Achard-Thiers s.	postmenopausal women, excess adrenocortical androgens; diabetes, hirsutism & masculinization
acquired circumscribed dermal melanocytosis of the face	Hori's nevus
acquired partial face-sparing lipodystrophy	Dunnigan s.
acrodermatitis enteropathica	AR; disorder of zinc uptake, vesiculopustulous dermatitis of head, body orifices, hands & feet with alopecia, diarrhea & steatorrhea
acrodermatitis papulosa infantum	Gianotti-Crosti s.
actinic prurigo	Hutchinson's prurigo
acute disseminated histiocytosis X	Letterer-Siwe d./s.
acute hemorrhagic edema of infancy	Finkelstein's d., Seidlmeyer's s.
Adams-Oliver s.	AD/AR; aplasia cutis congenita of scalp with terminal transverse limb defects
Adamson's fringe	bottom one-third of hair follicle
Addison's d.	adrenal insufficiency; hyperpigmentation, longitudinal nail bands, hypotension, altered serum electrolytes, vitiligo, weakness
adenosine deaminase deficiency	erythrocyte enzyme; thymic involution, decreased T cell survival, abnormal B cell function: mucocutaneous candidiasis, bacterial, protozoal & viral infections
adiposis dolorosa	Dercum's d.
Albright's s.	McCune-Albright s.
Alezzandrini's s.	unilateral tapetoretinal degeneration followed by alopecia areata, facial vitiligo, poliosis, sometimes deafness
alkaptonuria	AR, homogentisic acid accumulation due to deficient homogentisate 1,2-dioxygenase; elevated urine homogentisic acid, ochronosis, arthritis
Ambras s.	AD; hypertrichosis of face, ears, shoulders with facial dysmorphism & dental abnormalities
Ancell-Spiegler s.	multiple cylindromas
angiokeratoma corporis diffusum	Fabry's d.
angry back s.	excited skin s.
anhidrotic ectodermal dysplasia	Christ-Siemens-Touraine s.
anticardiolipin Ab s.	arterial & venous thromboses, elevated anticardiolipin antibody, livedo reticularis, SLE
antiphospholipid Ab s.	lupus anticoagulent s.
Apert's s.	acrocephalosyndactyly, associated with cutis verticis gyrata
Argyll-Robertson pupil	irregular, small, unequal pupils that react weakly to light; syphilis
Ascher s.	blepharochalasis, double lip & nontoxic thyroid enlargement
ataxia-telangiectasia	Louis-Bar s.
Auspitz sign	punctate bleeding at the sites of scale removal; psoriasis (except inverse or pustular psoriasis)
B¬¬k's s.	palmar hyperhidrosis
BADS s.	*b*lack locks, oculocutaneous *a*lbinism & *d*eafness of *s*ensineuronal type
Bannayan s., Bannayan-Zonana s.	Proteus s.
Baraitser's s.	atrichia, MR
Bart's s.	AD; aplasia cutis congenita (esp. of legs); dominant epidermolysis bullosa dystrophica

Bart-Pumphrey s.	AD; palmoplantar keratoderma with sensorineural deafness AKA PED type XV
basal cell nevus s.	Gorlin's s., Gorlin-Goltz s.
Basan's s.	AD; fine dermal ridges, single flexion crease, xerosis, long philtrum, thin upper lip, nail ridges
Bateman, herpes iris of	large lesions of EM with central bulla & marginal vesicle ring
Bateman's purpura	actinic (solar) purpura
Battle's sign	skin discoloration over mastoid area of skull; basilar skull fracture
Bazex's s.	1) AD; follicular atrophoderma of dorsal hands, abnormal hair & sweat glands, multiple BCC 2) acquired eczematous & psoriasiform lesions (ear, nose, dorsal hands, feet, knees), nail dystrophy & paronychial inflammation with SCC (GI, respiratory) (100%) AKA paraneoplastic acrokeratosis
Bean s.	AD, sporadic, rare; rubbery cutaneous venous malformations with GI bleeds & hemangiomas, anemia AKA blue rubber bleb nevus s.
Beane-Stevenson s.	AD, 10q26, FGFR2; cutis gyrata, craniofacial dysostosis
Beau's lines	transverse depression across nail plate; cytotoxic drugs, dysmenorrhea, MI, post fever, psoriasis
Becker's nevus	common, sharply demarcated hyperpigmented patch of shoulder, chest, scapula AKA Becker's melanosis
Beckwith-Wiedemann s.	AD with variable penetrance; sporadic; mid-facial capillary malformation, macroglossia, HSM, omphalocele, gigantism, hemihypertrophy, Wilms' tumor, adrenal cortical carcinoma, hepatoblastoma AKA EMG s. & exomphalos-macroglossia-gigantism s.
Bednar tumor	pigmented dermatofibrosarcoma protuberans
Behçet's d.	aphthosis (oral & genital), arthritis, cutaneous pustular vasculitis, meningoencephalitis, posterior uveitis, thrombophlebitis, intestinal inflammation
benign familial pemphigus	Hailey-Hailey d.
Bezold sign	inflammatory edema below apex of mastoid process; mastoiditis
BIDS s.	*b*rittle hair, *i*ntellectual impairment, *d*ecreased fertility, *s*hort stature
Biederman's sign	dark red color of anterior pillars of throat; syphilitics (some)
Bier spots	light macules on arms & legs of young adults; benign physiologic vascular anomaly
Biette's collarette	thin white ring of scales on papule surface; secondary syphilis
biotin dependent carboxylase deficiency	CNS disease; alopecia, ataxia, ***Candida*** dermatitis, decreased IgA, defective T cells, keratoconjunctivitis, seizures
biotinidase deficiency	AR; deficiency causes multiple carboxylase deficiency
bird-headed dwarfism	Seckel's s.
Birt-Hogg-Dube s.	fibrofolliculomas (>5) of the ear, forehead, nose, temporal region
Bj¬rnstad's s.	AR; pili torti & sensorineural deafness
Blaschko's lines	CHILD s., chondrodysplasia punctata, Delleman-Oorthuys s., focal dermal hypoplasia (Goltz's s.), hypomelanosis of Ito, incontinentia pigmenti, incontinentia pigmenti achromians, lichen striatus, linear & whorled nevoid hypermelanosis, linear epidermal nevus, linear lichen planus, linear nevus sebaceous, linear psoriasis, linear scleroderma, mosaic EHK, Proteus s., Schimmelpenning-Feverstein-Mims s.
Blaschko's lines, linear	ILVEN, lichen striatus, linear Darier's d., linear LP, linear porokeratosis, nevus comedonicus
Blaschko's lines, x-linked	CHILD s., Conradi-Hunermann s., focal dermal hypoplasia, hypohidrotic ectodermal dysplasia, incontinentia pigmenti, Menkes' kinky hair s., orofacial-digital s, Partington's s.
blastomycosis, N. American	Gilchrist's d.
blind loop s.	bowel bypass s.
Bloch-Sulzberger s.	XLD male lethal, Xp11; cutaneous, ocular, CNS & skeletal abnormalities AKA incontinentia pigmenti
Bloom's s.	AR, 15q26; photodistributed erythema with telangiectasia, chelitis, café-au-lait, craniofacial abnormalities, growth retardation, hypogonadism, leukemia, lymphoma,

	breast cancer, GI adenocarcinoma AKA congenital telangiectatic erythema
blue rubber bleb nevus s.	Bean s.
Borsieri's sign	fingernail drawn along skin produces white demarcation line which quickly turns red; scarlet fever (early)
Bourneville's s., Bourneville-Pringle s.	AD; adenoma sebaceum, brain hamartomas, café-au-lait spots, MR, seizures, shagreen patches, subungual fibromas, vitiligo AKA tuberous sclerosis
bowel bypass s.	(20%) post jejunal bypass; malaise, myalgia, polyarthralgia, rash, sterile skin pustules
Bowen's d.	SCC of genitals
Brachmann-de Lange s.	Cornelia de Lange s.
Brill-Zinsser d.	recurrent epidemic typhus (*Rickettsia prowazeki*)
Brocq, pseudopelade of	end-stage scarring alopecia caused by favus, LE, LP, sarcoidosis, scarring folliculitis, scleroderma
Brooke-Fordyce	hereditary benign cystic epitheliomas
Brooke-Spiegler s.	multiple trichoepitheliomas & cylindromas
Brunauer-Fuhs-Siemens PPK	AD; mild PPK AKA striate PPK, Wachters PPK, keratosis palmoplantaris varians
Brunsting-Perry pemphigoid	similar to cicatricial pemphigoid with predominant head & neck scarring involvement
Buckley's s.	hyperimmunoglobulin E s. with asthma & coarse facial features
Buerger's d.	thromboangiitis obliterans
burning feet s.	Goplans' d.
Burns' s.	KID s.
Buruli ulcer	*Mycobacterium ulcerans* skin ulcer
Buschke-Fischer-Brauer d.	keratosis punctata palmaris et plantaris
Buschke-Lowenstein tumors	dysplastic genital warts
Buschke-Ollendorff s.	AD; dermatofibrosis lenticularis disseminata, elastin nevi, osteopoikilosis
Bywater's lesions	digital pulp papules (leukocytoclastic vasculitis); autoimmune disease, especially RA
Calabar swellings	localized angioedema from adult worm migration through subcutis, usually around joints: loiasis
Campbell-DeMorgan spots	cherry angioma (senile angioma)
candidiasis, chronic mucocutaneous	severe combined immunodeficiency s., Nezelof's s., DiGeorge's s. **candidiasis, chronic oral**; onset any age, no inheritance pattern; oropharyngeal, sparing skin & nails **chronic candidiasis with endocrinopathy**; AR; onset childhood, one or more: hypoadrenalism, hypoparathyroidism hypothyroidism, hypogonadism, antibodies to endocrine glands, alopecia, vitiligo **chronic localized candidiasis**; (*Candida* granuloma) onset childhood, may be associated with endocrinopathy, mucosal, nail & skin, hyperkeratotic, granulomatous, vegetating *Candida* **chronic diffuse candidiasis**; AD/AR; nails, skin, mucous membranes, no endocrinopathy **chronic candidiasis with thymoma**; adult onset; myasthenia gravis, aplastic anemia, decreased neutrophils & immunoglobulins
Cannon's d.	white sponge nevus
Cantu's s.	AD; brown macules on face, forearms, feet with hyperkeratosis of palms & soles
carbon baby s.	rare; hyperpigmentation progressing from groin & face to entire skin surface birth-2 ½ years AKA universal acquired melanosis
carcinoid s.	pronounced flushing, with wheezing, diarrhea, abdominal pain; carcinoid cancer of bowel, bronchus, pancreas, thyroid, teratomas
cardio-facio-cutaneous s.	abnormal facies, cardiac anomalies, eczema, follicular hyperkeratosis, growth retardation, ichthyotic changes, MR, occasionally palmoplantar hyperkeratosis, splenomegaly
Caripito itch	irritating setae of genus *Hylesia* caterpillar AKA butterfly itch, moth dermatitis
Carney's s.	lentigines, myxomas (cardiac, skin, breast), endocrine abnormalities
Carrion's d.	Oroya fever
Casal's necklace	broad band of dermatosis around neck; pellagra (niacin deficiency)

