

Erdheim-Chester and Hormones

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ENDOCRINE NEOPLASIA & HORMONAL DISORDERS

ECD GROUP 10-10-15



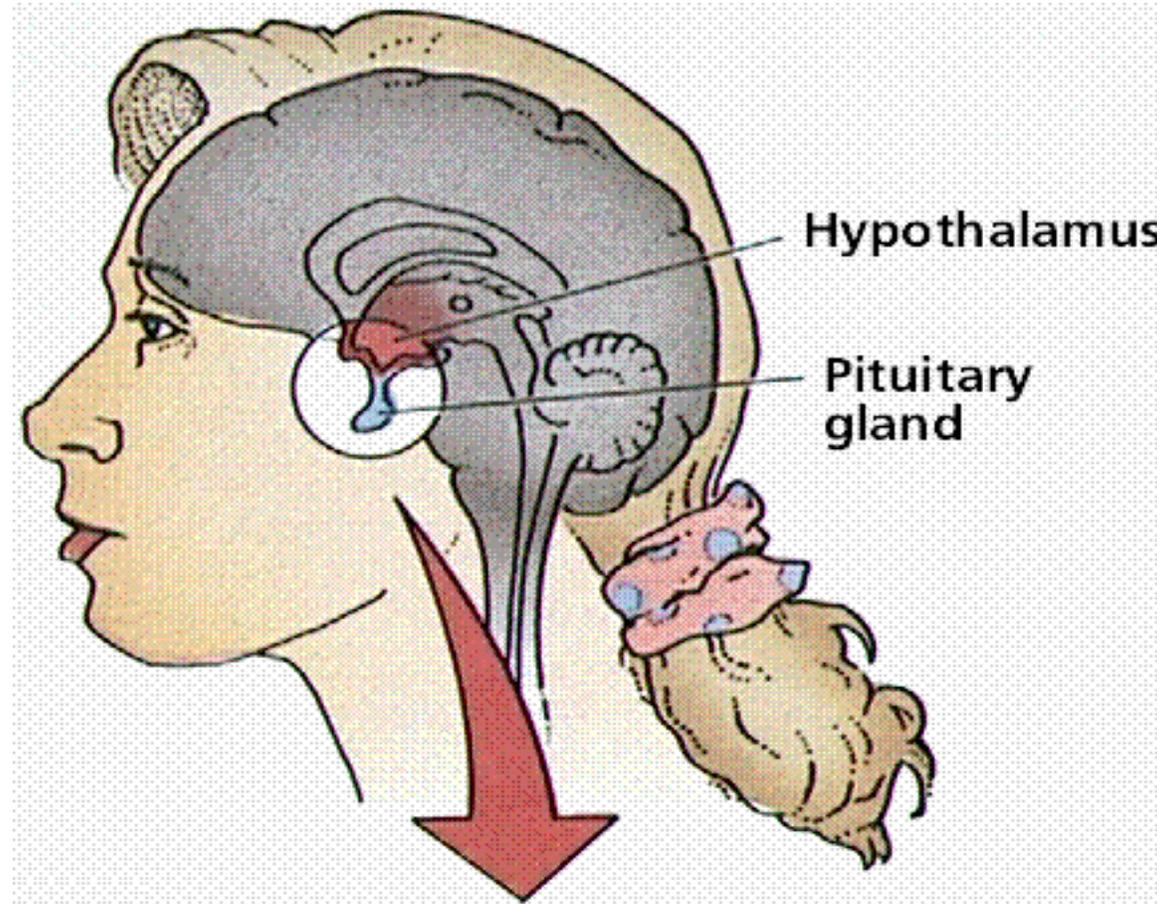
Glands affected by Erdheim-Chester

Pituitary (22%)

Diabetes insipidus (22%)

