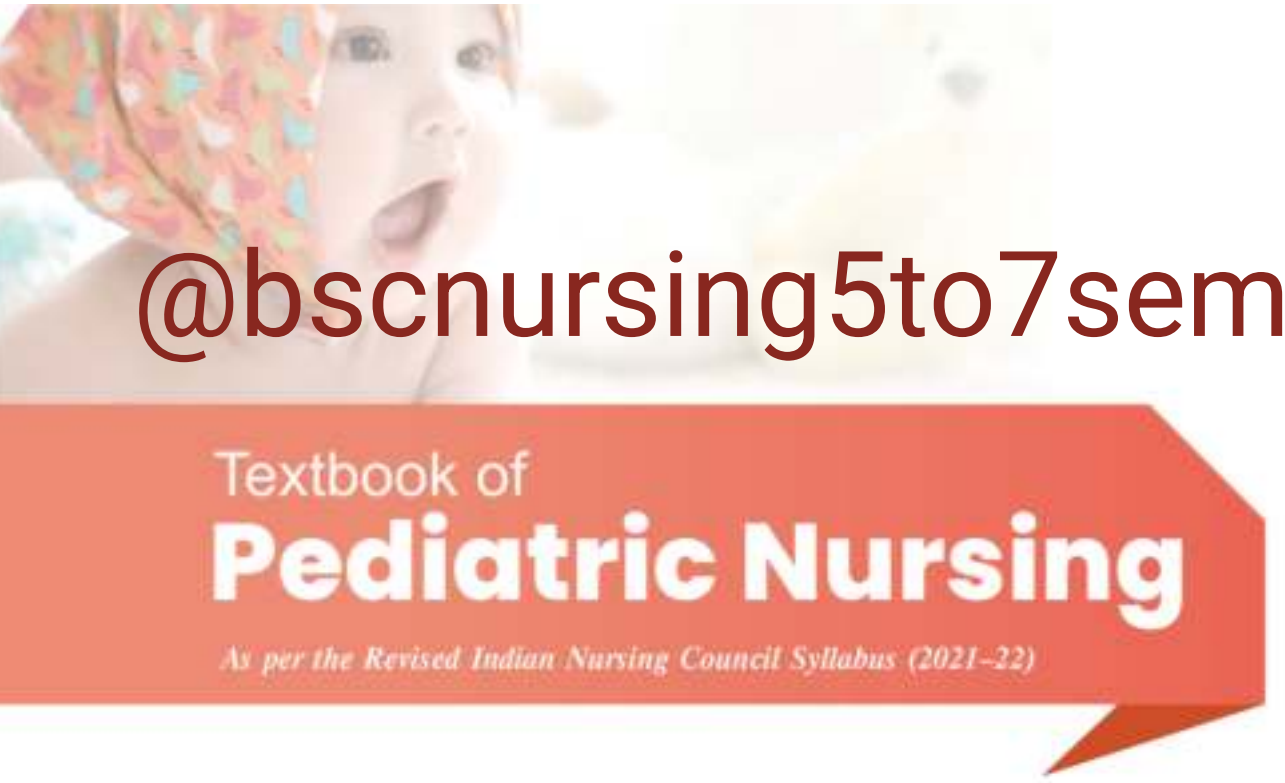

Textbook of Pediatric Nursing

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

Unit V

Childhood Diseases

Learning Objectives

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

- Describe the etiology, pathophysiology, clinical manifestation and nursing management of children with disorders of respiratory system.
- Describe the etiology, pathophysiology, clinical manifestation and nursing management of children with disorders of respiratory system and endocrine system.

Unit Outline

Chapter 12

Respiratory Disorders

Chapter Outline

- Introduction
- Upper Respiratory Tract Infection (URTI)
- Lower Respiratory Tract Infections (LRTI)
- Tuberculosis
- Bronchial Asthma
- Cystic Fibrosis

Common Respiratory Surgical Conditions

- Choanal Atresia
- Congenital Diaphragmatic Hernia (CDH)
- Esophageal Atresia with Tracheo Esophageal Fistula

INTRODUCTION

Respiratory diseases are the most common cause of mortality in children in underdeveloped economies and the commonest cause of morbidity in developed economies. Respiratory disorders include Upper Respiratory Tract Infections (URTI) and Lower Respiratory Tract Infections (LRTIs). URTIs are the most common infectious diseases. They include rhinitis (common cold), sinusitis, ear infections, acute pharyngitis or tonsillopharyngitis, epiglottitis, and laryngitis. Ear infections and pharyngitis cause the more severe complications, like deafness and acute rheumatic fever, respectively. The vast majority of URIs have a viral etiology.

The common Lower Respiratory Tract Infections (LRTIs) in children are croup syndromes, bronchiolitis and pneumonia.

Common congenital anomalies are choanal atresia and congenital diaphragmatic hernia and cystic fibrosis.

Common respiratory symptoms in children include:

- Cough, expectoration
- Respiratory sounds—snoring, grunting, rattling, stridor, wheezing
- Dyspnea, retractions, nasal flaring, use of accessory muscles

Diagnostic methods used to identify respiratory disorders are:

- Bronchoscopy- to visualize larynx, trachea, bronchi
- PFT- to evaluate respiratory system including history and physical examination, spirometry, ABG, Chest X-ray
- ABG – to evaluate acid base imbalance
- Imaging- X ray, CT scan- to identify pneumonia, TB
- Sweat chloride test- used to diagnose cystic fibrosis
- Spirometry – to measure lung function to diagnose asthma, cystic fibrosis
- Laryngoscopy

Let us discuss few important disorders in detail.

UPPER RESPIRATORY TRACT INFECTION (URTI)

Common URTIs include common cold and tonsillitis.

Common Cold (Rhinitis) or Nasopharyngitis

Common cold, nasopharyngitis or sniffles is viral disease of URT caused by adenovirus, influenza, rhino virus, RSV. The mode of transmission is droplet infection. Predisposing factors are chilling, exposure to cold air, overcrowding, allergy.

Clinical Features

Fever, nasal discharge, irritability, enlarged cervical lymph nodes, blocked eustachian tubes leading to otitis media, dry hacking cough, excessive lacrimation as lacrimal ducts in nose are blocked.

Complications

Otitis media, laryngitis, bronchiolitis, bronchopneumonia

Management

- Nasal saline drops to relieve decongestion
- Oral nasal decongestant, like pseudoephedrine
- Antiallergics- cetirizine
- Acetaminophen for fever
- Keep the child warm
- Protect from cold air

Tonsillitis

Definition: Tonsillitis is inflammation and infection of palatine tonsils and adenoids.

Cause: Tonsillitis is caused by Group A beta hemolytic streptococci and some viruses like Adenoviruses, Influenza virus, Epstein-Barr virus, Parainfluenza viruses, Enteroviruses, etc.

Clinical Features

- Edematous, enlarged red tonsils (Fig. 12.1)
- White patches of exudate on tonsils
- Dysphagia, dyspnea
- Enlarged cervical lymph nodes
- Sore throat, fever, earache

Complication

Quinsy- a severe sore throat that quickly gets worse. The signs of quinsy are:

- Swelling inside the mouth and throat
- Difficulty speaking
- Dysphagia, dyspnea
- Difficulty opening mouth

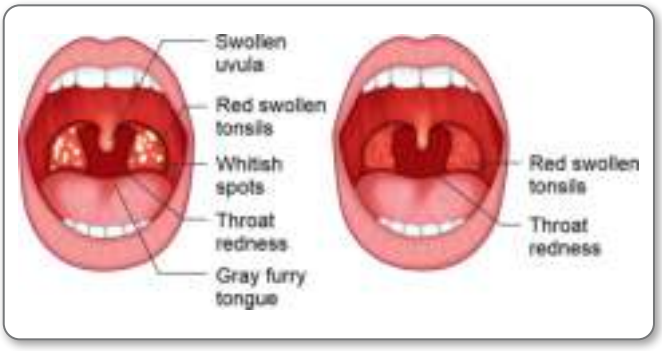

Figure 12.1: Tonsillitis

Diagnosis

Diagnosis can be done by visualization of oropharynx and throat culture.

Management

Medical management: Administer corticosteroids to reduce pharyngeal edema and antibiotics, e.g., penicillin (oral or I/M), clindamycin, vancomycin. Aspirin should not be given to children as it may cause Reyes syndrome.

Supportive management: Ensure that child:

- Gets maximum rest
- Drinks plenty of fluids
- Eats soft foods
- Takes warm liquids or cold foods, like popsicles to soothe the throat
- Isn't around cigarette smoke or anything else that could irritate the throat
- Sleeps in a room with a humidifier
- Gargles with saltwater
- Sucks on a lozenge
- Takes acetaminophen for pain

Surgical management: It includes Tonsillectomy. It is recommended for recurrent streptococcal infections and massive hypertrophy. Tonsillectomies are reserved for children >3 years of age due to excessive blood loss and a potential for the tonsils to grow back.

Nursing Interventions

Preoperative management

- Soft or liquid diet
- Cool mist vaporizer
- Throat lozenges, warm salt gargles
- Antibiotics, analgesics, antipyretics
- Obtain baseline CT, BT

Postoperative management

- Position on side or abdomen
- Avoid suctioning
- Administer acetaminophen for pain
- Provide cool clear liquids, popsicles, ice chips
- Avoid straw, highly seasoned, irritating foods, red colored or citrus fluids
- Ice collar for post operative sore throat
- Teach to avoid coughing
- Monitor for signs of bleeding, i.e.,
  - Frequent swallowing
  - Increased pulse, respiration
  - Decreased blood pressure
  - Excessive thirst, pallor
  - Vomiting of blood

LOWER RESPIRATORY TRACT INFECTIONS (LRTI)

The **LRTI** includes croup, bronchiolitis and pneumonia

Croup

In croup, a peculiar brassy cough is the main presenting feature. Croup includes acute epiglottis, laryngitis, laryngotracheobronchitis.

- **Epiglottitis:** It is caused by hemophilus influenza type B. The main clinical features are high fever, dysphagia, marked suprasternal and subcostal chest retractions, child sits leaning forward with neck extended. Diagnosis can be done by direct laryngoscopy to see red swollen epiglottis.
- **Laryngitis and laryngotracheobronchitis:** The main cause is para influenza, RSV, adenovirus and rhinovirus. Clinical features include mild cold followed by brassy cough, inspiratory stridor, chest retractions, anxiety and eventually cyanosis.

Management of Croup

- Maintain a patent airway.
- Assess respiratory status and monitor pulse oximetry; monitor for nasal flaring, sternal retraction, and inspiratory stridor.
- Monitor for adequate respiratory exchange; monitor for pallor or cyanosis.
- Elevate the head of the bed and provide rest.
- Provide humidified oxygen via a cool air or mist tent as prescribed for a hospitalized child. Nebulization with epinephrine (1:8)
- Instruct the parents to use a cool air vaporizer at home; other measures include having the child breathe in the cool night air or the air from an open freezer.
- Provide and encourage fluid intake; IV fluids
- Administer analgesics, corticosteroids (dexamethasone), antibiotics (ampicillin or chloramphenicol, ceftriaxone)
- Teach the parents to avoid administering cough syrups or cold medicines, which may dry and thicken secretions.
- Heliox (mixture of helium and oxygen) may be prescribed.
- Have resuscitation equipment available. Endotracheal intubation or tracheostomy if condition worsens

Bronchiolitis

Definition: It is defined as a first episode of expiratory wheeze of acute onset in a child less than 2 years of age who has signs of viral respiratory illness, like coryza, otitis media or fever with or without indications of respiratory distress, pneumonia and atopy. It commonly affects infants between ages of 1 and 6 months and occurs in winter and spring.

Causes: RSV, influenza virus, Parainfluenza, adenovirus, M. pneumoniae

Pathophysiology

Mode of transmission is direct contact with respiratory secretions. Virus invades epithelial cells of nasopharynx and spreads to lower respiratory tract. It causes increased mucus production. Diameter of bronchi is decreased, which causes increased airway resistance. There is hyperinflation, emphesamatous changes and atelectasis. Diminished ventilation and diffusion leads to respiratory acidosis.

Complications

Respiratory insufficiency, obstructed airway

Assessment

- Nasal drainage, decreased appetite, low grade fever
- History of URTI, Respiratory distress
- Sternal retractions, use of accessory muscles
- Dyspnea, cyanosis
- Tachypnea, tachycardia
- Paroxysmal cough, coryza, thick mucus
- Wheezing, crackles
- Prolonged expiration
- Fine rales and rhonchi on auscultation
- Increased AP diameter of chest

Diagnosis

- History and physical examination
- Bronchial mucus culture for RSV ELISA - a rapid test to detect antigen
- Chest X-ray shows hyperinflation, ABG analysis
- Elevated leukocyte count

Medical Management

- Aerosols: ribavirin (antiviral agent) by nebulization, 16 hours a day for 3–5 days
- Palivizumab- anti RSV monoclonal antibody
- Bronchodilators, e.g., salbutamol
- Nebulized epinephrine 0.5 mL/kg of 1:1000 solution diluted in 3 mL NS
- Corticosteroids
- RSVIG
- Alfa 2 interferons

Nursing Interventions

- Contact and respiratory isolation
- Strict hand washing
- Monitor vitals, pulse oximetry
- Keep child in cool mist tent
- Provide humidified oxygen
- Upright position
- Rest, IV Fluids

Pneumonia

Definition: Pneumonia is an inflammation and consolidation of the lung parenchyma caused by an infection.

Classification: It can be classified on the basis of location and type of organism (Table 12.1).

Table 12.1: Types of pneumonia

Location	Type of organism
• Lobar • Bronchopneumonia • Interstitial pneumonia	• Bacterial: caused by Klebsiella, E.coli, pneumococci, staphylococci, H. influenza and streptococci • Viral, e.g., RSV, influenza, adenovirus, herpes virus, CMV • Fungal, e.g., blastomycosis • Atypical organisms, e.g., Chlamydia and mycoplasma

Lobar pneumonia: A pathological state of lung where the alveolar air has been replaced by cellular exudate and transudate. All or a large segment of one or more pulmonary lobes is involved, the affected lungs are completely consolidated.

Bronchopneumonia: Begins in the terminal bronchioles and related alveoli which become clogged with mucopurulent exudates to form consolidated patches in nearby lobules also called lobular pneumonia.

Interstitial pneumonia: It is characterized by massive proliferation and desquamation of alveolar cells and thickening of alveolar walls.

Recurrent/persistent pneumonia: Two episodes of pneumonia in one year or more than 3 episodes at any time with radiographic clearance between 2 episodes of illness.

Pneumonitis: Localized inflammation of lung parenchyma due to non-infectious causes.

Aspiration pneumonia: It is caused by aspirating oral or gastric contents, either while eating, or after reflux or vomiting. The resulting lung inflammation is not an infection but can contribute to one, since the material aspirated may contain anaerobic bacteria or other unusual causes of pneumonia.

