

ENDOCRINE MANIFESTATIONS IN ERDHEIM CHESTER DISEASE

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Pituitary gland



- ▶ The **anterior pituitary** regulates other glands in the body and controls most of the hormonal secretions:
 - ▶ **Thyroid**
 - ▶ **Adrenals**
 - ▶ **Ovaries, testicles**
 - ▶ **Breasts**
 - ▶ **Growth**
- ▶ The **posterior pituitary** produces the **anti-diuretic hormone**
 - ▶ A lack in this hormone induces enhanced diuresis which in turn drives to an important thirst and a need to drink in order to maintain normal hydration
 - ▶ This deficit is called **diabetes insipidus**



Thyroid gland



- ▶ The **thyroid gland** secretes hormones that regulate many **metabolic processes**, including **growth** and **energy expenditure**



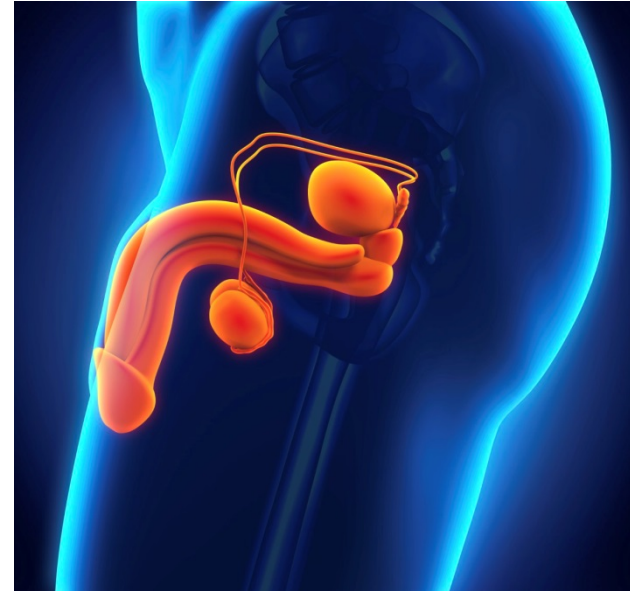
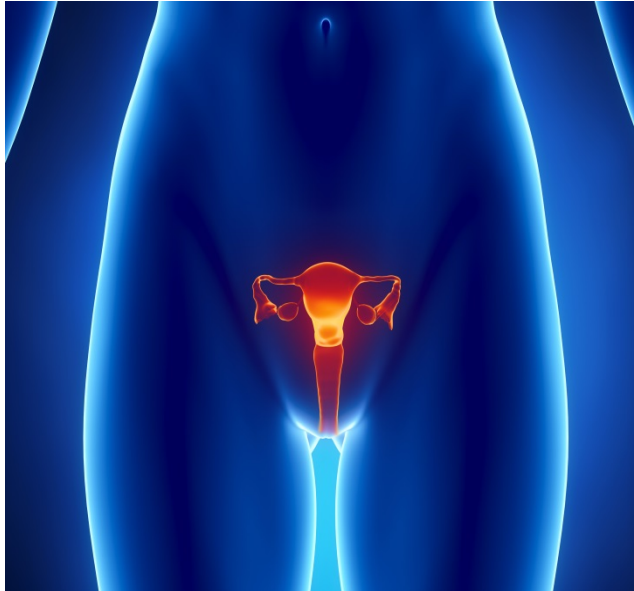
Adrenal glands



- ▶ **Adrenal glands produce several major hormones** important for
 - ▶ coping with **physical stresses** to body
 - ▶ maintaining adequate:
 - ▶ **blood pressure**
 - ▶ **blood volume**
 - ▶ **salt retention**
- ▶ These hormones are
 - ▶ **Cortisol** and **Aldosterone** which regulate blood pressure, salt retention and general well being
 - ▶ Mildly potent **male hormones**
 - ▶ **Adrenaline** and **noradrenaline**



