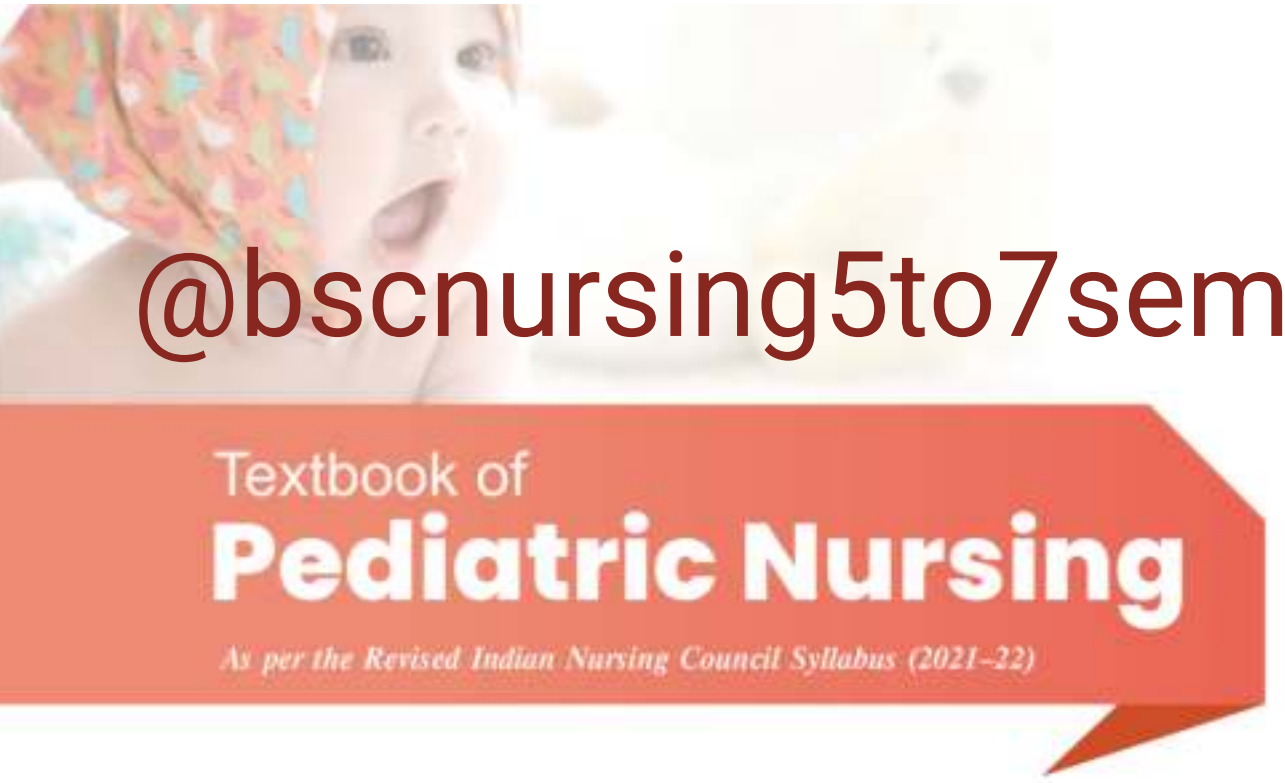


Textbook of Pediatric Nursing

As per the Revised Indian Nursing Council Syllabus (2021–22)

Unit VII

Disease Conditions–I

Learning Objectives

At the end of this unit, the students will be able to:

Describe the etiology, pathophysiology, clinical manifestations and nursing management of children with disorders of following systems:

- Cardiovascular system.
- Gastrointestinal system.
- Genitourinary system.
- Nervous system.

Unit Outline

Chapter 15

Cardiovascular Disorders

Chapter Outline

- Introduction
- Related Anatomy of Heart
- Congenital Heart Disease
- Nursing Care of a Child with Cardiac Surgery
- Congestive Heart Failure (CHF)
- Rheumatic Fever
- Infective Endocarditis
- Acquired Heart Disease

INTRODUCTION

Cardiovascular issues in children are complex, ranging from congenital defects that may present at birth to acquired heart disease. The presentation vary depending on the disorder and the child's age. The disorders may require episodic care or life-long medical management by a specialist. Managing a child with a cardiovascular disorder is very challenging and requires the nurse to have an in-depth understanding of disease processes, congenital heart defects, and treatment regimens as well as the ability to work with children of different age groups and families in varying states of emotional health.

RELATED ANATOMY OF HEART

Heart has four chambers, left separated from right. It has two types of valves, semilunar and atrioventricular valves. There are two outflow tracts for blood, i.e., through aorta and pulmonary artery. There are two circulation in series, systemic and pulmonary.

Left heart has high pressure, thick chamber, pumps blood to systemic circulation which has relatively high resistance and pressure. Right heart has low pressure, thin chamber, pumps blood to pulmonary circulation which has relatively low resistance and pressure. A chamber that has to pump against high pressure can fail with time. If a chamber that does not pump efficiently, can cause congestion of blood in the circulation that feeds it. This also causes strain on the chamber that is pumping against the congestion.

Deoxygenated blood returning from the upper body travels through the superior vena cava (SVC); and that returning from the lower body travels through the inferior vena cava (IVC). This blood mixes in the right atrium and then goes through the tricuspid valve into the right ventricle. It then goes through the pulmonic valve to the main pulmonary artery, continuing out to the right- and left-branch pulmonary arteries to the lungs. Once the blood is oxygenated, it returns to the left atrium by way of four pulmonary veins (two right and two left). It then travels through the mitral valve to the left ventricle, through the aortic valve, and out the ascending aorta (Fig. 15.1).

Fetal Circulation

Understanding fetal circulation (Fig. 15.2) is important to understand congenital heart defects. While in utero, the lungs exhibit high resistance to blood flow due to their fluid-filled status and constricted pulmonary arterioles. The lungs do not perform normal gas exchange functions; rather, the placenta performs the actions of oxygenation and carbon dioxide removal for the fetus. Therefore, the fetus requires blood to

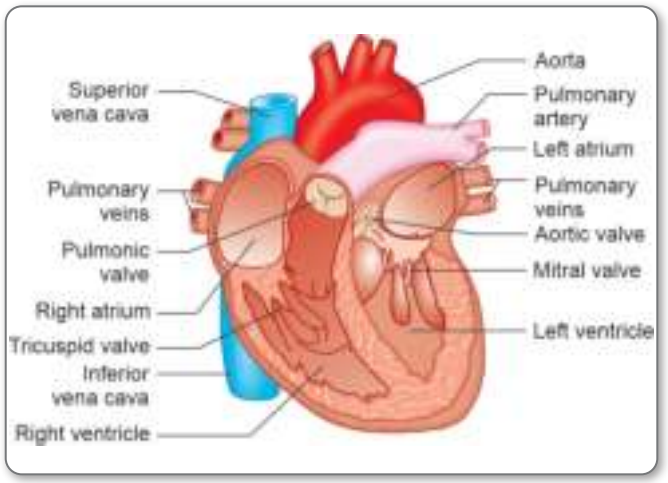


Figure 15.1: Anatomy of heart

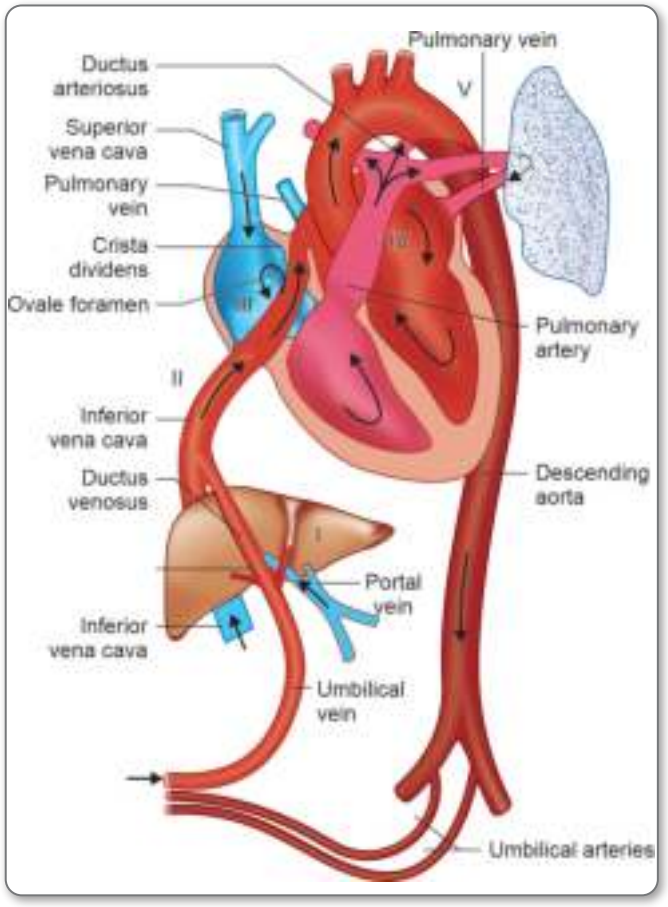


Figure 15.2: Fetal circulation

be mixed in the heart in order to provide oxygenated blood to the body. Three fetal shunts exist to facilitate this circulation:

1. The foramen ovale (opening between right and left atria)
2. The ductus arteriosus (between the pulmonary artery and aorta)
3. The ductus venosus (umbilical vein to inferior vena cava)

As the highly oxygenated blood returns from the placenta through the IVC, 40% of the flow is directed across the foramen ovale into the left atria. The foramen ovale is a flapped opening between the atria that allows for this oxygenated blood to bypass the lungs and move from the left atrium to the left ventricle and then to ascending aorta to the coronaries and upper body, which includes the brain.

The remaining IVC blood mixes in the right atrium with the deoxygenated blood that has returned from the head and upper extremities via the SVC. This blood then passes through the tricuspid valve, to the right ventricle, and then to pulmonary artery. But because of the higher pulmonary pressures in comparison with the lower systemic pressures, almost 90% of blood flowing from the right ventricle bypasses the lungs via the ductus arteriosus. **Ductus arteriosus** is a connection between the pulmonary artery and aorta. It allows blood to bypass the high-pressure pulmonary system and travel directly out from the aorta to the body. Only 10% or less of blood ejected from the right ventricle actually passes to the lungs for growth and development.