Castleman's d.	lymphoproliferative disorder presenting as isolated mediastinal mass or solitary cutaneous tumor AKA angiofollicular lymphoid hyperplasia
Chadwick's sign	dark blue or purple color of vaginal mucosa; pregnancy
Chagas' d., Chagas-Cruz d.	American trypanosomiasis, *Trypanosoma cruzi*, reduviid bug (vector) erythematous nodule (chagoma), at bite site, high fever, unilateral facial & eyelid edema (Roma™a's sign) regional LAD, HSM & meningoencephalic irritation
Chanarin-Dorfman s.	AR; disordered lipid metabolism, erythroderma, fine white scaling & lichenification over dorsal hands, myopathy, vacuolated leukocytes; increased in Middle Eastern or Mediterranean descent AKA neutral lipid storage d.
chancriform s.	nodule/chancre on distal extremity with chain of nodules extending proximally; sporotrichosis
Chediak-Higashi s.	AR, lethal; oculocutaneous albinism, absent elastase, increased cAMP, decreased cGMP, poor melanosome transport, ecchymoses, pigmented nevi, gray hair, pancytopenia, bleeding diathesis, lymphoma, HSM, giant lysosomal granules, histiocytic infiltration of organs, pulmonary pyogenic infections, leukocyte deficiencies (chemotaxis, adherence, killing, deformability)
CHILD s.	XLD; *c*ongenital *h*emidysplasia with *i*chthyosiform erythroderma & *l*imb *d*efects
CHIME s.	*c*oloboma, *h*eart defects, *i*chthyosiform dermatosis, *m*ental retardation & *e*ar anomalies AKA Zunich-Kaye s.
chondrodysplasia punctata	Conradi's d.
Christ-Siemens-Touraine s.	XLR; anodontia, cataracts, frontal bossing, MR, pseudorhagades, saddle nose, sebaceous gland hyperplasia, sparse dry hair, thickened lips, thin brittle nails AKA anhidrotic ectodermal dysplasia
chronic granulomatous d.	**XLR**, AR, AD; cutaneous/systemic pyogenic infections, defective oxidative metabolism of neutrophils and monocytes; abscesses, acne (severe), diarrhea, FTT, furuncles, hidradenitis suppurativa, infections, paronychia (chronic), perirectal ulcerative stomatitis
Churg-Strauss s.	allergic granulomatous angiitis; rare-vasculitis with asthma, peripheral eosinophilia & recurrent pneumonia
Clark's nevi	dysplastic melanocytic nevi
Clouston's s.	AD; alopecia, cataracts, nail dystrophy & clubbing, MR (some), palmoplantar hyperkeratosis, seizures (some), xerosis, French Canadians AKA hidrotic ectodermal dysplasia
Coat's d.	retinal telangiectasias & occasional skin telangiectasias
Cobb's s.	sporadic; dermatomal capillary malformation over spinal vascular malformation, angiokeratoma circumscriptum
Cockayne's s.	AR; aged appearance, cachexia, cataracts, growth arrest, microcephaly, neurological deterioration, photodistributed erythema, subcutaneous fat loss, early death
Cockayne-Touraine	dominant dystrophic epidermolysis bullosa
Coffin-Siris s.	AD; MR, sparse scalp hair, lax joints, bushy eyebrows, low nasal bridge, hypertrichosis
common variable immunodeficiency s.	late onset decreased immunoglobulins IgA, IgG, IgM; autoimmune diseases, alopecia areata, chronic giardiasis, eczema, recurrent otitis media, *Trichophyton rubrum*, URI infections, vitiligo
congenital self-healing reticulohistiocytosis	Hashimoto-Pritzker d.
congenital telangiectatic erythema	Bloom's s.
congenital total lipodystrophy	Lawrence-Seip s.
congenital varicella s.	chorioretinitis, cortical atrophy, cutaneous scars, limb hypoplasia
Conradi-H[illegible]nermann s.	X-dominant, male lethal ichthyosiform erythroderma in Blaschko's lines, cataracts & asymmetric limb defects AKA chondrodysplasia punctata
Consular d.	gnathostomiasis
contact urticaria s.	**stage 1**; localized urticaria restricted to the area of contact **stage 2**; generalized urticaria, including angioedema

	stage 3; urticaria associated with bronchial asthma **stage 4**; urticaria associated with anaphylactic reactions
COPS s.	*c*alcinosis cutis, *o*steoma cutis, *p*oikiloderma & *s*keletal abnormalities
Cornelia de Lange s.	de Lange s.
Corrigan's sign	purple line at junction of teeth & gum; chronic copper poisoning
Coulomb, pseudoscars of	stellate scars of the hands
Cowden's s., Cowden d.	AD; oral papillomatosis, palmoplantar keratoses, arched palate, fissured tongue, caries, GI hamartomatous polyps, GU cysts, breast cancer AKA multiple hamartoma s.
craniofacial dysostosis	Crouzon's d.
CREST s.	*c*alcinosis cutis, *R*aynaud's phenomenon, *e*sophageal dysfunction, *s*clerodactyly, *t*elangiectasia
cri du chat s.	partial 5p deletion; hypertelorism, microcephaly, MR, high-pitch cry, simian crease
Crocker, dermatitis repens of	acrodermatitis continua (Hallopeau)
Cronkhite-Canada s.	alopecia, diffuse palmar hyperpigmentation, spotty hypopigmentation of dorsal hands, nail dystrophy, GI polyposis & adenocarcinoma (15%), malabsorption with hypoproteinemia, electrolyte disturbance
Cross-McKusick-Breen s.	AR; oculocutaneous albinism, gingival hypertrophy, microphthalmus, small opaque corneas, oligophrenia with spasticity, high arched palate & scoliosis AKA oculocerebral-hypopigmentation s.
Crouzon's d.	AD, 10q26; craniofacial dysostosis
Crowe's sign	café-au-lait macules of axillae (axillary freckling); neurofibromatosis
Crow-Fukase s.	POEMS s.
Cullen's sign	blue periumbilical discoloration; acute pancreatic blood extravasation
Curth & Macklin, ichthyosis hystrix of	AD, very rare; resembles EHK with variable involvement, porcupine-like or verrucous hyperkeratosis
Cushing's s.	excessive glucocorticoids; characteristic habitus, cutis marmorata, easy bruising, ecchymoses, impaired wound healing, petechiae, striae, transparent epidermis, thinned dermis
cutis marmorata telangiectatica congenita	sporadic; atrophic reticulated vascular patch, ipsilateral hemiatrophy/hypertrophy, glaucoma, MR
Dabska's tumor	endovascular papillary angioendothelioma
Danoff s.	AD?; adrenocortical micronodular dysplasia, atrial myxoma, lentigines, spindle cell tumors
Darier's d., Darier-White d.	AD, 12q23-24.1, disrupted keratin tonofilament-desmosome complex; keratotic papules in seborrheic distribution, nail dystrophy, palmar pits, acrokeratoses AKA keratosis follicularis
Darier's sign	firm stroking of pigmented macule leads to mast cell mediator release & edema; urticaria pigmentosa (pathognomonic)
Davis Colley d.	keratosis punctata palmaris et plantaris
de Lange s.	brachycephaly, bushy eyebrows, carp mouth, coarse hair, depressed nose bridge, dwarfism, low set ears, MR, simian crease, webbed neck AKA Cornelia de Lange s.
De Sanctis-Cacchione s.	AR; subset of xeroderma pigmentosum with MR, retarded growth, gonadal hypoplasia, sometimes neurologic degeneration & ocular abnormality AKA xerodermic idiocy
DeBarsy's s.	cataracts, corneal opacities, ear dysplasia, growth retardation, joint hypermobility, microcephaly, MR, progeroid facies, pronounced nasolabial fold, skin wrinkling
Degos' acanthoma	benign epidermal tumor with glycogen containing epidermal cells AKA clear cell acanthoma
Degos' s.	multisystem lymphocytic vasculitis CNS & GI involvement AKA malignant atrophic papulosis
Delleman-Oorthuys s.	sporadic; cutis aplasia, skin tags, ocular defects, CNS defects
delusion of parasitosis	Ekbom's d.
Dennie-Morgan lines	accentuated folds of lower eyelid; atopic patients (non-diagnostic)
Dercum's d.	multiple painful lipomas; AKA adiposis dolorosa

dermatitis exfoliativa neonatorum	Ritter's d., Staphylococcal scalded skin s.
dermatomyositis	Wagner-Unverricht d.
Desmons' s.	KID s.
DiGeorge s.	teratogen caused; abnormal development of 3rd & 4th pharyngeal pouches; thymic & parathyroid hypoplasia, deficient cell mediated immunity; abnormal facies, congenital heart defects, decreased T cells, esophageal atresia, severe recurrent candidiasis infections
disseminated pagetoid reticulosis	Ketron-Goodman d.
Donohue's s.	absent subcutaneous fat, elfin face, hirsutism, thickened skin AKA leprechaunism
Dowling-Degos' d./s.	AD; brownish reticulate macules in flexural areas AKA reticulate pigmented anomaly of the flexures
Dowling-Meara EBS	epidermolysis bullosa herpetiformis
Down s.	trisomy 21; epicanthal fold, flat nose, short phalanges, widened spaces between 1st & 2nd digits of hands & feet, MR
Dubois' sign	short 5th digit; congenital syphilis
Dubokowitz's s.	broad nasal tip, eczematous lesions in infancy, epicanthal folds, micrognathia, MR, ptosis, sparse scalp hair, subcutaneous tissue diminished
Dukes' d.	mild febrile childhood illness with erythematous exanthem; Coxsackie ECHO virus group AKA scarlatinella, fourth d.
Duncan's d.	X-linked lymphoproliferative d.; abnormal immune response to EBV infection, early death or acquired dysgammaglobulinemia, chronic infectious mononucleosis, malignant lymphoma, T-lymphocyte depletion
Dunnigan s.	fat loss of limbs associated with DM AKA acquired partial face-sparing lipodystrophy
Dupuytren's d.	palmoplantar fibromatosis; dimpled palmar skin over 4th metacarpal, progressing to contracture
dyskeratosis congenita	Zinsser-Cole-Engman s.
dysplastic nevus s.	dysplastic nevi in patient at risk for familial or nonfamilial malignant melanoma
EEC s.	AD; *e*ctodermal dysplasia with *e*ctrodactyly, *c*left lip/palate, blepharitis, speckled irides, MR
Ehlers-Danlos s.	*see table at end of this section*
Ekbom's d.	delusion of parasitosis
Elejalde s.	AR; bronze skin after sun exposure, CNS dysfunction, hypotonia, MR, seizures, silver hair
Elliot's sign	induration of edge of syphilitic skin lesion
EMG s.	*e*xomphalos, *m*acroglossia, *g*igantism AKA Beckwith-Wiedemann s.
Enroth's sign	abnormal fullness of eyelids; Graves' disease
eosinophilic cellulitis	Well's s.
eosinophilic fasciitis	Schulman's s.
epidermal nevus s.	sporadic; ILVEN; café-au-lait macules, epidermal nevi, hemangiomas, kyphoscoliosis, limb deformities, lipodermoid tumors, MR, seizures AKA Schimmelpenning s., Solomon s.
Epstein's pearls	milia in oral cavity (palate) usually in infants
erythema multiforme major	Stevens-Johnson s.
erythema multiforme minor	Hebra's d.
erythrokeratoderma viriabilis	Mendes de Costa s.
erythroplasia of Queyrat	SCC of glans penis
erythropoietic porphyria	G∫nther d.
excited skin s.	patch testing; false + reactions (up to 40%), hypersensitivity caused by strong + reaction to one allergen AKA angry back s.
exomphalos-macroglossia-	Beckwith-Wiedemann s.

