ECHO Summary, 30/AUG/2024
Session Title: Adrenal and Pituitary Emergencies

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Definitions/Background

- Adrenal Insufficiency is an endocrine disorder due to a deficiency of adrenal cortex hormones that can be:
    - Primary adrenal insufficiency - mainly due to autoimmune destruction of the adrenal gland
    - Secondary - adrenal sufficiency due to decreased levels of adrenocorticotropic hormone (ACTH) and corticotropin-releasing hormone (CRH)
- Adrenal crisis: exacerbation of symptoms and associated with absolute hypotension SBP <100 mmHg unresponsive to volume repletion and vasopressors
- Pituitary apoplexy - acute life-threatening infarct or hemorrhage of the pituitary gland, which can precipitate adrenal crisis
    - Presents with acute severe headache (most common and early manifestation)
    - Ocular palsies (most commonly 3rd nerve palsy) can occur due to the involvement of cavernous sinus
    - Reduced visual acuity and visual defects due to optic chiasmal compression
    - Fever, neck stiffness, photophobia, or reduced consciousness similar to signs of subarachnoid hemorrhage or meningitis may occur

Figure 1. Where are these organs located?

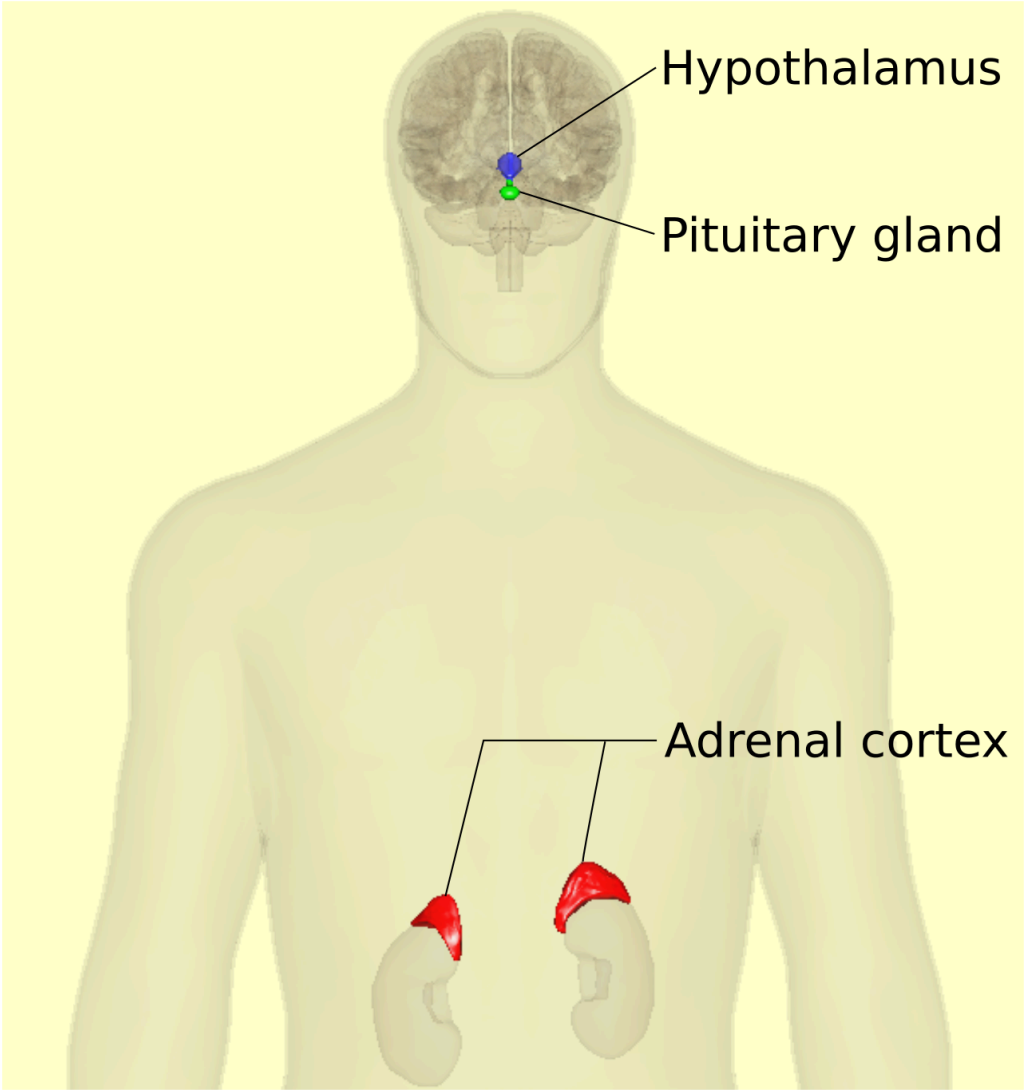

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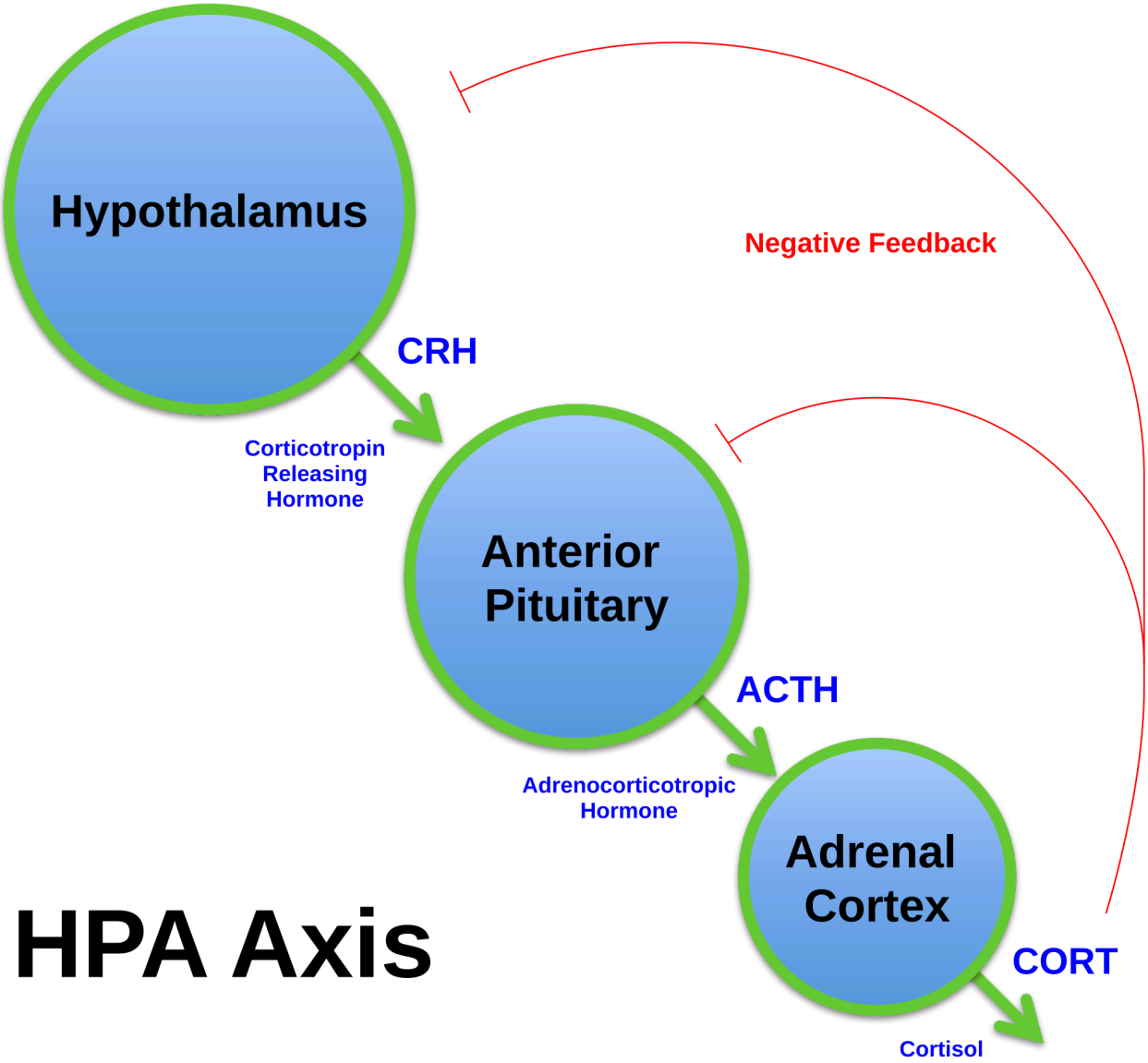
Figure 2. Hypothalamic pituitary axis (HPA).



By ShelleyAdams - This file was derived from: HPA Axis Diagram (Brian M Sweis 2012).png, CC BY-SA 3.0, https://commons.wikimedia.org/w/index.php?curid=53298307

