

Common Adrenal Disorders in Children



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Consultant

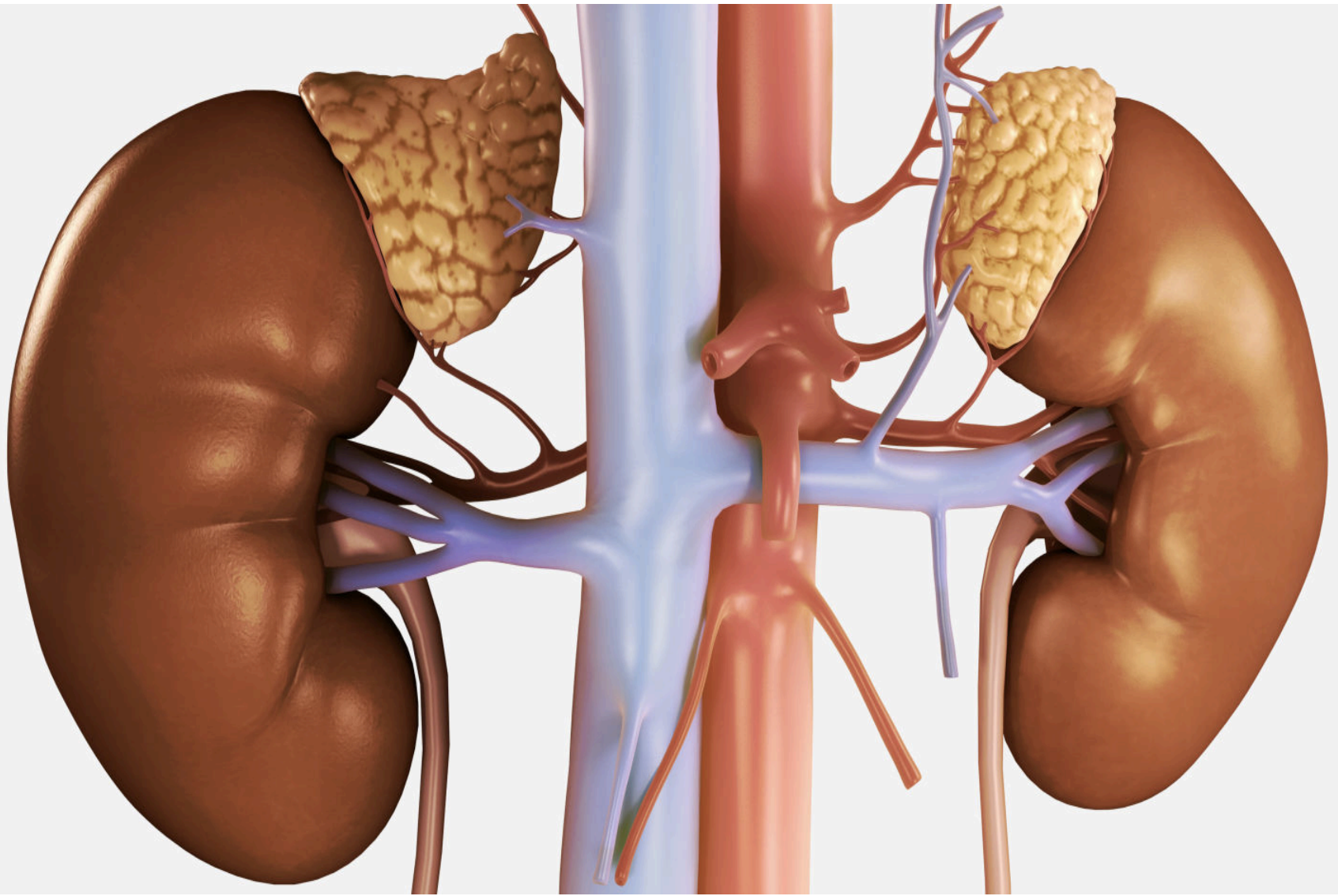
Assistant professor

Pediatric Endocrinology

King Saud University

Objectives

- Understand physiology of adrenal
- Know Causes of adrenal insufficiency
- Cushing Syndrome