gigantism s.	
Fabry's d.	XLR; storage disease of glycosphingolipid catabolism, deficiency of α-galactosidase A leading to accumulated ceramide trihexoside in CV & renal systems; telangiectases in bathing suit distribution, corneal opacities, burning pain of palms, soles & abdomen; leg edema, osteoporosis, retarded growth, delayed puberty AKA angiokeratoma corporis diffusum
familial atypical multiple mole melanoma s.	dysplastic nevi with GI & pancreatic malignancy AKA FAMMM s.
familial dysautonomia	Riley-Day s.
familial dyskeratotic comedones	AD; widespread comedonal lesions on extremities
familial Mediterranean fever	AR; fever, peritonitis, pleurisy, purpura, renal amyloidosis, synovitis, urticaria, vasculitis nodules
FAMMM s.	familial atypical multiple mole melanoma s.
Fanconi's s.	vitamin D resistant rickets, glucosuria, aminoaciduria, acidosis, hypouricemia, hypokalemia, flexion deformities, generalized ichthyosis & FTT
Farber's d.	lipogranulomatosis
Favre-Racouchot d.	open comedones in background of dermatoheliosis on temples AKA nodular elastosis with cysts & comedones
Felty's s.	anemia, frequent leg ulcers, leukopenia, pigmented lower extremity macules, RA, splenomegaly, thrombocytopenia
Feuerstein & Mims s.	epidermal nevus s. AKA Schimmelpenning s., Solomon s.
fifth d.	erythema infectiosum
Filipovitch's sign	yellow discoloration of palms & soles; typhoid fever AKA palmoplantar sign
Finkelstein's d.	acute hemorrhagic edema of infancy
Fisch's s.	deafness, partial heterochromia, premature graying hair
Fischer-Jacobsen-Clouston s.	AD; "drumstick fingers," growth retardation, hair abnormalities, MR, palmoplantar scaling, thickened nails AKA hidrotic ectodermal dysplasia, PED type X
Fitzpatrick sign	lateral compression produces dimpling; dermatofibroma
Flegel's d.	keratinous papules of calves AKA hyperkeratosis lenticularis perstans
focal dermal hypoplasia	Goltz s.
folliculitis decalvans	Quinquaud's d.
Fordyce's angiokeratoma	small benign blood vessel tumor on scrotum & labia majora
Fordyce's condition	benign ectopic sebaceous glands on oral mucosa, genital mucosa, esophagus or larynx
Forscheimer spots	pinpoint rose colored macules & petechiae on soft palate; rubella
fourth d.	Dukes' d.
fourth venereal d.	gangrenous & ulcerative balanoposthitis or granuloma inguinale
Fox-Fordyce d.	females (9x); axillae & pubic chronic, pruritic follicular eruption of apocrine glands
Fran÷ois s.	AR; nodules of hands, nose, ears, osteochondrodystrophy AKA oculomandibulofacial s.
Franceschetti-Jadassohn s.	AD, onset >2 years old; heat intolerance, hypohidrosis, palmoplantar hyperkeratosis, reticular hyperpigmentation, yellow teeth
Frey's s.	auriculotemporal s.; gustatory sweating in malar area following parotid gland damage
Futcher's line	pigmentary demarcation between dorsal & ventral forearm AKA type A lines, Voigt's lines
Gamborg-Nielson PPK	AD; diffuse glove & stocking keratoderma with constricting digital bands, nail abnormalities, angular chelitis, hyperhidrosis, developmental retardation AKA mal de Meleda
Gardner's s.	AD; colon polyps progressing to carcinoma, dental anomalies, desmoid tumors, epidermal cysts, fibromas, ocular fundus pigmentation, osteomas, retroperitoneal fibrosis, thyroid cancer
Gardner-Diamond s. (purpura)	autoerythrocyte sensitivity (thought to be psychogenic); painful bruises & ecchymoses on legs, arms, face, syncope, abdominal pain, vomiting

Gaucher's d.	AR; lysosomal storage disease, deficiency of β-glucocerebrosidase; café-au-lait macules, collodion baby, ichthyosis
Gianotti-Crosti s.	young children; HBV, coxsackie A16, EBV, CMV, parainfluenza virus, RSV, group A streptococcal; fever, malaise, HSM, copper red flat topped firm papules; face, extremities & buttocks-progress to plagues & scales AKA acrodermatitis papulosa infantum, infantile papular acrodermatitis, papular acrodermatitis of childhood
Gilchrist's d.	North American blastomycosis
Giroux-Barbeau s.	AD; erythrokeratoderma with ataxia
glucagonoma s.	glucagon-producing pancreatic tumor; dermatitis, necrolytic migratory erythema, anemia, carbohydrate intolerance, hypoaminoacidemia, stomatitis, weight loss
Goldenhar's s.	triad of accessory tragi, auricular fistulas, epibulbar dermoids or lipodermoids
Goldstein's sign	wide space between first two toes; cretinism, Down s.
Goltz s.	focal dermal hypoplasia
Good's s.	thymoma with acquired hypogammaglobulinemia
Goplans' d.	localized hypohidrosis & painful feet AKA burning feet s.
Gorham's d.	sporadic; venous malformations, lymphatic malformations, replacement of bone with fibrous tissue
Gorlin's s., Gorlin-Goltz s.	AD, 9q21; early BCC's, palmoplantar pits, cysts of mandible & maxilla, bone, intracranial calcification, eye & reproductive tract abnormalities, medulloblastomas, mental retardation, characteristic facies AKA basal cell nevus s.
Gorlin's sign	ability to touch tip of nose with tongue; Ehlers-Danlos s., normal persons
Gottron's papules	inflammatory papules over dorsal hand joints; dermatomyositis
Gottron's sign	violaceous erythema over knuckles, elbows, medial malleoli, patella; dermatomyositis (pathognomonic)
Gougerot-Blum s.	pigmented purpuric lichenoid dermatitis
Graham-Little s., Graham-Little-Feldman s.	cicatricial alopecia with follicular lichen planus of skin & scalp, may be associated with noncicatricial alopecia of axillae & pubic areas AKA Graham-Little-Piccardi-Lassueur s.
granuloma multiforme	Mkar d.
granulomatous chelitis	Melkersson's s., Melkersson-Rosenthal s.
Greenblatt's sign	linear depression over Poupart's ligament separating draining lymph nodes; lymphogranuloma venereum AKA groove sign of Greenblatt
Greither's s.	palmoplantar keratoderma with transgrediens to dorsal surfaces
Griscelli s.	AR; albinism with hypomelanosis, silver hair, pyogenic infection, HSM, thrombocytopenia, immune deficiency AKA hypopigmentation-immunodeficiency d.
Grisolle's sign	when skin is stretched, a papule is felt; smallpox (historical)
Grover's d.	pruritic erythematous papules typically middle aged or older males on trunk, shoulders, neck, thighs AKA transient acantholytic dermatosis
Gunther's d.	AR, 10q25.2-q26; uroporphyrinogen III cosynthetase; early; immediate photosensitivity with burning, edema, erosions late; mutilating scarring in sun exposed areas, cicatricial alopecia, hypertrichosis, brown teeth, photophobia, hemolytic anemia, splenomegaly AKA erythropoietic porphyria
Haarscheibe receptor	receptor associated with Merkel cells AKA Pinkus corpuscle, hederiform ending
Hailey-Hailey d.	AD, defect in tonofilament-desmosome complex, adhesion molecule abnormalities(?); recurrent vesicles & bullae in intertriginous areas AKA benign familial pemphigus
Hallermann-Streiff s.	dyscephaly, parrot nose, mandibular hypoplasia, proportionate nanism, hypotrichosis, bilateral congenital cataracts, and microphthalmia AKA oculomandibulofacial s.
Hallopeau, acrodermatitis continua of	sterile pustular eruption of fingers or toes that extends proximally
Hallopeau, pemphigus vegetans of	localized pemphigus vulgaris
Hallopeau-Siemens	generalized recessive dystrophic epidermolysis bullosa
Hand-Schʃller-Christian d.	Langerhan's cell histiocytosis

Hansen's d.	leprosy
harlequin s.	unilateral facial flushing & sweating
Hartnup's d.	AR, 2pter-q32.3, defective amino acid transport; photodistributed erythema, scale, ataxia, psychiatric disturbances, aminoaciduria, stomatitis
Hashimoto-Pritzker d.	congenital self-healing reticulohistiocytosis
Hatchcock's sign	tenderness elicited when running finger toward angle of jaw; mumps
Hebra's d.	erythema multiforme (minor)
Heck's s.	focal epithelial hyperplasia; mucosal HPV, primarily in native American children
Heerfordt s.	chronic sarcoid, anterior uveitis, parotid gland enlargement, facial nerve palsy
Heller, median canaliform dystrophy of	split midline nail with fir tree-like appearance, especially of thumbs
hemochromatosis	AR, 6, or acquired; deposition of hemosiderin in parenchymal cells, causing bronze skin pigmentation, dysfunction of liver, pancreas, heart, pituitary, arthropathy, diabetes, cirrhosis, HSM, hypogonadism, loss of body hair
hepatolenticular degeneration	Wilson's d.
hereditary hemorrhagic telangiectasia	Osler-Weber-Rendu s.
hereditary painful callosity s.	PPK with oral mucosa hyperkeratosis
heredopathia atactica polyneuritiformis	Refsum's d.
Herlitz variant	junctional epidermolysis bullosa AKA JEB-gravis, JEB-lethal
Hermansky-Pudlak s.	AR; pigment dilution, nevi, SK, SCC, BCC, ecchymoses, petechiae, cream colored hair & skin, photophobia, strabismus, hemorrhage, granulomatous colitis, cardiomyopathy
Hertogh's sign	lateral thinning of eyebrow hair; atopic dermatitis, hypothyroidism
Heubner's arteritis	endarteritis of medium & large arteries resulting in thrombotic infarction; syphilis
HID s.	sporadic; AKA hystrix-like ichthyosis with deafness
hidrotic ectodermal dysplasia	Clouston's s.
Higoumenakis sign	unilateral irregularly enlarged medial clavicle; late congenital syphilis
Hines & Bannick s.	hyperhidrosis & hypothermia associated with diencephalic epilepsy or hypothalamic storm
Hippocratic nail	onychogryphosis AKA Osler's toe, ram's horn nail
Hoffman-Zurhelle s.	nevus lipomatosis cutaneous superficialis
Hoigne reaction	psychotic symptoms secondary to procaine in procaine penicillin, pseudo-anaphylactic reaction; syphilis
Holmes-Adie s. with anhidrosis	Ross s.
homocystinuria	AR or non-genetic; developmental delay, failure to thrive, neurologic abnormalities, hematologic abnormalities
Hopf, acrokeratosis verruciformis of	AD; small warty papules on extensor surfaces with punctate keratoses of palm
Hori's nevus	acquired circumscribed dermal melanocytosis of the face
Howel-Evans' s.	AD; 17q23, palmoplantar keratoderma (tylosis) ages 5-15, later esophageal cancer
Hunt's s.	Ramsay-Hunt s.
Hunter s.	XLR; mucopolysaccharidosis; deficiency of iduronate-2-sulfatase, similar to Hurler s.
Huriez s.	AD; mild keratoderma with scleroatrophy & sclerodactyly, nail changes; SCCs of dorsal hand
Hurler s.	AR; mucopolysaccharidosis, deficiency of L-iduronidase; thick hyperpigmented inelastic skin, corneal clouding, death by age 10
Hutchinson's freckle	large lentigo with grossly irregular borders; may progress to lentigo maligna melanoma
Hutchinson's prurigo	actinic prurigo
Hutchinson's sign	diffusion of pigment from proximal nail matrix; melanoma
Hutchinson's teeth	widely spaced small notched upper incisors; congenital syphilis (pathognomonic)

