491 XXIII. Diseases of the Endocrines





# PITUITARY DWARFISM

o The achondroplastic dwarf has an orthopedic reason for having short limbs and Dwarfism a short spinal colum. The pituitary dwarf lacks growth hormone (an endocrine reason). Tumor causing hypopituitarism **o SYMPTOMS:** (1)✓ GH Deficiency #ADAM Low blood sugar

# Prevention

There is no preventing pituitary dwarfism

In some cases it may be caused by traumatic injury to the pituitary gland

Children with this disorder are smaller then others however, are just as smart and can lead long healthy lives

# Below average growth rate

# 2. Adult hypopituitarism

# (Simmonds's disease = progeria = pituitary old age = premature senility in females).

*Causes:* Nearly complete destruction or atrophy of the anterior lobe of pituitary as in:

- 1. Post-partum necrosis or infarction of pituitary at puerperium (Sheehan's syndrome).
- 2. Surgical removal or accidental trauma.
- 3. Chromophobe adenoma.
- 4. Craniopharyngioma.
- 5. Diffuse fibrosis (syphilitic or tuberculous).
- 6. Hypofunction (and hypoplasia) and late stages of gigantism and acromegaly.
- 7. Idiopathic.



### Effects:

- 1. Mental depression, weakness and loss of appetite  $\rightarrow$  cachexia.
- 2. Gradual atrophy of external and internal genitalia.
- 3. Loss of sexual desire; sterility.
- 4. Loss of axillary and pubic hair.
- 5. Low metabolic rate; sensitivity to cold; low blood pressure.
- 6. Hypoglycaemia (a flat glucose tolerance curve).
- 7. Associated fibrosis or atrophy of thyroid, Parathyroids, adrenal glands, ovaries and endometrium.



# HYPOPITUITARISM

Simmonds' disease [Panhypopituitarism] Complete absence of pituitary hormones Cachexia: most prominent feature Follows destruction of the pituitary by surgery, infection, injury, or a tumor

# Sheehan's syndrome

[Post-partum pituitary necrosis] A complication of delivery Results from severe blood loss and hypovolemia → Pituitary ischemia



Other clinical types:

3. Froehlich's syndrome (dystrophia-adiposo-genitalis).

Common; in children and at puberty; there is a hypothalamic lesion as well. *Causes:* 

• Chromophobe cell adenoma producing pressure-atrophy of eosinophil and basophil cells; and, implicating the posterior pituitary (& hypothalamus).

# Effects:

- 1. Sexual infantilism and external hypoplasia  $\rightarrow$  impotence and amenorrhea.
- 2. Excessive sudden adiposity with fluctuations (in breasts, hips and abdomen).
- 3. Atrophy of dermal connective tissue  $\rightarrow$  hairless thinned out skin.
- 4. Some mental retardation.
- 5. Low basal metabolic rate.



# 4. Laurence-Moon-Biedl syndrome: A variation of Froehlich's syndrome.

- In addition to :
  - 1. Adiposity.
    - 2. Genital dystrophy.
- There are:
  - 3. Poly-dactylism (six fingers and six toes).
  - 4. Retinitis pigmentosa  $\rightarrow$  partial blindness.
  - 5. Retarded mental development.
  - 6. A familial tendency.





# syndrome or Laurence-Moon-Biedl-Bardet



- . Obesity, hypogenitalism like in patients with Babinsky-Frelych's disease.
- 2. Decreased mental activity or debility.
- 3. Pigmental retinitis.
  - . Bones or inner organs abnormalities (polydactylia, syndactylia and others)



# 5. The Lorain Syndrome (Peter-Pan).

- A graceful attractive child who does not seem to grow up:
  - 1. Genital hypoplasia (undeveloped sexually).
  - 2. Small stature but the body is of normal proportions.
  - 3. Bright mentally.



# **Pituitary Dwarfism**

(Lorain-Levi syndrome)

- This is caused by severe deficiency of GH, and possibly of other hormones, in childhood.
- The individual is of small stature, height of adult is 3 ft. but is well proportioned, head is slightly larger in relation to body.
- Mental development is not affected.
- Puberty is delayed and there may be episodes of hypoglycaemia..

# 6. Brissaud's syndrome.

• Dwarfism + thyroid deficiency.

# *N.B.* 4

*Hypo-function of the <u>posterior pituitary</u>:* 

- This results in diabetes insipidus with
  - a. Polyuria at first (dilute urine up to 10 liters/day and
  - b. Sp. Gr. 1.002; with no sugar or albumin) and
  - c. Polydipsia (thirst) later on.

# Causes of diabetes insipidus:

- 1. Hypofunction of the posterior pituitary.
- 2. Traumatic injury of hypothalamus or interruption of its nerve-tracts (by operation).
- 3. Degeneration or disuse.

# 4. Tumours:

- **Primary** (Chromophobe adenoma; craniopharyngioma).
- Secondaries (from cancer of lung or breast).
- 5. Congenital (hereditary) or idiopathic.
- 6. Meningitis; encephalitis.
- 7. Tuberculosis; sarcoidosis.
- 8. Hand-Schuller-Christian disease.

# A pituitary tumour may be associated with a <u>hypothalamic syndrome:</u>

- 1. Polyuria.
- 2. Adiposity.
- 3. Pathological sleep (hypersomnelence).





# FLUID BALANCE: A MATTER FOR THE BRAIN AND KIDNEYS