Common Causes/Risk Factors

- Autoimmune destruction of the adrenal gland
- Infections (i.e. TB, HIV, syphilis)
- Drugs ketoconazole fluconazole, etomidate
- Adrenal and pituitary hemorrhage
- Tumors of the pituitary and adrenal[1]

Epidemiology

- Primary adrenal insufficiency (Addison's disease)
    - Incidence = 4-144 cases/million

    - Females at higher risk than males
    - Most commonly occurs in the 30-69 year age range
    - Twice the mortality rate of the general population!
- Secondary adrenal insufficiency
    - More common than Addison's because so many people are on chronic steroids
- 6-8% of chronic adrenal insufficiency patients will have a crisis each year
    - The mortality rate for adrenal crisis is 6%[2]

Risk Factors

Table 1. Risk factors for adrenal insufficiency/crisis and precipitating factors for adrenal crisis.[2]

Risk factors for adrenal insufficiency	Risk factors for adrenal crisis	Precipitating factors for adrenal crisis
● Anticoagulants → adrenal hemorrhage ● History of adrenal insufficiency ● Ongoing glucocorticoid therapy	● Asthma ● Autoimmune polyglandular disease ● Cardiac disease ● Diabetes mellitus ● Older age ● Previous adrenal crisis	● Abrupt cessation of chronic corticosteroids ● GI illnesses ● Infections ● Significant emotional distress ● Thyrotoxicosis ● Trauma

GI = gastrointestinal

Clinical features

Table 2. Clinical features for adrenal insufficiency/crisis.

