

# Alopecias

# Hair Follicle

- Basics
  - Adult human has 5 million follicles
  - All are present at birth
  - About 1 million on head and 100,000 alone on scalp

# Hair Follicle: cycling

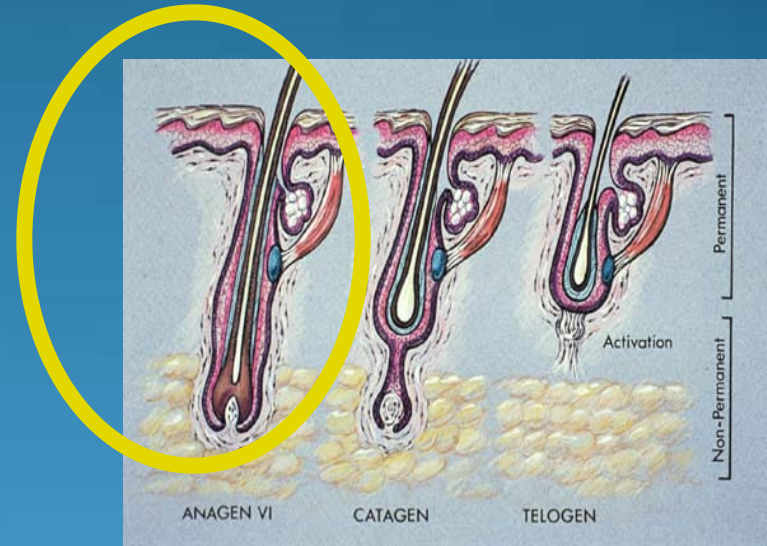
- Each hair follicle perpetually cycles through three stages
- Cycling is independent, not synchronous
- Many molecular signals orchestrate the transition of these stages

# Hair physiology

- With each hair cycle, remodeling of the follicle below the isthmus occurs (impermanant portion)
- Reservoir of pluripotent cells responsible for regeneration reside in the bulge region near the site of arrector pili attachment
- Langerhans cells are concentrated in the infundibular epithelium, bulge and sebaceous epithelium and initiate immune response
- When inflammation is limited to the lower follicle as in non scarring alopecias, the potential to regrow hair remains intact
- When inflammation affects the upper portion of the hair follicle, permanent functional disruption of the bulge area results in permanent alopecia

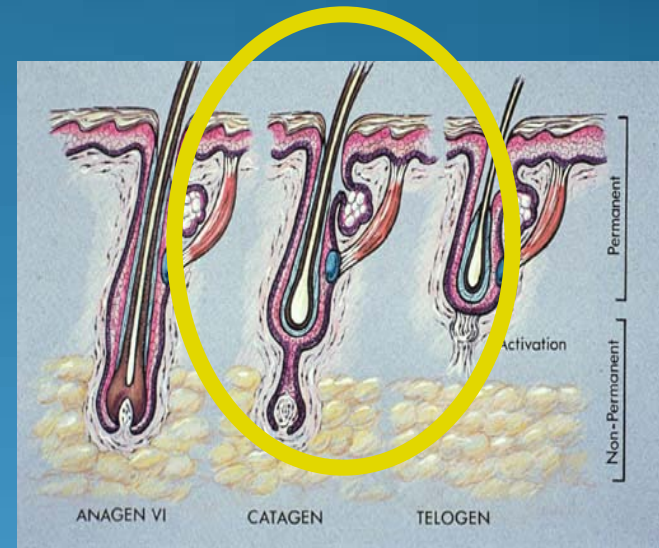
# Hair Follicle: cycling

- Anagen (90%)
  - Growth phase
  - 2-6 years
  - hair grows approximately 0.35 mm/day
  - longer anagen phase= longer hair
- Initiation:
  - insulin like growth factor 1
  - Fibroblast growth factor 7
  - Androgen???
- Cessation:
  - Fibroblast growth factor 5



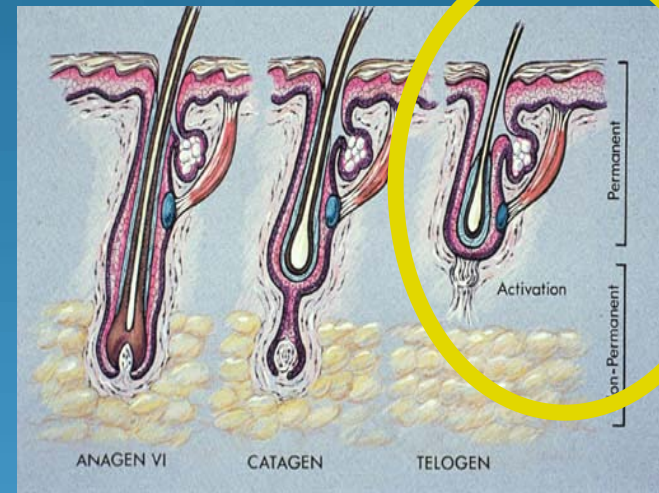
# Hair Follicle: cycling

- Catagen (<1%)
  - Involution phase
  - Lasts 2-3 weeks
  - Apoptosis of follicular keratinocytes
  - Dermal papillae condenses and rises to the bulge

