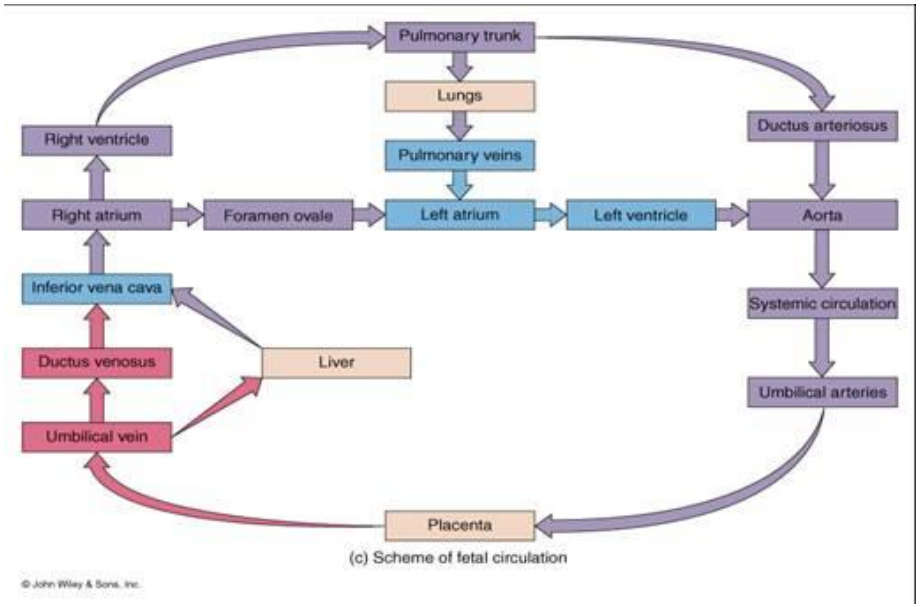
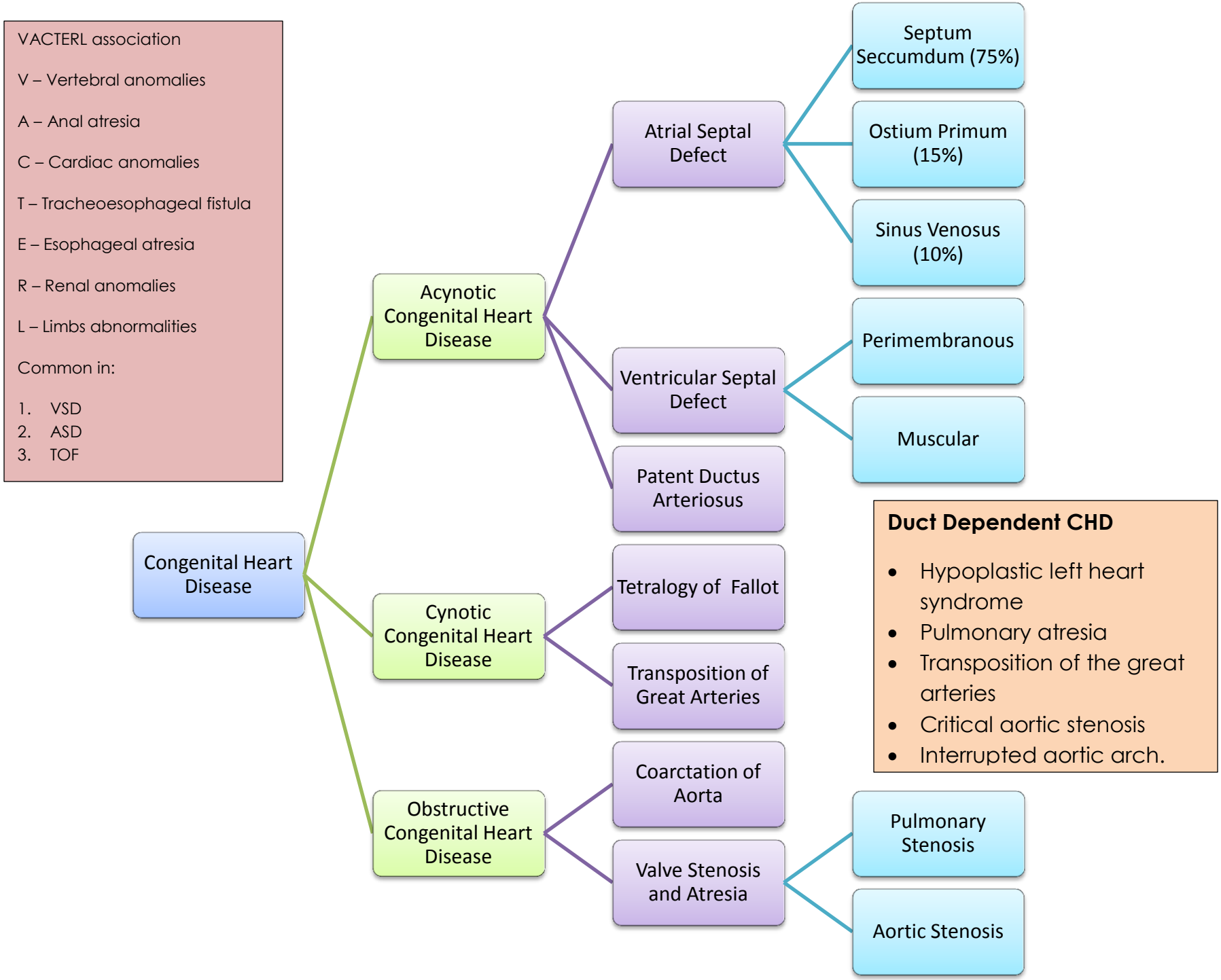
Pediatrics