Adrenal insufficiency	Incipient adrenal crisis	Adrenal crisis
● Abdominal pain ● Amenorrhea ● Anorexia ● Diarrhea/constipation ● Dizziness on standing ● Fatigue ● General malaise ● Hyperpigmentation ● Loss of appetite ● Myalgias/arthralgias ● Nausea/vomiting ● Postural dizziness ● Syncope	● Abdominal pain ● Anorexia ● Delirium ● Fatigue ● GI symptoms (i.e. nausea and vomiting) ● Hyperthermia ● Postural dizziness ● Syncope	● Other signs and symptoms of adrenal insufficiency PLUS signs and symptoms of shock*

• Vitiligo thinning axillary and pubic hair • Weight loss		

*Signs and symptoms of shock include: weak pulse, clammy skin, delayed capillary refill, altered mental status, hypotension, tachycardia

Diagnostics

- Random blood sugar - hypoglycemia
- Complete blood count (CBC) - eosinophilia, lymphocytosis, neutropenia
- Complete metabolic panel (CMP) - hyponatremia, hyperkalemia, increased urea
- Arterial blood gas (ABG) - metabolic acidosis
- ACTH levels - high in primary adrenal insufficiency, low or normal in secondary adrenal insufficiency,
- Plasma cortisol levels - serum cortisol < 5 mcg/dL is diagnostic
- The pituitary is responsible for making a LOT of other hormones; thus, patients can develop diabetes insipidus and syndrome of inappropriate ADH (SIADH). Consider testing urine electrolytes if you are worried about these[3]

Treatment

- ABCDE
    - Airway
        - Shock/poor perfusion may lead to altered mentation and subsequent airway compromise
        - Patients with pituitary apoplexy may have cerebral edema or intracranial hemorrhage → airway loss
        - Intubate if needed (but *resuscitate before you intubate*)
    - Breathing
        - Metabolic acidosis may lead to hyperpnea/Kussmaul respirations
        - Support breathing with oxygen therapy if there is hypoxia
        - Treat any underlying pneumonia that might have triggered an adrenal crisis
    - Circulation
        - Provide intravenous (IV) fluids followed by vasopressors for signs and symptoms of shock
        - Administer stress-dose steroids - hydrocortisone 100 mg IV followed by either 100 mg every 8 hours or 50 mg IV every 6 hours[2,3]
            - Child 0-3 years: 25 mg IV
            - Child 3-12 years: 50 mg IV[1]
    - Disability
        - Check blood glucose and administer supplemental dextrose if needed for hypoglycemia
        - Closely monitor Glasgow Coma Scale (GCS)

        - Complete a full neurologic examination, including cranial nerves, for patients with suspected pituitary apoplexy; they may have cranial nerve deficits[3]
        - Temperature regulation may be altered in patients with pituitary and adrenal conditions, so provide rewarming for those who are hypothermic and external cooling for those who are hyperthermic
    - Exposure - evaluate for any signs of trauma on exam that could be contributing to shock (i.e. are they actually bleeding somewhere and presenting with hemorrhagic shock? Or did some sort of trauma precipitate their adrenal crisis?)
- Adrenal crisis
    - Correct volume depletion
        - IV D5 30 ml/kg is preferred
        - Use caution with normal saline in patients with sodium <125 mEq/L; target a sodium increase of no > 10 mmol/day (to avoid osmotic demyelination syndrome)
    - Replace glucocorticoids (i.e. “stress-dose steroids). Continue IV hydrocortisone until the patient is able to start oral therapy
    - Correct other metabolic abnormalities
    - Start antibiotics +/- antimalarials, as it is difficult to exclude septic shock or severe malaria initially. These medications can be discontinued once these conditions are ruled out
    - Consult endocrinology
- Pituitary apoplexy
    - Consult neurosurgery or transfer for neurosurgery evaluation if pituitary apoplexy is diagnosed
    - Consult endocrinology
    - Consult ophthalmology if there are visual effects
    - Correct electrolyte derangements
    - Control headache with paracetamol; avoid over-sedating with opioids if possible[3]

Prognosis

- Patients with pituitary apoplexy may require surgical decompression, but there is no difference in mortality between surgically vs. medically managed patients[3]
    - *The major cause of morbidity and mortality in pituitary apoplexy patients is adrenal insufficency!*

Disposition

- Patients with adrenal insufficiency or pituitary apoplexy should be admitted (likely to an intensive care unit, where available)

Collaborating Partners

1. Ministry of Health of the Republic of Uganda
2. Seed Global Health

3. Techies Without Borders

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