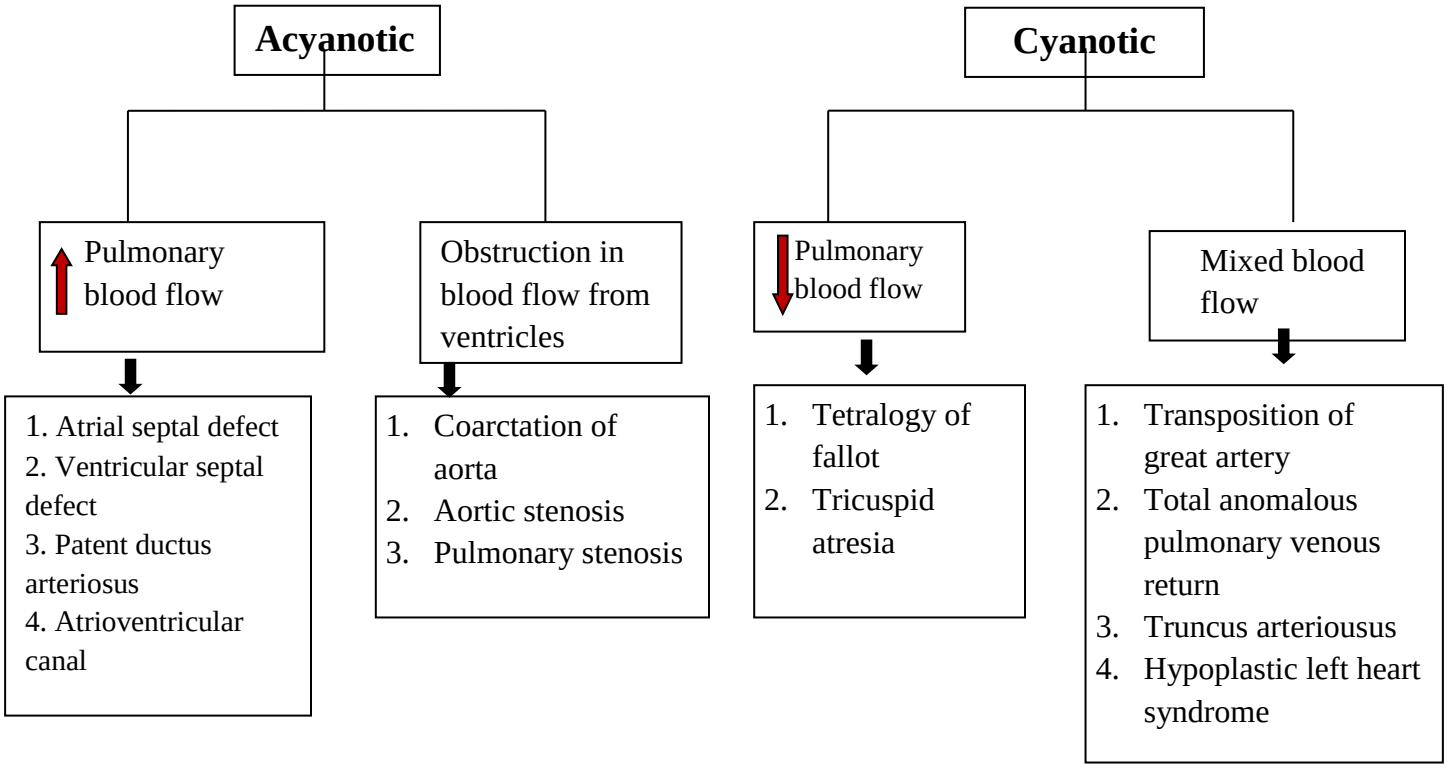
CONGENITAL HEART DISEASE

Congenital heart disease is the structural malformation of the heart or great vessels present at birth, it is the most common congenital malformations.

Etiology

- The exact cause of CHD is unknown in about 90 percent of cases.
- Heredity and consanguineous marriages are important etiological factors.
- Genetic disorders and chromosomal aberrations (trisomy 21, Turner's syndrome) are also known to predispose congenital heart disease.
- Other associated factors responsible for CHD include fetal and maternal teratogenic infections (rubella), teratogenic drug(thalidomide) intake, alcohol intake by the mother and irradiation in the first trimester of pregnancy,
- Maternal IDDM,
- High altitude,
- Fetal hypoxia,
- Birth asphyxia.

CLASSIFICATION OF CHD



CYANOTIC HEART DISEASE

I. Cyanotic heart disease with decreased pulmonary blood flow

In these defects there is some obstruction to the pulmonary blood flow on the right side of the heart and an abnormal communication between the right and left sides of the heart. The obstruction causes pressure in the right side of the heart to be abnormally high and blood therefore shunts from the

right to the left through the abnormal communication. This results in deoxygenated blood perfusing the systemic circulation and causes cyanosis.

1. TETRALOGY OF FALLOT

Introduction

Tetralogy of Fallot (TOF) is a congenital heart defect which is classically understood to involve four anatomical abnormalities (although only three of them are always present).

- 1672: Nicholas Steno, a 17th century Danish scholar who was equally famous as anatomist, geologist and theologian originally described fallot's tetralogy.
- In 1888, Etienne fallot, Louis Arthur described the four anatomical features of Fallot's tetralogy.
- Prof .Edward sandifast of Leyden described the clinical and anatomical featuers in a 12 year old toy.
- In 1947, Taussig described the various positions of the dyspnoea.

Incidence

- It is the most common cyanotic heart defect, representing 55-70%, and the most common cause of blue baby syndrome.
- It is commonest comprising 10% of congenital heart disease.
- Tetralogy of Fallot occurs in approximately 400 per million live births.
- It occurs slightly more often in males than in females

Definition

It is one of the commonest cyanotic congenital heart disease consisting of

1. Pulmonary stenosis (usually subvalvular)
2. Ventricular septal defect
3. Right ventricular hypertrophy and
4. Overriding of aorta.

Etiology:

The causes of most CHDs are unknown, although genetic studies suggest a multifactorial etiology (environmental or genetic factors or a combination)

- Certain conditions or factors that occur during pregnancy may raise the risk for having a child with tetralogy of Fallot These conditions and factors include:
  - German measles (rubella) and some other viral illnesses
  - Poor nutrition
  - Overuse of alcohol
  - Age (being older than 40)
  - Diabetes
- Mothers who take medications to control seizures (phenytion, valporic acid) and mothers with phenylketonuria (PKU) are also more likely to have a baby with tetralogy of Fallot.
- Heredity may play a role in causing tetralogy of Fallot. An adult who has tetralogy of Fallot may have an increased chance of having a baby with the condition.
- Children who have certain genetic disorders, such as Down syndrome and DiGeorge syndrome, often have congenital heart defects, including tetralogy of Fallot.

The syndromes that are associated with Fallot's tetralogy are:

1. Apert's syndrome: TOF, VSD, CA
2. Asymmentric Crying Face: TOF, VSD

3. Goldenhan Syndrome: Occulo-Cerebrovertebral ,TOF, VSD, ASD.
4. Silver Syndrome: TOF, VSD.
5. Delange Syndrome: VSD, TOF, PDA, DORV.

- Maternal drug associated with fallot's tetralogy:
  - Trimethadione - TGA, TOF, Hypoplastic left ventricles.
  - Thalidomide - TOF, VSD, ASD, truncus arteriosus.
- Embryology studies show that it is a result of anterior malalignment of the aorticopulmonary septum, resulting in the clinical combination of a VSD, pulmonary stenosis, and an overriding aorta. Right ventricular hypertrophy results from this combination, which causes resistance to blood flow from the right ventricle

CLASSIFICATION OF FALLOT'S TETRALLOGY:

Group I: *Severe fallot's tetralogy*- is associated wtih severe pulmonic stenosis or pulmonary atresia. The patient is severely cyanotic at birth and majority of patients die if there is no collateral blood supply.

Group II; *classical fallot's tetralogy* - the patient becomes symptomatic form six months of age.