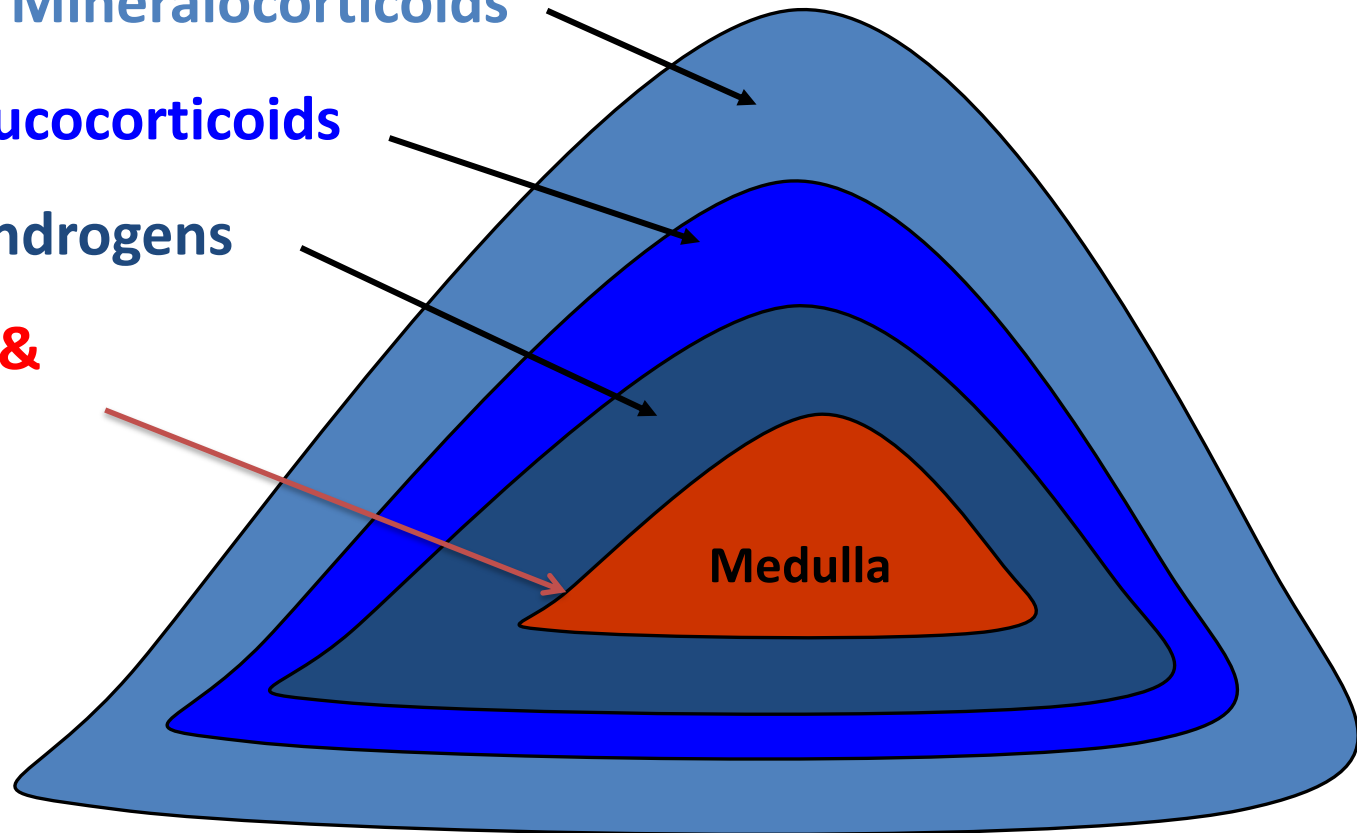
Adrenal Cortex

Zona **G**lomerulosa: Mineralocorticoids

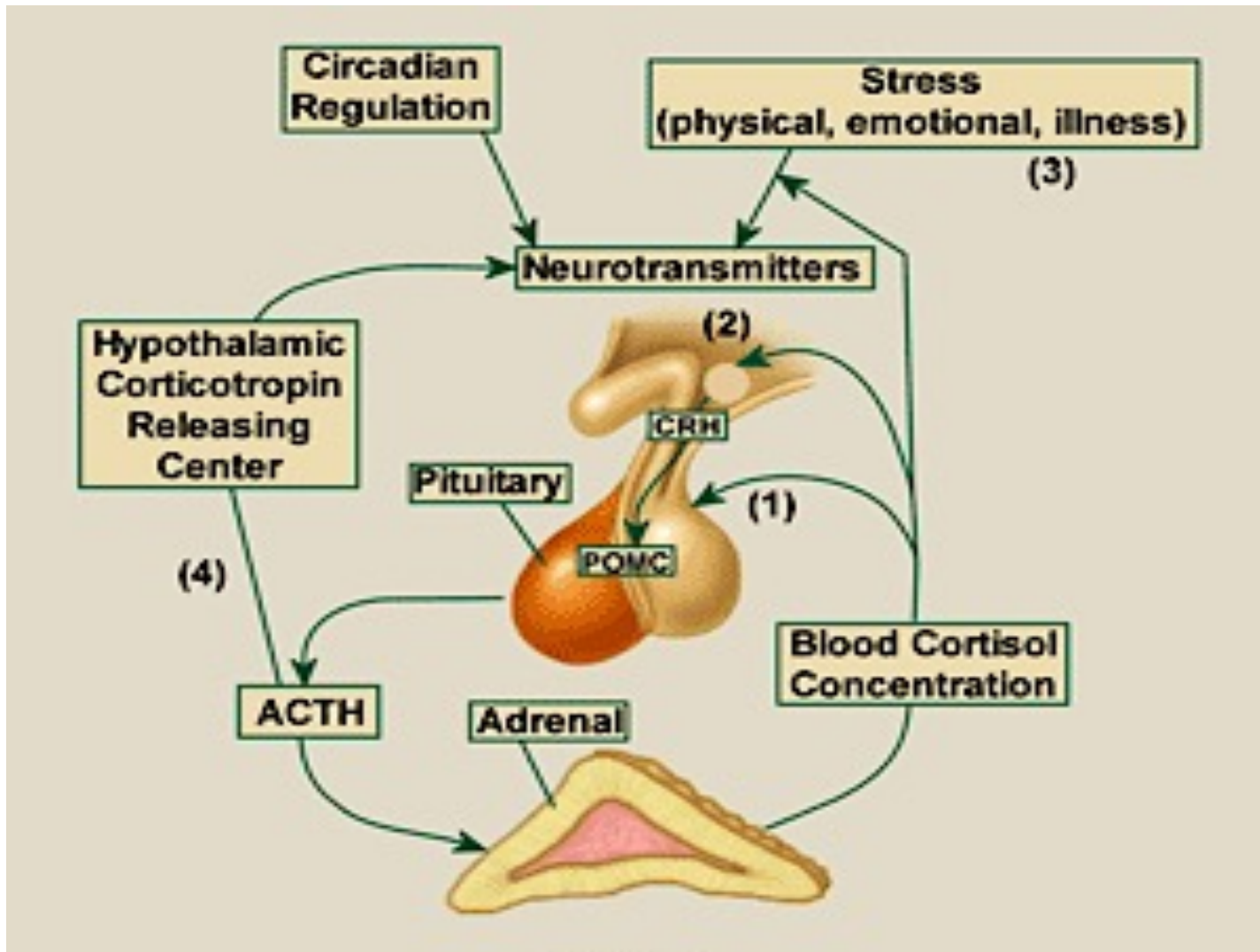
Zona **F**asiculata: Glucocorticoids

Zona **R**eticularis: Androgens

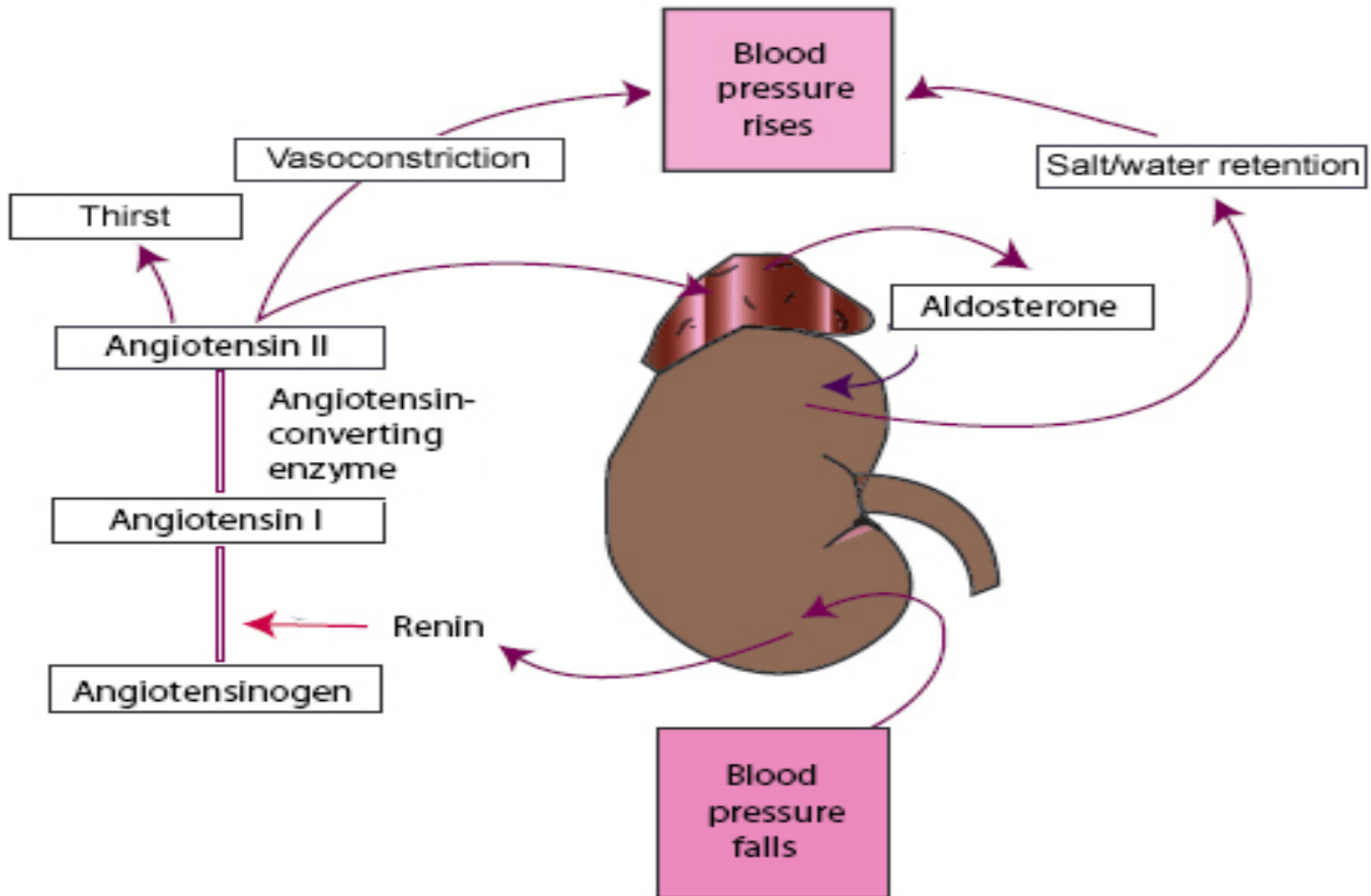
**Medula: adrenalin &
noradrenalin**

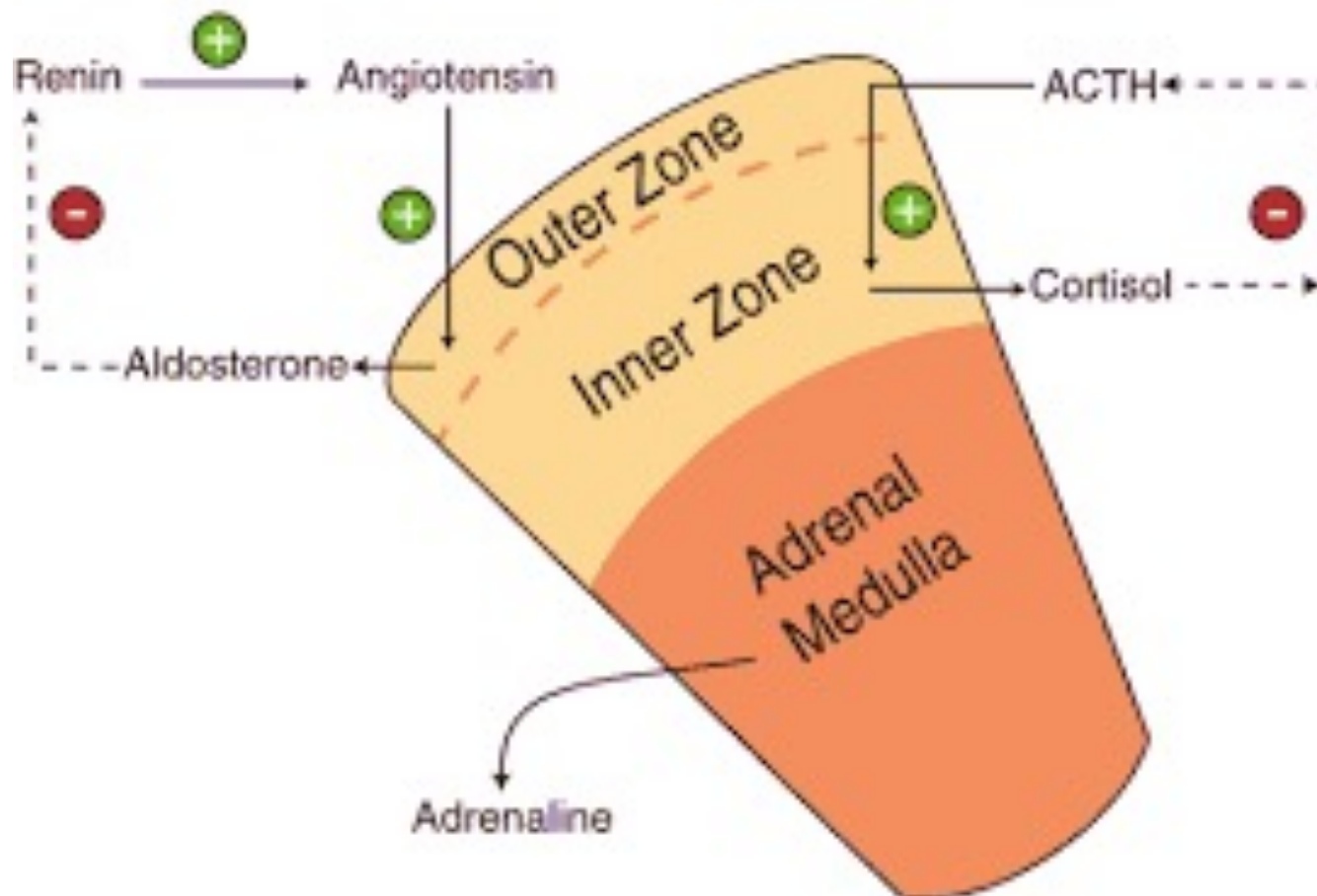


HPA axis

