Directions in Residency

Fall 2012

Official Publication of the Residents/Fellows Committee, American Academy of Dermatology

Boards' Fodder celebrates 10 years!

By Dean Monti, AAD managing editor, special publications

The idea was pitched 10 years ago in a Resident/Fellows Committee Meeting in February 2002, during the Academy's Annual Meeting in New Orleans. It was suggested that the Academy find new ways to alert dermatology residents to areas that were "high yield" for the mock and real board examinations. The answer was to provide charts of commonly asked and "highly askable" factoids relating to dermatology. Boards' Fodder had its premiere in AAD's *Resident Roundup* (now *Directions in Residency*) in fall 2002 with "Genes to Know," by Benjamin Solky, MD, and Brian Selkin, MD. Since that time, Boards' Fodder has proven to be one of the most popular features of this publication and a valuable study tool for residents for the past decade — indeed, the publication's longest running feature.

Considering that our audience changes every four years, it is notable that new residents have continued to carry the torch forward. Dr. Solky and his colleagues contributed to the feature through its nascent years, followed by Sharon Jacobs, MD, for several years. Then, somewhere along the line, word got around: Boards' Fodder was an opportunity for all up-and-coming residents to share information with other residents and showcase their individual resident talent in an Academy publication. As Boards' Fodder enters a new decade, contributions to the column are no longer the voice of one resident, but many, and contributions are on the rise. To celebrate the occasion, we are presenting three times the Fodder for this special issue.

Thank you all for your contributions over the years, which are helping us build an archive of study materials for current and future residents. As we forge ahead, *Directions in Residency* is working toward ways to bring this feature to you in new formats and, with your help, more often. DR


DERMATOLOGY

Resident ROUNDUP

AMA supports ACGME report calling for shorter resident work hours

Membership — don't leave your residency without it

additional resources

More resources than ever to help with boards study

If there's one thing we've discovered over the years, it's that residents can't get enough boards study materials. Recently the Resident/Fellows page at aad.org was updated and includes a new section, "Board prep," containing boards study resources.

The AAD offers a variety of resources to help you prepare.

- View the Boards' Fodder section in current and past issues of *Directions in Residency*, a quarterly publication for residents.
- Listen to *Dialogues in Dermatology*, the Academy's monthly audio subscription program in which leading dermatologists discuss current clinical issues.
- Read *Derm Clips*, a monthly publication that includes evidence-based summaries of clinical content from a broad range of medical journals.
- Watch Boards Blitz for two hours of board prep insight, tips, and resources that you can view while studying.
- Review the AAD's online Medical Student Core Curriculum to cover the basics of dermatology when preparing for the boards.
- Explore peer-recommended boards study tools that include books, study guides, websites, and quizzes.
- Board prep courses offered at AAD meetings.

You can also learn more about boards prep on the American Board of Dermatology site, www.abderm.org.

Directions in Residency is also looking for new ways to bring board materials to you. If you have not already done so, please take the *Directions in Residency* reader survey (see page 8). DR

SPECIAL BOARDS' FODDER EDITION!