Hutchinson-Gilford s.	progeria
Huygens' effect	condenser of microscope defocused to obtain a partial phase effect
hyalinosis cutis et mucosae	Urbach-Wiethe d.
hyper IgE s.	eczema, recurrent cutaneous & systemic infections, decreased neutrophil chemotaxis, reversible ichthyosis, fungal infections, CMC, urticaria, incontinentia pigmenti
hypereosinophilic s.	eosinophilia & eosinophilic infiltrate of organs
hyperkeratosis lenticularis perstans	Flegel's d.
hypervitaminosis A s.	dry lips & skin, hair loss, sticky skin sensation, extraosseous calcifications, embryotoxic & teratogenic effects from systemic retinoids
hypocomplementemic urticarial vasculitis s.	urticarial vasculitis, angioedema, eye inflammation, arthritis/arthralgia, mild renal d., obstructive pulmonary d., serum complement activation with hypocomplementemia, C1q precipitin
hypohidrotic ectodermal dysplasia	AR, similar to Christ-Siemens-Touraine s.
hypomelanosis of Ito	AD; guttate & whorled hypopigmentation, conductive hearing loss, ocular abnormalities, MR, seizures, skeletal deformities, female predominant AKA incontinentia pigmenti achromians
IBIDS s.	*i*chthyosis plus BIDS (*b*rittle hair, *i*ntellectual impairment, *d*ecreased fertility, *s*hort stature) AKA Tay's d.
IFAP s.	*i*chthyosis *f*ollicularis with *a*lopecia & *p*hotophobia
IgA deficiency s.	AR; 1: 600, atopy, asthma, autoantibodies, chronic gastroenteritis, DM, milk allergy, non-tropical sprue, PA, SLE, thyroiditis, URI
IgM deficiency s.	1:1000, infections; pneumococci, meningococci, verrucae, eczema, autoimmune features
incontinentia pigmenti	Bloch-Sulzberger s.
incontinentia pigmenti achromians	hypomelanosis of ITO
infantile papular acrodermatitis	Gianotti-Crosti s.
intestinal lipodystrophy	Whipple's d.
Jackson-Sertoli s.	AD, disruption of K17 expression; limited focal plantar keratoderma, woolly scalp hair, straight eyebrow hair, natal teeth AKA pachyonychia congenita type II, PED type II
Jadassohn-Lewandowsky s.	AKA pachyonychia congenita AD; **Type I**; thickened dystrophic nails, blisters around callosities, palmoplantar hyperhidrosis, leukokeratosis oris, follicular keratosis, laryngeal keratosis with hoarseness **Type II**; natal teeth and premature anodontia, steatocystoma multiples, follicular & palmoplantar keratosis without oral leukokeratosis **Type III**; corneal dystrophy, mucocutaneous keratosis
Jadassohn-Pellizari anetoderma	postinflammatory anetoderma
Janeway lesion	non-tender erythematous macule on proximal palms & soles; **endocarditis** (5%), gonococcemia, hemolytic anemia, SLE, typhoid fever
Jarisch-Herxheimer reaction	febrile reaction in patients treated with penicillin from unknown cause; syphilis
Jellinek's sign	brownish pigmentation on lid margins; hyperparathyroidism AKA Rasin's sign
Job s.	hyper IgE s. with red hair, atrophic nails, hyperextensible joints, cold abscesses
Johanson-Blizzard s.	AR; microcephaly, MR, congenital absent skin posterior midline scalp, sparse hair, ala nasi hypoplasia, café-au-lait macules, hypoplasia of nipples & areola
Johnston's s.	XLR/monogenic autosomal; hyperkeratotic collodion baby-type skin with arthrogryposis & posterior column hypoplasia
Jones-Mote reaction	cutaneous basophil hypersensitivity; may occur with allergic contact dermatitis
Kallmann's s.	anosmia & hypogonadotrophic hypogonadism, associated with X-linked ichthyosis
Kanagawa phenomenon	hemolysin produced by *Vibrio parahaemolyticus* associated with diarrhea

Kaposi-Irgang d.	lupus panniculitis, lupus profundus
Kasabach-Merritt s.	sporadic; infancy-platelet trapping in large hemangioma leading to anemia, consumption coagulopathy, CHF, DIC, GI bleed, thrombocytopenia
Kast's s.	Maffucci's s.
Kawasaki s.	infants/children; fever, edematous reddened palms & soles, polymorphous truncal exanthem, bilateral conjunctivitis, mucosal erythema & strawberry tongue, cervical LAD, ulcerative gingivitis, enlarged cervical lymph nodes, cardiac complications (25%) AKA mucocutaneous lymph node syndrome
keratoma hereditaris mutilans	Vohwinkel's s.
keratosis follicularis	Darier's d., Darier-White d.
keratosis punctata palmaris et plantaris	Buschke-Fischer-Brauer d.
Kerr's sign	alteration of the texture of skin below the somatic level of spinal cord lesion
Ketron-Goodman d.	generalized (disseminated) pagetoid reticulosis (mycosis fungoides)
KID s., keratitis-ichthyosis-deafness s.	sporadic, AR, AD; *k*eratitis, *i*chthyosiform erythroderma, & profound neurosensory *d*eafness alopecia, decreased sweating, malformed teeth, nail dystrophy, sometimes inflammatory corneal vascularization SCC may develop in childhood AKA Senter s., Desmons' s., PED type XVI
Kimura's d.	angiolymphoid hyperplasia with eosinophilia with different nature of proliferating vascular cells
Kindler-Weary s., Kindler s.	acral blistering, poikiloderma, reticulate hyperpigmentation, sclerodactyly, scleroatrophy
kinky-hair s.	Menkes s.
kissing d.	popular name; infectious mononucleosis
Kitamura, reticulate acropigmentation of	AD; reticulate, slightly depressed pitting brown hyperpigmentation initially on dorsal hand, & then generalizing
Klippel-Trenaunay-Weber s., Klippel-Trenaunay s.	sporadic, usually unilateral lower extremity lesions; angiokeratomas, AV fistulas, hemangiomas, hypertrophy of bone & soft tissue, lymphatic malformation, nevus flammeus, skin varices AKA Parkes-Weber s.
Koebner phenomenon	physical trauma leading to lesion spread; acquired perforating dermatosis, bullous pemphigoid, contact dermatitis, Darier's d., erythema multiforme, Grover's d., Hailey-Hailey d., lichen nitidus, lichen planus, porokeratosis of Mibelli, psoriasis (20%), pyoderma gangrenosum, sarcoid, verrucae, vitiligo (30%) AKA isomorphic phenomenon
Koebner, EBS of	generalized epidermolysis bullosa simplex
Koenen tumors	fibromas developing around fingers & toes; tuberous sclerosis
Koplik spots	white spots on buccal mucosa; coxsackievirus A16, echovirus 9, **measles**
Krisovski's sign	cicatricial lines radiating from the mouth; congenital syphilis
Kwashi shakes	Parkinsonian-like tremors in recovery phase; kwashiorkor
Kyrle's d.	AR(?); rare perforating disorder; papules with hyperkeratotic plugs coalescing to plaques
LAMB s.	*l*entigines, *a*trial myxoma, *m*ucocutaneous myxomas, & *b*lue nevi
Langer-Giedion s.	AAD, 8q24.11; redundant skin, sparse hair, bulbous nose, MR, hyperextensible joints
Langerhan's cell histiocytosis	malignant lymphoma, lung carcinoma, post-chemotherapy leukemia (esp. etoposide) AKA Hand-Schʃller-Christian d.
Laugier-Hunziger s.	lentigines of lips, mouth, genitalia, perineal, nail changes (pigmented bands, hyperpigmentation)
Lawrence-Seip s.	AR; near total fat loss with somatic hypertrophy, acanthosis nigricans, diabetes mellitus AKA congenital total lipodystrophy
Leiner's d.	C5 dysfunction leading to decreased serum phagocytosis (opsonic activity); eczema, seborrhea, erythroderma, diarrhea, recurrent gram negative infections, muscle wasting, & FTT in infants
LEOPARD s.	AD; *l*entigines, *e*lectrocardiographic abnormalities, *o*cular hypertelorism, *p*ulmonary stenosis, *a*bnormal genitalia, *r*etarded growth & development, *d*eafness
leprechaunism	Donohue's s.
leprosy	Hansen's d.

Leroy's s.	rare lipomucopolysaccharide disorder with skin thickening & bone changes
Leser-Trelat sign	multiple eruptive SK's with pruritus; acanthosis nigricans, cancer (breast, colon, lung, prostate, stomach), lymphoma, malignant melanoma, mycosis fungoides, primary lymphoma (brain)
Letterer-Siwe d./s.	AR; reticuloendotheliosis of childhood, eczema, hemorrhage, hepatosplenomegaly, progressive anemia AKA acute disseminated histiocytosis X
leukocyte alkaline phosphatase deficiency	AR; defective antibacterial & antifungal protection, increased IgE & eosinophils, normal chemotaxis; eczema, pulmonary infections
leukocyte myeloperoxidase deficiency	AR; defective antibacterial, antifungal protection
Lewis hunting response	alternating vasodilatation & vasoconstriction during cold exposure (especially of hands)
linear sebaceous nevus s.	nevus sebaceous of Jadassohn
lipoid proteinosis	Urbach-Wiethe d.
Lisch nodules	melanocytic iris hamartomas; neurofibromatosis
Loffler's s.	pulmonary infiltration & eosinophilia; rarely occurring with cutaneous larva migrans
Lofgren's s.	arthralgia, bilateral hilar adenopathy, cough, erythema nodosum, fever
Louis-Bar s.	AR, 11q22; ataxia, cutaneous & bulbar telangiectasias, café-au-lait macule, decreased IgA, IgE & lymphocytes, granuloma, lymphoma, nystagmus, respiratory infections, sclerodermoid changes, solar lentigines AKA ataxia-telangiectasia
Lovibond's angle	cuticle angle greater than 180° indicates *clubbing*
low sulphur hair s.	trichothiodystrophy
Lucio phenomenon/reaction	hemorrhagic infarcts; Latapi's lepromatosis
Lyell's s.	toxic epidermal necrolysis
Madelung's d.	benign symmetric lipomatosis
Maffucci's s.	sporadic; enchondromatosis, limb deformities, multiple cutaneous/visceral hemangiomas, venous malformations, short stature, sarcomas AKA Kast's s.
MAGIC s.	*m*outh *a*nd *g*enital ulcerations with *i*nflamed *c*artilage
Majocchi's d.	purpura annularis telangiectoides
Majocchi's granuloma	deep fungal infection producing granulomatous response AKA trichophytic granuloma
Mal de Meleda	AR; palmoplantar keratoderma with transgrediens
malignant atrophic papulosis	Degos' s.
Marfan s.	AD, sporadic 5 & 15 elastic degeneration; striae distensae, elastosis perforans serpiginosa, arachnodactyly, ocular defects, skeletal defects
Marinesco-Sj¬gren s.	AR; cerebellar ataxia, mental & growth retardation, cataracts, brittle fingernails, sparse incompletely keratinized hair
Marjolin's ulcer	carcinoma appearing in any type of skin scar
Maroteaux-Lamy s.	mucopolysaccharidosis; deficiency of N-acetylgalactosamine-4-sulfatase, dermatan sulfate in urine & metachromatic granules in leukocytes
Masson's pseudoangiosarcoma	intravascular papillary endothelial hyperplasia
mastocytosis s.	episodic s. in some patients with systemic mastocytosis; bone lesions, HSM, skin lesions
Mauserung phenomenon	stratum corneum shed in full-thickness sheets, leaving red tender base; bullous ichthyosis
McCune-Albright s.	sporadic; hyperthyroidism, precocious puberty, café-au-lait macule, polyostotic fibrous dysplasia, AKA Albright's s.
McDonald's acne	acne exacerbation from work near a deep fat fryer
Mee's lines	paired narrow white transverse nail lines; arsenic poisoning
Meischer's granuloma	actinic GA; annular elastolytic granuloma, erythema nodosum, Sweet's s. AKA actinic granuloma
Meischer's nevi	dome shaped nevi on face
Meissner's receptor	upper dermal papillae receptors; unknown function
Meleney's gangrene	progressive bacterial synergistic gangrene
Meleney's ulcer	Meleney's gangrene with burrowing necrotic fistulas through tissue planes

