The Annual General Pediatric Review & Self Assessment

Nicklaus Children's Hospital

DERMATOLOGY

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Disclosure of Relevant Relationship

Dr. Usmani has not had (in the past 24 months) any relevant conflicts of interest or relevant financial relationship with the manufacturers of products or services that will be discussed in this CME activity or in his presentation.

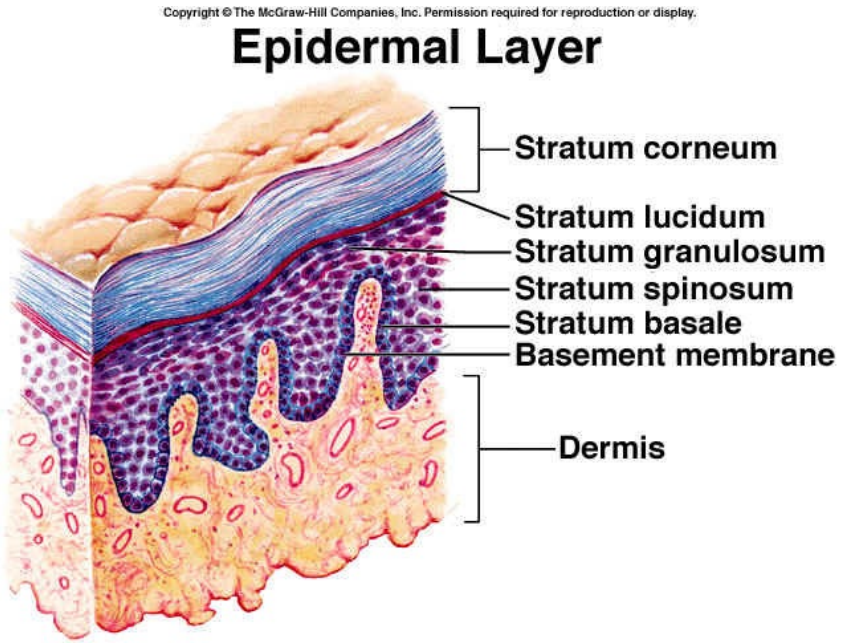
Dr. Usmani will support this presentation and clinical recommendations with the "best available evidence" from medical literature.

Dr. Usmani does not intend to discuss an unapproved/investigative use of a commercial product/device in this presentation.

SKIN facts

- Skin is the largest organ
- Main protein of the skin is keratin
- It renews every 28-32 days

- Skin has three layers:
  - The epidermis, the outermost layer of skin, provides a waterproof barrier and creates our skin tone.
  - The dermis, beneath the epidermis, contains tough connective tissue, hair follicles, and sweat glands.
  - The deeper subcutaneous tissue (hypodermis) is made of fat and connective tissue.

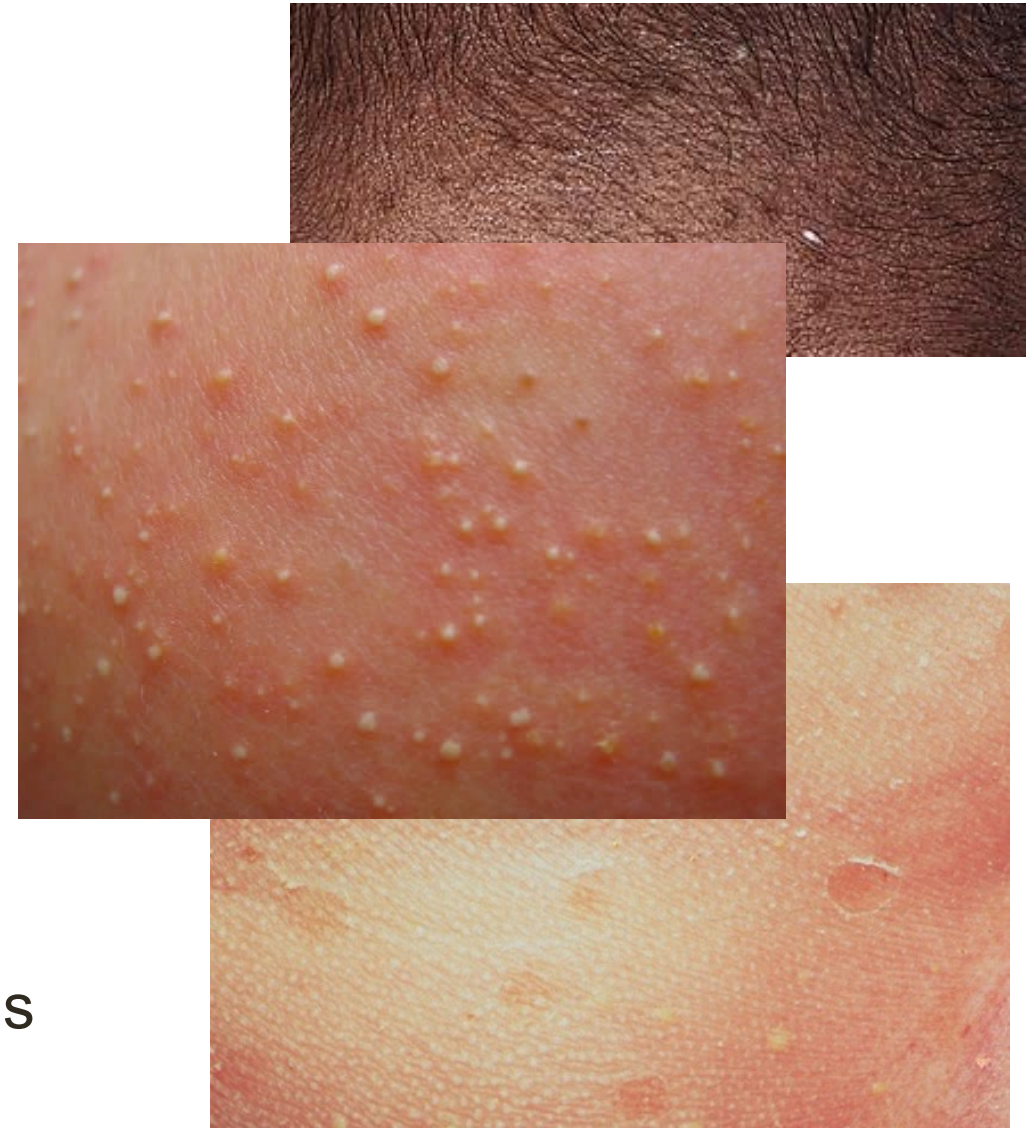


NEONATAL DERMATOLOGY

- Transient Neonatal Pustular Melanosis
- Erythema Toxicum Neonatorum
- Neonatal Cephalic Pustulosis (Neonatal Acne)
- Sclerema Neonatorum
- Subcutaneous Fat Necrosis of The Newborn
- Seborrheic Dermatitis
- Miliaria Crystallina (MC) or Miliaria Rubra (MR
- Aplasia Cutis Congenita (ACC
- Cutis Marmorata Telangiectasia Congenita (CMTC
- Sucking Blisters

NEONATAL DERMATOLOGY

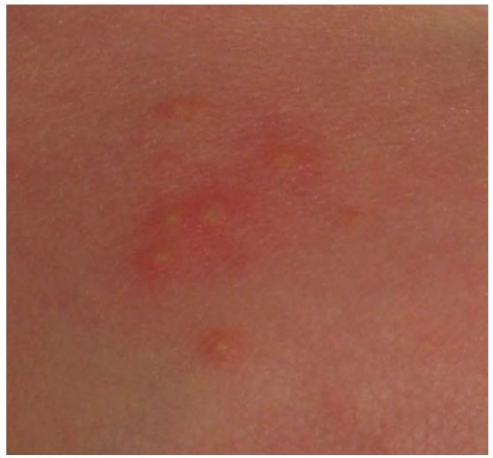
- Transient Neonatal Pustular Melanosis
  - Onset at birth
  - Common in darkly pigmented infants
  - Presents with small pustules or residual hyperpigmented macules with collarette of scale
  - Smear: neutrophils
  - Path:subcorneal pustules with neutrophils

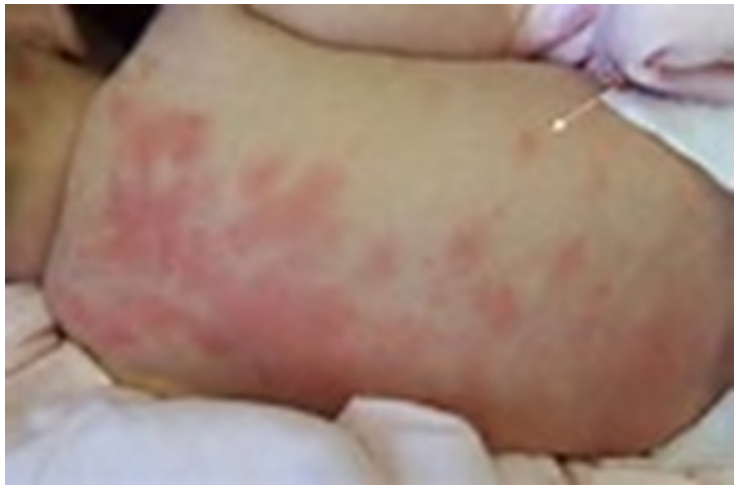


NEONATAL DERMATOLOGY

- Erythema Toxicum Neonatorum
  - Onset: 24-48 hours after birth (3 days to 2 weeks)
  - Appears in 50%newborns
  - Presents with blotchy erythematous macules, papules, pustules, and wheals
  - Smear: eosinophils
  - Path: subcorneal pustules with eosinophils associated with pilosebaceous units.
  - No treatment necessary

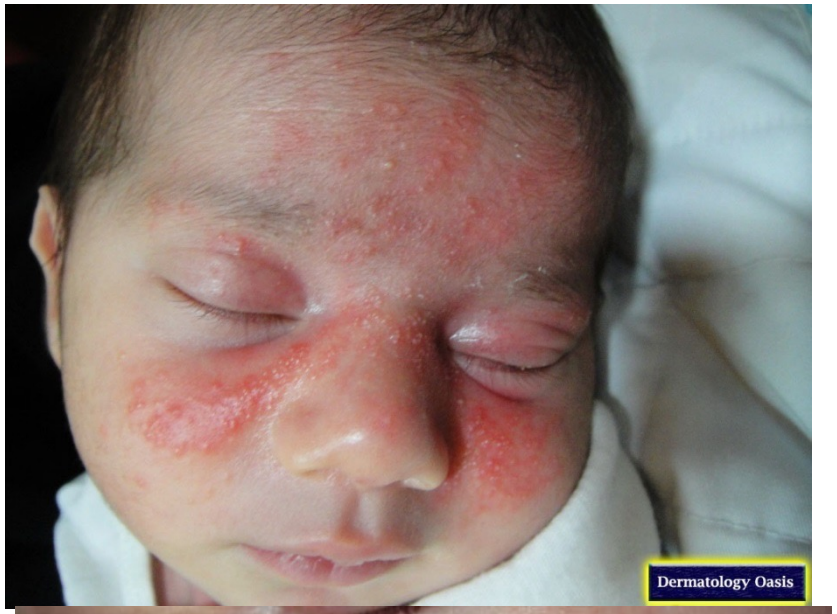
Erythema toxicum neonatorum

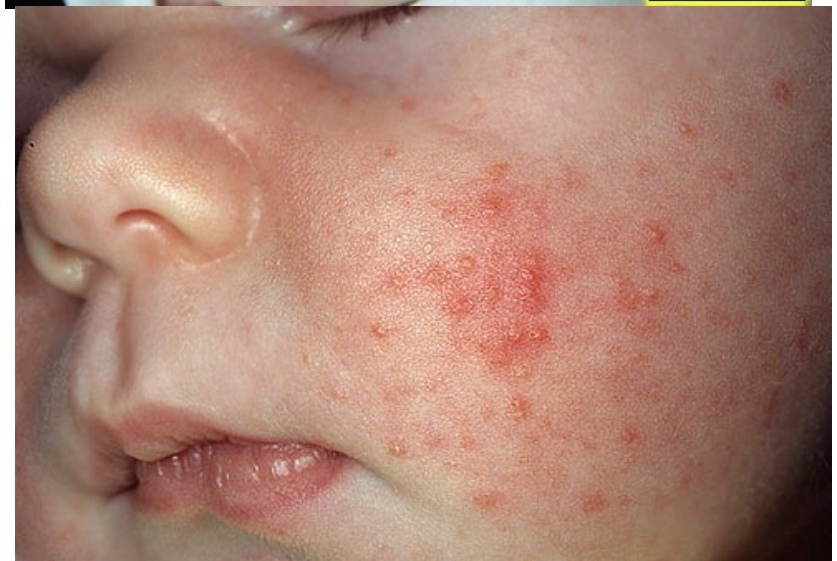




NEONATAL DERMATOLOGY

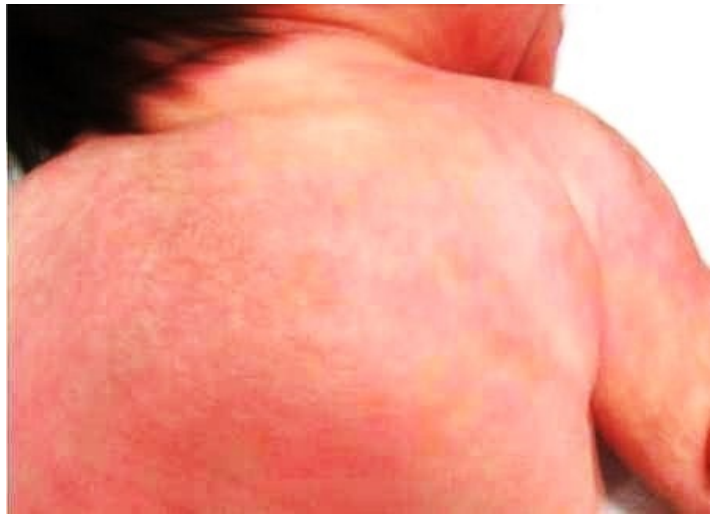
- Neonatal Cephalic Pustulosis (Neonatal Acne)
  - Onset: within first 30 days
  - Malassezia spp implicated in pathogenesis
  - Presents with comedones, papules, and pustules on face.
  - Path: follicular pustules with neutrophils

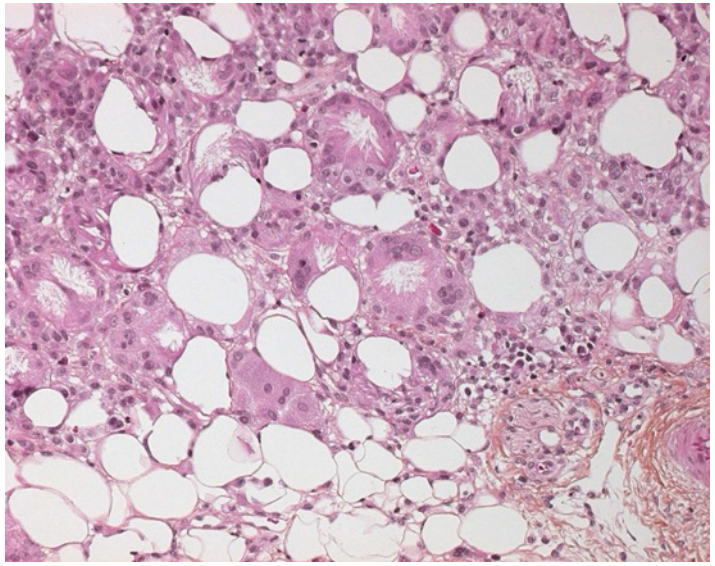




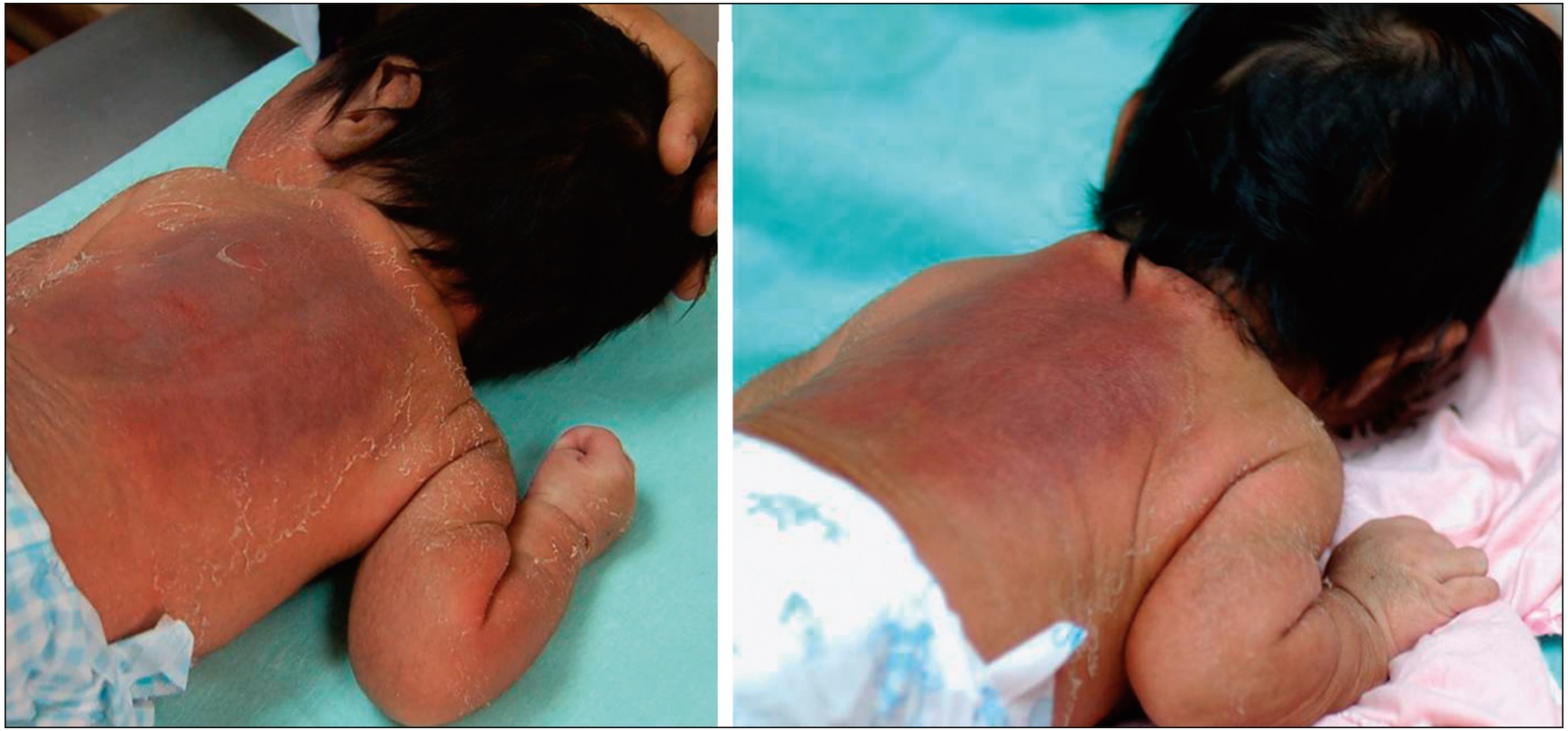
NEONATAL DERMATOLOGY

- Sclerema Neonatorum
  - Onset: within first week of life
  - It's a form of panniculitis in severely ill premature infants.
  - Often fatal
  - Presents with woody hardening of skin; spares genitalia, palms and soles.
  - Path: needle-shaped clefts with necrotic adipocytes with little surrounding inflammation





