

An Outline of Congenital Heart Disease

It include the following :-

Pink (Acyanotic)

Blue (Cyanotic)

Critical Left Sided Obstruction

Acyanotic Congenital Heart Disease

Normal pulmonary blood flow

- Valvular lesions
- Coarctation

Increased pulmonary blood flow

- Shunt lesions

Valvular lesions

- Stenosis (Aortic valve ,Pulmonary valve and Mitral valve)
- Regurgitation (Aortic valve ,Mitral valve and Pulmonary valve)
- Stenotic lesions – semilunar valves (Valvar dysplasia)
- Regurgitant lesions – semilunar valves (Valvar dysplasia ,Secondary to intervention for stenotic lesions ,Secondary to endocarditis)
- Mitral valve lesions (AV canal defects complete vs. partial ,Isolated MV lesions)

Coarctation

Narrowing that diminishes the aortic lumen and produces an Obstruction of the aortic arch ,Classically juxtaductal, although may occur anywhere along the aorta and May develop over time Femoral pulses should be checked routinely throughout childhood. Coarctation Characterized by weak femoral pulses, brachio-femoral delay in pulses and a systolic ejection type murmur heard loudest in the back.

Arm-leg blood pressure gradient (arm > leg) {Gradient greater than 30mmHg considered significant ,upper limb hypertension}

Treatment may be surgical or interventional (Balloon dilation or Stent implantation)

Shunt Lesions

Pink, oxygenated blood crosses from the left side of the heart to the right side of the heart ,Signs and symptoms dependent on the size of the lesion and relative vascular resistance – systemic vs. pulmonary ,Significant lesions marked by increased pulmonary flow.

Atrial septal defect (ASD)

Atrial level shunts result in right-sided volume overload ,Wide fixed split S2 characteristic finding of atrial level shunts ,Murmur due to increased flow across the pulmonary and possibly tricuspid valves . Significant symptoms uncommon even with large defects

Treatment either by Cath. Occluder or Surgery

Types:-

Secundum ASD

- Most common
- Defects at the level of the fossa ovalis, presumably secondary to deficiency, perforation or absence of the septum primum

Sinus Venosus ASD

- Involving the portion of septum adjacent to the entry of the systemic veins, outside the fossa ovalis
- Often associated with anomalous pulmonary veins

Primum ASD

- Endocardial cushion defect involving the inferior portion of the atrial septum, adjacent to the AV valves
- usually involving malformations of the AV valves

Ventricular septal defect (VSD)

Can occur anywhere along the ventricular septum .Defects in the perimembranous region are less likely to spontaneously close or even reduce in size and are more likely to require intervention.

Atrioventricular septal defects (AVSD/AV canal/endocardial cushion defects) will generally behave like large VSDs

Symptoms dependent on the size of the defect and the relative vascular resistance between the systemic and pulmonary systems {Congestive heart failure (CHF) signs and symptoms ,Timing of CHF in infants}

Hemodynamically significant lesions result in LA and LV dilation. Cardiac findings dependent largely on the size of the defect. Typical VSD murmur harsh, pansystolic, flat-topped murmur and intensity of the murmur generally inversely proportional to the size of the defect. Very large lesions may result in outflow tract or systolic ejection-type murmurs and possibly a diastolic rumble.

Treatment:- Medical therapy utilized to improved patient symptoms and optimize patient growth by use of diuretics, digoxin, after-load reduction and optimization of feeds .Surgical patch repair the mainstay of treatment of hemodynamically significant lesions

Patent Ductus Arteriosus

It occurs at the level of Great Artery. It results in Continuous or 'machinery-type' murmur ,Hemodynamically significant shunts result in left-sided volume overload ,Larger lesions in infants surgically ligated ,Smaller PDAs in older children generally closed via interventional catheter techniques

Cyanotic Congenital Heart Disease

Right to left shunting resulting in entry of desaturated blood into the systemic arterial system

Increased pulmonary blood flow

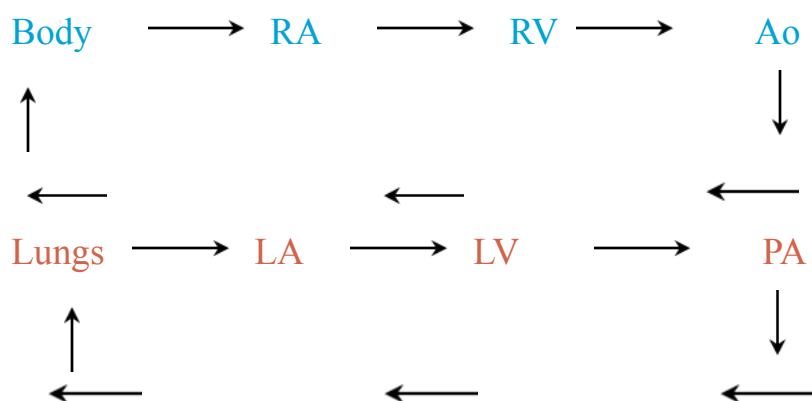
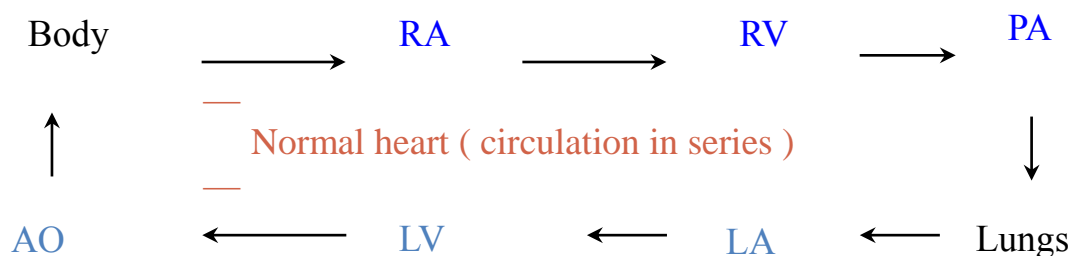
- Truncus arteriosus
- Transposition of the great arteries
- Total anomalous pulmonary venous return