These two shunts, the foramen ovale and ductus arteriosus, allow most of the poorly saturated blood from the upper and lower body to reach the right ventricle and the more highly saturated umbilical venous return to reach the left ventricle.

The third fetal shunt is the **ductus venosus**. As blood returns from the placenta through the umbilical vein, some goes into the hepatic veins to the liver and slightly more than half passes though the ductus venosus into the IVC. The ductus venosus is a shunt that allows oxygenated blood in the umbilical vein to bypass the liver. This highly oxygenated blood that is directed to the ductus venosus is then guided to the foramen ovale, to go to the left side of the heart and out to the body.

The changes that occur at birth, including hormonal levels, oxygen tension changes, pressure changes, and blood flow, lead to closure of these three shunts.

As amniotic fluid is absorbed in the lungs and they become a gas-filled system, pressures on the left side of the heart are now higher than on the right. As well, the volume of blood returning from the pulmonary veins directly to the left atrium significantly increases. These changes in volume and pressure allow for the flap of the foramen ovale to be pressed closed against the atrial septum immediately; and eventually over the subsequent weeks, the flap will become permanently closed.

The ductus arteriosus begins to constrict immediately at birth due to changing oxygen tension levels, decreasing prostaglandin levels, and a drop in pressure within the lumen of the ductus caused by changes in pulmonary vascular resistance, ultimately closing completely within a few days at the latest. Factors such as prematurity, congenital heart defects, or persistent pulmonary hypertension can change the expected closure times for these two shunts.

Permanent structural closure of ductus venosus is thought to occur within 2–3 weeks after birth.

Within a few days after birth and with no confounding factors, circulation will follow the same pattern as in an adult. It is important to note though that the pulmonary pressures in a newborn, although lower than systemic pressures immediately after birth, will continue to drop to their lowest level by approximately 4–6 weeks of age.

The common cardiac conditions seen in children are congenital heart diseases and acquired heart diseases like rheumatic fever, hypertension, and hyperlipidemia.

The child commonly presents with dyspnea, brow sweating, tachycardia, murmurs, cyanosis, and poor weight gain.

The common tests done to diagnose cardiac disorders are electrocardiogram (ECG), echocardiogram (ECHO), Chest X-ray, etc. Lab investigations include complete blood count (CBC) including serum calcium, sodium, and potassium. Sodium is an indicator of fluid status; hemoglobin and hematocrit determine the need for blood transfusions; and coagulation studies (prothrombin time [PT], partial thromboplastin time [PTT], international normalized ratio [INR]) are important before and after cardiac surgery to prevent unnecessary bleeding. Cyanotic children may develop a degree of polycythemia over time as the body develops a greater number of red blood cells in an effort to provide more oxygen carrying capacity to the body.

C-reactive protein (CRP) is an indicator of an active infectious process, and erythrocyte sedimentation rate (ESR) is an indicator of inflammation which occurs with rheumatic fever. B-Type natriuretic peptide (BNP) is a substance secreted from the ventricles in response to changes in pressure that occur when heart failure develops and worsens.

Let us discuss about some common cardiac disorders in children.

CONGENITAL HEART DISEASE

When caring for a child with a congenital heart defect, it is important to remember normal physiology, as this influences blood flow. The right side of the heart is a lower pressure system than the left side of the heart. This pressure difference allows blood to flow from one chamber to another, specifically from an area of high pressure to an area of low pressure; the blood will always follow the "path of least resistance." When venous blood from the right side of the heart mixes with blood on the left side, this is a "right-to-left" shunt that delivers deoxygenated blood to the body. Heart defects with this type of blood flow are termed **cyanotic heart disease**.

If the blood shunts left to right, then oxygenated blood from the left side mixes with blood in the right side of the heart and goes back to the lungs again. This is called **acyanotic heart disease.** The classification of congenital heart diseases is shown in Table 15.1.

Table 15.1: Classification of congenital heart diseases

Acyanotic (Left to right shunt)	Cyanotic (Right to left shunt)	Obstructive lesions
• There is increased pulmonary blood flow. • They do not result in cyanosis. • Prognosis is good • Surgery is done in one stage	• There is decreased pulmonary blood flow. They result in cyanosis. • Prognosis is guarded • Surgery is done in several stages	• Obstruction defects occur when heart valves, arteries, or veins are abnormally narrow or blocked.
Examples: • ASD: Atrial septal defect • VSD: Ventricle septal defect • PDA: Patent ductus arteriosus	**Examples:** • TOF: Tetralogy of fallot • TGA: Transposition of great arteries • Truncus: Arteriosus • TAPVC: total anomalous pulmonic venous connection • Tricuspid: Atresia	**Examples:** • Aortic stenosis • Pulmonic stenosis • COA: Coarctation of aorta

Etiology

Exact cause is unknown. It results from abnormal embryonic development or the persistence of fetal structure beyond the time of normal involution. Possible causes are:

- Fetal or maternal infection during first trimester (rubella)
- Chromosomal abnormalities (trisomy 21, 18, 13)
- Maternal insulin dependent diabetes
- Teratogenic effects of drugs (thalidomide) and alcohol
- Maternal age >40 at the time of first pregnancy
- Siblings with heart disease
- Parents with congential heart disease (CHD)

Congenital heart defects are associated with an increased incidence of some other symptoms, together being called the VACTERL association:

V – Vertebral anomalies
A – Anal atresia
C – Cardiovascular anomalies
T – Tracheoesophageal fistula
E – Esophageal atresia
R – Renal (Kidney) and/or radial anomalies
L – Limb defects

Assessment for Heart Disease

The assessment of a child for the presence or absence of heart disease can be done using "Nadas criteria" (Table 15.2). The criteria are divided into major and minor criteria. Presence of one major and two minor criteria are essential for indicating the presence of heart disease.

Table 15.2: Nadas criteria for assessment of heart disease

Major criteria	Minor criteria
• Systolic murmur grade 3 or more especially when associated with a thrill • Diastolic murmur • Cyanosis • Congestive cardiac failure	• Systolic murmur less than grade 3 • Abnormal S2 • Abnormal ECG • Abnormal chest X-ray • Abnormal BP

Acyanotic Defects (Left to Right Shunt)

This includes ASD, VSD and PDA.

Atrial Septal Defect (ASD)

ASD is an abnormal opening in the septum between the left atrium and the right atrium (Fig. 15.3).

Types

- Ostium Secundum type is located in the center of the atrial septum (most common).
- Ostium Primum type is a large gap at the base of the septum frequently associated with deformities of the mitral and tricuspid valves and/or a small, high ventricular septal defect (endocardial cushion defects).

Pathophysiology

The pressure in the left atrium (LA) is greater than the pressure in the right atrium (RA) and promotes the flow of oxygenated blood from the LA to the RA.

The oxygenated blood that flows through the defect enters the RA and mixes with the systemic venous blood returning to the lung.

The major hemodynamic abnormality is volume overload of the RV.

If the pulmonary resistance is great, this may increase right atrial pressure thus causing a reversal of the shunt, with deoxygenated blood flowing from the RA to the LA. This situation will produce cyanosis.

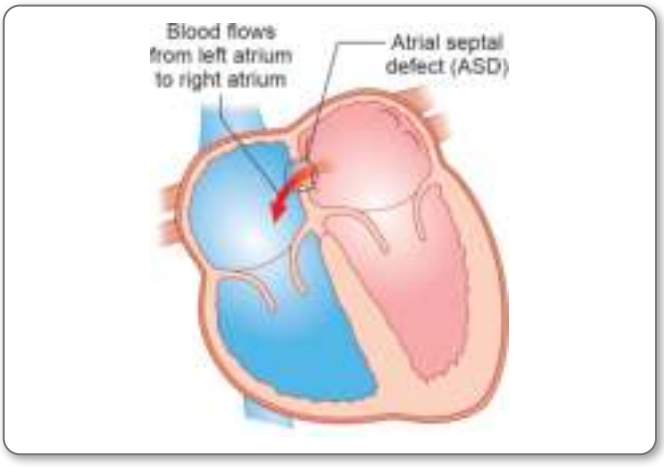


Figure 15.3: Atrial septal defect (ASD)

Clinical Manifestations

Ostium secundum type is generally asymptomatic even when the defect is large.

Ostium primum type is generally asymptomatic, although the symptoms like slow weight gain, tiredness, dyspnea with exertion, frequent respiratory infections and CHF may occur.

Diagnosis

- **Auscultation:** Systolic, medium pitched ejection murmur heard best at the 2nd left interspace.
- Fixed widely spaced S2
- **Chest X-ray:** Prominent main pulmonary artery, right atrial and right ventricular enlargement, increase in vascular markings of the lungs
- **ECG:** Right ventricular hypertrophy and right axis deviation (ostium secundum defect); left axis deviation, P wave changes indicating atrial enlargement and prolonged PR interval are common in ostium primum defects.
- Echo, Cardiac catheterization, Angiocardiography

Complications

- Infective endocarditis
- Cardiac failure
- Pulmonary hypertension
- Coronary artery disease
- Atrial fibrillation

Treatment

Defect size of 0–8 mm: Spontaneous closure by 18 months

If the defect is 8 mm or larger with evidence of increased pulmonary blood flow, the child is referred to for closure immediately. Surgical closure involves a median sternotomy incision and cardiopulmonary bypass by suture or patch. These children are typically home within 2–3 days of cardiac surgery.

Ventricular Septal Defect (VSD)

VSD is an abnormal opening in the septum between the RV and LV (Fig. 15.4). It may vary in size from small defects to very large defects and may occur in either the membranous or muscular portion of ventricular septum.

Pathophysiology

The pressure in the LV is greater than the pressure in the RV and promotes the flow of oxygenated blood from the LV to the RV.

The oxygenated blood that flows through the defect mixes with the blood returning from the RA. The blood flow through the shunt recirculates through the lung, thus increasing the total blood flow through the lung.

The major hemodynamic abnormalities are as follows:

- Increased right ventricular and pulmonary arterial pressure.