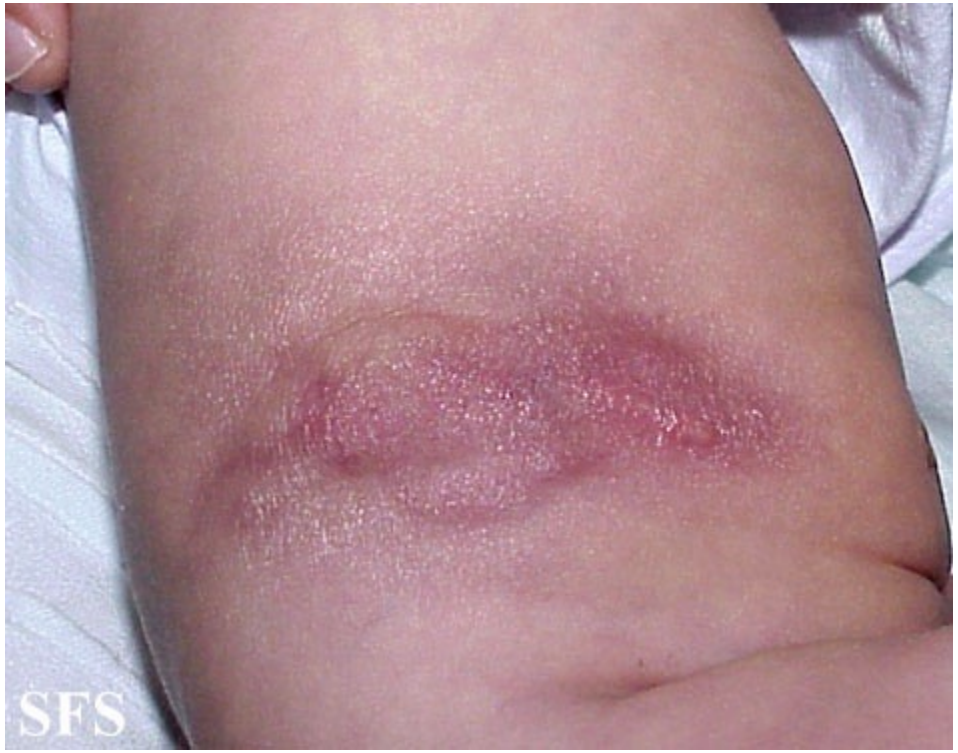
NEONATAL DERMATOLOGY



NEONATAL DERMATOLOGY

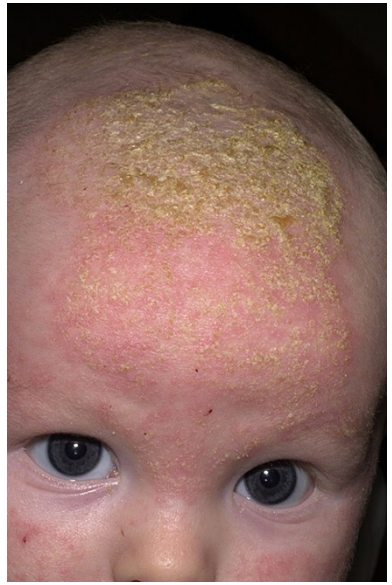
- Subcutaneous Fat Necrosis of The Newborn
  - Onset: within first week of life
  - Localized form of sclerema neonatorum in healthy infants
  - Presents with indurated subcutaneous nodules favoring cheeks, shoulders, back, buttocks, and thighs.
  - Associated:
    - hypothermia, perinatal hypoxemia (from preeclampsia, meconium aspiration etc.), hypoglycemia.
  - Calcification may occur, +/- profound hypercalcemia with resolution.
    - Check Ca+ levels 1 month after resolution of lesions
  - Path: panniculitis with prominent inflammatory infiltrate, needle-shaped clefts and fat necrosis

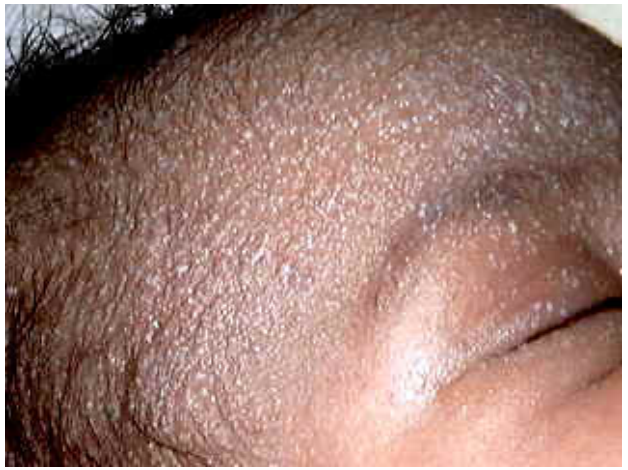
NEONATAL DERMATOLOGY



NEONATAL DERMATOLOGY

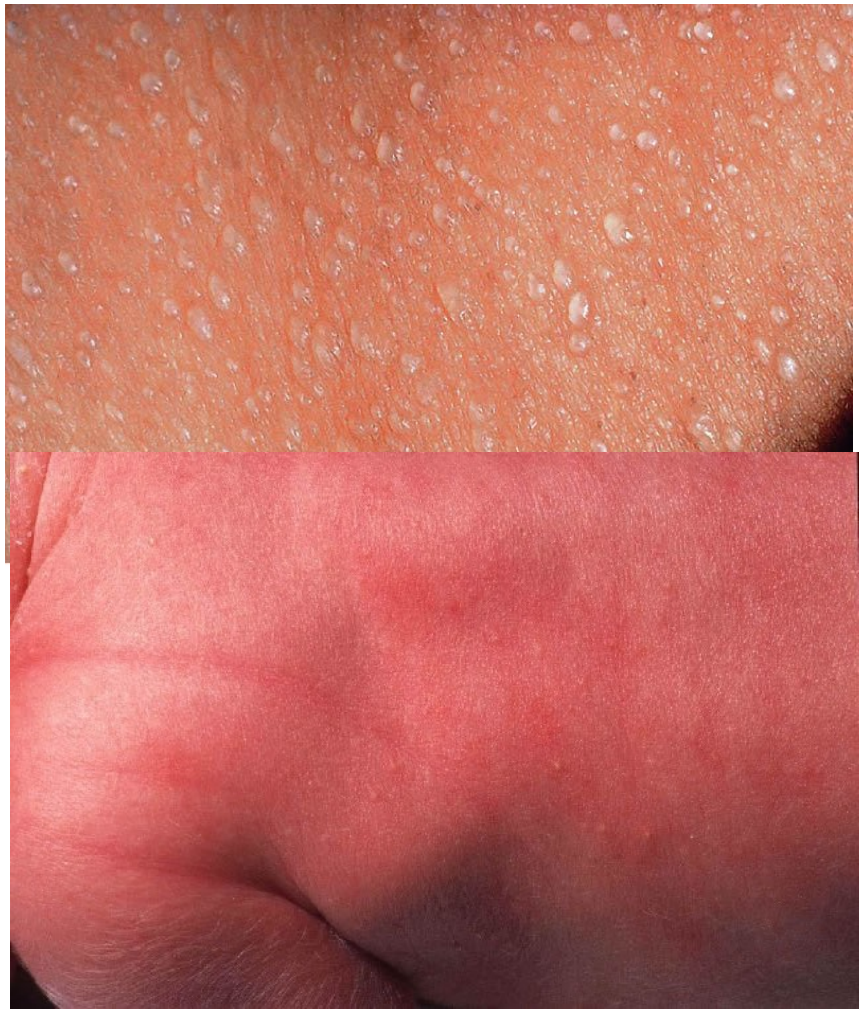
- Seborrheic Dermatitis
  - Onset: typically 1 week after birth
  - Lasts several months, mostly resolves by 1 year of life
  - Presents with ill-defined erythematous patches with waxy scales over scalp (cradle cap); can be present in the axillae and groin; lesions may appear psoriasiform





NEONATAL DERMATOLOGY

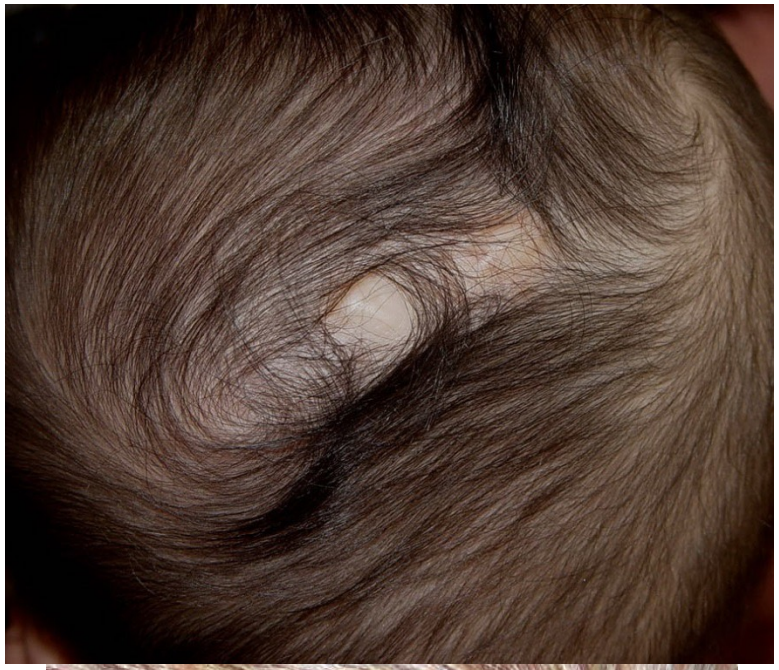
- Miliaria Crystallina (MC) or Miliaria Rubra (MR)
  - Onset: within first few weeks; due to obstructed sweat glands and associated with inc temperature (i.e. occlusion)
  - Presents with clear vesicles on head, neck, and upper trunk (MC) or erythematous vesicles in intertriginous areas or occluded areas (MR)

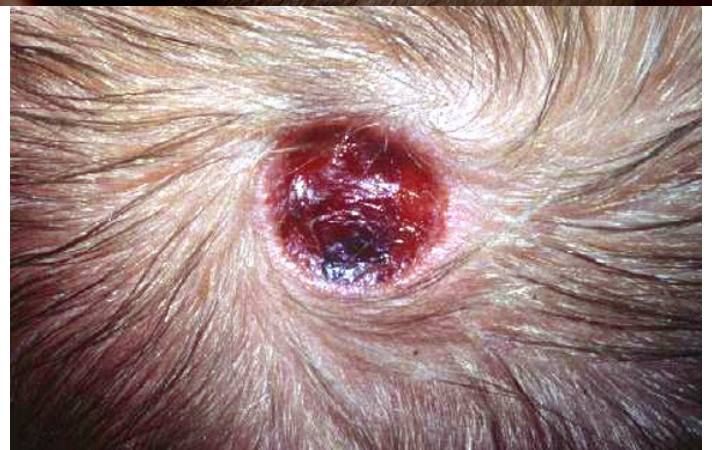


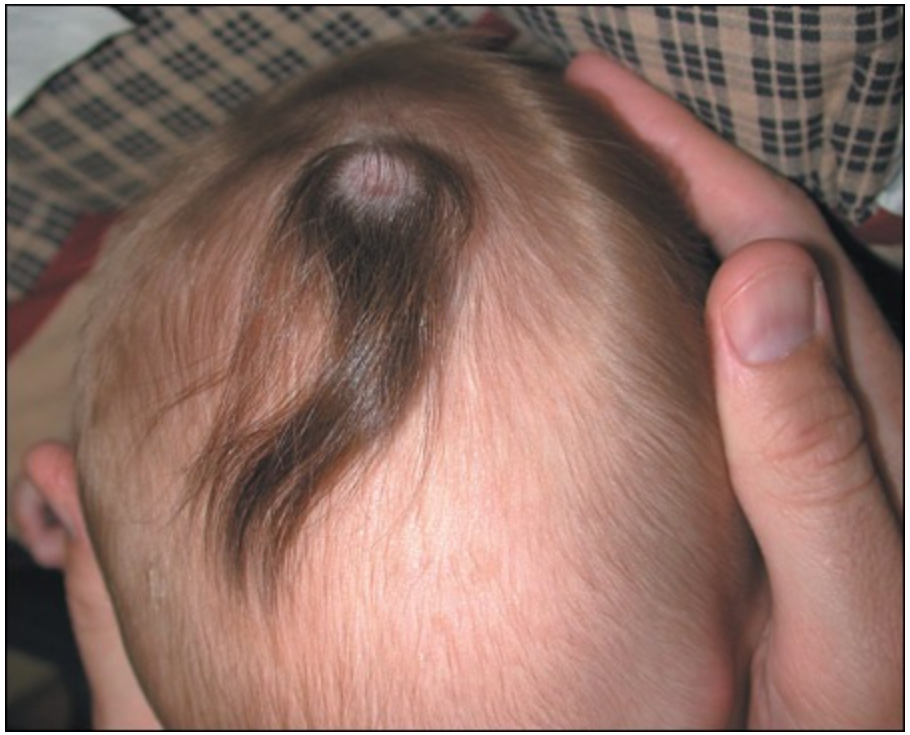
NEONATAL DERMATOLOGY

- Aplasia Cutis Congenita (ACC)
  - Onset: before birth
  - Localized defect in epidermis, dermis, and/or fat; variable appearance, typically along midline.
  - Presents with erosion, ulceration, scar or membranous defect.
  - Hair collar sign: ring of dark long hair around the lesion is a marker of underlying neural tube defect.
  - Typically isolated abnormality.
  - Can be associated with following syndromes
    - Bart Syndrome
    - Adams-Oliver Syndrome
    - Seitles Syndrome

NEONATAL DERMATOLOGY

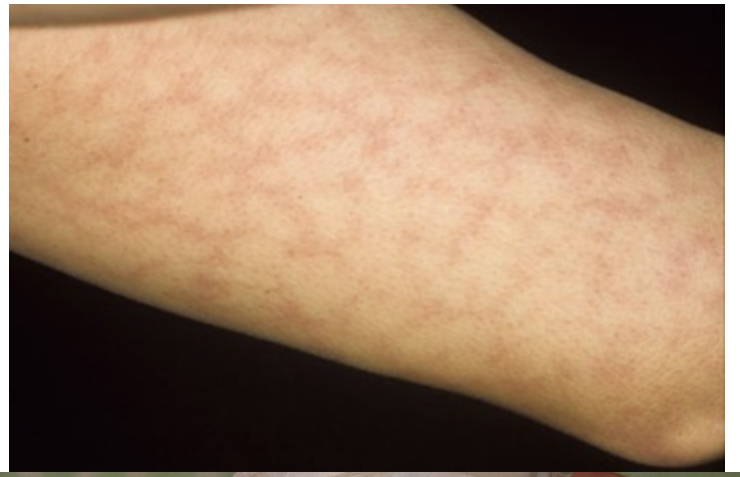


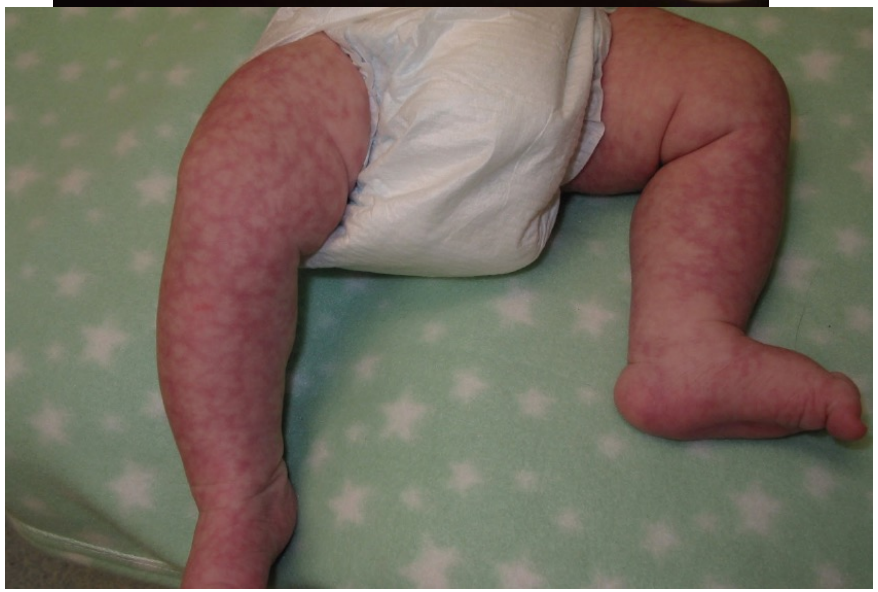




NEONATAL DERMATOLOGY

- Cutis Marmorata Telangiectasia Congenita (CMTC)
  - Onset: at birth, improves with age
  - Presents with blanching reticulated vascular pattern on trunk/extremities with segmental distribution
  - Associated with anomalies in ½ of pts(varicosities, nevus flammeus, macrocephaly, ulceration, hypoplasia,and or hypertrophy of soft tissue and bones).



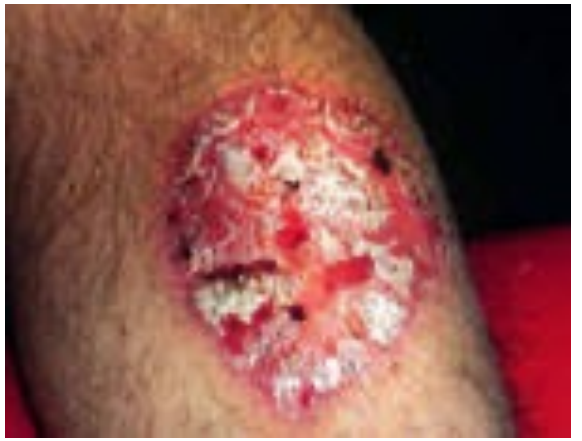


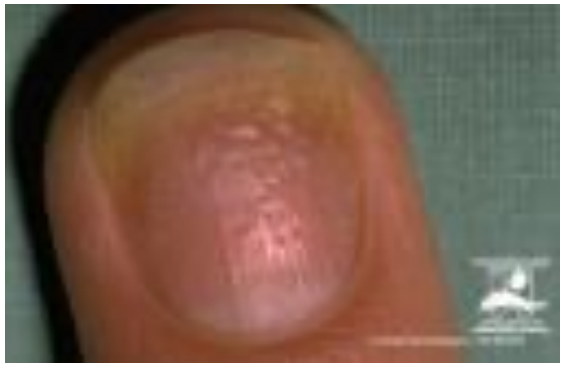
PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

- Psoriasis
- Pityriasis Lichenoides (PLEVA, PLC)
- Acropustulosis of Infancy
- Pityriasis Rosea
- Lichen Striatus
- Atopic Dermatitis (Eczema)
- Diaper Dermatitis
- Tinea infections (corporis, pedis, cruris)

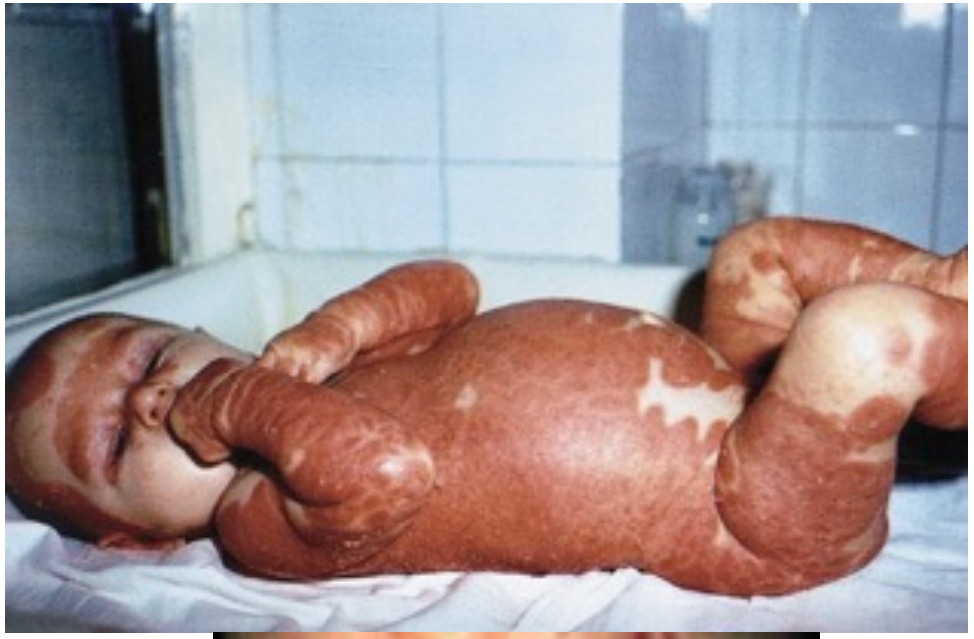
PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

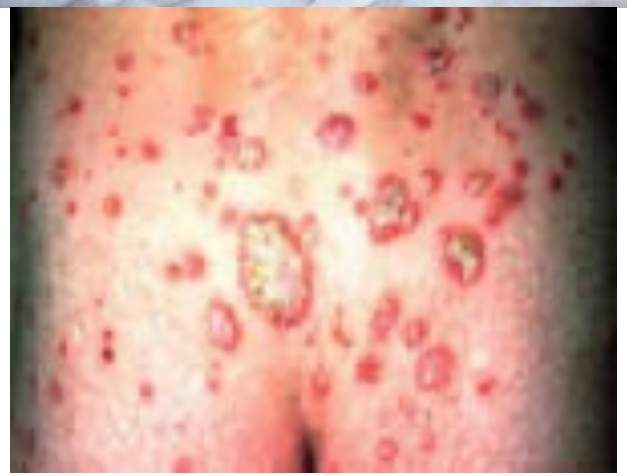
- Psoriasis
  - 25% presents before 15
  - Erythematous well demarcated plaques with micaceous scale
  - Associted with psoriatic arthritis, nail involvement (pitting, oil spots, hyperkeratosis)
  - Guttate psoriasis is common in children, presents with nummular plaques scattered on the body; triggers include strep infections, viral infecions, stress and trauma

