NEONATAL DERMATOLOGY

Advanced Desert Dermatology

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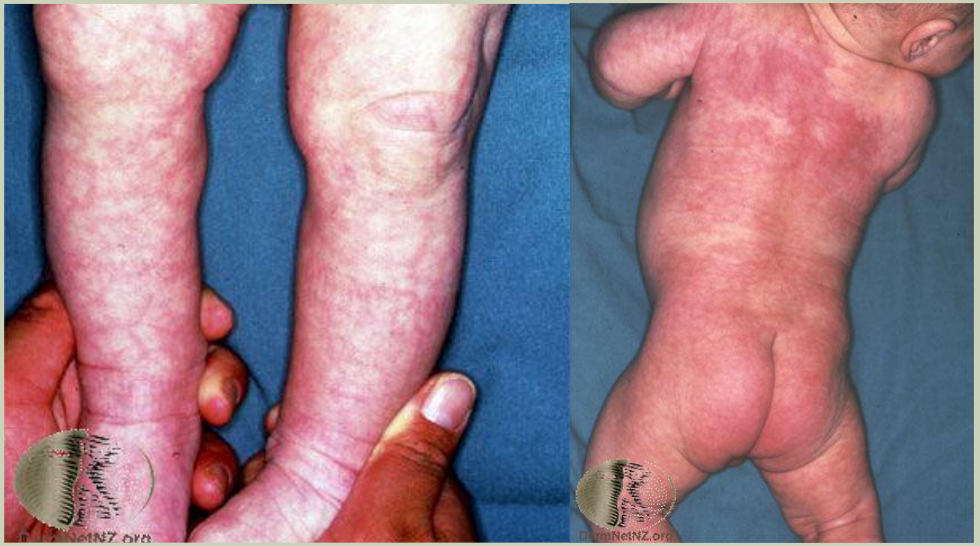
DISCLOSURES

- No relevant financial relationships to disclose
- Off-label use of acitretin in ichthyoses will be discussed

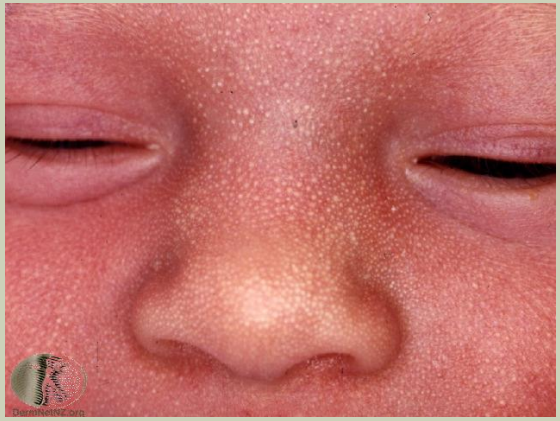
PHYSIOLOGIC

- Vernix caseosa
  - Creamy biofilm
  - Present at birth
  - Opsonizing, antibacterial, antifungal, antiparasitic activity
- Cutis marmorata
  - Reticular, blanchable vascular mottling on extremities > trunk/face
  - Response to cold
    - Disappears on re-warming
  - Associations (if persistent)
    - Down syndrome
    - Trisomy 18
    - Cornelia de Lange syndrome



