- shunt usually of small magnitude

complications :-
- infective Endocarditis
- Rupture of aneurysmal fistula
- pulm. HTN.
- congestive failure.

Acyanotic Congenital Heart disease ~~cout~~ a shunt.

I.] Lft heart Malformations :-

1) Congenital Aortic stenosis :-

types of malformations :-
- congenital valvular aortic stenosis.
- discrete subaortic stenosis.
- supravalvular aortic stenosis.

a:) valvular aortic stenosis :-
- more common in males.
- Congenital bicuspid aortic valve, ~~c~~ is not necessarily stenotic is one of most ~~imp~~ common congenital malformations.
- The bicuspid valves may become stenotic ~~c~~ time & be the site of inf. endocarditis, the lesion may be difficult to distinguish in adults from acquired rheumatic / degenerative calcific aortic stenosis.

the dynamics of blood flow associated ē congenitally formed rigid aortic valve leads to thickening of cusps & in later life to calcification.

- Hemodynamically significant obstruc^n causes concentric hypertrophy of LV wall & dilation of ascending aorta.

H(t)- medical- prophylaxis against Inf. endocarditis
- if cardiac reserve ↓ed- digitalis
- diuretics & Na restric^n

- if stenosis severe- avoid strenuous physical activity.
- In critical obstruc^n (aortic valve area < 0.5 cm^2)
→ Aortic valve replacement.
- in asymptomatic children / adolescents / young adults ē critical aortic stenosis ēout valvular calcification → Aortic balloon valvulaplasty.

b.) Subaortic stenosis:- The most common form of subaortic stenosis is idiopathic hypertrophic variety, also kn hypertrophic cardiomyopathy.
- Resembles valvular aortic stenosis.

- Lesion usually consists of a membranous diaphragm or fibrous ring encircling LV outflow tract just beneath base of aortic valve.
- Doppler shows turbulence proximal to aortic valve & also detects & quantitates pressure gradient & severity of aortic regurgitation.

Tt:- excision of membrane / fibrous ridge.

c) Supravalvular Aortic stenosis:-
- Consists of localized or diffuse narrowing of ascending aorta originating just above level of coronary arteries at superior margin of sinuses of valsalva.
- The coronaries are subjected to elevated systolic pressures from the LV & are often dilated & tortuous. & are susceptible to premature atherosclerosis.

2] Co-arctation of Aorta:- Narrowing / constriction of lumen of aorta may occur anywhere along its length but is most common distal to origin of lt subclavian artery.
- Aneurysmal arterial dilation of circle of willis produces high risk of sudden rupture & death.

- Headache, epistaxis, cold extremities, claudication
& eds may occur.
- hypertension in UE & delayed pulsations in femoral arteries are detected.
- enlarged & pulsatile collateral vessels may be palpated in IC spaces anteriorly, in axillae & post. in interscapular area.
- UE & thorax more developed than LE
- ECG reveals LV hypertrophy.
- X ray shows dilated left subclavian artery & (over left mediastinal border) & dilated ascending aorta.
- there is indentation of aorta at site of co-arcta & pre & post- stenotic dilatation (sign of 3) along left paramediastinal shadow.

complications - development of cerebral aneurysm & hemorrhage.
- rupture of aorta.
- premature coronary atherosclerosis.
- LV failure.
- IE.

Tt:- resection & end to end anastomosis

Rt outflow tract obstruction:-

1) pulmonary stenosis c intact ventricular septum

- obstruction to Rv outflow may be localized to supravalvular, valvular or subvalvular levels.
- Multiple sites of narrowing of peripheral pulm. arteries are a feature of rubella embryopathy.
- Severity of obstructing lesion rather than the site of narrowing is the most imp. determinant of clinical course.

In presence of (N) cardiac output, a peak systolic transvalvular pressure gradient between 50 & 80 mmHg is considered to be moderate stenosis, levels below & above that rang are classified as mild & severe resp.

- Symptoms vary c degree of obstruction.
- Fatigue, dyspnoea, Rv failure & syncope may limit activity of older pts., in whom moderate/severe obstruction may prevent an augmentation of cardiac output c exs.
- In pts c severe obstruction, systolic pressure in Rv may exceed that in LV since ventricular septum is intact.

Rv ejection is prolonged c mod./severe stenosis sound of pulm. valve closure is delayed &

Atresia

w hypertrophy reduces compliance of that chamber & forward RA contraction is reqd.

+/t:- balloon valvuloplasty.

CYNOTIC CHD Ē ↑ed pulm. bld flow:-

1) Complete transposition of gr8 vessels.:-

- Aorta rises from RV, pulm. art. from LV.
- some communication must exist between them after birth to sustain life.
- most pts have inter atrial communication.
  - 2/3 have a PDA.
  - 1/3 have VSD.
- pulm vascular obstruction develops by 1-2 yrs of age in pts Ē associated large ventricular septal defect or large PDA in absence of obstruction to ~~outflow~~ LV outflow

+/t:- balloon / blade catheter or surgical creation or enlargement of in interatrial communication in the neonate.

- systemic - pulm. artery anastomosis may be indicated in pt. Ē severe obstruction to LV outflow & ↓ pulm. bld. flow.

- intra-cardiac repair accomplished by re-arranging the venous returns (Mustard or Senning operation) so that the systemic venous blood is directed to the mitral valve & ∴ to LV & pulm. artery, while the pulm. venous blood is diverted through tricuspid valve & RV to aorta.

2) Single ventricle:-

- Both AV valves or a common A-V valve opens to a single ventricular chamber.
- Associated anomalies:- pulmonic valvular or subvalvular stenosis.
  - subaortic stenosis.

Tt:- creation of pathways from systemic veins to pulm. arteries.

CYANOTIC CHD ↓ed pulm. bld. flow:-

1) Tricuspid Atresia:- Atresia of tricuspid valve, an interatrial communication & hypoplasia of RV & pulm. artery. (sometimes VSD also)

- There is severe cyanosis dtb obligatory admixture of systemic & pulm. venous blood in LV
- ECG shows RA enlargement, LV hypertrophy.
- Tt:- Atrial septostomy & palliative operations to ↑ pulm. bld. flow by anastomosis of a systemic artery or vein to a pulm. art.

2) Ebstein's anomaly =
- downward displacement of tricuspid valve into RV, due to anomalous attachment of the tricuspid leaflets.
- tricuspid valve tissue is dysplastic & results in tricuspid regurgitation → this produces an 'atrialized' portion of RV lying between AV ring & origin of the valve ≤ is continuous ē the RA.
- ASD also present.
- cynosis coz of R → L shunt.

t/t:- replacement of tricuspid valve when leaflets are tethered or repair of native valve.

3) TOF :- VSD + obstruction to RV outflow, + overriding of aortic over VSD + RV hypertrophy.

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DISEASES OF THE HEART VALVES.

MITRAL STENOSIS :-

- more in females, - rheumatic in origin generally. rarely congenital.
- in Rheumatic stenosis → valve leaflets diffusely thickened by fibrous tissue &/or calcific deposits.
- Mital commissures fuse, chordae tendinae fuse & shorten cusps become rigid → lead to narrowing.
- calcification of stenotic mitral valve immobilises leaflets & narrows orifice further.
- Thrombus formation & arterial embolisation may arise from calcific valve itself or from dilated LA in pts ē atrial fibrillation.