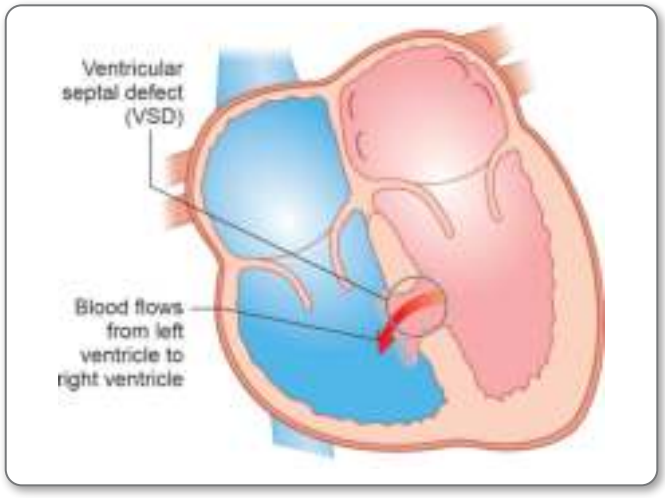


Figure 15.4: Ventricular septal defect (VSD)

- Increased blood flow to the RV, pulmonary arteries, LA and LV.

If pulmonary resistance is great, this may increase right ventricular pressure, thus causing a reversal of the shunt with deoxygenated blood flowing from the RV to the LV (this situation, termed Eisenmenger's complex will produce cyanosis).

Clinical Manifestations

Small VSDs: Are usually asymptomatic, may close spontaneously

Large VSDs: May develop symptoms as early as 1–2 months of age like slow weight gain, feeding difficulties, pale, frequent respiratory infections, tachypnea, excessive sweating, congestive heart failure.

Diagnosis

- Chest X-ray, to see if there is a large heart with fluid in the lungs.
- ECG shows signs of an enlarged left ventricle.
- Echocardiogram is done for definitive diagnosis.

Complications

- Infective endocarditis
- Congestive heart failure
- Aortic insufficiency
- Arrhythmias
- Failure to thrive in infancy

Treatment

- Digitalis (digoxin) and diuretics
- Prophylactic antibiotics
- Medical management of congestive heart failure if it occurs in infancy
- Spontaneous closure of defect occurs by 6 months to 2 years
- **Surgical closure of VSD:** VSDs are typically closed using the heart lung machine and by placing a patch over the defect. The patch may be a piece of fabric (Dacron) or the patient's own tissue (pericardium) and it is secured with fine sutures. Purse string suture for smaller defect. Patients with pulmonary arterial hypertension require early surgery (before 2 years of age) to avoid irreversible pulmonary bed changes.

Patent Ductus Arteriosus (PDA)

PDA is the persistence of a fetal connection between the pulmonary artery and aorta. The ductus arteriosus, a blood vessel between pulmonary artery and aorta, fails to close after birth (Fig. 15.5).

Normally functional closure occurs soon after birth, i.e., 96 hours and anatomical closure occurs between 1 and 3 months.

Causes

- Maternal exposure to rubella during the first trimester
- Living at high altitude, long term exposure to low blood O_2 tension
- Increased sensitivity of DA to prostaglandins
- Hypoxia, acidosis
- Defective smooth muscle
- Associated with coarctation of the aorta, transposition of the great vessels, VSD, pulmonary and aortic stenosis

Risk Factors

- Premature infants
- Respiratory distress syndrome
- Prophylactic use of surfactant
- Lack of antenatal steroids
- Sepsis
- Liberal fluid therapy

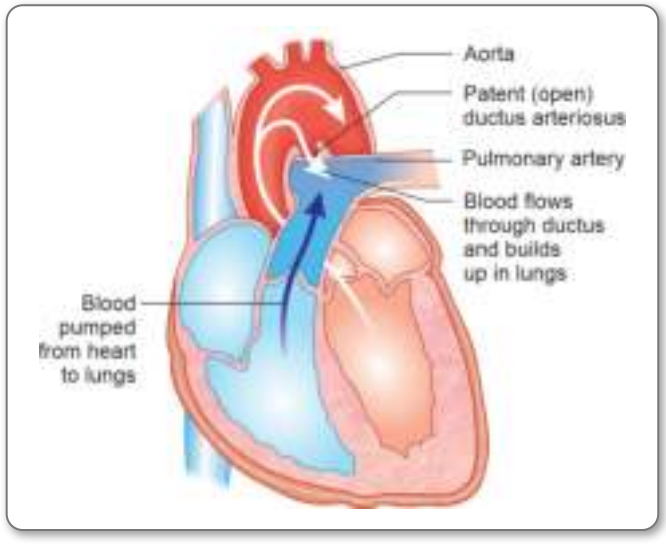


Figure 15.5: Patent ductus arteriosus

Pathophysiology

During fetal life, the ductus arteriosus (DA) allows most of the right ventricular blood to bypass the non-functioning lungs by directing blood from the pulmonary artery to the aorta.

After birth, with the initiation of respiration, DA is no longer necessary. It should functionally close within several hours after birth and anatomically close within several hours after birth.

The smooth muscle in the wall of the ductus arteriosus contracts to obliterate the lumen.

Within several weeks after birth, degenerative changes occur in the ductus arteriosus and it becomes a cord of fibrous connective tissue (ligamentum arteriosum).

When the DA remains patent, oxygenated blood from the higher pressure systemic circuit (aorta) flows to the lower pressure pulmonary circuit (pulmonary artery) through the PDA.

The volume of blood that the heart must pump in order to meet the demand of the peripheral tissues is increased. A greater volume burden is placed on the lungs and also on the left heart. This is an acyanotic defect, as the blood flowing from the aorta is fully oxygenated.

Clinical Manifestations

Small PDA is usually asymptomatic.

Large PDA may develop symptoms in very early infancy like slow weight gain, feeding difficulties, frequent respiratory infections and congestive heart failure.

Other symptoms include systolic or continuous machinery murmur, bounding pulse, widened pulse pressure.

Diagnosis

Auscultation: Continuous machinery like murmur at the left intra-clavicular area is heard in most children.

Wide pulse pressure and/or bounding posterior tibial and dorsalis pedis pulses.

Chest X-ray: Gross cardiomegaly and increased pulmonary vasculature

ECG: May demonstrate left ventricular hypertrophy

Complications

CHF, infective endocarditis

Treatment

- **Prophylactic treatment**
  - **Indomethacin:** It changes oxygen concentration of tissue and enhances tissue changes that close the defect. It is a Prostaglandin inhibitor. It is given in first 24 hours of life for baby <1000 g. Side effect is that it decreases cerebral and renal blood flow.
    Indomethacin – Total 3 doses, IV
    0.2 mg/kg stat
    <2 days – 0.1 mg/kg 12 hourly 2 doses
    2–7 days – 0.2 mg/kg/dose 12 hourly 2 doses
    >7 days – 0.25 mg/kg/dose 12 hourly 2 doses
  - **Ibuprofen:** Prostaglandin synthesis inhibitor
    - 10 mg/kg stat
    - 5 mg/kg 2 doses at 24-hour intervals PO
    - It has less side effects than indomethacin
- Fluid restriction
- Diuretics: Lasix
- Avoid hypoxia and acidosis
- Prophylactic antibiotics
- Surgical ligation (clipping) is performed. Endovascular coils can be placed in the PDA. Surgical division of the PDA is done in early infancy if CHF develops and cannot be controlled and electively by 2–3 years of age.

Cyanotic Defects (Right to Left Shunt)

This includes TOF, TGA, Truncus Arteriosus, TAPVC, and Tricuspid Atresia.

Tetralogy of Fallot (TOF)

It consists of four abnormalities (Fig. 15.6):

1. VSD
2. Overriding aorta: An aorta that arises from both ventricles, rather than exclusively from the left ventricle.
3. Right ventricular hypertrophy: Thickened muscular wall of the right ventricle. It is acquired due to increased pressure within the right ventricle.
4. Right ventricular outflow stenosis or atresia (Pulmonic stenosis)

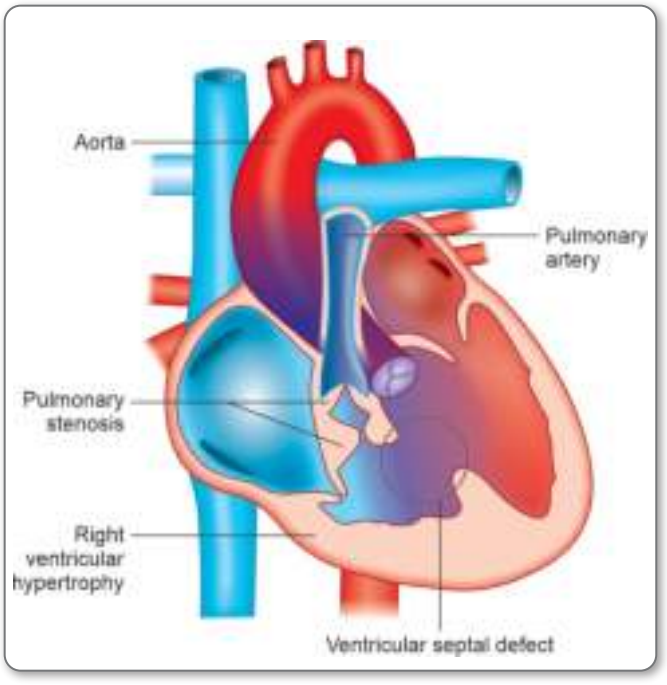


Figure 15.6: Tetralogy of Fallot

Pathophysiology

Obstruction of the blood flow from the right ventricle to the pulmonary circulation is caused by obstruction at the pulmonary valve level or the infundibular area of the right ventricle below the pulmonary valve. Deoxygenated blood is shunted from the right ventricle through the VSD directly into the aorta. The right ventricle is hypertrophied because of high right ventricular pressure.

Clinical Manifestations

Clinical manifestations depend on the size of VSD and the degree of right ventricular outflow obstruction.