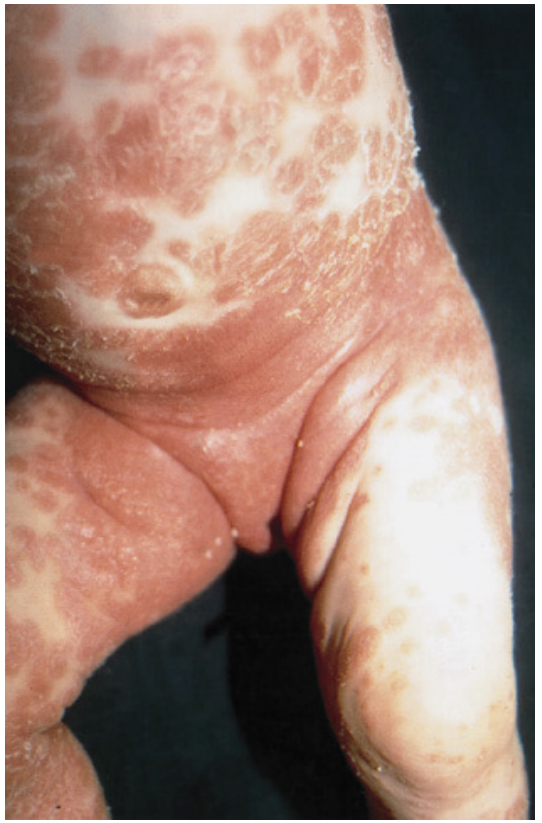




PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

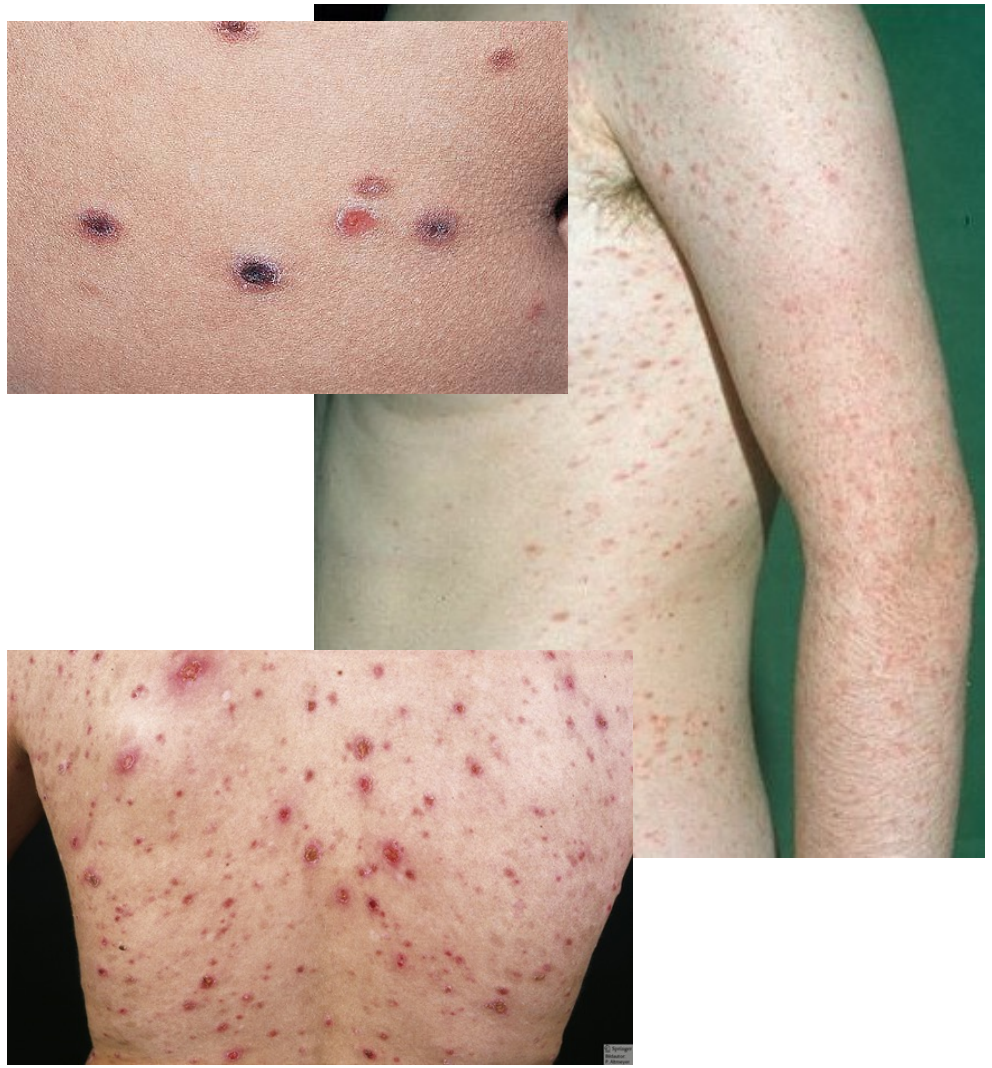






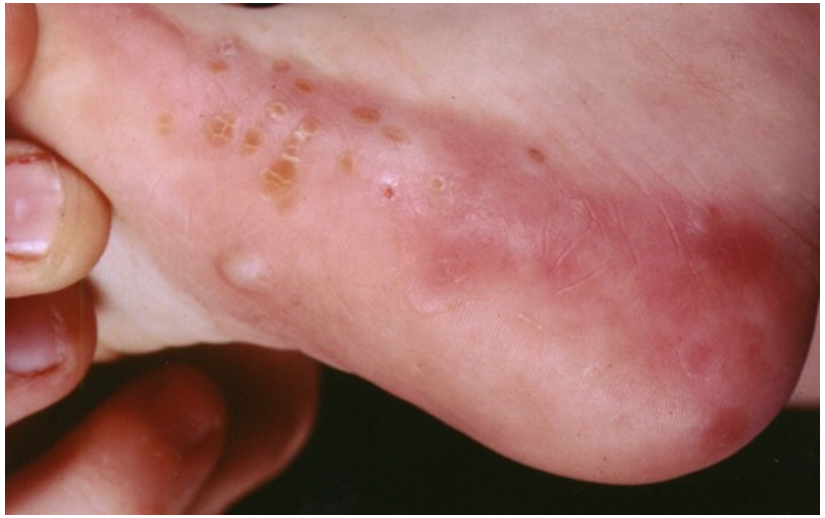
PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

- Pityriasis Lichenoides
  - PLEVA: abrupt onset of erythematous papules and vesicles with crusted or necrotic centers, often involuting within weeks to months, tx w/ erythromycin, phototherapy and steroids
  - PLC: reddish-brown papules with adherent scales, heals dyschromia; more chronic lasts months to years



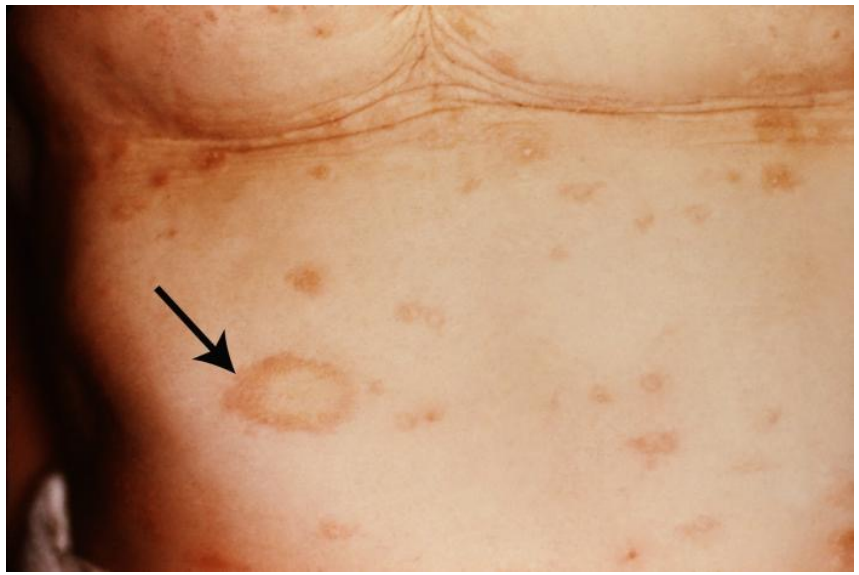
PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

- Acropustulosis of Infancy
  - Onset: 6 months to 2 years; resolves by age 3
  - Presents with recurrent crops of pruritic pustules on palms, soles and distal extremeties
  - Tx: topical steroids



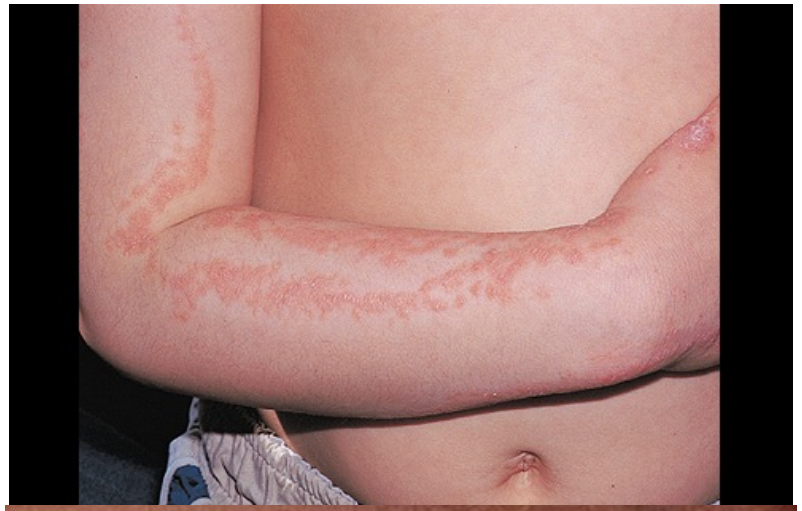
PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

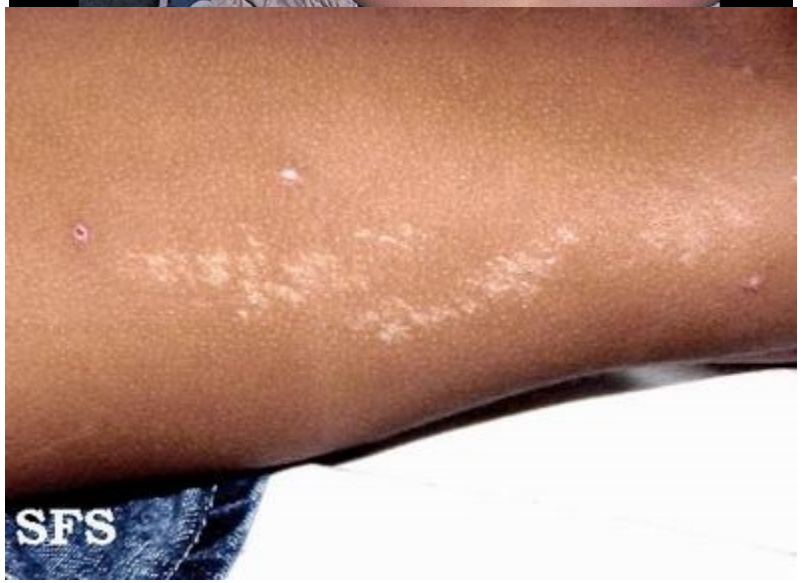
- Pityriasis Rosea
  - Self limited eruption
  - Viral pathogen HHV 7, HHV 6
  - Presents with initial Herald Patch
  - Followed by salmon-colored oval patches and plaques with inner scale along long lines of Langer's lines of cleavage (Christmas tree pattern)
  - Variants: inverse pattern (flexural accentuation); papular PR (younger children, darker-skinned pts)



PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

- Lichen Striatus
  - Self-limited, linear inflammatory red scaly papules spreads down extremity or trunk and typically follows lines of Blaschko's.
  - Associated with hypopigmentation
  - Resolves in few years

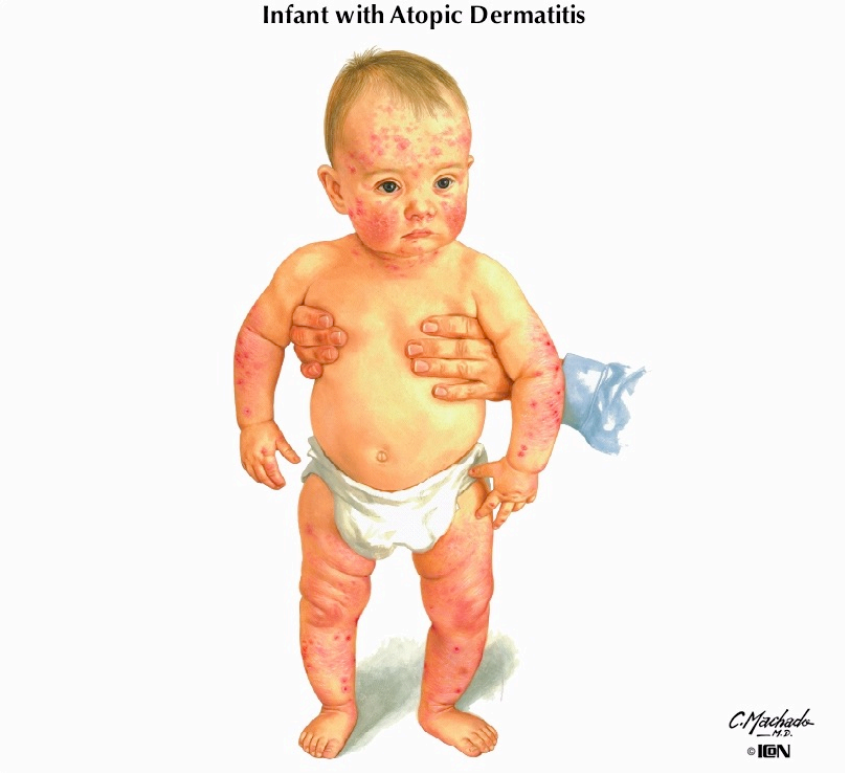




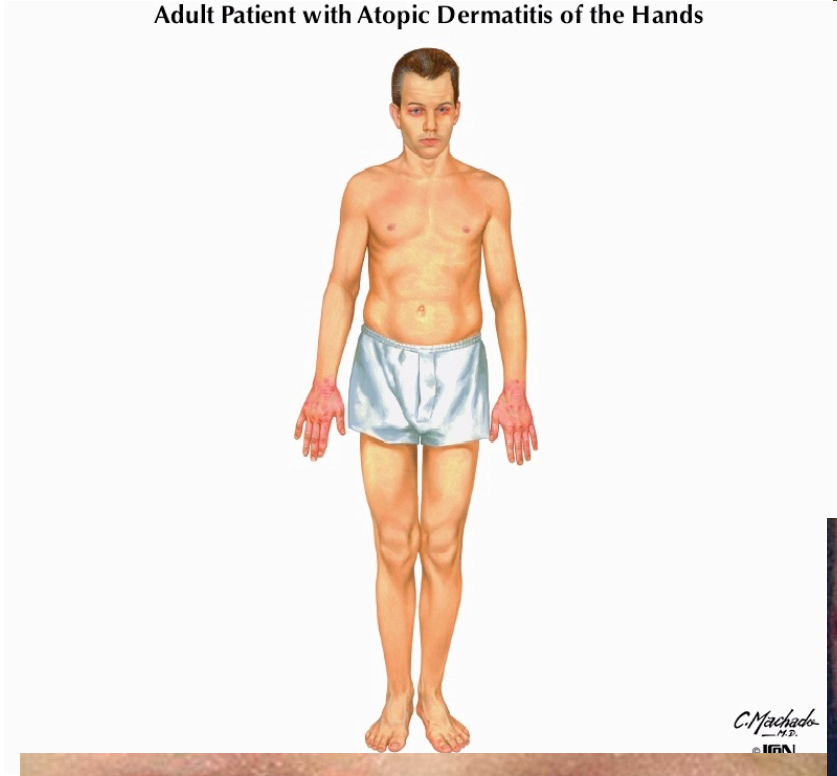
PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

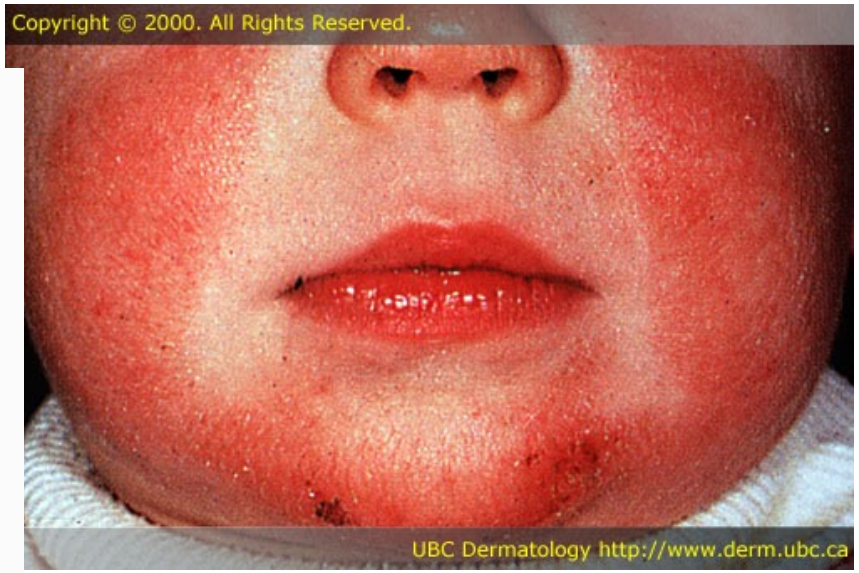
- Atopic Dermatitis
  - Occurs in 10-15% of children; presents at 2-3 months of age
  - Pathogenesis: multifactorial but includes inc secretion of TH2 cytokines (IL4, IL5)
  - Triad of atopy: AD, allergic rhinitis, asthma
  - Intrinsic type (inc IgE) or Extrinsic type (nl IgE)
  - Some patients can have food allergies
- Presents with eczematous lesions, xerosis, and lichenification
- Distribution varies with age
  - Infants: face, scalp, extensors
  - Children: antecubital/popliteal fossae, neck, wrists, ankles
  - Adults: typically chronic hand eczema

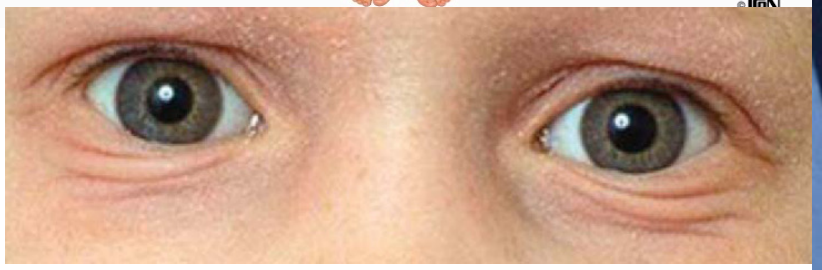
PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

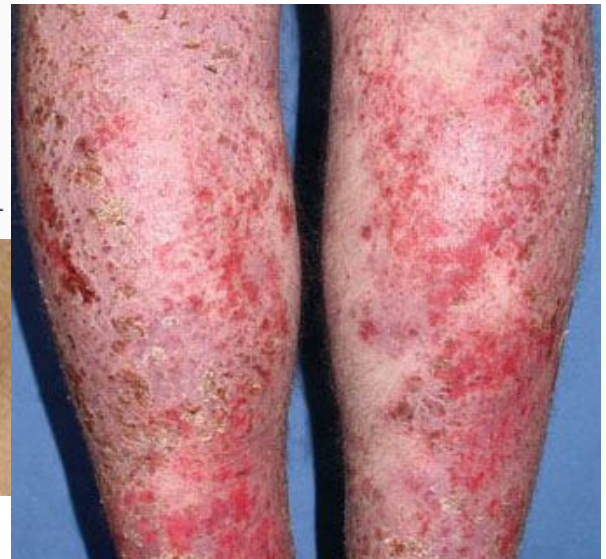


PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES



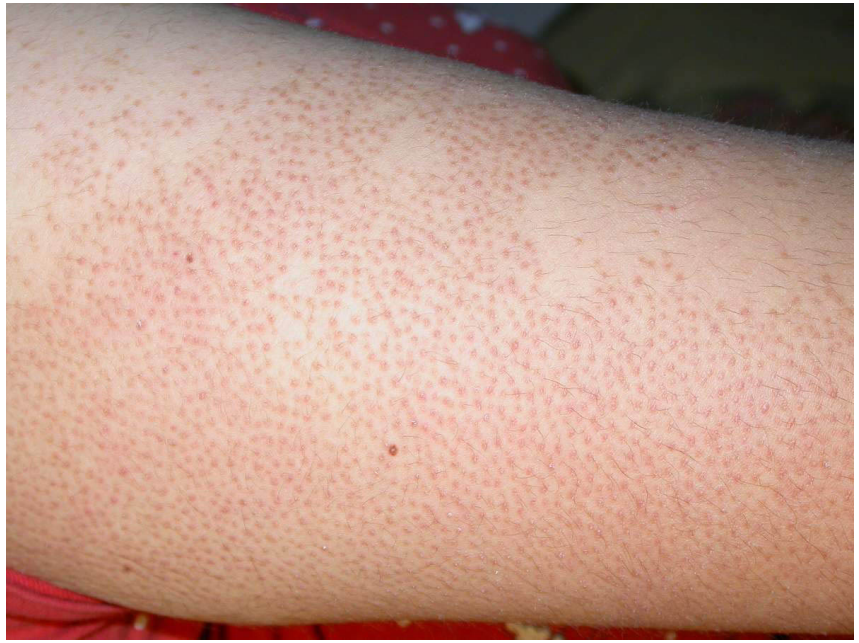






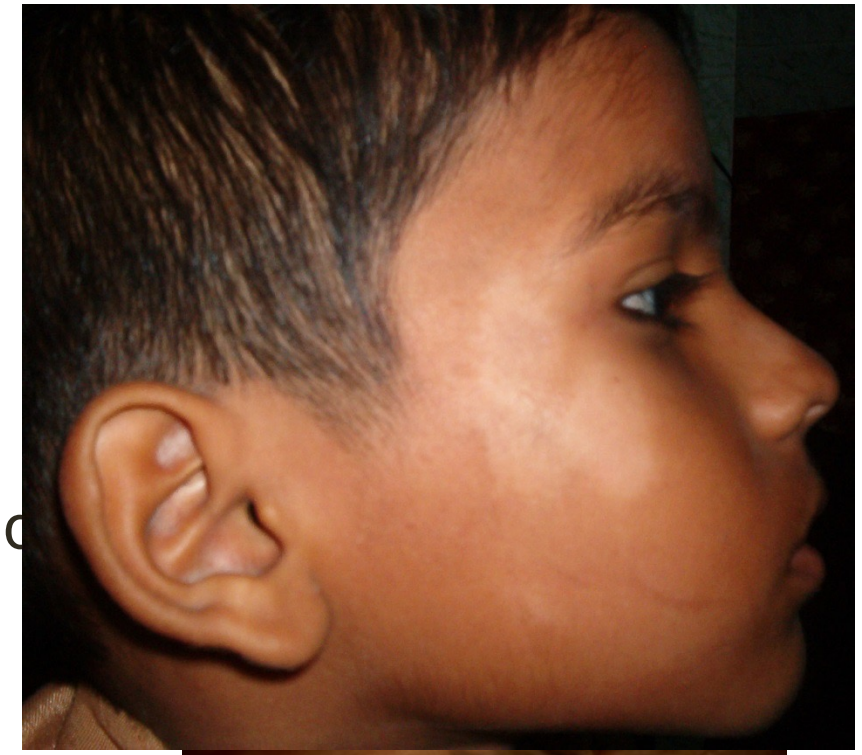
PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

