• ectodermal dysplasia

Ectodermal dysplasia is an X-linked recessive condition characterized by abnormal development of the skin and associated structures *(hair, nails, teeth, and sweat glands)*. It involves all structures that are derived from the **ectoderm.** It affects males more than females. Common clinical findings include **hypothrichosis** *(decrease in hair)*, **anhidrosis** *(no sweat glands, leading to heat intolerance),* **anodontia** or **oligodontia** *(complete or partial absence of teeth),* **depressed bridge of nose, lack of sweat glands,** and the **child appears much older than what he or she is**. There is no treatment for the disease, however dentures can be fabricated for these patients. Keep in mind that they will need to be replaced periodically to accommodate the patient's jaw growth. **See pictures #28 and #29 in booklet**

Cleidocranial dysplasia is an autosomal dominant condition of bony development characterized by **hypoplasia or aplasia of the clavicles,** cranial bossing, ocular hypertelorism, and dental abnormalities which include **retained primary teeth,** malaligned teeth, the presence of **multiple supernumerary** teeth, and **unerupted** teeth. **Important:** The dentition itself, as observed by radiographs alone, often suggests the diagnosis. **See picture #30 in booklet**

Pierre Robin syndrome is an inherited disorder that presents the following in the neonate: severe **micrognathia**, **mandibular hypoplasia,** severe **glossoptosis** *(posterior displacement of the tongue),* and **high-arched** or **cleft palate.** This condition is also characterized by respiratory problems.

Peutz-Jeghers syndrome *(PJS)* is a genetic condition marked by hyperpigmentation *(freckling)* of the lips and sometimes other parts of the face, hands, and feet followed by the development of benign polyps called hamartomas throughout the intestines but primarily in the small intestine. **See picture #35 in booklet**

Osteopetrosis *(also called Albers-Schonberg disease or marble bone disease)* is an uncommon bone condition that may be inherited as an autosomal dominant *(less serious)* or recessive trait *(more serious)*. The characteristic feature of osteopetrosis is an **absence of physiologic bone resorption owing to reduced osteoclastic activity.** The lack of bone resorption results in accumulation of bone mass and manifests itself in skeletal disturbances, including bone cavity occlusion, decreased hematopoietic activity, and growth retardation. Bone pain is the most frequent symptom. Blindness and deafness from sclerosis of ostia, anemia from sclerosis of bone marrow, and osteomyelitis due to diminished vascularity are also seen. **Dental findings** include delayed eruption, congenitally absent teeth, unerupted and malformed teeth, and enamel hypoplasia.