Inside *this* issue

Oral Disease, Part 2

by Helena Pasieka, MD

	CLINICAL	PATHOLOGY	TREATMENT	ASSOCIATIONS
SALIVARY GLAND DISEASES				
Mucocele	Painless submucosal swelling. Color ranges from clear/blue/colorless depending on depth of lesion. Lower labial mucosa most common, but can occur anywhere where there are minor salivary glands.	Collection of mucus surrounded by macrophages and granulation tissue. Look for the inflamed minor salivary gland as a clue.	Superficial mucoceles often resolve w/o intervention by spontaneous rupture. However, most require surgical excision.	
Cheilitis glandularis	Ranges from slight hypertrophy of lower lip to nodular enlargement with eversion. Most commonly in adult men.	Localized dense accumulations of inflammatory cells within and around the mucous glands in a background of actinic chelitis.	Vermilionectomy of the lower lip, with or without cosmetic debulking is the standard of care. Injections of corticosteroids can provide symptomatic relief..	
Sjögren's syndrome	Slow onset of variable degrees of eye and oral dryness. Increased dental carries and difficulty wearing dentures. Increased candidiasis.	Salivary glands with focal aggregates of >50 lymphocytes adjacent to normal-appearing acini. In the parotid gland characteristic epimyoepithelial islands are seen.	Symptomatic care and management of the associated complications. Salivary stimulation (sugarless gum, hard candies, pilocarpine) or artificial saliva.	
Salivary gland tumors	Submucosal, painless, rubbery firm swelling often noted on the posterior hard palate or anterior soft palate.	Varies depending on the specific type of salivary tumor.	Benign salivary gland tumors are conservatively excised. Malignant tumors are more extensively removed +/- radiotherapy as adjunct.	
HEMATOLOGIC/ONCOLOGIC DISEASE				
Chemotherapy-induced mucositis	Multiple oval or irregularly shaped ulcers, usually on the gingivae, lateral tongue, or buccal mucosa. Usually appear 4-7 days after chemotherapy administered.	Lichenoid mucositis.	Usually resolves in 2-3 weeks cessation of chemotherapy. Palifermin (keratinocyte growth factor) may reduce severity for those on high-dose chemotherapy. Meticulous oral hygiene and symptom management.	
Leukemia	Many oral manifestations, most commonly bruising or hemorrhage related to thrombocytopenia. Also, pallor of anemia, increased viral, fungal and bacterial infections related to leukopenia. Diffuse, firm, non-tender gingival enlargement can be caused by infiltration of the gingival connective tissue by leukemic cells.	Infiltration of leukemic cells into gingival connective tissue (most commonly with monocytic or myelomonocytic leukemias).	Multi-agent chemotherapy and peripheral blood stem cell or bone marrow transplantation are most commonly used to treat acute leukemia.	
Lymphoma	Slow growing, painless, soft or rubbery, with purplish swelling. Most commonly on the palate and the buccal vestibule. Overlying telangiectasia sometimes seen. Ulceration possible, mimicking SCC.	Infiltration of atypical lymphocytes, usually of B-cell type.	Chemotherapy and/or monoclonal antibodies (e.g. rituximab).	HIV, other ilmmunosuppression, older age.
Melanoma	Most commonly on hard palate, maxillary attached gingiva. Has same features as cutaneous melanoma.	Proliferation in epithelium and infiltration of the connective tissue by atypical melanocytes, with or without melanin production. Can use Melan-A or S100 to stain.	Wide surgical excision with (-) margins. Sentinel lymph node biopsy for prognostication. Minimal radial growth phase on mucosa, so they differ from cutaneous melanomas in that they present in the vertical growth phase. Chemotherapy and XRT of little utility. Worse prognosis than cutaneous lesions, w/5year survival rate of ~15%, and median from dx of < 2 years.	

Helena Pasieka, MD, is a second year resident in the department of dermatology at Johns Hopkins University.

Oral Disease, Part 2 (continued)