Risk Factors

- Low birth weight
- Vitamin A deficiency
- Lack of breast feeding
- Passive smoking
- Poor socioeconomic status
- Large family size
- Family history of bronchitis
- Immunocompromised client
- Underlying lung disease
- Altered consciousness
- Endotracheal intubation
- Malnutrition
- Immobilization
- Overcrowding

Causes

- Viral – RSV, influenza, adenovirus, herpes virus, CMV
- Bacterial – Klebsiella, E.coli, pneumococci, staphylococci, H. influenzae and streptococci
- Atypical organisms – Chlamydia and mycoplasma
- Pneumocystitis carini
- Fungi – blastomycosis
- Aspiration of food, oily nose drops and liquid paraffin
- Kerosene poisoning
- Hypersensitivity pneumonitis

Pathophysiology

Four stages of lobar pneumonia are:

1. **Stage of congestion:** (First 24h), the lung is grossly doughy in consistency, and, microscopically, it is characterized by vascular congestion and alveolar edema. Many bacteria and a few neutrophils are present
2. **Stage of red hepatization:** (2–3 d), so called because of its similarity to the consistency of liver, is characterized by the presence of many erythrocytes, neutrophils, desquamated epithelial cells, and fibrin within the alveoli
3. **Stage of gray hepatization:** (2–3 d), the lung is gray-brown to yellow because of fibrinopurulent exudate, disintegration of red cells, and hemosiderin.
4. Stage of resolution is characterized by resorption and restoration of the pulmonary architecture. Fibrinous inflammation may extend to and across the pleural space, causing a rub heard by auscultation and it may lead to resolution or to organization and pleural adhesions.

Clinical Manifestations

- Cough, tachypnea
- Fever associated with chills, malaise
- Sharp or stabbing chest pain worsened by deep breathing or coughing
- Rapid, shallow breathing
- Shortness of breath
- Retractions, chest indrawing
- Nasal flaring
- Pallor to cyanosis
- Excessive sweating and clammy skin
- Loss of appetite
- Fatigue, myalgia, rhinitis, sore throat, wheezing, stridor

Diagnosis

- **Percussion:** To identify an area of consolidation
- **Chest X-ray:** Lobar consolidation
- Gram's stain and culture of the sputum
- Nasopharyngeal culture, blood cultures
- Bronchoalveolar lavage
- CBC to check white blood cell count; if high, this suggests bacterial infection
- Arterial blood gases, Thoracic CT
- Pulmonary ventilation and perfusion scan
- Pleural fluid culture if there is fluid in the space surrounding the lungs

Table 12.2: Clinical classification to facilitate treatment of Pneumonia

Signs and symptoms	Classification	Therapy	Where to treat
Cough or cold No fast breathing and chest indrawing	No pneumonia	Home remedies. Monitor for chest indrawing, increased RR	Home
Respiratory Rate 50 or more – 2–12 months 40 or more – 12 months to 5 years	Pneumonia	Oral Co-trimoxazole Amoxicillin/ ampicillin 25–50 mg/kg/day	Home
Chest indrawing, no cyanosis, able to drink	Severe pneumonia	IV/IM penicillin 25000 units/kg/ dose, 6 hrly, OD	Hospital
Cyanosis, severe chest indrawing and inability to feed	Very severe pneumonia	Oxygen, IV chloramphenicol 25 mg/kg/day	Hospital

Medical Management

Clinical classification to facilitate treatment of Pneumonia is shown in Table 12.2.

Medical management includes administration of following group of drugs:

- Antibiotics
- Antipyretics
- Antitussives
- Mucolytics
- Expectorants
- Bronchodilators

Complications

- Persistent effusions and empyemas are the most common serious complications of bacterial pneumonia
- Pulmonary abscess
- Respiratory distress
- Sepsis

Nursing Interventions

- Promote rest and comfort
- Prevent spread of infection
- Administer antibiotics, antipyretics, bronchodilators
- Institute oxygen therapy
- Promote hydration and good pulmonary hygiene

TUBERCULOSIS

Introduction

TB is a chronic infection caused by the bacteria Mycobacterium tuberculosis. It usually involves the lungs, but other organs of the body can also be involved. It is the most common cause of infection-related death worldwide.

Risk Factors

Overcrowding, poor housing, and inadequate ventilation predispose individuals to the development of TB. Patients receiving steroid therapy, cancer chemotherapy, and with hematologic malignancies, poor immunity, malnutrition and HIV infection are at increased risk for developing TB.

Mode of Transmission

It is transmitted from person to person by airborne droplets. Usually this infection is passed on as a result of very close contact, so family members of an infected person are endangered if the person continues to live in the same household and has not undergone proper treatment.

Pathophysiology

TB occurs when individuals inhale bacteria aerosolized by infected persons. The organism is slow growing and tolerates the intracellular environment, where it may remain metabolically inert for years before reactivation and disease. The main determinant of the pathogenicity of TB is its ability to escape host defense mechanisms, including macrophages and delayed hypersensitivity responses. Upon inhalation, the bacilli are deposited (usually in the midlung zone) into the distal respiratory bronchiole or alveoli. The alveolar macrophages phagocytose the inhaled bacilli. However, these macrophages are unable to kill the mycobacteria, and the bacilli continue to multiply unhindered.

Clinical Features

Fever, night sweats, anorexia, nonproductive cough, failure to thrive, and difficulty gaining weight may occur.

Lymphadenopathy in extrapulmonary TB

Revised National TB Control Program

National Tuberculosis Program (NTP) is in operation since 1962. In 1992, a joint Government of India/World Health Organization review found that despite the existence of the NTP, TB patients were not being accurately diagnosed and that the majority of diagnosed patients did not complete treatment. Based on the recommendations of the review, the Revised National Tuberculosis Control Program (RNTCP), incorporating the internationally recommended DOTS strategy, was developed. In 1993, RNTCP was started in pilot areas covering a population of 18 million and large-scale implementation of the RNTCP began in 1998.

Diagnosis

Diagnosis is based on a combination of clinical presentation (fever and/or cough for more than 3 weeks, with or without weight loss or no weight gain):

- Sputum examination
- Chest X-ray (PA view)
- The tuberculin skin test (TST) which is a widely used diagnostic test–The recommended TST is the Mantoux test. The dosage of 0.1 mL or 5 tuberculin units [TU] of purified protein derivative (PPD) is injected intradermally into the volar aspect of the forearm using a 27-gauge needle. It is positive if induration >10 mm after 48–72 hours)
- History of contact with a suspected or diagnosed case of active TB disease within the last 2 years.

Treatment of Pediatric TB

DOTS is the recommended strategy for treatment of TB and all Pediatric TB patients should be registered under RNTCP. Treatment categories and regimens for childhood Tuberculosis are shown in Table 12.3.

Pulmonary TB refers to disease involving lung parenchyma. Extra Pulmonary TB refers to disease involving sites other than lung parenchyma. If both pulmonary and extra pulmonary sites are affected, it will be considered Pulmonary for registration purposes. Extra Pulmonary TB involving several sites should be defined by most severe site.

Smear positive: Any sample (sputum, induced sputum, gastric lavage, broncho-alveolar lavage) positive for acid fast bacilli.

New case: A patient who has had no previous ATT or for less than 4 weeks.

Table 12.3: Treatment categories and regimens for childhood tuberculosis

Category of treatment	Type of patients	TB treatment regimens	
		Intensive phase	Continuation phase
New cases	• New smear-positive pulmonary Tuberculosis (PTB) • New smear-negative PTB • New extra-pulmonary TB	$2H_3R_3Z_3E_3$*	$4H_3R_3$
Previously treated cases	• Relapse, failure to respond or treatment after default • Re-treatment Others	$2S_3H_3R_3Z_3E_3$ + $1H_3R_3Z_3E_3$	$5H_3R_3E_3$

H = Isoniazid, R = Rifampin, Z = Pyrazinamide, E = Ethambutol, S = Streptomycin

**The number before the letters refers to the number of months of treatment. The subscript after the letters refers to the number of doses per week.*

Relapse: Patient declared cured/completed therapy in past and has evidence of recurrence.

Treatment after default: A patient who has taken treatment for at least 4 weeks and comes after interruption of treatment for 2 months and has active disease.

Failure to respond: A case of pediatric TB who fails to have bacteriological conversion to negative status or fails to respond clinically/or deteriorates after 12 weeks of compliant intensive phase shall be deemed to have failed response provided alternative diagnoses/reasons for non-response have been ruled out.

Others: Cases who are smear negative or extra pulmonary but considered to have relapse, failure to respond or treatment after default or any other case which do not fit the above definitions.

In patients with TB meningitis on Category I treatment, the four drugs used during the intensive phase can either be HRZE or HRZS. The present evidence suggests that Ethambutol can be used in children.

Children who show poor or no response at 8 weeks of intensive phase may be given benefit of extension of IP for one more month. In patients with TB Meningitis, spinal TB, miliary/disseminated TB and osteo-articular TB, the continuation phase shall be extended by 3 months making the total duration of treatment to a total of 9 months. A further extension may be done for 3 more months in continuation phase (making the total duration of treatment to 12 months) on a case to case basis in case of delayed response and as per the discretion of the treating physician.

Under Revised National Tuberculosis Program (RNTCP), all patients shall be covered under directly observed intermittent (thrice weekly) therapy. The supervised therapy is considered the most optimal treatment and is followed under RNTCP. It is important to ensure completion of treatment in every case put on treatment to prevent emergence of resistance, particularly to Rifampicin. In the rare circumstances where a patient is given daily therapy, observation and completion of therapy remain as important. It is the duty of the prescriber to ensure appropriate and complete treatment in all cases.

Prevention

The prevention of TB depends on prompt identification and treatment of patients with TB. Other strategies include patient education, treatment of latent infection, and vaccination with BCG injection. Thoroughly educate patients regarding compliance to therapy, adverse effects of medications, maintaining good living standards, and follow-up care. Educate to avoid contact with those who have the active disease. Using medications as a preventive measure in high-risk cases. BCG vaccination is available for the prevention of disseminated TB.

BRONCHIAL ASTHMA

Definition

Bronchial asthma is a disease characterized by an increased responsiveness of the airways to various stimuli. Airway inflammation may cause bronchospasm, which causes symptoms including wheezing, dyspnea, chest tightness, and cough, particularly at night or after exercise.

Types

Extrinsic (atopic): IgE mediated, caused by sensitivity to specific external allergens, e.g., smoke, pollen, dust.

Intrinsic (nonatopic): Non-IgE mediated, caused by reaction to internal, non-allergic factors, e.g., stress, exercise, fatigue, endocrine changes, anxiety, humidity variation, temperature variation, URTI.

Nocturnal asthma: Circadian variation in lung function and inflammatory mediator release in the circulation and airways can cause nocturnal asthma.

Risk Factors

- Family H/O asthma and atopic diseases
- Bronchiolitis during infancy
- Sensitization to allergens during childhood
- Passive smoking, aspirin
- Elevated IgE, Eosinophilia
- Concurrent allergic rhinitis, onset before 1 year
- House dampness, furred animal in home
- LBW

Pathophysiology

Airway obstruction in asthma is caused by 1. Edema and inflammation of mucous membrane lining the airways 2. Excessive secretion of mucous inflammatory cells and cellular debris and 3. Spasm of smooth muscle of bronchi.

Inhalation of allergen leads to early and late reaction ultimately causing bronchoconstriction.

Early reaction starts within 10 mins. There is release of histamine, leukotrienes, prostaglandins and bradykinins from mast cells. This leads to bronchoconstriction, mucosal edema (Fig. 12.2). This phase can be inhibited by beta 2 agonists (albuterol, terbutaline).

Late phase: Develops 3–4 hours later with peak at 8–12 hours. This phase presents as clinical asthma. There is release of mast cell mediators leading to inflammatory reaction, mucosal edema, Increased airway resistance during exhalation, air trapping, hyperinflated lungs, increased work of breathing, ventilation perfusion mismatch. Inadequately perfused lungs leads to low PaO_2, hypoxemia. Rise in $PaCO_2$ leads to respiratory acidosis.

Figure 12.2: Normal and asthmatic bronchiole

Triggers of an attack of asthma include:

- Allergy- mold, dust, pollen
- Viral or bacterial infections
- Exercise,
- Pollutants, Weather changes
- Emotional factors
- Food, Endocrine factors

Clinical Manifestations

- Expiratory Wheezing
- Cough with or without sputum (phlegm) production
- Intercostal retractions - Pulling in of the skin between the ribs when breathing
- Shortness of breath that gets worse with exercise or activity
- Cyanosis
- Tachypnea and tachycardia
- Use of accessory muscles
- Chest tightness, Sweating

Infants and young children suffering a severe episode display the following characteristics:

- Breathless during rest
- Not interested in feeding
- Sit upright
- Talk in words (not sentences)
- Usually agitated

Diagnosis

- **History and physical examination:** Wheezing, increased RR, HR
- **ABG analysis:** Decreased PaO_2 and increased $PaCO_2$
- Increased serum immunoglobulin E (allergic reaction)
- Absolute eosinophil counts- increased
- **Chest X-ray:** Hyperinflated lungs with air trapping during an attack
- **PFTs:** Decreased forced expiratory volumes (FEV) and increased residual volume and total lung capacity.
- **PEFR, peak expiratory flow rate:** Measured with peak expiratory flowmeter
- **Allergy test:** Skin test and RAST (radio-allergo-sorbent test) may identify allergens

Management

Management of asthma includes:

- Environmental control or eliminating the triggers
- Medical management
  - Management of chronic asthma
  - Management of acute asthma
- Parent education
- Treatment of comorbid conditions
- Environmental control/ eliminating triggers
  - Control of indoor and outdoor air pollution
  - Avoidance of tobacco smoke
  - Avoid overcrowding in house and classrooms
  - Sun dry the bedding weekly
  - Avoid carpet, wood burning smoke, strong odors
  - Clean filter of AC at regular interval
  - Stay indoor esp midday and afternoon when pollen and mold counts are high
  - Wet mopping of floors to be done
  - Avoid suspected foods

Medical Management

Pharmacologic asthma management includes the use of quick relief medications and long-term control agents (Table 12.4).

The severity of asthma can be classified based on symptoms, night time symptoms and PEFR findings (Table 12.5). According to the severity the treatment of asthma is shown in Table 12.6.

Table 12.4: Pharmacological management of asthma

Quick-Relief Medications Used to treat symptoms and exacerbations (Rescue Medications)	Long-Term Control Used to achieve and maintain control of inflammation (Medications to Prevent Attacks)
• Short-acting ß2 agonists/ SABA- (for bronchodilation) - terbutaline, albuterol • Anticholinergics (for relief of acute bronchospasm) – ipratropium (inhaled) • Systemic corticosteroids (for anti-inflammatory action to treat reversible airflow obstruction)- beclomethasone, budesonide, fluticasone, triamcinolone	• Inhaled corticosteroids (for anti-inflammatory action) • Antiallergy medications (to prevent an adverse response on exposure to an allergen) • Anti-allergy NSAID – cromolyn- stops mast cell from releasing histamine • Long-acting ß2 agonists/LABA (for long-acting bronchodilation)- salmeterol, used with corticosteroids • Leukotriene modifiers (to prevent bronchospasm and inflammatory cell infiltration)- oral-Montelukast sodium, Zafirlukast or Zileuton • Monoclonal antibody (blocks binding of IgE to mast cells to inhibit inflammation)- omalizumab

Table 12.5: Classification of asthma according to severity

Classification	Symptoms/day	Symptoms/night	PEF or FEV_1 PEF variability
STEP 1 intermittent	<1 time a week asymptomatic and normal PEF between attacks	≤2 times a month	>80% <20%
STEP 2 mild persistent	>1 time a week but <1 time a day attacks may affect activity	>2 times a month	>80% 20–30%
STEP 3 moderate persistent	Daily attacks affect activity	>1 times a week	60%–80% >30%
STEP 4 severe persistent	Continuous limited physical activity	Frequent	<60% >30%

Table 12.6: Stepwise treatment of asthma

	Long-term prevention
Step 4 severe persistent	Inhaled short acting β-agonist as required + Inhaled corticosteroids: Budesonide/Beclomethasone, 400 µg twice daily may increase up to 2000 µg/day in selected cases + Long acting bronchodilator: Long acting inhaled β_2 agonist and/or sustained release theophylline + Oral corticosteroids low dose alternate day (if no relief with above treatment)
Step 3 moderate persistent	Inhaled short acting β-agonist as required + Inhaled corticosteroids: Budesonide/Beclomethasone, 400–800 µg divided twice daily If needed long acting bronchodilator: Long acting inhaled β_2 agonist salmeterol 50 µg once/twice daily and/or sustained release theophylline

Contd...

