

# Erdheim-Chester and Hormones

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ENDOCRINE NEOPLASIA & HORMONAL DISORDERS

ECD GROUP 10-10-15



# Glands affected by Erdheim-Chester

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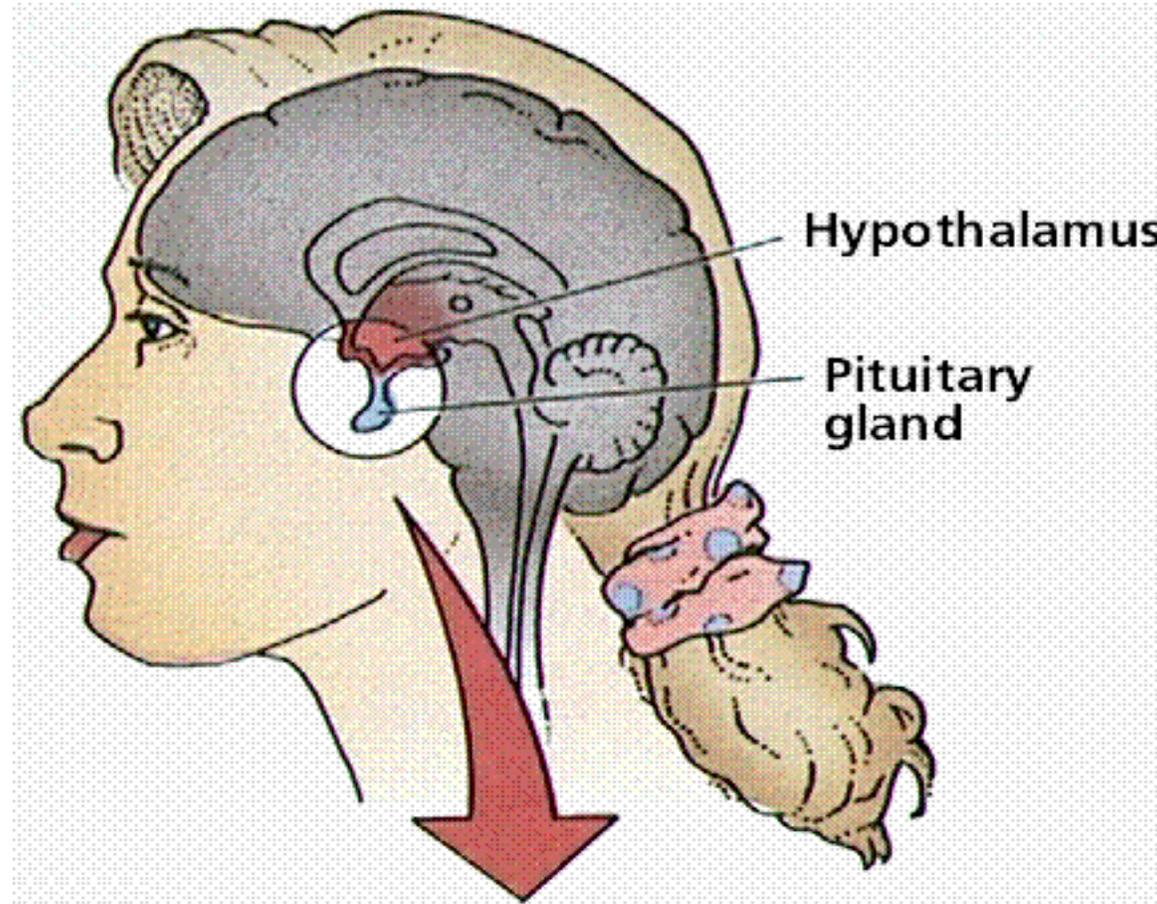
Pituitary (22%)

Diabetes insipidus (22%)

Bones (95%)