by Helena Pasieka, MD

	CLINICAL	PATHOLOGY	TREATMENT	ASSOCIATIONS
MANIFESTATIONS OF SYSTEMIC DISEASE				
Amyloidosis	Firm macroglossia, often with scalloped edge. Xerostomia or dysgeusia can be seen before the onset of tongue enlargement.	Homogeneous eosinophilic accumulation. (+) congo red stain.		
Pernicious anemia	Gradual onset of smooth, 'beefy-red' tongue, w/ill-defined areas of erythema which can coalesce into diffuse dorsal tongue involvement causing a smooth, beefy-red appearance.	Absence of filiform papillae.	Vitamin B12 injections provide rapid improvement.	
Crohn's disease	Linear fissures +/- ulcers of the vestibule or "cobblestone" ulcers of the buccal mucosa. Sometimes scarring. Can also have cheilitis granulomatosa both clinically and histologically.	Non-necrotizing granulomatous inflammation.	Respond to therapy for bowel lesions. Topical or intralesional corticosteroids also effective.	
Pyostomatitis vegetans	"Snail track" arrangement of multiple tiny, creamy-yellow pustules set against a bright erythematous background. Fragile pustules lead to shallow erosions and ulcerations. Labial, gingival and buccal mucosa are most commonly involved; tongue is usually spared.	Intra- or subepithelial microabscesses containing eosinophils and neutrophils.	Management of the underlying GI disease often results in improvement of oral lesions.	Inflammatory bowel disease.
HIV				
Kaposi's sarcoma	Multiple violaceous macules, plaques, or nodules. Most commonly on palate, but can be anywhere in mouth.	Infiltration of the dermis w/slit-like vascular spaces, and dilated vessels. Many extravasated RBCs and proliferating spindle cells seen.	May improve or resolve with improved immune status (i.e., initiation of HAART).	
Oral hairy leukoplakia	White, shaggy, corrugated protrusions on lateral tongue. Cannot be dislodged with tongue depressor.	Irregular keratin projections, parakeratosis, acanthosis, and groups of pale epithelial cells.	Antiretroviral therapy may lead to regression.	Epstein-Barr virus in HIV. Predictor of rapid decline and progression to AIDS.
Candidiasis	Thick white or cream-colored deposits on tongue or posterior oropharynx with "cottage cheese-like" appearance. Can be dislodged with tongue depressor. Also can have fissuring at the corners of the mouth.	Pseudohyphae and budding yeast sometimes seen in H&E. More easily seen with PAS or GMS stain.	Improvement of immune status. Anti-candidal treatment, such as clotrimazole troches or PO fluconazole.	
HSV	Grouped vesicles on erythematous base, becoming ulcerated. Can coalesce into larger lesions. Look for scalloped border.	Epidermal necrosis and ballooning degeneration. Infected cells are multinucleated w/glassy nuclear contents, marginalized nuclear chromatin, and nuclear molding.	Suppressive therapy with antiviral medications. Severely immunosuppressed may need IV acyclovir.	
CMV	Ulcerations anywhere in mouth. Appear like aphthae, may be slightly larger.	Vascular dilation with large cytomegalic endothelial cells. "Owls eye" appearance due to halo around intranuclear inclusion bodies.	Antiviral drugs (ganciclovir and valganciclovir).	Development on mucosal surface usually a sign of disseminated disease.
SYNDROME ASSOCIATIONS				
Odontogenic keratocysts of the jaw	Often incidentally noted radiolucent or mixed radiolucent/radiopaque lesions of the mandible. Asymptomatic; rarely mild facial swelling and discomfort.	Lining of stratified squamous epithelium with basal layer of the epithelium exhibiting palisaded cuboidal to columnar cells. The luminal surface often with a corrugated morphology and parakeratosis.	Opinions differ and range from wide local excision to marsupialization to curettage.	Nevoid basal cell carcinoma syndrome w/mutations in *PTCH (*Hedgehog signaling pathway).
Multiple osteomas of the jaw	Asymptomatic facial deformity.	Same as typical solitary osteomas.	Surgical removal, screening for malignancy.	Often the earliest marker of Garner syndrome occurring >80% around puberty.
Multiple endocrine neoplasia syndrome type 2B	Multiple mucosal neuromas involving lips and tongue are often the first dermatologic manifestation.	"Plexiform neuromas" of hyperplastic bundles of nerves surrounded by a thickened perineurium.	Screening for malignancy: Medullary thyroid carcinoma with pheochromocytoma in 50% of cases, and digestive neurofibromatosis.	

Sources:
1. Bolognia JL, Jorizzo JL, Rapini RP eds. Dermatology. 2nd ed. Mosby; 2007.
2. Rapini R. Practical Dermatopathology. Mosby; 2005.

Special thanks to Dr. Gary Warnock.

