Pediatric Endocrine Dysfunction

Chapter 52
Debra Mercer BSN, RN, RRT

Mechanisms of Hormonal Regulation

The endocrine system is composed of various glands located throughout the body. These glands can synthesize and release hormones. General functions include:
1. Differentiation of the reproductive and central nervous systems in the developing fetus
2. Stimulation of growth and development
3. Maintenance of an optimal internal environment throughout life
4. Initiation of corrective and adaptive responses when emergency demands occur
Endocrine Glands

- Anterior lobe (adenohypophysis) releases or withholds 7 hormones
- These hormones influence somatic and sexual development.
- Clinical manifestations depend on the hormones involved and the age of onset.

Pituitary Gland
Disorders Of The Pituitary Function

- Hypopituitarism
- Pituitary hyperfunction
- Precocious puberty
- Diabetes insipidus
- Syndrome of inappropriate antidiuretic hormone secretion (SIADH)

HYPOPITUITARISM

- Primarily associated with a deficient secretion of somatotropin (growth hormone (GH)).
- In 50% of cases no lesion is evident and the cause is unknown.
- Primary site of dysfunction appears to be in the hypothalamus.
- Treated more frequently in boys than in girls.
- Frequently associated with other pituitary hormone deficiencies.
Therapeutic Management of GH Deficiency

- Treat underlying disease
- Definitive treatment of GH deficiency is replacement of GH.
- Biosynthetic GH prepared by recombinant deoxyribonucleic acid (DNA) technology is the treatment of choice.
- May require additional hormones depending on any other disorders associated with GH deficiency.

Hyopituitarism

Many clinical pictures with diverse symptomology

Unrecognized → Death

Unrecognized

Unrecognized → Death

Unrecognized

Unrecognized → Death

Unrecognized

Unrecognized → Death

Unrecognized

Unrecognized → Death

Unrecognized

Unrecognized → Death

Unrecognized
Creutzfeldt-Jakob Disease (CJD)

- Rare neurodegenerative condition reported in some patients after receiving human GH from cadavers
- Does not occur with biosynthetic GH
- Blood banks will not accept donation from former human GH recipients because of risk of infection with CJD

Nursing Considerations

- Family support needs
- Child’s body image
- Preparing child for daily injections
- Treatment very expensive ($20,000 to $30,000/year)

Pituitary Hyperfunction

- Excess GH before closure of epiphyseal shafts results in overgrowth of long bones
- Reach heights of 8 feet or more
- Vertical growth plus increased muscle
- Weight generally in proportion to height
Acromegaly

Diagnostic Evaluation
- History of excessive growth through childhood
- Evidence of increased levels of GH
- Enlargement of bones
- Joint changes
- Endocrine studies
**Therapeutic Management**
- Surgical treatment to remove tumor
- Radiation and radioactive implants
- Hormone replacement therapy after surgery in some cases

**Nursing Considerations**
- Early identification of children with excessive growth rates
- Early treatment for improved outcomes
- Emotional support
- Body image concerns

**Precocious Puberty**
- Defined as sexual development before age 9 in boys or before age 8 in girls
- Occurs more frequently in girls
- Potential causes
  - Disorder of gonads, adrenal glands, or hypothalamic-pituitary gonadal axis
  - No causative factor in 80% to 90% of girls and 50% boys
Types of Precocious Puberty

- True or complete precocious puberty
- Precocious pseudopuberty
- Incomplete puberty

Therapeutic Management

- Treatment of specific cause if known
- May be treated with leuprolide (Lupron)
  - Slows prepubertal growth to normal rates
  - Treatment is discontinued at age for normal pubertal changes to resume
- Psychologic support for child and family

Diabetes Insipidus (DI)

- The principal disorder of the posterior pituitary
- Results from hyposecretion of ADH
- Produces uncontrolled diuresis
- Primary causes: familial or idiopathic
- Secondary causes: trauma, tumors, CNS infection, aneurysm
Clinical Manifestations of DI

• Cardinal signs: polyuria and polydipsia
• First sign is often enuresis
• Infants: irritability relieved with feedings of water but not milk; dehydration often occurs

Therapeutic Management of DI

• Instruct parents in difference between diabetes insipidus and diabetes mellitus
• Daily hormone replacement of vasopressin
• Drug of choice: DDAVP
  – Nasal spray or IV administration
  – Requires treatment for life

Syndrome of Inappropriate Antidiuretic Hormone (SIADH)

• Produced by hypersecretion of the posterior pituitary (increased ADH)
• S/S: fluid retention and hypotonicity
• Kidneys unable to reabsorb water
• Anorexia, nausea/vomiting, irritability, personality changes
• Symptoms disappear when ADH is decreased
Disorders Of Thyroid Function

- Juvenile hypothyroidism
- Goiter
- Lymphocytic thyroiditis
- Hyperthyroidism

Disorders of Thyroid Function

- Thyroid hormone regulates BMR
- Thyroid secretes two types of hormones
  - Thyroid hormone, which is made up of:
    - Thyroxin (T4)
    - Triiodothyronine (T3)
  - Thyrocalcitonin
- May have hypothyroidism or hyperthyroidism
- May have disturbance in secretion of TSH
Juvenile Hypothyroidism