- **Cyanosis:** Initially, the shunt through the VSD is mainly from left to right. Many infants with this defect are not cyanotic at birth, but they develop cyanosis as they grow and as the stenosis becomes relatively more severe. Cyanosis may first be observed only with exertion and crying, but during the first years of life, the child may become cyanotic at rest. Infundibular stenosis may be minimal so that cyanosis never develops.
- Clubbing of fingers and toes.
- Squatting/knee chest position during episodes of cyanosis
- Slow weight gain.
- Dyspnea on exertion.
- Hypoxic spells, transient cerebral ischemia.

Diagnosis

- **Auscultation:** Single second sound
- Systolic ejection murmur at the second and third interspaces to the left of the sternum.
- **Inspection:** Prominent left chest with right ventricular heave.
- **Chest X-ray:** Heart size normal, pulmonary segment small and concave (boot-shaped heart), diminished pulmonary vascular markings
- **ECG:** Right axis deviation, right ventricular hypertrophy
- Cardiac catheterization
- Angiocardiography
- **Lab data:** Polycythemia, increased hematocrit.

Complications

- Congestive heart failure may occur in newborn but is uncommon beyond infancy.
- Infective endocarditis
- Cerebral vascular accident due to thrombosis or severe hypoxia.
- Brain abscess
- Iron deficiency anemia

Treatment

- **Knee chest position:** Older children will often squat during a tet spell, which increases systemic vascular resistance and allows for a temporary reversal of the shunt
- Oxygen and morphine administration to improve oxygenation during tet spells
- Propranolol (inderal) to prevent tet spells
- Prophylactic antibiotics
- Vasopressors, correct anemia and acidosis
- Feeding the child slowly
- Giving smaller, more frequent meals
- Decreasing the child's anxiety by remaining calm
- Minimizing crying by trying to anticipate the child's needs
- Recruiting others to care for the child to prevent parental exhaustion and burn-out

Surgical Management

Goal: To improve oxygenation of arterial blood.

- **Palliative:**
  - **Blalock-taussig shunt:** Anastomosis between the right or left subclavian artery and the right pulmonary artery. (Fig. 15.7)
  - **Waterson shunt (RPA):** Anastomosis between the posterior lateral aspect of the ascending aorta and the right pulmonary artery.
  - **Potts shunt (LPA):** A descending aorta to the left pulmonary artery anastomosis. A direct side to side anastomosis.
- **Total correction:**
  - Removal of shunt is advised if previously performed. With cardiopulmonary bypass, VSD is repaired and the right ventricular outflow obstruction is relieved. Total correction is advocated for infants in whom pulmonary arteries are of sufficient size.
- **Specific points:**
  - Teach parents to recognize tet spells to get emergency treatment.
  - If child has undergone BT procedure, do not use the arm on the operative side for measuring blood pressure, inserting IV lines or drawing blood samples because blood perfusion on this side diminishes greatly until collateral circulation develops.

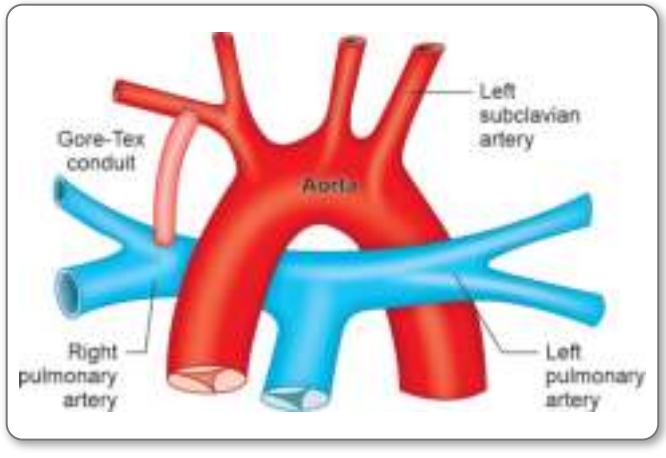


Figure 15.7: Blalock-Taussig shunt

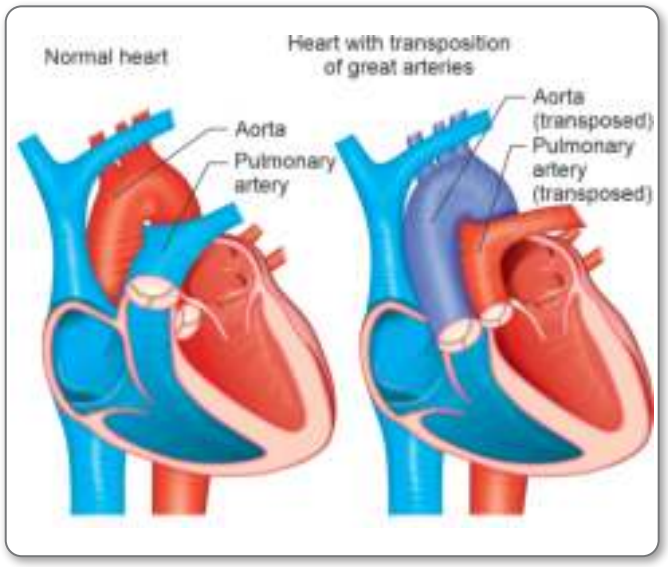


Figure 15.8: Transposition of great arteries

Transposition of Great Arteries (TGA)

TGA is a congenital heart defect in which the two major vessels that carry blood away from the heart, i.e., the aorta and the pulmonary artery are switched (transposed). TGA occurs when the pulmonary artery originates posteriorly from the left ventricle and the aorta originates anteriorly from the right ventricle (Fig. 15.8).

Pathophysiology

- This defect results in two separate circulations; the right heart manages the systemic circulation and the left heart manages the pulmonary circulation.
- In order for life to be sustained, there must be an accompanying defect which provides for the mixing of oxygenated and deoxygenated blood between the two circulations.
- The mixing of oxygenated and deoxygenated blood occurs through one or more of the following shunts: ASD, VSD, PDA, Patent foramen ovale.

Clinical Manifestations

Clinical manifestations are influenced by the extent of inter-circulatory mixing. Cyanosis usually develops shortly after birth. Congestive heart failure is manifested by tachypnea, cardiomegaly, hepatomegaly, tiredness, and slow weight gain and clubbing of fingers and toes.

Diagnosis

- **Auscultation:** Murmurs may be absent in infancy
- **Chest X-ray:** Cardiomegaly, narrow mediastinum, egg-shaped cardiac silhouette, increased vascular markings
- **ECG:** Right axis deviation, right or biventricular hypertrophy
- **Lab tests:** Polycythemia, increased hemoglobin and hematocrit
- Echocardiogram
- Cardiac catheterization
- Angiocardiography

Complications

- Congestive heart failure
- Infective endocarditis
- Brain abscess
- Cerebral vascular accident due to thrombosis or severe hypoxia

Treatment

- Management of CHF
- Prostaglandin E
- Prophylactic antibiotics

Palliative Procedure

- **Rashkind procedure:** The creation of an ASD with a balloon catheter during cardiac catheterization
- **Blalock–Hanlon procedure:** Surgical creation of ASD
- **Pulmonary artery banding:** Indicated for infants with VSDs with large pulmonary blood flow
- **Systemic pulmonary anastomosis:** Indicated for infants with severe pulmonary stenosis

Complete Correction

- **Mustard procedure:** With cardiopulmonary bypass, the atrial septum is removed and a baffle of Dacron and/or pericardium is sutured in place in such a way that the pulmonary venous blood is directed toward the right ventricle and the systemic venous blood is directed toward the left ventricle.
- **Senning procedure:** With the Senning surgical repair, a baffle – or conduit - is created within the atria that reroutes the deoxygenated blood coming from the inferior and superior venae cavae to the mitral valve and therefore to the pulmonary circulation.
- **Rastelli procedure:** It is the surgery of choice for transposition with VSD and left ventricular outflow tract obstruction. With cardiopulmonary bypass, the VSD is closed in such a way that the left ventricle communicates with the aorta. The pulmonary artery is ligated and the right ventricle is connected to the distal portion of the pulmonary artery by means of a valve bearing tubular graft.
- **Arterial switch operation:** The aorta and pulmonary artery are switched back to their normal positions. The aorta is connected to the left ventricle, and the pulmonary artery is connected to the right ventricle. The coronary arteries, which carry the oxygen-rich blood that nourishes the heart muscle, also need to be re-attached to the new aorta.

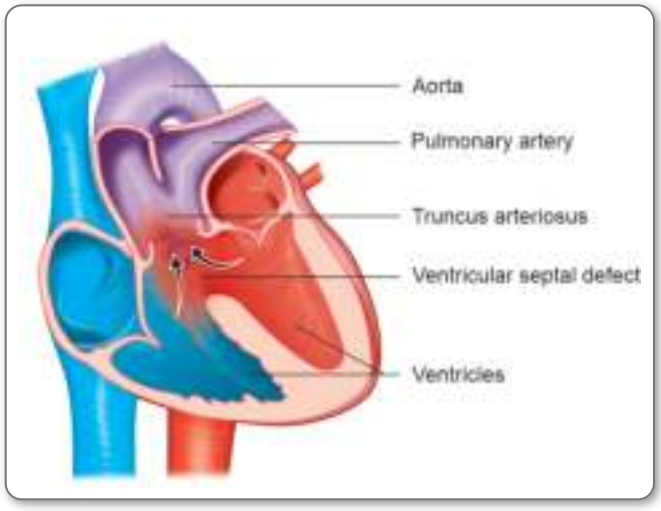


Figure 15.9: Truncus arteriosus

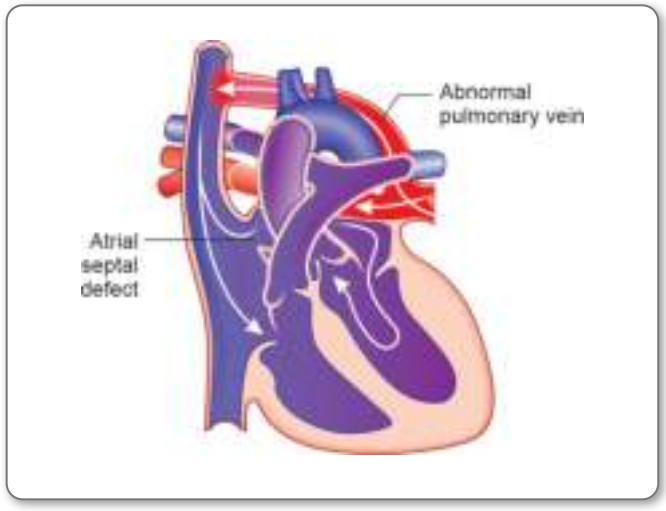


Figure 15.10: Total anomalous pulmonary venous connection

Truncus Arteriosus

In Truncus arteriosus, there is failure of normal septation and embryonic division of pulmonary artery and aorta resulting in a single vessel that overrides both ventricles giving rise directly to the pulmonary and systemic circulations (Fig. 15.9). Blood from both the ventricles enters the common artery and flows either to the lungs or the aortic arch and body.

