Pediatric endocrine and metabolic disorders NCLEX Review

The endocrine system is comprised of several glands throughout the body. Glands release chemical messengers called hormones that control and regulate the activity of target cells and organs. Hormones influence growth, development, and digestion and regulate metabolism and reproduction. Glands release the hormones into the blood to a stimulus, another hormone, or a threshold. Glands stop releasing hormones when they receive the signal to turn off the hormone production by a process called direct feedback. The endocrine system maintains homeostasis. Feedback tells a gland to increase or decrease the hormone production so the body returns to homeostasis. When the concentration of a hormone reaches a threshold, hormone production is turned off.

Thyroid Gland

The thyroid gland is located in the anterior neck, overlying the trachea. The thyroid gland makes:

- Thyroxine (T4): Regulates carbohydrate metabolism, lipids, proteins, and growth and development.
- Triiodothyronine (T3):Regulates carbohydrate metabolism, lipids, proteins, and growth and development.
- Calcitonin: Regulates blood calcium and phosphate release from the bones.

The pituitary gland is a pea-sized gland connected with the hypothalamus and divided into two parts:

- Anterior pituitary that produces

1. Thyroid-stimulating hormone (TSH):Stimulates the thyroid gland to produce hormones.
2. Growth hormone (GH): Increases protein synthesis, increases fat mobilization, and decreases the use of carbohydrate, all of which encourages tissue growth.
3. Follicle-stimulating hormone (FSH):Stimulates graafian follicles to mature and secrete estrogen (female) and stimulates the seminiferous tubules development (males).
4. Luteinizing hormone (LH):Causes the rupture of the follicle resulting in the release of the ovum (female). Stimulates production of testosterone (males).
5. Prolactin hormone: Simulates the secretion of breast milk.

- Posterior pituitary that produces

1. Oxytocin hormone: Stimulates uterus contraction and the letdown lactating reflex.
2. Antidiuretic hormone (ADH):Regulates the concentration of fluids in the body by altering the permeability of the collecting ducts and distal convoluted tubules in the kidneys.

Adrenal Glands

- Adrenal glands are located at the top of each kidney in the retro peritoneum. Adrenal glands are comprised of two parts: Adrenal cortex:

1. Aldosterone: Responsible for renal reabsorption of sodium and excretion of potassium.
2. Cortisol:Maintains glucose control, increases hepatic gluconeogenesis (the making of glucose), and manages the body's stress response.
3. Androgens:Male sex hormones that promote secondary sex characteristics.

- Adrenal medulla:

1. Catecholamines:

a. Epinephrine:Increases heart and respiratory rate, and blood pressure and dilates airways and an increase in the metabolic rate.

b. Norepinephrine: Increases heart and respiratory rate, and blood pressure and dilates airways and an increase in the metabolic rate.

Parathyroid Glands

Parathyroid glands are composed of usually four or more small glands located on the posterior side of the thyroid gland. Parathyroid glands produce:

- Parathyroid hormone (PTH):Maintains the calcium level in the blood and inhibits phosphorus. If the serum calcium level falls, PTH is released causing bones to break down, releasing calcium into the blood and kidneys to retain calcium and increase phosphate excretion.

Pancreas

The pancreas contains a cluster of cells called the islets of Langerhans. These cells produce:

- Insulin: Produced by the beta cells, insulin increases the cellular use of glucose.
- Glucagon: Produced by the alpha cells, glucagon increases glucose when the blood level of glucose is low.
- Somatostatin: Produced by the delta cells, somatostatin inhibits the release of corticotrophin and growth hormone.

Gonads

Gonads are sex glands and include:

- Ovaries (female) are located on the uterus and produce eggs and also produce

1. Estrogen: Promotes secondary female characteristics, regulates menstrual cycle.
2. Progesterone: Supports pregnancy and prepares the breasts for lactation.

- Testes (male) are located in the scrotum and produce testosterone which stimulates production of spermatozoa and maintains the secondary male sex characteristics.