Congenital Heart Disease

Fetal Circulation

1. Lest atrium has low pressure due to low blood return from lungs
2. Right atrium has higher pressure because it receives blood from
    a. Systemic venous return
    b. Placenta (umbilical vein)
3. Because of this, flap valve of Foramen Ovale is held open leading to flow of blood across the atrium septum from right atrium to left atrium, then into left ventricle and being pumped to the rest of fetal body
4. With first breath;
    a. Pulmonary vascular resistance falls
    b. Volume of blood flowing into the lungs increases by six folds
5. Results in the rise of left atrial pressure. Meanwhile blood return into the right atrium falls as placenta has been excluded after delivery
6. Pressure changes lead to closure of flap valve of Foramen of Ovale.
7. Ductus Arteriosus will eventually close after few hours to days following delivery




VACTERL association
V – Vertebral anomalies
A – Anal atresia
C – Cardiac anomalies
T – Tracheoesophageal fistula
E – Esophageal atresia
R – Renal anomalies
L – Limbs abnormalities
Common in:
1. VSD
2. ASD
3. TOF
Congenital Heart Disease
Acynotic Congenital Heart Disease
Cynotic Congenital Heart Disease
Obstructive Congenital Heart Disease
Atrial Septal Defect
Septum Seccumdum (75%)
Ostium Primum (15%)
Sinus Venosus (10%)
Ventricular Septal Defect
Perimembranous
Muscular
Patent Ductus Arteriosus
Tetralogy of Fallot
Transposition of Great Arteries
Coarctation of Aorta
Valve Stenosis and Atresia
Pulmonary Stenosis
Aortic Stenosis
Duct Dependent CHD
Hypoplastic left heart syndrome
Pulmonary atresia
Transposition of the great arteries
Critical aortic stenosis
Interrupted aortic arch.