Long-term prevention	
Step 2 mild persistent	Inhaled short acting β-agonist as required + Inhaled corticosteroids Budesonide/Beclomethasone, 200-400 µg or cromolyn or sustained release theophylline or leucotriene modifiers
Step 1 intermittent	Inhaled short acting β-agonist as required for symptoms relief. If they are needed more than 3 times a week move to step 2

*If Fluticasone is used the dose is half that of Budesonide/Beclomethasone

Delivery devices and best route of administration: In pediatric asthma, inhaled treatment is the cornerstone of asthma management. Inhaler devices are currently used to deliver inhaled corticosteroids (ICSs) fall into the following 4 categories:

1. **Pressurized metered dose inhaler (pMDI):** Propellant is used to dispense steroid when canister is pressed manually
2. **Dry powder inhaler (DPI):** Does not require hand-breath coordination to operate
3. **Breath-actuated pMDI:** Propellant is used to dispense steroid when patient inhales
4. Nebulized solution devices

The inhaler device must be chosen on the basis of age, cost, safety, convenience, and efficacy of drug delivery.

Nursing Interventions

Parent Education

- Identify triggers to asthma attacks
- Demonstrate use of a metered-dose inhaler and peak flow meter
- Emphasize drugs can control but not cure asthma
- Maintain diary of events—daytime cough, nocturnal cough, wheezing
- Recognize early S/S of respiratory infection and hypoxia

Complications

- Pneumothorax
- Pneumomediastinum
- Subcutaneous emphysema
- Atelectasis
- Secondary bacterial pneumonia
- Status asthamaticus

CYSTIC FIBROSIS

Definition: Cystic fibrosis is an autosomal recessive disorder which involves generalized dysfunction of the exocrine glands producing abnormal viscid mucus. It affects multiple organ systems, e.g., pulmonary, GI system, sweat glands, salivary glands, etc.

Pathophysiology

Cystic fibrosis is caused by defects in the cystic fibrosis gene, which codes for a protein transmembrane conductance regulator (CFTR) that functions as a chloride channel.

Failure of chloride transport across epithelial cells on mucosal surfaces leads to dehydration of secretions that results in mucus that is stickier to bacteria, which promotes infection and inflammation. Secretions in the respiratory tract, pancreas, GI tract, sweat glands, and other exocrine tissues have increased viscosity, which makes them difficult to clear (Fig. 12.3).

In the respiratory system, there is thick mucus in trachea, bronchioles which leads to airway obstruction, atelectasis, reduced area for gas exchange. Mucus serves as medium for bacterial growth leading to secondary respiratory infections.

Pancreatic ducts become clogged with mucus, there is no release of pancreatic enzymes (lipase, amylase, trypsin); decreased absorption of fats and proteins. Unabsorbed food fractions are excreted as steatorrhea. Inability to reabsorb leads to FTT.

There is biliary duct obstruction, portal HTN and esophageal varices.

Assessment

System wise clinical manifestations of cystic fibrosis are shown in Table 12.7 and organs affected in Figure 12.3.

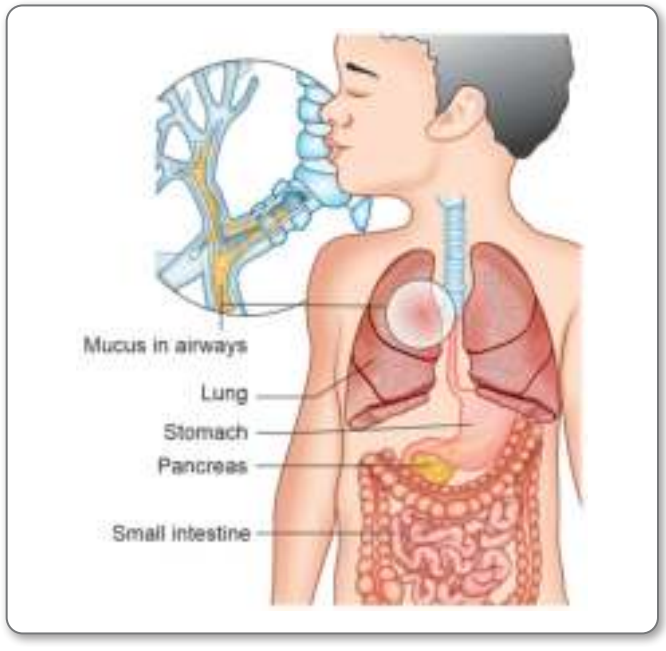

Figure 12.3: Organs affected by cystic fibrosis

Table 12.7: Clinical manifestations of cystic fibrosis

GI Tract	Respiratory tract	Genital tract
• Bulky greasy foul smelling stools • Thin arms and legs • Distended abdomen • FTT • Meconium ileus in new born • Salty taste on child's skin • pancreatitis • Voracious appetite • Rectal prolapse	• Dyspnea, tachypnea • Chronic productive cough • Repeated URTIs • Barrell chest, clubbing of digits	• Cervical mucus thick: Infertility • Obstruction of vas deferens: Sterility, azoospermia

Diagnosis

- **Sweat chloride test:** Using pilocarpine iontophoresis, Cl >60 mEq/L on 2 occasions
- **PFT:** Reduced VC, elevated residual volume
- **Liver enzymes test:** Hepatic insufficiency
- **Pancreatic enzymes:** Decreased or absent
- **Sputum culture:** Staphylococcus
- **Serum albumin:** Nutritional status
- Elevated fecal fat

Medical Management

- Aerosol therapy- recombinant human Dnase (rh Dnase (enzyme), 2.5 mg OD or BD. It helps thin airway mucus and reduces adhesiveness of CF sputum.
- Antibiotics- fluoroquinolones, cephalosporins, amino-glycoside
- Mucolytic agents- N-acetyl cystein (mucomyst)
- Bronchodilators
- Inhaled steroid therapy
- Oral pancreatic enzymes

Nursing Management

- Institute airway clearing techniques
  - Chest physiotherapy, postural drainage, breathing exercises
  - Oxygen
  - Avoid cough suppressants, antihistamines
  - Good oral hygiene
- For Meconium ileus - administer gastrografin enema, it is of high osmolality, draws water into gut, and helps in expulsion of meconium.
- Genetic counselling
- Nutritional management:
  - Encourage high calorie, high protein diet, fats as tolerated, and increased salt intake
  - Increased fluids must be provided. Oral pancreatic enzymes in form of capsules, which can be sprinkled over food (3000–10,000IU per meal) can be given.
  - Fat soluble vitamins- A, D, E, K may be administered.

COMMON RESPIRATORY SURGICAL CONDITIONS

CHOANAL ATRESIA

Congenital failure of nasal cavities to open posteriorly into the nasopharynx is Choanal atresia (Fig. 12.4).

Etiology

Failure of resorption of buccopharyngeal membrane during embryonic development.

Symptoms

A newborns generally prefers to breathe through nose. Typically, infants breathe through mouth only when they cry. Babies with choanal atresia have difficulty breathing unless they are crying. Choanal atresia may affect one or both sides of the nasal airway. Bilateral Choanal atresia blocking both sides of the nose causes acute breathing problems with cyanosis and breathing failure and may need resuscitation at delivery. More than half of infants have a blockage on only one side, which causes less severe problems. Child may exhibit:

- Chest retractions
- Dyspnea following birth, which may result in cyanosis, unless infant is crying
- Persistent one-sided nasal blockage or discharge

Diagnosis: There is inability to pass an 8 F catheter through the nasal cavity more than 5.5 cm from the alar rim. There is

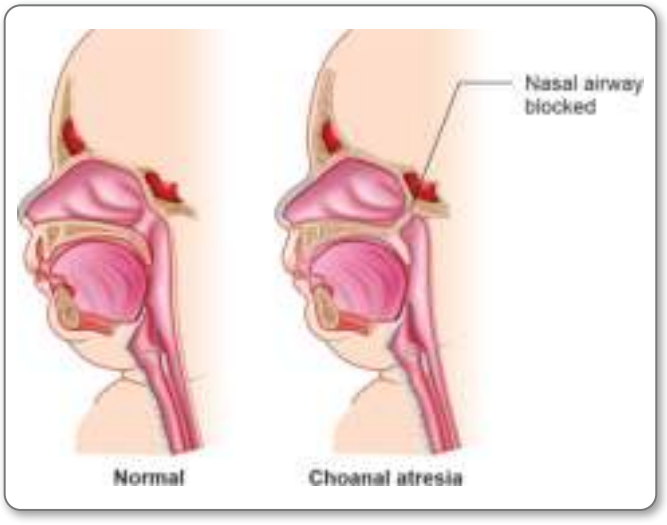

Figure 12.4: Choanal atresia

lack of movement of a thin wasp of cotton under the nostrils while the mouth is close. There is absence of fog on the mirror when it is placed under the nostrils.

A physical examination may show an obstruction of the nose. Tests that may be done include: CT scan, Endoscopy of the nose, Sinus X-ray.

Management

The immediate concern is to resuscitate the baby if necessary. An airway may need to be placed so that the infant can breathe. In some cases, intubation or tracheostomy may be needed.

An infant can learn to mouth breathe, which can delay the need for immediate surgery.

Surgery to remove the obstruction cures the problem. Surgery may be delayed if the infant can tolerate mouth breathing. The surgery may be done through the nose (transnasal) or through the mouth (transpalatal). Transnasal endoscopic repair is less invasive. Transpalatal repair involves removal of posterior hard palate. Stents are placed to prevent restenosis and left in place for 3–6 weeks postoperatively.

Prognosis: Full recovery is expected.

Complications: Possible complications include:

- Aspiration while feeding and attempting to breathe through the mouth
- Respiratory arrest
- Renarrowing of the area after surgery

CONGENITAL DIAPHRAGMATIC HERNIA (CDH)

Introduction

CDH is a congenital malformation of the diaphragm. The most common type of CDH is a Bochdalek hernia; other types include Morgagni's hernia, diaphragm eventration and central tendon defects of the diaphragm. This anomaly of the diaphragm allows the abdominal organs to push into the chest causing hinderance to proper lung formation (Fig. 12.5).

The major cause of death in infants is due to two complications, i.e., pulmonary hypoplasia and pulmonary hypertension. Newborns with CDH often have severe respiratory distress which can be life-threatening unless treated appropriately.

Types

- **Bochdalek hernia:** It is also known as a postero-lateral diaphragmatic hernia, the most common manifestation of CDH, accounting for more than 95% of cases. There is hole in the postero-lateral corner of the diaphragm which allows passage of the abdominal viscera into the chest cavity.
- **Morgagni's hernia:** This rare anterior defect of the diaphragm is variably referred to as Morgagni's, retrosternal, or parasternal hernia. It accounts for approximately 2% of all CDH cases, it is characterized by herniation through the foramina of Morgagni which are located immediately

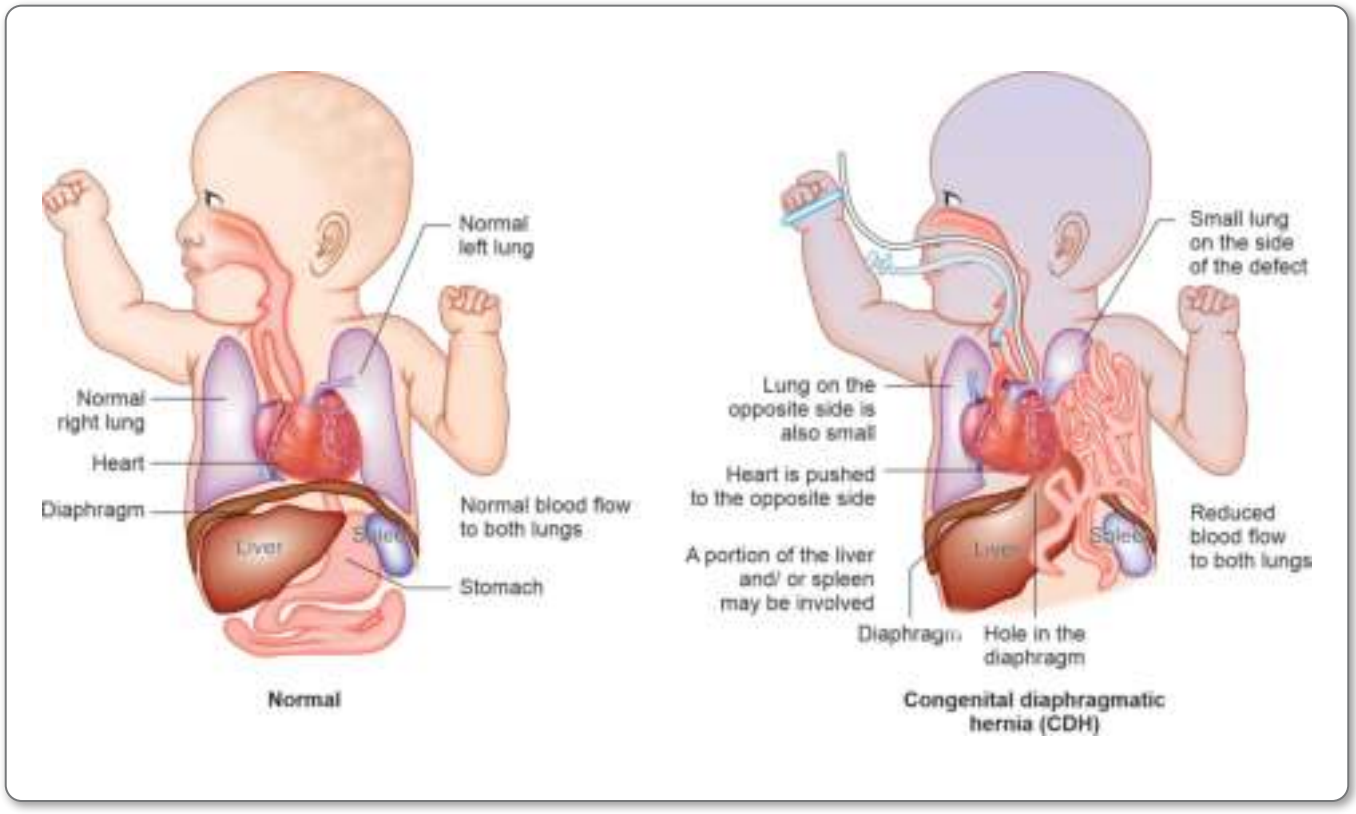

Figure 12.5: Congenital diaphragmatic hernia

adjacent to the xiphoid process of the sternum. The majority of hernias occur on the right side of the body and are generally asymptomatic. However newborns may present with respiratory distress at birth similar to Bochdalek hernia. Additionally, recurrent chest infections and gastrointestinal symptoms have been reported in those with previously undiagnosed Morgagni's hernia.

