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

Hypothalamus

Pituitary gland
Pituitary Gland

Anatomy of Pituitary

- Optic nerve crossing
- Stalk of the pituitary gland
- Anterior pituitary
- Posterior pituitary
- Pituitary fossa
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

- Bronze Pigmentation of Skin
- Hypoglycemia
- Changes in Distribution of Body Hair
- Postural Hypotension
- GI Disturbances
- Weight Loss
- Weakness

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

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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

Intolerance to Cold
Receding Hairline
Facial & Eyelid Edema
Dull-Blank Expression
Extreme Fatigue
Thick Tongue - Slow Speech
Anorexia
Brittle Nails & Hair

Late Clinical Manifestations
- Subnormal Temp
- Bradycardia
- Weight Gain
- ↓ LOC
- Thickened Skin
- Cardiovascular Complications

Hair Loss
Apathy
Lethargy
Dry Skin (Coarse & Scaly)
Muscle Aches & Weakness
Constipation
Menstrual Disturbances
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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