II. CONGENITAL HYPOTHYROIDISM

Congenital hypothyroidism is a lack of, or too little, thyroid hormone during fetal development or following birth resulting from an underdeveloped or absent thyroid gland caused by the mother's iodine deficiency or taking antithyroid medication during pregnancy or from autoimmune thyroiditis. Congenital hypothyroidism affects growth of the nervous system and bone and affects mental development if left untreated.

NCLEX!!! Treatment must begin within 3 months of age to ensure normal development.

Assessment:

- Fatigue due to slow metabolism
- Noisy respiration due to an enlarged tongue
- Hypothermia due to slow metabolism
- Short, thick extremities and neck
- Brittle nails due to low levels of thyroid hormone that helps growth and development
- Thin dry hair from lack of thyroid hormone
- Dry skin from lack of thyroid hormone
- Slow cognitive function due to slow metabolism
- Weight gain due to low levels of thyroid hormone that causes fatigue, sluggishness
- Cognitive impairment due to untreated condition

Diagnostics:

- Thyroid scan: Decreased uptake of iodine or absence of thyroid.
- Serum TSH: Increase TSH unless the cause is due to a decrease production of TSH by the pituitary gland.
- Serum thyroid hormone: Decrease T3, T4.
- Radiograph: Absence of tibial or femoral epiphyseal line.
- Electrocardiogram: Flat or inverted T waves and bradycardia

Treatment:

- Replacement hormone (levothyroxine, liothyronine).
- Serum measurements of T3 and T4 will need to be performed after 6 to 8 weeks to determine if the patient is taking the correct dose.
- The patient needs to be aware this is lifetime replacement.
- Vitamin D supplement to prevent rickets that might result from rapid bone growth.

Nursing Interventions:

- Ensure that the newborn is screened for congenital hypothyroidism so that treatment can begin within 3 months of birth.
- Monitor vital signs because treatment may cause tachycardia and hypertension.
- Monitor for irritability, sweating, and fever that indicate the dose is too high.
- Monitor for lethargy, constipation, decreased appetite, and fatigue that indicates the dose is too low.
- Provide a warm environment.
- Low-calorie diet.
- Increase fluids and fiber to prevent constipation.

- Take thyroid replacement hormone each morning to avoid insomnia.
- Monitor for signs of thyrotoxicosis (an increase in T3) (nausea, vomiting, diarrhea, sweating, tachycardia).
- Explain to the family the side effects of thyroid hormone replacement and review the signs of hyperthyroidism and hypothyroidism. Also teach the family that treatment is lifelong.

III. CUSHING SYNDROME

Cushing syndrome occurs when the adrenal cortex secretes an excess of glucocorticoids or an excess secretion of adrenocorticotropic hormone (ACTH) by the pituitary gland as a result of either a pituitary tumor or adrenal tumor or from ongoing glucocorticoid therapy.

Assessment:

- Moon face during excess cortisol production
- Weight gain
- Buffalo hump (fat pad located in the upper back) from excessive corticosteroids
- Osteoporosis from an excess of corticosteroids, which weaken the bones
- Bruising
- Delayed wound healing
- Sleep disturbance
- Reddish purple striae on the abdomen
- Changes in mental status from excessive steroids

Diagnostics:

- Dexamethasone suppression test: A dose of glucocorticoid is given to test the hypothalamus-pituitary-adrenal axis. If there is suppression of cortisol with the dose, it indicates a pituitary origin of the excess cortisol. If no suppression occurs, the etiology is an adrenal or ectopic tumor.
- A24-hour urine collection: Increase in cortisol from excess production.
- Computed tomograph (CT) scan: Presence of a pituitary tumor or adrenal tumor.
- Serum: Increase blood glucose due to overproduction of steroids.
- Serum: Increase sodium due to excess fluid loss.
- Serum: Decrease potassium.

Treatment:

- Surgical removal of the pituitary tumor or adrenal tumor

NCLEX!!! The child is highly susceptible to infections; therefore it is critical to prevent the child from being exposed to infections

Nursing Interventions:

- Weigh daily to monitor fluid status.
- Monitor input and output to ensure adequate hydration.
- Monitor for glucose and acetone in urine because elevated levels of corticosteroids may produce hyperglycemia.