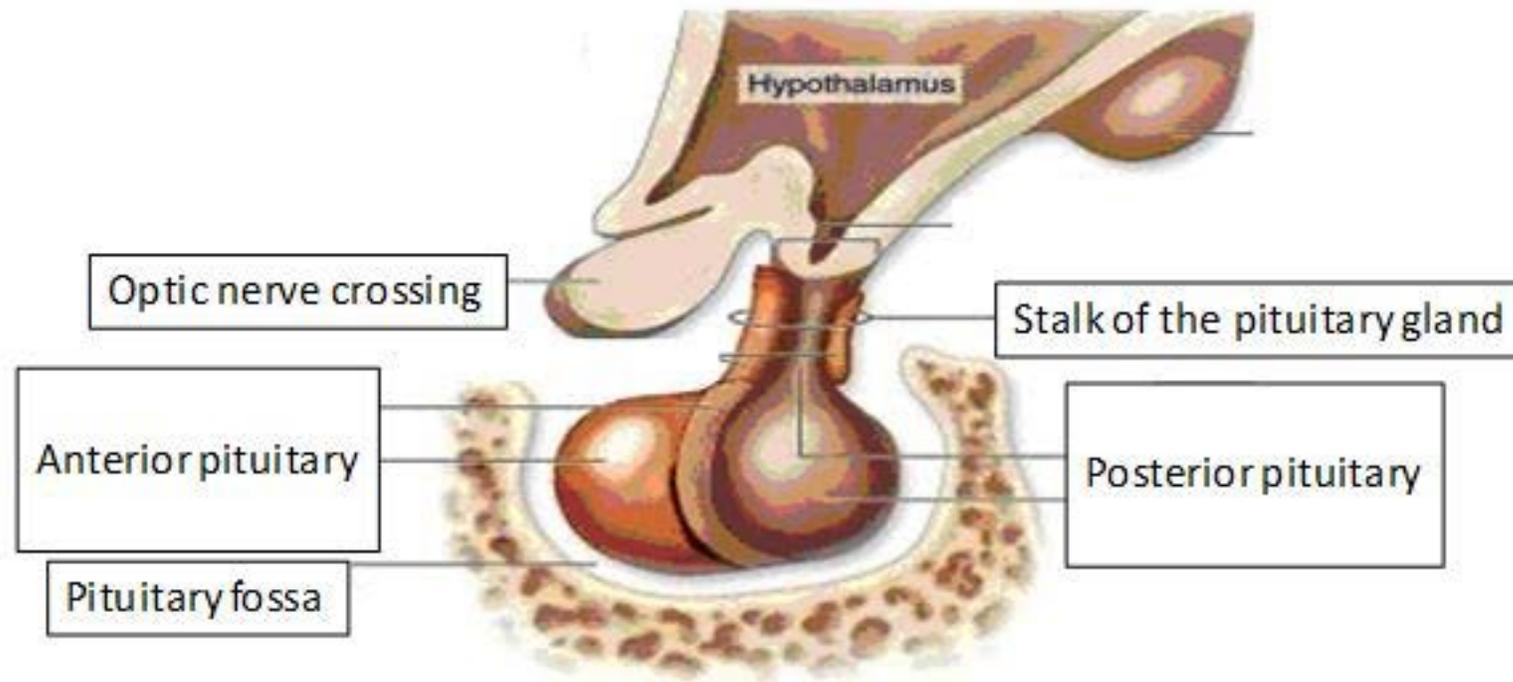
Bones (95%)

Pituitary Gland



Pituitary Gland

Anatomy of Pituitary



Diabetes Insipidus

Lack of hormone vasopressin (DDAVP)

Pituitary stops making it due to ECD

Symptoms:

- Extra Thirsty (gallons)
- Excessive Urination

Diabetes Insipidus- Diagnosis

- Urine electrolytes
- Urine osmolality
- Serum electrolytes
- Other pituitary hormones (thyroid, sex, growth and adrenocortical)
- Usually persists despite medical therapy for ECD

Diabetes Insipidus- Treatment

Treatment goal:

- to keep pt from staying in the bathroom urinating
- Sleep well

Drink fluids - water

DDAVP

- Intranasal
- Tablets
- Subcutaneous (in skin) or IV (vein) [hospital]

Call MD office if excessive thirst or urination

Pituitary Hormones- “Master” Gland

Controls other target organs

Anterior Pituitary makes 6 hormones:

Thyroid Stimulating Hormone (TSH)

Adrenocorticotrophic hormone (ACTH)

Luteinizing hormone (LH)

Follicle stimulating hormone (FSH)

Growth hormone

Prolactin

ACTH deficiency- Adrenal Insufficiency

Controls the cortisol secreted from 2 glands on top of your kidney called adrenals