Group III: *Pink fallot's tetralogy* - patient becomes symptomatic in between 4-5 years of age.

Associated anomalies:

In addition, tetralogy of Fallot may present with other anatomical anomalies, including:

1. ASD
2. PDA
3. Absence of pulmonic valves.
4. Absence of left pulmonary artery.
5. Aortic regurgitation
6. Persistence of left superior vena cava drains in to coronary sinus and sometimes drains into left atrium and the patient always has cyanosis even after surgical correction.
7. Conus coronary : an aberrant branch arises from right coronary artery passes through the infundibulum which has to be preserved during surgery.
8. Right sided aortic arch seen in 25% of the patients of fallot's tetralogy. Right sided aortic arch is associated with severe fallot tetralogy.
9. TAPVR.

Pathophysiology

- ✓ Due to structural defect there is right to left shunt causing cyanosis. The most vital abnormalities are pulmonary stenosis and VSD.
- ✓ Obstruction of blood flow from the right ventricle due to pulmonary valve stenosis results in shunting of deoxygenated blood through the VSD into the left ventricle, then to the aorta causes cyanosis.(degree of cyanosis depends on right ventricular outflow tract obstruction and the size of the VSD)

- ✓ Pulmonary stenosis causes reduced pulmonary blood flow and an increase in work load for the right ventricle, resulting in right ventricular hypertrophy.
- ✓ Finally the condition is complicated by persisitent arterial unsaturation, poor pulmonary vascularity, polycythemia to compensate cyanosis.

Pathological anatomy: the disease, fallot's tetralogy can also be called fallot's duology, where the main defects are pulmonic stenosis and VSD.

1.**Pulmonary stenosis**

- A narrowing of the right ventricular outflow tract and can occur at the pulmonary valve (valvular stenosis) or just below the pulmonary valve (infundibular stenosis).
- Infundibular pulmonic stenosis is mostly caused by overgrowth of the heart muscle wall.
- The pulmonic stenosis is the major cause of the malformations, with the other associated malformations acting as compensatory mechanisms to the pulmonic stenosis.
- The degree of stenosis varies between individuals with TOF, and is the primary determinant of symptoms and severity.

2. Overriding aorta

- An aortic valve with biventricular connection, that is, it is situated above the ventricular septal defect and connected to both the right and the left ventricle.
- The degree to which the aorta is attached to the right ventricle is referred to as its degree of "override."
- The aortic root can be displaced toward the front (anteriorly) or directly above the septal defect, but it is always abnormally located to the right of the root of the pulmonary artery.

1. The enlarged right ventricle drags the aorta from the left side to right side.
2. Left ventricular pressures and size are decreased due to reduction in blood which yields to the pressure of right ventricles.

3. ventricular septal defect (VSD)

A hole between the two bottom chambers (ventricles) of the heart. The defect is centered around the most superior aspect of the ventricular septum (the outlet septum), and in the majority of cases is single and large.

4. Right ventricular hypertrophy

- The right ventricle is more muscular than normal, causing a characteristic boot-shaped appearance as seen by chest X-ray.
- The right ventricular wall increases in size to deal with the increased obstruction to the right outflow tract. This feature is now generally agreed to be a secondary anomaly, as the level of hypertrophy generally increases with age. It is due to pulmonic stenosis.

Development of collaterals:

There are three types of developmental collaterals mainly seen in severe fallot's tetralogy associated with severe pulmonic stenosis or pulmonary stenosis.

Type I: collaterals develop in between the bronchial artery.
Type II: collaterals directly arise from aorta and connect the pulmonary artery branches.
Type III: indirectly collaterals – they arise from the branches of aorta mainly from subclavian artery and connect the pulmonary artery branches.

Pathology of lungs:
The size of the lungs is decreased, and the sizes of the alveoli are also decreased due to insufficient blood flow through the pulmonary artery.

CLINICAL MANIFESTATIONS:
Infants:

- Some infants may be acutely cyanotic at birth; others may have mild cyanosis that progresses over the first year of life as the pulmonic stenosis worsens.
- There is a characteristics murmur.
- There are acute episodes of cyanosis and hypoxia, called blue spells or tet spells.
- Anoxic spells occur when the infant's oxygen requirements exceed the blood supply, usually during crying or after feeding.
- Patients are at risk for emboli, seizures, and loss of consciousness or sudden death following an anoxic spell.

Signs and Symptoms:

1. Dyspnoea: develops at 6 months of age in majority of cases. It is not associated with cough.
2. Cyanosis: may be present from the birth in severe fallot's tetralogy or it may be present from 6 months of age in classical fallot's tetralogy.
3. Cyanosis aggravated by exertion.
4. A common clinical manifestation years ago was squatting, a posture characteristically assumed by older children to increase systemic vascular resistance and to encourage increased pulmonary blood flow. Squatting is rarely seen currently because TOF is now surgically repaired during the first year of life (*reasons for squatting – when the patient is walking or doing exercise, the peripheral vascular resistance is decreased due to vasodilation resulting in more right to left shunt and the patient becomes cyanotic).*
   Mode of relief
   a. The right to left shunt is decreased due increase of peripheral vascular resistance due to folding of vessels if the patient squats.
   b. Decreased flow of impure blood with metabolites from lower limbs of the heart.
5. *Cyanotic spells*: the child is severely cyanosed due to exertion and the patient may develop convulsions and loss of consciousness.
6. Growth retardation decreases height & weight
7. Hypoxia

8. Associated chromosomal anomaly.
9. On examination of the patient: marfan or down's syndrome features
10. Cardiac enlargement and CCF

Diagnostic evaluation:

- **X- ray chest**:

1. *Boot shaped heart* due to the small pulmonary arteries and right ventricular hypertrophy
2. Right atrial and ventricular dilatation and increased pulmonary marking.

- **ECG:**

1. Right ventricular hypertrophy and right axis deviation.

- **Doppler echo:** in addition to above defects, the pressure gradient in between right ventricle and pulmonary aretery and the amount of blood flow in to the aorta can be assessed. Other associated anomalies can be identified.
- **Cardiac catheterisation**:

1. The catheter can be negotiated from right atrium in to right ventricle and left atrium through VSD.
2. The pressure in right ventricle is increased to systemic level.
3. The pressure in pulmonary artery is decreased.
4. The oxygen content in the aorta is decreased.

- **Angiocardiography**: the amount of blood flow in to aorta and pulmonary artery and the severity can be assessed.
- **Blood examination**:
    1. polcythemia is present
    2. if the RBC count exceed more than 8 millions or if the hematocrit exceeds more than 60% the patients can develop spontaneous thrombosis.
    3. Microcytic, hypochromic anaemia may be present.

MANAGEMENT

If the patient is severely cyanosed with group I Fallot's , the patient should be given prostaglandin infusion to keep the ductus arteriosus patent for augmentation of blood supply to pulmonary artery.

SURGICAL MANAGEMENT

Palliative shunt: in infants who cannot undergo primary repair, a palliative procedure to increase oxygen saturation may be performed. The preferred procedure is the **Blalock-Taussing** or **Modified Blalock- Taussing shunt**, which provides blood flow to the pulmonary arteries from the left or right subclavian artery. In general , however, shunts are avoided because they may result in pulmonary artery distortion.

1. **Blalock Taussig operation**:

Anastomosis is done in between subclavian artery with ipsilateral pulmonary artery. In left sided aortic arch the anastomosis is done in between right subclavian artery and pulmonary artery.

In right aortic arch- left subclavian artery and pulmonary artery

2. **Gore tex shunt**: interposition shunt is placed in between the subclavian artery and ipsilateral pulmonary artery is the treatment of choice in children.

a. Left-sided shunt for left side aorta.
b. Right-sided shunt for right side aorta

3. **Waterston's operation**: anastomosis is done in between the ascending aorta and pulmonary artery. This operation is abandoned because of complications.