- Allow for adequate rest to allow the body to stabilize.
- Avoid skin because elevated levels of corticosteroids can delay wound healing.
- Bone densitometry to assess for osteoporosis because corticosteroids can leech calcium from the bone.
- Following surgery:

1. Assist in early ambulation, deep breathing, coughing to facilitate mucus mobilization, decrease risk for emboli.
2. Monitor incision site for drainage, erythema, signs of infection.
3. Elevate the head of bed 30 degrees to reduce intracranial pressure.

- Explain to the family to maintain a high-calcium, high-protein, high potassium, low-carbohydrate, low-sodium, and low-calorie diet to aid in wound repair and replace calcium.
- Administer pain medication as needed

IV. DIABETES MELLITUS

Certain foods are converted into glucose, which is the primary energy supply. Insulin from beta cells of the pancreas transports glucose into cells for cell metabolism.

Diabetes mellitus occurs when beta cells either are unable to produce insulin (type 1 diabetes mellitus) or produces an insufficient amount of insulin (type 2 diabetes mellitus). As a result, glucose doesn't enter cells and remains in the blood.

Increased glucose levels in the blood signal the body to increase the intake of fluid to flush glucose out of the body in urine, resulting in increased thirst and increased urination in the patient. Cells become starved for energy because of the lack of glucose and signal the body to eat causing the patient to experience an increase in hunger.

There are three types of diabetes mellitus:

1. Type 1: Known as insulin dependent diabetes mellitus (IDDM). Beta cells are destroyed by an autoimmune process. There is a genetic predisposition, although coxsackie B, mumps, and congenital rubella viruses injure beta cells and can result in type 1 diabetes.
2. Type 2: Known as noninsulin dependent diabetes mellitus (NIDDM). Beta cells produce insufficient insulin.
3. Gestational diabetes mellitus: Insufficient insulin is produced by the mother during pregnancy. Patients with gestational diabetes mellitus recover following pregnancy; however, they are at risk for developing type 2 diabetes mellitus later in life.

NCLEX!!! Patients with type 1 and type 2 diabetes mellitus are at risk for vision loss (diabetic retinopathy), damaged blood vessels and nerves (diabetic neuropathy), and kidney damage (nephropathy). However, complications can be minimized by maintaining a normal blood glucose level through consistent monitoring, administering insulin, and dieting.

Assessment:

Type 1:

- Fast onset because no insulin is being produced.
- Increased appetite (polyphagia) because cells are starved for energy and are signaling a need for more food.
- Increased thirst (polydipsia) from the body attempting to rid itself of glucose.
- Increased urination (polyuria) from the body attempting to rid itself of glucose.
- Weight loss because glucose is unable to enter cells.
- Frequent infections as bacteria feeds on the excess glucose.
- Delayed healing because elevated glucose levels in the blood hinder healing process.

Type 2:

- Slow onset because some insulin is being produced.
- Increased thirst (polydipsia) from the body attempting to rid itself of glucose.
- Increased urination (polyuria) from the body attempting to rid itself of glucose.
- Candidal infection as bacteria feeds on the excess glucose.
- Delayed healing because elevated glucose levels in the blood hinder healing process.

Diagnostics:

- Urine test: Increase glucose in urine (glucosuria).
- Fasting plasma blood glucose test: Aplasma glucose level of ?126 mg/dL (or 7.0 mmol/L) on three different tests.
- Oral glucose tolerance Test (OGTT): Aplasma glucose of ?200 mg/dL (or 11.1 mmol/L) 2 hours after ingesting 75 g oral glucose.
- Random plasma glucose test: Aplasma of ?200 mg/dLor 11.1 mmol/L.
- Glycosylated hemoglobin A1C: ?6.0%.