- **Congenital**
  - Congenital hypoplastic thyroid gland
- **Acquired**
  - Partial or complete thyroidectomy for CA or thyrotoxicosis
  - Following radiation for Hodgkin or other malignancy
- Rarely occurs from dietary insufficiency in US
Therapeutic Management

- Oral thyroid hormone replacement
- Prompt treatment needed for brain growth in infant
- May administer in increasing amounts over 4 to 8 weeks to reach euthyroidism
- Compliance with medication regimen is crucial

Goiter

- Hypertrophy of the thyroid gland
- Congenital
  - Usually results from maternal ingestion of antithyroid drugs during pregnancy
- Acquired
  - Result of neoplasm, inflammatory disease, dietary deficiency (but rarely in children), or increased secretion of pituitary thyrotropic hormone
Lymphocytic Thyroiditis

- Hashimoto disease or juvenile autoimmune thyroiditis
- Most common cause of thyroid disease in children and teens
- Accounts for largest percent of juvenile hypothyroidism

Hyperthyroidism (Graves Disease)

- Most common cause of hyperthyroidism in childhood is Graves disease
- Believed to be caused by serum thyroid-stimulating immunoglobulin, but no specific etiology
- Enlarged thyroid gland and exophthalmos
- Peak incidence 12 to 14 years of age, but may be present at birth
- Familial association
Graves Disease Management

- Therapy is controversial
- Goal of therapy: to retard rate of hormone secretion
- Treatments
  - Antithyroid drugs (PTU and methimazole)
  - Subtotal thyroidectomy
  - Ablation with radiiodine

Thyrotoxicosis

- Thyroid "crisis" or "storm"
- May occur from sudden release of hormone
- Unusual in children, but can be life threatening
- May be precipitated by infection, surgery, or discontinuation of antithyroid therapy
- Treatment of thyroid storm
  - Antithyroid drugs
  - Propranolol

Nursing Considerations

- Identify children with hypothyroidism
- Alert for signs and symptoms
- Child needs quiet environment, rest periods
- Help family cope with emotional lability associated with disorder
- Dietary requirements to meet child’s increased metabolic rate
- Medications: side effects
Disorders of Parathyroid Function

- Hypoparathyroidism
- Hyperparathyroidism
- Acute Adrenocortical Insufficiency
- Cushing syndrome
- Addison disease
- Congenital Adrenal Hyperplasia (CAH)

Disorders of Parathyroid Function

- Parathyroid glands secrete parathormone (PTH)
- Function of PTH: to maintain serum calcium by:
  - Increasing release of calcium and phosphate from bone demineralization
  - Increase absorption of calcium and excretion of phosphate by the kidneys
  - Promote calcium absorption in GI tract

Hypoparathyroidism

- Autoimmune hypoparathyroidism
  - Deficient production of PTH
- Pseudohypoparathyroidism
  - Production of PTH is increased but end organs are unresponsive to the hormone
  - Thought to be inherited as X-linked-dominant trait with variable expressivity
Hyperparathyroidism

- Primary: adenoma of the gland
- Secondary: chronic renal disease, congenital anomalies of urinary tract
- Common factor is hypercalcemia
- Rare in children

Disorders of Adrenal Function

- Acute adrenocortical insufficiency
- Addison disease
- Cushing syndrome
- Congenital adrenal hyperplasia
- Hyperaldosteronism
Disorders of Adrenal Function

- Adrenal cortex secretes three groups of "steroids"
  - Glucocorticoids (cortisol, corticosterone)
  - Mineralocorticoids (aldosterone)
  - Sex steroids (androgens, estrogens, and progestins)
- Altered levels of these produce significant dysfunction
- Adrenal medulla secretes catecholamines: epinephrine and norepinephrine
- Catecholamine-secreting tumors are the primary cause of adrenal medullary hyperfunction

Acute Adrenocortical Insufficiency

- Adrenal crisis
- Etiologic factors: hemorrhage into the gland from trauma, fulminating infections, abrupt withdrawal of exogenous cortisone, failure to increase cortisone during times of stress
- Diagnosis generally based on clinical symptoms

Chronic Adrenocortical Insufficiency: Addison Disease

- Rare in children
- When it occurs, usually result of neoplasms or lesion of adrenal glands or idiopathic cause
- Symptoms appear gradually after 90% adrenal tissue is nonfunctional
Cushing Syndrome

- A characteristic group of manifestations caused by excessive circulating free cortisol
- May be caused by excessive or prolonged steroid therapy
- Condition is reversible once steroids are discontinued
- Abrupt withdrawal of steroids may precipitate acute adrenal insufficiency