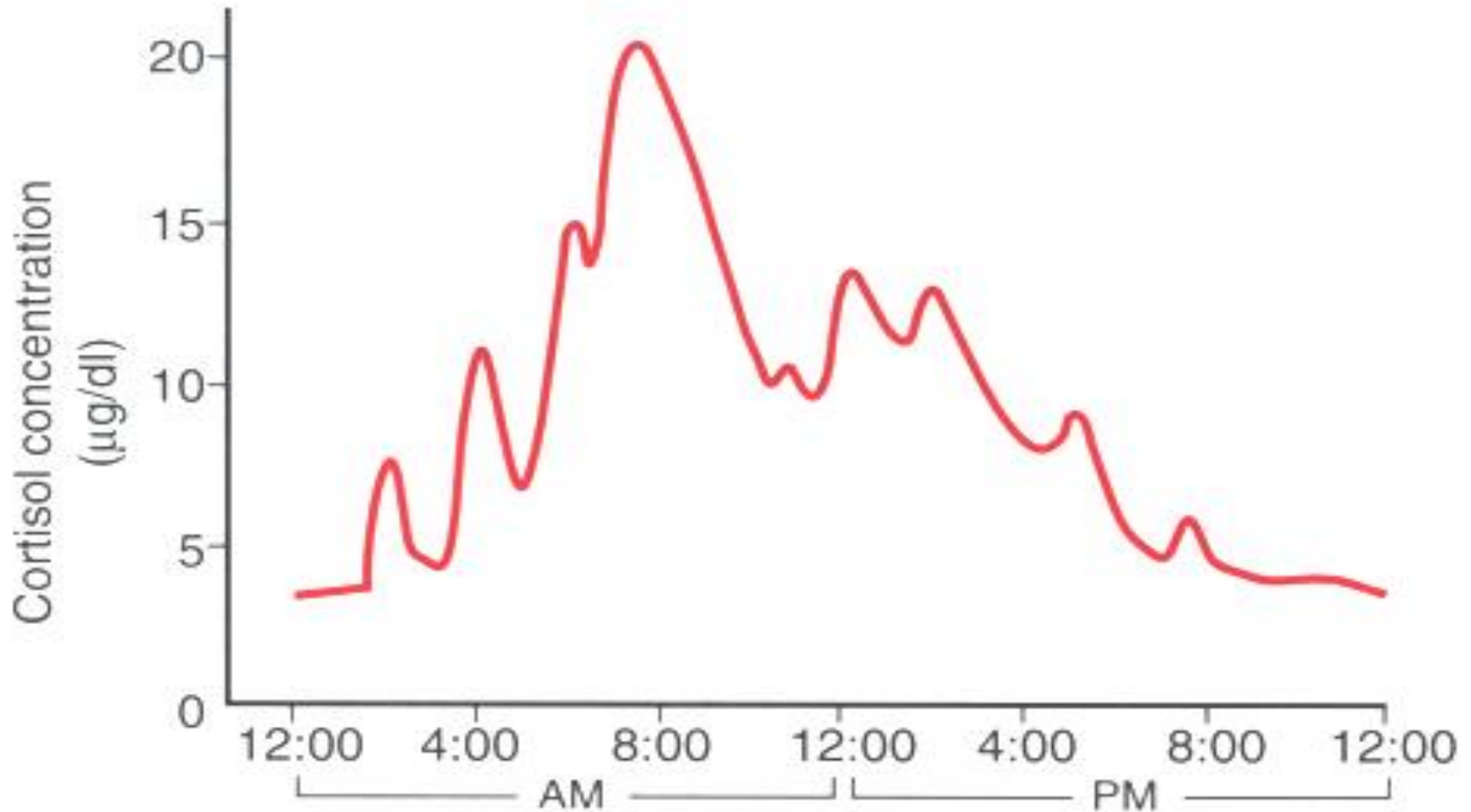


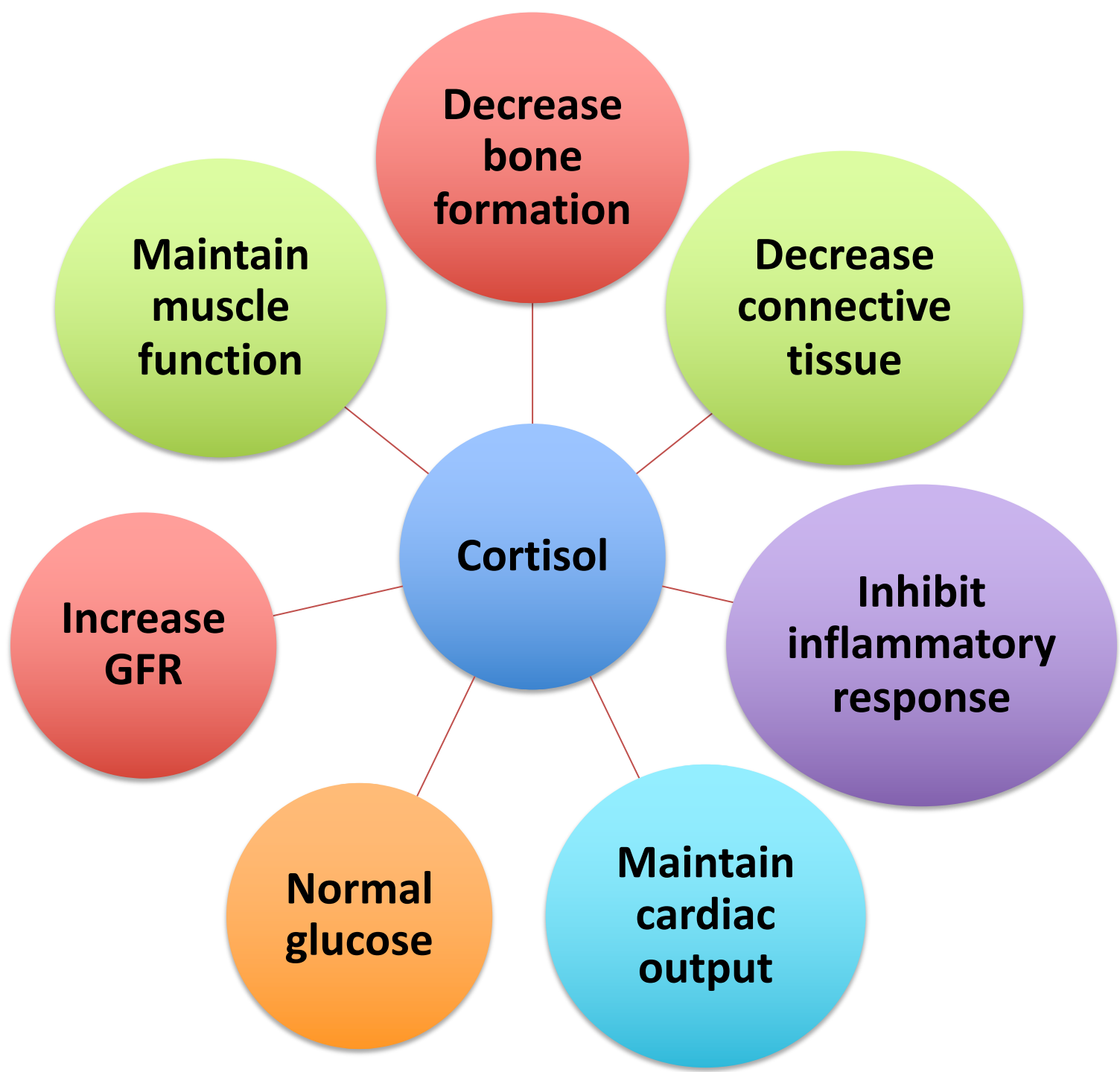
Renin-angiotensin system





Cortisol level during the day





**Decrease
bone
formation**

**Decrease
connective
tissue**

**Inhibit
inflammatory
response**

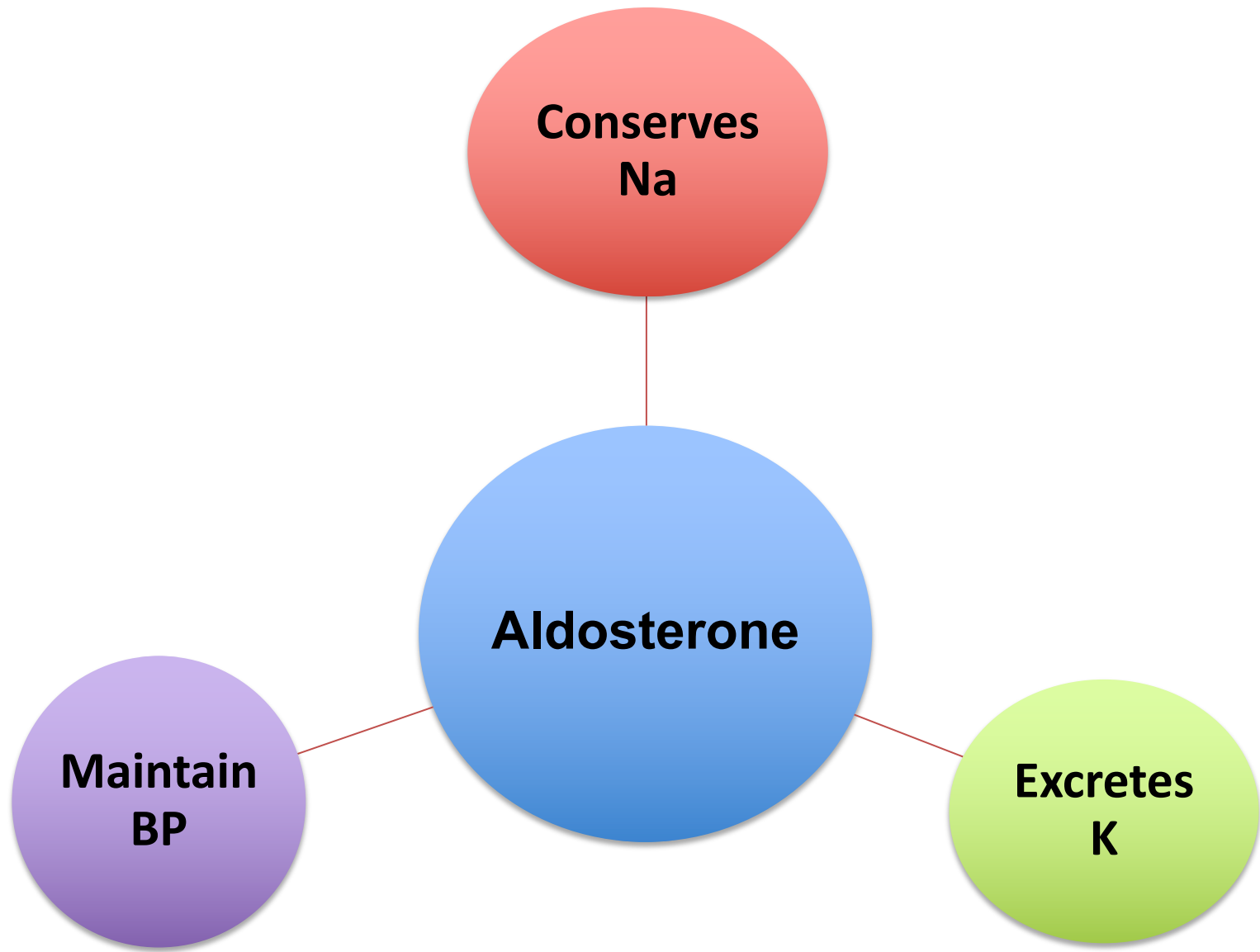
**Maintain
cardiac
output**

**Normal
glucose**

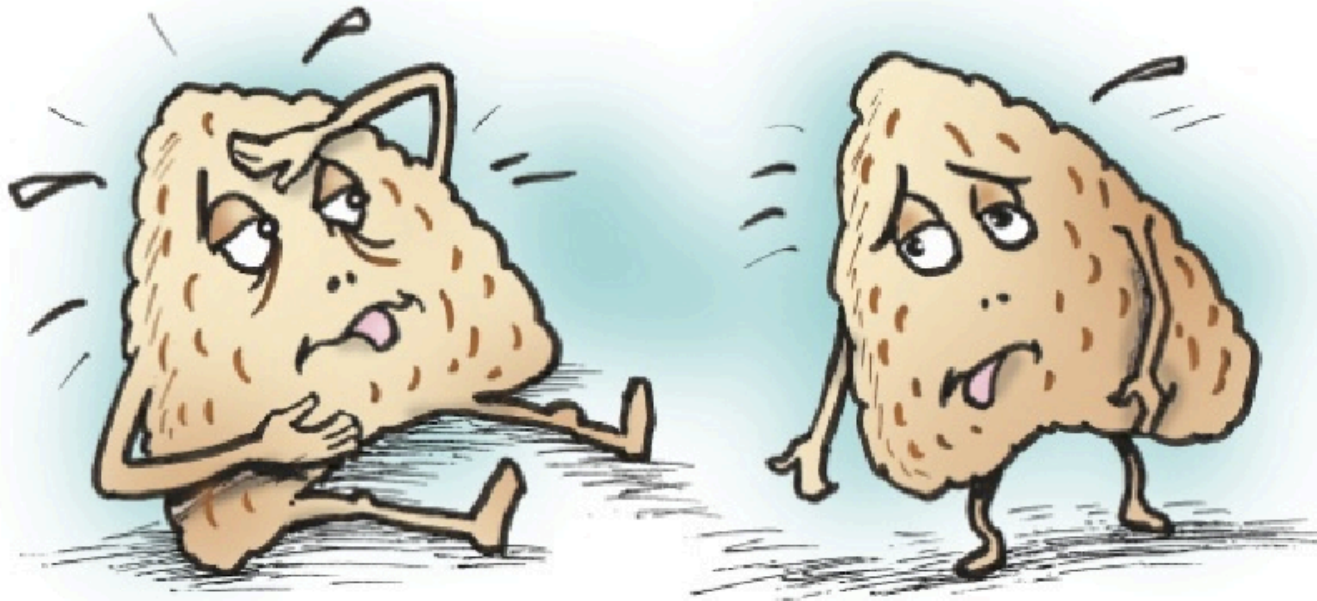