Congenital heart disease presents with

1. **Antenatal Cardiac Ultrasound Diagnosis**
    a. 70% infant who need surgical intervention before the age of 6 months being diagnosed antenatally
    b. Detail fetal scan done routinely at 18-20 weeks of gestation
        i. Any abnormality, refer to pediatric cardiologist for Echocardiography
        ii. Echo also been done in high risk patient
            1. Suspected Down's syndrome baby
            2. Mother with congenital heart disease
            3. Mother who had history of delivering baby with congenital heart disease
2. **Cardiac Murmur**
    a. Not all murmurs are of congenital heart disease
    b. 30% of infant have innocent murmur. **Innocent murmur hallmarks are 4S's**
        i. As**S**ymptomatic
        ii. **S**ystolic murmur
        iii. **S**oft blowing murmur
        iv. Left **S**ternal edge
    c. Other hallmarks of innocent murmur include
        i. Normal heart sound with no added sound
        ii. Absence of thrill
        iii. Absence of radiation
    d. Flow murmur may be present in
        i. Fever
        ii. Anemia
3. **Heart Failure**
    a. **Clinical manifestations**
        i. **Symptoms**
            1. Breathlessness (often during feeding)
            2. Sweating
            3. Poor feeding
            4. Recurrent chest infection
        ii. **Signs**
            1. Failure to thrive
            2. Tachypnea
            3. Tachycardia
            4. Cardiac murmur
            5. Gallop rhythm
            6. Cardiomegaly; displaced apex beat
            7. Hepatomegaly
            8. Pulmonary edema; bibasal crepitation
            9. Cold periphery
    b. **Eisenmenger Syndrome**
        i. Irreversible raised in pulmonary vascular resistance
        ii. Leads to increase pulmonary pressure and flow
        iii. Shunting of blood from previously left to right, now become right to left
4. **Shock**
5. **Cyanosis**

Acynotic Congenital Heart Disease			
Types	**Clinical Features**	**Investigations**	**Management**

Atrial Septal Defect	**Symptoms**
	• None (commonly)
	• Recurrent chest infection
	• Arrhythmias (during adulthood)
	Signs
	• Fixed and widely split second heart sound
	  o Equal right ventricular stroke volume during inspiration and expiration
	• Ejection systolic murmur
	  o Best heard at upper left sternal edge (increase pulmonary blood flow)
		Chest X-ray
		• Cardiomegaly
		• Enlarged pulmonary arteries
		• Plethoric lungs field
		ECG
		• Right bundle branch block
		• Right axis deviation
		Echocardiography
		• Delineate anatomy
		• Mainstay of investigation
			Large ASD
			• Cardiac catheterization
			  o Occlusion devices
			
			Partial AVSD
			• Surgical correction done at 3-5 years of age
			• To prevent cardiac failure and arrhythmias in later life

Ventricular Septal Defect	**Small VSD ≤ 3mm**
	Symptoms
	• Asymptomatic
	Signs
	• Loud pansystolic murmur
	  o Lower left sternal edge
	• Quite p2 sound
	
	Large VSD >3mm
	Symptoms
	• Heart failure
	• Recurrent chest infection
	Signs
	• Tachypnea
	• Tachycardia
	• Active precordium
	• Soft pansystolic murmur
	• Apical mid-diastolic murmur
	• Load P2 sound
		Small VSD
		• Normal chest X-ray, ECG
		• Echocardiogram
		  o Detect anomaly
		  o Hemodynamic change detect using Doppler U/S
		  o Absence of pulmonary hypertension
		Large VSD
		• **Chest Xray**
		  o Cardiomegaly
		  o Dilated pulmonary arteries
		  o Plethoric lung fields
		  o Pulmonary edema
		• **ECG**
		  o Biventricular hypertrophy by the age of 2 months
		• **Echo**
		  o Mainstay of investigation
			• **Medical**
			  o Diuretics
			  o Captopril
			• Additional calorie intake
			• Surgical intervention at 3-6 months of life because
			  o Prevent heart failure
			  o Prevent Eisenmenger syndrome

Patent Ductus Arteriosus
• Failure to close of Ductus Arteriosus byv 1 month after expected date of delivery
	Symptoms
	• Assymtomatic
	Signs
	• Continuous murmur
	  o Beneath clavicle
	  o Radiated to the back
	• Increased pulse pressure
	• Collapsing/ bounding pulse
		Chest X-ray
		• Cardiomegaly
		• Dilated pulmonary arteries
		• Plethoric lungs field
		ECG
		• Can be left ventricular hypertrophy due to left to right shunt
		• Can be right ventricular hypertrophy in ES
			Closure is recommended to prevent
			• Bacterial endocarditis
			• Pulmonary vascular disease
			Medical treatment
			• NSAIDs – Indomethecin
			Surgical
			• Cardiac catheterization – occlusive devices at 1 year of age