4. **Pott's operation**: anastomosis is done between ascending aorta and left branch of pulmonary artery. It is not preferred because of its complications.

PERMANENT OPERATION / COMPLETE REPAIR:

Elective repair is usually performed in the first year of life. Indications for the repair include increasing cyanosis and the development of hypercyanotic spells. Complete repair involves the closure of the VSD and resection of the infundibular stenosis, with a pericardial patch to enlarge the right ventricular flow tract. The procedure requires a median sternotomy and the use of cardiopulmonary bypass.

The broad steps are:

1. Resection of infundibular area taking care not to injure conus coronary.
2. Closure OF VSD.
3. Trabeculectomy.

INDICATIONS

1. In symptomatic patients- the total correction can be done in between 3 to 4 months for proper development of pulmonary artery and lungs.
2. Asymptomatic and mildly cyanosed patient- correction can be done in between 3-24 months.
3. Mildly cyanosed patient with Blalock Taussig shunt – total correction can be done in between 1-2 years
4. Asymptomatic and acyanotic patient –total correction can be done in between 1-2 years of age.

Post operative complication

- Residual shunt
- Residual right ventricular outflow obstruction
- Poor ventricular function
- Dysrhythmias

PROGNOSIS:

the operative morality for total correction of TOF is less than 5%. With improved surgical techniques there is a lower incidence of dysrrhythmias and sudden death.; surgical heart block is rare. CHF may occur postoperatively

2. TRICUSPID ATRESIA

Tricuspid atresia is the congenital absence of tricuspid valve resulting in no communication between the right atrium and the right ventricle. The right ventricle is hypoplastic so the total systemic venous return enters the left heart by means of foramen ovale or an ASD, resulting cyanosis.

Incidence

Tricuspid atresia accounts for 1% to 3% of CHDs, it is the third most common CHD.

PATHOPHYSIOLOGY

- It is a complex lesion with many variations
- In this lesion the tricuspid valve does not develop
- An ASD or patent foramen ovale must be present for the fetus or infant to survive
- The right ventricle is hypoplastic (underdeveloped)
- The VSD can be of varying size
- The pulmonary artery may be in normal position or transposed with aorta
- There may be pulmonary stenosis of varying degrees
- The new born may depend on the ductus arteriosus for pulmonary blood flow

ALTERED HEMODYNAMICS

- The desaturated blood enters the right atrium and is shunted right to left through the patent foramen ovale/ ASD into the left atrium, since it cannot flow into the right ventricle because the tricuspid valve is arteric or absent
- In the left atrium desaturated blood mixes with the saturated blood
- From the left atrium it flows through the mitral valve, into the left ventricle.
- Some of the mixed saturated blood flows out the aorta and to the systemic circulation
- Some will flow through the VSD and into the right ventricular chamber, into the pulmonary artery, and to the lungs to become oxygenated

  For children with severe pulmonary stenosis and no VSD or other complex anatomy, the PDA is critical to ensure pulmonary blood flow.

CLINICAL MANIFESTATIONS

Clinical presentation depends on the state of the pulmonary blood flow, which may be diminished or increased**.**

- Cyanosis is usually seen in the new born period.
- A single first heart sound is heard because there is no closure of the tricuspid valve.
- A systolic murmur of the VSD or a PDA murmur may be heard (if patent)

- Increased respiratory effort
- Metabolic acidosis
- Tachycardia and dyspnea
- Feeding difficulties.
- In older children, cyanosis, polycythemia, easy fatigability, exertional dyspnea, and chronic hypoxemia with clubbing occurs

Diagnostic Evaluation

- Auscultation: murmurs vary depending on the associated lesions; single second heart sound.
- Chest X-ray: decreased pulmonary vascular markings
  - Left ventricular, right ventricular and right atrial hypertrophy.
- ECG: Left ventricular, right ventricular and right atrial hypertrophy.
- Two-dimensional echocardiogram identifies the atretic tricuspid valve and hypoplastic RV; Doppler study and color flow mapping documents the right-to-left atrial shunt and the size of the PDA or VSD.
- Cardiac catheterization may be necessary to delineate anatomy.

MANAGEMENT

Medical Management

- For the neonate whose pulmonary blood flow depends on the patency of the ductus arteriousus, a continuous infusion of *prostaglandin E_1* is started at 0.1 mg/kg of body weight per minute until surgical intervention can be arranged.
- Intubate and ventilate as needed.
- Inotropic support as needed.
- Infective endocarditis prophylaxis (lifelong).

Surgical Management

Surgical management of tricuspid atresia is often done in stages

Palliative surgery

- This is the FIRST surgery in a three stage effort to palliate this defect
  - The first stage involves early pulmonary arterial band (usually under the age of 3 months)for patient who have increased pulmonary blood flow.
  - Modified blalock Taussing shunt (BT shunt) is indicated in neonates if pulmonary blood flow is insufficient. This involves:
    - Dacron ^{R} or Gore-Tex^{R} conduit from right or left subclavian artery to corresponding pulmonary artery.
    - Blood flow from the subclavian artery will therefore flow to the pulmonary system and increase pulmonary blood flow and oxygenation.
  - No treatment is required if pulmonary blood flow is balanced.
- SECOND surgery: ages 4 to 9 months of age once the pulmonary vascular resistance has decreased to normal pressure

- Bidirectional Glenn shunt: end-to-side anastomosis of the SVC to the right Pulmonary artery. This surgery allows effective palliation until the age of 4-6 years

- THIRD surgery: ages 18 months to 3 years:
  - Modified Fontan procedure: is required for the elimination of cyanosis. Here all the systemic venous return (from both vena cava) is routed to the pulmonary artery.

Post operative complications

- CHF.
- Dysrhythmias
- Systemic venous hypertension
- Ventricular dusfunction
- Persistent pleural effusion (especially after Stage II and Stage III repairs).
- Thrombus formation in the systemic venous system.
- Infective endocarditis.
- Rarely, heart block.
- Protein-losing enteropathy

PROGNOSIS

Surgical mortality is less than 5%, the rate increase when the anatomy is more complicate and other risk factors are present

3. TRANSPOSITION OF THE GREAT ARTERIES

Definition

Transposition of the great arteries (TGA) occurs when the PA arises from the left ventricle and the aorta arises from the right ventricle.

INCIDENCE

It accounts for 5% to 7% of CHDs. Associated lesions include VSD, ASD, PDA, Pulmonary stenosis, and Coarctation of aorta. It is more common in males than in females. About 45% of affected children have a coexisting VSD. Incidence is significantly high in the history of diabetes in grandparents and babies having large birth weight.

ETIOLOGY

- Improper septation and rotation of the common truncal vessel in the fetal life causes this defect

ALTERED HEMODYNAMICS

- In this defect the pulmonary and systemic circulations exist in parallel
- Desaturated systemic venous blood returns to the right atrium, flows into the right ventricle, and pumps the desaturated blood into the aorta and back to the body

- Saturated pulmonary venous blood returns to the left atrium(from the lung) flows into the left ventricle, and is pumped through the pulmonary artery and back through the lungs
- Survival depends on the mixing of these two circulations through the fetal structures- the foramen ovale and ductus arteriosus.
- This allows oxygenated blood to be delivered to the body and the deoxygenated blood to return to the lungs for oxygenation.

CLINICAL MANIFESTATIONS

Symptoms evident soon after birth; clinical scenario is influenced by the extent of intercirculatory mixing.

- Infants with minimum communication are severely cyanotic and have depressed function at birth.
- Hypoxemia with minimal response to oxygen administration.
- Tachypnea.
- Cardiomegaly
- Metabolic acidosis.
- CHF.
- Feeding difficulties.

DIAGNOSTIC EVALUATION

- ***Auscultation***: varies; no murmur, or a murmur related to an associated defect, single S_2(second sound).
- ***Chest X-ray:***
  - varies; neonate chest X-ray usually normal
  - Cardiomegaly with a narrow mediastinum and plethoric lung fields.
  - The cardiac silhouette has an egg on side appearance
- ***ECG***: RVH or biventricular hypertrophy.
- ***Two-dimensional echocardiogram*** with Doppler study and colour flow mapping identifies the structural abnormalities: transposed vessels, coronary artery pattern, and degree of mixing across the atrial septum plus associated lesions.