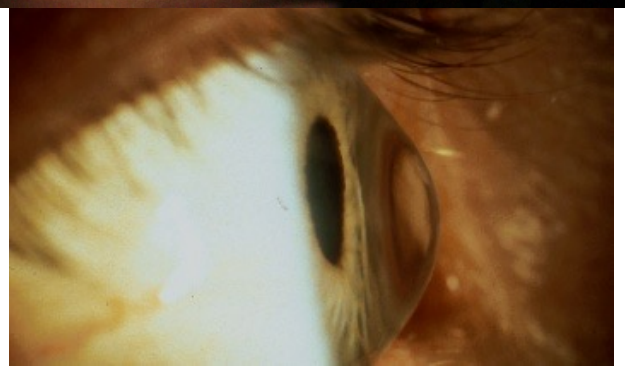
- Keratosis Pilaris
  - Excessive keratinization causing horny follicular plugs on upper arms, thighs, and cheeks
  - Associated with atopy



PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

- Pityriasis Alba
  - Hypopigmented patches with minimal scale
  - Maybe the only manifestation of AD
  - Complications: keratoconus, eyelic dermatitis, increase risk of infection





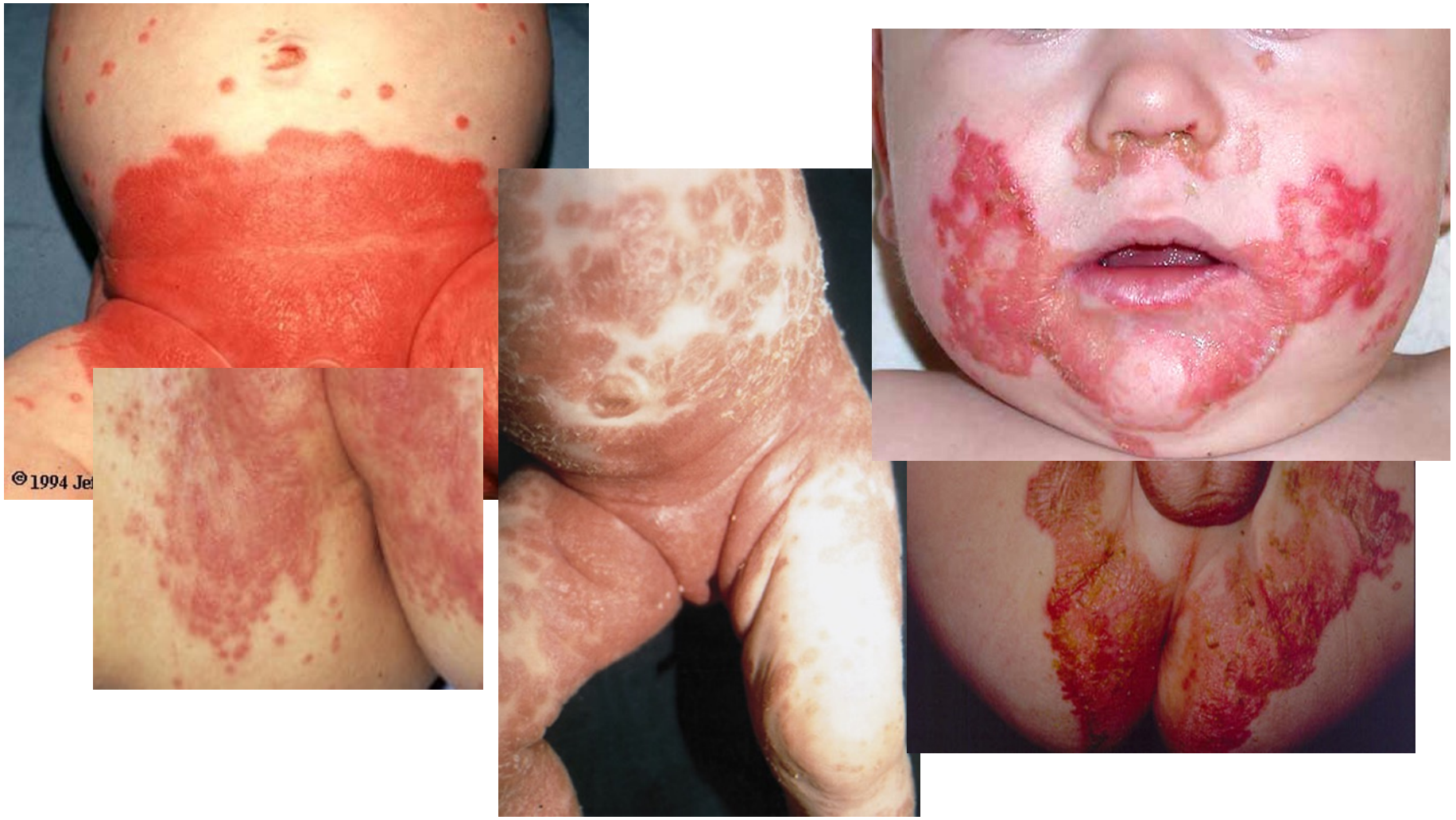
PAPULOSQUAMOUS AND ECZEMATOUS DERMATOSES

- Diaper Dermatitis- dermatitis in the confines of diaper area. Commonly caused due to
  - Overhydration of the skin
  - Maceration
  - Prolonged contact with urine and feces
  - Retained diaper soaps
  - Topical preparations
  - More than 3 diarrheal stools per day
  - Adverse effects of antibiotics
  - Early sign of Biotin deficiency
- Most common cause is Candida. (C. albicans-92%)

DIFFERENTIAL DIAGNOSIS FOR DIAPER DERMATITIS

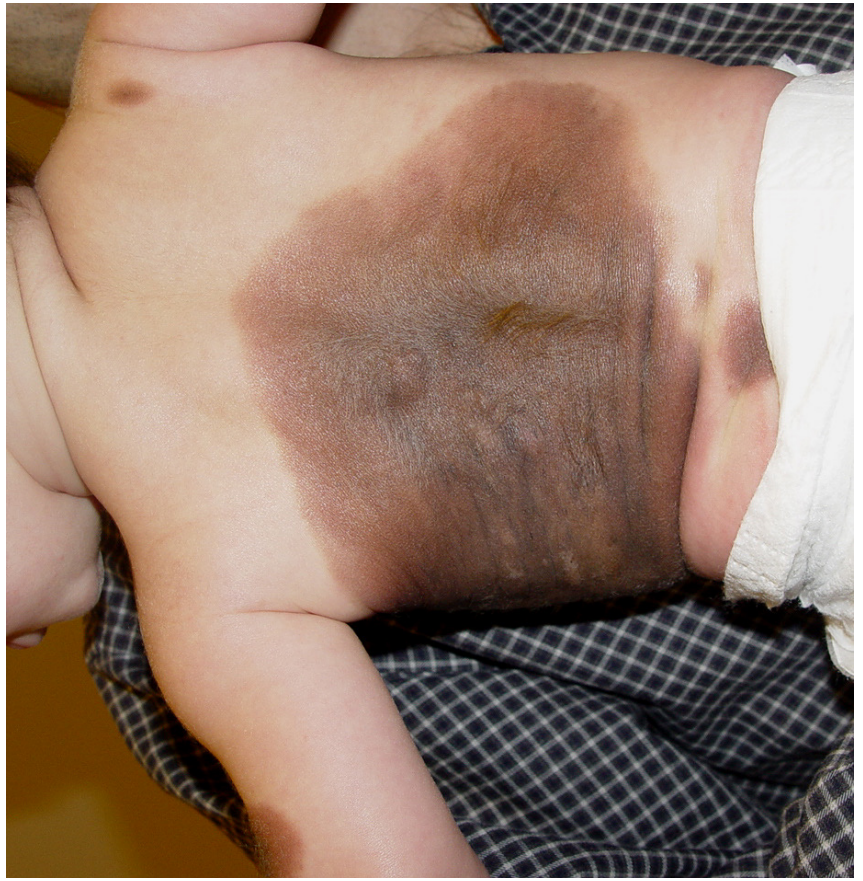
- Candidal Dermatitis
- Irritant Dermatitis
- Seborrheic Dermatitis
- Psoriasis
- Allergic Contact Dermatitis
- Atopic Dermatitis
- Miliaria
- Granuloma Gluteale Infantum
- Perianal Pseudoverrucous Nodules
- Cystic Fibrosis
- Acrodermatitis Enteropathica
- Multiple Carboxylase Deficiency
- Biotin Deficiency
- Langerhans Cell Histiocytosis
- Kawasaki Disease
- Perianal strep
- Bullous Impetigo
- Scabies
- Congenital syphilis

DIFFERENTIAL DIAGNOSIS FOR DIAPER DERMATITIS



PIGMENTED LESIONS

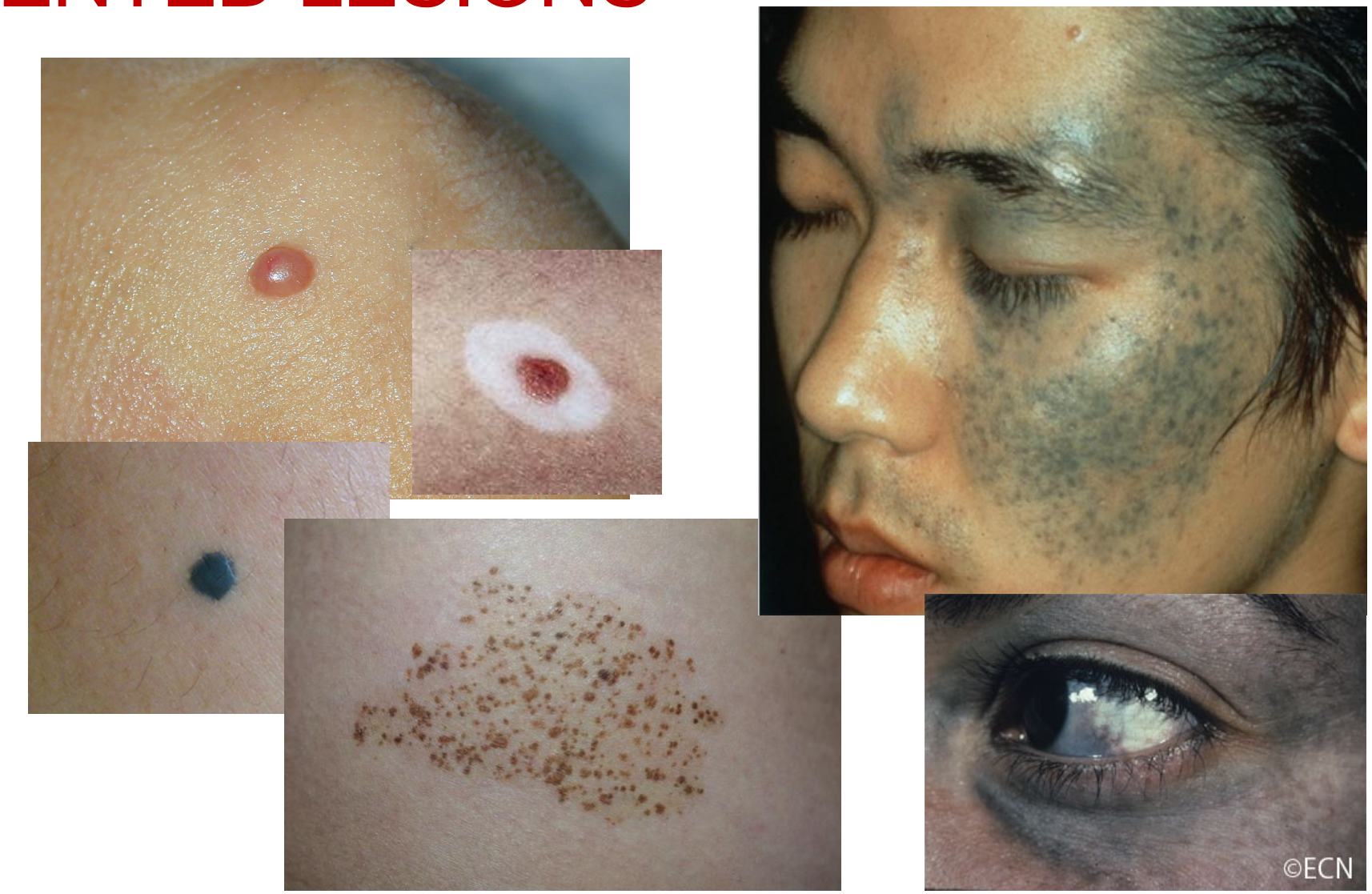
- Cafe au lait macules (CALM)
  - Dark brown macules and patch
  - > 5 or 1 lesion bigger than 1.5 cm : association with NF and McCune Albright syndromes
- Congenital Nevus
  - Onset at birth or 1st year
  - Small (<1.5 cm)
  - Medium (1.5-20 cm)
  - Large (>20cm
  - Increased risk of melanoma (3-12%)
  - On scalp r/o neurocutaneous melanosis



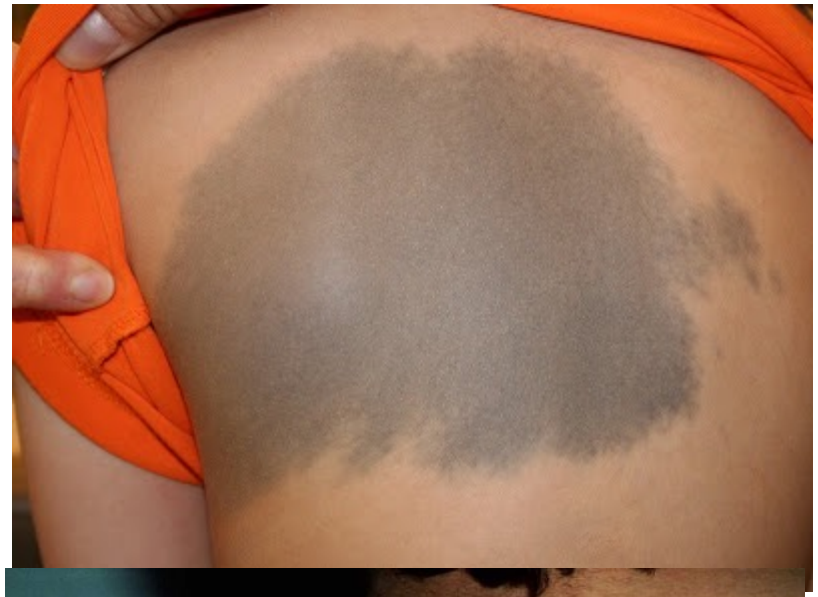
PIGMENTED LESIONS

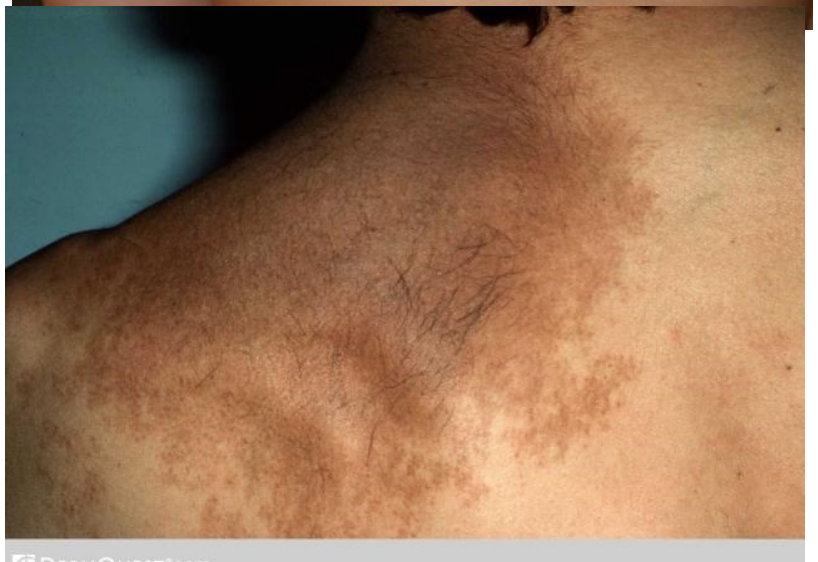
- Spitz nevus
  - Dome shaped tan papule occurs within 1st two decades
- Nevus spilus
  - Tan patch with scattered macules
- Halo nevus
  - Melanocytic nevus with surrounding hypopigmentation
  - May appear in vitiligo
  - r/o melanoma
- Blue Nevus
- Becker's Nevus
  - Smooth muscle hamartoma; increased number of hair and darkens at puberty
- Nevus of Ito
- Nevus of Ota
- Mongolian Spot