Symptoms

Cyanosis, LVH, dyspnea, marked activity intolerance and growth retardation.

In case of harsh murmur audible, congestive heart failure usually develops.

Management

- **Palliative treatment:** Banding both pulmonary arteries to decrease the amount of blood going to lungs.
- **Corrective treatment:** Closing VSD so truncus originates from LV and creating pathway from RV.

Total Anomalous Pulmonary Venous Connection (TAPVC)

TAPVC is a defect in which all four pulmonary veins drain into systemic veins or the right atrium with or without pulmonary venous obstruction (Fig. 15.10). Systemic and pulmonary venous blood mix in the right atrium. An atrial defect or foramen ovale is important in left ventricular output both in fetal and in newborn circulation. Because all pulmonary venous return connects to the systemic venous system, right atrial and right ventricular enlargement occurs, and, if significant pulmonary venous obstruction develops, right ventricular hypertrophy occurs. If the foramen ovale is restrictive, right atrial pressure elevates, and systemic and pulmonary venous congestion both occur. Pulmonary blood flow increases, and pulmonary artery hypertension may occur.

Physical examination findings include severe cyanosis with significant respiratory distress.

ECG reveals significant right ventricular hypertrophy in most of these patients. Other tests done are Chest X-ray, Echo.

Surgical Care

The goal of surgery is to redirect pulmonary vein flow to the left atrium. In patients with a supracardiac or infracardiac connection, the common pulmonary vein is opened and connected side to side to the left atrium. The foramen ovale is closed, and the ascending or descending vein is ligated.

The pulmonary vasodilators like inhaled nitric oxide, magnesium sulphate and alprostadil can be used to treat elevated pulmonary vascular resistance in the postoperative period.

Tricuspid Atresia

Tricuspid atresia is a condition in which there is atresia of the tricuspid valve so that there is no communication between the right atrium and the right ventricle, interatrial septal defect and a hypoplastic right ventricle (Fig. 15.11).

Pathophysiology

Blood from the systemic circulation is shunted from the right atrium through an interatrial communication to the left atrium and then to the left ventricle. Pulmonary blood flow is established either through a PDA, bronchial circulation or a VSD.

Clinical Manifestations

- Severe cyanosis in the neonatal period
- Respiratory distress
- Clubbing, hypoxic spells
- Delayed weight gain
- Right heart failure may occur

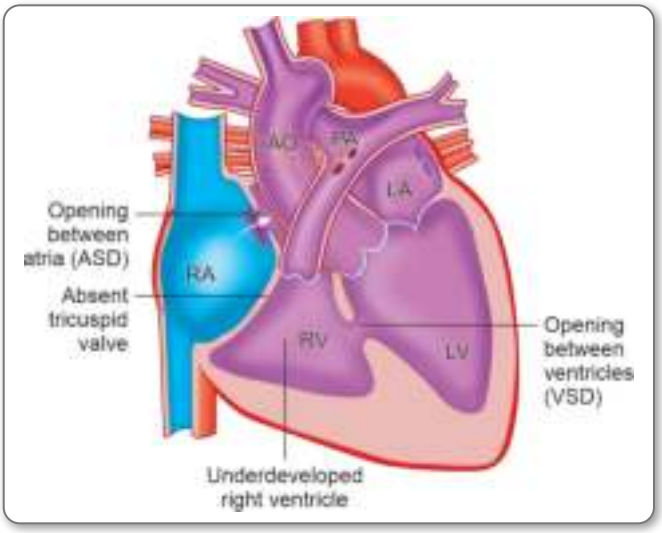


Figure 15.11: Tricuspid atresia

Diagnosis

- **Auscultation:** First sound single and accentuated, commonly no murmurs.
- **Chest X-ray:** Patients with diminished pulmonary blood flow have a normal to mildly increased cardiac silhouette, concavity in the region of main pulmonary artery and diminished pulmonary vascular markings
- **ECG:** Right atrial, left atrial, left ventricular hypertrophy, left axis deviation.
- Echocardiogram
- Cardiac catheterization
- Angiocardiography

Complications

- Cerebrovascular accidents
- Brain abscess
- Bacterial endocarditis

Treatment

- **Prostaglandin E:** To maintain ductal patency
- **Surgical treatment:** Shunt (or bypass) from the aorta to the pulmonary arteries.
- **Fontan operation:** Where the systemic venous return is connected to the pulmonary arterial tree

Obstructive Lesions

This includes Aortic Stenosis, Pulmonic Stenosis and Coarctation of Aorta.

Aortic Stenosis

Aortic valve stenosis occurs when there is obstruction to the left ventricular outflow at the level of the valve (Fig. 15.12A). This is the most common form of aortic stenosis, others being hypertrophic subaortic stenosis and supravalvular stenosis.

Pathophysiology

Blood flows from the left ventricle through the obstructed aortic valve into the aorta. Left ventricular pressure increases to overcome the resistance of the obstructed valve. Myocardial ischemia may occur as a result of an imbalance between the increased oxygen requirements of the hypertrophied left ventricle and the amount of oxygen that can be supplied to the myocardium.

Clinical Manifestations

Rarely symptomatic during infancy; in severe cases, infants may demonstrate evidence of decreased cardiac output, such as faint peripheral pulses or exercise intolerance. Older children may experience chest pain, dyspnea and fatigue with exertion, narrow pulse pressure and weak peripheral pulses.

Diagnosis

- **Auscultation:** Harsh, low pitched systolic ejection murmur, maximal at the 2nd right intercostal space, radiates to apex, back and neck. Ejection click at 4th interspace to the left of the sternum, single or narrowly split S2.
- **Chest X-ray:** Dilated ascending aorta and varying degrees of left ventricular enlargement
- **ECG:** Normal or left ventricular hypertrophy, T wave inversion in severe stenosis, ST segment depression indicates myocardial ischemia
- Cardiac catheterization
- Angiography

Complications

- Congestive heart failure (CHF)
- Myocardial infarction
- Bacterial endocarditis
- Sudden death

Treatment

Aortic valvulotomy or prosthetic valve replacement.

Pulmonary Stenosis

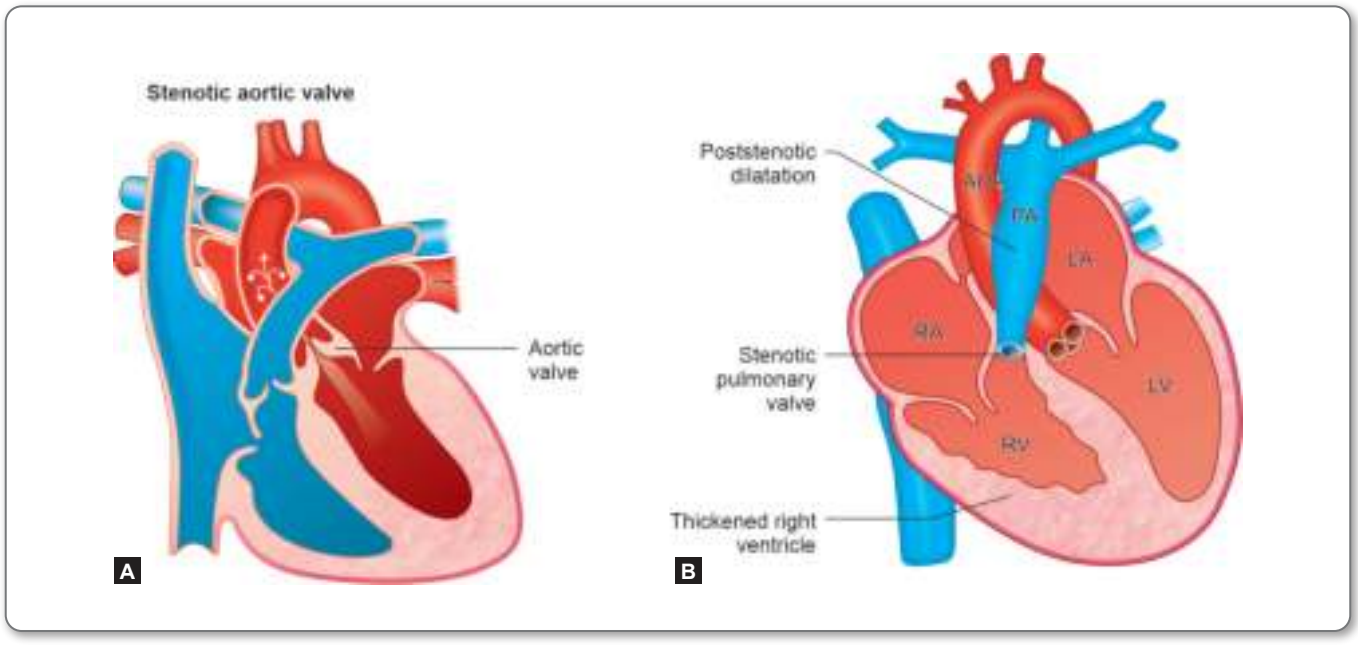
Pulmonary stenosis refers to any lesion that obstructs the flow of blood from the right ventricle. (Fig. 15.12B).

Pathophysiology

Blood flows from the RV through the obstructed pulmonary valve into the pulmonary artery. RV pressure increases to maintain normal cardiac output. RV hypertrophy develops. Right-sided heart failure occurs in severe cases.

Clinical Manifestations

- Generally asymptomatic, child may have decreased exercise tolerance. With severe obstruction, child may have dyspnea, generalized cyanosis.
- Pericardial pain.



