Eponyms in Dermatology

by Heather Kiraly Orkwis, DO. (Updated July 2015)*

Aleppo/Baghdad/Delhi boil = lesion of cutaneous leishmaniasis

Asboe-Hansen sign = extension of intact blister when pressure is applied to roof; seen in pemphigus vulgaris

Auspitz's sign = punctate bleeding points within lesion upon scratching; seen in psoriasis

Bateman purpura = actinic (solar) purpura

Bazex syndrome, acquired = acrokeratosis paraneoplastica

Bazex syndrome (Bazex–Dupré–Christol Syndrome), XLD = follicular atrophoderma, multiple BCCs, hypotrichosis, localized hypohidrosis

Bazin's disease = erythema induratum, associated with TB

Becker nevus/melanosis = large, unilateral hyperpigmented patch, with secondary hypetrichosis often hairy on upper extremity or chest

Beckwith-Wiedemann syndrome = exomphalos-macroglossia-gigantism syndrome (p57/KIP2)

Behcet's disease = triad of apthous ulcers, genital ulcers, ocular inflammation (+HLA-B51; Silk Road Disease)

Bloch-Sulzberger disease =Incontinentia Pigmenti (NEMO; X-linked dominant)

Bockhart's impetigo = follicular impetigo

Bourneville's disease = Tuberous Sclerosis (Epiloia) (TSC1, TSC2)

Bowen's disease = squamous cell carcinoma in situ

Buruli ulcer = M. ulcerans (named after Buruli region of Nile River, Africa)

Buschke-Lowenstein tumor = verrucous carcinoma of glans penis and prepuce (HPV 6, 11)

Buschke-Ollendorff syndrome = dermatofibrosis lenticularis disseminata, osteopoikilosis (LEMD3)

Calabar swellings = localized angioedema in tissue from migrating loiasis

Carney Complex = NAME syndrome, LAMB syndrome (PRKAR1A)

Carvajal syndrome = left sided cardiomyopathy, woolly hair, keratoderma (desmoplakin)

Cobb syndrome = cutaneomeningospinal angiomatosis

Civatte bodies = degenerated, apoptotic keratinocytes seen in lichen planus

Conradi-Hünermann syndrome = XLD chondrodysplasia punctata (EBP gene)

Crowe's sign = axillary or inguinal freckling seen in neurofibromatosis

Darier disease = Keratosis follicularis (ATP2A2)

Darier's sign = urtication following rubbing of macule/papule in mastocytosis (urticaria pigmentosa)

Dennie-Morgan lines = crescentic creases of lower eyelids due to stagnation of venous blood, seen in atopic dermatitis

Degos' disease = malignant atrophic papulosis

Dercum disease = adiposis dolorosa; mostly obese menopausal women, consisting of multiple exquisitely tender lipomas

Favre-Racouchot = open comedones and solar elastosis

Gianotti-Crosti syndrome = papular acrodermatitis of childhood

Goltz syndrome = focal dermal hypoplasia (PORCN)

Gorlin syndrome = nevoid basal cell carcinoma syndrome (patched-1)

Gottron's papules = erythematous eruption over knuckles, elbows, knees, seen in dermatomyositis

Graham-Little-Piccardi-Lasseur syndrome = variant of LPP: cicatricial alopecia of scalp, non-scarring alopecia of axilla and groin, follicular lichen planus eruption

Grover's disease = transient acantholytic dermatosis

Hailey-Hailey disease = familial benign pemphigus (ATP2C1)

Heck's disease = oral focal epithelial hyperplasia (HPV 13, 32)

Griscelli syndrome = pigmentary dilution, T- and B-cell immunodeficiency, recurrent infection, progressive CNS deterioration (myosin VA)

Hermansky-Pudlak syndrome = pigment dilution, bleeding diathesis, lysosomal membrane defect (HPS1)

Howel-Evans syndrome = non-transgradiens PPK, esophageal carcinoma (TOC)

Hughes' triad = antiphosholipid antibody syndrome (fetal loss, thrombosis, thrombocytopenia)

Hutchinson-Gilford syndrome = progeria (lamin A)

Hutchinson's sign = pigmentation of proximal nail fold, suggestive of melanoma

Janeway lesions = non-painful hemorrhagic macules or nodules of palms and soles, seen in infective endocarditis

Kasabach-Merritt syndrome = consumptive coagulopathy within a kaposiform hemangioendothelioma or tufted angioma

Klippel-Trenaunay syndrome = angio-osteohypertrophy syndrome; port-wine stain, soft tissue and bony hypertrophy, venous and lymphatic malformations

Koplik's spots = small, white spots on erythematous buccal mucosa, seen in early measles