**Increase
GFR**

**Maintain
muscle
function**

Cortisol



Adrenal Dysfunction



Adrenal insufficiency

Primary

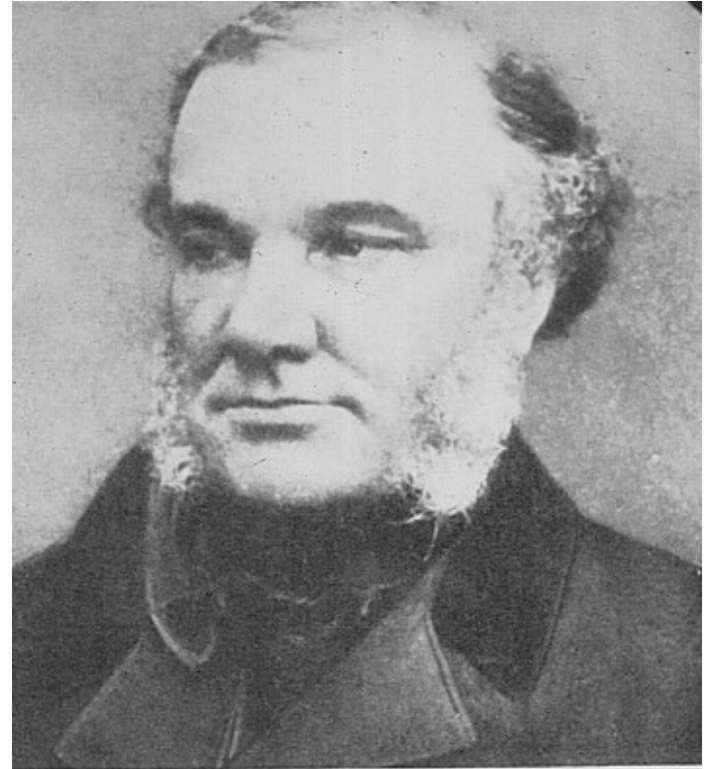
- **Acquired:**
 - Addison disease (autoimmune)
 - Infection (TB, sepsis)
 - Hemorrhage
- **Congenital:**
 - Congenital adrenal hyperplasia
- **Metabolic:**
 - Adrenoleukodystrophy