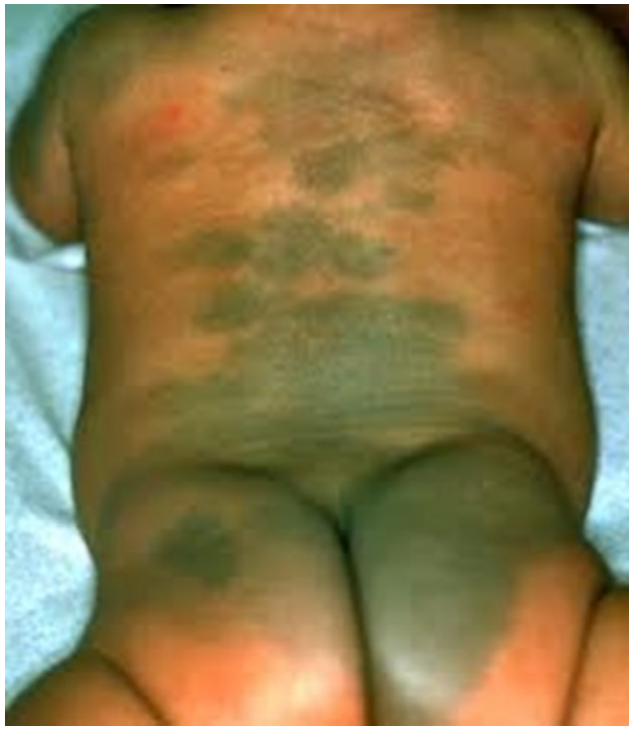
PIGMENTED LESIONS



PIGMENTED LESIONS

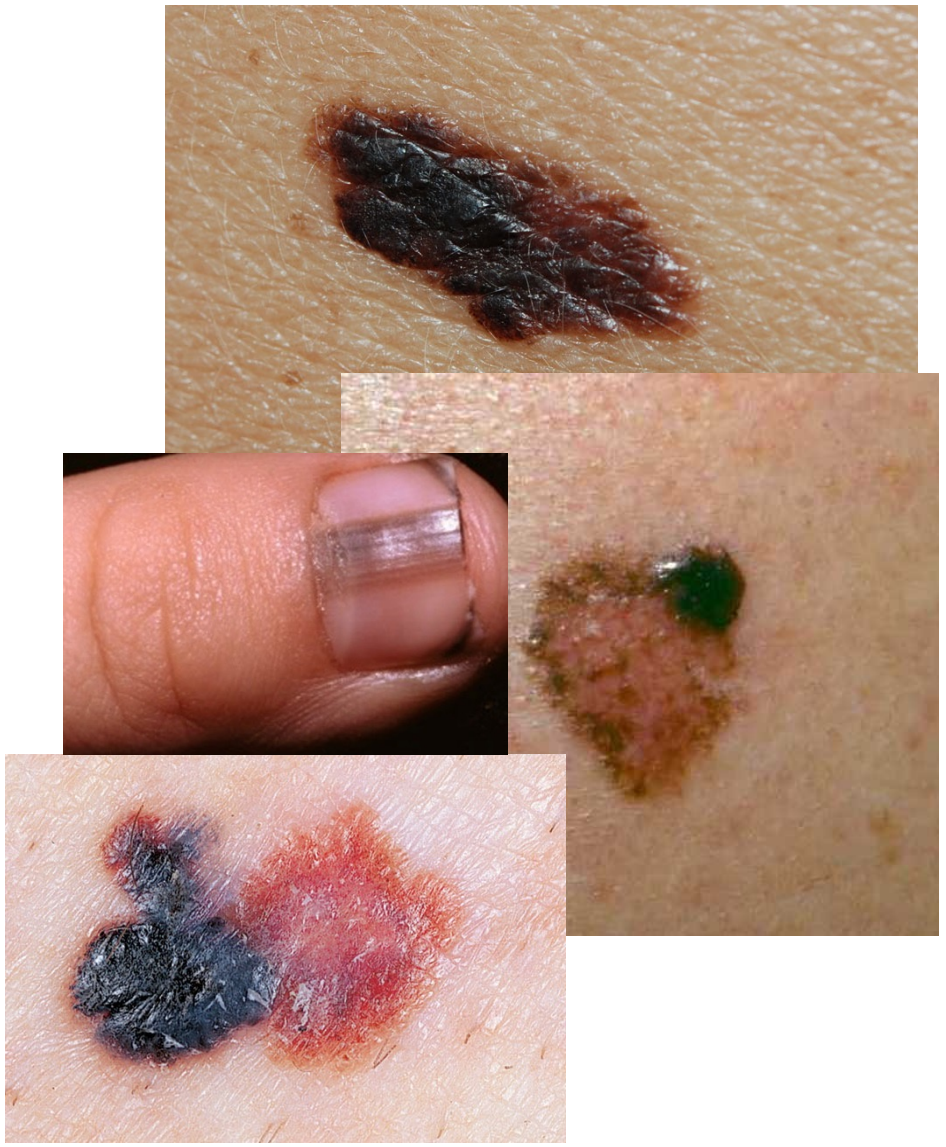




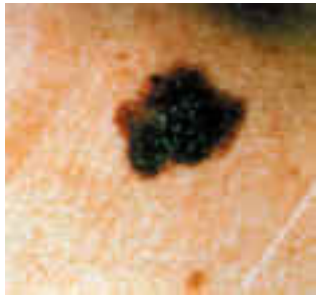


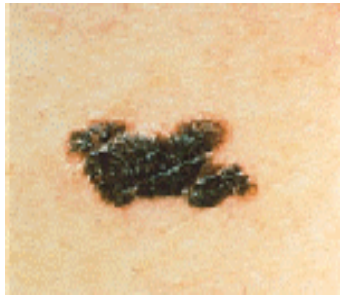
PIGMENTED LESIONS

- Melanoma
  - 0.3-0.4% of melanoma in prepubertal children
  - Increased risk with
    - fair skin, blue eyes, blonde/red hair,
    - CDKN2A or p16 mutation,
    - xeroderma pigmentosum,
    - dysplastic nevus syndrome,
    - large congenital nevus, or
    - neurocutaneous melanosis
    - Increased sunexposure/tanning beds

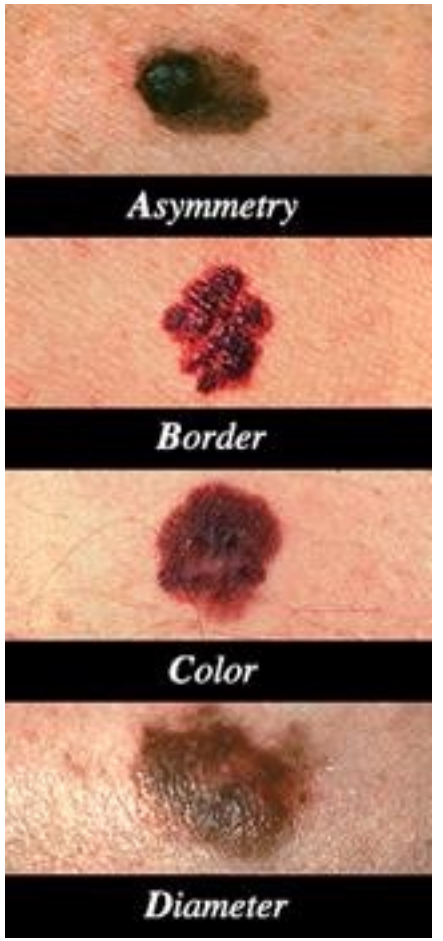


Identifying Melanoma

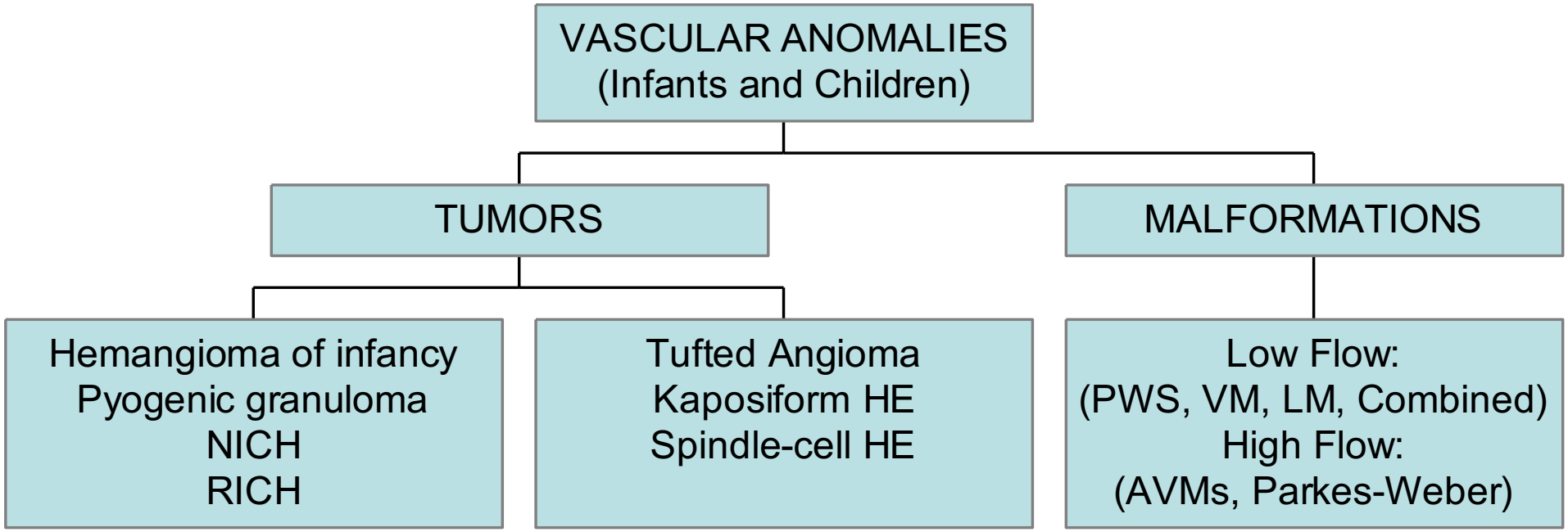




- The Skin Cancer Foundation and the American Academy of Dermatology recommend using the ABCDE method to help detect melanoma.

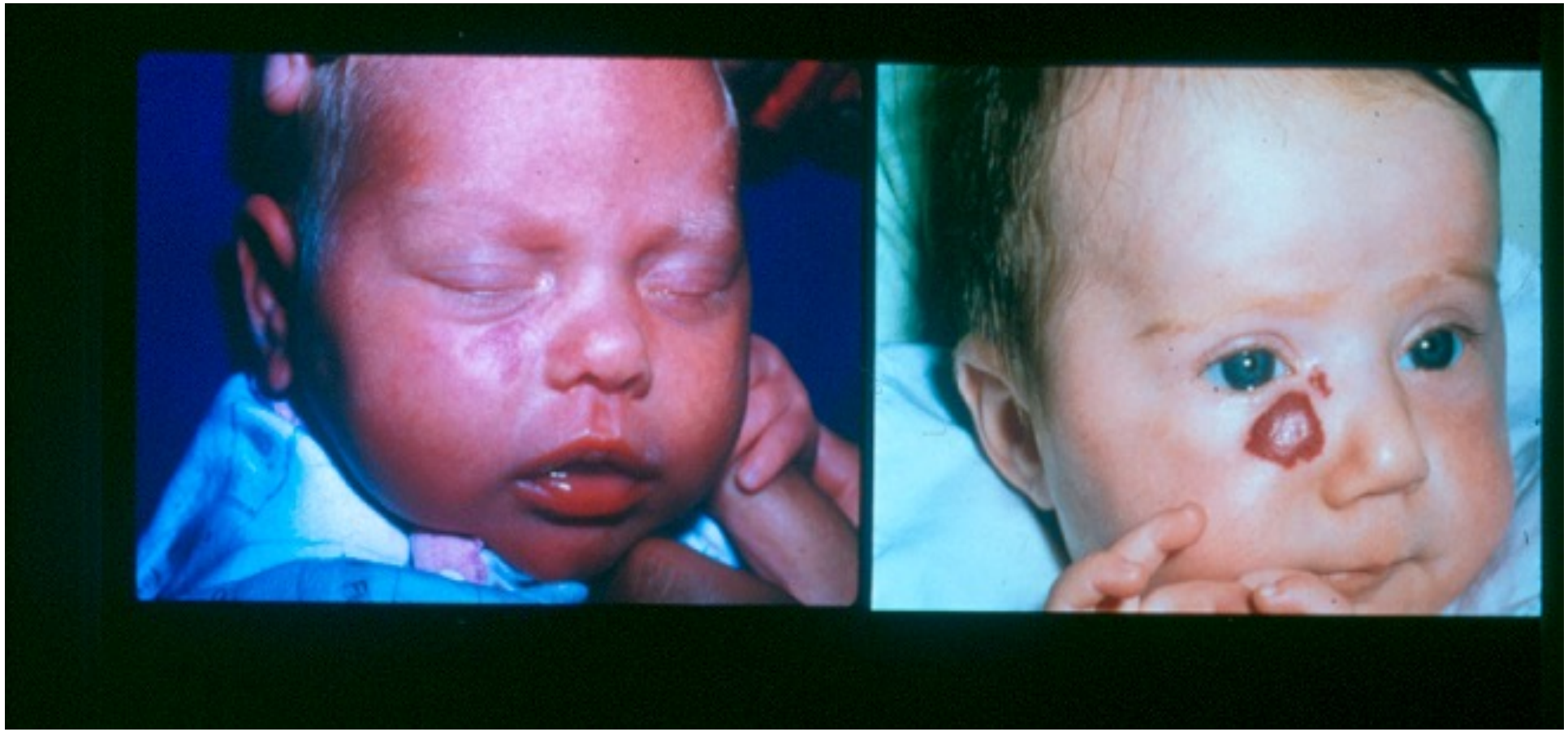


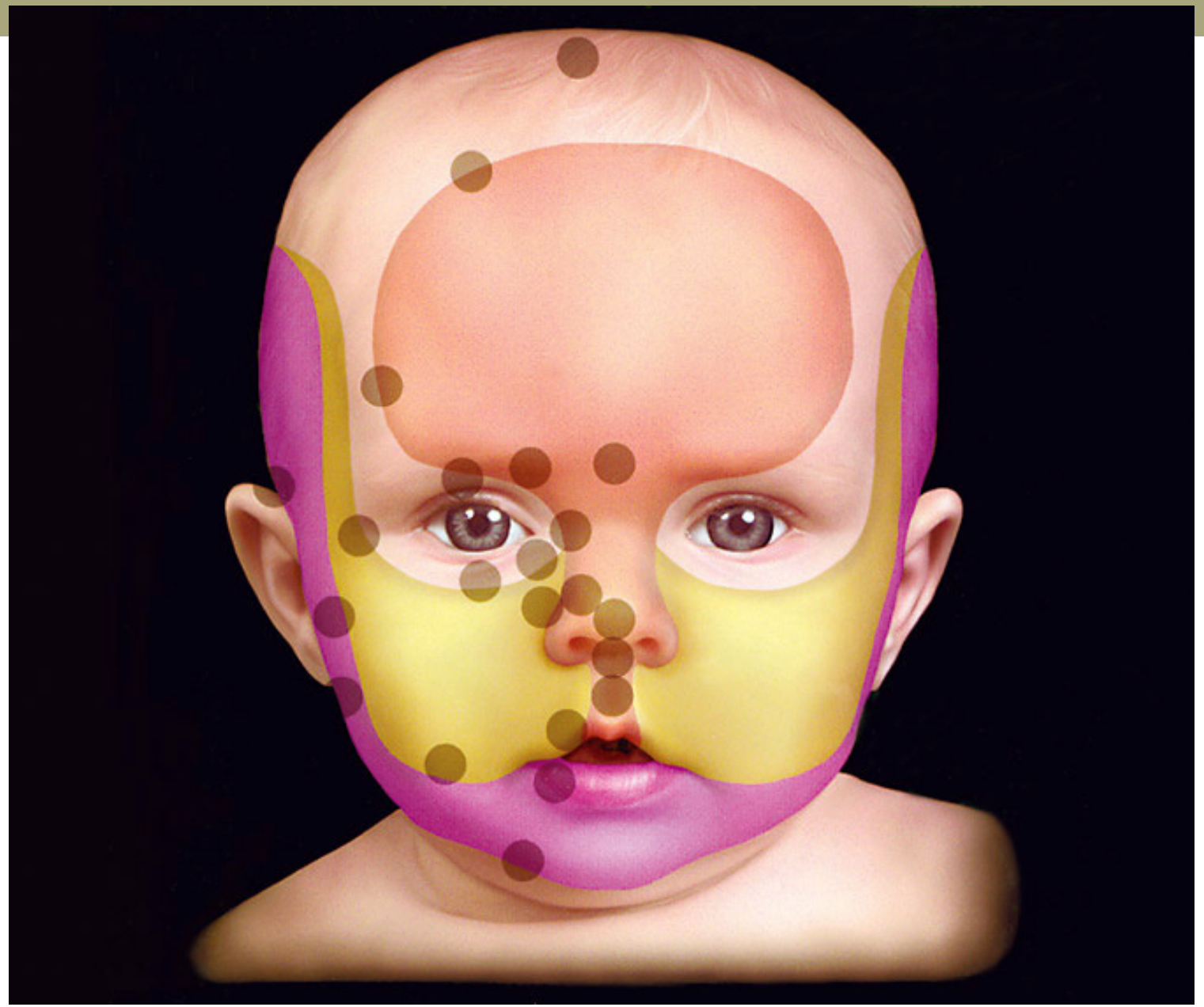
Vascular Anomalies:
Revised ISSVA Classification



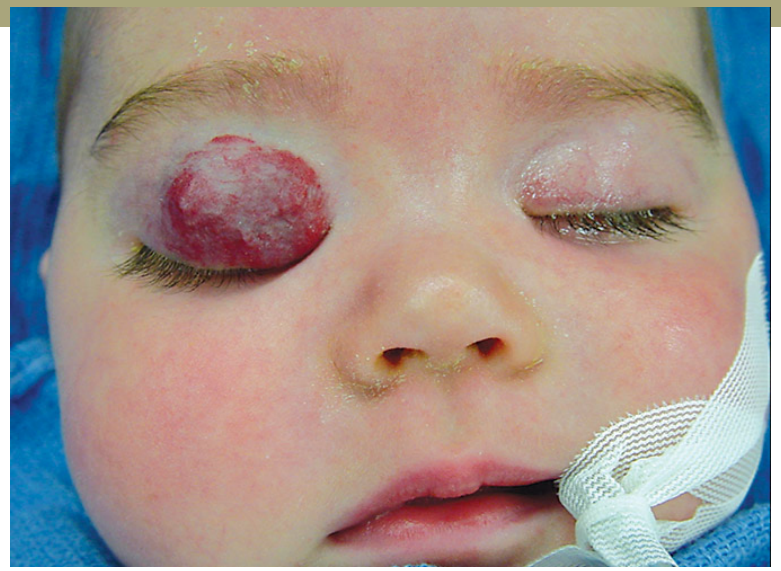
International Society for Study of vascular anomalies 1996

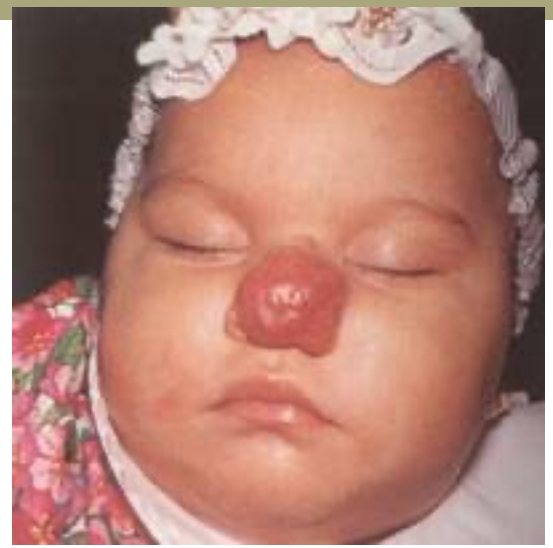
Hemangiomas of Infancy (HOI)

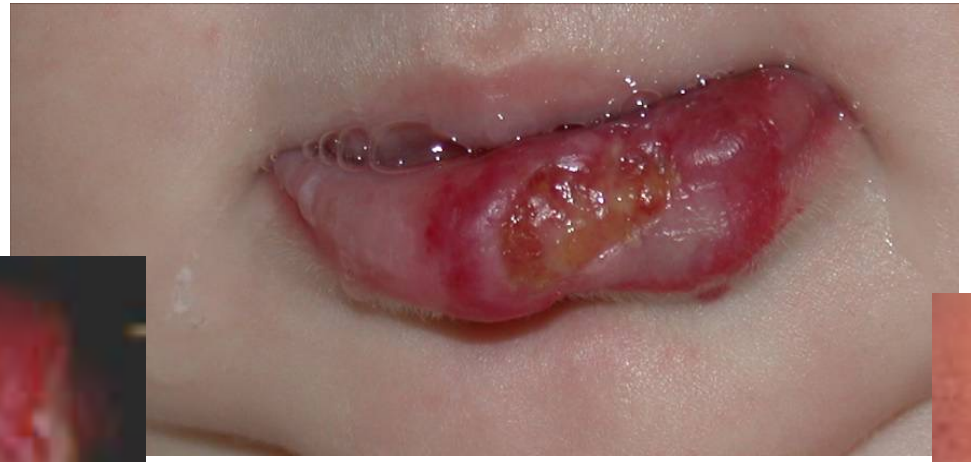


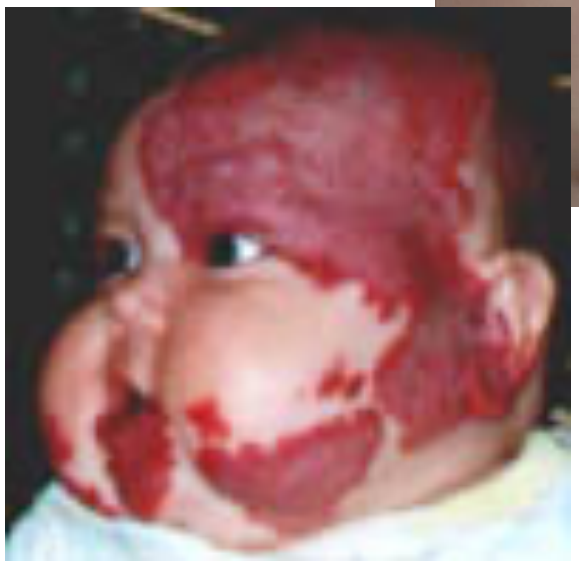


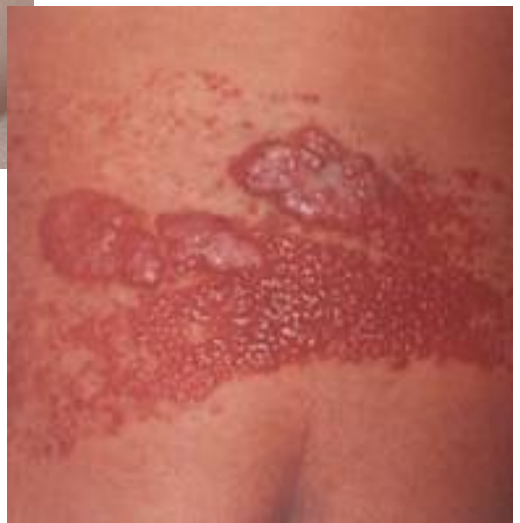
Predilection for embryologic fusion lines