Pathophysiology

The diaphragm initially develops as a septum between the heart and liver, progresses posterolaterally, and closes at the left Bochdalek foramen at approximately 8–10 weeks' gestation. In CDH the herniation of viscera usually occurs during the pseudoglandular stage of lung development. Lung compression results in pulmonary hypoplasia that is most severe on the ipsilateral side, although both lungs may be abnormal. Pulmonary hypoplasia is associated with fewer bronchial generations, alveoli, and arterial generations.

Infants born with diaphragmatic hernia experience respiratory failure due to both pulmonary hypertension and pulmonary hypoplasia. The first condition is a restriction of blood flow through the lungs thought to be caused by defects in the lung. Pulmonary hypoplasia or decreased lung volume is directly related to the abdominal organs present in the chest cavity which causes the lungs to be severely undersized, especially on the side of the hernia.

Signs and Symptoms

Infants frequently exhibit a scaphoid abdomen, barrel-shaped chest, and signs of respiratory distress (retractions, cyanosis, grunting respirations). In left-sided posterolateral hernia, auscultation of the lungs reveals poor air entry on the left, with a shift of cardiac sounds over the right chest. In patients with severe defects, pneumothorax signs (poor air entry, poor perfusion) may also be found.

Diagnosis

- **Arterial blood gas:** Obtain frequent ABG measurements to assess for pH, $PaCO_2$, and PaO_2.
- **Chromosome studies:** Obtain chromosome studies because of the frequent association with chromosomal anomalies.
- **Serum electrolytes:** As with all critically ill neonates, monitor levels of serum electrolytes, ionized calcium, and glucose initially.
- **Chest radiography:** Typical findings in a left-sided posterolateral CDH include air- or fluid-filled loops of the bowel in the left hemithorax and shift of the cardiac silhouette to the right.
- Examine the chest radiograph for evidence of pneumothorax.
- **Cardiac ultrasonography:** To find associated cardiac anomalies.
- **Pulse oximetry:** Continuous pulse oximetry is valuable in the diagnosis and management of Persistent Pulmonary Hypertension (PPHN).
- **Renal ultrasonography:** To determine genitourinary anomalies
- **Cranial ultrasonography:** To identify CNS defects (neural tube defects, hydrocephalus) which may be associated with CDH.

Management

- Endotracheal intubation and mechanical ventilation is required in all infants with severe CDH who present in the first hours of life.
- If the diagnosis is known at the time of delivery, avoid bag-and-mask ventilation in the delivery room because the stomach and intestines become distended with air and further compromise pulmonary function.
- A nasogastric tube should be placed as soon as possible to provide intestinal decompression and connect it to continuous suction to prevent bowel distension and further lung compression.
- The goal is to expand the lung but to avoid overdistension; therefore, inspiratory pressures should be kept as low as possible.
- Consider the use of high-frequency ventilation (HFV) if high inspiratory pressures are required.
- Avoid high peak inspiratory pressures and stay alert to the possibility of early pneumothorax if the infant does not stabilize.
- Place arterial catheter in the umbilical artery or in a peripheral artery (radial, posterior tibial) for blood pressure and frequent ABG monitoring.
- Central venous catheter placement is done via the umbilical vein to allow for administration of inotropic agents and hypertonic solutions such as calcium gluconate.
- Extracorporeal membrane oxygenation
- Venoarterial or venovenous extracorporeal membrane oxygenation (ECMO) support: It is an adaptation of cardiopulmonary bypass and involves a surgical team; insertion of catheters into the internal carotid artery, internal jugular vein, or both; systemic heparinization; and oxygenation through the use of a membrane lung.

Fetal Surgery

Fetal intervention involves occluding the fetal trachea. The fetal lung secretes fluid by active ion transport throughout gestation, and this lung fluid provides a template for lung growth. Occlusion of the fetal trachea traps this fluid and stimulates lung growth, by retention of growth factors within the lung and stimulation of local growth factors by the gentle distension provided by the fluid.

Postnatal Surgical Care

- Reduction of the herniated viscera and closure of the diaphragmatic defect should be emergently performed following birth.
- However, a delayed surgical approach that enables preoperative stabilization decreases morbidity and mortality.
- The ideal time to repair a CDH is unknown.
- Some suggest that if repair happens 24 hours after stabilization, it is ideal, but delay of up to 7–10 days is typically well tolerated

Postoperative Care

- Assess Vitals
- Maintain NG decompression
- Administer IV fluids
- ABG assessment
- Care of chest tubes, chest tube drainage is necessary when a tension pneumothorax is present
- Balanced intrathoracic drainage, in which a closed gated pressure system is used to maintain intrathoracic pressure within the normal physiologic range, may minimize risk of pulmonary injury.

Medical therapy is directed toward stabilizing blood pressure and circulating volume, pulmonary distress, and hypoxemia. The common groups of drugs used are:

- **Vasoactive agents:** *Dopamine, dobutamine*
- **Opioid analgesics:** Fentanyl
- **Neuromuscular relaxing agents:** Pancuronium (Pavulon) Vecuronium (Norcuron)
- **Pulmonary vasodilating agents:** Nitric oxide is an important mediator of vascular tone that was recently approved as a therapeutic modality for infants with PPHN. It is delivered as an inhaled gas.

ESOPHAGEAL ATRESIA WITH TRACHEO ESOPHAGEAL FISTULA

Definition: Esophageal atresia (EA) is a congenital disorder in which esophagus ends in a blind pouch; there is no entry route to the stomach. Tracheoesophageal fistula (TEF) is an abnormal open connection between trachea and esophagus.

Etiology

- More predominant in males
- Polyhydramnios
- LBW or premature babies
- Congenital anomalies
- Family history of twinning
- Use of drugs, like thalidomide and hormones, like estrogen or progesterone

Pathophysiology

- Esophagus develops from first layer of embryonic gut.
- During 4th and 5th week of gestation foregut lengthens and separates into 2 parallel channels, i.e., trachea and esophagus that are joined only at larynx.
- In EA with TEF abnormal development of trachea and esophagus occurs due to altered cellular growth during the embryonic period. Fistulas may be present when the esophagus is patent, when it is narrowed or when it is not joined to its distal portion.
- Oral intake enters lungs or air enters stomach.

Types of EA with TEF

There are 5 types of TEF (Fig. 12.6)

Type A: It is a pure esophageal atresia which consists of a blind pouch at each end, widely separated and with no communication to the trachea.

Type B: It is an extremely rare anomaly in which the upper pouch of esophagus is connected to trachea with a fistula. Thus the oral secretions enter trachea and cause coughing. Feeds may enter trachea.

Type C: proximal esophagus ends in blind pouch leading to drooling of secretions, distal pouch communicates with trachea by a fistula. This leads to air entry from trachea to stomach and causes abdominal distension.

Type D is K type fistula and Type E is H type fistula. Oral intake enters lungs or air enters stomach.

Clinical Manifestations

- Regurgitation and vomiting commonly seen in type C
- Fluid returns through nose and mouth
- Abdominal distension- type C
- **C**oughing, **c**hoking, **c**yanosis (three Cs) at the time of feeding- type E
- Frothy saliva in mouth and nose, drooling
- Abdominal distension

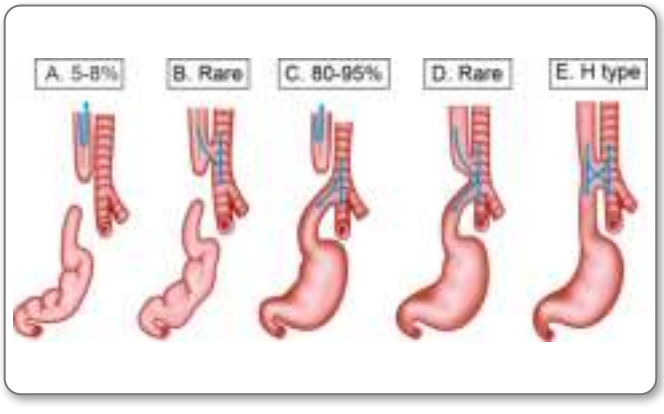

Figure 12.6: Common types of EA with TEF

Diagnosis

- Arrest of red rubber catheter of 6-8 Fr size at 10 cm from gum margin when passed orally- esophageal atresia
- 5–10 mL of air is injected through orogastric catheter. Chest and abdomen radiography is done to look for air. If air enters abdomen, it is TEF. If no air in abdomen then it is EA or EA with proximal TEF
- Bronchoscopy to visualize fistula
- Presence of polyhydramnios antenatally - amniotic fluid normally swallowed by fetus does not reach GI tract leading to polyhydramnios.
- Presenting symptoms – 3 Cs (Coughing, choking, cyanosis)

Associated Problems

- Aspiration of saliva and feeds leading to pneumonia
- Reflux of gastric secretions through tracheal communication leading to chemical pneumonitis
- VACTERL syndrome (vertebral, anorectal, cardiac, tracheoesophageal, renal and limb anomalies)

Management

Surgical management includes primary repair or staged intervention (Table 12.8).

Table 12.8: Surgical management of TEF

1. Immediate primary repair under tension	2. Delayed surgical intervention
Indications • No pulmonary complications • Weight of child above 2–2.5 kg • No major congenital anomalies present • Baby is otherwise healthy • Gap between proximal and distal esophagus is less than 2 cm	**Indications** • Associated pneumonia, sepsis, cardiac malformation, RDS • Severe prematurity • Gap is more than 2.5 cm
Steps of surgery • Right posterolateral thoracotomy through 4th intercostal space. Mobilization of proximal pouch after ligating vena azygous. Identification and ligation of fistula at the tracheal end. • End to end anastomosis of proximal and distal esophagus by single layer of 4/0 synthetic suture • Transanastomotic sterile feeding tube is left in stomach, passed through nose. • Closure of chest with drain connected to underwater seal bottle • Gastrostomy may be done	**Stages of surgery** • Left cervical esophagostomy to allow drainage of saliva and gastrostomy for feeding • Right thoracotomy and ligation of TEF • Replacement of gap between 2 pouches at 6-8 months of age by isolated segment of colon or by gastric tube when the weight of the child is above 10 kg.

Preoperative Care

- Maintain patent airway, 30° head end elevation
- Perform gentle oropharynx suctioning to minimize aspiration of saliva
- Administer oxygen in case of cyanosis
- Keep child NPO
- Administer IV fluids
- No pacifier to be given
- Prepare for gastrostomy tube insertion to decompress stomach and prevent aspiration of gastric contents from fistula. Administer gastrostomy tube feedings. Maintain the patency of the tube, do not clamp it.

Postoperative Care

- Maintain patent airway
  - Put the child in supine position with head elevated position for optimum ventilation.
  - Perform oropharyngeal suctioning as necessary.
  - Administer oxygen
  - Salbutamol nebulization 6 hourly
  - Maintain care of chest tubes.
- Prevent infection
  - Care of incision site – monitor for inflammation
  - Administer antibiotics
  - Monitor vitals
- Maintain fluid and electrolyte balance
  - Administer IV fluids, record intake output
  - Record weight daily
- Provide adequate nutrition
  - Stimulate non-nutritive sucking with pacifier/sham feedings to meet oral needs
  - Gastrostomy feedings after 48 hours, initially with water, if tolerated well, start milk
  - Monitor for swallowing ability
  - Provide progressive oral feeds after contrast study on 4th or 5th post-op day.
  - Gradually increase feeds and elevate gastrostomy tube, feed slowly, burp frequently.
  - Dilation of esophagus with soft red rubber catheter after 14 days

Complications of Surgery

Early

- Pneumothorax
- Anastomotic leak- manifested as saliva in chest tube
- Recurrence of fistula – anastomotic leak and inflammation will lead to rejoining of trachea and esophagus
- Pulmonary infections due to aspiration of gastric contents into trachea

Late

- Anastomotic stricture due to tension or ischemia, braided silk suture, GER and anastomotic leak. Management is dilation of esophagus.

- Esophageal motility disorders leading to dysphagia.
- Gatroesophageal reflux due to traction on lower esophageal sphincter and altered angle of Hiss. Practice anti GER measures. Surgical management is fundoplication.
- Tracheal stenosis.
- Tracheomalacia – weakness in the tracheal wall that occurs when a dilated proximal pouch compresses trachea early in fetal life, decreased cartilaginous rings and compression by aorta and innominate artery. Diameter of trachea is narrowed which may lead to reflex apnea and airway obstruction.

Discharge Teaching

Teach about signs of respiratory distress which may indicate constriction of esophagus like poor feeding, dysphagia, drooling and regurgitation

- Teach gastrostomy feedings and skin care
- Tell to avoid foods, which can cause choking, e.g., hot dogs, large pieces of meat; cut food into small pieces

Nursing Management of Child with Respiratory Disorders

Nursing Assessment

- History and physical examination specifically for coughing, wheezing, grunting, cyanosis, retractions.
- Assess respiratory status, psychosocial status.

Nursing Diagnosis

- Ineffective breathing pattern related to inflammatory process.
- Ineffective airway clearance related to mechanical obstruction, inflammation and increased secretions.
- Ineffective thermal regulation R/T disease process
- Fear and anxiety related to difficulty breathing, unfamiliar procedures and hospital environment
- High risk for infection related to presence of infective organisms
- Activity intolerance related to inflammatory process, imbalance between oxygen supply and demand
- Altered family process related to hospitalization of child

Nursing Management

Goal: Maintain Normal Respiratory Functions

- Position for maximum ventilation
- Avoid constricting clothing or bedding
- Provide supplemental oxygen
- Assess oxygen saturation
- Promote rest and sleep
- Encourage relaxation techniques

Goal: Maintain Patent Airway

- Position child in proper body alignment
- Suction secretions as needed
- Assist child in expectorating sputum
- Keep nares patent, instill normal saline drops
- Perform chest physiotherapy
- Administer expectorants, if prescribed

Goal: Prevent Infection

- Maintain sterile technique
- Isolate child as indicated
- Provide nutritious diet
- Teach child and family manifestations of illness
- Administer antibiotics as prescribed

Goal: Maintain Adequate Energy Level

- Continue breast feeding, if possible
- Assess child's physical level of tolerance
- Assist child in activities of daily living
- Provide diversional play activities
- Provide sleep and rest periods, provide comfortable position
- Balance rest and activity when ambulatory

Goal: Reduce Anxiety and Increase Ability to Cope

- Recognize parental concern and need for information and support
- Explore family's feelings and problems
- Provide support as needed
- Encourage family-centered care and encourage family to become involved in their child's care

Summary

The common respiratory diseases afflicting pediatric population are URTI, pulmonary tuberculosis, asthma and pneumonia. These diseases are associated with significant morbidity and mortality. Morbidity involves loss of school and work. It also implies significant financial costs for the patients in the form of hours of work lost, expenditure incurred for the diagnostic tests, and treatment prescribed. Thus, health personnel need to proactively manage these conditions for healthy India which may include lifestyle and environmental changes.