Cyanotic Congenital Heart Disease			
Types	**Clinical Features**	**Investigations**	**Management**

Tetralogy of Fallot
• Large VSD
• Overriding Aorta
• Subpulmonary stenosis
• Right ventricular hypertrophy
	Symptoms
	• Hypercynotic spell
	  ○ Rapid increase in cyanosis
	  ○ Irritable
	  ○ Inconsolable crying
	  ○ Breathlessness
	  ○ Pallor due to acidosis
	• Squatting on exercise
	Signs
	• Clubbing of fingers and toes in older children
	• Loud harsh ejection systolic murmur
	  ○ Left sternal edge at first day of life
		Chest X-ray
		• Relatively small heart
		• Uptilted apex (boot shaped)
		• Pulmonary artery bay
		• Oligemic lung fields
		ECG
		• Right ventricular hypertrophy in older children
		Echo
		• Demonstrates cardinal signs
		• Cardiac catheterization may be required to show detailed anatomy of coronary artery
			• **Definitive treatment at 6 months of life**
			  ○ Closing of VSD
			  ○ Dilation of pulmonary stenosis
			• **Neonatal cyanosis**
			  ○ Modified Blalock Taussig shunt
			  ▪ Artificial tube between subclavian and pulmonary arteries
			  ○ Dilation of pulmonary stenosis
			• **Hypercyanotic spell more than 15 minutes**
			  ○ Sedation and pain relief (morphine)
			  ○ IV propranolol
			  ○ Fluid resuscitation
			  ○ Bicarbonate to relief acidosis
			  ○ Intubation may be required

Transposition of Great Arteries
• Aorta connected to right ventricle
• Pulmonary artery connected to left ventricle
• Incompatible with life, only compatible if has
  ○ VSD
  ○ ASD
  ○ PDA
	Symptoms
	• Profound cyanosis
	  ○ At 2 days of life after closure of ductus arteriosus
	  ○ Less severe with has associated anomalies
	Signs
	• Cyanosis
	• Load single second heart sound
	• Usually murmur is absent
		Chest X-ray
		• Egg on side cardiac contour
		• Narrow pedicle
		• Plethoric lung fields
		ECG
		• Usually normal
		Echo
		• Demonstrate abnormal arterial connection
		• Other associated abnormalities
			• Maintain patency of DA
			  ○ Misoprostol
			• Balloon arterial septostomy
			• Surgical switching of arteries is done as early as neonatal life

Obstructive Congenital Heart Disease			
Types	**Clinical Features**	**Investigations**	**Management**

Aortic Stenosis
• Can be associated with
  ○ Mitral valve stenosis
  ○ Coarctation of aorta
	Symptoms
	• Severe heart failure symptoms and shock
	Signs
	• Small volume
	• Slow rising pulse
	• Carotid thrills
	• Ejection systolic murmur
	  ○ Upper right sternal edge
	  ○ Radiated to the neck
	• Delayed and soft A2 sound
	• Apical ejection click
		Chest X-ray
		• Prominent left ventricle
		• Post-stenotic ascending aortic dilation
		ECG
		• Left ventricular hypertrophy
		Echo
			• Palliative during early age, until childhood
			• Balloon valvotomy with pressure gradient >64mmHg
			• Valve replacement

Pulmonary Stenosis	**Symptoms**
	• Asymptomatic usually
	Signs
	• Ejection systolic murmur
	  ○ Upper left sternal edge
	  ○ Thrill may present
	• Ejection click at upper left sternal edge
	• Parasternal heaving
		Chest X-ray
		• Post- stenotic pulmonary dilation
		ECG
		• Right ventricular hypertrophy
		Echo
			• Transcatheter balloon dilation when pressure gradient >64mmHg

Coarctation of Aorta	**Symptoms** • Severe heart failure and shock after 2 days of life due to closure of duct **Signs** • Sick baby • Severe heart failure • Absent femoral pulse • Radio-femoral delay • Severe metabolic acidosis	**Chest X-ray** • Cardiomegaly • Rib notching • Collateral arteries in teenagers **Normal ECG**	• Surgical repair