Secondary

- HPA suppression
- Pituitary:
 - Congenital
 - Tumor
 - Trauma
 - Infection

Primary Adrenal Insufficiency

- 1st described in 1855 by Dr. Thomas Addison
- Refers to acquired primary adrenal insufficiency
 - Usually autoimmune (~80%)



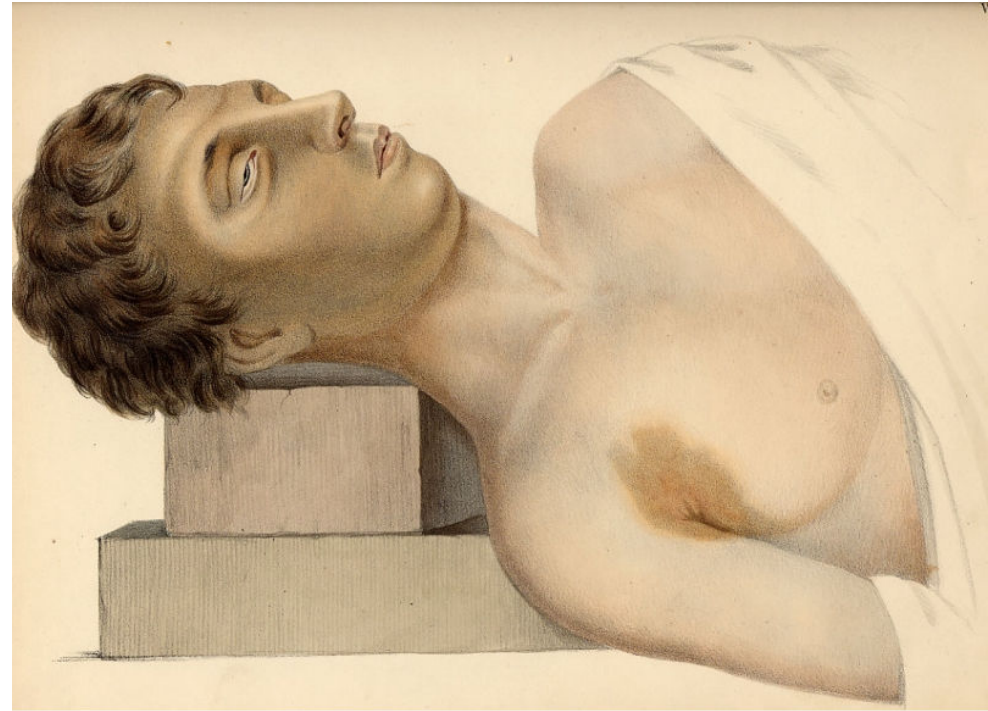
Thomas Addison

Symptoms

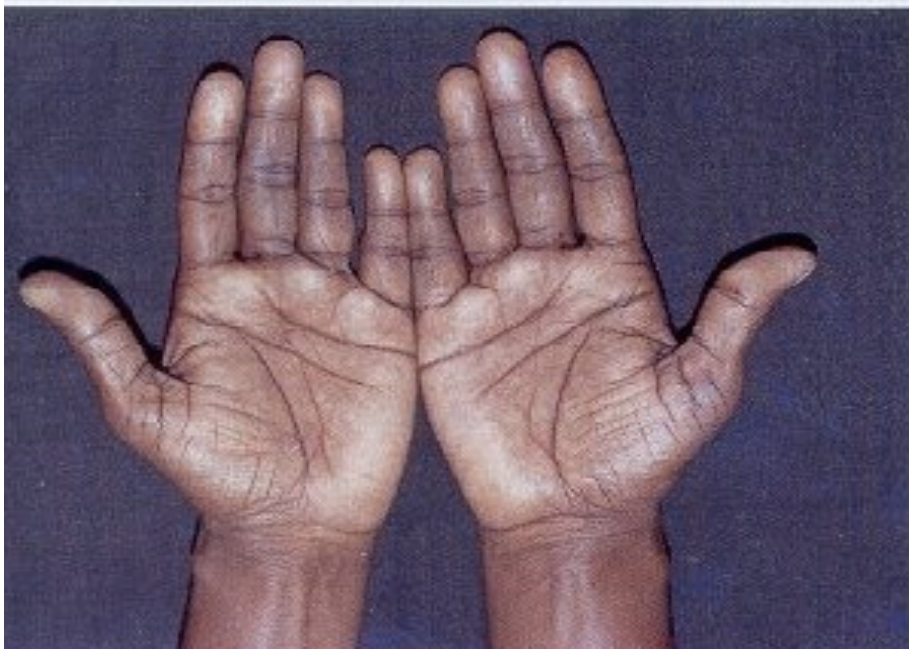
- Fatigue
- Weakness
- Skin & mucous membrane hyperpigmentation
- Weight loss
- Poor appetite
- Nausea, vomiting
- Abdominal pain
- Salt craving

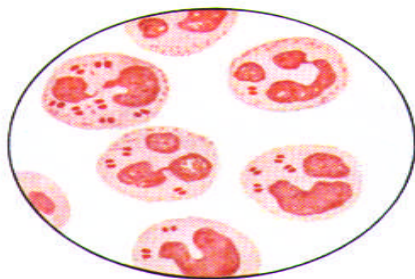
Physical findings

- Hyperpigmentation
- Hypotension
- Dehydration
- Orthostatic changes
- Weak pulses
- Shock
- Loss of axillary/pubic hair (women)