# Pituitary Gland

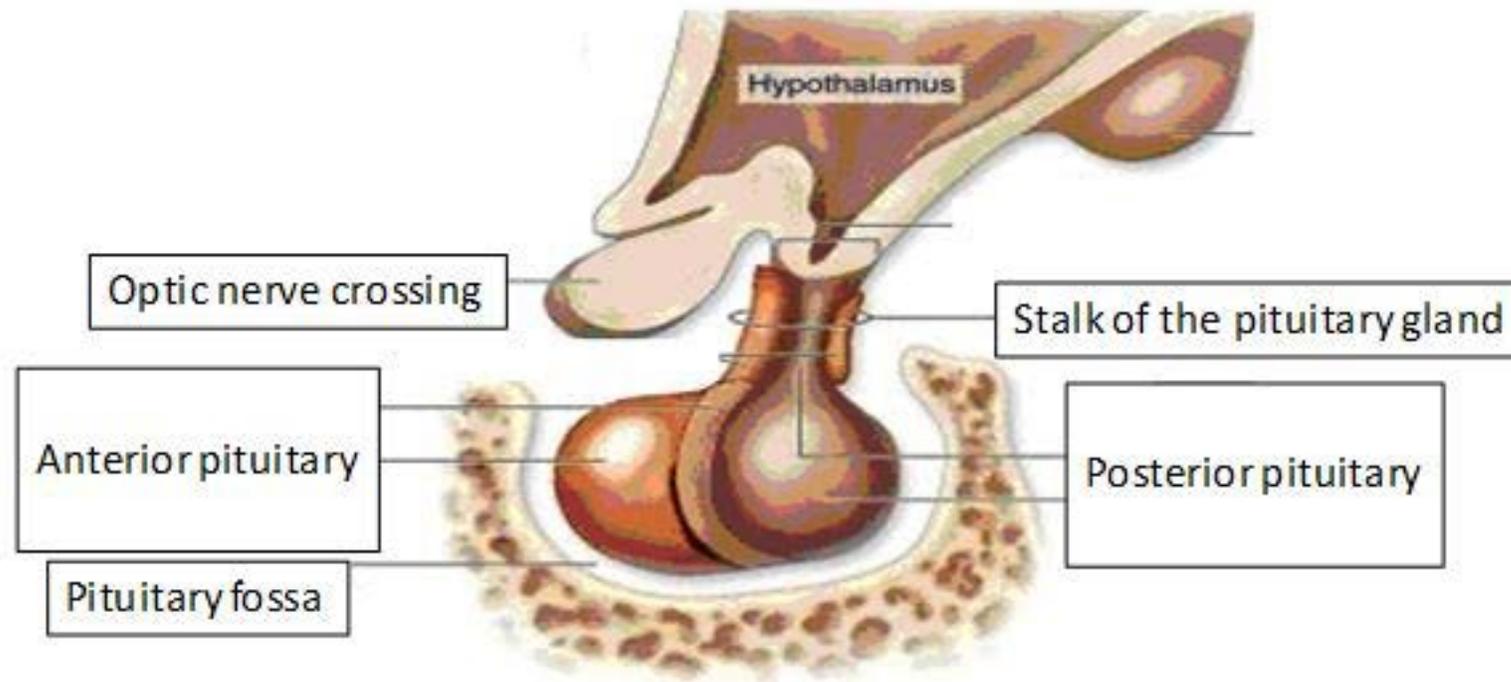
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# Pituitary Gland

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Anatomy of Pituitary



# Diabetes Insipidus

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Lack of hormone vasopressin (DDAVP)

Pituitary stops making it due to ECD

Symptoms:

- Extra Thirsty (gallons)
- Excessive Urination

# Diabetes Insipidus- Diagnosis

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

# Diabetes Insipidus- Treatment

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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

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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

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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

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low blood pressure when stand up / fast heart beat