Inborn Errors of Metabolism

by Kristina Burke, MD, and Erin Adams, MD

DISORDER	DEFECT	SKIN FINDINGS	OTHER
Alkaptonuria (Endogenous ochronosis) **AR**	Homogentistic acid oxidase Disorder of phenylalanine and tyrosine metabolism. Homogentistic acid accumulates	Blue-grey pigmentation of nose, ears, axillae, genitalia and cartilage. Blue sclera (Osler's sign), dark urine (pH >7), black cerumen	- Large joint arthropathy - Intervertebral disc calcification - Mitral/aortic valve disease - Nephrolithiasis
Fabry's disease (Angiokeratoma Corporis Diffusum) **XLR** A lysosomal storage disease	α-galactosidase A (glycolipids accumulate in skin, heart, kidneys)	Angiokeratomas (esp lower extremities, scrotum, penis, and lower trunk), whorl-like corneal opacities, edema, hypohldrosis	- Renal failure, cardiovascular events, strokes - Acroparesthesias and painful crises - Maltese cross in urine - Enzyme replacement available
Fucosidosis **AR** A lysosomal storage disease	α-L- fucosidase	Angiokeratomas, coarse features, facial dysmorphism	- Mental retardation (MR), neurologic deterioration
Gaucher disease **AR** A lysosomal storage disease	Acid-β-glucosidase (glucocerebrosidase) Leads to accumulation of glucocerebroside in histiocytes (Gaucher's cells)	Type 1: diffuse hyperpigmentation, petechiae, pingueculae of sclera Type 2: congenital ichthyosis, collodian baby	ALL: hepatosplenomegaly (HSM) - Type I: Adult type, Ashkenazi Jews, no CNS involvement - Type 2: infantile, rapid neuro deterioration, aspiration pneumonia - Type 3: juvenile chronic neuropathic
Phenylketonuria **AR**	Phenylalanine hydroxylase (Phenylalanine not oxidized to tyrosine)	Pigmentary dilution of skin, hair, eyes (fair complexion, blond hair, blue eyes), pseudoscleroderma, eczematous dermatitis	- MR, seizures - Phenylpyruvic acid in urine (musty odor) - Screened for at birth - Dietary restriction
Tyrosinemia II (Richner-Hanhart) **AR**	Tyrosine aminotransferase (hepatic) TAT gene	Painful palmoplantar keratoderma	- Herpetiform keratitis, blindness - MR - Corneal ulcers
Homocystinuria **AR**	Cystathionine β-synthase	Fair complexion, malar flush, livedo reticularis, leg ulcers Sparse, fine hair Marfinoid habitus	- Thromboembolic events (50% by 30yo) = common cause of death - Ectopia lentis (downward) - Osteoporosis - MR, developmental delay
Niemann-Pick disease **AR** A lysosomal storage disease	Type A and B: Sphingomyelinase (SMPD1) Type C: NPC1 and 2	Type A and B: ochre to brownish-yellow discoloration of skin, papular lesions face and upper extremities, xanthomas	Type A: severe, CNS deterioration, HSM, failure to thrive Type B: spares CNS, survival to adulthood Type C: childhood, HSM, developmental delay, psychomotor deterioration
Trimethylaminuria "Fish odor syndrome"	Mutation of flavin-containing monooxygenase type 3 (FMO3) gene	Skin, urine, and sweat smell like "rotting fish"	- Smell due to accumulation of trimethylamine - Avoid choline in diet
Lesch-Nyhan (juvenile gout) **XLR**	HPRT1 gene leading to hypoxanthine-guanine phosphoribosyl transferase (HGPRT) deficiency	Loss of tissue around mouth and fingers (due to self-mutilation) Tophaceous deposits (hyperuricemia)	- MR, choreoathoid movements, self-mutilation - Orange crystals in diaper
Wilson's disease (Hepatolenticular degeneration) **AR**	Defect in ATP7B gene (hepatic copper transporting ATPase)	Blue lunulae, Kayser-Fleischer rings (copper deposition in Descemet's membrane), greenish discoloration of face, neck and genitalia, pretibial hyperpigmentation	- HSM, cardiomyopathy, renal tubular acidosis - Progressive neurologic dysfunction (dysrthria, ataxia, dementia) - Lab: low ceruloplasmin - Tx: penicillamine, trientine, zinc supplement
Hartnup disease **AR**	SLC6A19 gene (neutral amino acid transporter)	Pellegra-like dermatitis (photosensitive eruption on face, arms, neck, legs)	- Cerebellar ataxia, MR - Tends to improve with age - Defect in tryptophan transport

Kristina Burke, MD, is a third year resident at Walter Reed National Military Medical Center in Bethesda, Md.