Melkersson's s., Melkersson-Rosenthal s.	AD; triad of recurrent noninflammatory orofacial swelling, relapsing facial paralysis & fissured tongue (lingua plicata) AKA granulomatous chelitis
Mendes de Costa s.	AD; ichthyosis with transient migratory macular erythroderma & fixed hyperkeratotic plaques AKA erythrokeratoderma viriabilis
Menkes' s., Menkes' kinky hair s.	XLR, copper transport abnormality, tyrosinase deficiency; characteristic facies, trichorrhexis nodosa, pili torti, monilethrix, severe cerebral degeneration & arterial change, death in infancy
Mibelli, porokeratosis of	AD, rare; plaques with coronoid lamellae usually on acral surfaces AKA classic porokeratosis
Michelin tire baby	appearance of rolls of fatty tissue; generalized congenital smooth muscle hamartoma, generalized nevus lipomatosus
MIDAS s.	*mi*crophthalmia, *d*ermal *a*plasia, *s*clerocornea
Miescher-Melkersson-Rosenthal s.	Melkersson's s.
Milroy's d.	primary (essential) lymphedema
Mkar d.	granuloma multiforme
Mondor's d.	thrombophlebitis of large subcutaneous veins of lateral chest & breast
Mongolian spots	blue-brown pigmented patch on lower back & buttocks of infants; Asians, blacks, inborn error of metabolism; GMI type 1 gangliosidosis
monilethrix	AD; beaded hair that breaks less than an inch long
Montgomery's s.	xanthoma disseminatum
Moon's molars	abnormal teeth; congenital syphilis AKA mulberry molars
Morquio's s.	AR; mucopolysaccharidosis, excretion of keratan sulfate in urine, genu valgum, pectus carinatum, deafness, corneal clouding, platyspondyly, short neck & trunk
Morton's neuroma	3rd or 4th inter-metatarsal foot space fibrosis & vascular proliferation with nerve entrapment
Moynahan's s.	multiple symmetric lentigines, congenital mitral valve stenosis, dwarfism, genital hypoplasia, MR AKA progressive cardiomyopathic lentiginosis
Mucha-Habermann d.	PLEVA
Muckle-Wells s.	AD; chronic relapsing urticaria, fever, arthralgias, deafness, renal amyloidosis
mucocutaneous lymph node s.	Kawasaki d.
mucopolysaccharidoses	*see table at end of this section*
Muehrcke's lines	paired white parallel transverse nail bands; hypoalbuminemia
Muir-Torre s.	AD, 2p; multiple sebaceous tumors; adenoma, multiple GI carcinomas, hyperplasia, BCC, KA, GU & GI carcinoma. AKA Torre s.
multiple carboxylase deficiency	deficiency of holocarboxylase synthetase or biotinidase, causing deficiency of carboxylase; alopecia, ataxia, developmental delay, hyperammonemia, hypotonia, metabolic ketoacidosis, organic aciduria, seizures, rash
multiple hamartoma s.	Cowden's s.
multiple lentigines s.	LEOPARD s., Moynahan s.
multiple mucosal neuroma s.	Sipple's s., multiple endocrine neoplasia type 2b
mutilating keratoderma	Vohwinkel's s.
Naegeli-Franceschetti-Jadassohn s.	AD; reticulate hyperpigmentation hypohidrosis, severe enamel defects with loss of dentition
nail-patella s.	hereditary osteo-onychodysplasia; absent patella, clinodactyly, micronychia, triangular lunulae
NAME s.	AD; *n*evi, *a*trial myxoma, *m*yxoid neurofibromas, *e*phelides, plus testicular tumors, adrenocortical d., pituitary adenomas
Nekam d.	keratosis lichenoides chronica
NERD s.	*n*odules, *e*osinophilia, *r*heumatism, *d*ermatitis; articular nodules, dermographism, episodic hand & foot edema, eosinophilia, generalized pruritic dermatitis, urticaria

Netherton's s.	AR; ichthyosis linearis circumflexa (pathognomonic), trichorrhexis invaginata (hair shaft defect), atopic diathesis, sometimes MR & aminoaciduria
Neu-Laxova s.	AR, fatal; abnormal face, eclabion, ectropion, IUGR, limb deformities, microcephaly, severe hyperkeratosis
Neumann, pemphigus vegetans of	more extensive pemphigus vulgaris than pemphigus vegetans of Hallopeau
neutral lipid storage d.	Chanarin-Dorfman s.
nevus elasticus	pseudoxanthoma elasticum
nevus fuscocaeruleus opththalmomaxillaris	unilateral, usually facial, slate-gray macules AKA nevus of Ota
nevus lipomatosus cutaneous superficialis	Hoffman-Zurhelle s.
nevus sebaceous of Jadassohn	linear sebaceous nevus s.
Nezelof's s.	AR, XL, spontaneous; thymic dysplasia with normal immunoglobulins, absent T cell function, chronic infections, chronic mucocutaneous *Candida*, purine nucleoside phosphorylase deficiency (some)
Nicolaides-Baraitser s.	brachydactyly, MR, prominent lower lip, sparse hair, short metacarpals
Niemann-Pick d., Niemann's d.	Five types; lysosomal storage disease deficiency of sphingomyelin phosphodiesterase with sphingomyelin accumulation in reticuloendothelial system
Nikolsky's sign	separation of dermal/epidermal layer with stroking; bullous impetigo, bullous pemphigoid, epidermolysis bullosa, GVH d., intracutaneous bulla formation, pemphigus erythematosus, pemphigus foliaceus, pemphigus vulgaris, SSSS, Stevens-Johnson s., **toxic epidermal necrolysis**
Nissl arteritis	endarteritis of small arteries & arterioles resulting in thrombotic infarction; syphilis
Noonan's s.	congenital heart disease, hypogonadism, ptosis, short stature, webbed neck
occipital horn s.	XLR form of cutis laxa
oculocerebral-hypopigmentation s.	Cross-McKusick-Breen s.
oculomandibulofacial s.	Fran÷ois s., Hallermann-Streiff s., mandibulo-oculofacial dyscephaly
Ofuji's d.	eosinophilic pustular folliculitis; sterile annular pustules-face, trunk, extremities
Ogna, EBS of	Norwegian cases of EBS
Olmsted's s.	massive, mutilating keratoderma with hyperkeratotic plaques & severe nail dystrophy with alopecia, follicular hyperkeratosis, oral leukokeratoses, psychomotor delay, short stature
Omenn's s.	AR; combined T & B cell immunodeficiency with alopecia, diffuse erythema, FTT, hyperkeratosis & recurrent infections
Osler's nodes	painful erythematous or hemorrhagic macules, papules or nodules on distal fingers; gonococcemia, hemolytic anemia, SLE, typhoid fever (*see also Janeway lesion*)
Osler's sign	small painful erythematous swellings (Osler's nodes) in skin of hands & feet; subacute bacterial endocarditis (10%), SLE
Osler's toe	onychogryphosis AKA Hippocratic nail, ram's horn nail
Osler-Weber-Rendu s.	AD, 9q33; punctate telangiectasias of ears, feet, hands, lips, tongue; epistaxis, GI & GU telangiectasias GI hemorrhage, recurrent epistaxis in childhood, pulmonary & hepatic AV fistulas, CNS aneurysms, AV malformations AKA hereditary hemorrhagic telangiectasia
osteogenesis imperfecta	*see table at end of section*
Ostertag s.	AD; hereditary systemic amyloid, hepatomegaly, hypertension, nephropathy
Ota, nevus of	unilateral, usually facial, slate-gray macules AKA nevus fuscocaeruleus opththalmomaxillaris
pachydermoperiostosis	Touraine-Solente-Gol— s.
pachyonychia congenita	Jadassohn-Lewandowsky s.
Pacinian corpuscles	receptors in deep dermis or subcutis especially in digits, associated with blood vessels, serving as rapidly adapting mechanoreceptors to vibrational stimuli

PACK s.	primary biliary cirrhosis, anti-centromere antibody, CREST (calcinosis cutis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly, telangiectasia) and keratoconjunctivitis sicca
pagetoid reticulosis	Woringer-Kolopp d.
painful bruising s.	women; purpuric painful ecchymoses with emotional stress, without preceding trauma
Pallister-Killian s.	circumscribed hypopigmentation on cranium, sparse eyebrows & eyelashes, sparse scalp hair, severe MR, hearing loss, seizures, ptosis, high forehead, hypertelorism, facial defects
Papillon-Lefℵvre s.	AR; palmoplantar keratoderma with transgrediens, keratotic plaques of elbows, knees, periodontitis, tooth loss, falx calcification
papular purpuric gloves and socks s.	Parvovirus B-19 in adults
paraneoplastic acrokeratosis	Bazex's s.
parasitic melanoderma	vagabond's d.
Parkes-Weber s.	Klippel-Tr—naunay-Weber s.
Parrot's lines	depressed linear scars radiating from anus, mouth, & nose like wheel spokes; congenital syphilis AKA rhagades
Parry-Romberg s.	facial hemiatrophy, hyperpigmentation & atrophy of dermis, subcutaneous fat, muscle & bone
Partingtons s.	FTT, hemiplegia, recurrent pneumonia, hyperpigmentation (generalized reticulate), seizures
Pasini	dominant dystrophic epidermolysis bullosa albopapuloid
Pastia's lines	linear petechiae; Kawasaki d., scarlet fever
Paterson's s., Paterson-Brown Kelly s., Paterson-Kelly s.	Plummer-Vinson s.
peeling skin s.	AR; cycles of spontaneous desquamation of full thickness stratum corneum sheets, generalized hyperkeratosis, palmoplantar hyperkeratosis with pruritus, underlying erythroderma
PEP s.	POEMS s.
Peruvian wart	vascular papules & nodules developing in crops; Carrion's d. AKA verruga peruana
Peutz-Jeghers s.	AD; lentigines (around mouth, eyes, lips, oral mucosa, hands, feet), GI polyps, GI malignancies (3%), cancer (breast, pancreas, reproductive organs), colic, intussusception, GI bleeding
Peyronie's d.	induration of corpora cavernosa of penis, producing fibrous chordee
phenylketonuria	AR; hyperphenylalaninemia due to phenylalanine 4-monooxygenase deficiency; eczema, hypopigmentation of hair & skin, MR, seizures, tumors, mousy odor
PIBIDS	*p*hotosensitivity plus IBIDS (*i*chthyosis, *b*rittle hair, *i*ntellectual impairment, *d*ecreased fertility, *s*hort stature)
piebaldism	AD; c-kit on 4q12, depigmented patches, white forelock
pigmented purpuric lichenoid dermatitis	Gougerot-Blum s.
Pinkus corpuscle	see *Haarscheibe receptor*
plasma cell balanitis (vulvitis)	Zoon's balanitis (vulvitis)
PLEVA	Mucha-Habermann d.
Plummer-Vinson s.	angular chelitis, dysphagia, hypochromic anemia, koilonychia & painful tongue AKA Paterson's s., Paterson-Brown Kelly s., Paterson-Kelly s., sideropenic dysphagia, Vinson's s.
POEMS s.	*p*olyneuropathy, *o*rganomegaly, *e*ndocrinopathy, *M* protein & *s*kin changes AKA Crow-Fukase s.
Pohl-Pinkus marks	hair shaft constrictions, acquired trichodystrophy; antimitotic drugs, emotional stress, systemic d.

