Medical condition, caused by problems arising in the pituitary gland, in which the body does not produce enough growth hormone.

**Phases of Growth**

- **Thyroid hormone:** birth through infancy
- **Growth hormone:** school age adolescence
- **Sex hormone (also GH, Insulin):** adolescence (puberty)

**Causes**

- 25% of the patients with growth hormone deficiency had an organic etiology;
  - CNS tumor (e.g. Craniopharyngiomas)
  - CNS malformation
  - Septo-optic dysplasia
  - Leukemia
  - CNS radiation
  - CNS trauma
  - Histiocytosis
  - CNS infection
- 75% is idiopathic

**GH deficiency**

**History**
- Normal BWT
- Deceleration after 2 years of age
- Hypoglycemia
- Micropenis

**Examination**
- Severe short stature
- Proportionate stature
- Low muscle to fat ration
- Micropenis
- Midfacial defects
- Low growth velocity

**Lab**

**1st test**
- IGF1 (Low for age and sex)
- IGFBP3 (low for age and sex)

**Other tests**
- Low peak GH<10mcg on 2 occasions
- MRI of the brain
- Full pituitary hormone assessment

**Short stature**

- Is defined as a standing height below the 2.5 percentile for sex and age

**Growth failure**

- Is a pathologic state of abnormally low growth rate over time, whereas short stature is often a normal variant
Common causes of short stature

- Familial short stature
- Constitutional delay of growth and development
- Idiopathic short stature
- SGA without catch up growth after 2 years
- Chronic illnesses

Uncommon causes of short stature

- GH deficiency
- Tuner syndrome
- Cushing syndrome
- Hypothyroidism
- GH insensitivity
- Noonan syndrome
- Russul -silver syndrome
- Down syndrome
- Craniopharangioma….. ect

Lab tests for short stature

- Insulin like growth factor-I (IGF-I), and IGF binding protein-3 (IGFBP-3)
- Karyotype
- Measuring serum levels of GH (Basal and provocative)
- CBP & ESR
- Antiendomysial immunoglobulin
- Thyroid function test
- Serum transferrin and prealbumin concentrations for under nutrition

Imaging studies

- AP X-ray of left hand and wrist for assessment of bone age.

Morbidity and mortality

- Average adult height for untreated patients with severe isolated GH deficiency is 143 cm in men and 130 cm in women.
- 5% of children with GH deficiency also have episodes of hypoglycemia, particularly in infancy, which resolve with GH therapy

History of growth hormone therapy

- 1887 Gigantism dt GH
- 1922 Injection of pituitary extract
- 1957 HGH
- 1985 was linked to CJD
- 1971 Structure of GH
- 1981 Synthetic GH by DNA tech
Treatment

- Growth hormone replacement is used to treat growth hormone deficiency
- It usually causes significant increase in growth velocity (averaging 10-11 cm/y during first y of therapy). Response wanes each y, but growth velocity continues at faster than pretreatment rates

Complications

- Benign intracranial hypertension
- Edema and even carpal tunnel syndrome
- Skeletal and joint problems;
  - slipped capital femoral epiphysis
  - progression of scoliosis
- Prepubertal gynecomastia
- No evidence suggests an association between growth hormone therapy and leukemia in otherwise healthy children.

Prognosis

- With treatment most children with growth hormone deficiency reach normal adult stature.

Diabetes Insipidus

Deficient secretion antidiuretic hormone (ADH)—by the pituitary gland (central DI)

(2) renal tubular unresponsiveness to ADH

Pathogenesis

ADH deficiency or resistance
- Inability to reabsorb water in the collecting tubules of the kidney
- Na & electrolyte absorption is unaffected

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Pure water loss
- Diluted urine
- Hypermaturemia

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Serum hyperosmolarity
- Polydipsia

Etiology

Non Genetic
- Head trauma, tumor,
- Neurosurgical procedures.
- Destructive lesions of the pituitary, hypothalamus, or both

Genetic
- Wolfram syndrome AR
- X-linked nephrogenic DI
- AD & AR DI
Age of presentation

- Central DI
  - AR present before 1 year
  - AD present after 1 year
- Nephrogenic DI
  - AD, AR & X linked all present with one week of life
- Secondary DI
  - Occurs at a random order

Clinical presentation

- Nonspecific presenting features (poor feeding, failure to thrive, irritability).
- Vigorous suck with vomiting
- Fever without apparent cause
- Constipation
- Excessively wet diapers from urination, Nocturia

Physical examination

- Irritable infant with a dripping wet diaper, along with detectable signs of dehydration

Complications

- Growth failure
- Nocturia and enuresis
- Hypernatremic dehydration
- Seizures
- Mental retardation
- Hypovolemic shock

Differential diagnosis

- Diabetes Mellitus, Type 1
- Head Trauma
- Histiocytosis
- Hypercalcemia
- Medullary cystic disease
- Sickle cell anemia
- Hypokalemia

Lab tests

- Urine specific gravity of the first morning
- Dilute urine with a relatively high serum sodium and osmolarity
- 24-hour urine collection
- Serum K and Ca to exclude polyuria secondary to Hypokalemia and Hypercalcemia
- Cranial MRI; to exclude pituitary cysts, hypoplasia, and mass
- Water deprivation test
Treatment of Central DI

- Intravenous fluid in case of dehydration especially during illness
- Central DI; treatment of choice is desmopressin, Aqueous vasopressin are available for intravenous (IV) use in emergency circumstances.
- Other useful medications include chlorpropamide and thiazide diuretics
- Over dose of desmopressin may cause; hyponatremia and seizures

Treatment of Nephrogenic DI

- Thiazide diuretics
- Amiloride, and indomethacin or aspirin are useful when coupled with a low-solute diet.

Diet and Activity

- Breast milk diet to decrease solute load.
- Protein should account for 6% of caloric intake
- Sodium should be reduced to 0.7 mEq/kg/day

Prognosis

- Central DI has an excellent prognosis with replacement with desmopressin
- In Nephrogenic DI; increase risk of attention deficit, hyperactivity, learning disorders, and psychomotor delay, due to early presentation and poor response to treatment