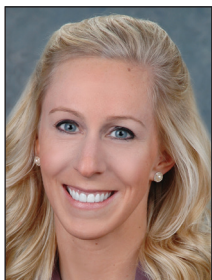


Neonatal Ichthyosis

by Alyx Rosen, MD and Kate Oberlin, MD

Disease	Gene Defect	Clinical Presentation	Additional Manifestations	Prognosis/ Outcome
Harlequin ichthyosis	AR ABCA12	Thick membrane with deep cracks and fissures forming plate-like scales, ectropion, eclabium	Premature delivery; respiratory distress; ear deformities; fluid imbalance	Survival rate ~50%; oral retinoids may help prolong survival
Congenital ichthyosiform erythroderma (CIE)	AR TGM-1 ALOX12B ALOXE3	Generalized erythema and fine scale; collodion membrane at birth; involves flexures; nail dystrophy	Heat intolerance; scarring alopecia; rare ectropion	No improvement with age
Lamellar ichthyosis	AR TGM-1 ABCA12	Thick, brown scale involving flexural areas and trunk; eclabium, ectropion	Heat intolerance; hypernatremia	No improvement with age
Epidermolytic hyperkeratosis (EHK); Bullous CIE	AD K1/K10	Widespread blistering and erythema at birth; involves flexures, palmoplantar keratoderma	Failure to thrive; hypernatremia; recurrent infections	Evolves into verrucous hyperkeratotic plaques
Ichthyosis vulgaris	AD Filaggrin	Generalized fine, adherent scale sparing flexures	Atopic diathesis; keratosis pilaris	Improves with age
X-linked ichthyosis	XLR Steroid sulfatase	Generalized fine scale and desquamation at birth	Corneal opacities; cryptorchidism; prolonged labor	Brown "dirty" scales sparing flexures
Netherton syndrome	AR SPINK5	Trichorrhexis invaginata, ichthyosis linearis circumflexa, atopic dermatitis	Failure to thrive; infections; elevated IgE; food allergies	Pruritus; eczematous plaques
Refsum disease	AR PEX7 PHYH	Skin resembles ichthyosis vulgaris with mild scaling	Retinitis pigmentosa; cerebellar ataxia	Accumulation of phytanic acid; deafness
Sjögren-Larsson syndrome	AR FALDH	Generalized erythema and scale at birth, then dark scales on trunk	Parafoveal glistening white dots on retina	Mental retardation, seizures and spastic paralysis
Conradi-Hünemann-Happle syndrome	XLD EBP gene	Severe ichthyosiform erythroderma and scale following lines of Blaschko	Chondrodysplasia punctata; cataracts; deafness; alopecia	Follicular atrophoderma and scarring
KID syndrome	AD GJB2	Keratitis, ichthyosis, deafness, keratoderma	Photophobia; leukonychia	Increased risk of cutaneous SCC
Trichothiodystrophy (PIBIDS)	AR ERCC2 ERCC3	Photosensitivity, ichthyosis, brittle hair "tiger tail", intellectual impairment, decreased fertility, short	Sulfur deficiency in hair; diffuse alopecia	Photosensitivity decreases with age
CHILD syndrome	XLR NSDHL	Congenital hemidysplasia with ichthyosiform erythroderma, limb defects	Ipsilateral organ aplasia; stippled epiphyses	Erythema fades into hyperkeratosis



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DDx "Collodion Baby": Lamellar Ichthyosis, Congenital Ichthyosiform Erythroderma (CIE), Netherton syndrome, Harlequin Ichthyosis, Self-healing collodion baby (SHCB), Trichothiodystrophy (PIBIDS), Sjögren-Larsson syndrome, Infantile Gaucher disease, Neutral lipid storage disease with ichthyosis, Hay-Wells syndrome, Conradi-Hünemann-Happle syndrome

References:

- Bolognia, Jean, Joseph Jorizzo, and Julie Schaffer. "Chapter 57." *Dermatology*. 3rd ed. Philadelphia: Elsevier Saunders, 2012. 837-870. Print.
- Cohen, Bernard. "Chapter 2." *Pediatric Dermatology*. 4th ed. Philadelphia: Elsevier Saunders, 2013. 25-39. Print.
- Schachner, Lawrence, Ronald Hansen. "Chapter 8." *Pediatric Dermatology*. 4th ed. Philadelphia: Elsevier Mosby, 2011. 567-643. Print.