poikiloderma congenitale	Rothmund-Thompson s.
polycystic ovary s.	Stein-Leventhal s.
popliteal web s.	congenital popliteal webs, cleft palate, pits (lower lip), toenail dysplasia
porphyria cutanea tarda	AD, 1p34, uroporphyrinogen decarboxylase gene, & sporadic/acquired; delayed photosensitivity, facial hypertrichosis, **scarring alopecia**, **milia**, **hypermelanosis**
porphyria, acute intermittent	AD 11q24, porphobilinogen deaminase; no skin features, acute attacks with seizures, peripheral neuropathy, weakness, abdominal pain, tachycardia, hyponatremia secondary to ADH secretion
porphyria, erythropoietic	AR, 10q25.2-q26; uroporphyrinogen III cosynthetase; early; immediate photosensitivity with burning, edema, erosions late; mutilating scarring in sun exposed areas, scarring alopecia, hypertrichosis, brown teeth, photophobia, hemolytic anemia, splenomegaly AKA G∫nther's d.
porphyria, hepatoerythropoietic	AR, 1p34, uroporphyrinogen decarboxylase gene, homozygous form of familial PCT; onset infancy, very rare, severe photosensitivity, hemolytic anemia, splenomegaly, dark urine, hypertrichosis, hyperpigmentation, sclerodermoid change, mutilating scars
porphyria, variegate	AD 14q32, protoporphyrinogen oxidase; bullae, erosions, scarring, milia, hypertrichosis, acute attacks precipitated by drugs, infection fever, alcohol, pregnancy
porphyria; erythropoietic coproporphyria	Extremely rare, little is known; elevated PROTO & COPRO in red blood cells.
porphyria; erythropoietic protoporphyria	AD, 18pter-p11.2, ferrochelatase deficiency; onset 1-4 years old. early; burning erythematous plaques in sun distribution. late; waxy thickened scarring with cholelithiasis, jaundice, anemia
porphyria; hereditary coproporphyria	AD, 9, coproporphyrinogen oxidase gene; onset young adults, delayed photosensitivity, acute attacks similar to PCT precipitating factors, usually less severe
postphlebitic s.	complications of deep venous thrombosis; chronic venous insufficiency, persistent edema, pain, purpura, increased cutaneous pigmentation, eczematoid dermatitis, pruritus, ulceration, & indurated cellulitis
Pott's d.	scrotal cancer in chimney sweeps from polycyclic aromatic hydrocarbons
Preus s.	arched palate, cataracts, dolichocephaly, generalized hypopigmentation, growth retardation, hypochromic anemia, psychomotor retardation, small teeth,
primary (essential) lymphedema	Milroy's d.
progeria	unknown inheritance; thin, atrophic skin, mottled hyperpigmentation, sparse hair, large cranium, micrognathia, osteoporosis, premature atherosclerosis, CHF, MI, short stature, short life span
progressive cardiomyopathic lentiginosis	Moynahan's s.
progressive pigmented purpuric dermatosis	Schamberg's d.
prolidase deficiency	AR; aminoacidopathy,deficiency of X-Pro dipeptidase, urinary excretion of imidodipeptides; chronic skin lesions, impaired motor & cognitive development, frequent infections, bone abnormalities
proteus s.	AD, sporadic; AV malformations, capillary malformation, growth & mental retardation, linear epidermal nevi, intracranial tumors, large at birth, lipomas, lymphatic-venous malformations, macrocephaly, macrodactyly, pigmented penile macules, scoliosis, soft tissue & bony hypertrophy of extremities, subcutaneous masses AKA Bannayan s.
pseudo-Darier's sign	urticarial wheal, induration, piloerection with stroking; congenital smooth muscle hamartoma
pseudo-Hutchinson's sign	discoloration of nail matrix; subungual hematoma
pseudopelade of Brocq	end stage cicatricial alopecia & fibrosis from; favus, folliculitis (scarring), LE, lichen planus, sarcoidosis, scleroderma
pseudoxanthoma elasticum	AR; basophilic degeneration of elastic tissue; flexural yellow macules & papules forming plaques, lax inelastic redundant skin, angioid streaks (retina), arterial insufficiency of

	lower extremities, calcified arteries, coronary insufficiency, hypertension, mitral valve prolapse, GI hemorrhage AKA nevus elasticus
purpura annularis telangiectoides	Majocchi's d.
Quincke pulsation	flushing of nail beds synchronous with heartbeat; aortic regurgitation
Quinquaud's d.	folliculitis decalvans
Rabson-Mendenhall s.	acanthosis nigricans, dental dysplasia, dystrophic nails, premature puberty
Raeder's s.	Horner's s., plus frontal/temporal headache & lacrimal sweating
Ramsay-Hunt s.	herpes zoster of facial & auditory nerves, external ear with ipsilateral facial paralysis occasional deafness, tinnitus, vertigo AKA geniculate neuralgia, herpes zoster auricularis, neuralgia facialis vera, otic neuralgia, Hunt's s.
Rapp-Hodgkin s.	AD, AR?; absent dermatoglyphics, cleft lip/palate, coarse scalp hair, dry skin, epiphora, ectropion AKA anhidrotic ectodermal dysplasia
Rasin's sign	brownish pigmentation on lid margins; hyperparathyroidism AKA Jellinek's sign
Raynaud's sign	acrocyanosis
Reed's s.	familial leiomyomatosis cutis et uteri
Refsum's d.	AR; phytanic oxidase deficiency; arrhythmias, ataxia, bony anomalies, deafness, hyperkeratosis, lenticular opacity, retinitis pigmentosa AKA heredopathia atactica polyneuritiformis
Reiter's s.	seronegative asymmetric arthropathy with one or more; cervicitis, circinate balanitis, conjunctivitis, dysentery, keratoderma blennorrhagicum, stomatitis, urethritis; males 9:1, HLA-B27 (80%)
REM s. , reticular erythematous mucinosis s.	women; photosensitive reticulated erythematous macules & papules with dermal mucin
Rendu-Osler-Weber s.	hereditary hemorrhagic telangiectasia AKA Osler-Weber-Rendu s.
reticulate pigmented anomaly of the flexures	Dowling-Degos' d.
Reye's s.	acute noninflammatory encephalopathy (lethargy, confusion, vomiting), hepatitis preceded by varicella (20-40%) & usually aspirin AKA infantile digital fibromatosis
Richner-Hanhart s.	AR; 16q22, AKA tyrosinemia type II; tyrosine aminotransferase deficiency, MR, palmoplantar keratoderma, severe keratitis
Richter's s.	development of large cell lymphoma in patient with chronic lymphocytic lymphoma
Riley-Day s.	AR; defective lacrimation, skin blotching, emotional instability, motor incoordination, absence of pain sensation leading to burns & bitten tongue, hyporeflexia, erythema of face & trunk, cyanosis of extremities, corneal anesthesia, hyperhidrosis, hypertension AKA familial dysautonomia
Riley-Smith s.	AD; multiple lymphatic venous malformations & pseudo-papillomas, macrocephaly
Ritter's d.	*S. aureus* infection elaborating exfoliatin, leading to denuded skin AKA staphylococcal scalded skin s.
Riyadh chromosome breakage s.	depigmentation, MR, silver hair
Roberts phocomelia s.	upper limb reduction malformation, flexion contractures of knees, silver hair, IUGR, MR
Romana's sign	unilateral bipalpebral edema; Chagas' d., oculoglandular s.
Rombo s.	AD; atrophoderma vermiculatum, BCC, hypotrichosis, milia, peripheral vasodilation with cyanosis
Rosai-Dorfman s.	sinus histiocytosis with massive LAD
Rosenthal-Kloepfer s.	corneal leukomata, acromegaloid appearance, cutis verticis gyrata
Ross s.	progressive segmental anhidrosis with tonic pupils, absent DTRs
Roth's spots	conjunctival petechiae; subacute bacterial endocarditis
Rothmann-Makai s.	lipogranulomatosis subcutanea; idiopathic lobular panniculitis with fat cell necrosis, lipophagic granuloma, cysts
Rothmund-Thomson s.	AR, 8; mostly females (some have C1q deficiency); atrophic hyperpigmented reticulated telangiectatic cutaneous plaques, alopecia, bone defects, cancer, dental dysplasia,

	hypogonadism, hypoparathyroidism, nail dystrophy, photosensitivity AKA poikiloderma congenitale
Rothschild's sign	loss of hair from lateral third of eyebrows; hypothyroidism (non-specific)
Rowell's s.	erythema multiforme-like lesions occurring in patients with SLE & La/SS-B autoantibodies
Rozychi's s.	achalasia, congenital deafness, leukoderma, muscle wasting
rubber man s.	Ehlers-Danlos s.
Rubinstein-Taybi s., Rubinstein's s.	mental & motor retardation, broad thumbs & great toe, keloid formation, short stature, characteristic facies, high palate, beaked nose, large foramen magnum, vertebral abnormalities
Rud's s.	AR (?); associated with recessive X-linked ichthyosis, hypogonadism, MR, obesity, retinitis pigmentosa [may be the same disease as X-linked recessive ichthyosis]
Ruffini's corpuscle	rare, subcutaneous acral skin receptor; unknown function
runting s.	GVH with diarrhea, dermatitis, HSM, hemolytic anemia, pancytopenia
Russell's sign	callous (or calluses) on the dorsum of dominant hand; purging eating disorder
Russell-Silver s., Russell's s.	café-au-lait spots, incurved fifth fingers, lateral asymmetry, low birth weight, precocious puberty, short stature, syndactyly, triangular-shaped face, turned down corners of mouth,
Ruvalcaba-Myhre-Smith s.	bony & craniofacial abnormalities, genital macules, intestinal polyposis
Sabinas s.	AR; brittle hair, MR, nail dystrophy, ocular dysplasia, xerosis
Sanfilippo's s.	mucopolysaccharidosis, excretion of heparan sulfate in urine; generalized hirsutism, hepatomegaly, macrocephaly, MR, death before age 20
SAPHO s.	*s*ynovitis, *a*cne, *p*ustulosis, *h*yperostosis, *o*steitis
Satoyoshi's s.	early onset alopecia areata, malabsorption, painful muscle spasms, short stature, skeletal defects
scalded skin s.	AKA dermatitis exfoliativa neonatorum, Ritter's d.
SCARF s.	XLR (?); **s**keletal abnormalities, **c**raniostenosis, **c**utis laxa, **a**mbiguous genitalia, **r**etardation, **f**acial anomalies
scarlatinella	Dukes' d.
Sch←–fer's s., Sch←–fer-Branauer s.	pachyonychia congenita with physical & mental retardation
Schamberg's d.	progressive pigmented purpuric dermatosis; tan macules with minute petechiae on lower extremities
Schimmelpenning s.	sporadic, AD; AKA epidermal nevus s.
Schimmelpenning-Feverstein-Mims s.	systemic nevi in Blaschko's lines, mild MR, skeletal abnormalities
Schinzel-Giedion s.	AR: narrow, deep set triangular nails, telangiectasias of nose & cheeks, dermatoglyphic changes, simian crease, hypertrichosis
Schnitzler's s.	monoclonal IgM, arthralgia, bone pain, fever, lymphadenopathy, hepatomegaly, hyperostosis, nonpruritic urticarial vasculitis
Schnyder's s.	progressive partial symmetrical erythrokeratoderma with deafness
Schönlein-Henoch s.	nonthrombocytopenic purpura due to vasculitis in children; arthropathy, arthritis, erythema, GI symptoms, renal disease, urticaria AKA Henoch-Schönlein purpura
Schopf-Schulz-Passarge s.	AR, diffuse symmetric PPK; fragile nails, sparse hair, eyelid cysts, hypodontia, hypotrichosis, longitudinal & oblique nail furrows AKA PED type XIX
Schultz-Charlton phenomenon	(historic test) intradermal injection of 0.1 mL of antitoxin into area of scarlet fever rash producing blanching at site of injection; scarlet fever
Schwachman's s.	AR; exocrine pancreatic insufficiency, growth retardation, impaired neutrophil chemotaxis, neutropenia, recurrent infections, skeletal defects, with ichthyosiform or eczematous change
Schweninger-Buzzi anetoderma	idiopathic anetoderma without preceding lesions
Seckel's s.	AR; hypodontia, pancytopenia, simian crease, skeletal defects, trident hands AKA bird-headed dwarfism