Erin Adams, MD, is a staff dermatologist at Walter Reed National Military Medical Center and also academic staff for the National Capital Consortium Dermatology Residency Program.

Inborn Errors of Metabolism (continued)

by Dr. Burke, MD and Dr. Adams, MD

DISORDER	DEFECT	SKIN FINDINGS	OTHER
Prolidase deficiency	Deficiency of the enzyme prolidase	Skin fragility, lower extremity ulceration, telangiectasias, poliosis	- Mental deficiency, recurrent infections, syndromic facies
Citrullinemia	Type 1: argininosuccinic acid synthetase (ASS1 gene) Type 2: SLC25A13 gene	Resembles zinc deficiency Erythematous, erosive, scaling patches periorally, lower abdomen, and diaper area	- Clears with arginine supplementation
Farber disease A lysosomal storage disease	Ceramidase deficiency	Periarticular swelling, rubbery SQ nodules	- Onset first month of life, death by age 2 - Weak, hoarse cry; pulmonary failure, MR
Adrenoleukodystrophy (Schilder's disease) X-linked	ALD gene	Hyperpigmentation, mild ichthyosis, sparse hair with trichorrhexis nodosa-like features	- Progressive demyelination of cerebral white matter
CADASIL Cerebral autosomal dominant arteriopathy w/ subcortical infarcts and leukoencephalopathy	NOTCH 3 gene	Findings on skin biopsy (eosinophilic granular material in arterial walls)	- Depression, migraine headaches - Multiple cerebral infarcts leading to early dementia - Most common hereditary stroke disorder
Lafora disease (Lafora progressive myoclonic epilepsy)	EPM2A- encoding laforin EMP2B – encodes a ubiquitin ligase	Few – rarely see papulonodular lesions over ears, plaques on arms	- Progressive epilepsy syndrome - Dementia and ataxia - Best site to biopsy = axilla (Lafora bodies around eccrine ducts)
Alagille syndrome **AD**	JAG 1	-Xanthomas, jaundice -Unusual facies	- Congenital intrahepatic biliary hypoplasia w/ cholestasis and pruritus. - Hyperlipidemia - Butterfly-shaped vertebra
Sitosterolemia **AR** "phytosterolemia"	ABCG5 (encoding sterolin-1) or ABCG8 (encoding sterolin-2)	Tuberous and tendinous xanthomas during the first decade of life	- Elevated plasma levels of plant sterols - Arthritis, premature vascular disease, high risk of fatal cardiac events during teenage years
Hurler syndrome **AR** "gargoylism" A lysosomal storage disease	Deficiency of α-L-iduronidase	Diffuse fine lanugo hair, extensive blue pigmentation Facial dysmorphism, large tongue, thick lips	- MR, HSM, corneal opacities, umbilical hernia - Dental abnormalities, persistent rhinitis
Hunter syndrome **XLR** A lysosomal storage disease	Deficiency of iduronate-2-sulfatase	Skin-colored pebbly lesions of the upper back, neck, chest, proximal extremities	- Dysotosis multiplex
Tangier Disease **AR** (Familial α-lipoprotein deficiency)	ATP-binding cassette (ABCA1) transport protein: almost complete absence of plasma HDL and massive deposition of cholesterol esters in tissues	Tonsils are yellow and enlarged. Maculopapular eruption over trunk and abdomen	- HSM, lymph node enlargement, peripheral neuropathy, corneal infiltration in adults - Premature coronary artery disease

References:
1. Bolognia JL, Jorizzo JL, Rapini RP, editors. *Bolognia Textbook of Dermatology. 2 nd ed.* Spain: Mosby Elsevier publishing; 2008: chapters 48 and 62.
2. James WD, Berger TG, Elston DM, eds. *Andrews' Diseases of the Skin: Clinical Dermatology*. 11th ed. Philadelphia, Pa: Saunders Elsevier; 2011: chap 26.
3. Spitz JL. *Genodermatoses: A clinical guide to genetic skin disorders*. Philadelphia: Lippincott Williams & Wilkins; 2005: chapters 8 and 11.