MANAGEMENT

Medical Management

- A continuous PGE_1 infusion is begun to maintain ductal patency and support mixing of oxygenated and deoxygenated blood at the level of ductus and provide an oxygen saturation of 75% or to maintain cardiac output
- ***Balloon Atrial septostomy (Rashkind's procedure)*** via interventional cardiac catheterisation may be performed on some infants if there is no adequate intraatrial mixing to enhance mixing of blood It is successful only upto the age of 6-12 weeks [a balloon tipped catheter is inserted in the femoral vein and guided through the right atrium and foramen ovale to the left atrium. The balloon is then inflated with saline and the catheter is pulled back to the right atrium. This results in a man made atrial septal defect through which oxygenated blood will shunt from left to right, so increasing systemic oxygenation]
- Treat pulmonary overcirculation with digoxin and diuretics as needed.
- Intubate and ventilate as needed.
- Inotropic support as needed.

- Infective endocarditis prophylaxis (lifelong).

Surgical Management

- ***Arterial switch operation (Jatene)***procedure of choice
  - Ideally performed during the first week of life.
  - The aorta and PA are switched back to their anatomically correct ventricle above the level of the valve.
  - Coronary arteries are transferred to the new aorta.
  - Associated lesions are also repaired at this time.
- ***Rastelli operation***is the operative choice in infants with TGA, VSD, and severe pulmonary stenosis.
  - Repaired during the first year of life.
  - VSD patch repaired to include LV to aortic outflow continuity with pulmonary blood flow provided via an RV to PA homograft.
- ***Atrial switch operation: Mustard or Senning procedure:***
  - Rerouting of atrial blood flow: RA→ mitral valve→LV→ PA and LA →tricuspid valve →RV →Aorta
  - Restores oxygenated blood into the systemic system and deoxygenated blood guided to the pulmonary system.
  - Disadvantages:
    - RV is left as the systemic ventricle -- will develop RV dysfunction.
    - Increased incidence of atrial dysrhythmias and baffle obstruction.

COMPLICATIONS

- Severe hypoxia.
- Aortic regurgitation
- Dysrhythmias
- RV dysfunction.
- Left ventricular failure
- Spasm or kinking of the reimplatented coronary arteries resulting in myocardial ischemia or death.

Prognosis

Operative mortality is less than 2%

4. Total Anomalous Pulmonary Venous Connection

DEFINITION

Total anomalous pulmonary venous connection (**TAPVC**), also known as **total anomalous pulmonary venous drainage** (**TAPVD**) and **total anomalous pulmonary venous return** (**TAPVR**), is a rare cyanotic congenital heart defect (**CHD**) in which there is ***no connection between the pulmonary veins and the left atrium***; the pulmonary veins instead connect directly into the right atrium or to a systemic vein (Innominate, superior vena cava (SVC), Azygus, inferior vena cava (IVC) , hepatic vein or portal vein) by an alternative pathway (vertical vein). Because all venous blood returns to the right atrium, the survival of baby depends upon the obligatory connection between the

left and right atria. A patent foramen ovale or an atrial septal defect *must* be present, or else the condition is fatal due to a lack of systemic blood flow.

Incidence

The incidence of total anomalous pulmonary venous drainage is 0.008% of live births, 2.2% in patients with congenital heart disease.

Classification

There are four types:

1. ***Supracardiac*** (50%): blood drains to one of the innominate veins (brachiocephalic veins) or the superior vena cava
2. ***Cardiac*** (20%): blood drains into coronary sinus or directly into right atrium, easily repaired.
3. ***Infradiaphragmatic*** (20%): blood drains into portal or hepatic veins. It is often associated with obstruction to the pulmonary venous drainage. It carries highest morbidity and mortality.
4. ***Mixed*** (10%)

Physiologically TAPVC can be divided into

a) Patients with pulmonary venous obstruction and
b) Patients without pulmonary venous obstruction.

PATHOPHYSIOLOGY

- The right atrium receives all the blood that normally would flow into the left atrium
- As a result the right side of the heart hypertrophies, whereas the left side, especially the left atrium, may remain small.
- The blood flow to the left atrium is through an associated patent foramen ovale or atrial septal defect allows systemic venous blood to shunt from the hight pressure right atrium to left atrium and into the left side of heart.
- As a result the oxygen saturation in the pulmonary artery is higher or identical to that in the aorta because of mixing of blood in the right atrium.
- If the pulmonary blood flow is large, pulmonary venous return is also large, and the amount of saturated blood is relatively high.
- If there is obstruction to pulmonary venous drainage, pulmonary venous return is impeded,pulmonary venous pressure rises and pulmonary interstitial edema develops and eventually contributes to CHF.

CLINICAL MANIFESTATION

- Cyanosis- most infants develop cyanosis in early life. The degree of cayanosis is inversely related to the amount of pulmonary blood flow- the more pulmonary blood, the less cyanosis.
- congestive heart failure
- the patients are irritable and have failure to thrive
- increased respiratory effort
- systolic ejection murmur at left upper sternal border
- cardiomegaly

- right axis deviation on ECG
- Snowman sign or `figure of 8 configuration` on chest radiograph
- right ventricular hypertrophy
- Without intervention, cardiac failure will progress to death

DIAGNOSIS

1. **Chest X-Ray :**

- The chest radiographs of patients who have unobstructed types of TAPVR typically exhibit right atrial and right ventricular hypertrophy with increased pulmonary blood flow.
- In patients whose return is to the left brachiocephalic vein, there may be a characteristic enlargement of the superior mediastinum, bilaterally in a *figure-8 or snowman shape*.

2. **ECG-** An electrocardiogram will usually show right-axis deviation with right atrial P-wave abnormality and right ventricular hypertrophy.

Obstructed TAPVC presents with relatively normal cardiac size and features of pulmonary venous hypertension, ground glass opacity.

3. **Echocardiography :** Two-dimensional echocardiography has been shown to establish the diagnosis accurately.
4. **CT Angio/ conventional catheterization :** may be necessary if echocardiography is inconclusive in determining the site(s) of the pulmonary venous connections.

SURGICAL MANAGEMENT

- Surgical correction of TAPVR is indicated during infancy.
- Before surgery, infants may be stabilized with prostaglandin E_1 to dilate the ductus venosus and the ductus arteriosus, although with significant obstruction this is usually not effective. If surgery cannot be performed urgently, extracorporeal membrane oxygenation (ECMO) may be required to maintain oxygenation.
- Precise description of the drainage sites is important in determining the suitable surgical approach
- In TAPVC without obstruction, surgical redirection can be performed within the first month of life. The operation is performed under general anesthesia. *The four pulmonary veins are reconnected to the left atrium, and any associated heart defects such as atrial septal defect, ventricular septal defect, patent foramen ovale, and/or patent ductus arteriosus are surgically closed. The anomalous pulmonary venous connection is ligated* With obstruction, surgery should be undertaken emergently.
- Early results are generally good, even for critically ill neonates.

POST OPERATIVE COMPLICATIONS

- Residual pulmonary vein obstruction
- Pulmonary artery hypertension
- Dysrhythmias
- Bleeding
- Heart block
- Persistent heart failure.

PROGNOSIS

Mortality for all types is less than 10% and is lowest for cardiac type; morbidity increases with the presence of pulmonary vein obstruction.

5. Truncus arteriosus

Failure of normal separation and division of the embryonic bulbar trunk into the pulmonary artery and the aorta, resulting in a single vessel that overrides both ventricles. Blood from both ventricles mixes in the common great artery, causing desaturation and hypoxaemia blood ejected from the heart flows prefentially to the lower pressure pulmonary arteries, causing increased pulmonary blood flow and reduced systemic blood flow.