Etiologies of Cushing Syndrome

- Pituitary: excess ACTH
- Adrenal: hypersecretion of glucocorticoids
- Ectopic: extrapituitary neoplasm
- Iatrogenic: administration of excessive steroids
- Food dependent: inappropriate adrenal response to secretion of polypeptide
Cushingoid Appearance

Cushing Syndrome

- Therapeutic management
  - Surgery
  - Replacement of growth hormone, ADH, TH, gonadotropins, and steroids
- Nursing considerations

Congenital Adrenal Hyperplasia (CAH)

- Caused by excessive secretion of androgens by the adrenal cortex
- Pathophysiology: six major types of biochemical defects, interference in the biosynthesis of cortisol in fetal life
- Karyotype for positive sex determination for those clients with ambiguous genitalia
- Management includes cortisone to suppress elevated ACTH levels
Hyperaldosteronism
- May be caused by adrenal tumor
- May be caused by adrenogenital syndromes
- Results in hypertension, hypokalemia, and polyuria that fails to respond to ADH administration

Pheochromocytoma
- Adrenal tumor that secretes catecholamines
- May occur around adrenal medulla, along paraganglia of aorta, or thoracolumbar sympathetic chain
- Children often have bilateral, multiple, benign tumors
- Increased production of catecholamines; may mimic other disorders
Disorders of Pancreatic Hormone Function

- Diabetes mellitus
  A. Etiology
  B. Pathophysiology
  C. Diagnostics
  D. Therapeutic management
  E. Nursing Care

Pancreatic Hormone Function

- Function of islets of Langerhans
  - Alpha cells produce glucagon
  - Beta cells produce insulin
  - Delta cells produce somatostatin (believed to regulate insulin and glucagon)
Diabetes Mellitus (DM)

- Characterized by a total or partial deficiency of the hormone insulin
- The most common endocrine disorder of childhood
- Peak incidence in early adolescence

Three Major Groups of DM

- Type 1
- Type 2
- Maturity onset diabetes of the young (MODY)

Type 1 Diabetes

- Characterized by destruction of beta cells, usually leading to absolute insulin deficiency. Typically, onset in childhood and adolescence, but can occur at any age
- Most DM of childhood is type
Type 2 Diabetes

- Arises because of insulin resistance
- Onset usually after age 40
- Native American, Hispanic, and African-American children are at increased risk of type 2 DM
- Affected people may require insulin injections

MODY

- Transmitted as autosomal dominant disorder with formation of structurally abnormal insulin with decreased biologic activity
- Onset is generally before age 25

Ketoacidosis

- When glucose is unavailable for cellular metabolism, the body breaks down alternate sources of energy. Ketones are released, and excess ketones are eliminated in urine (ketonuria) or by the lungs (acetone breath)
- Ketones in the blood are strong acids that lower serum pH and produce ketoacidosis
Kussmaul Respirations

- Hyperventilation characteristic of metabolic acidosis, resulting from respiratory system's attempt to eliminate excess CO₂ by increased depth and rate

Diabetic Ketoacidosis: DKA

- Pediatric emergency
- Results from progressive deterioration with dehydration, electrolyte imbalance, acidosis, coma; may cause death

Therapeutic Management of Type 1 DM

- Insulin therapy
- Glucose monitoring: goal range 80-120 mg/dl
- Lab measurement of hemoglobin A₁
- Urine testing for ketones
  - Not routinely used except:
  - Helpful to test q3h during illness and whenever glucose is ≥240 mg/dl when illness not present
Insulin Therapy

- Highly purified pork preparations and human insulin
- Rapid, short, intermediate, long acting
- All are packaged in the strength of 100 units/ml
- Amount of insulin to give is based on capillary blood glucose levels
- Regular insulin best administered 30 min ac SQ, Lispro best administered 10-15 minutes ac SQ

Nutrition

- Children require no special foods or supplements
- They need consistent intake and timing of food, especially carbohydrates
- Balanced diet incorporates six basic food groups: milk, meat, vegetables, fat, fruit, bread
- Fiber is important because it diminishes rise in blood glucose after meals
Exercise

- Lowers blood glucose levels
- Regular exercise aids in the body’s use of food
- Decreases insulin requirements

Hypoglycemia

- Occurs with bursts of physical activity, activity without additional food, delayed, omitted or incompletely consumed meals or snacks
- Should carry a source of glucose
- Glucose needs to be followed by a complex carbohydrate like crackers/bread with peanut butter
- Somogyi effect: rebound hyperglycemia from the release of stress hormones

Nursing Care Management

- Assess: daily blood glucose levels
- Educational needs include pathophysiology of diabetes, meal planning, insulin, injection procedure, glucose monitoring, urine testing, hyperglycemia/hypoglycemia, exercise, self-management, facilitating family support, yearly eye exam, record keeping
- Increase dietary intake with increased activity and exercise
Possible Nursing Diagnoses for Pediatric Client with DM

- Risk for injury related to insulin deficiency
- Risk for injury related to hypoglycemia
- Deficient knowledge (diabetes management) related to care of child with newly diagnosed diabetes mellitus

Acanthosis Nigricans