Assess Yourself

1. Name the causative organism for tonsillitis and bronchiolitis.
2. Name antitubercular drugs used in the management of pediatric TB. Mention their side effects.
3. Discuss the diagnostic tests and symptoms of a child with cystic fibrosis.
4. Explain the causes and medical management for a child with bronchial asthma.
5. Define pneumonia. Discuss pathophysiology and nursing management.
6. Draw diagram of 5 types of tracheoesophageal fistula.

Chapter 13

Endocrine Disorders

Chapter Outline

INTRODUCTION

The work of the pituitary gland is directed by the **hypothalamus**. Pituitary gland is divided into the anterior lobe (adenohypophysis), the posterior lobe (neurohypophysis), and the intermediate lobe. Altogether, these divisions store and release nine different hormones (Fig. 13.1). Four of these: the antidiuretic hormone (ADH), thyrotropin, corticotropin, and somatotropin are prominently involved in childhood illnesses (Table 13.1).

FUNCTIONS OF ENDOCRINE SYSTEM

- Regulation of normal growth
- Maintenance of body metabolism
- Stress management
- Fluid and electrolyte balance
- Bone mineral homeostasis
- Sex differentiation
- Puberty
- Glucose metabolism

CAUSES OF ENDOCRINE DISORDERS

Causes of endocrine disorders are typically grouped into two categories:

1. Endocrine insufficiency results when a gland produces too much or too little of an endocrine hormone, it is called hormone imbalance.
2. Endocrine disease occurs due to the development of lesions (such as nodules or tumors) in the endocrine system, which may or may not affect hormone levels.

ANTIDIURETIC HORMONE (ADH) DISORDERS

Diabetes Insipidus

Diabetes insipidus (DI) is a disease in which there is decreased release of ADH (vasopressin) by the pituitary gland. This causes a reduced amount of reabsorption of fluid in the kidney tubules (Fig. 13.2). Urine becomes extremely dilute, and majority of fluid is lost from the body. Diabetes insipidus may be an X-linked dominant trait, or it may be transmitted by an autosomal recessive gene. It may also result from a lesion, tumor, or injury to the posterior pituitary, or it may have an unknown cause.

Assessment

The child with diabetes insipidus experiences **polydipsia** that is relieved only by drinking large amounts of water; there is accompanying **polyuria**. The specific gravity of the urine will

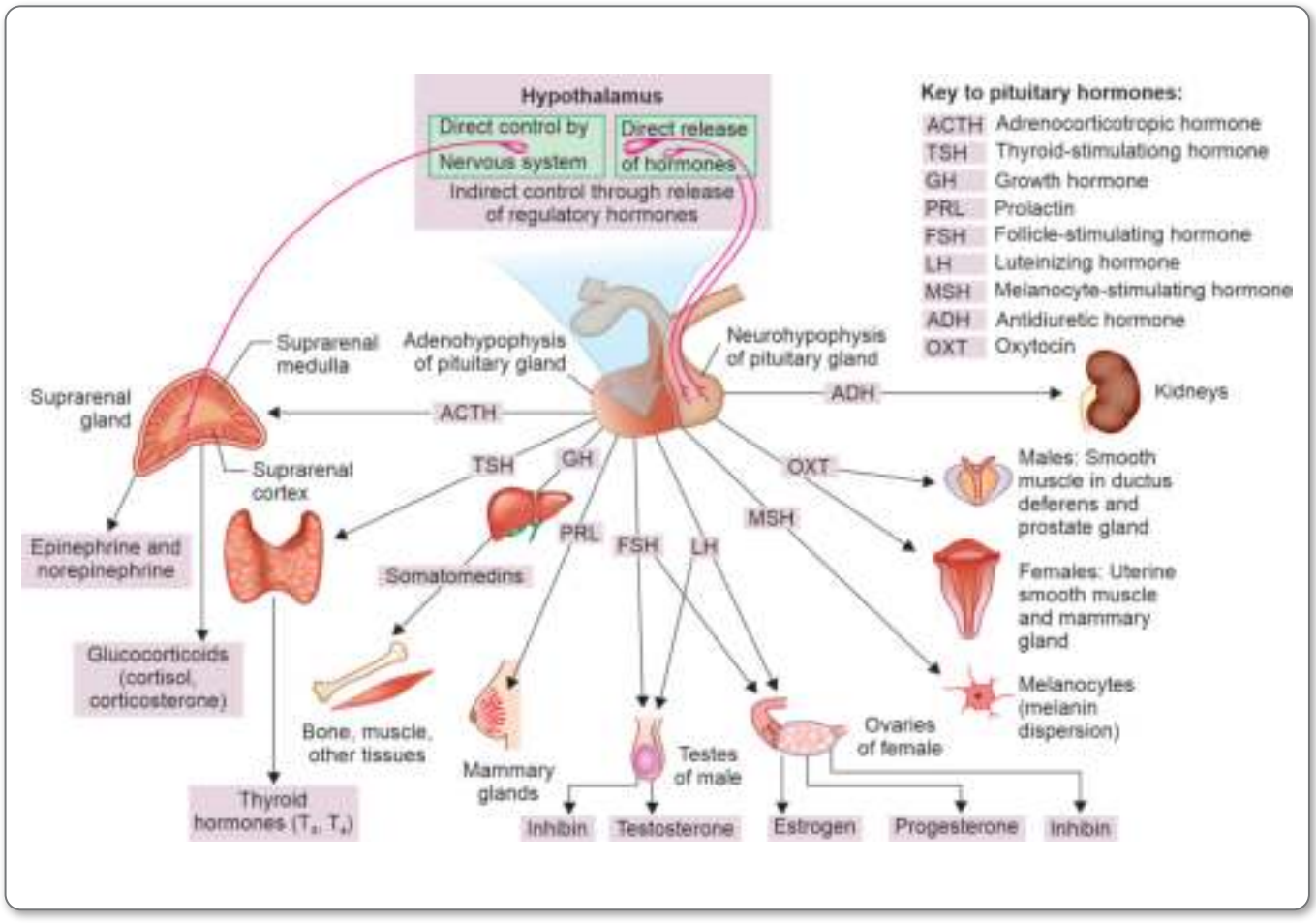

Figure 13.1: Glands and hormones

Table 13.1: Common pituitary hormones

Gland	Hormones	Target Organs	Hormones	Disorder
Posterior Pituitary (neurohypophysis)	ADH (vasopressin)	Kidneys		Diabetes Insipidus, SIADH
Pituitary (posterior)	Oxytocin	Uterus		
Anterior Pituitary (adenohypophysis)	Growth hormone (somatotropin)	Acts on all body cells (Liver, bone cells, muscle)		Gigantism, Acromegaly, Dwarfism
	ACTH (Corticotropin)	Adrenal gland (cortex)	Glucocorticoids, mineralocorticoids, androgens	Adrenal insufficiency, Cushing's disorder
		Adrenal Medulla	Epinephrine, Norepinephrine	Pheochromocytoma
	TSH	Thyroid Gland	T3, T4	Hypothyroidism, Hyperthyroidism
	FSH and LH	Ovary and Testes	Progesterone, Estrogen, Testosterone	
	Prolactin	Mammary glands		
Pancreas			Insulin Glucagon	IDDM

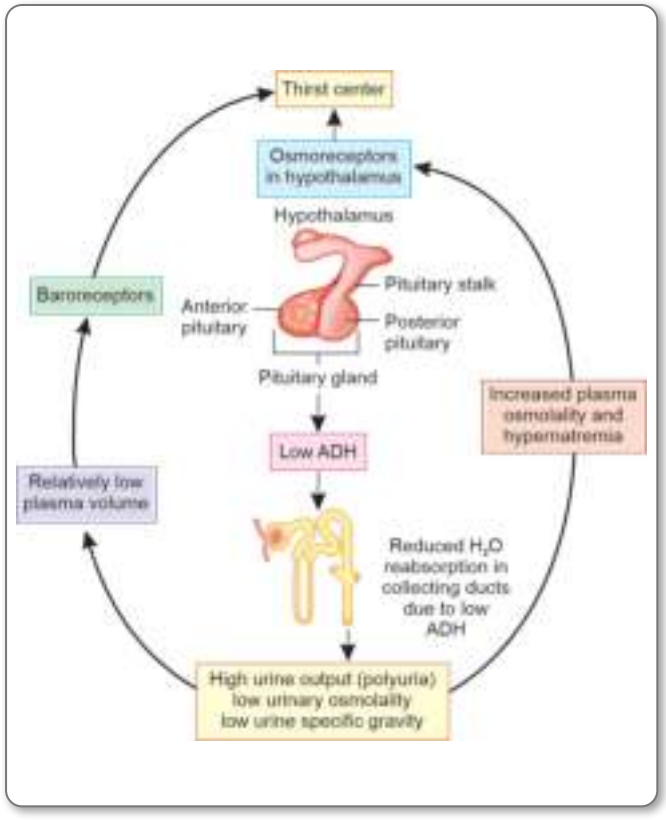

Figure 13.2: Pathophysiology of diabetes insipidus

be as low as 1.001 to 1.005 (normal values is between 1.010 to 1.030). Urine output may reach 4 to 10 L in a 24-hour period (normal range, 1 to 2 L), depending on age.

Because so much fluid is lost, sodium becomes concentrated or hypernatremia occurs with symptoms of irritability, weakness, lethargy, fever, headache, and seizures. The signs and symptoms usually appear gradually. Parents may notice the polyuria first as bed-wetting in a toilet-trained child or weight loss because of the large loss of fluid. If the condition remains untreated, the child is in danger of losing large quantity of water that can lead to dehydration and death.

Diagnosis

MRI, CT scanning, or an ultrasound study of the skull reveal whether a lesion or tumor is present. A further test is the administration of vasopressin (Pitressin) to rule out kidney disease. For this:

- Establish baseline urine output
- Vasopressin is administered. The drug decreases the blood pressure, alerting the kidney to retain more fluid in order to maintain vascular pressure.
- If there is fault in pituitary gland and kidneys are normal then urine output is decreased
- If the fault is with the kidneys, urine will remain dilute and excessive in amount because the diseased kidneys cannot concentrate fluid.

Management

Surgery is indicated in case of tumor. If the cause is idiopathic, the condition can be controlled by the administration of desmopressin (DDAVP), an arginine vasopressin. In an emergency, DDAVP is given intravenously. For long-term use, it is given intranasally or orally. If desmopressin is given as an intranasal spray, this may cause nasal irritation. Intranasal route will not be effective if the child has an upper respiratory tract infection with swollen mucous membranes. Caution children that they will notice an increasing urine output just before the next dose.

Syndrome of Inappropriate Antidiuretic Hormone (SIADH)

SIADH is characterized by **overproduction** of ADH by the posterior pituitary gland. This results in a decrease in urine production, which leads to water intoxication. As sodium levels fall in proportion to water, the child develops hyponatremia. SIADH can be caused by CNS infections such as bacterial meningitis, long-term positive pressure ventilation, or pituitary compression due to edema or a tumor.

Mild symptoms of hyponatremia are weight gain, urine with increased specific gravity, nausea, and vomiting. As the hyponatremia becomes severe, coma or seizures occur from brain edema.

Management consists of restriction of fluid and supplementation of sodium by IV fluids if needed. The following drugs may be used:

- **Vasopressin receptor antagonist:** Conivaptan and tolvaptan
- **Loop diuretics:** Lasix
- **Tetracyclines:** Demeclocycline.

Difference between DI and SIADH is shown in Table 13.2.

ADRENAL DISORDERS

Adrenal Insufficiency

There are two adrenal glands, located above the kidneys. Adrenal gland consists of adrenal cortex and adrenal medulla (Fig.13.3). Adrenal cortex secretes:

- **Mineralocorticoids:** Aldosterone for sodium and fluid balance,
- **Glucocorticoids:** Cortisol for blood glucose regulation, and

Table 13.2: Difference between DI and SIADH

Diabetes Insipidus	SIADH
Low ADH, Low water in body	High ADH, water intoxication
High urine output (polyuria)	Low urine output (oliguria)
Complication-Hypovolemic Shock	Complication-Seizures
Treatment- DDAVP	Treatment-Fluid Restriction

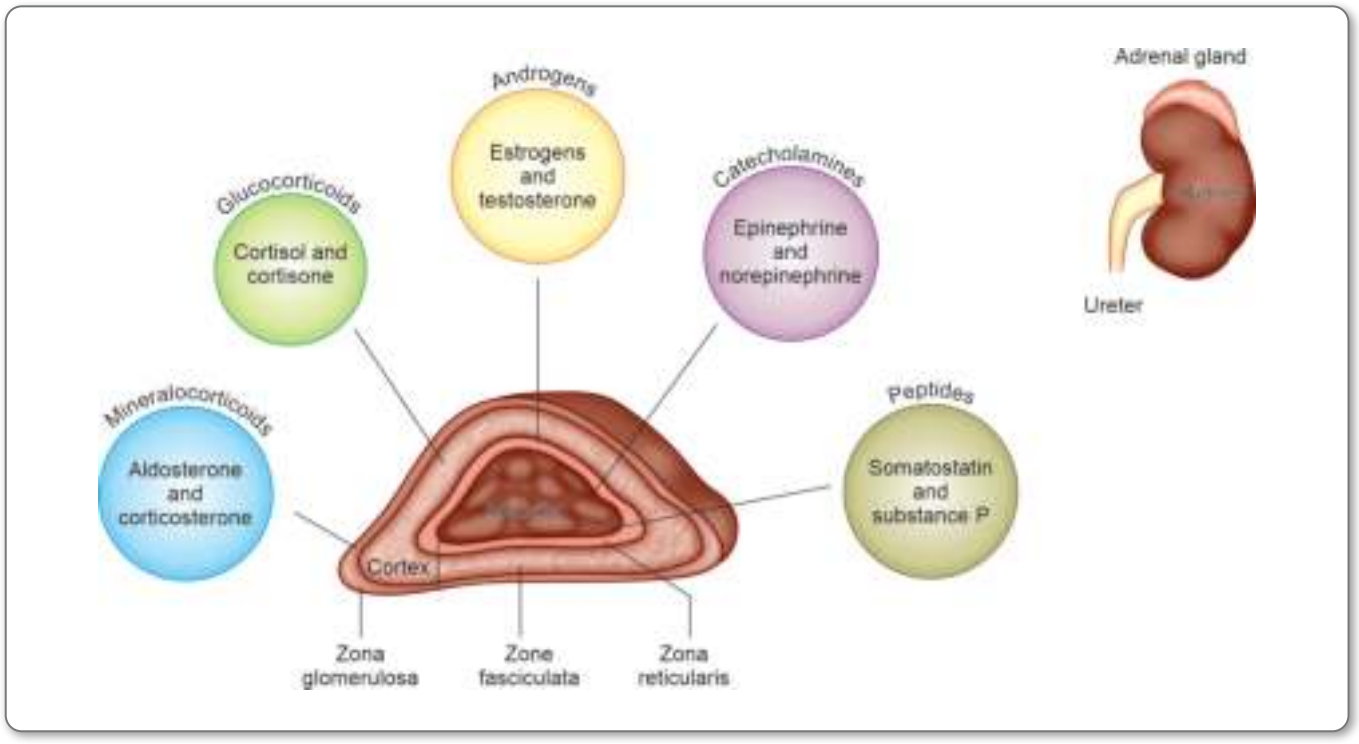

Figure 13.3: Adrenal gland hormones

- **Androgens:** (DHEA, androstenedione) for sexual development.