Clinical Course:Hemangiomas (HOI)

- Up to 6-12m rapid rate of growth
- Growth complete by 12-18 months - **plateau phase**
- involution occurs slowly
- 10% rule / year
- 30% will involute by 3 y age
- 50% will involute by 5 y age
- Virtually all HOI are replaced by age 10-12

Risk Factors for Systemic Involvement

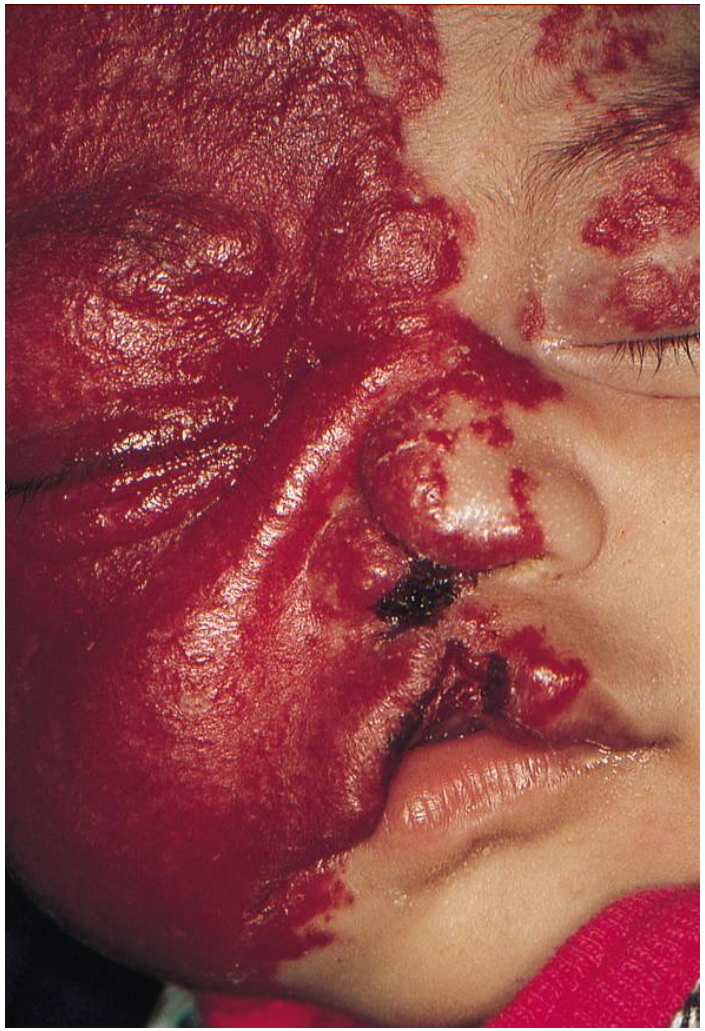
- Multiple hemangiomas
  - Need hepatic ultrasound, CBC, stool guaiac, serial exams
- Segmental hemangiomas
  - Facial need eye exam, echo, MRI/MRA of head
  - Lumbosacral need MRI of spine, GI/GU exam
- Beard hemangiomas
  - ENT evaluation, laryngoscopy

HOI vs. Congenital Hemangiomas

Congenital

- Rapidly involuting congenital hemangioma (RICH)
- Non-involuting congenital hemangioma (NICH)
- Both types stain differently from HOI
  - GLUT-1 negative
  - Merosin, Lewis Y antigen

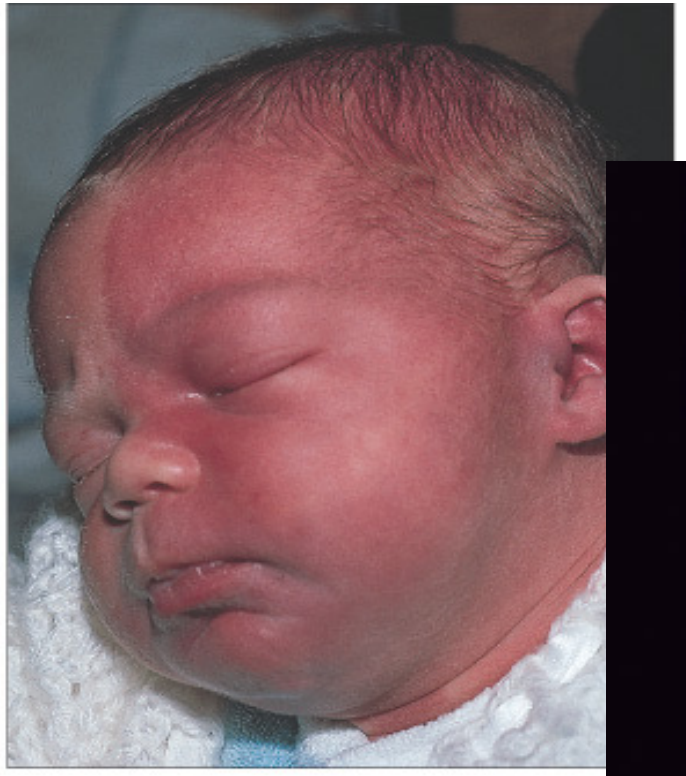
PHACES Syndrome

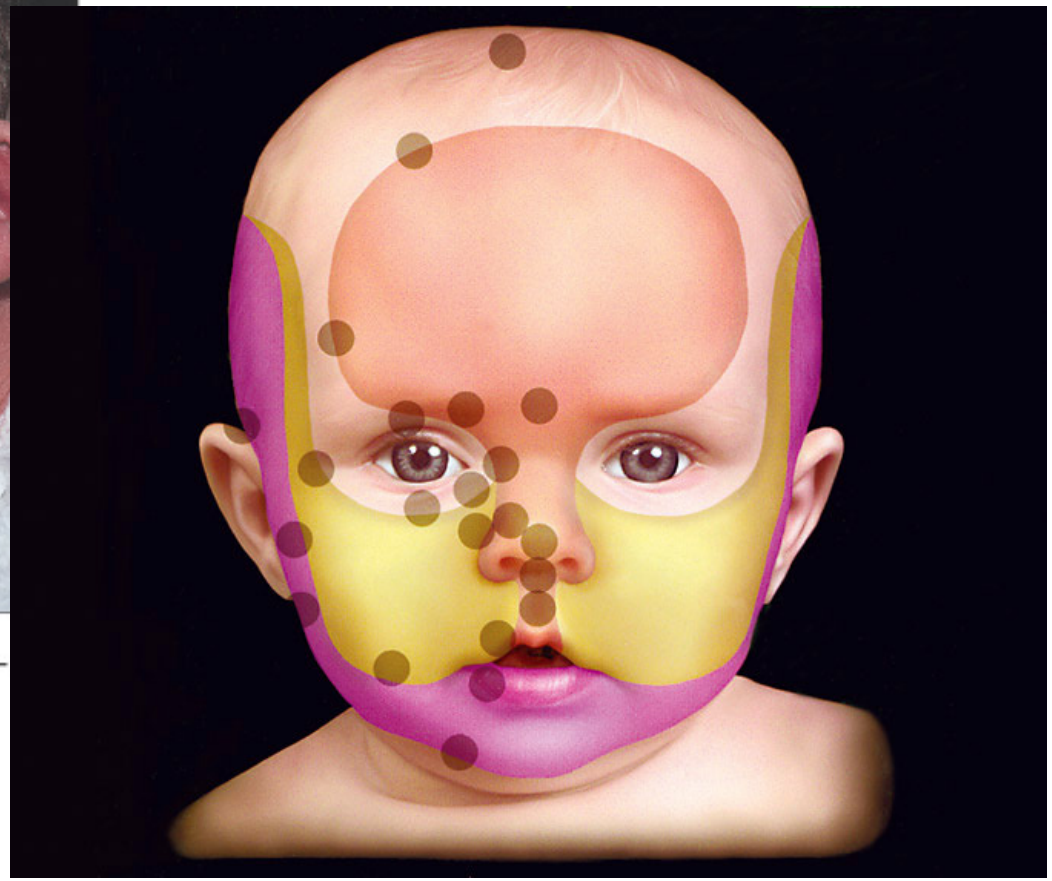


PHACES Syndrome

- P - posterior fossa malformations
  Dandy-walker malformation
- H - hemangiomas
  - Large and facial involving > 1 dermatome require
- A - arterial anomalies
  - Lacking or aneurysms of carotid vessels
- C - coarctation of the aorta and cardiac defects
- E - eye abnormalities
  - Cataracts, optic nerve hypoplasia
- S - sternal clefting and supraumbilical abdominal raphe

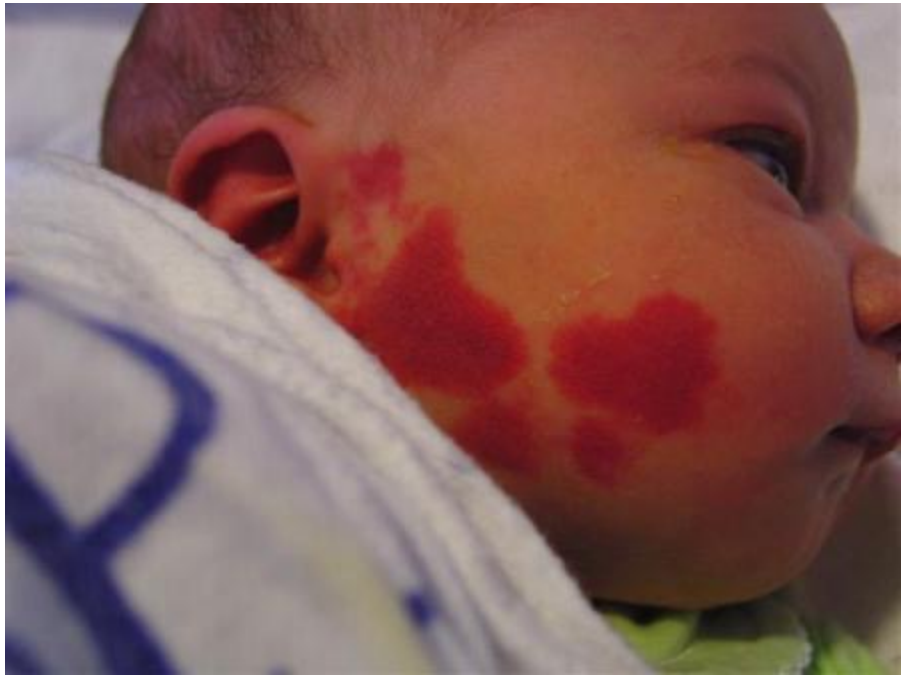
PWS-Port wine stain





STURGE-WEBER Syndrome

Ophtalmic distribution of the trigeminal nerve



-Tram-track malformation in brain (seizure)
-Glaucoma/cataracts

Klippel-Trenaunay Syndrome

- PWS involving a limb
- Underlying venous varicosities
- Soft tissue hypertrophy
- Bone overgrowth

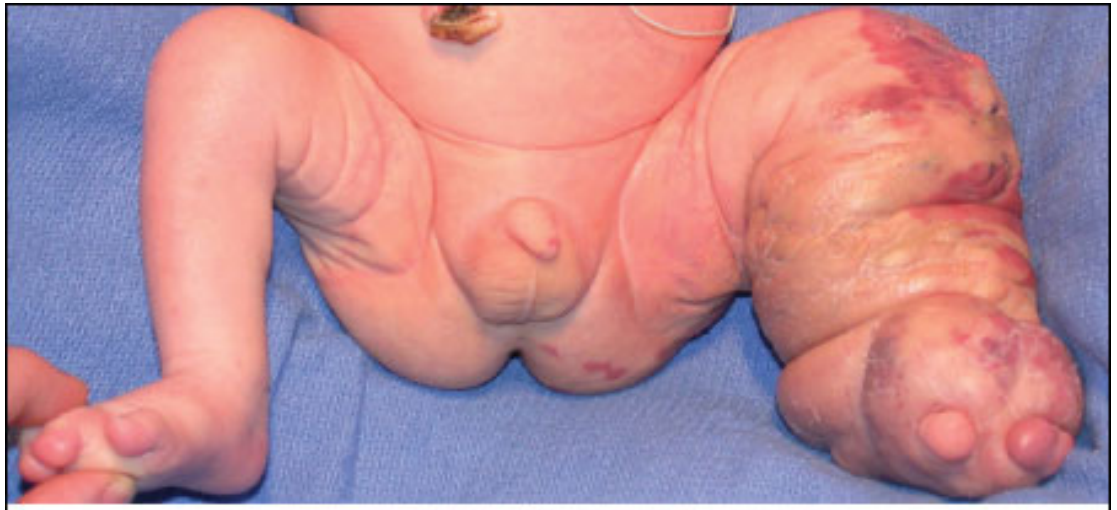
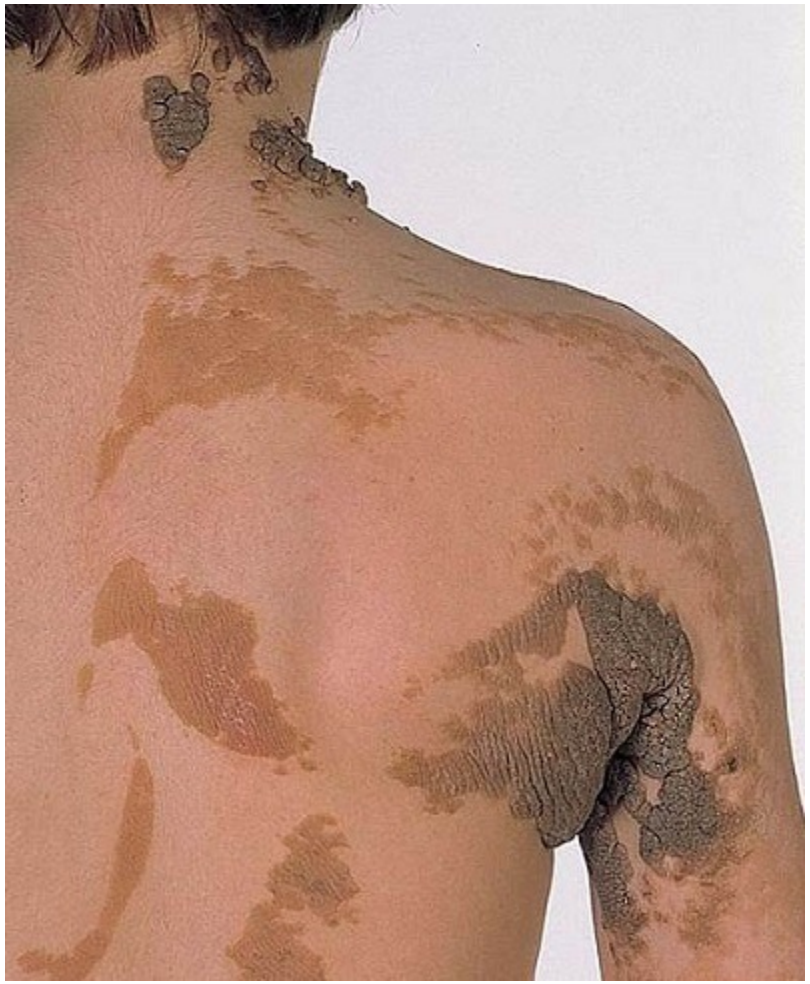


Figure 1

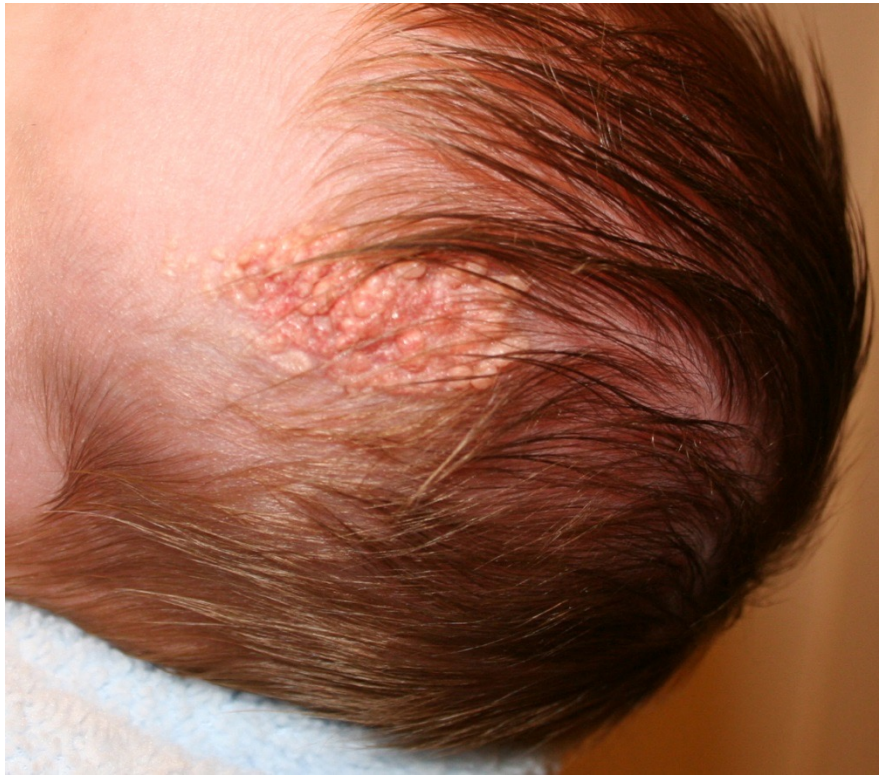
EPIDERMAL, APPENDAGEAL AND DERMAL TUMORS

- Epidermal Nevus (EN)
  - Onset: typically at birth
  - Hamartoma of epidermis and papillary dermis
  - Presnts as hyperpigmented verrocous plaques in Blaschko's lines
  - ILVEN: inflammatory
  - Epidermal Nevus synd (Schimmelpenning sydrome)Underlying CNS, ocular, cardiac and skeletal defects



EPIDERMAL, APPENDAGEAL AND DERMAL TUMORS

- Nevus Sebaceous
  - Presents as solitary yellow-orange plaque typically on the scalp
  - Mutation in PTCH gene
  - Associated with benign tumors (trichoblastoma, syringocysadenoma) and malignant tumors (BCC)

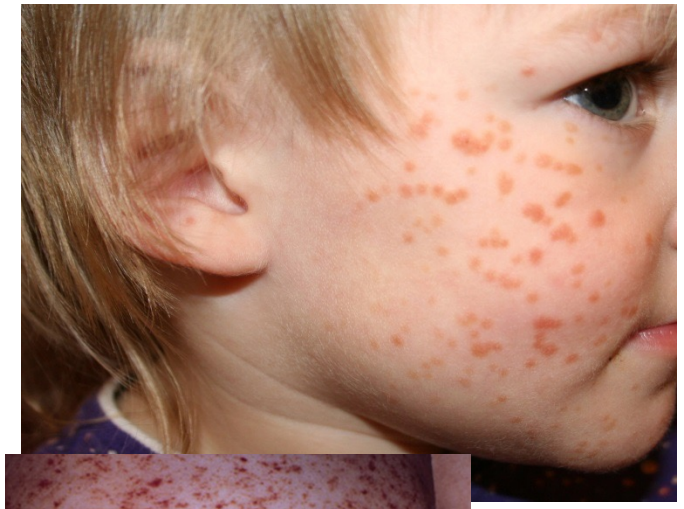


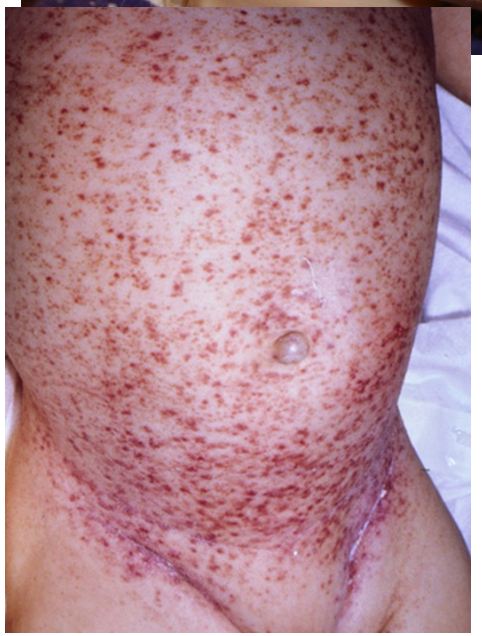
EPIDERMAL, APPENDAGEAL AND DERMAL TUMORS