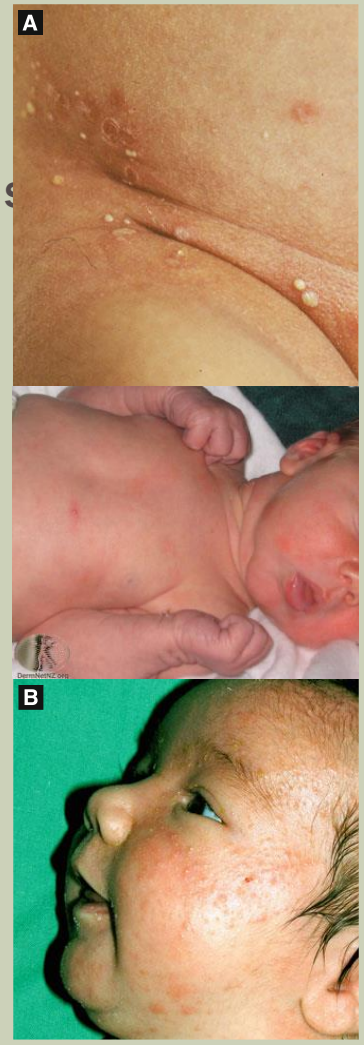
PHYSIOLOGIC

- Milia
  - Hard palate – Bohn's nodules
  - Oral mucosa – Epstein pearls
  - Associations
    - Bazex-Dupre-Christol syndrome (XLD)
      - BCCs, follicular atrophoderma, hypohidrosis, hypotrichosis
    - Rombo syndrome
      - BCCs, vermiculate atrophoderma, trichoepitheliomas
    - Oro-facial-digital syndrome (type 1, XLD)
    - Basal cell nevus (Gorlin) syndrome
    - Brooke-Spiegler syndrome
    - Pachyonychia congenita type II (Jackson-Lawler)
    - Atrichia with papular lesions
    - Down syndrome
    - Secondary
      - Porphyria cutanea tarda
      - Epidermolysis bullosa



TRANSIENT, NON-INFECTIOUS

- Transient neonatal pustular melanosis
  - Birth
  - Pustules → hyperpigmented macules with collarette of s
  - Resolve within 4 weeks
  - Neutrophils
- Erythema toxicum neonatorum
  - Full term
  - 24-48 hours
  - Erythematous macules, papules, pustules, wheals
  - Eosinophils
- Neonatal acne (neonatal cephalic pustulosis)
  - First 30 days
  - *Malassezia globosa & sympoidalis* overgrowth

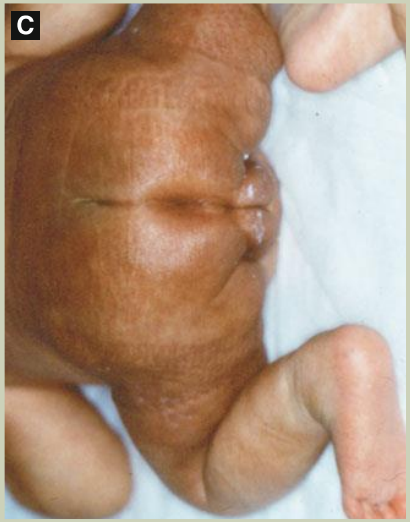


TRANSIENT, NON-INFECTIOUS

- Miliaria
  - First weeks
  - Eccrine gland obstruction due to heat, moisture, occlusion
  - Crystallina
    - Clear vesicles on head, neck, upper trunk
  - Rubra
    - Erythematous papulovesicles in intertriginous areas
  - Profunda
- Sucking blister
  - Solitary blister on hand, wrist, lip

TRANSIENT, NON-INFECTIOUS

- Subcutaneous fat necrosis of newborn
  - Healthy infants
  - Perinatal hypoxemia, hypothermia, hypoglycemia
  - 1st week
  - Localized, indurated subcutaneous nodules
    - Cheeks, shoulders, back, buttocks, thighs
  - Hypercalcemia
    - Monitor x 1 month after clinical resolution
  - Panniculitis, necrosis, needle-shape clefts, prominent inflammation



- Sclerema neonatorum
  - Ill/premature infants
  - 1st week
  - High mortality rate
  - Diffuse woody induration
    - Spares genitalia, palms, soles
  - Panniculitis, necrosis, needle-shaped clefts, minimal inflammation

TRANSIENT, BENIGN RASHES

*COURTESY OF AAD BASIC DERM CURRICULUM

	Erythema toxicum neonatorum	Benign cephalic pustulosis	Transient neonatal pustular melanosis	Miliaria	Seborrheic dermatitis
Onset	24-48 hours	2nd and 3rd week of life	Present at birth	Days to weeks	3 weeks to 12 months
Morphology	Erythematous macules, papules and pustules on erythematous base	Inflammatory papules and pustules; no true comedones	Vesicles, superficial pustules and pigmented macules, rim of scale	Small vesicles, vesicles with surrounding erythema, papules, small pustules	Erythema, greasy scales, and salmon-colored oval scaly patches
Distribution	Face, trunk, buttocks, and proximal extremities	Forehead, nose, and cheeks	Chin, neck, forehead, chest, buttocks, and less often on palms and soles	Forehead, neck and upper trunk	Scalp, face, forehead, eyebrows, trunk, intertriginous and flexural areas
Duration	5-7 days	6-12 months of age	Few days, pigmentation 3-6 months	Few days	Weeks to months

DIAPER DERMATITIS

- *Candida* infection
- Irritant contact dermatitis
- Allergic contact dermatitis
- Seborrheic dermatitis
- Psoriasis
- Atopic dermatitis
- Miliaria
- Granuloma gluteale infantum
- Jacquet's erosive dermatitis
- Perianal pseudoverrucous papules & nodules
- Acrodermatitis enteropathica
- Biotin/multiple carboxylase deficiency
- Cystic fibrosis
- Langerhans cell histiocytosis
- Kawasaki disease
- Perianal *Strep*
- Bullous impetigo
- Scabies
- Congenital syphilis