Gonads = ovaries / testicles



- ▶ **Gonads** have 2 functions
 - ▶ Secretion of the **sexual hormones**: **estradiol** and **progesterone** in women, **testosterone** in men
 - ▶ Production of **gametes** to ensure **reproduction**: **oocytes** in women and **spermatozooids** in men



ECD and endocrine manifestations

- ▶ **All the glands** can be infiltrated by the histiocytosis
 - ▶ Pituitary (24%)
 - ▶ Testicles (29%)
 - ▶ Adrenals (39%)
 - ▶ Thyroid
 - ▶ Breast

- ▶ **Hormonal dysfunctions are very frequent and have important implications**
 - ▶ Diabetes insipidus
 - ▶ Fatigue, headrush
 - ▶ Excess body weight
 - ▶ Low muscular strength
 - ▶ Impotency, infertility
 - ▶ Depression, mood changes
 - ▶ Increase in cardiovascular risk
 - ▶ Bone demineralization



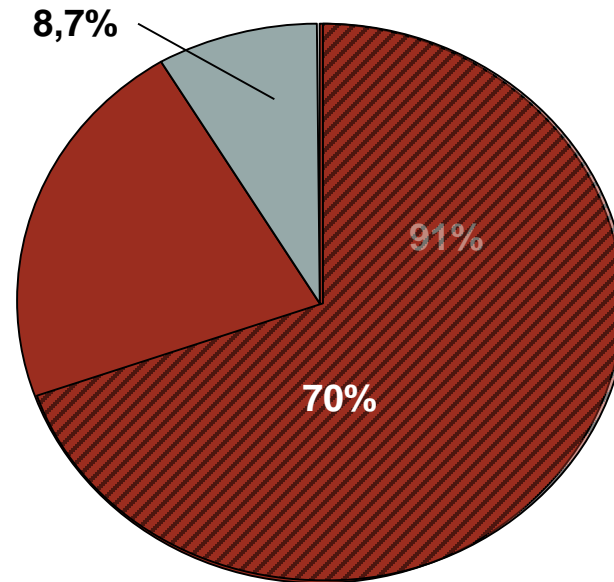
Endocrine manifestations

Hormonal dysfunction	% of patients
Growth hormone deficiency	79%
Testicular deficiency	53%
Diabetes insipidus	33%
Gonadal function deficiency	22%
Thyroid deficiency	20%
Cortisol deficiency	4%
NONE	1%

- ▶ **Diabetes insipidus**
 - ▶ is often one of the first signs of ECD and the first endocrine manifestation
 - ▶ is permanent
- ▶ **New deficits** can appear during follow-up
- ▶ Men can have **alteration in sperm count**, so if there might be a parental project, **conservation of sperm** as soon as possible is mandatory

▶ **Results**

Anterior pituitary deficits



■ ≥ 1 deficit

■ No anterior pituitary dysfunction

■ ≥ 2 deficits



Recommendations

	CLINICAL EVALUATION	MORPHOLOGICAL EVALUATION	HORMONAL EVALUATION
PITUITARY	<p>Search for signs of anterior pituitary deficits</p> <p>24hours diuresis and water intake</p>	Pituitary MRI	Evaluation of anterior and posterior pituitary functions
GONADS	Evaluation of testicular volume and search of palpable testicular nodules	<p>Gonadal sonography</p> <p>If man with testicular infiltration > sperm cryopreservation</p>	Evaluation of gonadal function (ovaries, testicles)
THYROID	Search of a goitre and of nodules	<p>Thyroid sonography</p> <p>if clinical anomalies</p>	Evaluation of thyroid function
ADRENAL	Search of signs of adrenal deficiency	Abdominal or adrenal CT scan	Evaluation of adrenal function
BREAST	Search for lumps † ‡	<p>Mammography +/- mammary sonography</p> <p>if presence of clinical lumps</p>	-
METABOLISM	<p>Blood pressure</p> <p>Electrocardiogram</p>	-	<p>Blood glucose</p> <p>Lipid profile</p>