Secondary adrenal insufficiency – result is lack of cortisol

ACTH deficiency- Symptoms

low blood pressure when stand up / fast heart beat

fatigue

Nausea

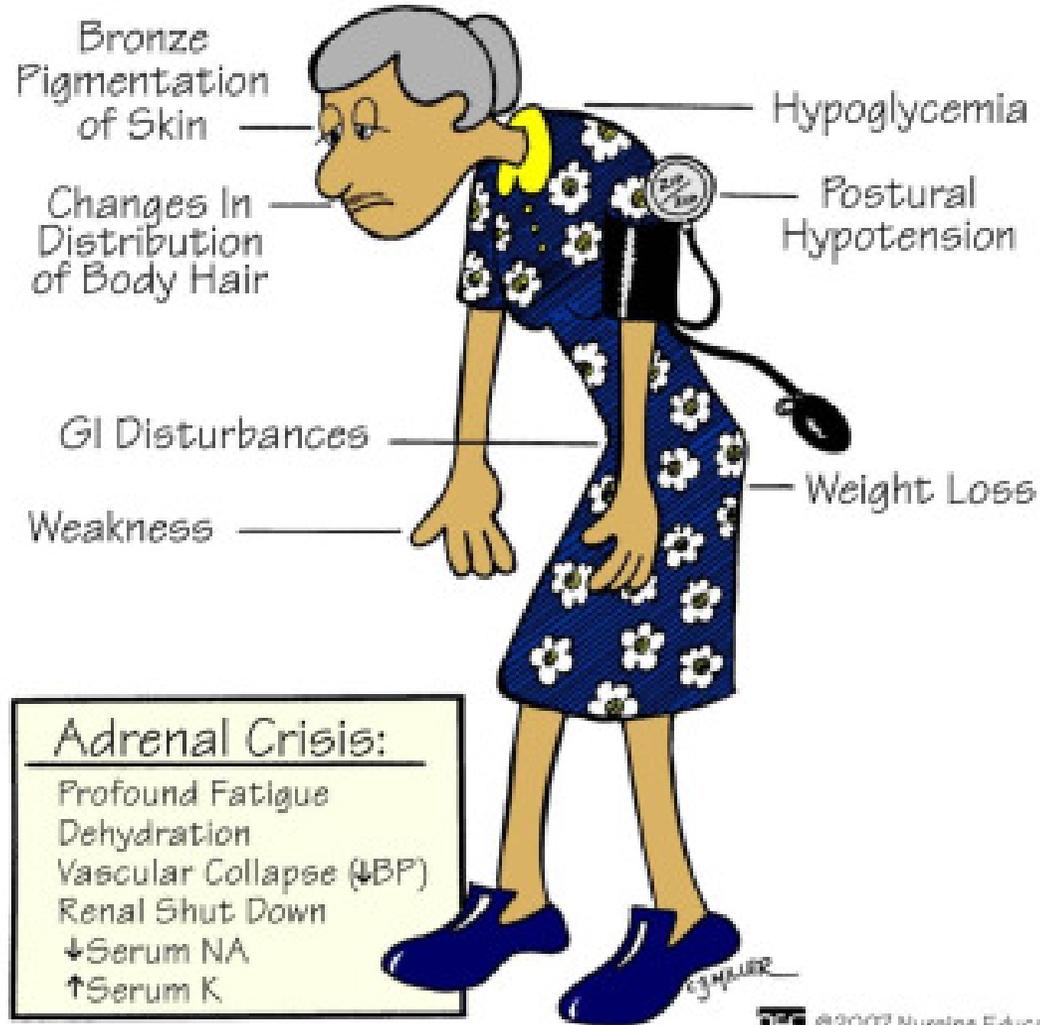
Weight loss

Decreased libido

Hypoglycemia

eosinophilia

ADDISON'S DISEASE



blah.

Adrenal Insufficiency - Treatment

Replace with steroids

Hydrocortisone

Prednisone

Dexamethasone

TSH deficiency - Hypothyroidism

Fatigue

Cold intolerance

Decreased appetite

Constipation

Facial puffiness

Dry skin

Slow heart beat

Low blood counts

HYPOTHYROIDISM



Hypothyroidism - Treatment

Replace with thyroid hormone (T4)

Name brand or generic

- levothyroxine

Gonadotropin deficiency - Hypogonadism

Low FSH / low LH – secondary hypogonadism

Women:

Low estrogen –

- menopausal – irregular periods or none; infertility; vaginal dryness; hot flashes

Men:

testicular hypofunction;

infertility and low testosterone

Decreased energy; libido; hot flashes

Treatment of Hypogonadism

Women:

Replace with Estrogen +/- Progesterone

Men:

Testosterone

Growth Hormone Deficiency

Children – short stature

Adults

- Body composition changes: increased fat mass with a decrease in lean body mass
- Decreased bone mineral density

Prolactin deficiency

Prolactin is for lactation

Inability to lactate

No consequence otherwise

Treatment of the Hormone Deficiencies

Adrenal Insufficiency

- Steroids – hydrocortisone, prednisone or dexamethasone

Hypothyroidism

- Thyroid hormone - levothyroxine

Gonadal Hormone Replacement

- Estrogen/Progesterone or Testosterone

Prolactin is not replaced

Growth Hormone depends on state of ECD, but often not replaced

Thank you!

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