- Langerhans Cell Histiocytosis
  - Clonal proliferative dz of Langerhans cell (comma shaped nuclei, S100+, CD1a+, intracytoplasmic Birbeck granules seen on EM)
  - Current classification based on the organ systems involved. Historically divided into 4 groups
- Letterer-Siwe Disease
  - Multisystem involvement
  - Acute disseminated form
  - Onset before 2 yrs of age
  - Small, pink papules, pustules, vesicles with scale/crust/petechiae in seborrheic distribution
- Hand-Schuler-Christian Disease
  - Onset between 2-6 years of age
  - Typical triad: diabetes insipidus, bone lesions, exophthalmos
  - Osteolytic bone lesions (cranium)

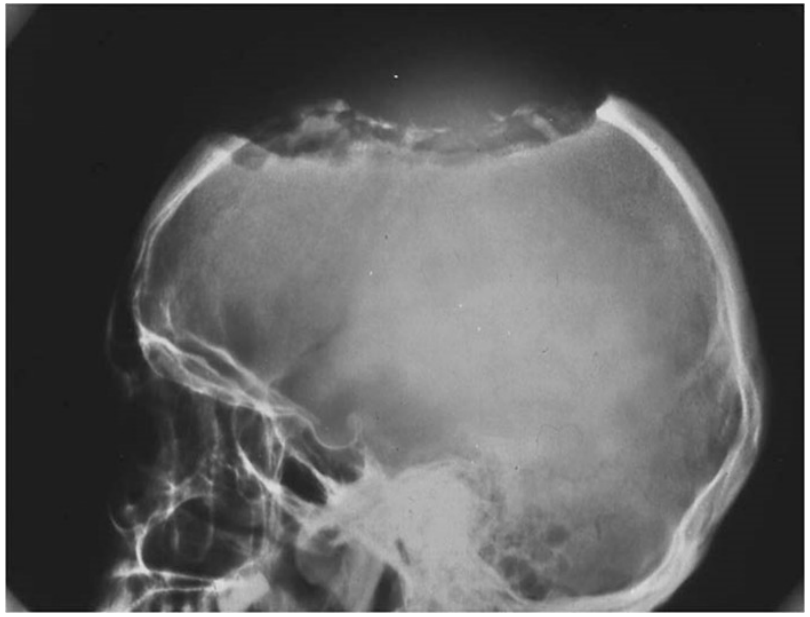
EPIDERMAL, APPENDAGEAL AND DERMAL TUMORS

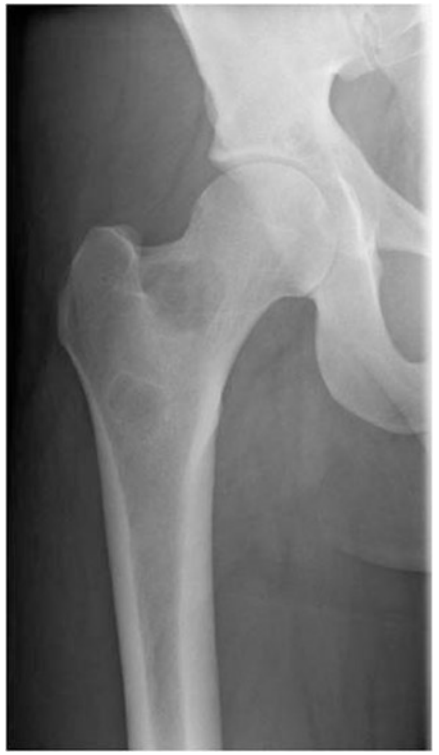
- Eosinophilic Granuloma
  - Onset in older children
  - Localized LCH variant
  - Asymptomatic granulomatous lesions involving bone (cranium), spontaneeous fracture
- Congenital Self-Healing Reticulohistiocytosis
  - Aka Hashimoto-Pritzker dz
  - Onset at birth or soon after
  - Limited to the skin- widespread , red-brown papulonodules
  - Self healing within weeks to months

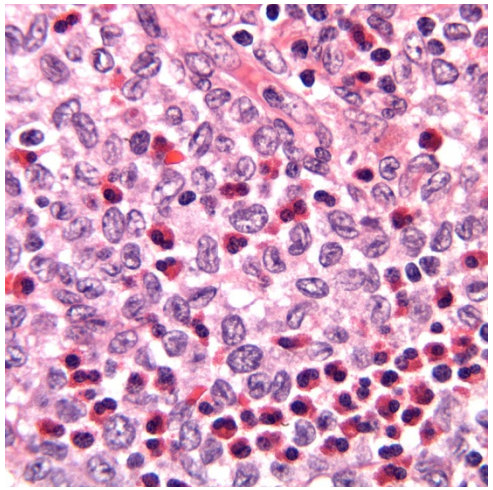


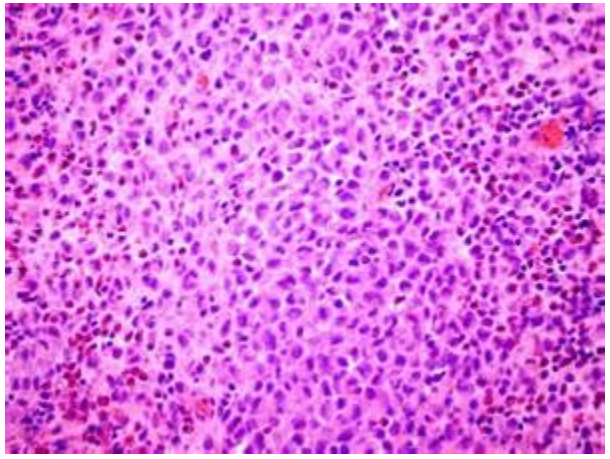


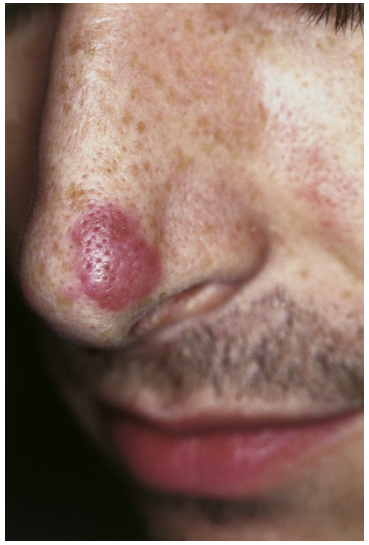
EPIDERMAL, APPENDAGEAL AND DERMAL TUMORS









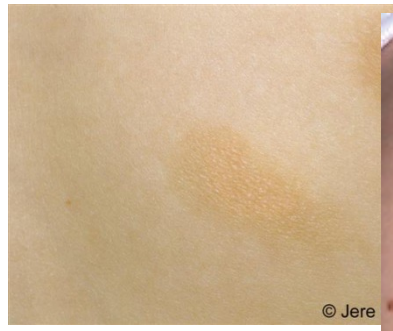


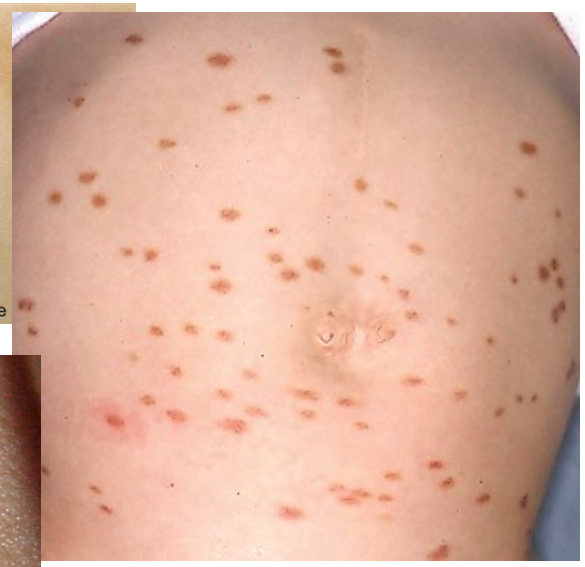
EPIDERMAL, APPENDAGEAL AND DERMAL TUMORS

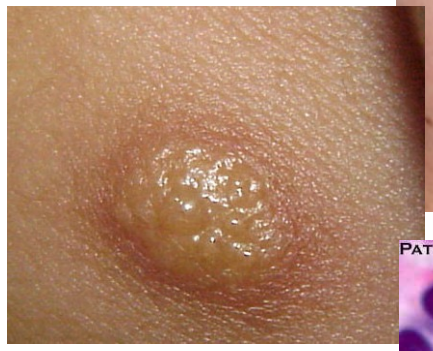
- Mastocytosis
  - d/o of mast cell hyperplasia
  - Childhood- onset before puberty (50% before age 2)
  - C-kit alteration (proto-oncogene, tyrosine kinase subfamily
  - Avoid mast cell degranulators: asprin, alcohol, quinine, opiates, polymixin B sulfate, amphotericin B, tubocuraine, scopolamine
- Solitary Mastocytoma
  - Tan to brown minimally infiltrated plaque or nodule
  - Spontaneous resolution
  - +Darrier's sign
- Urticaria Pigmentosa (UP)
  - Onset early childhood , may occur in adults
  - Hyperpigmentes to pink pruritic macules or papules on the trunk
  - +Darrier's sign
  - Variant: Bullous UP

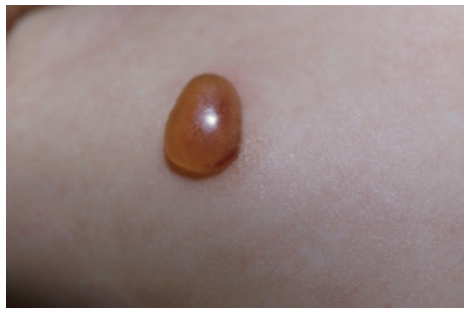
EPIDERMAL, APPENDAGEAL AND DERMAL TUMORS

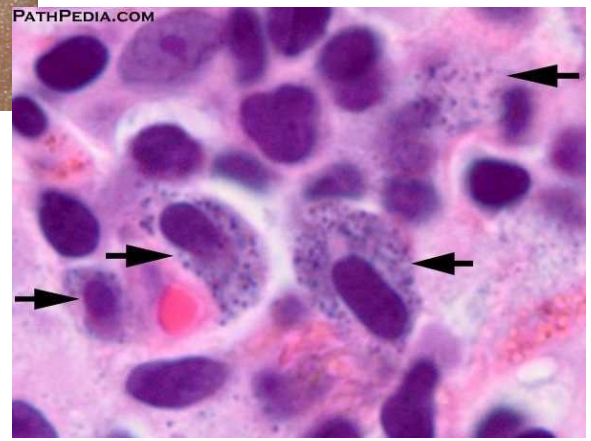
- Diffuse Cutaneous Mastocytosis
  - Doughy or boggy skin texture with lichenification and yellow hue
  - Extreme pruritus, friction may cause bullae
  - Sys sign: bronchospasm, diarrhea
- Telengiectasia Macularis Eruptiva Perstans (TMEP)
  - Persistent eruption of macules and papules with red-brown hue and telengiectasia
  - Rare in childhood





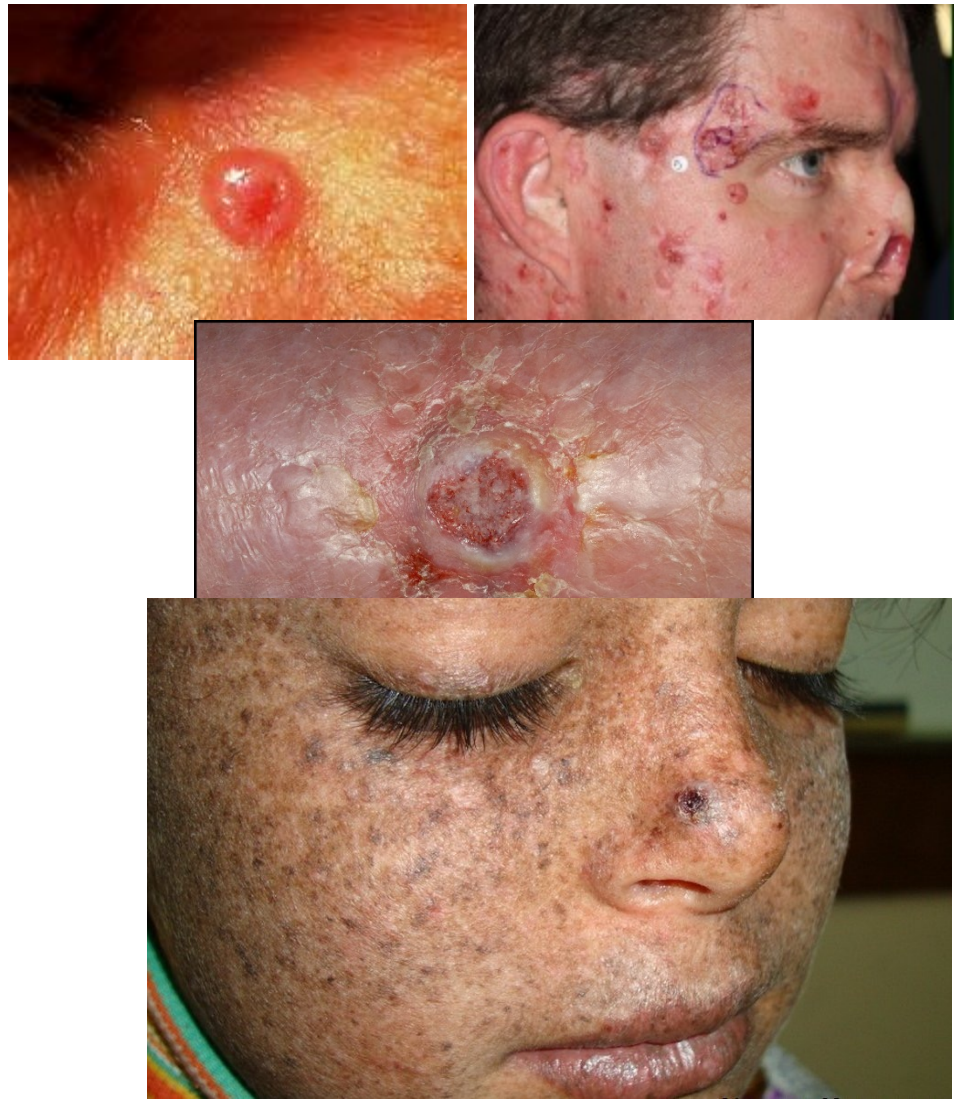






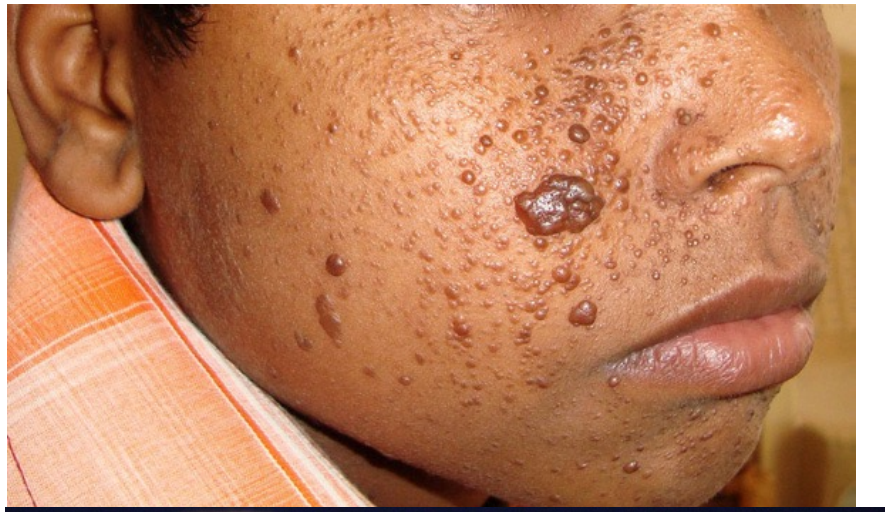
EPIDERMAL, APPENDAGEAL AND DERMAL TUMORS

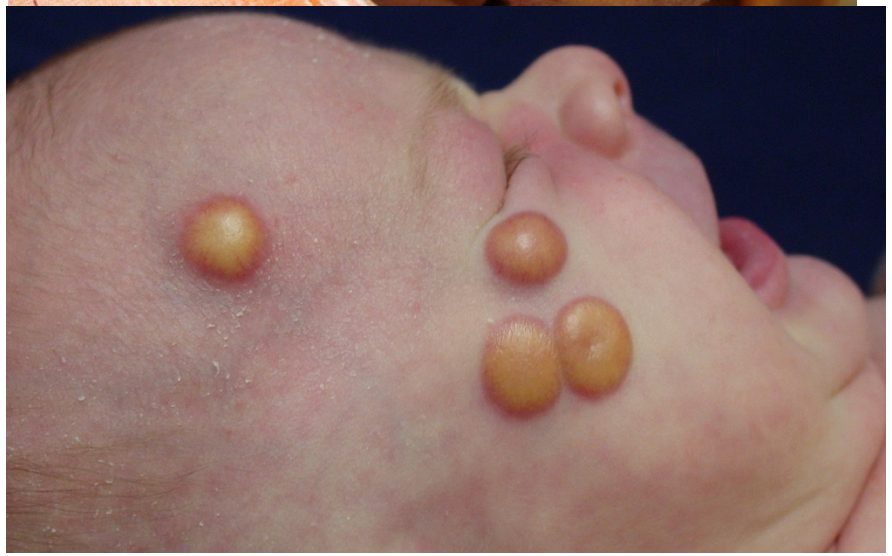
- Basal cell cancer
  - In children with Xeroderma pigmenatosum(XP), Basal Cell Nevus syndrome(Gorlin Syndrome)
- Squamous Cell Cancer
  - In children with XP, dystrophic EB, and albinism



EPIDERMAL, APPENDAGEAL AND DERMAL TUMORS

- Angiofibroma
  - In tuberous sclerosis
- Dermoid Cysts
  - Subcutaneous nodules seen along embryonic fusion planes commonly around eyes or nasal root
- Juvenile Xanthogranuloma
  - Pink-red nodules with yellowish hue on head/neck >trunk/ext
  - 2 types, regress in children
  - 0.5% ocular involvement
  - Assoc with NF-1 and juvenile myelomonocytic leukemia



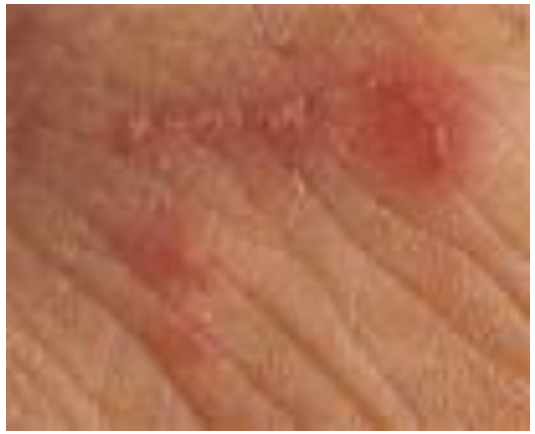


CHILDHOOD INFECTIOUS DISEASE

- Acute Hemorrhagic Edema of Infancy (Finkelsteins disease)
- Erythema Infectiosum (slapped cheek or 5th dz)
- Gianotti-Crosti Syndrome
- Hand-Foot-Mouth Disease
- Henoch-Schonlein Purpura (HSP)
- Herpangina
- Kawasaki Disease (Mucocutaneous Lymph Node Syndrome)
- Measles (Rubeola/1st Dz)
- Infectious Mononucleosis
- Papular Purpuric Gloves and Socks Syndrome)
- Roseola (exanthem Subitum/6th Dz)
- Rubella (German Measles or 3rd Dz)
- Scarlet Fever (2nd Dz)
- Unilateral Laterothoracic Exanthem
- Varicella (Chickenpox)

Scabies

- Papules with Silvery lines known as burrows on hands, flexor surfaces of wrists and genitalia
- Pruritis worst at night (during 1st infection)
- Dx scraping
- TX:
  -
  - 8- to 14-hour application of 5% permethrin cream
  - gamma benzene hexachloride lotion
  - 10% crotamiton cream
  - Ivermectin
- All family members and close contacts must be treated