Secretans s.	traumatic edema of dorsal hand (factitial)
Seeligmuller's sign	mydriasis of side of face affected by neuralgia
Seidlmeyer's s.	acute hemorrhagic edema of infancy
Seip's s.	accelerated osseous maturation, lipodystrophy, muscular hypertrophy
Senear-Usher s.	pemphigus erythematosus
Senter s.	KID s.
serum sickness s.	develops 8-12 days after administration of serum proteins; albuminuria, arthralgia, fever, hypocomplementemia, LAD, leukopenia, nephritis, splenomegaly, urticaria
severe combined immunodeficiency s. (SCIDs)	XLR, sporadic, AR; decreased humoral & cell-mediated immunity, absence of delayed hypersensitivity, lack of immunoglobulins, lymphocytopenia, GVH in utero, eczema, recurrent infections, recurrent Candidiasis, diarrhea, FTT, death by age 2
Sezary s.	CTCL with generalized exfoliative erythroderma, intense pruritus, LAD, Sezary cells (skin, lymph nodes, blood)
Shab-Waardernburg s.	Waardenburg's s.
Shapira's s.	ataxia, developmental delay, hair defects (sparse, brittle, light color), short stature
Shprintzen's s.	marfanoid features with craniosynostosis AKA Shprintzen-Goldberg s.
Shulman's s.	eosinophilic fasciitis
sideropenic dysphagia	Plummer-Vinson s.
Siegert's sign	short, medially curved 5th fingers; Down syndrome
Siemens, ichthyosis bullosa of	AD, keratin 2e mutations; similar to EHK
Siemerling-Creutzfeldt d.	adrenal atrophy, early death, hyperpigmentation, leukodystrophy
Silex's sign	furrows radiating from mouth; congenital syphilis
Silver-Russell s.	Russell-Silver s.
Silvestrini-Corda s.	elevated levels of circulating estrogens from liver failure; atrichia, eunuchoid body, gynecomastia, hypogonadism, sterility
Sipple's s.	AD, sporadic; mucosal neuromas, medullary carcinoma of thyroid, pheochromocytoma, marfanoid body habitus AKA multiple mucosal neuroma s., multiple endocrine neoplasia type IIa
sister chromatid exchanges	Bloom's s., Cockayne s., dyskeratosis congenita, Fanconi's anemia
Sister Marie Joseph sign	umbilical metastasis; colon, **gastric**, ovarian
sixth d.	exanthema subitum (HHV 6)
Sjögren's s.	keratoconjunctivitis sicca, xerostomia with connective tissue disease; polymyositis, RA, scleroderma, SLE
Sjögren-Larsson s.	AR; congenital oligophrenia, ichthyosis, spastic pyramidal symptoms
Sneddon's s.	livedo vasculitis with cerebral infarction; aphasia, hemiplegia, &/or hemianopsia
Sneddon-Wilkinson d.	subcorneal pustular dermatosis
Sobye's massage	BID facial massage; rosacea
Solomon s.	epidermal nevus s.
Soret band	390-410 nm radiation band; absorbed by porphyrins
Sotos' s.	abnormal facies, genital lentigines, macrocephaly, skeletal defects
Spiegler-Fendt sarcoid	pseudolymphoma
Spitz's nevi	dysplastic nevi
Splendore-Hoeppli phenomenon	refractile amorphous eosinophilic matrix; immunoglobulin to *S. aureus*, *P. aeruginosa,* coagulase-negative staphylococci, streptococcal species, *Escherichia coli*, *Proteus* species
staphylococcal scalded skin s.	Ritter's d.
steatocystoma multiplex	AD; presents at puberty, numerous cysts over sternum, chest, axillae, proximal arms
Steijlen's s.	atrichia, MR, palmoplantar keratoderma, tooth loss
Stein-Leventhal s.	anovulation, hirsutism, oligomenorrhea, polycystic ovaries AKA polycystic ovary s.
Stevens-Johnson s.	erythema multiforme minor with mucocutaneous & systemic lesions AKA erythema multiforme major

Stewart-Treves s.	lymphangiosarcoma following lymphedema/lymphadenectomy, usually after radical mastectomy
stiff hand s.	fibrosis of hand leading to stiffness; diabetes
Still's d.	juvenile RA
Sturge-Weber s.	sporadic; usually unilateral nevus flammeus over trigeminal nerve, progressive tissue & bone hypertrophy beneath nevus, vascular malformation in leptomeninges, hemiparesis, MR, seizures
subcorneal pustular dermatosis	Sneddon-Wilkinson d.
Sucquet-Hoyer canal	contains glomus cells giving rise to glomus tumors
Sulzberger-Garbe s.	exudative discoid & lichenoid dermatitis
Sutton's nevus	halo nevus
sweat retention s.	occlusion of sweat ducts; pruritus, dermatitis, miliaria
Sweet's s.	acute febrile neutrophilic dermatosis
Takayasu's d.	vasculitis of aortic arch & its major branches
Tangier d.	AR, lipoprotein & lipid metabolism disorder; absence of HDL, deficient apolipoproteins A-I & A-II, low to normal LDL, high triglycerides, accumulation of cholesteryl esters, enlarged orange tonsils, pharyngeal mucosa, rectal mucosa, recurrent peripheral neuropathy, splenomegaly, corneal infiltration
Tay's d.	(P)IBIDS = trichothiodystrophy
Tay's s.	bone defects, café-au-lait spots, hypersplenism, lentigines, MR, physical retardation, vitiligo
Texier's d.	allergic reaction to Vitamin K injection
Tietze's s.	AD; albinism, with normal eye pigment, deaf-mutism, eyebrow hypoplasia
TORCH s.	*t*oxoplasmosis, *o*ther (syphilis, bacterial sepsis), *r*ubella, *C*MV, *h*erpes; chorioretinitis, deafness, HSM, jaundice, microcephaly, purpura, thrombocytopenia
Torre's s.	Muir-Torre s.
Touraine-Solente-Gol— s.	AD; cutis verticis gyrata, short stature, thin yellow nails AKA pachydermoperiostosis
toxic epidermal necrolysis	Lyell's s.
toxic shock s.	desquamation 1-2 weeks after onset, disorientation, fever, GI upset, hepatic changes, hypotension, mucous membrane hyperemia, myalgia, rash, renal abnormalities, thrombocytopenia
transient acantholytic dermatosis	Grover's d.
trench mouth	Vincent's infection
trichomegaly	excessive eyelash &eyebrow hair growth associated with dwarfism, MR, retinal pigment degeneration
trichothiodystrophy	Tay's d., (P)IBIDS
triparanol s.	use of triparanol causing alopecia, poliosis, ichthyosis, irreversible cataracts, impotence
Trousseau's s.	hypercoagulable state secondary to malignancy of pancreas, stomach, lung, prostate, colon, ovaries, gallbladder leading to thrombophlebitis
tuberous sclerosis	Bourneville's s., Bourneville-Pringle s.
Turcot's s.	familial adenomatous polyps, CNS tumors
Turner s.	XO or 45X; disorder of gonadal differentiation, short stature undifferentiated gonads, neck webbing, low posterior hair line, cardiac defects, sterility
Turner's sign	blood causing blue discolored skin at costovertebral angle; acute hemorrhagic pancreatitis
twenty nail s.	trachyonychia
Tyndall effect	blue appearance of melanin in dermal lesions due to selective light absorption
Tyndall effect/phenomenon	light absorption in deep dermis lending blue color to lesions
Tyson's glands	sebaceous glands of prepuce
Ullrich-Turner s.	45X, female phenotype; lymphedema, melanocytic nevi, mild MR, short stature, sexual