DIAPER DERMATITIS

- *Candida*
  - Satellite papules/pustules
  - Scrotal involvement
- Irritant dermatitis
  - Spares inguinal folds
- Allergic contact dermatitis
  - Rare
  - Topicals or foods
- Seborrheic dermatitis
  - 1 week – 1 year
  - Also on scalp
- Psoriasis
  - Involves inguinal folds
  - Minimal scale
  - Most common presentation in infancy

DIAPER DERMATITIS

- Granuloma gluteale infantum
  - Granulomatous nodules
  - Secondary to inflammation, maceration, *Candida* infection, halogenated topical steroids
- Jacquet erosive dermatitis
  - Rare
  - Erythematous, papular & erosive with elevated borders
  - Pain with urination
  - Secondary to irritant dermatitis, moisture, *Candida*, Hirschsprung disease
- Perianal pseudoverrucous nodules
  - Urinary/fecal incontinence

DIAPER DERMATITIS

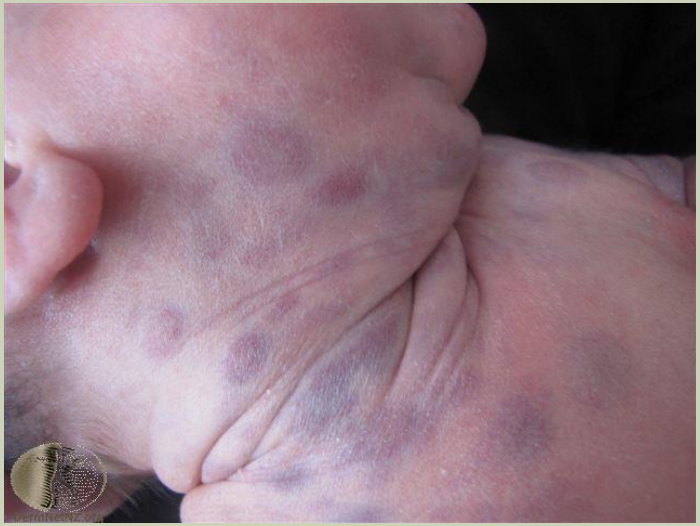
- Acrodermatitis enteropathica
  - Eczematous & erosive patches/plaques, flaccid bullae
  - Also periorificial & acral
  - Zinc deficiency
    - Alkaline phosphatase
  - Healthy, breastfed infants
    - Low zinc level in maternal milk
  - Upon weaning from breast milk
    - Premature infants
    - Inherited form (AR)
  - Acquired form
    - Inadequate nutrition
    - Poor absorption
- Biotin/multiple carboxylase deficiency
  - Neonatal form (AR)
    - Holocarboxylase synthetase deficiency
    - Alopecia & erythroderma
    - Fatal if untreated
  - Infantile form
    - Biotinidase deficiency
    - Alopecia, seizures, hearing loss
  - Treat both with biotin

DIAPER DERMATITIS

- Langerhans cell histiocytosis
  - Hashimoto-Pritzker disease
  - Yellow-brown, crusted papules & petechiae/purpura
  - Seborrheic distribution
- Kawasaki disease
  - Tender
  - Desquamation
- Perianal Strep
  - Well demarcated perianal erythema
  - Preceding *Strep* URI
- Bullous impetigo
  - Honey-colored crusts, flaccid bullae
- Scabies
- Congenital syphilis
  - Red-brown, papulosquamous
  - May be erosive/bullous

CONGENITAL INFECTIONS

- Toxoplasmosis
  - Truncal “Blueberry muffin” lesions
    - Red-blue papulonodules
    - Due to dermal hematopoiesis
  - Ocular & CNS abnormalities
  - Thrombocytopenia
  - Intracranial calcifications
- Rubella
  - Blueberry muffin lesions
  - Cataracts, deafness
  - Congenital heart disease
  - CNS abnormalities
  - Hepatosplenomegaly
  - Maternal infection during first 12-16 weeks of gestation