INCIDENCE

It accounts for 1% to 4% of all CHD's

ETIOLOGY

A persistent truncus arteriosus results from failure of an aorticopulmonary septum to develop, which normally divides into the pulmonary artery and pulmonary valve and the aorta and aortic valve. This failure in division results in a single large vessel and single valve, which give rise to the pulmonary, systemic coronary circulation. the ventricular septum fails to develop at the same time, and therefore an associated VSD is present. the common truncal arteriosus vessel overrides the VSD and receives blood from both the right and left ventricles. The truncal valve is not a normal semilunar valve and can be stenotic or regurgitant.

Types

There are four classifications of truncus arteriosus related to the site of origin of the pulmonary artery from the common truncal vessel.

Type-1

A single pulmonary trunk arises near the base of the truncus and divides into the left and right pulmonary arteries.

Type -2

The left and right pulmonary arteries arise separately but in close proximity and at the same level the back of the truncus.

Type -3

The pulmonary arteries arise independently from the sides of the truncus.

PATHOPHYSIOLOGY/ ALTERED HEMODYNAMICS

Desaturated blood enters the right atrium and flows through the tricuspid valve into the right ventricle. Saturated blood from the left atrium flows through the mitral valve in the right ventricle.

⬇

The desaturated and saturated blood mixes in the ventricles at the level of the VSD and common ventricular outflow tract.

⬇

The common great vessel sends this blood to the systemic, pulmonary, and coronary circulations. The amount of pulmonary blood flow depends on the size of the pulmonary arteries and the pulmonary vascular resistance. Generally, resistance to pulmonary blood flow is less than systemic vascular resistance resulting in preferential blood flow to the lungs

⬇

Oxygen saturation depends on the volume of the pulmonary blood flow; the greater this flow the more symptoms of Pulmonary vascular disease, CHF, decreased cardiac output, disease and potential for coronary artery ischemia. The ventricles are under pressure and volume overload.

CLINICAL FEATURES

- Most infants are symptomatic with moderate to severe CHF and variable cyanosis.
- Poor growth
- Activity intolerance
- A harsh systolic murmur is herd and may be accompanied by a thrill.
- A diastolic murmur of truncal valve insufficiency may be heard.
- The infant may also have bounding pulse and a widened pulse pressure because of truncal valve insufficiency.
- The volume of pulmonary blood flow determines the severity of symptoms.
  - Unrestricted flow to the pulmonary artery results in pulmonary congestion and severe CHF
  - If pulmonary stenosis is present, pulmonary blood flow is limited and cyanosis increases.

DIAGNOSTIC EVALUATION

- History and physical examination
- ECG- right and left ventricular hypertrophy
- chest X-ray- increased pulmonary vascular markings
- Two dimensional echocardiogram.

MANAGEMENT

Medical

Medical management is aimed at reducing the effects of CHF and preventing polycythemia CHF is treated with digoxin and diuretics.

Surgical Treatment

1. open heart
2. sternototmy
3. Early repair in the first few months of life corrective repair involves
   - ***Closing the VSD***, so that the truncus arteriosus receives the outflow from the left ventricle to aorta.
   - Excising the pulmonary arteries from the aorta, and attaching them to the right ventricle by means of a homograft (segments of cadaver aorta and pulmonary artery that are treated with antibiotics and cryopreserved) are preferred over synthetic conduits to establish continuity between the right ventricle and pulmonary artery. Homografts are more flexible and easier to use during the procedure and appear less prone to obstruction.

Complications

- ✓ Persistant heart failure
- ✓ Bleeding
- ✓ Pulmonary artery hypertension
- ✓ Dysrhythmias
- ✓ Residual VSD

Because conduits are not living tissue, they will not grow along with the child and may also become narrowed with calcification. These children require additional procedures to replace the conduit as its size becomes inadequate in relation to the children's growth. A future truncal valve repair or replacement may be needed.

PROGNOSIS

Mortality is greater than 10%. The mortality risk is higher with truncal valve stenosis or insufficiency or other associated problems.

6. HYPOPLASTIC LEFT HEART SYNDROME

HLHS is a constellation of left heart abnormalities that include the following:

- marked hypoplasia of the left ventricle and ascending aorta
- the aortic and mitral valves are atretic, hypoplasric, or stenostic.
- a large patent ductus arteriosus supplies blood to the sysytemic circulation

Incidence

Hypoplastic left heart syndrome accounts for 1% of all CHDs. It is the most common cause of death from cardiac defects in the first month of life. it is more seen in males than in females.

Etiology

Inadequate development of the left side of the heart results in only one effective ventricle. this may be due to

- abnormal partitioning of the truncus arteriosus which result in a small aortic outflow tract, hypoplastic valve annulus, and hypoplastic aortic isthmus.
- Fetal aortic valve stenosis has been shown to reduce the growth rate of the left ventricle (10), and the concomitant outflow leads to abnormal development of this chamber

Pathophysiology/ altered hemodynamics

- Saturated pulmonary venous return is unable to flow from the left atrium through the rest of the left side of the heart.
- It is shunted left to right through a patent foramen ovale into the right atrium, where it mixes with the desaturated blood.
- Mixed saturated blood travels through the right ventricle to the main pulmonary artery.
- A portion of blood flows to the branch pulmonary arteries and to the lungs. a portion flows from the pulmonary artery through the PDA to the descending aorta.
- From the aorta this mixed saturated blood provides systemic and coronary blood supply. the coronary blood supply is from retrograde flow in the ascending aorta to the coronary arteries

Clinical Manifestations

- Although cyanosis may not always be obvious in the 1st 48 hr of life, a grayish-blue color of the skin is soon apparent and denotes a mix of cyanosis and poor perfusion. If the ductus arteriosus partially closes, signs of poor systemic perfusion and shock predominate.
- Signs of heart failure usually appear within the 1st few days or weeks of life and include dyspnea, hepatomegaly, and low cardiac output.
- All of the peripheral pulses may be weak or absent.
- Cardiac enlargement is usual, with a palpable right ventricular parasternal lift
- Extracardiac anomalies, particularly of the kidneys and central nervous system, may be present.
- Neonate may appear completely well initially, but becomes critically ill when the PDA closes.
- Once the PDA begins to close:
    - Tachypnea due to CHF.
    - Decreased urine output.
    - Poor feeding and feeding intolerance.
    - Lethargic; change in level of alertness.
    - Pallor; gray.
    - Weak peripheral pulses.
    - Cyanosis.

Diagnostic Evaluation

- Auscultation: single S_2; usually no heart murmur is present, but occasionally a soft systolic ejection murmur may be heard.
- Chest X-ray: cardiac silhouette varies (normal to increased size); increased pulmonary markings and pulmonary edema.
- ECG: RV hypertrophy; decrease electrical forces in V_5 and V_6.
- Two-dimensional echocardiogram with Doppler study and color flow mapping identifies the structural abnormalities and the altered blood flow patterns.
- A cardiac catheterization is usually not needed for initial diagnosis. It may be performed if a balloon atrial septostomy is needed to improve oxygenation.

Management

Nearly all neonates with hypoplastic left heart syndrome will die within the first month of life without surgical intervention. Medical management is done to stabilize the child before surgery.

Medical Management

- PGE_1 infusion is needed to maintain ductal patency and ensure adequate systemic blood flow.
- correction of fluid and electrolyte imbalances.
- Inotropic support as needed (dopamine, dobutamine).
- Intubate and ventilate as needed.
- Assess hepatic, renal, and neurologic function.
- Infective endocarditis prophylaxis (lifelong).
- Refer for surgical intervention.

Cardiac catheterization

- May need balloon atrial septostomy to allow unrestrictive LA to RA blood flow.