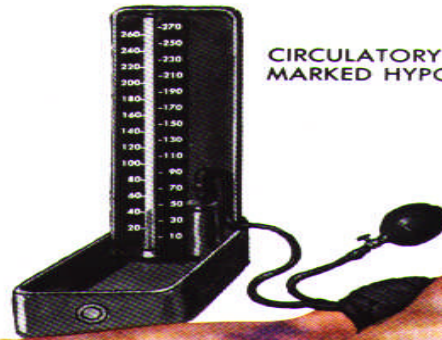


Hardin Library for the Health Sciences. John Martin Rare Book Room

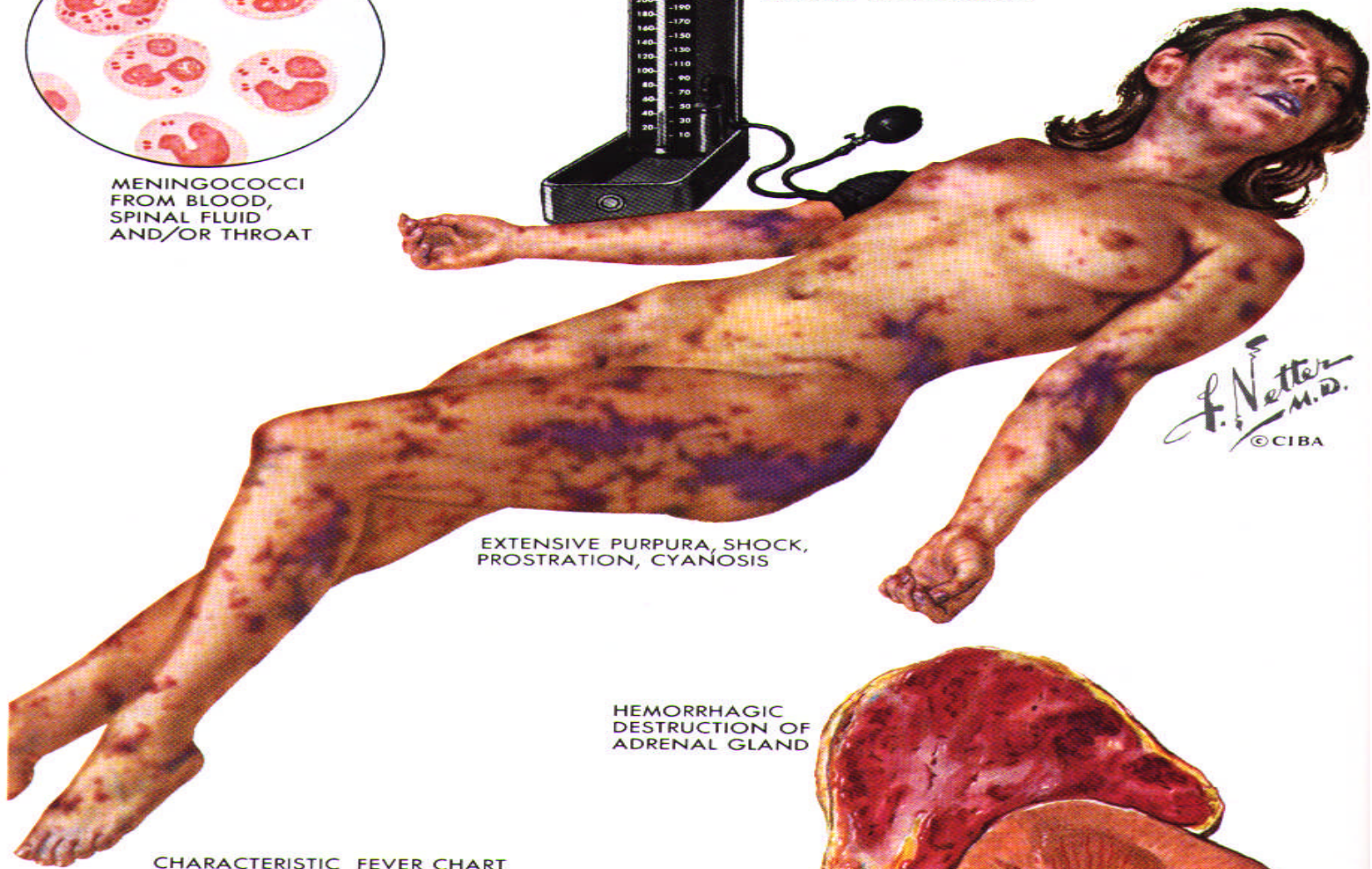




MENINGOCOCCI
FROM BLOOD,
SPINAL FLUID
AND/OR THROAT



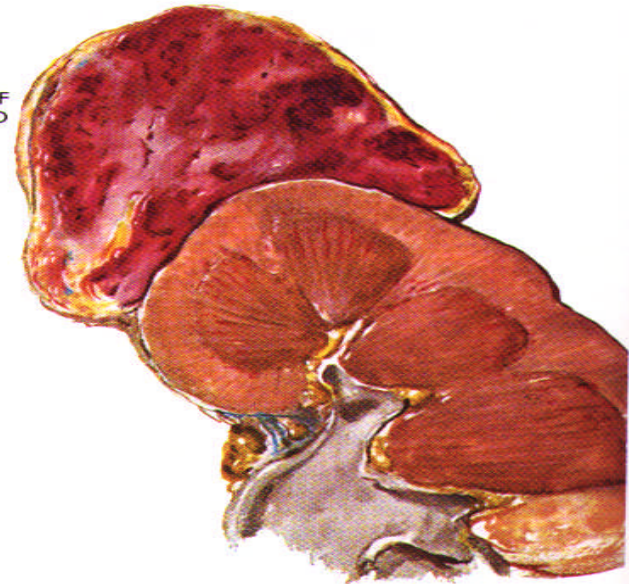
CIRCULATORY COLLAPSE;
MARKED HYPOTENSION



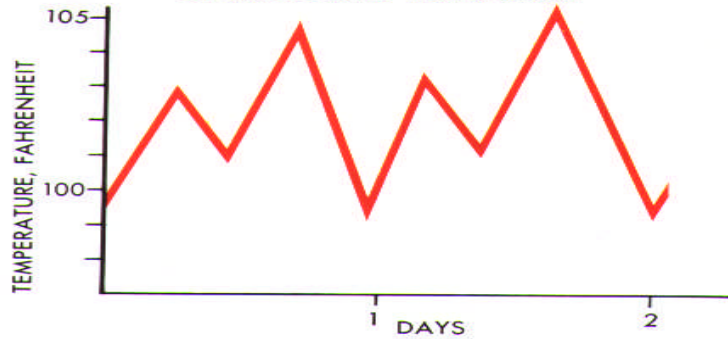
EXTENSIVE PURPURA, SHOCK,
PROSTRATION, CYANOSIS

F. Netter
M.D.
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HEMORRHAGIC
DESTRUCTION OF
ADRENAL GLAND



CHARACTERISTIC FEVER CHART



Laboratory findings

- Na ↓
- K ↑
- Glucose ↓
- Cortisol ↓
- ACTH ↑
- Renin ↑
- Aldosterone ↓

Diagnosis

- Am cortisol, ACTH
- ACTH stimulation test
- Adrenal antibodies

Treatment

- Hydrocortisone
- +/- Fludrocortisone

For life

STRESS MANAGEMENT

Sick day management

fever of 38.5 C - 39.4 C or moderate illness/stress, give a **double** dose

fever > 39.5 C or severe illness, give a **triple** dose

Continue the double or triple doses during the duration of stress.

Intubation & surgeries

- Hydrocortisone 50mg/m² IV

Adrenal crisis

- Life threatening complication
- Severe vomiting and diarrhoea followed by dehydration
- Low BP & shock
- Hypoglycemia
- Loss of consciousness
- Treatment: IV fluids resuscitation +IV hydrocortisone

CONGENITAL ADRENAL HYPERPLASIA

CAH

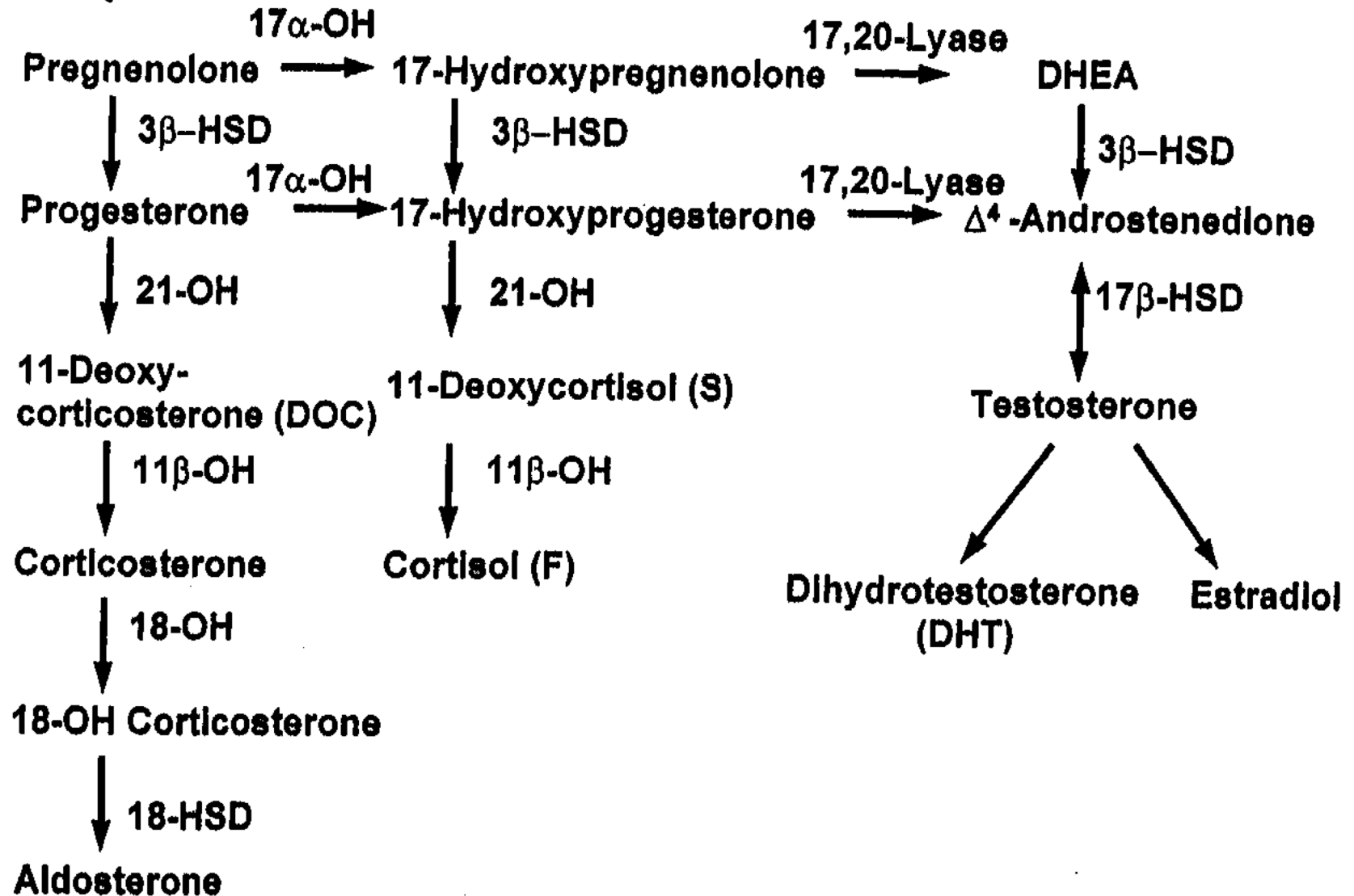
- deficiency of one of several enzymes necessary for steroid synthesis
- Autosomal Recessive (M=F)
- 21-hydroxylase is the commonest form