Kveim-Sitzbach test = skin test with human sarcoid tissue injected into a patient suspected of having sarcoidosis; positive results are a sarcoid granuloma at the site

Kyrle's disease = chronic generalized dermatosis with papules with central keratotic plugs (DM, renal disease)

Leser-Trélat sign = abrupt onset of multiple seborrheic keratoses, associated with internal malignancy

Lichtenberg's figures = branching pattern of cutaneous marks pathognomonic for lightning injury

Lofgren syndrome = erythema nodosum, bihilar lymphadenopathy (sarcoidosis)

Louis-Bar syndrome = ataxia telangiectasia mutated (ATM) gene

Lovibond's angle = 160° angle between proximal nail fold and the nail plate

Lyell's syndrome = toxic epidermal necrolysis (TEN)

Madelung's disease = benign symmetric lipomatosis (Launois-Bensuade syndrome, horse-collar appearance)

Maffucci syndrome = superficial and deep venous malformations, enchondromas, chondrosarcoma (PTHR1)

Majocchi's disease = purpura annularis telangiectoides

Majocchi granuloma = deep dermatophyte infection of hair follicle

Mal de Meleda = keratoderma palmoplantaris transgrediens (SLURP1)

Marfan syndrome = tall stature, arachno-dactyly, ectopia lentis, progressive aneurysmal dilation of ascending aorta, CHF (fibrillin 1)

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Marjolin's ulcer = aggressive SCC arising in site of chronic injury or burn

McCune-Albright Syndrome = Albright syndrome; "Coast of Maine" café-au-lait macule(s), polyostotic fibrous dysplasia, precocious puberty (GNAS1)

Milroy's disease = congenital lower limb lymphedema (FLT4)

Montgomery syndrome = xanthoma disseminatum

Mucha Habermann disease = pityriasis lichenoides et varioliformis acuta (PLEVA)

Muckle-Wells syndrome = recurrent fevers and urticaria, progressive deafness, secondary amyloidosis (cryopyrin)

Muir-Torre syndrome = DNA mismatch repair defect, sebaceous tumors, adenocarci-noma of the colon (MLH1, MSH2)

Naxos disease = right sided cardiomyopathy, woolly hair, non-epidermolytic PPK (plakoglobin)

Netherton syndrome = ichthyosis linearis circumflexa (SPINK5)

Nikolsky's sign = normal epidermis easily separated when pressed firmly with a sliding motion, seen in pemphigus vulgaris, staphylococcal scalded skin syndrome

Ollendorf's sign = secondary syphilis papule tender to touch with blunt probe

Osler-Weber-Rendu syndrome = Hereditary Hemorrhagic Telangiectasia syndrome (HHT1, HHT2)

Papillon-Lefèvre syndrome = palmoplantar keratoderma with periodontosis (cathepsin C)

Parry-Romberg syndrome = acquired progressive hemifacial atrophy (morphea variant)

Refsum syndrome = phytanic acid storage disease (PAHX, PEX7)

Richner-Hanhart syndrome = Tyrosinemia type II (tyrosinase aminotransferase)

Ritter's disease = staphylococcal scalded skin syndrome

Russell's sign = dorsal hand with dry skin and calluses, seen with bulimia/purging

Schnitzler's syndrome = nonpruritic urticaria, arthralgias, IgM monoclonal protein

Senear-Usher syndrome = pemphigus erythematosus; variant of P. foliaceous confined to seborrheic sites

Sezary syndrome = generalized exfoliative erythroderma (part of CTCL)

Shulman's syndrome = eosinophilic fasciitis (dry river bed)

Sjögren-Larsson syndrome = ichthyosis with erythroderma, spastic di-tetraplegia with scissor gait, mental retardation, atypical retinitis pigmentosa (FALDH)

Sneddon's syndrome = livedo reticularis, HTN, CVA associated with antiphospholipid antibodies

Sturge-Weber syndrome =encephalotrigeminal angiomatosis

Tyndall effect = blue tinging of subcutaneous lesions due to short wavelength colors (blue/violet) scattering

Urbach-Wiethe disease = lipoid proteinosis (ECM1)

Vohwinkel syndrome = PPK mutilans, keratoderma hereditaria mutilans (connexin 26, loricrin)

Von Recklinghausen disease = neurofibromatosis I (neurofibromin)

Well's syndrome = eosinophilic cellulitis, 'flame figures' on dermatopathology

Zinsser-Engman-Cole syndrome = Dyskeratosis Congenita (dyskerin)

References

1. Bolognia JL, Jorizzo JL, Schaffer JV, editors. *Dermatology. 3rd ed.* China: Elsevier publishing; 2012.
2. Spitz JL. *Genodermatoses: A clinical guide to genetic skin disorders.* Philadelphia, Pa: Lippincott Williams & Wilkins; 2005.

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