Surgical Management

two surgical courses are available

- Palliative, staged repair:
    - Stage I Norwood procedure (neonate):
        - *reconstruction of the hypoplastic aorta* using the PA and an aortic or pulmonary allograft to create a new aorta
        - atrial septectomy,
        - ligation of ductus arteriosus
        - placement of a BT shunt, to provide pulmonary blood flow.
    - Stage II*: bidirectional Glenn shunt* (ages 6 to 9 months): transect the SVC off the right atrium and directly suture end to side to right PA; ligate BT shunt.
    - Stage III: *Fontan procedure* (ages 18 months to 3 years): IVC to PA connection (extracardiac conduit or intracardiac baffle).

- ***Cardiac transplantation***, as a single definitive correction has been successful, with an 85% of operative survival rate and an 81% 5- year survival rate. the scarsity of the neonatal hearts however, greatly limits the number of infants who may receive transplants.

COMPLICATIONS

Norwood procedure

- Bleeding, low cardiac output syndrome, and arrhythmia
- Aortic arch obstruction at the site of surgical anastomosis
- Progressive cyanosis caused by limited blood flow through the shunt

Fontan procedure

- Transient superior vena cava syndrome
- Persistent pleural or pericardial effusion

Others

- Cyanosis.
- Metabolic acidosis.
- Thrombus formation in the systemic venous system.
- Infective endocarditis.
- Cardiovascular collapse.
- Multisystem failure.
- Death.

Prognosis of surgical treatment

- Success rate (survival to discharge):
  - Stage1:75%
  - Stage2:95%
  - Stage3:70%
- 5-year survival: 70%

Quality-of-life outcomes

- Behavioural abnormalities, learning disabilities, lower intelligence scores
- Multifactors:
  1. Possible associated abnormalities of CNS
  2. Hemodynamic instability in the pre-op period
  3. Intra-op perfusion --> neurologic injury

Risk factors with HLHS

- Prematurity/low birth weight
- Chromosomal and other extracardiac anomalies
- Additional intracardiac lesions/anatomic variants
- Obstructed pulmonary venous return

TYPICAL MURMUR ASSOCIATED WITH CONGENITAL HEART DEFECTS

SOUND HEARD	CHARACTERISTICS AND LOCATION	MAY INDICATE
Pansystolic murmur.	Maximum in the 4th, 5th and 6th intercostals spaces at left sterna border; high, harsh murmur heard throughout systole; may be associated with palpable thrill heard in the apical area.	VSD
Split $S_{2\ On}$ expiration systolic Ejection Murmur.	Split is heard in 2nd, 3rd, or 4th intercostals space; murmur maximum in 2nd and 3rd intercostals space.	ASD
Loud aortic closure sound systolic murmur.	Aortic sound heard at 2nd left intercostals space.	TPGA
Single S2 systolic murmur.		TOF
Continuous Murmur.	Murmur maximal in 2nd and 3rd intercostals spaces at lefet sterna border.	PDA
	Maximum in 2nd left intercostals space envelops S2 with late systolic accentuation; terminate in late or mid-diastole; radiate to 1st intercostals space.	

NURSING CARE OF THE CHILD WITH CONGENITAL HEART DISEASE

Nursing Assessment

1. Obtain a thorough nursing history.
2. Discuss the care plan with the health care team (cardiologist, cardiac surgeon, nursing case manager, social worker, nutritionist). Discuss the care plan with the patient, parents, and other caregivers.
3. Measure and record height and weight. Plot on a growth chart.
4. Record vital signs and oxygen saturations.
    a. Measure vital signs at a time when the infant/child is quiet.
    b. Choose appropriate-size blood pressure (BP) cuff.
    c. Check four extremity BP Ã— 1.
5. Assess and record:
    a. Skin color: pink, cyanotic, mottled.

b. Mucous membranes: moist, dry, cyanotic.
c. Extremities: check peripheral pulses for quality and symmetry; dependent edema; capillary refill; color and temperature.

6. Assess for clubbing (cyanotic heart disease).
7. Assess chest wall for deformities; prominent precordial activity.
8. Assess respiratory pattern.
    a. Before disturbing the child, stand back and count the respiratory rate.
    b. Loosen or remove clothing to directly observe chest movement.
    c. Assess for signs of respiratory distress: increased respiratory rate, grunting, retractions, nasal flaring.
    d. Auscultate for crackles, wheezing, congestion, stridor.
9. Assess heart sounds.
    a. Determine rate (bradycardia, tachycardia, or normal for age) and rhythm (regular or irregular).
    b. Identify murmur (type, location, and grade).
10. Assess fluid status.
    a. Daily weights.
    b. Strict intake and output (number of wet diapers; urine output).
11. Assess and record the child's level of activity.
    a. Observe the infant while feeding. Does the infant need frequent breaks or does he or she fall asleep during feeding? Assess for sweating, color change, or respiratory distress while feeding.
    b. Observe the child at play. Is play interrupted to rest? Ask the parent if the child keeps up with peers while at play.
    c. Assess and record findings relevant to the child's developmental level: age-appropriate behavior, cognitive skills, gross and fine motor skills.

NURSING DIAGNOSIS- PRE -OPERATIVE

1. Impaired Gas Exchange related to altered pulmonary blood flow or pulmonary congestion
2. Decreased Cardiac Output related to decreased myocardial function
3. Activity Intolerance related to hypoxia or decreased myocardial function
4. Imbalanced Nutrition: Less Than Body Requirements related to excessive energy demands required by increased cardiac workload
5. Risk for Infection related to chronic illness
6. Fear and Anxiety related to life-threatening illness

Nursing Interventions

Relieving Respiratory Distress

1. Position the child in a reclining, semi-upright position.
2. Suction oral and nasal secretions as needed.
3. Identify target oxygen saturations and administer oxygen as prescribed.
4. Administer prescribed medications and document response to medications (improved, no change, or worsening respiratory status).

- Diuretics.
- Bronchodilators.

5. May need to change oral feedings to nasogastric feedings because of increased risk of aspiration with respiratory distress.

Improving Cardiac Output

1. Organize nursing care and medication schedule to provide periods of uninterrupted rest.
2. Provide play or educational activities that can be done in bed with minimal exertion.
3. Maintain normothermia.
4. Administer medications as prescribed.
    a. Diuretics (furosemide, spironolactone):
        i. Give the medication at the same time each day. For older children, do not give a dose right before bedtime.
        ii. Monitor the effectiveness of the dose: measure and record urine output.
    b. Digoxin:
        i. Check heart rate for 1 minute. Withhold the dose and notify the physician for bradycardia (heart rate less than 90 beats/minute [bpm]).
        ii. Lead II rhythm strip may be ordered for PR interval monitoring. Prolonged PR interval indicates first-degree heart block (dose of digoxin may be withheld).
        iii. Give medication at the same time each day. For infants and children, digoxin is usually divided and given twice per day.
        iv. Monitor serum electrolytes. Increased incidence of digoxin toxicity associated with hypokalemia.
    c. Afterload-reducing medications (captopril, enalapril):
        i. When initiating medication for the first time: check BP immediately before and 1 hour after dose.
        ii. Monitor for signs of hypotension: syncope, light-headedness, faint pulses.
        iii. Withhold medication and notify the physician according to ordered parameters

Improving Oxygenation and Activity Tolerance

1. Place pulse oximeter probe (continuous monitoring or measure with vital signs) on finger, earlobe, or toe.
2. Administer oxygen as needed.
3. Titrate amount of oxygen to reach target oxygen saturations.
4. Assess response to oxygen therapy: increase in baseline oxygen saturations, improved work of breathing, and change in patient comfort.
5. Explain to the child how oxygen will help. If possible, give the child the choice for face mask oxygen or nasal cannula oxygen.

Providing Adequate Nutrition

1. For the infant:
    a) Small, frequent feedings.
    b) Fortified formula or breast milk (up to 30 cal/oz).
    c) Limit oral feeding time to 15 to 20 minutes.
    d) Supplement oral feeds with nasogastric feedings as needed to provide weight gain (ie, continuous nasogastric feedings at night with ad-lib by-mouth feeds during the day).
2. For the child:
    a) Small, frequent meals.
    b) High-calorie, nutritional supplements.
    c) Determine child's likes and dislikes and plan meals accordingly.
    d) Allow the parents to bring the child's favorite foods to the hospital.
3. Report feeding intolerance: nausea, vomiting, diarrhea.
4. Document daily weight (same time of day, same scale, same clothing).
5. Record accurate inputs and outputs; assess for fluid retention.
6. Fluid restriction not usually needed for children; manage excess fluid with diuretics.