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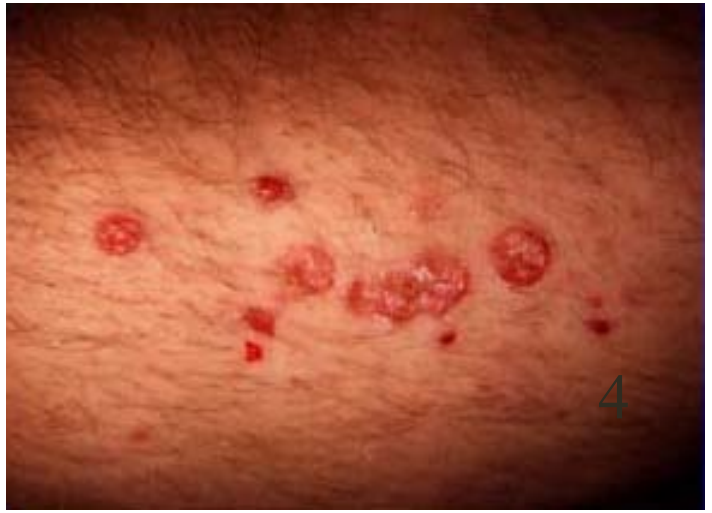
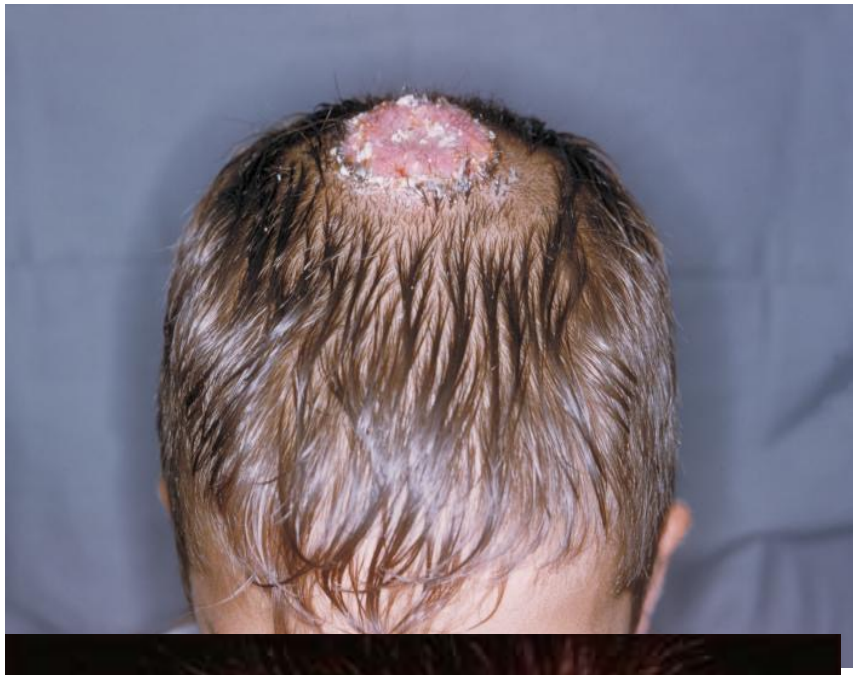
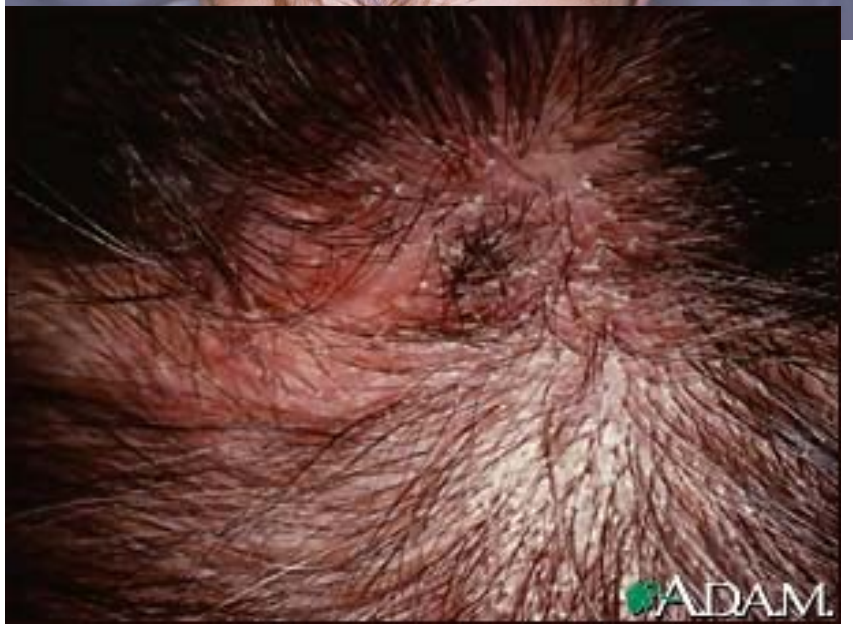


4

Tinea Capitis

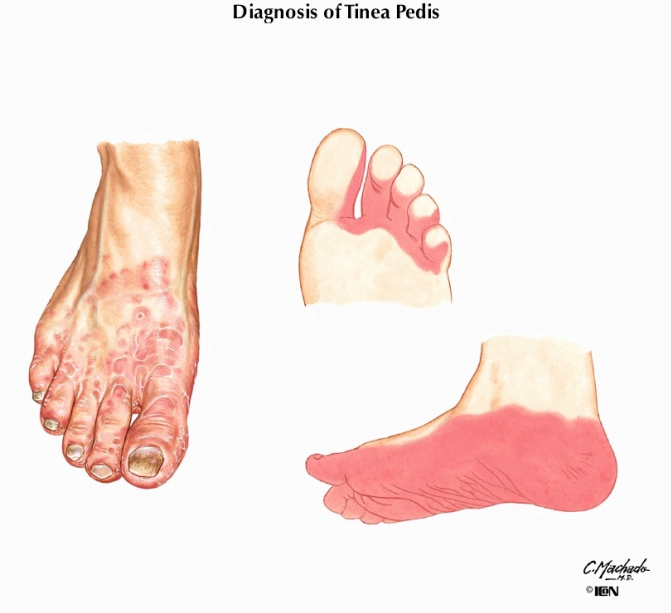
- *Trichophyton tonsurans* 90% of cases
- Varies considerably in clinical appearance
- “Black-dot“ appearance , combination of inflammation with hair breakage and loss.
- Kerion highly inflamatory presentation
- Griseofulvin PO 10-25mg/kg (6weeks)
- Terbinafine 3-6mg/kg/day (4 weeks)





Tinea Corporis and Pedis

- Itchy, red, circular lesion
- A ring that spreads along its borders and is clearing in the center



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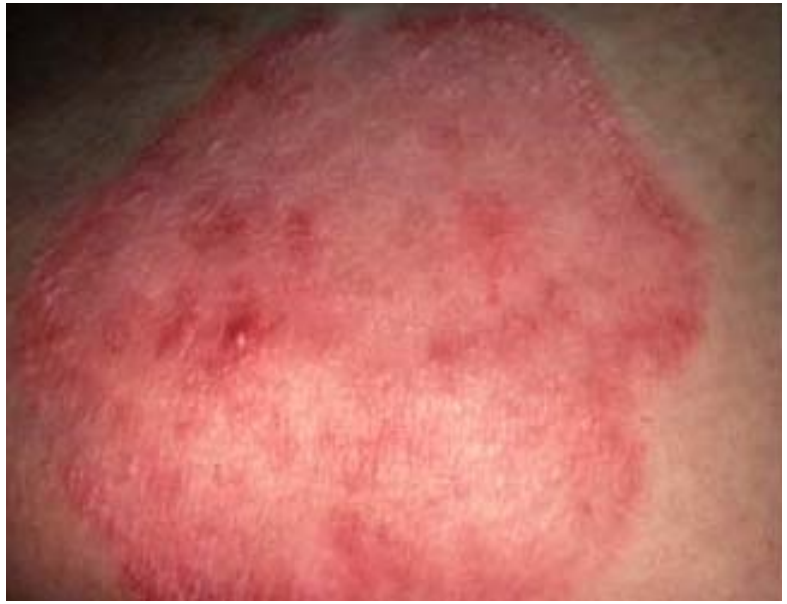
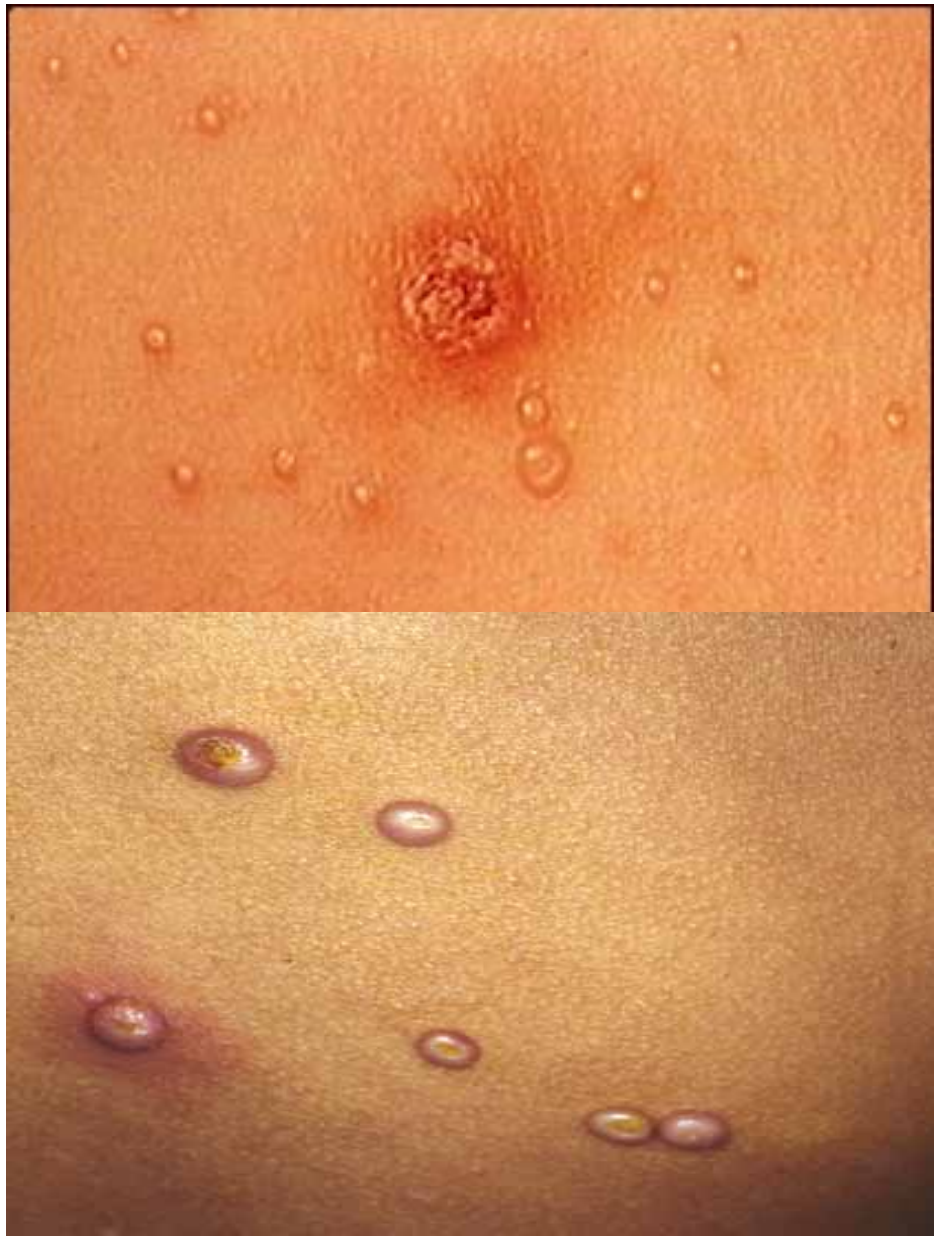


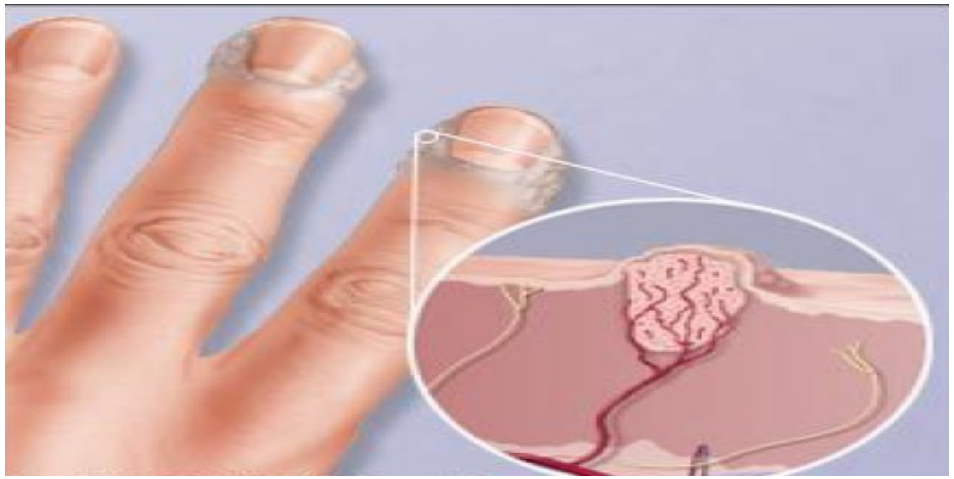
5

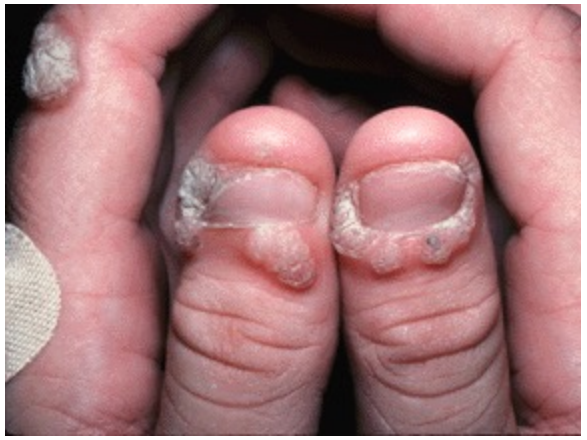
Molluscum Contagiosum

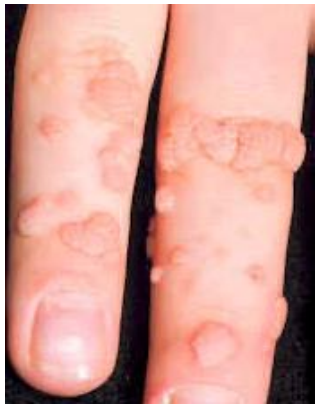
- Poxvirus
- Last 12-18 m before resolution
- Translucent, umbilicated white papules
- Curettage,freezing
- Imiquimod
- Retinoids

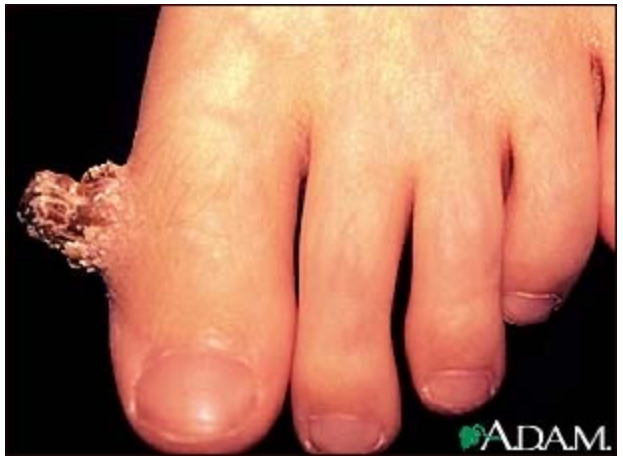


Verruca Vulgaris- HPV









CONGENITAL INFECTIONS OF THE NEWBORN

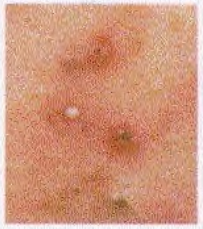
- Cytomegalovirus (CMV)
- Herpes Simplex Virus(HSV)
- Rubella
- Toxoplasmosis
- Varicella
- Syphilis

ACNE

TYPES OF LESIONS

Noninflammatory lesions

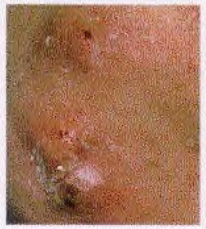
Closed comedones



Open comedones

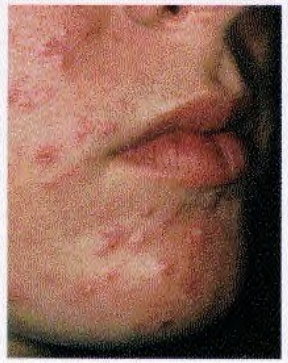
Inflammatory lesions

Papules/pustules

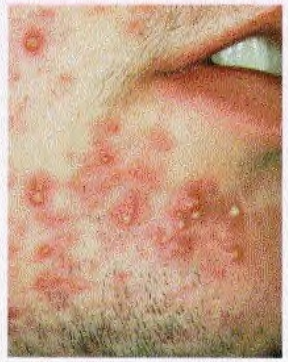


Nodules

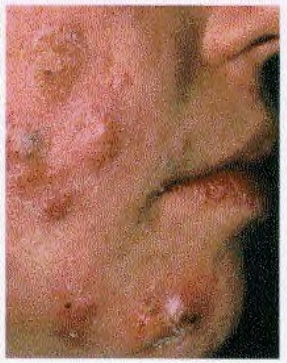
ACNE CLASSIFICATION AND GRADING



Mild
Papules/pustules +/++
Nodules 0



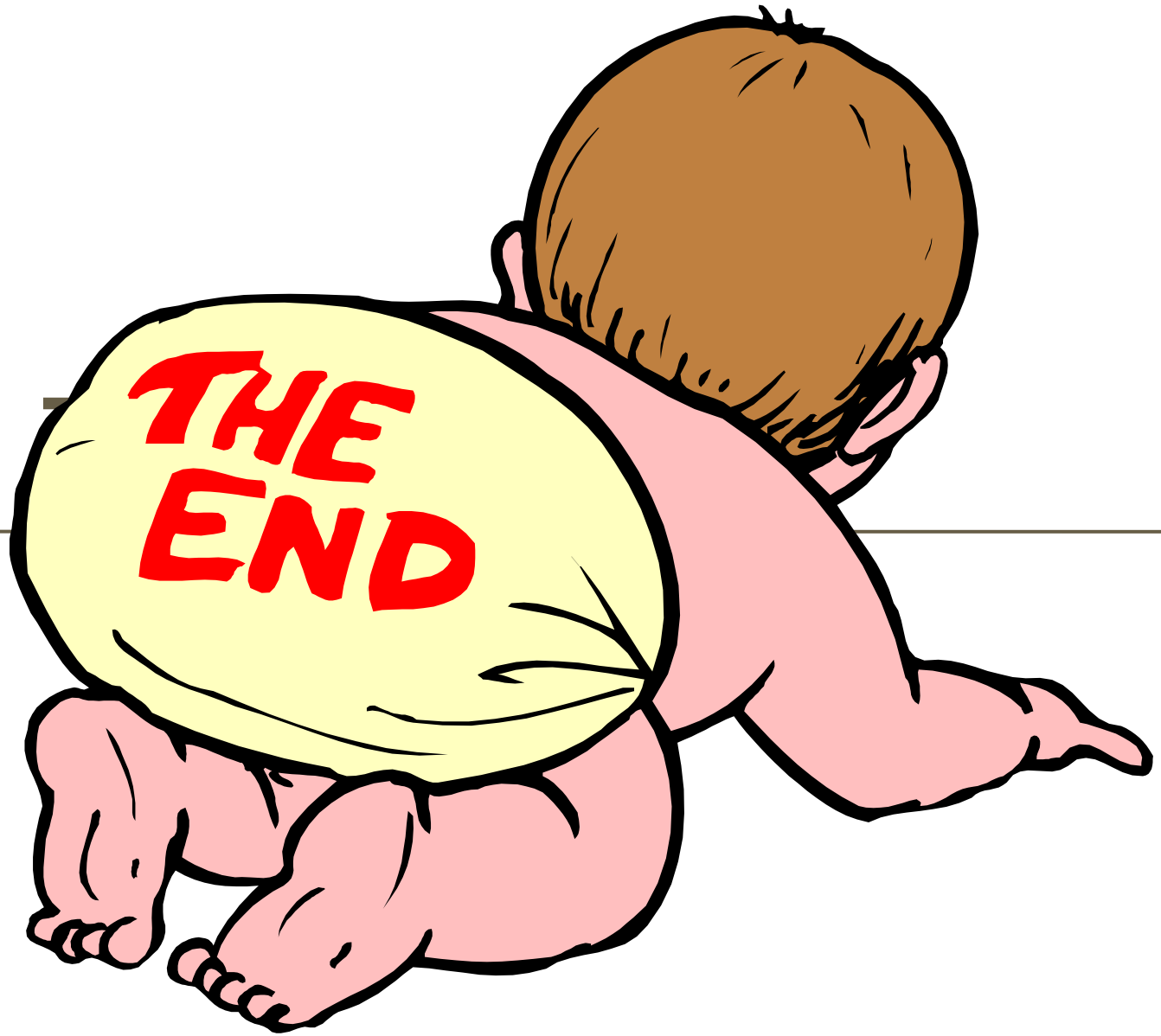
Moderate
Papules/pustules ++/+++
Nodules +/++



Severe
Papules/pustules +++/++++
Nodules +++

SEVERITY GRADING OF INFLAMMATORY LESIONS

Severity	Papules/pustules	Nodules	Additional factors that determine severity
Mild	Few to several	None	Psychosocial circumstances
Moderate	Several to many	Few to several	Occupational difficulties
Severe	Numerous and/or extensive	Many	Inadequate therapeutic responsiveness


THE
END