Decreased pulmonary blood flow

- Tetralogy of Fallot/pulmonary atresia
- Tricuspid atresia
- Critical pulmonary stenosis

Transposition of the great arteries

There is anatomic reversal of the relationship of the great arteries: the aorta arises entirely or largely from the RV, and the PA arises entirely or largely from the LV (ventriculoarterial discordant connection)



Normal		D-TGA		L-TGA	
LA	RA	LA	RA	LA	RA
LV	RV	LV	RV	RV	LV
Ao	PA	PA	Ao	Ao	PA
AV concordance		AV concordance		AV discordance	

Treatment options :-

Must bring oxygenated blood into the systemic circulation {Great artery level shunt – PDA, Atrial level shunt – PFO}, Prostaglandin E1 (PGE) {Re-opens and maintains patency of the ductus arteriosus}, Balloon atrial septostomy (BAS) {Increase intracardiac shunting across the atrial septum} and Arterial switch repair {Transection and switching of great arteries, Movement of coronary arteries via tissue buttons, Patching of neo-pulmonary artery, Closure of ASD and Ligation of PDA}.

Total Anomalous Pulmonary Venous Return (TAPVD)

Combined systemic and pulmonary venous returns to the RA and circulates to the systemic and pulmonary systems via the ASD and TV ,Pulmonary veins communicate with systemic vein and fail to connect to left atrium. It results in cyanosis and increased pulmonary flow ,Dilated RA and RV . Reasonably stable in the absence of obstruction along the pulmonary venous pathway .Not a PGE dependent lesion .Surgical correction by Rerouting/baffling of pulmonary veins to the LA.

Truncus arteriosus {common single outflow tract with pulmonary arteries originating from the ascending aorta, abnormal truncal valve ,large VSD ,not a PGE dependent lesion and the Surgical treatment include Closure of VSD to include truncus on LV side with Placement of RV-PA conduit with removal of PA's from the truncus and reanatomosis to the conduit }

Tetralogy of Fallot {Pulmonary stenosis, Overriding aorta, RVH and VSD }

Pulmonary atresia/VSD {Tetralogy of Fallot with atretic pulmonary valve ,Variable pulmonary artery anatomy and Generally a PGE dependent lesion }

Critical pulmonary stenosis {Severe pulmonary stenosis with inadequate pulmonary flow ,Pulmonary atresia/intact ventricular septum and PGE dependent lesion }

Tricuspid atresia {tricuspid atresia , severely hypoplastic RV ,VSD ,ASD – large ,pulmonary stenosis (Variable) and Generally a PGE dependent lesion }.

Cyanotic Heart Disease

Decreased blood flow due to RVOT obstruction may require augmentation of pulmonary blood flow via creation of a surgical systemic to pulmonary shunt called Blalock-Taussig Shunt (BTS)

Blalock-Taussig Shunt (BT Shunt):- It perform to prolong the lives of children with Tetralogy of Fallot and became known as the 'blue baby

operation'. *The original operation* consisted of anastomosing the right subclavian artery to the right pulmonary artery, thus reliably increasing the amount of blood flow to the lungs which would also grow over with the patient. Currently, *a modified BT* shunt is generally performed, using a Gortex tube graft to anastomose the subclavian artery to the pulmonary artery and is still often used for children with Tetralogy of Fallot as well as other cyanotic heart lesions where pulmonary blood flow is limited.

Critical Left-Sided Obstruction

Neonatal presentation

- Critical aortic stenosis
- Coarctation
- Hypoplastic left heart syndrome

Cardiogenic shock

- PGE dependent lesions

Coarctation of the aorta:- Critical narrowing of the “juxtaductal” aorta, Blood cannot get past the obstruction, Characterized by weak or absent pulses particularly in the lower limbs and Initiation of PGE lifesaving. Surgical correction following initiation of PGE and stabilization.

Critical Aortic Stenosis:- Critical Inadequate forward flow to maintain cardiac output may lead to shock and it characterized by Weak pulses throughout, Low blood pressure, Variable murmur and Acyanotic. PGE dependent lesion. The treatment include Balloon valvuloplasty or surgical valvuloplasty

Hypoplastic Left Heart Syndrome (HLHS):- It characterized by Mitral atresia, Aortic atresia, Hypoplastic left ventricle and Hypoplastic ascending aorta. Initially cyanotic. With closure of the PDA lead to SHOCK (Tachycardia, tachypnea, low blood pressure, weak pulses, poor perfusion, cyanotic/grey colour). It need PGE. The surgical treatment include Norwood procedure (Stage I single ventricle palliation) and Heart Transplant.