Figures 15.12A and B: **A.** Aortic stenosis; **B.** Pulmonary stenosis

Diagnosis

- **Auscultation:** Systolic ejection murmur over pulmonic area, ejection click, widely split S2.
- **Chest X-ray:** Right ventricular enlargement, normal pulmonary vascularity and normal left side. In severe stenosis, right atrial hypertrophy is also observed.
- **ECG:** Moderate or severe cases demonstrate right ventricular hypertrophy.
- Other tests are Cardiac catheterization, Angiocardiography and Ultrasound

Complications

- Anoxic spells in infants with severe lesions
- Bacterial endocarditis
- Sudden death at any age

Treatment

- Valvulotomy

Coarctation of Aorta (COA)

COA is a narrowing or constriction of the vessel at any point (Fig. 15.13). Most commonly the constriction is located just distal to the origin of the left subclavian artery in the vicinity of the ductus arteriosus.

Pathophysiology

The narrowing of the aorta obstructs blood flow through the constricted segment of the aorta, thus increasing left ventricular pressure and workload. Collateral vessels develop arising chiefly from the branches of the subclavian and intercostal arteries, bypassing the coarcted segment of the aorta and supplying circulation to the lower extremities.

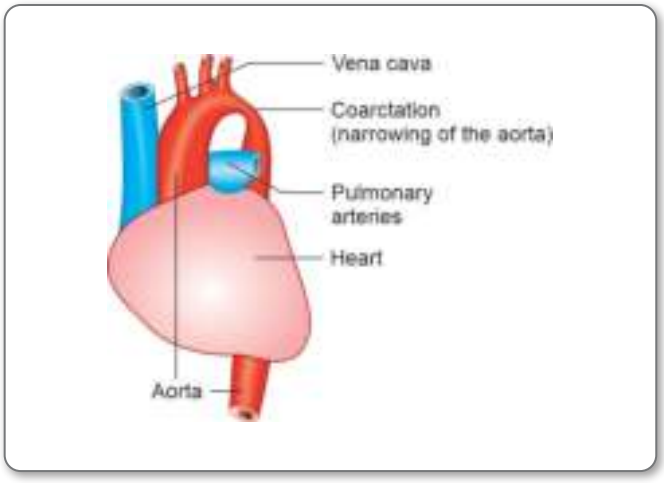


Figure 15.13: Coarctation of aorta

Clinical Manifestations

- Occasional fatigue, headache, nosebleed, leg cramps.
- Absent or reduced femoral pulses.
- Hypertension in upper extremities and diminished blood pressure in lower extremities
- Growth failure, tachypnea, dyspnea, severe congestive heart failure.

Diagnosis

- **Auscultation:** Nonspecific systolic murmur heard along the left sternal border
- **Chest X-ray:** Prominent aorta, rib notching, cardiomegaly
- **ECG:** Left ventricular hypertrophy
- Cardiac catheterization, Angiocardiography, Ultrasound

Complications

- Cerebral hemorrhage
- Aortic rupture
- Infective endocarditis
- Congestive heart failure

Treatment

- **Prostaglandin E infusion:** To reopen or maintain PDA
- **Inotropic agents:** Dopamine, dobutamine
- Diuretics
- Prophylactic antibiotics to prevent infective endocarditis
- Balloon angioplasty
- **End to end anastomosis:** With surgery, the narrowed segment of the aorta is removed, then repaired by anastomosis (placing the two free ends of the aorta back together) if the segment with the coarctation was short. If a longer segment must be removed, a Dacron graft (a synthetic material) is used to fill the gap. Surgery is done within 3–6 years if asymptomatic or within first 6 months if symptomatic.
- **Subclavian flap aortoplasty:** The distal subclavian artery is divided and the flap of the proximal portion is used to expand the coarcted aorta.

NURSING CARE OF A CHILD WITH CARDIAC SURGERY

Preoperative Care

A child must be stable before cardiac surgery. They must exhibit no signs of an upper respiratory tract infection or have a fever.

- Assess baseline vital signs (blood pressure, temperature, pulse, respirations, and oxygen saturation)
- Record height and weight.
- Check laboratory values, like hematology, and coagulation values. Blood products must be typed and cross-matched.
- Ensure that consents for surgery and anesthesia have been signed by the family and the appropriate provider.
- Discuss about surgical procedure and postoperative course with the child and family in a developmentally appropriate manner.
- Explain about equipment such as a cardiac monitor, oxygen, IV equipment, chest tubes and ventilator.

Postoperative Care

- Risk for ineffective cardiopulmonary tissue perfusion related to depressed myocardial function after surgery
  - Administer IV fluids
  - An indwelling urinary catheter is inserted at the time of surgery so urine output can be carefully measured and recorded postoperatively. Preferred urine output after surgery is approximately 0.5 to 1 mL/kg/hr for infants.
  - Laboratory tests such as arterial blood gases, hemoglobin, hematocrit, clotting time, and electrolytes (particularly sodium, potassium, calcium, and magnesium) should be monitored closely to assess postoperative hemodynamic status.
  - IV inotropic agents such as milrinone or dopamine may be added to enhance cardiac output and decrease afterload.
- Risk for excess or deficient fluid volume related to fluid shifts during cardiac surgery
  - Children may develop relative hypovolemia after cardiac surgery because of increased production of aldosterone by the adrenal glands and an increase in antidiuretic hormone secretion by the pituitary gland in response to stress. Cardiopulmonary bypass also causes fluids to shift from the intravascular system to the interstitial spaces because of an inflammatory response that causes leaky capillaries. After surgery, as this fluid returns by osmosis to the vessels and is excreted with the help of diuretics, balance is restored.
  - Monitor central venous pressures carefully to evaluate preload to determine which type of therapy is necessary. IV fluids may be given if the circulating volume is low as indicated by a low CVP. It is usually preferred that the product given have a high oncotic pressure, as with red blood cells or albumin, in order to prevent further leakage across dilated capillaries. If the CVP is high, diuretics and restricted IV fluid intake will be necessary.
- Impaired gas exchange related to atelectasis/pleural effusion
  - Perform endotracheal suctioning when child is on ventilator to prevent secretion accumulation in the lungs or obstruction of the endotracheal tube.
  - Assist with chest physiotherapy (percussion and vibration) to remove lung secretions.
  - As soon as the ET tube and ventilator are removed, encourage the child to cough and breathe deeply or use an incentive spirometer at hourly intervals to mobilize secretions.
  - Administer prescribed analgesia. Teach children how to use a pillow as a splint while coughing and deep breathing.
  - Suggest games such as blowing cotton balls or bubbles, or using the incentive spirometer, to help achieve lung expansion.
- Risk for ineffective peripheral tissue perfusion related to dysrhythmias.

  After cardiac surgery, child is at risk for dysrhythmias due to damage to the conduction system, inflammation of or injury to the heart itself, or electrolyte imbalance. In anticipation of possible dysrhythmias, children may have two pacing wires attached to the surface of the heart and brought out through the chest wall. These are temporary pacemaker wires that can be attached to a pacemaker at the bedside in the event the child experiences a dysrhythmia or

heart block. If no evidence of abnormal rhythms has been seen after several days, the pacing wires will be pulled out.

- Perform cardiac monitoring. Monitor ECG to identify any dysrhythmia.
- Identify need for permanent pacemaker: If a child is dependent on the pacemaker due to heart block and normal conduction does not spontaneously return after 14 days, the child is typically taken to the operating room for placement of a permanent pacemaker.

  The pacemaker is placed in the abdominal cavity in infants and small children. For older children and adolescents, the pacemaker is placed in the left shoulder area, and the wires are threaded transvenously to the right heart. Currently, permanent pacemaker batteries last approximately 5 years. Having a permanent pacemaker requires monthly transmissions to document pacemaker function and will limit the child's ability to participate in contact sports.

  Once a child is hemodynamically stable, the child may be moved from the ICU to a routine patient care unit.

  Once on the floor, general nursing care includes the following:
  - Watching for blood loss
  - Preventing infection
  - Preventing/managing pain
  - Maintaining lung expansion
  - Encouraging mobility
  - Encouraging healthy eating
  - Including the family in care
- Risk for infection related to surgical incision
  - Continue antibiotic therapy for 24–48 hours after chest closure.
  - Frequently monitor the temperature and assess the surgical incision site and the points of insertion of the thoracotomy tubes or central catheters for drainage and erythema. This helps in identifying infection.
  - Infants must wear shirts and bibs when feeding to prevent formula from dropping on the incision site. Older children should wear T-shirts at all times. This not only helps with food but also keeps them from scratching the site or pulling at the butterfly closures.
- Parental anxiety related to lack of knowledge of postoperative routine and exercises
  - Encourage parents to participate in their child's care during this period. Parents can encourage their child to routinely perform breathing exercises, eat and drink throughout the day, and get out of bed.
  - Caution parents not to pick up an infant under the arms to move or hold the child because this pulls on the chest incision. Show them how to lift an infant by placing their hands behind the neck and scooping under the buttocks instead. Teach parents to help an older child out of bed by wrapping their arms around the child's chest. Do not pull on the child's arms.

CONGESTIVE HEART FAILURE (CHF)

Definition

Heart failure is a syndrome manifesting as the inability of the heart to fill with or eject blood due to any structural or functional cardiac conditions.

Causes

Common causes of heart failure include:

- Congenital heart diseases
- Left heart obstructive disease
  - Hypoplastic left heart syndrome
  - Aortic Stenosis
  - Aortic Coarctation
- Large shunt with pulmonary overflow
  - Ventricular Septal Defect
- Pump failure
  - Myocarditis
  - Cardiomyopathy

Pathophysiology

CHF occurs when the heart can no longer meet the metabolic demands of the body at normal physiologic venous pressures and cardiac output is decreased.

Cardiac output is the volume of blood pumped by the ventricles each minute. The formula for calculating cardiac output is heart rate multiplied by stroke volume. Stroke volume is affected by the following factors:

- **Preload:** The volume of blood in the ventricles at the point just before contraction; it is an indicator of circulating blood volume.
- **Contractility:** Ability to modulate the rate and force of fiber shortening
- **Afterload:** Amount of resistance met by the ventricles upon ejection
- **Compliance:** The ability of the ventricles to stretch and fill

Infants and children can easily modify their heart rate in an effort to increase cardiac output, but they are not as able to change their stroke volume. Much of the therapy for pediatric heart failure is aimed at altering those factors that affect stroke volume, in particular, reducing preload and afterload and increasing contractility to promote better cardiac output.