Treatment:

Type 1:

- Regular monitoring of blood glucose.
- Administer insulin to maintain normal blood glucose levels .
- Maintain a diabetic diet.
- Administer:

Rapid acting:

1. Aspart
2. Lispro
3. Glulisine

Short acting: Regular insulin

Intermediate: NPH

Long acting:

- Glargine

- Lantus

Inhaled insulin

Exubera: Ashort-acting insulin for before-meal control

Type 2:

- Maintain ideal body weight through diet and exercise.
- Regular monitoring of blood glucose.
- Administer oral sulfonylureas to stimulate secretion of insulin from the pancreas.
- Administer oral Biguanides to reduce blood glucose production by the liver:

Metformin

- Administer thiazolidinediones to sensitize peripheral tissues to insulin:

Rosiglitazone

Pioglitazone

- Administer meglitinide analogs to stimulate section of insulin from the pancreas:

Repaglinide

- Administer D-phenylalanine derivative to stimulate insulin production:

Nateglinide

- Administer alpha-glucosidase inhibitors to delay absorption of carbohydrates in the intestine:

Acarbose

Miglitol

- Administer DPP4 (dipeptidyl peptidase 4) inhibitors to slow the inactivation of incretin hormones; GLP-I that assists insulin product in the pancreas:

Sitagliptin

- Administer incretin mimetics to assist insulin production in the pancreas and help regulate liver production of glucose. It also decreases appetite and increases the time glucose remains in the stomach before entering the small intestine for absorption.
- Administer amylin analog that causes glucose to enter the bloodstream slowly and can cause weight loss:

Pramlintide

Nursing Interventions:

- Educate the family and child about the disease and the importance of maintaining normal glucose levels.
- Demonstrate blood glucose monitoring.
- Review diet and food choices, including portion sizes.
- Encourage exercise. Discuss coping skills to reduce stress.
- Teach self-injection of insulin (type 1).
- Teach importance of daily medications and self-care including insulin injection. Explain to the family and patient the signs and symptoms and intervention for hypoglycemia, diabetic ketoacidosis, and hyperglycemia.

V. GALACTOSEMIA

Galactosemia is the inability of the patient to metabolize galactose in carbohydrate to glucose due to the missing hepatic enzyme GALT, resulting in the buildup of galactose in the blood and causing liver dysfunction.

Assessment:

- Vomiting following intake of milk
- Weight loss
- Diarrhea
- Jaundice
- Lethargy

Diagnostics:

- Serum: Increased galactose.
- Blood: Decreased or absent GALT activity in erythrocytes.
- Urine: Increased galactose.

Treatment:

- Galactose (lactose)-free diet (avoid dairy products, canned and frozen foods that contain lactose, cakes, cookies, pies, food coloring)

Nursing Interventions:

- Explain to the family and child the importance of avoiding dietary galactose.

VI. GRAVES DISEASE (HYPERTHYROIDISM)

Graves disease is an overproduction of T3 and T4 by the thyroid gland that can be caused by an autoimmune disease, a benign tumor (adenomas) resulting in an enlarged thyroid gland (goiter), or an overproduction of TSH by the pituitary gland caused by a pituitary tumor.

The prognosis is good if the cause is treated; however, this is a chronic disease. Signs such as bulging eyes (exophthalmos) are not reversible. Furthermore, thyroid surgery may result in complications.

Assessment:

- Enlarged thyroid gland (goiter) caused by tumor.
- Protrusion of the eyeballs (exophthalmos) due to lymphocytic infiltration that pushes out the eyeball.
- Irritability.
- Sweating (diaphoresis): Excess thyroid hormone raises the metabolic rate.
- Increased appetite due to increased metabolism.
- Hyperactivity due to high levels of thyroid hormone.
- Weight loss due to increased metabolism.
- Insomnia due to increased metabolism.

Diagnostics:

- Serum: Increased serum T3 and T4.
- Radioimmunoassay: Increased T4.
- Serum: Increased TRH and TSH if pituitary gland is the cause of hyperthyroidism.
- Serum: Presence of antibodies if cause is Graves disease.
- Thyroid scan: Enlarged thyroid

Treatment:

- For mild cases and for young patients, administer antithyroid medication such as propylthiouracil (PTU) and methimazole (Tapazole) to block synthesis of T3 and T4.
- For severe cases where the size of the thyroid gland interferes with swallowing or breathing, the thyroid gland is surgically reduced in size or removed. The patient must be on lifelong thyroid replacement therapy.