CONGENITAL INFECTIONS

- CMV
  - Most common infectious cause of deafness & MR
  - Blueberry muffin lesions, petechiae
  - Owl's eye intranuclear inclusion bodies
- HSV
  - Vesicles, erosions, scarring
  - Temporal lobe encephalitis
  - Microcephaly, chorioretinitis
  - Mostly perinatal HSV-2
  - 50-75% mortality if untreated
- Varicella
  - Congenital
    - Cicatricial skin lesions
    - First 20 weeks gestation
  - Neonatal
    - 5 days before to 2 days after delivery

CONGENITAL INFECTIONS

- Syphilis
  - 14+ weeks gestation
  - Early (birth to 2 years)
    - Rhagades
    - Papulosquamous macules/papules
    - Bullous lesions
    - Snuffles
    - Lymphadenopathy
    - Splenomegaly
  - Late
    - Parrot's lines
    - Hutchinson's teeth
    - Mulberry molars
    - Higoumenakis sign
    - Saddle nose
    - Saber shins
    - Interstitial keratitis
    - Gummas
    - Tabes dorsalis

RASHES REQUIRING WORKUP & TREATMENT

*COURTESY OF AAD BASIC DERM CURRICULUM

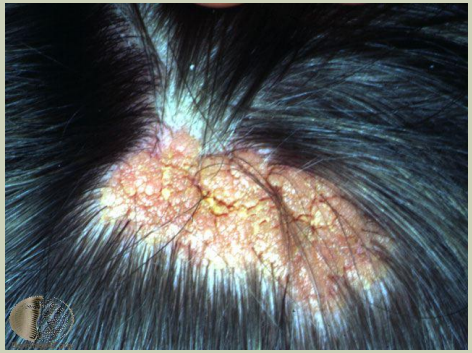
	Neonatal HSV	Neonatal LCH
Onset	Perinatal, postnatal	Birth, neonatal period
Morphology	Clusters of 2-4 mm vesicules with surrounding erythema (vesicles progress to pustules, and later crusting) Mouth: small ulcers	Multiple reddish-brown papules, later become crusts. Other morphologies also seen.
Distribution	Any where in the body	Head, neck, and distal extremities. Rarely extracutaneous.
Diagnosis	Tzanck smear, culture, PCR, rapid immunofluorescence assays or enzyme immunoassays	Skin biopsy CD-1A positive staining and Birbeck granules on electron microscopy
Work-up	Rule out CNS involvement and eye involvement	CBC, serum chemistries, liver function tests, coagulation studies, and urine osmolarity.
Prognosis	Good for early diagnosis, untreated disseminated neonatal HSV 80% mortality	Usually good, resolution in 3-4 months. Patients should be followed for progression/recurrence. 3% risk of mortality, 10% chance of relapse.

OTHER INFECTIONS

- Staph scalded skin syndrome
  - Group 2 phage *Staph aureus*
  - Exfoliative toxin A/B → cleave desmoglein 1
  - Immature renal function, lack of specific immunity
- Omphalitis
  - Periumbilical erythema, edema, tenderness, focal purulent discharge
- Mastitis
  - Term infants
  - 3 weeks
  - Neonatal breast abscess due to *S. aureus*

CONGENITAL NEVI & DEFECTS

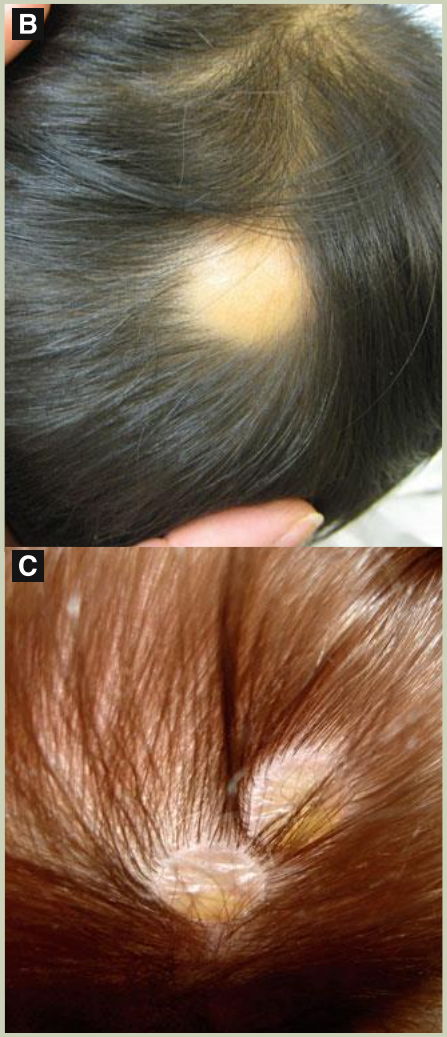
- Nevus sebaceous
  - Waxy, yellow-orange-tan, hairless plaque on face/scalp
  - Most prominent in neonates & again in puberty
  - Tumor development
    - Trichoblastoma, syringocystadenoma papilliferum
    - BCC