In 2013, the In-Training Examination will be given on Thursday, February 21 in US and Canadian dermatology training programs, and on Monday, February 25 in overseas international dermatology training programs. Information will be emailed to training programs in the fall and the deadline for registering online is November 15. For more information, call the Board office at 313-874-1089.

Do you have an interesting Boards' Fodder? We're expanding our Boards' Fodder archives and you can be a part of it. Contact Dean Monti, managing editor, special publications, at dmonti@aad.org.

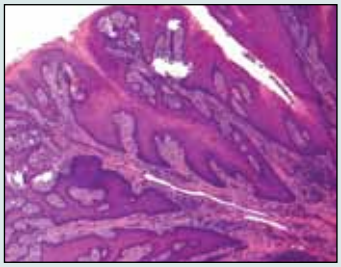
If you enjoyed this special Boards' Fodder edition, let us know. Write to dmonti@aad.org.

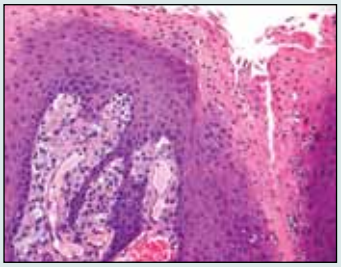
Need more Boards' Fodder? Visit the Directions in Residency *archive listed under "Publications" at www.aad.org.*

Making a case We're hoping to ramp up our archives of Race for the Case. Perhaps you have a clever quiz and a trio of photos you feel would fit in nicely on this page? If you would like to contribute, contact Allison Evans, aevans@aad.org for details on submission.

Race for the Case

By Karolyn Wanat, MD





A 44-year-old woman presented to a community STD clinic with an asymptomatic but growing lesion that occurred after shaving in the suprapubic area. She was otherwise completely healthy. A biopsy was performed.

1) What is the diagnosis?
2) What is the most important histopathologic feature (shown above)?
3) Are there any laboratory abnormalities in these patients?
4) What genodermatoses can these be associated with?

Respond today with the correct diagnosis to Allison Evans, staff editor at the AAD, at aevans@aad.org, and you might win a Starbucks gift card and get your photo in *Directions*.

Race for the Case victory lap

In the last issue, we presented a 57-year-old Caucasian man with history of hypertension controlled with medication who presents with abrupt onset of a new pruritic dermatitis. The questions and correct answers are

1) Diagnosis and what would you expect to see on pathology? : Drug-induced subacute cutaneous lupus erythematosus; on pathology, one would expect to see an interface dermatitis with baso-vacuolar change, possibly eosinophils
2) Most common medication associated with this eruption? Hydrochlorothiazide
3) HLA most commonly associated with this condition? HLA-DR3
4) In the majority of cases, what is the auto-antibody most commonly associated with this? SSA, anti-Ro

Congratulations to Van Hoang, MD, winner of the summer 2012 Race for the Case! She's a third-year dermatology resident at the Montefiore Albert Einstein Dermatology Residency Program in New York. Other than studying dermatology factoids (which she obviously excels at!), she enjoys doing yoga, running in Central Park, and trying new restaurants in New York City. Congratulations and thanks for racing with us!

Van Hoang, MD

Help make us 'appy : AAD seeking new mobile apps

The Academy would like to hear from residents about mobile apps that you use professionally or recommend as health care resources for patients, as well as app ideas or apps in development. Please visit www.aad.org/app-info to share the name of the app, what type of app it is (iPhone, iPad, Android), and how you use it professionally.