fatigue

Nausea

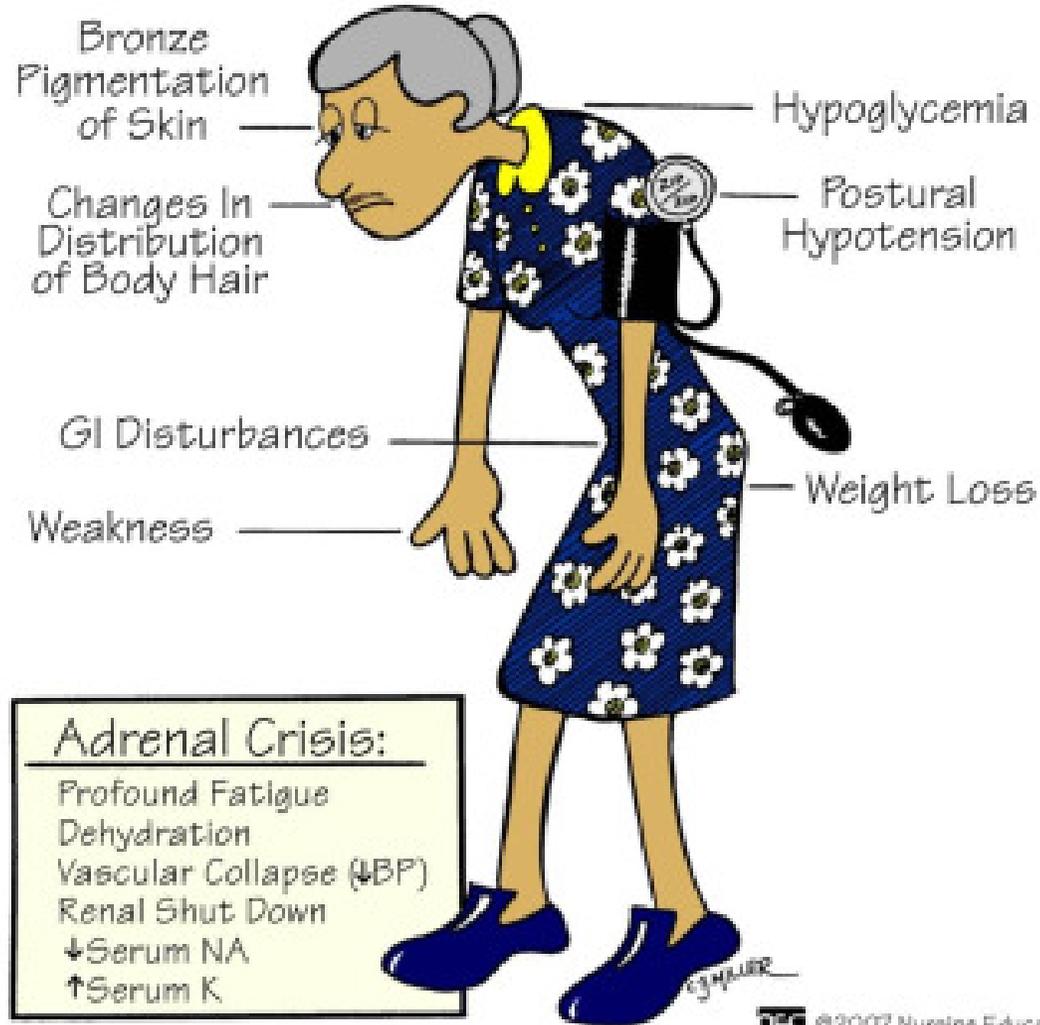
Weight loss

Decreased libido

Hypoglycemia

eosinophilia

# ADDISON'S DISEASE



**Adrenal Crisis:**  
Profound Fatigue  
Dehydration  
Vascular Collapse (↓BP)  
Renal Shut Down  
↓Serum NA  
↑Serum K

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blah.

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# Adrenal Insufficiency - Treatment

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Replace with steroids

Hydrocortisone

Prednisone

Dexamethasone

# TSH deficiency - Hypothyroidism

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Fatigue

Cold intolerance

Decreased appetite

Constipation

Facial puffiness

Dry skin

Slow heart beat

Low blood counts

# HYPOTHYROIDISM



# Hypothyroidism - Treatment

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Replace with thyroid hormone (T4)

Name brand or generic

- levothyroxine

# Gonadotropin deficiency - Hypogonadism

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**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

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## Women:

Replace with Estrogen +/- Progesterone

## Men:

Testosterone

# Growth Hormone Deficiency

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Children – short stature

## Adults

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

# Prolactin deficiency

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Prolactin is for lactation

Inability to lactate

No consequence otherwise

# Treatment of the Hormone Deficiencies

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## 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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