	infantilism, webbed neck
uncombable hair s.	AD; pili torti & canaliculi; blond dry thick shiny hair
unilateral nevoid telangiectasia	syndrome of generalized essential telangiectasia of vascular nevus under influence of estrogen
Unna's nevi	exophytic nevi with silhouette of fibroepithelial polyps
Unna-Thost s.	AD; diffuse palmoplantar keratoderma
Unverricht's d.	Baltic myoclonic epilepsy
Urbach-Wiethe d.	AR; infiltrative hyaline deposits in skin, mucous membranes & internal organs AKA lipoid proteinosis, hyalinosis cutis et mucosae
vagabond's d., vagrant's d.	skin discoloration due to chronic *Pediculus humanus corporis* bites AKA parasitic melanoderma
Van Lohuizen's s.	cutis marmorata telangiectasia congenita
Venus' necklace	hypopigmented macules on liner pigmented reticulae patches; secondary syphilis AKA leukoderma colli syphiliticum
Verbov-Sharland s.	palmoplantar keratoderma with neurosensory deafness
Vincent's infection	acute necrotizing ulcerative gingivitis AKA trench mouth
Vinson's s.	Plummer-Vinson s.
Vogt triad	epilepsy, MR, skin abnormalities (adenoma sebaceum); <1/3 of patients have full triad
Vogt-Koyanagi-Harada s., Vogt-Koyanagi s.	alopecia, bilateral uveitis, choroiditis, deafness, headache, meningism, poliosis, retinal detachment, vision loss, vitiligo, vomiting, sometimes glaucoma or vertigo
Vohwinkel's s.	AD; palmoplantar keratoderma with digital pseudoainhum, scarring alopecia & high frequency hearing loss AKA keratoma hereditaris mutilans, mutilating keratoderma
Voigt's lines	pigment demarcation lines of dorsolateral arms; blacks & Asians AKA Futcher's lines
von Hippel-Lindau d.	AD, 3p25-26; hereditary phakomatosis; angiomatous lesions (kidneys, liver, pancreas), capillary malformation (rare), café au lait macules, pheochromocytoma, renal cell cancer, vascular malformations in cerebellum & brain stem, retinal hemangioblastoma
von Recklinghausen's d.	AD 17q11.2; neurofibromatosis type I
Vorner's s.	clinical appearance of Unna-Thost s. with epidermolytic hyperkeratosis on biopsy
Waardenburg's s.	AD, 2q35; white forelock, neurosensory deafness, wide nasal bridge, heterochromia of iris
Wachters PPK	Brunauer-Fuhs-Siemens PPK
Wagner-Unverricht d.	dermatomyositis
Watson's s.	deletion of NF1 gene; variant of neurofibromatosis 1, multiple café-au-lait macules, neurofibromas (few), MR, pulmonary valvular stenosis, short stature
Weber-Christian s.	idiopathic lobular panniculitis; relapsing febrile nodular nonsuppurative panniculitis-arthralgias, fever, malaise
Weber-Cockayne s.	localized epidermolysis bullosa simplex
Wegener's granulomatosis	+ C-ANCA, facial and periauricular lesions; malignant pyoderma
Weil's d.	*Leptospira interrogans serovar icterohemorrhagiae*; hepatorenal failure-jaundice, oliguria, purpura
Well's s.	eosinophilic cellulitis
Werner's s.	AR, 8p12; premature aging with baldness, cataracts, muscular atrophy, osteoarthritis, scleroderma, subcutaneous calcification, telangiectasias, diabetes mellitus tendency, neoplasms, short stature
Westerhof's s.	AD; café-au-lait macules, growth retardation, hyper & hypopigmented macules, MR
Whipple's d.	abdominal pain, anorexia, arthritis, CNS disturbance, diarrhea, fever, LAD, skin pigment changes, steatorrhea AKA intestinal lipodystrophy
Wickham's striae	reticulate white lines on top of papules & buccal mucosa; lichen planus
Williams s.	AD; dysmorphic facies, supravalvular aortic stenosis, velvety skin
Wilson's d.	AR, q13; copper metabolism defect of ceruloplasmin, copper accumulates in liver, brain, kidney, cornea, azure lunulae, hyperpigmented legs, Kayser-Fleischer ring AKA hepatolenticular degeneration

Wimberger sign	osteochondritis of medial proximal tibial metaphysis; congenital syphilis AKA cat bite sign
Winterbottom's sign	enlarged posterior cervical lymph nodes; trypanosomiasis (Gambian)
Wiskott-Aldrich s.	XLR, immunodeficiency; autoimmune phenomena, cyclic neutropenia, decreased chemotaxis, eczema, increased malignancy, recurrent pyogenic infections, thrombocytopenia, decreased IgA & IgE, normal/increased/decreased IgM, platelet dysfunction
Woolf's s.	piebaldism with deafness
Woringer-Kolopp d.	localized pagetoid reticulosis
Woronoff's ring	white blanching skin ring (leukoderma); psoriasis
Wyburn-Mason's s.	sporadic; facial nevus flammeus, ataxia, AV malformation of optic nerve & retina, enlarged facial veins, MR, nystagmus, seizures
xeroderma pigmentosa	AR; deficient enzyme in excisional repair of UV-damaged DNA; extreme UV photosensitivity; keratoses, malignancies (BCC, SCC), papillomas, telangiectasia
xerodermic idiocy	De Sanctis-Cacchione s.
X-linked hypogammaglobulinemia	decreased IgM, IgG, IgA, C1q, cutaneous & systemic pyogenic infections, chronic echovirus infection with dermatomyositis-like finding, eczema, URI, osteomyelitis, pneumonia, joint infections, large joint arthritis, no B cells
yellow nail s.	lymphedema with smooth, thickened, curved, yellow discolored nails
Zinsser-Cole-Engman s.	XLR/AD; reticular pigmentation progressing to atrophy & telangiectasia, bullous conjunctivitis, esophageal strictures, leukoplakia (mucous membranes), mental deficiency, nail dystrophy, palmoplantar hyperkeratosis & hyperhidrosis, pancytopenia, skeletal disorders, thrombocytopenia AKA dyskeratosis congenita
Ziprowski-Margolis s.	XLR, males; Xg26.3-q27.1; deaf-mutism, heterochromic irides, piebald-like hypomelanosis of skin & hair
Zoon's balanitis (vulvitis)	variant of lichen planus (?) of genitalia AKA plasma cell balanitis (vulvitis)
Zunich-Kaye s.	CHIME s.

Classifications of syndromes

Autosomal dominant	acrokeratosis verruciformis of Hopf, Adams-Oliver s., albinism, albinism and deafness, anonychia ectrodactyly, Bannayan's s., Bart's s., basal cell nevus s., Basan's s., Beckwith-Wiedemann s., blue rubber bleb nevus s., bullous ichthyosiform erythroderma, Buschke-Ollendorff s., Clouston's s., cold hypersensitivity, congenital scalp defect, Cowden's s., cutis laxa, Darier's d., distichiasis and lymphedema, dyskeratosis congenita, EB-Cockayne, EB-dystrophica, EB-simplex, EEC s., Ehlers-Danlos s., epidermolysis bullosa simplex, epidermolysis bullosa dystrophica, epitheliomas, erythrokeratoderma viriabilis, familial angioedema, familial dyskeratotic comedones, familial localized heat urticaria, familial Mediterranean fever, familial pachydermoperiostosis, Gardner's s., glomus tumors, Hailey-Hailey d., hereditary hemorrhagic telangiectasia, hereditary koilonychia, hereditary sclerosing poikiloderma, hidrotic ectodermal dysplasia, Howel-Evans' s., Huriez s., hypertrichosis universalis, hypomelanosis of Ito, ichthyosis hystrix gravior, ichthyosis vulgaris, incontinentia pigmenti achromians, Jadassohn-Lewandowsky s., keratoderma palmaris et plantaris, keratoderma with esophageal cancer, LEOPARD s., leukonychia totalis, lipoatrophic diabetes, lymphedema and distichiasis, lymphedema-hereditary, Maffucci's s., Marfan's s., Marie-Unna hair dystrophy, melanoma, Melkersson's s., milia and decreased hair density, monilethrix, Muir-Torre s., multiple benign ring-shaped skin creases, multiple cylindromas, multiple leiomyomata, multiple lipomatosis, Naegeli-Franceschetti-Jadassohn s., Naegeli's s., nail-patella s., NAME s., neurofibromatosis, Osler-Weber-Rendu s., pachydermoperiostosis, pachyonychia congenita, Peutz-Jeghers s., piebaldism, pili annulati, porphyria cutanea tarda, porphyria-acute intermittent, porphyria-variegate, porphyria- erythropoietic protoporphyria, porphyria-hereditary coproporphyria, Rapp-Hodgkin s., Riley-Smith s., sclerotylosis, uncombable hair s., steatocystoma multiplex, trichorhinophalangeal s., tuberous sclerosis, urticaria-deafness-amyloidosis, Vohwinkel's s., von Hippel-Lindau d., von Recklinghausen's d., Waardenburg's s., Westerhof s., woolly hair

autosomal recessive	acrodermatitis enteropathica, albinism, alkaptonuria, arginosuccinic aciduria, aspartylglycoaminuria, ataxia-telangiectasia, biotinidase deficiency, Bj¬rnstad's s., Bloom's s., cartilage-hair hypoplasia, cerebrotendinous xanthomatosis, Chanarin-Dorfman s., Chediak-Higashi s., circumscribed keratoderma, Cockayne's s., Conradi's d., Cornelia de Lange s., Cross-McKusick-Breen s., cutis laxa, De Sanctis-Cacchione s., Desmons-Britten s., EB-dystrophica, EB-letalis, EB-junctional, Ehlers-Danlos s., epidermodysplasia verruciformis, erythropoietic porphyria, familial dysautonomia, Fanconi's s., Farber's lipogranulomatosis, fucosidosis type II, Gaucher's d., Hallermann-Streiff s., harlequin fetus, Hartnup's d., hemochromatosis, Hermansky-Pudlak s., homocystinuria, Hunter's s., Hurler s., hypohidrotic ectodermal dysplasia, ichthyosiform erythroderma (non-bullous), ichthyosis-lamellar, IgA deficiency s., juvenile fibromatosis, KID s., keratitis-ichthyosis-deafness s., keratoderma palmaris et plantaris, lamellar ichthyosis, Lawrence-Seip s., Letterer-Siwe d., leukocyte alkaline phosphatase deficiency, leukocyte myeloperoxidase deficiency, lipoid proteinosis, Mal de Meleda, Marinesco-Sj¬gren s., Morquio's s., mucopolysaccharidoses, multiple sulfatase deficiency, Netherton's s., Neu-Laxova s., Nezelof s., Niemann-Pick d., Omenn's s., Papillon-Lefℵvre s., peeling skin s., phenylketonuria, porphyria-erythropoietic, porphyria-hepatoerythropoietic, progeria, prolidase deficiency, pseudoxanthoma elasticum, Refsum's d., Richner-Hanhart s., Rothmund-Thomson s., Rud's s., Sabinas s., Schwachman's s., Seip-Lawrence s., sialidosis-juvenile type II, Sjögren-Larson s., Swiss type aggamaglobulinemia, Tangier d., trichorrhexis invaginata, trichothiodystrophy, tyrosinemia II, Urbach-Wiethe d., vitamin D-resistant rickets (type II0 with alopecia, Werner's s., Wilson's d., Wilson's d., xeroderma pigmentosum
chemotactic abnormalities	**neutrophils**; lazy leukocyte s., Chediak-Higashi s., hyperimmunoglobulin E s., Shwachman s. **leukocytes**; Wiskott-Aldrich s.
chromosomal fragility	ataxia-telangiectasia, Bloom's s., Cockayne s., Fanconi's anemia
Lyonization (functional mosaicism)	Conradi s., incontinentia pigmenti
non-Mendelian	Delleman-Oorthuys s., Klippel-Trenaunay-Weber s., McCune-Albright s., Neurocutaneous melanosis, proteus s., Schimmelpenning s., Sturge-Weber s.
X-linked dominant	Albright's d., atrichia with keratin cysts, Bloch-Sulzberger s., chondrodystrophia congenita punctata (Conradi-Hunermann type), congenital hemidysplasia with ichthyosis and limb defects (CHILD s.), Conradi-H∫nermann s., craniofrontal dysplasia, focal dermal hypoplasia (Goltz s.), incontinentia pigmenti, keratosis follicularis spinulosa de Calvans, orofacialdigital s., streblocactly, type II vitamin D resistance
X-linked recessive	anhidrotic ectodermal dysplasia (Christ-Siemens-Touraine s.), Bruton's X-linked aggamaglobulinemia, chronic granulomatous d., Crandall's s., cutis verticis gyrata with thyroid aplasia, dyskeratosis congenita (Zinsser-Cole-Engman s.), Ehlers-Danlos s. type V & IX, Fabry's d., keloids, keratosis pilaris decalvans, Mendes de Costa s., Menkes Kinky hair s., Hunter's s., ichthyosis follicularis, keratosis follicularis spinulosa decalvans, Lesch-Nyhan s., occipital horn s., properdin dysfunction, renal dysplasia, severe combined immunodeficiency s., rhabdomyomatous mesenchymal hamartoma, SCIDS, torticollis, Wiskott-Aldrich s., X-linked ichthyosis, Ziprowski-Margolis s.