Preventing Infection

1. Maintain routine childhood immunization schedule. With the exception of RSV (Synagis) and influenza, immunizations should not be given for 6 weeks after cardiovascular surgery.
2. Administer yearly influenza vaccine.
3. Administer RSV immunization for children younger than age 2 with complex CHD and those at risk for CHF or pulmonary hypertension.
4. Prevent exposure to communicable diseases.
5. Good hand washing.
6. Report fevers.
7. Report signs of URI: runny nose, cough, increase in nasal secretions.
8. Report signs of GI illness: diarrhoea, abdominal pain, irritability.

Reducing Fear and Anxiety

1. Educate the patient and family.
2. Provide the family with contact phone numbers: how to schedule a follow-up visit; how to reach a cardiologist during the work week, evenings, weekends, and holidays.

Family Education and Health Maintenance

1. Instruct the family in necessary measures to maintain the child's health:
    a) Complete immunization.
    b) Adequate diet and rest.
    c) Prevention and control of infections.
    d) Regular medical and dental checkups. The child should be protected against infective endocarditis when undergoing certain dental procedures.
    e) Regular cardiac checkups.

2. Teach the family about the defect and its treatment.
    a) Provide patients and families with written and verbal information regarding the CHD. Offer appropriate Internet resources for information about CHD and medical and surgical treatment options.
    b) Signs and symptoms of CHF .
    c) Signs of hypercyanotic spells associated with cyanotic defects and need to place child in knee-chest position.
    d) Need to prevent dehydration, which increases risk of thrombotic complications.
    e) Emergency precautions related to hypercyanotic spells, pulmonary edema, cardiac arrest (if appropriate).
    f) Special home care equipment, monitors, oxygen.
3. Encourage the parents and other people (teachers, peers) to treat the child in as normal a manner as possible.
    a) Avoid overprotection and overindulgence.
    b) Avoid rejection.
    c) Promote growth and development with modifications. Facilitate performance of the usual developmental tasks within the limits of the child's physiologic state.
    d) Prevent adults from projecting their fears and anxieties onto the child.
    e) Help family deal with its anger, guilt, and concerns related to the disabled child.
4. Initiate a community health nursing referral if indicated.
5. Stress the need for follow-up care.
6. Encourage attendance in support groups for patients and families.

Evaluation: Expected Outcomes

1. Improved oxygenation evidenced by easy, comfortable respirations
2. Improved cardiac output demonstrated by stable vital signs, adequate peripheral perfusion, and adequate urine output
3. Increased activity level
4. Maximal nutritional status demonstrated by weight gain and increase in growth curve percentile
5. No signs or symptoms of infection
6. Parents discuss diagnosis and treatment together and with child

PREOPERATIVE PREPERATION

Pre-operative preparation includes:

- ❖ Psychological preparation
- ❖ Pre-operative assessment
- ❖ Legal assessment
- ❖ Physical preparation

Psychological preparation

1. Discuss surgical procedures with parents and child and evaluate their understanding.

2. Secure a favourite blanket, toy or other object to the bed or stretcher when the child is taken to the operative room.
3. Encourage the parents to visit the child prior to surgery no matter how early in the day it is scheduled. Have parents accompany the child as far as possible.
4. Assure child that parents will be nearby, waiting for him or her after the operation is over, if this is indeed the plan. Tell parents where to wait during surgery.
5. The parents and child should tour the intensive care unit and other units in which the child will be during the hospitalization. this allows them to be familiar with the environment.
6. The sequence of events surrounding the day of surgery when and where to arrive and where to wait during the procedure should be reviewed.
7. Parents should be assured that they will receive updates abouts their condition throughout the procedure and will be permitted to visit soon after the surgery is completed
8.

Pre-operative assessment

It is performed to determine baseline data and identify conditions that may interfere with the administration of anaesthesia or produce problems post operatively.

Infants should be positioned on the examination table for the entire examination. toddlers and small children may lie on parents lap for the initial part of the examination, and they can be moved to the examination table for abdominal, inguinal, genital and rectal examinations when required. having the parent by examination table reduces the fear and anxiety and should be encouraged.

Ethical considerations

- Written informed consent of the parent or legal guardian is required.
- adults who have reached the age of consent(18 yrs).

Physical preparation

- Monitor temperature, pulse, respirations and blood pressure. any abnormal vital signs are reported to the surgeon.
- pre-operative fasting

  Fasting guide lines before surgery
  - 2 hours clear non-particulate and uncarbonated fluids.
  - 4 hours breast milk.
  - 6 hours solid food, infant formula or other milk.
- Make certain that all other prescribed pre-operative procedures have been completed(enema, insertion of nasogastric tube)
- Give a through bath prior to the surgery (using chlorhexine scrub or any other antiseptic solution).
- Dress child in clean hospital gown or other attire according to institutional procedure.

- Observe for loosened teeth, dental appliances such as partial plates or braces and report if present. Dental appliances are removed if possible and given to the parents. Presence of loose teeth is reported to the anaesthetist.
- Remove makeup and nail polish pre operatively.

Premedication

- Premedication is given 30 minutes before surgery.
- All preoperative preparation is completed before the premedication is given administration of these medications is recorded in the patients charts before the child is transferred to the operating room.
- The child should remain in bed following administration of the medication to promote maximum effect and to prevent falls from dizziness.
- The side rails are raised and the child is instructed not to get out of bed.

Preoperative Procedures

- Childs medical and family history
- Childs normal routine
- Observations
  - Temperature, pulse, BP, SaO_2
  - Urinalysis
  - Height
  - Weight
  - Date of last menstrual period (adolescent girls)
- Chest X-ray
- 12 lead electrocardiogram(ECG)
- Echocardiography (ECHO)
- Blood samples
  - Full blood count(FBC)
  - Urea and electrolytes
  - Glucose
  - Creatinine
  - Calcium
  - Magnesium
  - Clotting studies
  - Albumin
  - C- reactive protein (CRP)
  - Liver function tests (LFTs)
- Cardiac catheterization

IMMEDIATE POST-OPERATIVE CARE

1. **Airway and Breathing**

1. The child will be intubated and ventilated in the immediate postoperative period.
2. The child will be extubated when
   -cardiovascularly stabe
   - making good respiratory effort with good gas exchange on minimal ventilator support.
3. Continuous monitoring and hourly recordings of the following observations are essential in the immediate post operative period.
   -Respiratory rate
   -chest expansion
   -chest auscultation
   -appropriate ventilator observations
4. The nurse should also regularly assess
   -chest expansion
   -chest auscultation
   -respiratory effort

POSTOPERATIVE NURSING DIAGNOSIS

1. **Nursing diagnosis**

Decreased cardiac output related to effects of decreased preload/hypovolemia, postoperative bleeding, increased after load, altered myocardial contractility, arrhythmias, or cardiac defects

Outcome evaluation

child will demonstrate a cardiac output adequate to maintain perfusion of body, as evidenced by stable vital signs, level of consciousness within normal limits, hemodynamic stability and adequate tissue perfusion.

Intervention

- obtain baseline and frequent assessment of vital signs(BP.HR,RR)
- Assess for sighs and symptoms of decreased cardiac output such as hypotension, edema, cool extrimities, delayed capillary refill, weak peripheral pulseslow urine putput and mental status changes.
- monitor intake and output,hemodynamic parameters, and laboratory values.
- administer oxygen as ordered. monitor oxygen saturation and arterial blood gas values.

2. **Nursing diagnosis**

Impaired gas exchange related to unexpanded lung space and collection of lung excretions.