Neonatal Infectious Dermatoses

by Alyx Rosen, MD, and Kate Oberlin, MD

Disease	Clinical Presentation	Extracutaneous Manifestations	Work-Up	Treatment
Syphilis	Macules or papules on palms and soles, trunk, extremities; also erosions, bullae and petechiae; mucous patches	IUGR, HSM, osteochondritis, "snuffles" or rhinitis	Maternal and neonate serology; darkfield examination	High dose penicillin G to prevent late complications
Candidiasis	Diffuse red eruption with satellite red papules and pustules on the folds and scrotum	Oral thrush, paronychia, systemic infection	Gram stain or KOH reveals pseudohyphae or budding yeast	Topical nystatin, ciclopirox, imidazoles, PO/IV if septic
Toxoplasmosis	Varies depending on timing of infection; non-specific, diffuse macules or papules, ecchymosis	Chorioretinitis, hydrocephalus, deafness, preterm birth, IUGR	Serologic testing for antigen, PCR, or antibody (IgG, IgM)	Pyrimethamine with Sulfadiazine and leucovorin
Rubella	"Blueberry muffin" macules due to extramedullary hematopoiesis	Cataracts, congenital heart disease, deafness, HSM, microcephaly	Viral cultures of pharynx, CSF, urine, conjunctiva, rubella antibody	Neuroimaging; supportive care
CMV	Generalized petechiae or purpura within 48 hours of birth, "blueberry muffin" macules	IUGR, deafness, HSM, thrombocytopenia, chorioretinitis	Viral culture, PCR, antibody testing	Ganciclovir, valganciclovir, foscarnet
Scabies	Vesicles and papules with rare burrows on the palms, soles, trunk and genitalia	Pruritus, irritability, poor feeding	Mineral oil prep demonstrating mite or eggs	Permethrin cream
HIV	Thrush, widespread seborrheic dermatitis	Bacterial and viral infections	Serology testing, PCR	Zidovudine; anti-retroviral therapy
HSV	Grouped vesicles on red base; diffuse erosions	Ocular and CNS involvement, sepsis	Viral cultures, PCR, Tzanck prep	Intravenous acyclovir
Varicella	Cicatricial dermatomal lesions with hypoplasia (congenital); generalized papules (neonatal)	Ocular and CNS abnormalities	Antibody testing, PCR, Tzanck prep	Varicella-zoster immunoglobulin; acyclovir
Impetigo neonatorum	Pustules, vesicles or bullae on an erythematous base in folds and groin; second week of life	Rarely complicated by sepsis, osteomyelitis and pneumonia	Gram stain with Gram-positive cocci in clusters and neutrophils	Dicloxacillin or cephalixin for a 10-day course
Staphylococcal Scalded Skin Syndrome (SSSS)	Scarlatiniform eruption with erythroderma in intertriginous sites; periorificial crust, +Nikolsky sign, spares mucosa	Irritability, fever, skin tenderness, rhinorrhea	Histology (frozen) with granular layer cleavage; bacterial cultures	β -lactamase-resistant antibiotics; decolonization

Abbreviations: HSM=hepatosplenomegaly; CMV=Cytomegalovirus; HSV=Herpes simplex virus; HIV=Human immunodeficiency virus; CNS=Central nervous system; IUGR=intrauterine growth restriction

DDx of "Blueberry muffin baby": CMV, varicella, zoster, rubella, toxoplasmosis, langerhans cell histiocytosis, syphilis, leukemia cutis, neuroblastoma, parvovirus, hemolytic anemia, twin-twin transfusion, myelodysplasia, rhabdomyosarcoma, neonatal lupus

References:

1. Bologna, Jean, Joseph Jorizzo, and Julie Schaffer. "Chapters 80-81." *Dermatology*. 3rd ed. Philadelphia: Elsevier Saunders, 2012. Page 1321-1366. Print.
2. Cohen, Bernard. "Chapter 2." *Pediatric Dermatology*. 4th ed. Philadelphia: Elsevier Saunders, 2013. 25-39. Print.
3. Schachner, Lawrence, Ronald Hansen. "Chapter 6." *Pediatric Dermatology*. 4th ed. Philadelphia: Elsevier Mosby, 2011. 299-373. Print.

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