- Epidermal nevus
  - Bullous congenital ichthyosiform erythroderma risk in offspring
- Accessory tragus
  - First branchial arch
  - Associated with oculoauriculovertebral (Goldenhar) syndrome
- Supernumary nipples
  - Occur along embryological milk lines

CONGENITAL NEVI & DEFECTS

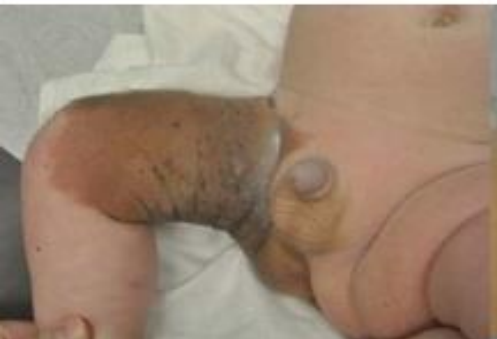
- Aplasia cutis congenita (ACC)
  - Ovoid erosion/ulceration/scar/membranous defect
  - Hair collar sign
  - Associations
    - Adams-Oliver syndrome
      - Scalp ACC, CMTC, limb & cardiac defects
    - Bart syndrome
      - Lower extremity ACC, dominant dystrophic EB
    - Seitles syndrome
      - Bilateral temporal ACC, leonine facies, abnormal lashes, upward slanting brows
    - Scalp
      - Teratogens – methimazole, varicella, HSV
      - Trisomy 13, ectodermal dysplasias, amniotic band syndrome
    - Extensive – increased AFP

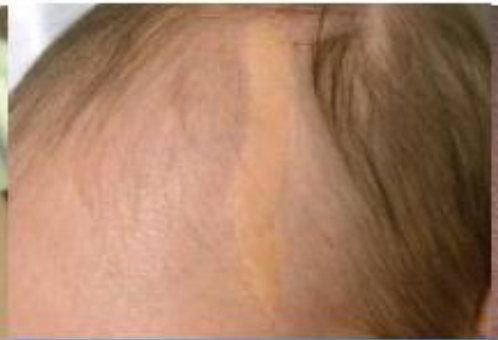
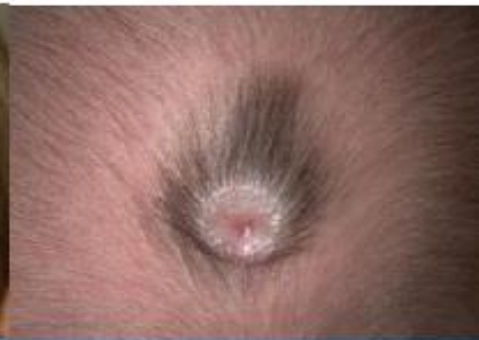


CONGENITAL NEVI & DEFECTS

*COURTESY OF AAD BASIC DERM CURRICULUM

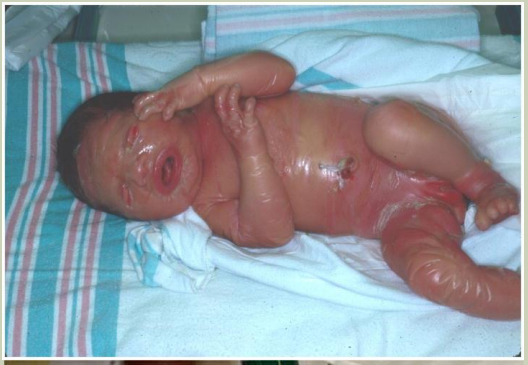
Congenital melanocytic nevus	Dermal melanocytosis	Nevus sebaceous	Aplasia cutis congenita
Proliferation of benign melanocytes	Entrapment of melanocytes in dermis	Organoid hamartoma	Absence of skin present at birth
Pigmented macules, papules or plaques; +/- hair	Blue-gray pigmentation, ill defined	Smooth, yellow-orange hairless patch	Presents as superficial erosion, ulceration or scar
Risk of melanoma in large/giant CMN	Benign, predilection buttocks/lower back	More pronounced around adolescence	Atypical/large or hair collar sign indicate need for imaging