Nursing Interventions:

1. Monitor vital signs.
2. Provide cool environment.
3. Provide a quiet environment.
4. Protect the patient’s eyes with dark glasses and artificial tears if the patient has exophthalmos.
5. Provide a diet high in carbohydrates, protein, calories, vitamins, and minerals.
6. After surgery:

- Monitor for laryngeal edema following surgery (hoarseness or inability to speak clearly).
- Keep oxygen, suction, and a tracheotomy set near bed in case the neck swells and breathing is impaired.
- Keep calcium gluconate near the patient’s bed following surgery, which is the treatment for tetany, to maintain the serum calcium level in normal range.
- Place the patient in a semi-Fowler position to decrease tension on the neck following surgery.

- Support the patient's head and neck with pillows following surgery.
- Monitor for muscle spasms and tremors (tetany) caused by manipulation of the parathyroid glands during surgery.
- Check drainage and hemorrhage from incision line. Red flags are frank hemorrhage and purulent, foul-smelling drainage.
- Monitor signs of hypocalcemia (tingling of hands and fingers).
- Check for Chvostek sign (tapping of the facial nerve causes twitching of the facial muscles). These signs are positive when the parathyroid glands have been manipulated during thyroid surgery, in which case they secrete too much phosphorus and not enough calcium. Because muscles and the heart need calcium for work, a low calcium level may cause spasms of muscle, which is easily detected by Chvostek sign and Trousseau sign. The treatment is intravenous calcium, administered quickly.
- Check for Trousseau sign (inflate blood pressure cuff on the arm and muscles contract).

VII. MAPLE SYRUP URINE DISEASE

In maple syrup urine disease the branched-chain amino acids are defective or absent due to a genetic disorder resulting in an increase in branched-chain amino acids and ketoacids (by-products), causing a burnt sugar smell in urine.

Asssessment:

- Maple syrup odor from urine
- Seizures
- Difficulty feeding
- Moro reflex absent
- Abnormal respirations

Diagnostics:

- Serum: Increased branched-chain amino acids.
- Urine: Increased branched-chain amino acids.
- Blood gases: Acidosis.

Treatment:

- Increase dietary thiamine.
- Avoid dietary isoleucine, valine, and leucine.
- Hemodialysis to remove branched-chain amino acids from the body

Nursing Interventions:

- Perform urine and blood test following the first day of feeding.
- Assess diapers for maple syrup odor.
- Teach parents the importance of avoiding foods that contain isoleucine, valine, and leucine.

VIII. PHENYLKETONURIA

Phenylketonuria (PKU) is a genetic disorder that occurs because of a dysfunctional phenylalanine hydroxylase enzyme that is used to convert phenylalanine to tyrosine, resulting in an accumulation of phenylalanine in the body that can cause mental retardation.

Assessment:

- Family history of PKU
- Mental retardation as early as 4 months of age
- Dry skin
- Macrocephaly
- Irritable
- Hyperactive
- Musty skin odor
- Seizures (later years)

NCLEX!!! The child has normal blood phenylalanine levels at birth; however levels increase after birth and can result in irreversible damage by 2 years of age if not detected and treated.

Diagnostics:

- Guthrie screening test: Increased level in blood of phenylalanine 4 days after birth.
- Chromatography: Increased level in blood of phenylalanine 4 days after birth.

Treatment:

- Maintain blood levels of phenylalanine between 3 mg/dL and 9 mg/dL by restricting dietary phenylalanine (protein-rich foods).
- Administer enzymatic hydrolysate of casein (Lofenalac, Pregestimil) in place of milk.

Nursing Interventions:

- Explain to the family that the child should avoid eggs, meat, fish, poultry, breads, aspartame, and cheese for the child's entire life.
- The blood level of phenylalanine must be tested throughout the child's life to ensure that it remains within the desired level.

NCLEX!!! Be alert for signs of phenylalanine deficiency (anorexia, skin rashes, anemia, diarrhea, lethargy) that might occur from too little phenylalanine in the diet.