

**Newborn Reproductive**  
*Nursing 201: Nursing Care of Special Populations*

**Congenital Adrenal Hyperplasia:**

- Autosomal \_\_\_\_\_ inherited disorder
- Insufficient supply of enzyme required to synthesize \_\_\_\_\_ and \_\_\_\_\_.
- 90% of cases caused by deficiency of \_\_\_\_\_
- Incidence: 1 in 15-20,000 live births

**Hormone Review**

- **Cortisol:** adrenal hormone that maintains homeostasis, “stress hormone,” returns to normal after stressful event
  - Affects: glucose, fat/protein/carb metabolism, immune response, anti-inflammatory action, blood pressure, heart and blood vessel tone, CNS activation
- **Glucocorticoid:** essential for the utilization of carbs, fat, and protein by the body and for normal response to stress; anti-inflamm affects; decrease immune response
- **Mineralocorticoid:** regulates Na and water
- **Aldosterone:** regulates Na and K, controls BP, balances fluid and electrolytes
- **Androgen:** “male hormones”; male traits and female reproduction (necessary for synthesis of estrogen)
- **Adrenocorticotrophic hormone (ACTH)-** hormone formed in the pituitary gland; stimulated to produce cortisol
- **21- hydroxylase-** deficiency blocks the production of adrenal mineralocorticoids and glucocorticoids

**Cause:**

- 21-Hydroxylase deficiency blocks the production of mineralocorticoids and glucocorticoids
- \_\_\_\_\_ Cortisol
- \_\_\_\_\_ ACTH production by anterior pituitary to stimulate the adrenal gland production
- Prolonged \_\_\_\_\_ of ACTH causes hyperplasia of adrenal glands to produce excess \_\_\_\_\_.
- Male characteristics appear early or inappropriately

**Signs and Symptoms:**

Males:

Females:

**Complications:**

- Complications:
  - Hyponatremia
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  - Hyperkalemia
  - Hypotension
  - Dehydration
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  - Shock
  - Hypoglycemia
  - Short adult stature
  - Adult testicular tumors in males

**Mild Form:**

- Symptoms appear later (normal genitals at birth)
- Early puberty (toddler/preschool age)
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  - Males with normal fertility, females with lower
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**Diagnostics:**

- 21- hydroxylase deficiency detected on NB metabolic screening
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**Management:**

- STOP excess adrenal secretion of androgens
- Maintain normal Growth and Development
- FOR LIFE.....
  
- Infants may require Na replacement
- Females: controversial to correct genitalia at infancy or older
  - May need gender determination by karyotyping
  - Probable second surgery during puberty if corrected at infancy
  
- Monitor for Acute Adrenal Crisis
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  - Infant:
  
  - Treatment:
    - IV steroids (hydrocortisone)
    - Fluid Resuscitation (D5NS)
    - Correct electrolyte imbalance
    - Maintain oral hormone supplements
    - **In an emergency:**