GENODERMATOSES PRESENTING IN NEONATAL PERIOD

- Collodion baby
  - Lamellar ichthyosis
  - Congenital ichthyosiform erythroderma
  - Self-healing collodion baby
- Harlequin fetus (AR)
  - ABCA12 gene
  - Universally fatal historically
    - Supportive care
  - Acitretin
    - Increased survival
    - Improved mobility
    - Encourages softening & shedding of encasement
      - Eclabium – able to latch & feed
    - Start early (by day 7) → significant improvement in 1 month

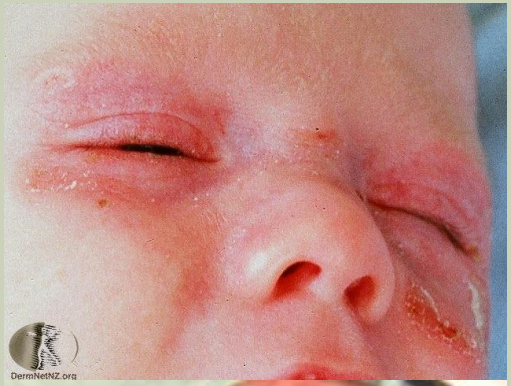


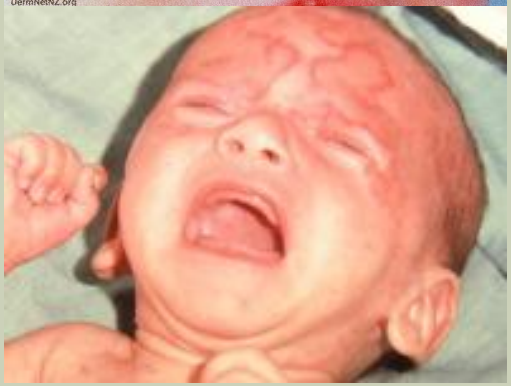
GENODERMATOSES PRESENTING IN NEONATAL PERIOD

- Epidermolysis bullosa
- Incontinentia pigmenti
- Oculocutaneous albinism
- Neurofibromatosis type 1
- Cutis laxa
- Ehlers-Danlos syndrome
- Hereditary palmoplantar keratodermas
- Piebaldism
- Congenital erythropoietic porphyria
- Ectodermal dysplasias
- Multiple lentigines syndromes
- Phakomatosis pigmentovascularis

IMMUNOLOGIC

- Neonatal lupus erythematosus
  - Birth to first few weeks
  - SCLE lesions periorbitally & on extremities
  - Transplacental exposure to maternal antibodies
    - Anti-Ro
    - Anti-La, Anti-U1RNP
  - Congenital heart block (50%)
    - EKG
    - 2/3 require permanent pacemaker
    - 20% mortality
  - Thrombocytopenis, neutropenia, transaminitis
  - Cutaneous lesions resolve in 6-8 months
- Neonatal pemphigus
- Langerhans cell histiocytosis
- Immunodeficiency syndromes





REFERENCES

- Sharma, YK, Sadana, DJ, Rizvi A, Dash K. A comprehensive classification and abbreviated update of neonatal dermatological entities. Indian J Paediatr Dermatol. 2015;16:122-135.
- Jain, S. Dermatology – Illustrated Study Guide and Comprehensive Review. 28-31. New York: Springer; 2012:28-31.
- Pride HB, Tollefson M, Silverman R. What's new in pediatric dermatology?: Part I. Diagnosis and pathogenesis. J Am Acad Dermatol. 2013 Jun;68(6):885.e1-12. doi: 10.1016/j.jaad.2013.03.001
- Pride HB, Tollefson M, Silverman R. What's new in pediatric dermatology?: Part II. Treatment. J Am Acad Dermatol. 2013 Jun;68(6):899.e1-11. doi: 10.1016/j.jaad.2013.03.002
- Craiglow BG. Ichthyosis in the Newborn. Seminars in perinatology. 2013;37(1):26-31. doi:10.1053/j.semperi.2012.11.001
- Ahmed, H, O'Toole, EA. Recent Advances in the Genetics and Management of Harlequin Ichthyosis. Pediatr Dermatol. 2014;31:539-546. doi:10.1111/pde.12383
- Prado R, Ellis LZ, Gamble R, Funk T, Arbuckle HA, Bruckner AL. Collodion Baby: An update with a focus on practical management. J Am Acad Dermatol. 2012;67:1362-1374.
- Harvey HB, Shaw MG, Morrell DS. Perinatal management of harlequin ichthyosis: a case report and literature review. J Perinatol. 2010;30:66-72.

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