Outcome Evaluation

Childs respiratory rate remains within age-appropriate parameters; absence of rales or other adventitious breath sounds; chest tubes function normally.

Interventions

- suction as necessary while child is receiving ventilator assistance to prevent pooling of secretions in the respiratory tract.
- as soon as the ET tube and ventilator are removed , encourage the child to cough and deep breath or use an incentive spirometer at hourly intervals to help mobilize secretrions.
- chest physiotherapy with percussion and vibration mat be prescribed to keep lung secritions mobile.
- blowing cotton balls and blowing up a balloon are exercises to help achieve lung expansion**.**

3. **Nursing diagnosis**

   Risk for infection related to surgical incision and tube sites

Outcome evaluation

Childs temperature remains at or below 100.4 0Faxillary. Incision site is clean, dry and without evidence of erythemea or foul drainage.

Intervention

- frequently monitor temperature post operatively to assess for infection.
- use strict aseptic technique when changing when changing the incisional dressing to avoid introducing pathogens
- frequently assess the dressing over the surgical incision and the points of insertion of the thoracotomy tubes for drainage and erythema
- as ordered by physician begin a prophylactic course of a broad spectrum antibiotic

4. **Nursing diagnosis**

   Hypothermia related to cooling during surgery

Outcome evaluation

Childs temperature is above 96.8^{o} F axillary. Capillary refill is less than 5seconds.

Interventions

- monitor the temperature of the child
- cover the child with a hyperthermia blanket, warm blankets, or radiant heat may be necessary to elevate the temperature to normal

5. **Nursing diagnosis**

   Risk for excess or deficit fluid volume related to fluid shifts accompanying cardiac surgery.

Outcome evaluation

Child maintains weight, skin turgor is good and CVP or pulmonary artery pressure is within established parameters.

[Child tend to develop hypervolemia after cardiac surgery because of increase production of aldosterone by the adrenal glands and an increase in antidiuretic hormone secretion by the pituitary gland in response to stress. Also if cardiopulmonary bypass was used, some fluid may have been shifted from the intravascular system to the interstitial spaces during surgery. after surgery this fluid returns by osmosis to the vessels causiong hypervolemia. on the other hand an individual child may have experienced excessive bleeding because of the heparin used during surgery and may subsequently develop hypovolemia.]

Interventions

- monitor CVP or PA pressure to evaluate a childs hemodynamic status.
- monitor IV fluid administration carefully to prevent fluid overload.
- oral fluid intke is withheld for atleast the first 24 hours after surgery.
- once bowel sounds have returned, oral fluids can be introduced gradually.

6. Nursing diagnosis

Parental anxiety related to lack of knowledge of post operative routine and exercises

Outcome evaluation:

Family members accurately state plans for child's post operative recovery, relate less anxiety after teaching and support

Interventions

- encourage parents to do whatever they want ever they want to do for their childs care during this period.
- offering the child sips of water or helping the child with bathing helps parents see that their child is returning to usual activities and doing well.
- urge parents to read to children or play music to them as a way to provide quite rest periods
- show them how to lift an infant by placing their hands under the shoulders and buttocks.
- at hospital discharge, parents need clear explanation of the activities in which the child will and will not be able to participate.

POTENTIAL COMPLICATIONS AND ITS MANAGEMENT

These may be detected by:

- unequal chest expansion
- unequal air entry
- a decrease in child's saturations
- increase respiratory efforts in the self ventilating child

Potential complications are as follows:

1. Pneumothorax

2. Haemothorax
3. Chylothorax
4. Pleural Effusion
5. Consolidation

2. Circulation

- Continuous monitoring and hourly recording of the following observations are essential in the immediate post operative period.
  - Heart rate and rhythm
  - Blood pressure
  - Central venous pressure
  - Core and peripheral temperature
  - Drainage loss, volume and quality
  - Urine output
- In addition children who are prone to pulmonary hypertension will also have continuous observation and hourly recording of
  - Left atrial pressure
  - Pulmonary artery pressure

Potential complications

1. Dysrhythmias

These may be caused by:

- Inflammation and edema caused around atrial or ventricular incisions or around the sinoatrial or atrioventricular nodes
- Surgical damage to the SA node, AV node or the conduction pathways
- Hypoxia of the SA node, AV node or the conduction pathways
- Acidosis
- Electrolyte disturbances

These may be treated by:

- Correcting any hypoxia
- Correcting any acidosis
- Correcting any electrolyte disturbances
- Use of antidysrhythmics eg adenosine, amiodarone, flecanide
- Cardioversion
- pacing

2. Bleeding

These may be caused by:

- Failure to fully reverse the anticoagulation given whilst on cardiopulmonary bypass
- An open vessel

Nursing observations will reflect:

- Tachycardia
- Hypotension
- Low central venous pressure
- Excess, bloody drainage
- Poor peripheral perfusion

Treatment involves the following:

- ✓ Check clotting profile, if clotting is extended administer the appropriate clotting factor.
- ✓ If clotting is normal, call the surgeon and the theatre team, re-open the chest and seal the bleeding point.
- ✓ Volume, eg. Blood may need to be give to support the child's circulation until the above measures have been instigated.

3. Cardiac tamponade

4. There is **collection of excess fluid in the pericardium**, this will 'compress' the heart, restricting its function, and lead directly to cardiac arrest. In the postoperative cardiac patient, Cardiac tamponade is usually caused by bleeding into the pericardium.

Treatment is as follows:

- Children who are tamponading will usually develop dysrhythmias and lose cardiac output very quickly. Full resuscitation may be intigated. The surgeon and the theatre team must be called immediately to reopen the childs chest and evacuate the excess blood from the pericardium.

5. Reduced cardiac output/ hypotension

This is caused either by hypovolaemia or decreased cardiac function.

Hypovolaemia nursing observations will reflect:

- ➢ Tachycardia
- ➢ Hypotension
- ➢ Low central venous pressure
- ➢ Poor peripheral perfusion
- ➢ Reduced urine output

Treatment is:

- ➢ 10 ml/kg colloid, review effect and repeat bolus if necessary.
- ➢ Human albumin solution (4.5%) is readily available and is the colloid of choice for acute incidents of hypovolaemia.
- ➢ Blood or fresh frozen plasma should be considered if the child has a low haemoglobin or deranged clotting.

Decreased cardiac function Nursing observations will reflect:

- ✓ Tachycardia
- ✓ Hypotension
- ✓ Normal, high or low central venous pressure
- ✓ Poor peripheral perfusion
- ✓ Reduced urine output

Treatment is:

- ✓ Continous inotrop infusion to improve contractility eg. Dobutamine, dopamine, adrenaline , noradrenaline.
- ✓ Afterload reduction eg. milrinone
- ✓ Optimised preload.eg. noradrenaline

6. Pulmonary hypertensive crisis

NEUROLOGICAL

Cerebral insult may occur due to:

Hypoxia, hypotension, cerebral bleed (related to anticoagulation for bypass), air embolism and thrombus (r/t bypass procedure)

It is therefore important ot perform an early neurological assessment on children folloeing cardiac surgery.

Nursing observations will asses:

- Puipil reaction
- Posture and muscle tone
- Seizure activity
- Consciousness level

Any abnormality or change in the child's neurological status must be referred to the medical staff for further investigation and management.

RENAL

Renal insult may occur due to

Hypoxia, hypotension

Nursing observations will assess:

- Hourly urine output
- Hourly fluid balance
- Serum creatinine, urea and electrolytes.

Regular frusemide may be given to maintain a good urine output (> 1ml/kg/h) and to optimise cardiac function.

FLUIDS AND NUTRITION

- ➢ Fluids are strictly restricted to 50% of normal maintenance requirements following open heart surgery.
- ➢ The childs fluid balance should be calculated hourly and the electrolytes and blood sugar level should be checked regularly.
- ➢ A nasogatric tube should be passed and enteral nutrition should be commenced, ideally within 3-4 hours of the child returning from the theatre.