Adrenal insufficiency is of 2 types: Congenital and acquired.

Causes of Adrenal Insufficiency

The most frequent cause of acquired primary adrenal insufficiency is autoimmune and is associated with the presence of antibodies that are associated with damage to the adrenal gland. Genetic disorders can also cause primary adrenal insufficiency. Other causes include infections, abnormal bleeding into the adrenal gland, adrenal tumors, and surgical removal of the adrenal gland. Babies can be born with congenital primary adrenal insufficiency because of the inability of the adrenal gland to make enough cortisol and/or aldosterone.

Symptoms of Adrenal insufficiency

The symptoms of adrenal insufficiency include fatigue, muscle weakness, decreased appetite, and weight loss. Infants may fail to regain their birth weight and have trouble feeding. Some individuals experience nausea, vomiting, and diarrhea. In older children, symptoms can include dizziness, sweating, low blood sugar, and low blood pressure. Individuals with primary adrenal insufficiency may have salt craving and darkening of the skin.

Diagnosis

The most common way to diagnose primary adrenal deficiency is to obtain a fasting blood sample early in the morning to check both cortisol and ACTH levels.

- In primary adrenal insufficiency, the cortisol level will be low with an elevated ACTH level.
- In secondary adrenal insufficiency, the cortisol level is low with an ACTH level that is low or normal but not high.

Sometimes, an ACTH stimulation test will be needed to confirm the diagnosis.

Blood work includes measurement of blood sodium, potassium, glucose, and plasma renin activity. In some instances, imaging studies, such as ultrasound, MRI, or CT scans, may be helpful.

Management

Treatment includes hormone replacement. Oral hydrocortisone or other similar medications are used to replace cortisol and need to be taken 2 to 3 times a day. Patients with aldosterone deficiency usually take fludrocortisone to help maintain salt balance. The hydrocortisone dose needs to be increased at times of significant body stress because child's body cannot make more hydrocortisone. This is called stress dosing. Examples of stress include fever, severe diarrhea, severe vomiting, severe trauma, or surgery. If a child is not able to take oral medications because of vomiting or being unconscious, hydrocortisone injections, (e.g., Solu-Cortef, Hydrocortisone sodium succinate) can be used; an emergency hydrocortisone injection kit for intramuscular injections should be available for such situations. It is important for parents to learn how and when to administer intramuscular hydrocortisone injections. With appropriate treatment,

children with adrenal insufficiency can lead a normal life and have a normal life span.

Nursing Management

Goal: Maintain adequate hormone levels for optimized ability to create energy and respond to stress and electrolyte balance to regulate blood pressure.

Interventions

- **Assess vitals:** Temperature, blood pressure and heart rate. Watch for orthostatic changes and hyperpyrexia
- Monitor weight, intake and output
- Encourage oral fluids
- Minimize stress and assist with activities/provide rest periods
- **Monitor nutrition:** Aldosterone deficiency causes the kidneys to excrete sodium which may result in salt cravings. Encourage patients to increase salt intake and supplements as necessary to prevent hyponatremia. Reinforce to eat high protein/low carb snacks and meals as tolerated followed by rest periods to prevent fatigue due to hypoglycemia and to facilitate digestion.
  - Monitor EKG for signs of hyperkalemia. Lack of aldosterone means increased sodium excretion and increased potassium retention. Signs of hyperkalemia will include peaked T-waves and prolonged QRS complex.
- Administer medications
  - **Kayexalate:** Orally or by enema to reduce potassium levels
  - **Cortef or Cortone and prednisone:** Orally or IV to increase cortisol levels
  - **Florinef:** Given orally to promote replacement and retention of sodium and water

Pheochromocytoma

Pheochromocytoma is a rare catecholamine-secreting tumor of adrenal medulla. It is a type of neuroendocrine tumor that grows from cells called chromaffin cells which are found in the adrenal glands. However, the tumor may develop anywhere in the body. Release of catecholamines (norepinephrine and epinephrine) into the circulation by these tumors causes significant hypertension.

Clinical Manifestations

The clinical manifestations of a pheochromocytoma result from excessive catecholamine secretion by the tumor. Catecholamines typically secreted are norepinephrine and epinephrine. Stimulation of alpha-adrenergic receptors results in elevated blood pressure, increased cardiac contractility, glycogenolysis, gluconeogenesis, and intestinal relaxation. Stimulation of beta-adrenergic receptors results in an increase in heart rate and contractility.

The classic clinical presentation includes paroxysmal attacks of headaches, pallor, palpitations, and diaphoresis. Other symptoms include-hypertension, nausea, vomiting, visual disturbances, polyuria, polydipsia, convulsions, etc.

Diagnosis

Diagnostic tests for pheochromocytoma include the following:

- Plasma metanephrine testing
- 24-hour urinary collection for catecholamines and metanephrines

Management

Medical management includes avoiding stimuli which precipitate hypertensive crisis, alpha adrenergic blocking agents, rest, non-stressful environment and high calorie diet.

Definitive treatment is with surgical removal, i.e., **adrenalectomy**. Chemotherapy and radiotherapy in metastatic and unresectable pheochromocytoma.

GROWTH HORMONE INSUFFICIENCY

Growth hormone, also called somatotropin, is a polypeptide hormone which stimulates growth and cell reproduction. The decrease or excess level of growth hormone in the blood is known as growth hormone insufficiency.

Growth Hormone Deficiency

Growth hormone deficiency (GHD) is a rare disorder characterized by the inadequate secretion of growth hormone (GH) from the anterior pituitary gland. Growth represents general health of a child.

Causes of Growth Hormone Deficiency

- Congenital GHD results from genetic error, and may be associated with brain structure defects. Several congenital defects have been identified (Fig. 13.4):
  - Dwarfisms
  - **Turner syndrome:** Characterized by short stature, ovarian dysgenesis (underdeveloped, degenerate ovaries), and, in some cases, unusual physical appearance
  - Russell-Silver syndrome characterized by unilateral poor growth, short stature, characteristic facies, mental delays.
  - **Prader-Willi syndrome:** Characterized by below-normal intelligence; small stature; hypogonadism, failure to thrive in infancy followed by obesity and insatiable appetite, bizarre binge-type eating behaviors.
  - **Down syndrome:** Characterized by growth retardation and mental retardation
  - Cystic fibrosis occurs in 1 in 2,000 live births.
- Acquired GHD can occur as a result of many different causes including brain trauma (perinatal or postnatal),

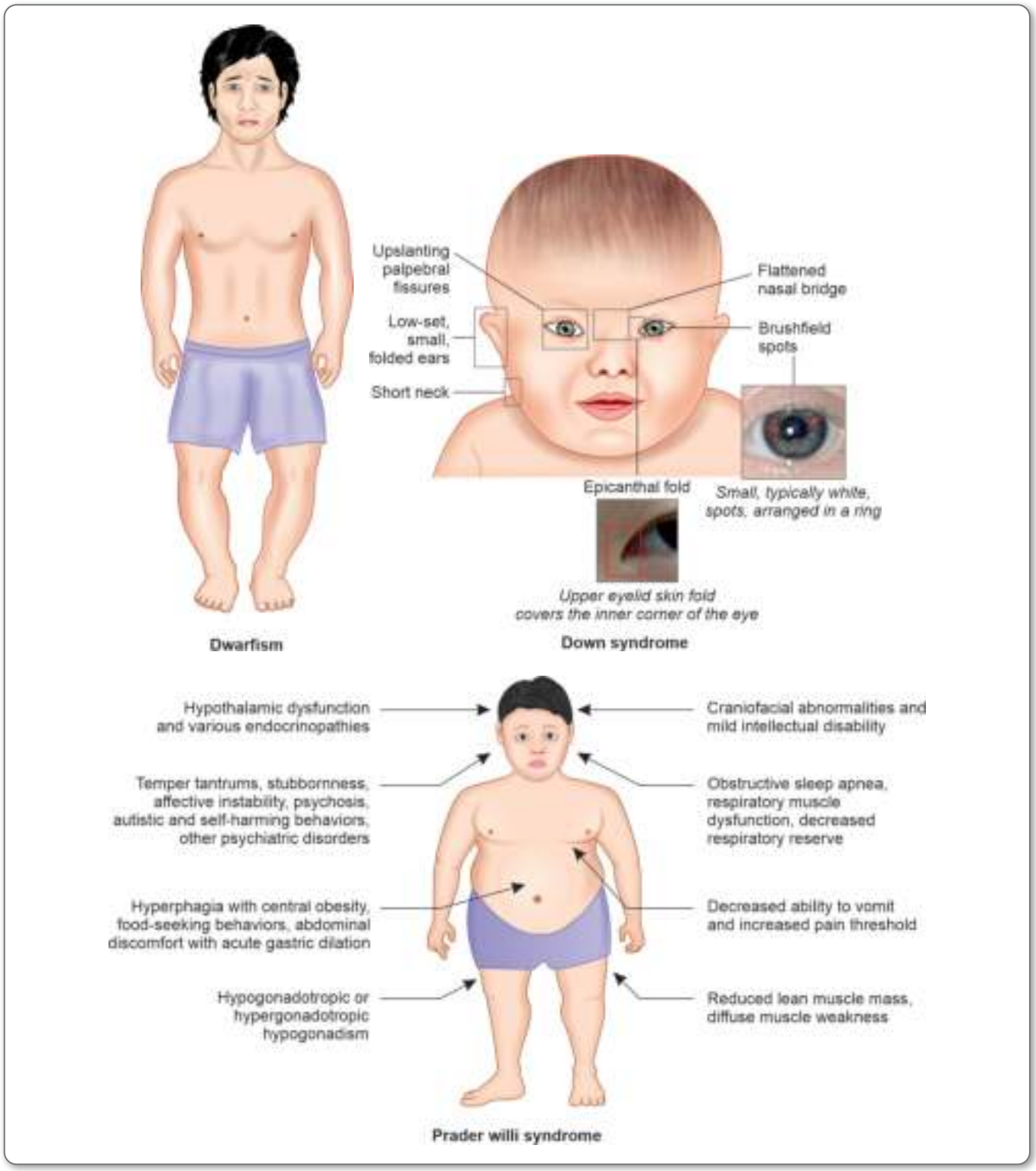

Figure 13.4: Examples of growth hormone deficiency

CNS infection, tumors of the hypothalamus or pituitary (pituitary adenoma, craniopharyngioma), radiation therapy, infiltrative diseases (Langerhans cell histiocytosis, sarcoidosis, tuberculosis)

- Idiopathic or unknown cause

Clinical Manifestations

- Hypoglycemia
- Prolonged jaundice
- Microphallus (small penis) usually in the neonate

- Growth velocity usually less than the fifth percentile for chronologic age
- Delayed skeletal maturation "bone age at least 1 year delayed from chronologic age"
- Frequently delayed eruption of primary and secondary teeth (not as severe as in hypothyroidism)
- Short stature
- Overweight for height, truncal obesity
- Crowding of midfacial structures, frontal bossing, depressed nasal bridge and prominent philtrum
- Delayed sexual development

Diagnosis

Testing is very important in determining whether the child with growth retardation does indeed have growth hormone deficiency. Rule out organic, nonendocrine causes of short stature, (i.e., chronic illness, nutritional deficiencies, genetic disorders, psychosocial factors).

- Calculate growth velocity (deviated growth pattern from the growth curve)
- Height <3rd percentile
- Prepubertal growth velocity <4 cm/year
- Bone age < chronological age
- Resumption of growth following GH administration
- Chromosome testing of females to rule out turner syndrome.
- Thyroid function tests to rule out hypothyroidism.
- GH secretion laboratory indicators: IGF-1 (insulin-like growth factor 1), IGF-binding protein 3 are decreased.
- Subnormal secretion of GH in response to two provocative stimuli: Peak GH levels <10 ng/mL during provocative stimulation tests
  - Insulin-induced hypoglycemia
  - Abnormal stimulatory response to arginine infusion, L-dopa, clonidine, or glucagon; all of which have specific actions resulting in GH secretion from pituitary. Pharmacologic agent is given, followed by blood sampling for GH response; GH levels less than 10 ng/mL is abnormal.
- FreeT4, TSH, cortisol, celiac antibodies, etc. are measured to rule out underlying organic causes of short stature.
- MRI of head to rule out the cause.

Children with severe GHD should be re-tested after completing growth to see if they meet the requirements for GH therapy as an adult.

Management

Goal of treatment is to restore normal growth and development as well as to maximize growth potential and prevent hypoglycemia.

- Growth monitoring
- Children with GHD should be started on recombinant human growth hormone given as subcutaneous injection as soon as the disorder is recognized to optimize growth potential. Initial dosage of Recombinant GH is 0.07–0.1 IU/kg/day. The dosage is gradually increased to its highest dose during puberty, and discontinued at or near completion of skeletal maturation when the patient may require retesting to see if GH is needed as an adult.

Nursing Management

- Evaluate growth pattern
  - Perform frequent and accurate measurements of height and weight.
  - Accurately plot measurements on growth curve for absolute chronologic age.
  - Assess growth pattern for any deviation from normal.
  - Report any child whose pattern deviates from the expected pattern for age.
- Health Education
  - Teach method of injecting GH through written and verbal instructions. Give demonstration and encourage return demonstration.
  - Encourage rotation of sites in the subcutaneous tissue of the upper arms or thighs to prevent skin irritation and hypertrophy.
  - Encourage regular follow-up for growth evaluation and maintenance of therapy. Document growth every 3–6 months while on therapy.
  - If poor growth response, evaluate for appropriate dose, compliance, and injection technique. There may be initial catch-up growth that will be exhibited by a growth velocity above normal.
  - Instruct patient and 0 family to report adverse effects: severe headache, hip and knee pain, limp, increased thirst and urination.
  - Review medication dosage and injection technique periodically.
  - Discuss potential lifelong therapy, which may be necessary depending on the degree of deficiency. Many hypopituitary patients require lifelong replacement therapy.
- Encourage social interaction
  - Encourage the child to verbalize feelings regarding short stature.
  - Have the child describe what he or she likes about certain people to help him/her understand that friendships and social values are based on personality traits rather than absolute height.
  - Suggest involvement in activities that do not use height as an advantage, such as music, art, and gymnastics.
- Strengthen self esteem
  - Help child and parents to identify age-appropriate behaviors and develop a plan for maintaining consistent behaviors in the home and socially.
  - Make sure parents have realistic expectations of child.
  - Encourage the use of positive feedback rather than punishment.

Figure 13.5: Gigantism

Growth Hormone Excess

An overproduction of GH usually is caused by a benign tumor of the anterior pituitary (an adenoma). If the overproduction occurs before the epiphyseal lines of the long bones have closed, excessive or overgrowth will result, also called as **Gigantism (Fig. 13.5)**. Weight will become excessive also, but it is proportional to height. The skull circumference typically exceeds usual, and the fontanels may close late or not at all.

After epiphyseal lines close, **acromegaly** (enlargement of the bones of the head and soft parts of the hands and feet) begins to be evident. The tongue can become so enlarged and thickened that it protrudes from the mouth, giving the child a dull, apathetic appearance and making it difficult to articulate words. If the condition remains untreated, a child may reach a height of more than 8 ft.