Cholesterol

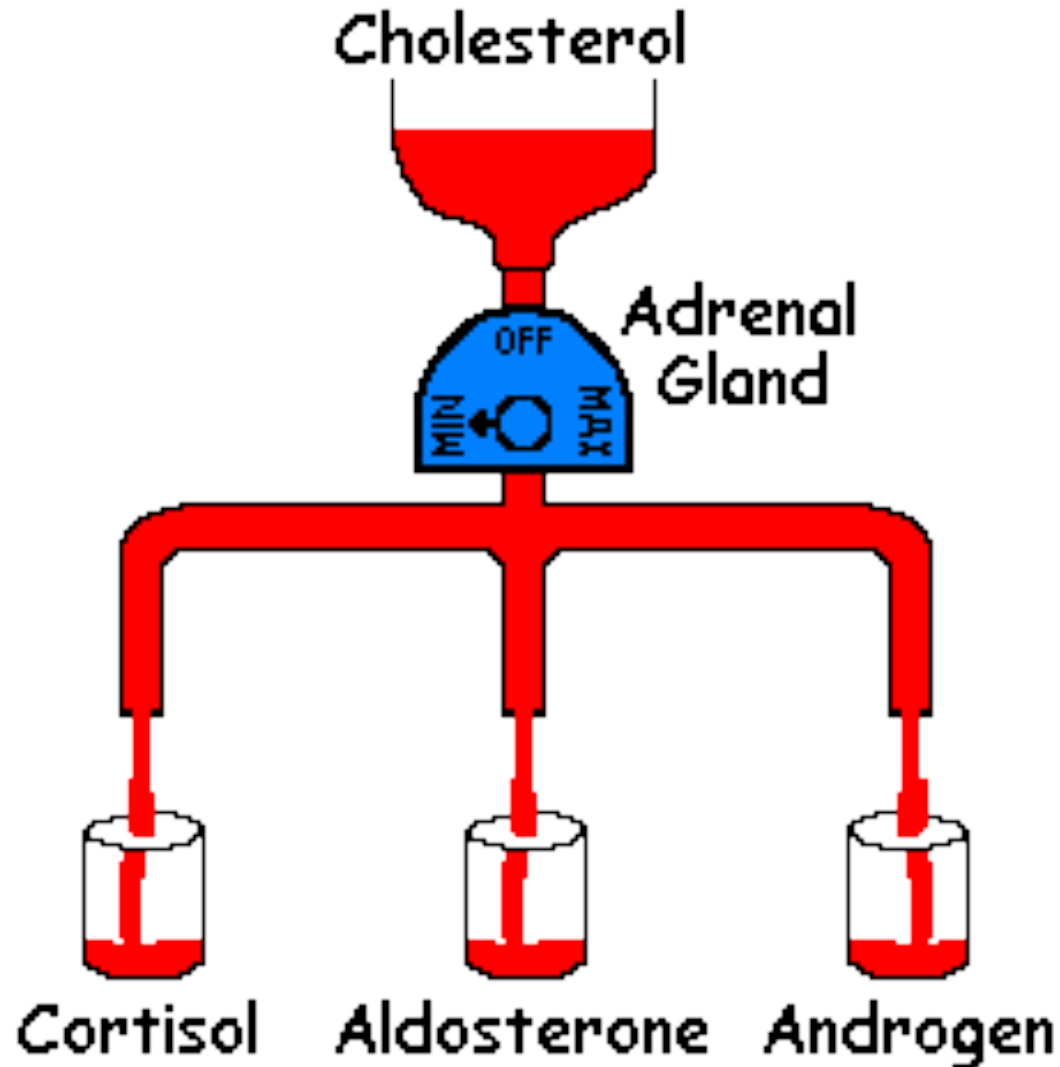
MINERALOCORTICOIDS

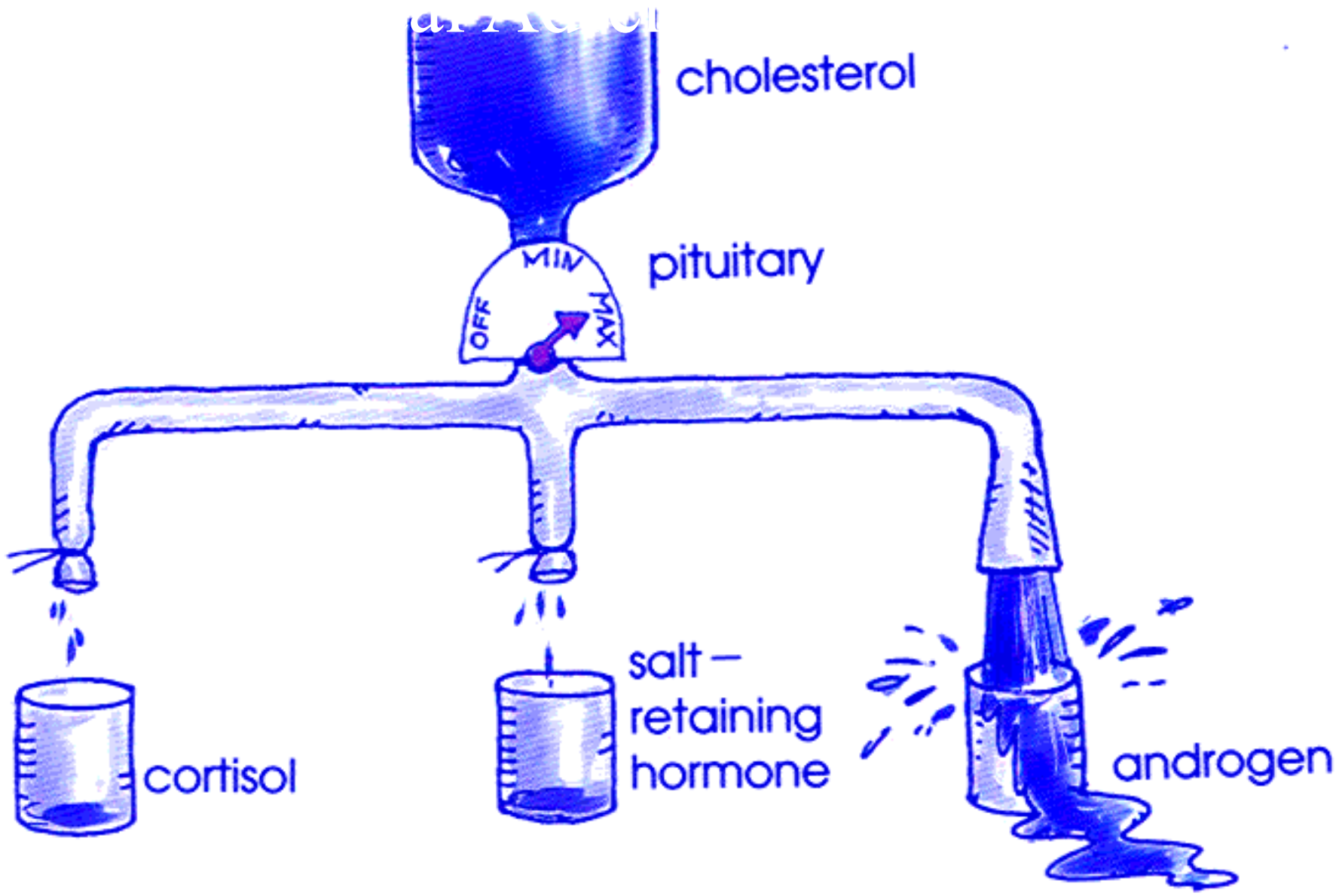
GLUCOCORTICOIDS

SEX HORMONES



Congenital Adrenal Hyperplasia



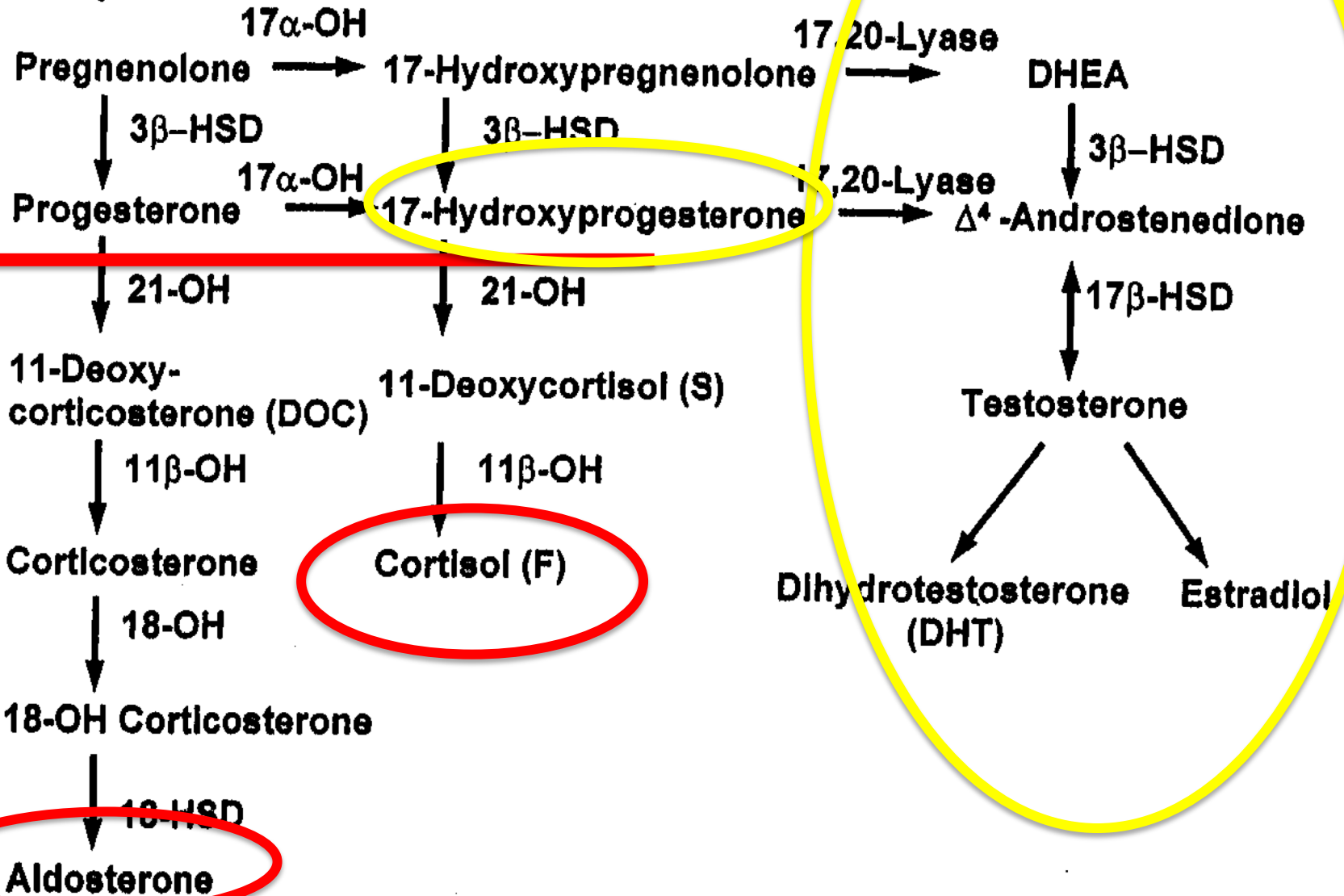


Cholesterol

MINERALOCORTICOIDS

GLUCOCORTICOIDS

SEX HORMONES

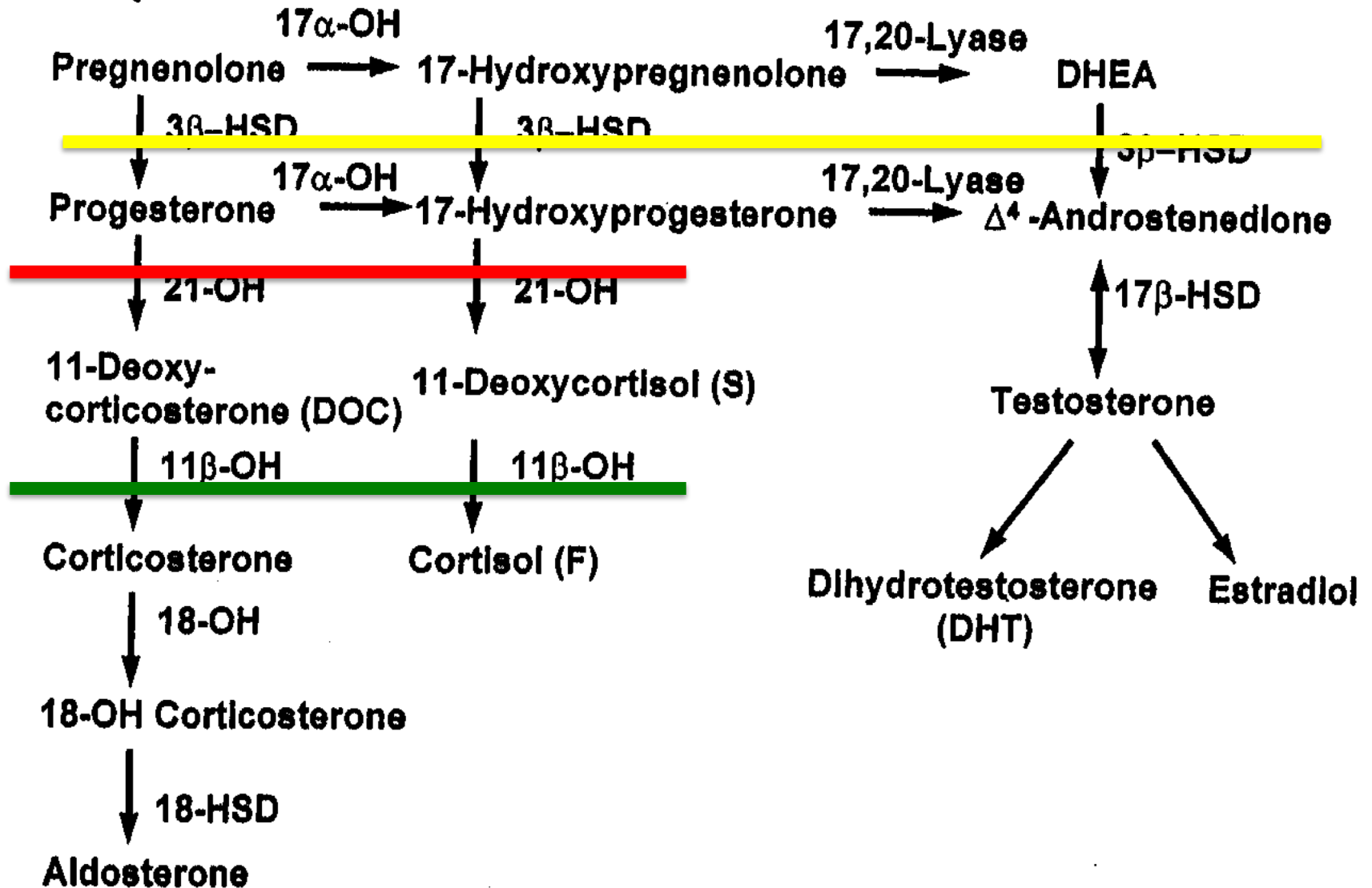


Cholesterol

MINERALOCORTICOIDS

GLUCOCORTICOIDS

SEX HORMONES



Presentations of 21-Hydroxylase CAH

Early

- **Complete enzyme defect**

Late

- **Partial enzyme defect**

Early Presentations of 21-Hydroxylase CAH

Boys

- Normal genitalia

Girls

- Ambiguous genitalia

2nd week of life if genitalia looks normal or if not recognized:

- Dehydration
- Shock
- Salt-loss presentations with electrolytes imbalance
 - Hyponatremia
 - Hyperkalemia
 - Hypoglycaemia
- Hyper-pigmentations

**Salt
loosing
crisis**











Diagnosis

- Serum electrolytes & glucose
 - Low Na & high K
 - Fasting hypoglycemia
- Elevated plasma Renin & ACTH levels
- Low Cortisol
- Low Aldosterone
- High 17 OHP
- High androgens especially testosterone level
- Chromosomes
- Pelvic US

Management

- **Life-long** Hydrocortisone
- Fludrocortisone 0.05 - 0.2 mg/day
- Triple hydrocortisone during stress
- **During adrenal crisis intravenous hydrocortisone and IV fluid**
- Corrective Surgery for female external genitalia
- Monitor growth

Newborn screening for CAH

- Neonatal screening by filter paper on 2nd day of life
- 17 Hydroxyprogestrone blood level (17 OHP)

903/W961/11/98

Baby's Surname

PRICE

Fill all circles



4 Kg

Late Presentations of 21-Hydroxylase Non-classical CAH

- Residual enzyme activity
- Non salt losing CAH

**later in
childhood**

- **early pubic hair**
- **precocious puberty**
- **accelerated growth**

**adolescence or
adulthood**

- **Virilization**
- **oligomenorrhea**
- **infertility**



Questions