When the heart begins to perform inadequately, several physiologic responses occur. The initial neurohormonal response to decreased cardiac output is peripheral vasoconstriction and fluid retention. The sympathetic nervous system provides a rapid response by increasing heart rate, stimulate myocardial contractility, and promote regional vasoconstriction. Activation of the renin–angiotensin–aldosterone system (RAAS) occurs, which stimulates renal fluid retention to expand vascular volume. The body's goal is to maintain adequate blood pressure

and circulating blood volume. Stroke volume and cardiac output initially improve, but as the underlying etiology is addressed, the heart eventually becomes overloaded from the extra volume and cannot maintain adequate contractility against the increased afterload.

In addition, the vasoconstriction and redistribution of blood flow to ensure adequate perfusion of vital organs (brain and heart) occur at the expense of skin, intestinal, and renal blood flow. This leads to lactic acidosis secondary to anaerobic metabolism, fatigue, an intolerance to oral intake, decreased urine output, and electrolyte imbalance.

As the demands on the heart exceed the normal range of physiologic compensatory mechanisms, signs of congestive heart failure occur. These signs include tachycardia; venous congestion; high catecholamine levels; and, ultimately, insufficient cardiac output with poor perfusion and end-organ compromise.

Clinical Manifestations

Infants and young children may present with poor feeding and failure to thrive, whereas older children may complain of fatigue, exercise intolerance, and breathlessness. The infant will tire easily, may be diaphoretic, or may have difficulty sucking because of the dyspnea and the energy required to eat. A child may gain weight from fluid overload, but more frequently, an infant will lose weight because their caloric expenditure is greater than the caloric intake.

Right-sided failure leads to hepatomegaly, increased venous pressure noted in jugular venous distention in older children, or periorbital edema.

Left-sided failure results in increased pulmonary pressures, rales, tachypnea, and shortness of breath. With left-sided heart failure, back pressure causes blood to accumulate in the pulmonary system, causing orthopnea. The child may demonstrate use of accessory muscles to support their breathing, which causes intercostal, substernal, and/or suprasternal retractions. The sound of rales (crackles) can be heard on auscultation.

Diagnosis

- Pulse oximetry, Echo, ECG
- CBC count and hemoglobin concentration, electrolyte levels, calcium level, BUN level, creatinine level, and renal and hepatic function. The CBC count is evaluated for signs of anemia or infection.
- Brain natriuretic peptide (BNP) or *N*-terminal prohormone BNP (NT-proBNP) levels are elevated due to ventricular dilation.

Medical Management

The goals of medical management for CHF include reducing the preload, enhancing cardiac contractility, reducing the afterload, improving oxygen delivery, and enhancing nutrition.

Preload reduction is achieved with diuretics, (e.g., furosemide, thiazides, metolazone). Venous dilators, (e.g., nitroglycerin) can be administered.

Contractility can be supported with intravenous agents, (e.g., dopamine) or mixed agents, (e.g., dobutamine, inamrinone, milrinone) and digoxin.

Afterload reduction is obtained by administration of ACE inhibitors or other agents such as hydralazine, nitroprusside, and alprostadil. Care must be taken when planning a medication schedule for a patient on a diuretic and an ACE inhibitor. If the two medications reach peak effect at the same time, the child is at risk for hypotension. Therefore, the nurse must ensure the medications are given 1 to 2 hours apart in order to prevent this complication.

Nursing Management

- Altered cardiac output related to inadequate heart function
  - To improve cardiac output, administer digoxin. Dose not more than 0.05 mg. Monitor HR, check pulse for 1 full minute, withhold if pulse is <90-110. Monitor serum K levels as Hypokalemia enhances digoxin toxicity. Monitor for signs of digoxin toxicity, like nausea, vomiting, anorexia, bradycardia, dysrhythmias. Check ECG.
- Altered fluid balance, greater than body requirements due to poor myocardial function and fluid retention.
  - Monitor I/O, weight
  - Administer diuretics
  - Provide skin care
  - Change position frequently
- Ineffective breathing pattern
  - Auscultate lung sounds, assess vitals
  - Monitor SpO_2 levels by pulse oximetry
  - Position for maximal lung expansion
  - Administer humidified oxygen
- Risk for imbalanced nutrition, less than body requirements, related to easy fatigability
  - Give small frequent, high calorie feeds
  - Administer gavage feeds
  - Give planned care
  - Provide thermoneutral environment
- Risk for infection
  - Follow standard precautions
  - Strict hand washing before and after every procedure
  - Administer antibiotics
  - Restrict visitors
- Anxiety related to disease condition and possible outcome
  - Provide comfortable environment
  - Explanation of procedures and equipment should be done
  - Provide counseling to caregivers, encourage questioning

RHEUMATIC FEVER

Definition

Rheumatic fever is an inflammatory disease which may develop after an infection with **streptococcus bacteria** such as strep throat or scarlet fever and can involve the heart, joints, skin, and brain. Rheumatic fever primarily affects children between ages 6 and 15 and occurs approximately 20 days after strep throat or scarlet fever.

Causes

- Altered host resistance to streptococcal infections
- Malnutrition
- Crowded living condition

Pathophysiology

Rheumatic fever is an autoimmune disease which occurs after an untreated Group A streptococcal infection, typically a throat infection. Person suffering from rheumatic disease will develop antibodies against that streptococci. The antibodies formed against these proteins sometimes cross-react with normal tissue causing damage and leading to clinical symptoms. Streptococcal antigen and human myocardium are identical antigenically. These antibodies have capacity to attack connective tissue in heart and muscle since they are antigenically similar. Antibodies produce characteristic lesions at specific tissue sites especially in the connective tissue, heart and joints. It results in antigen antibody complexes that ultimately destroy heart tissue. RHD refers to cardiac manifestations of rheumatic fever like pancarditis (myocarditis, pericarditis, endocarditis) during the early acute phase or chronic valvular disease later.

Manifestations/Diagnosis

Jones Criteria for the Diagnosis of Rheumatic Fever

Diagnosis of rheumatic fever requires two major manifestations or one major and two minor manifestations along with evidence of preceding Streptococcus infection.

Major Criteria (Fig. 15.14)

- **Carditis:** Inflammation of the heart muscle which can manifest as congestive heart failure with shortness of breath, pericarditis with a rub, or a new heart murmur.
- **Migratory polyarthritis:** A temporary migrating inflammation of the large joints, usually starting in the legs and migrating upwards.
- **Sydenham's chorea (St. Vitus' dance):** A characteristic series of rapid movements without purpose of the face and arms. This can occur very late in the disease.
- **Erythema marginatum:** A long lasting rash that begins on the trunk or arms as macules and spread outward to form a snakelike ring while clearing in the middle.

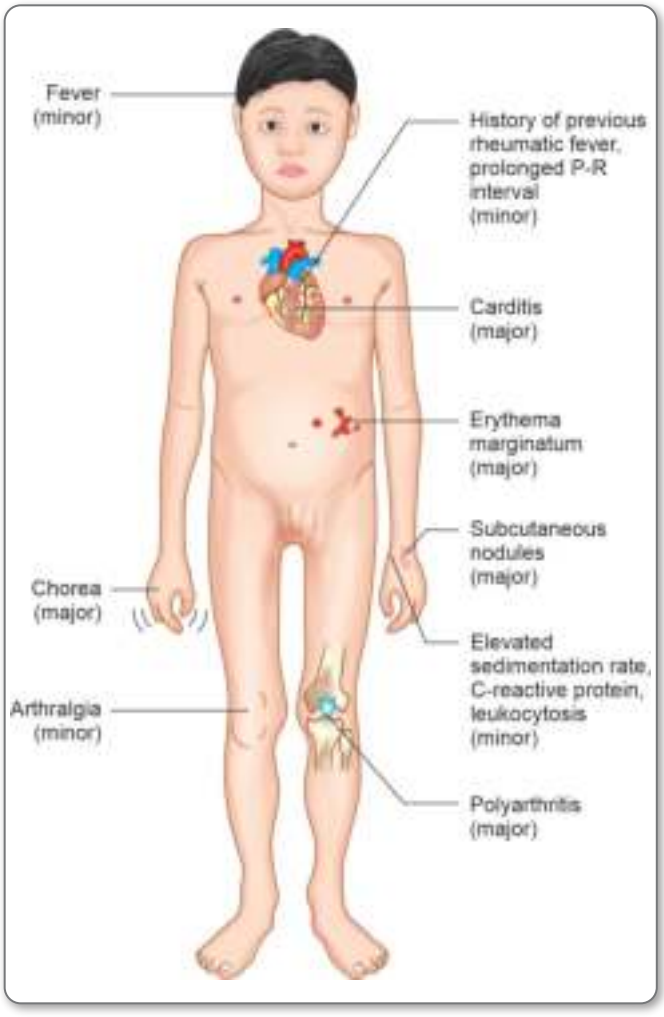


Figure 15.14: Symptoms of rheumatic fever

- **Subcutaneous nodules (a form of Aschoff bodies):** Painless, firm collections of collagen fibers on the back of the wrist, the outside elbow, and the front of the knees. These now occur infrequently.

Minor Criteria

- **Clinical criteria include:**
  - **Fever:** Temperature elevation
  - **Arthralgia:** Joint pain without swelling
  - **Laboratory abnormalities:** Increased Erythrocyte sedimentation rate, increased C-reactive protein, leukocytosis
  - **Electrocardiogram abnormalities:** A prolonged PR interval
  - Previous rheumatic fever or rheumatic heart disease
- **Essential criteria include:**
  - Evidence of Group A Strep infection: positive culture for Group A Strep
  - Elevated or rising Antistreptolysin O titre
  - Positive throat culture
  - Recent scarlet fever

Diagnosis

Important laboratory findings include the presence of an elevated antistreptolysin-O (ASO) titer which is evidence that the child had a recent streptococcal infection. The child will have a fever >38.5°C, and inflammatory markers such as ESR and CRP will also be elevated. Echocardiogram is imperative to evaluate the extent of cardiac involvement. If a child meets the Jones diagnostic criteria and all other potential diagnoses have been ruled out, the child should be treated for rheumatic fever.