Diagnosis

- Clinical examination
- Growth assessment
- X-ray
- CT/MRI
- GH levels >400 ng/mL

Management

- Laser resection of adenoma
- Radiation
- GH antagonist such as Bromocriptine (orally) - 10–20 mg/day or Octreotide (injection) to slow GH production

DISORDERS OF PANCREAS

The pancreas has both endocrine (ductless) and exocrine (with duct) types of tissue. The *islets of Langerhans* form the endocrine portion, alpha islet cells secrete glucagon, and beta islet cells secrete insulin. Insulin is essential for carbohydrate, fats and protein metabolism.

The term diabetes mellitus describes a metabolic disorder of multiple etiologies characterized by chronic hyperglycemia with disturbances of carbohydrate, fat and protein metabolism resulting from defects of insulin secretion, insulin action or both.

Type 1 Diabetes (IDDM)

Diabetes mellitus (DM) is a chronic metabolic disorder caused by an absolute or relative deficiency of insulin, an anabolic hormone. Insulin is produced by the beta cells of the islets of Langerhans located in the pancreas, and the absence, destruction, or other loss of these cells results in type 1 diabetes (insulin-dependent diabetes mellitus [IDDM]). Comparison of Type 1 and 2 is shown in Table 13.3.

Causes

Most cases (95%) of type 1 diabetes mellitus are the result of environmental factors interacting with a genetically susceptible person. This interaction leads to the development of autoimmune disease directed at the insulin-producing cells of the pancreatic islets of Langerhans. These cells are progressively destroyed, with insulin deficiency usually developing after the destruction of 90% of islet cells.

Human leukocyte antigen (HLA) class II molecules DR3 and DR4 are associated strongly with type 1 diabetes mellitus.

Dietary factors are also relevant. Breastfed infants have a lower risk for insulin-dependent diabetes mellitus (IDDM)

Other causes: Include congenital absence of the pancreas or islet cells, Pancreatectomy and secondary to pancreatic damage, (i.e., cystic fibrosis, chronic pancreatitis, thalassemia major, hemolytic-uremic syndrome)

Pathophysiology (Figure 13.6)

- Insulin is essential to process carbohydrates, fat, and protein. Functions of insulin:
  - **Helps in glycogenesis:** Insulin reduces blood glucose levels by allowing glucose to enter muscle cells and by stimulating the conversion of glucose to glycogen (glycogenesis) as a carbohydrate store.
  - **Prevents glycogenolysis:** Insulin also inhibits the release of stored glucose from liver glycogen (glycogenolysis) and slows the breakdown of fat to triglycerides, free fatty acids, and ketones. It also stimulates fat storage.
  - **Prevents gluconeogenesis:** Additionally, insulin inhibits the breakdown of protein and fat for glucose production (gluconeogenesis) in both liver and kidneys.

Table 13.3: Comparison of Type 1 and Type 2 diabetes

Assessment	Type 1	Type 2
Age at onset	5–7 years or at puberty	40–65 years (may occur in adolescents as maturity-onset diabetes of youth [MODY])
Type of onset	Abrupt	Gradual
Weight changes	Marked weight loss often initial sign	Associated with obesity
Other symptoms	Polydipsia and polyphagia	Polydipsia
	Polyuria (often begins as bed-wetting)	Polyuria
	Fatigue (marks fall in school)	Fatigue
	Blurred vision (marks fall in school)	Blurred vision
	Mood changes (may cause behavior problems in school)	Mood changes
Therapy	Hypoglycemia agents never effective; insulin required	Diet, oral hypoglycemic agents, or insulin
	No dietary foods used; should count carbohydrates plus evaluate blood glucose levels to help determine insulin dosage.	Nutrition concentrates on no excess weight gain and balanced intake of carbohydrates, protein, and fat
	Commonsense foot care for growing children	Meticulous skin and foot care necessary
Period of remission	Period of remission for 1–12 months ("honeymoon period") generally after initial diagnosis	Not demonstrable

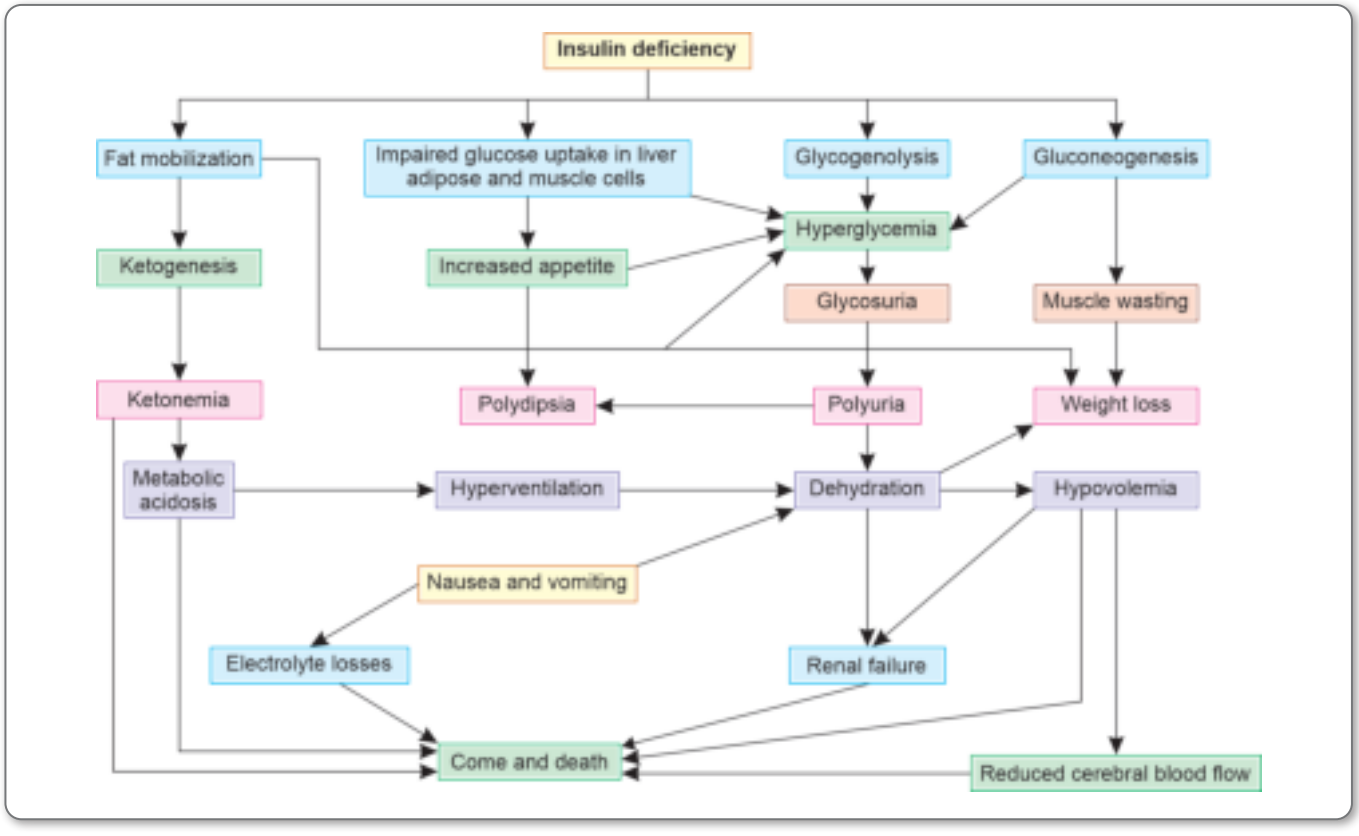

Figure 13.6: Pathophysiology of diabetes mellitus type 1

- Hyperglycemia (random blood glucose concentration more than 200 mg/dL or 11 mmol/L) results when insulin deficiency leads to uninhibited gluconeogenesis and prevents the use and storage of circulating glucose.
- The kidneys cannot reabsorb the excess glucose load, causing glycosuria, osmotic diuresis, thirst, and dehydration.
- Increased fat and protein breakdown leads to ketone production and weight loss.

- Without insulin, a child with type 1 diabetes mellitus wastes away and eventually dies due to diabetic ketoacidosis (DKA).
- Glucose is the only energy source for erythrocytes, kidney, medulla and brain.

Signs and Symptoms

- **Hyperglycemia:** It occurs secondary to osmotic diuresis and glycosuria.
- **Glycosuria:** It leads to increased urinary frequency and polyuria, which is particularly troublesome at night (e.g., nocturia) and often leads to enuresis in a previously continent child. These symptoms are easy to overlook in infants because of their naturally high fluid intake and diaper/napkin use.
- **Polydipsia:** Secondary to the osmotic diuresis causing dehydration.
- **Weight loss:** Insulin deficiency leads to uninhibited gluconeogenesis, causing breakdown of protein and fat leading to weight loss. Failure to thrive and wasting may be the first symptoms noted in an infant or toddler and may precede frank hyperglycemia.
- **Symptoms of ketoacidosis:** Severe dehydration, Smell of ketones, acidotic breathing, (i.e., Kussmaul respiration) masked as respiratory distress, abdominal pain, vomiting, drowsiness and coma.
- **Other non-specific findings:** Hyperglycemia impairs immunity and renders a child more susceptible to recurrent infection, particularly of the urinary tract, skin, and respiratory tract. Candidiasis may develop, especially in groin and flexural areas.
- **Symptoms in infants:** Severe monilial diaper/napkin rash, unexplained malaise, poor weight gain or weight loss, increased thirst, vomiting and dehydration, with a constantly wet napkin/diaper

Diagnosis

- **Urine glucose:** A positive urine glucose test suggests but is not diagnostic for type 1 diabetes mellitus (T_1DM). Diagnosis must be confirmed by test results showing elevated blood glucose levels.
- **Urine ketones:** Ketones in the urine confirm lipolysis and gluconeogenesis, which are normal during periods of starvation. With hyperglycemia and heavy glycosuria, ketonuria is a marker of insulin deficiency and potential DKA.
- **Blood glucose:** A random whole-blood glucose concentration of more than 200 mg/dL (11 mmol/L) is diagnostic for diabetes. A fasting whole-blood glucose concentration exceeds 120 mg/dL (7 mmol/L). Blood glucose tests using capillary blood samples, reagent sticks, and blood glucose meters are the usual methods for monitoring day-to-day diabetes control.
- **Glycated hemoglobin:** Glycosylated hemoglobin derivatives (HbA1a, HbA1b, HbA1c) are the result of a nonenzymatic reaction between glucose and hemoglobin. A strong correlation exists between average blood-glucose concentrations over an 8-week to 10-week period and the proportion of glycated hemoglobin. Measurement of HbA1c levels is the best method for medium-term to long-term diabetic control monitoring. Check HbA1c levels every 3 months. Normal value for HbA1c is between 7–9%. Values less than 7% are associated with an increased risk of severe hypoglycemia; values more than 9% carry an increased risk of long-term complications.
- Oral glucose tolerance test (OGTT):
  - Obtain a fasting blood sugar level, then administer an oral glucose load (2 g/kg for children aged <3 years, 1.75 g/kg for children aged 3–10 years [max 50 g], or 75 g for children aged >10 years).
  - Check the blood glucose concentration again after 2 hours.
  - A fasting whole-blood glucose level higher than 120 mg/dL (6.7 mmol/L) or a 2-hour value higher than 200 mg/dL (11 mmol/L) indicates diabetes.
  - However, mild elevations may not indicate diabetes when the patient has no symptoms and no diabetes-related antibodies.
- **Lipid profile:** Lipid profiles are usually abnormal at diagnosis because of increased circulating triglycerides caused by gluconeogenesis. Apart from hypertriglyceridemia, primary lipid disorders rarely result in diabetes. Hyperlipidemia with poor metabolic control is common.

Treatment

- All children with type 1 diabetes mellitus (T_1DM) require insulin therapy.
- Only children with significant dehydration, persistent vomiting, or metabolic derangement, or with serious intercurrent illness, require inpatient management and intravenous rehydration.
- Train the child or family to check blood glucose levels, to administer insulin injections, and to recognize and treat hypoglycemia.
- Insulin has 3 basic formulations:
  - Short-acting (e.g., regular, soluble, lispro, aspart, glulisine)
  - Medium-acting or intermediate-acting, (e.g., isophane, lente, detemir), and
  - Long-acting, (e.g., ultralente, glargine).

Insulin Lispro (Humalog): Onset of action is 10–30 min, peak activity is 1–2 hours, and duration of action is 2–4 hours.

Pediatric: 0.5–1 U/kg/d SC initially. Adjust doses to achieve premeal and bedtime blood glucose levels of: <5 years: 100–200 mg/dL (5.5-10 mMol/L) ≥5 years: 80–140 mg/dL (4–7.5 mMol/L).

Regular insulin (Humulin R, Novolin R): Onset of action is 0.25–1 hours, peak activity is 1.5–4 hours, and duration of action is 5–9 hours.

Injection technique: Teach parents that when insulins are mixed in one syringe, the regular or short-acting insulin should be drawn into the syringe first. Then, if mixing accidentally occurs in the bottle, the time of effectiveness of the short-acting insulin (which needs to be kept short-acting for emergency treatment) will not be lengthened by the addition of the intermediate-acting insulin.

Insulin is always injected SC except in emergencies, when half the required dose may be given IV. Subcutaneous tissue injection sites used most frequently in children include those of the upper outer arms and the outer aspects of the thighs (Fig. 13.7). The abdominal subcutaneous tissue injection sites commonly used in adults can be adequate sites, but most children dislike this site because abdominal skin is tender. Encourage children or parents to rotate sites in a pattern based on their planned activity. Absorption, for example, is increased if the muscles under the injection site are exercised, so it is best to choose sites that will not be exercised soon after the injection. If a child will be jogging after an injection, for example, the thigh probably should not be used. Similarly, if the child will be playing tennis, the injection probably should not be given in the dominant arm.

- **Dietary management:** The following are the most recent consensus recommendations:
  - Carbohydrates should provide 50–55% of daily energy intake. (No more than 10% of carbohydrates should be from sucrose or other refined carbohydrates).
  - Fat should provide 30–35% of daily energy intake.
  - Protein should provide 10–15% of daily energy intake.

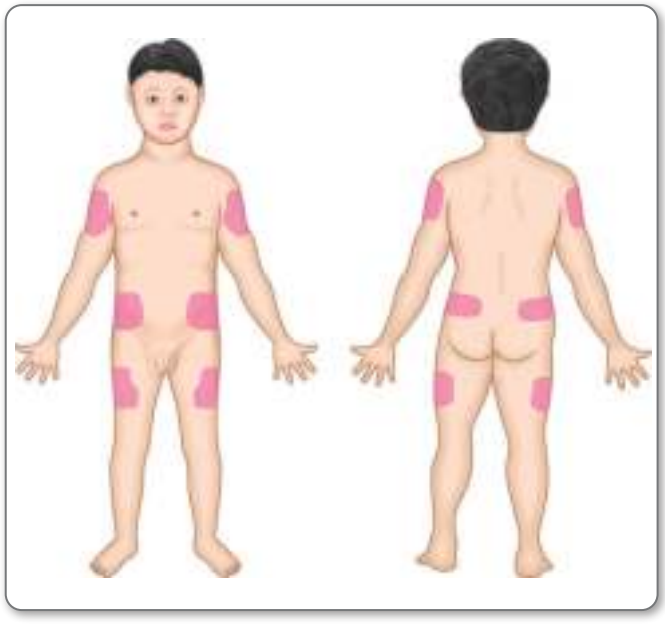

Figure 13.7: Insulin injection sites

The complications of type 1 diabetes mellitus can be divided into 3 major categories:

1. Acute complications,
2. Long-term complications, and
3. Complications caused by associated autoimmune diseases.

Acute complications reflect the difficulties of maintaining a balance between insulin therapy, dietary intake, and exercise. Acute complications include hypoglycemia, hyperglycemia, and DKA.

