

## GENODERMATOSES

- I. Ichthyoses
  1. Genetically determined conditions with thick scaling plates
  2. Two mechanisms:
    - a Increased cohesiveness (I. vulgaris, X-Linked Ich).
    - b Increased cellular production (lamellar Ich., epidermolytic hyperkeratosis)
  3. Infants often present with membrane ("colloidian baby")
  4. Symptoms:
    - a pruritus
    - b cosmesis
    - c bacterial 2<sup>o</sup> infections
    - d odor
    - e ectropion
    - f fluid, electrolyte imbalance
  5. Treatment:
    - a antibiotics
    - b moisturization (LacHydrin)
- II. Neurofibromatosis (von Recklinghausen's)
  - A. Autosomal dominant
  - B. cafe-au-lait macules (at least 6 greater than 1 cm.)
  - C. neurofibromas
    1. typical

- 2. plexiform-may eventuate in neurosarcoma
  - D. pheochromocytoma
  - E. acoustic neuroma
  - F. \*\*Crowe's sign--axillary freckling
  - G. \*\*Lisch nodules
- III. Tuberous Sclerosis
- A. Autosomal dominant
  - B. Ash leaf macules
  - C. Adenoma sebaceum (angiofibromas)
  - D. \*\*Shagreen patch
  - E. mental retardation
- IV. Peutz Jaegher's
- A. Autosomal dominant
  - B. Multiple lentigines
  - C. Small intestinal polyps (pre adenocarcinomatous)
- V. Gardiner's
- A. Autosomal dominant
  - B. multiple cysts
  - C. colonic polyps (premalignant)
- VI. Osler-Weber-Rendu
- A. Autosomal dominant
  - B. Familial hemorrhagic telangiectasia
  - C. Telangiectases on lips, oral mucosa, skin

D. Bleeding from CNS, GI tract