Treatment

- Strict bed rest during acute phase
- **Diet:** High calorie, high protein, salt restricted, vitamins and minerals
- Anti-inflammatory medications such as aspirin or corticosteroids for joint pain and inflammation.
- **Antibiotics:** Penicillin to prevent damage from future attacks
- **Analgesics:** Aspirin for arthritis pain
- **Chorea:** Institute safety measures for chorea, maintain a calm environment, phenobarbitone 15–30 mg TDS, valium, promethazine, chlorpromazine, benadryl. Reduce stimulation, avoid the use of forks or glass and assist in walking to prevent injury.
- Penicillin
  - Procaine penicillin G: Therapeutic dose is 40,000 units/kg, IM, OD for 10 days after blood cultures have been obtained.

Prevention

- Identify streptococcal sore throat, pharyngitis,
- Prophylactic management involves long acting benzathine penicillin for q 21 days. It is to be given till patient is 25–30-year-old or lifelong. Dose is 6 lacs units for children up to 6 years, 1.2 mega units for older. Side effect is fever after 24–36 hours.
- For patients who are sensitive to penicillin, give erythromycin 40 mg/kg/24 hours to be given once a day or Oral sulfadiazine 0.5–1 g once a day.
- Valvular surgery for damaged valves.

Nursing Management

Nursing diagnosis: Risk for nonadherence to drug therapy related to knowledge deficit about importance of long-term therapy.

Intervention: Preventive maintenance doses of penicillin are started once initial treatment is completed. These doses are given either as a monthly IM injection of penicillin G benzathine or twice-daily oral tablets of penicillin V potassium. Although the IM injection is more painful but it is considered more effective due to the potential lack of compliance with twice-daily oral medications. Preventive therapy continues until the child is at least 21 years of age as patients who have had acute rheumatic fever are highly susceptible to recurrent rheumatic fever. If the child incurred valve damage, preventive therapy will continue until at least 40 years of age.

INFECTIVE ENDOCARDITIS

Infective endocarditis (IE) is defined as an infection of the endocardial surfaces of the heart—primarily of 1 or more heart valves, the mural endocardium, or a septal defect. Its intracardiac effects include severe valvular insufficiency, intractable congestive heart failure, and myocardial abscesses. If left untreated, IE is inevitably fatal.

Causes

- *P aeruginosa,*
- Enteric gram-negative rods, (e.g., *E. coli, Proteus mirabilis*)
- *Streptococcus pneumoniae*

Signs and Symptoms

- Fever, often low-grade and intermittent
- Heart murmurs
- Petechiae
- **Subungual (splinter) hemorrhages:** Dark red, linear lesions in the nail beds
- **Osler nodes:** Tender subcutaneous nodules usually found on the distal pads of the digits
- **Janeway lesions:** Nontender maculae on the palms and soles
- **Roth spots:** Retinal hemorrhages with small, clear centers (rare)

Diagnosis

- Two blood cultures positive for organisms typically found in patients with IE
- Echocardiogram positive for IE

Management

Antibiotics remain the mainstay of treatment for IE. It includes:

- Penicillin G
- Gentamicin
- Ceftazidime, cefepime
- Vancomycin (if penicillin allergy or high-level penicillin G resistance)

Nursing Management

- Provide a calm, quiet environment for the patient and document his vital signs every 2 to 4 hours. Prepare him

for insertion of a new IV line or a peripherally inserted central catheter for medication administration.
- Continually check the patient's SpO_2. Monitor him for a worsened or returning fever and assess for signs and symptoms of heart failure, such as dyspnea, orthopnea, and crackles. Also perform multisystem assessments to identify systemic embolization that may occur in the brain, spleen, bowel, extremities, or kidneys.
- Before discharge, teach him the importance of checking with his primary care provider about taking prophylactic antibiotics before dental work or procedures that involve the respiratory, genitourinary, or gastrointestinal tract. Also tell him to notify his dentist and all other health care providers about his condition.

ACQUIRED HEART DISEASE

Acquired heart disease is a term used to define heart disorders that develop after birth. Kawasaki disease is the leading cause of acquired heart disease in children.

Kawasaki Disease

Kawasaki disease (mucocutaneous lymph node syndrome) is defined as an acute febrile syndrome associated with generalized vasculitis (inflammation of blood vessels) affecting all blood vessels throughout the body, including the coronary arteries. The vasculitis is a principal and life-threatening symptom because it can lead to the formation of coronary aneurysms which will predispose the child to thrombus formation and a high risk of myocardial infarction.

It has an unknown etiology. Although there does seem to be a genetic predisposition to the syndrome, there is also a strong suggestion of an infectious precursor. It occurs more commonly in winter and spring, with males affected more than females; 76% of children affected are less than 5 years old. It is more common among Americans of Asian or Pacific Island descent.

Kawasaki disease is a diagnosis of exclusion as there is no imaging or laboratory test that can diagnose it. Clinical criteria have been established to help with the diagnosis of Kawasaki disease.

Assessment

Kawasaki disease can be divided into an acute phase (week 1) and subacute phase (weeks 2 and 3). There are certain criteria that a child must meet for this diagnosis, including:

- Prolonged fever (>100.4°F [39°C]) of 5 or more days
- Four or more of the following symptoms:
  - Changes in hands and feet (erythema, edema, peeling)
  - Polymorphous exanthema (diffuse maculopapular rash of the trunk and extremities)
  - Bilateral conjunctivitis without exudates
  - Changes in lips and mouth (erythema, strawberry tongue, dry, cracked lips)
  - Cervical lymphadenopathy (>1.5 cm diameter, usually unilateral)

Changes in laboratory data may also be noted such as thrombocytosis, leukocytosis, elevated ESR and CRP, elevated liver enzymes, and mild anemia. The changes in these infectious and inflammatory markers mandate a thorough investigation into possible causes for the child's symptoms including viral infections, scarlet fever, staphylococcal scalded skin syndrome, juvenile rheumatoid arthritis, or Rocky Mountain spotted fever to name a few. If no definitive cause is determined and the child's symptoms fit the criteria listed, the child is given the diagnosis of Kawasaki disease.

During the acute phase, children are very irritable and uncomfortable from the fever and inflammatory process that causes joint pain. After approximately 10 days, a subacute phase begins. The skin desquamates, particularly on the palms and soles. The platelet count rises, increasing the possibility of clot formation. If an aneurysm forms in a coronary artery, accumulating thrombi can dislodge and lead to obstructed blood flow distally in the coronary artery which will lead to myocardial ischemia and infarction.

Therapeutic Management

Treatment for Kawasaki disease is focused on addressing the immediate symptoms and preventing any long-term consequences. Primarily, supportive measures are necessary as these children are irritable and not taking adequate oral intake so antipyretics for the fever as well as IV fluids are necessary. Kawasaki disease also causes dilation of the coronary arteries and coronary artery aneurysms in 15–25% of untreated children, and these coronary changes can be fatal. Evidence has shown that treating these children within the first 7 to 10 days of the fever with high-dose intravenous immunoglobulin (IVIG) and high-dose aspirin therapy has demonstrated a reduction in the appearance of coronary artery ectasia and aneurysms due to their anti-inflammatory properties. Children are treated with 2 g/kg of IVIG. This dose may be repeated if the fever does not break. High-dose aspirin at 80–100 mg/kg divided four times a day is also continued for 48–72 hours after the fever breaks. The aspirin is then continued at a low dose of 3 to 5 mg/kg/day once a day for 6–8 weeks with no evidence of coronary artery abnormalities. If abnormalities occur, aspirin may be continued indefinitely.

Nursing Management

Nursing Diagnosis: Parental anxiety related to a lack of knowledge regarding Kawasaki disease

Intervention: Inform parents about the disease process and provide support. Inform about follow up echocardiograms for evaluating aneurysms.

Nursing Diagnosis: Discomfort related to swelling of lymph nodes and inflammation of joints

Intervention: Administer antipyretics such as acetaminophen to reduce both the pain and fever. Providing additional comfort measures such as rocking and holding or distraction may help. Try to protect edematous areas from pressure; make certain clothing is not constricting and irritating areas of rash. Applying lip balm helps protect lips from drying and cracking.

Because the child's fever remains high, offer extra fluid to help maintain hydration and reduce mouth tenderness.

Nursing Diagnosis: Inadequate oral intake related to sore mouth

Intervention: Monitor and record the monitor and record child's intake and output. Encourage the child to continue brushing his or her teeth (use a soft toothbrush or a commercial swab), even though the oral mucous membrane is tender, to prevent tooth decay or ulcer formation. Soft, non-irritating foods may be better tolerated. Avoid foods that require chewing or acidic fluids, such as orange juice, which might sting.

Summary

The majority of children who suffer from heart disease have a congenital heart defect. It includes holes in the walls of the heart, narrowed valves, valve leaks, abnormal connections of chambers and vessels, absent or hypoplastic chambers, narrowed or hypoplastic vessels, and sometimes a combination of many defects. Children with obesity are at a significantly increased risk of cardiovascular disease. After a child has been diagnosed with a heart problem, the most effective strategy to treat that problem is decided. Various factors such as the nature and severity of the problem, is taken into account to choose from a variety of treatment options like medications, interventional cardiac catheterization, or surgery.

Assess Yourself

1. Classify congenital heart diseases.
2. Discuss the Jones criteria for diagnosis in case of Rheumatic fever.
3. Explain the medical and nursing management of a child with Congestive heart failure.
4. The three fetal shunts that exist in fetal circulation are ....., ... and .....
5. A child assumes knee chest position in which disorder?
6. Which medicine is used to close PDA? Functional closure of PDA occurs at which age?
7. List the four defects seen in TOF.
8. Name the surgeries done in the following cases:
   a. TGA .....
   b. TOF .....
   c. Tricuspid Atresia .....
9. Hypertension in upper extremities and diminished blood pressure in lower extremities is seen in .....