Hypoglycemia

- If insulin is injected in a treated diabetic child who has not eaten adequate amounts of carbohydrates, blood glucose levels progressively fall.
- The brain depends on glucose as a fuel. As glucose levels drop below 65 mg/dL (3.2 mmol/L) counter-regulatory hormones, (e.g., glucagon, cortisol, epinephrine) are released, and symptoms of hypoglycemia develop. These symptoms include sweatiness, shaking, confusion, behavioral changes, and eventually, coma when blood glucose levels fall below 30–40 mg/dL.
  - Mild hypoglycemia can be managed by giving rapidly absorbed oral carbohydrate or glucose.
  - For a comatose patient, administer an IM injection of the hormone glucagon, which stimulates the release of liver glycogen and releases glucose into the circulation.
  - Where appropriate, an alternative therapy is IV glucose (preferably no more than a 10% glucose solution). All treatments for hypoglycemia provide recovery in approximately 10 minutes.

Hyperglycemia

In a child with diabetes, blood sugar levels rise if insulin is insufficient for a given glucose load. The renal threshold for glucose reabsorption is exceeded when blood glucose levels exceed 180 mg/dL (10 mmol/L), causing glycosuria with the typical symptoms of polyuria and polydipsia. All children with diabetes experience episodes of hyperglycemia. Persistent hyperglycemia in very young children (<4y) may lead to later intellectual impairment.

Diabetic Ketoacidosis (DKA)

DKA is a life-threatening medical emergency. In the absence of insulin, body starts breaking fats as fuel and produces lots of ketones quickly which make blood acidic. DKA usually follows increasing hyperglycemia and symptoms of osmotic diuresis. Children are more likely to present with nausea, vomiting, and abdominal pain and symptoms similar to food poisoning.

Injection-site hypertrophy: If children repeatedly inject their insulin into the same area, subcutaneous tissue swelling may

develop, causing unsightly lumps and adversely affecting insulin absorption. Injection sites must be rotated to avoid injection site hypertrophy.

Long-term complications: These arise from the damaging effects of prolonged hyperglycemia and other metabolic consequences of insulin deficiency on various tissues. These include: Retinopathy, Cataracts, Hypertension, Progressive renal failure, Early coronary artery disease, Peripheral vascular disease, Neuropathy (both peripheral and autonomic) and increased risk of infection.

Nursing Management

Growth measurement: The measurement of height, weight and body mass index is an integral component of diabetes care for children and adolescents. Anthropometric measurements should be plotted on an appropriate centile chart. Changes in growth or significant changes in weight, or pubertal delay, may reflect changing glycaemic control. In such cases, comorbidities such as coeliac disease or thyroid dysfunction should also be considered.

- Dietary advice and meal planning should be revised regularly to meet changes in appetite and insulin regimens, and to ensure optimal growth. Prevention of overweight and obesity is a key strategy in the management of type 1 diabetes.
- Regular review and adjustment of insulin doses (and basal rates on pumps) is required in children and adolescents, because insulin requirements can change rapidly with growth and puberty. In particular, significant insulin resistance may occur during puberty, and insulin requirements typically increase (>1 unit/kg/day). Post-pubertally, insulin doses usually decline.

DISORDERS OF THYROID GLAND

Thyroid gland secretes thyroxine (T_4) and triiodothyronine (T_3). The action of these hormones promotes cellular growth and differentiation, protein synthesis, and lipid metabolism (cholesterol turnover). The secretion of thyroid hormone is regulated by hypothalamus and pituitary gland. Function of the thyroid gland is controlled by a negative feedback mechanism utilizing TSH released from the pituitary gland (Fig. 13.8). Disorders of the thyroid gland are broadly classified as hypothyroidism and hyperthyroidism.

Hypothyroidism

Normally, the thyroid gland should be fully developed in all babies by the 22nd week of pregnancy. However, this development can go wrong and lead to underdevelopment or absence of the thyroid gland. This condition is called hypothyroidism. Severe hypothyroidism in an infant is called **cretinism.** In children there are two main ways that hypothyroidism can occur:

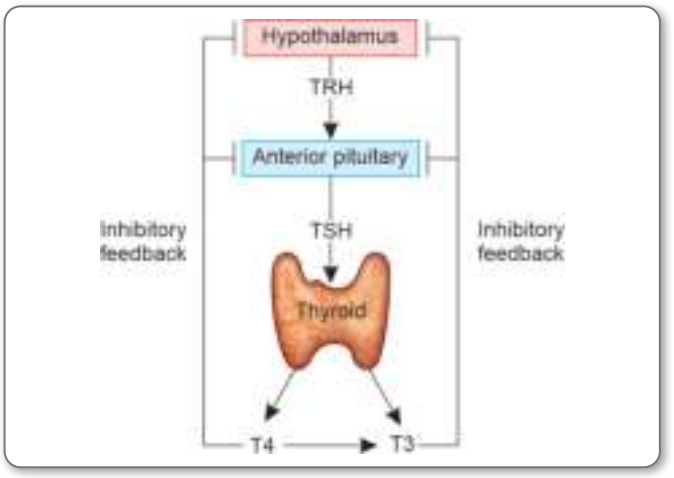

Figure 13.8: Thyroid gland hormones

- Hypothyroidism which is present from birth is called congenital hypothyroidism.
- Hypothyroidism that develops later in a child's life is called late onset or acquired hypothyroidism.

Risk Factors

Around 40% of children with acquired hypothyroidism have relatives who also have some type of thyroid gland problem. Girls and women are much more commonly affected than boys or men. In addition, the following conditions are considered a risk factor for hypothyroidism: Down's Syndrome, Turner Syndrome, Metabolic and Blood Conditions, and Pituitary Gland Problems.

Clinical Manifestations

Neonates (for congenital hypothyroidism): Neonates presents with very subtle physical signs, if any

- Markedly open posterior fontanelle
- Prolonged physiologic jaundice
- Feeding difficulties
- Skin cool to touch, mottled
- Poor muscle tone/Hypotonia
- Umbilical hernia

After Age 6 Months (for congenital hypothyroidism)

- Growth failure
- Large, protruding tongue
- Coarse facial features
- Open posterior fontanel
- Umbilical hernia
- Hypotonia
- Large abdomen
- Poor feeding and constipation

Acquired Cases

- Growth retardation, slow growth rate, increased weight gain
- Puffy appearance of face

- Lethargy, obedient, nonaggressive, somnolent
- Cold intolerance
- Delayed skeletal maturation, delayed dental development and delayed puberty
- Possible poor school performance
- Constipation

Diagnosis

- Neonatal screening of T_4 and TSH (TSH>100mU/mL)
- Thyroid nuclear scan: Reduced uptake
- Abnormal growth rate
- Bone age X-ray; delayed
- Thyroid antibodies elevated in autoimmune thyroiditis

Management

- Replacement of thyroid hormone: levothyroxine (Synthroid, Levothroid).
  - Infants 10–15 mcg/kg/day
  - Older children 4–8 mcg/kg/day
- Cortisol replacement
- Therapy goal is to maintain normalcy of thyroid function tests (free T_4, T_4 and T_3 concentrations) in the upper half of normal range.

Complications

- Mental retardation in neonate who is undiagnosed or untreated
- Short stature, growth failure, and delayed physical maturation and development in the older child

Nursing Management

- Assessment
  - Assess neonate for clinical manifestations listed above.
  - Perform behavioral assessment to include sleeping, eating, bowel patterns, level of alertness, and school performance.
  - Assess growth patterns: Growth velocity (rate of growth over time), weight gain, and head circumference.
- Promote growth and development
  - Teach parents to administer thyroid hormone replacement daily.
  - Mix the medicine with small amount of fluid and give with dropper or syringe, or crush tablet and place in teaspoon of infant food.
  - Monitor growth and developmental milestones at regular intervals.
- Alleviate parental anxiety
  - Encourage the parents to verbalize feelings about child and the condition.
  - Educate the parents as to the importance of the therapy so child will grow and develop normally.
  - Stress that with replacement therapy, the child can participate in all usual activities.
- Family Education and Health Maintenance
  - Encourage follow-up for blood studies and evaluation of neurologic development to ensure adequate treatment and prevent mental retardation.
  - Make sure the family understands that therapy is usually life-long.
  - Support the parents and refer the child to special testing and therapy if mental retardation is suspected.
  - Teach family the importance of avoiding over dosage of levothyroxine and to be alert for signs of over dosage, i.e., weight loss, restlessness, heat intolerance, fatigue, muscle weakness, tachycardia.

Hyperthyroidism

Hyperthyroidism is a disorder of the thyroid gland in which there is a high circulating level of T_4. Elevated circulating thyroid hormone increases the body's metabolic rate, causing an increase in excitability of the neuromuscular, cardiovascular and sympathetic nervous system.

Etiology

- **Graves' disease:** It is caused by stimulation of TSH receptor located on the thyroid gland by an antibody, which is known as TSH receptor antibody (TRAb). Furthermore, this may lead to hyperplasia and hyperfunction of the thyroid gland. Autoimmune process leads to release of thyrotropin receptor antibodies of a stimulating nature which stimulates thyroid gland to produce and secrete T_4.
- May also be caused by ingestion or overdosage of thyroid medication (iatrogenic)
- Subacute (viral) thyroiditis
- Chronic lymphocytic thyroiditis
- Bacterial thyroiditis
- Pituitary adenoma

Clinical Manifestations

- Thyromegaly (enlargement of the thyroid)
- Polyphagia with weight loss
- Exophthalmos, proptosis, lid retraction
- Hyperactivity, restlessness, nervousness, hand tremors, sleeping disturbances, emotional lability, inability to concentrate, decreased school performance.
- Heat intolerance, excessive diaphoresis
- Fatigue, muscle weakness
- Tachycardia, wide pulse pressure
- Tall stature, underweight for height

Difference between symptoms of hypothyroidism and hyperthyroidism is explained in Figure 13.9.

Diagnostic Evaluation

- Serum thyroid function test shows elevated T_4, T_3 resin uptake with a suppressed TSH

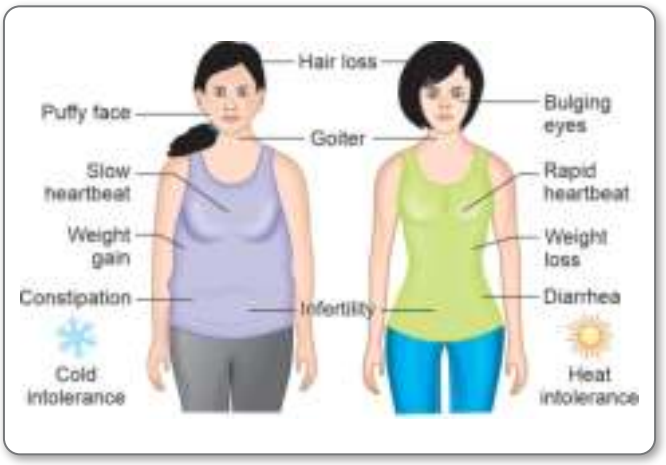

Figure 13.9: Difference between hypothyroidism and hyperthyroidism

- Microsomal antibodies positive
- Thyroid radionuclide scan of goiter is done to rule out cold nodules that could indicate thyroid carcinoma

Management

- **Antithyroid drugs:** The major drug prescribed to reduce the secretion of thyroxine is Carbimazole although very occasionally the drug Propylthiouracil or methimazole (Tapazole) is used. Carbimazole is generally well tolerated but can produce side effects in about 5% of children. Adverse effects include headache, nausea, diarrhea, skin rash, itching, liver disease, jaundice, arthralgia and, rarely, agranulocytosis.
- Propranolol (Inderal), a β-adrenergic blocking agent, for cardiac effects.
- Radioactive ablation of thyroid gland using radioiodine preferred over thyroidectomy. This is chosen when medical management is ineffective and results in permanent hypothyroidism that requires treatment.
- The thyroid gland can be removed by surgery (thyroidectomy). Thyroid hormone replacement (a daily tablet) will then be needed in the long term to replace what the thyroid gland is no longer making.

Nursing Management

Assessment

- Perform physical assessment. Assess temperature, heart rate, BP, height, and weight.
- Obtain history related to development of symptoms, any changes in behavior, school performance, emotions, or sleep patterns.
- Assess for presence of goiter pain with swallowing or talking, palpation of thyroid.

Improve Activity Tolerance

- Administer and teach parents to administer medications and comply with treatment to gradually lower the metabolic rate and improve activity tolerance.
- Assess activity tolerance periodically. Ascertain if fatigue is present at rest, with activities of daily living, or with exercise.
- Promote relaxation and rest between activities.
- Ensuring nutritional intake
  - Encourage high-calorie, nutritious diet to maintain weight.
  - Advise parents that effective treatment will lower metabolic rate and facilitate appropriate weight gain.
  - Periodically assess growth and development parameters.
- Normalizing sleep pattern
  - Assess sleep pattern, including naps and sleeping through the night.
  - Adjust schedule to allow maximum amount of rest until sleeping through the night.
  - Allow short naps as needed.
- Allaying anxiety
  - Encourage parents and child to verbalize fears related to the thyroid ablation.
  - Teach them about the procedure and clear misconceptions.
  - Ablation is accomplished through radioactive destruction of thyroid tissue by administration of a radioactive iodine pill.
  - The thyroid gland is the only tissue in the body that absorbs iodine; therefore, the radiation only destroys thyroid tissue.
  - Radiation will be eliminated through the child's urine and feces; precautions for disposal must be followed according to nuclear medicine department policies.
  - Explain that the effect of radioactive ablation will be hypothyroidism, which can be managed with lifelong treatment of T_4 replacement.
- Health Education
  - Teach about medications and stress the importance of compliance.
- Advise about periodic blood testing and monitoring for adverse effects of antithyroid medication.
  - Educate family about possibility of over suppression, from which hypothyroidism could develop.
  - Encourage follow-up to monitor treatment through blood tests, growth and development evaluation, and size of thyroid gland.

Summary

The endocrine system creates and controls child's hormones. The hormones in child's body regulate everything from their body temperature to their mood, growth, and more.

Hormones are secreted by endocrine glands, such as the pituitary gland, thyroid, ovaries, or adrenal glands. These hormones are sent through the blood stream to various tissues and organs in the body.

When there is an imbalance of child's hormones, it can cause serious problems. The common risk factors include obesity and genetics. Symptoms of an endocrine disorder vary widely depending on the specific gland involved. Hormone therapy is generally the first line of defense for treating pediatric endocrine disorders.

Assess Yourself

1. List the hormones secreted by anterior and posterior pituitary.
2. Discuss the nursing interventions for a child with Diabetes Mellitus Type 1.
3. Compare the clinical manifestations of a child with hypothyroidism and hyperthyroidism.
4. Define – acromegaly, cretinism, ketoacidosis, pheochromocytoma.