Eponyms in Dermatology

by Heather Kiraly Orkwis, DO

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

Bazex syndrome = acrokeratosis paraneoplastica (acquired)

Bazex syndrome = follicular atrophoderma, multiple BCCs, hypotrichosis, localized hypohidrosis (X-linked dominant)

Bazin's disease = erythema induratum, associated with TB

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 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 = X-linked dominant chondrodysplasia punctata

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 equisitely tender lipomas

Gianotti-Crosti syndrome = Papular acrodermatitis of childhood

Goltz syndrome = Focal dermal hypoplasia (PORCN)

Gorlin syndrome = Nevoid basal cell carcinoma syndrome (PATCHED1)

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-transgrediens PPK, esophageal carcinoma (TOC)

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

Hutchinson-Gilford syndrome = Progeria (LAMIN A)

Hutchinson's sign = pigment in paronychial area suggestive of melanoma

Janeway lesions = nonpainful 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 multiple seborrheic keratoses, associated with internal malignancy

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

Louis-Bar syndrome = Ataxia-Telangiectasia (ATM)

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

Lyell's syndrome = Toxic Epidermal Necrolysis

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, arachnodactyly, ectopia lentis, progressive aneurysmal dilation of ascending aorta, CHF (FIBRILLIN 1)

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, adenocarcinoma 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 next to blister easily separated when pressed with a sliding motion, seen in pemphigus vulgaris, staphylococcal scalded skin

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

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

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 dermpath

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

References

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Message from the Chair

Jeremy Brauer, MD

The ABD Board exam. For many, the last exam of our formal medical education — the last hurdle before embarking on the career we've dreamed of for nearly a decade or more. Upon reflection, those are probably the only positive thoughts I have about that exam .... Well, and that it's behind me!

While the board exam is likely the furthest thing from your mind at the start of residency, as you get to know your new co-residents, new computer systems, not to mention learn an entirely new vocabulary, the seed of the board exam has already been subtly planted. As the months of that first year move along and you're finally getting comfortable with the daily routine and developing differential diagnoses, the plant suddenly sprouts and the flowers bloom. The ABD — and likely your attendings and/or program directors — begin to not-so-gently remind you that you should be studying toward the in-service examination that will be administered in the late winter or early spring. While daunting at first, the in-service is a good way to gauge your knowledge base and provides a tangible goal for you to study and work towards as you progress through residency.

So now that the seed/plant metaphor is exhausted, let's get to some useful "deep thoughts" for all the different years of residency from friends and colleagues who recently completed and passed the exam:

"Study regularly and maintain your sanity and you'll be fine!"

"Make subject folders and organize your notes in advance, so you're not doing it in the last few months of residency when all you want to just do is study…"

"Images, images, images — get a good atlas early and review the AAD slide set."

"It's important to see as many clinical presentations of the same diagnosis as possible…"

"Accumulate a 'best of' collection of dermatopathology slides, and look into attending the regional and national courses."

"Practice the timing with the dermatopathology section — as long as it remains timed — since it can add an unnecessary dimension of stress."

"Take advantage of journal clubs and other materials made available for reviewing the latest in the literature."

At the end of the day, we are part of such an amazing specialty. You will pass the boards as long as you take your training and studying seriously — just don't forget to have fun and enjoy it all! DR

Talk back to Directions

A special message from Jeremy Brauer, chair of the Resident/Fellows Committee.

We're working toward improving and enhancing *Directions in Residency*, the official AAD publication of the Resident/Fellows Committee. That's why we hope you take a few minutes to fill out our readership survery, which was sent to all residents last month via email.

We want *Directions in Residency* to reflect your preferences and reading habits, and so we need your help. With only 15 questions, this short survey will help give us information to use going forward. Additionally, because this is a publication that is both for residents and by residents, it is your opportunity to make suggestions that can influence the future of this publication.

If you have not already done so, please make sure to complete this survey, which will provide the necessary data to help improve the publication. There is also room for your comments and suggestions, and we strongly encourage them.

To access this survey, please go to www.aad.org/directionsSurvey. If you have any questions or comments regarding this survey, please feel free to contact the managing editor, Dean Monti, at dmonti@aad.org. Thank you in advance for completing this survey. DR

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AAD Directions in Residency

Fall 2012