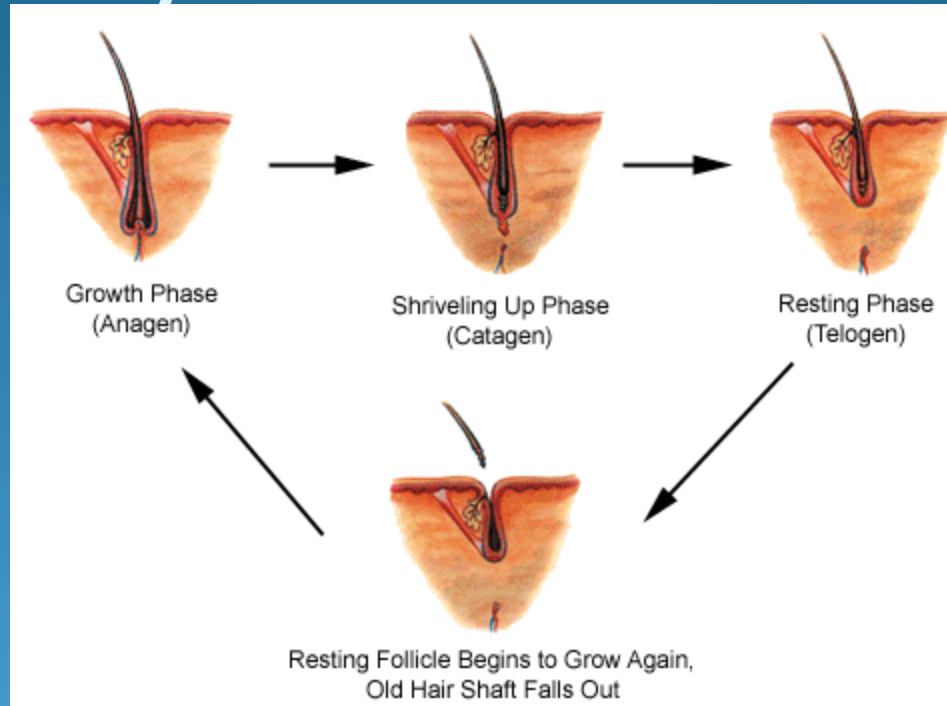


# Hair Follicle: cycling

- Telogen (10%)
  - Resting/ shedding phase
  - Lasts 2-3 months
  - 50 -150 scalp hairs per day



# Hair Cycles





# Evaluating Hair Loss

- Tests:
  - Hair pull
  - Hair pluck
  - Hair analysis (microscope)
  - Hair collection and count
  - Hair shave window
  - Scalp biopsy:
    - Vertical
    - Horizontal
    - DIF

# Non-scarring Alopecias

# Androgenetic Alopecia

- Pathogenesis
  - Androgen dependent
  - Increased 5 alpha reductase activity on testosterone/ DHEA --> increased DHT--> TGF-beta 1 production from dermal papilla cells which mediates hair growth suppression
  - Higher levels of 5 alpha reductase and androgen receptors in frontal hair follicles vs occipital follicles
  - Progressive miniaturization (terminal--> vellus)
    - decrease dermal papilla size, decrease in size of hair matrix, decrease in hair shaft diameter, decrease in anagen phase, more rapid cycling, follicle becomes more superficial with finer, shorter, less pigmented hair

# Androgenetic Alopecia

- Histology
  - Decrease in anagen and increase in telogen follicles
  - Miniaturized follicles and shaft diameter variability
  - Vascular or fibromucinous fibrous tract remnant below miniaturized or telogen follicles

# Androgenetic Alopecia

## (Male Pattern Baldness)

- Clinical
  - Begins in the teens, 20s or early 30s
  - Thought to be autosomal dominant with incomplete penetrance and polygenic inheritance
  - affects 50% of men
- Exam
  - Whisker or kinky hair may be the first sign
  - Most frequent pattern is bitemporal recession and vertex balding
  - Anterior hairline recedes on each side in the Geheimratswinkeln (“professor angles”)
  - Normal hair pull

# Androgenetic Alopecia



Hamilton



In male pattern baldness, hair recedes in an 'm' shape, the crown bald patch eventually meeting the top points to form a horseshoe shape



ADAM

# Male Androgenetic Alopecia

- Treatment
  - Topical minoxidil
    - promotes survival of dermal papilla cells, induces and prolongs anagen phase, and results in enlargement of shaft diameter (vellus-->terminal hairs)
  - Finasteride (men aged 18-41)
    - Type 2 5-alpha reductase inhibitor
    - Prolongs anagen phase
    - 1 mg daily can prevent further hair loss and increase hair counts
    - hair patterning on the temples is not improved
  - Hair transplantation of occipital hair follicles to anterior scalp

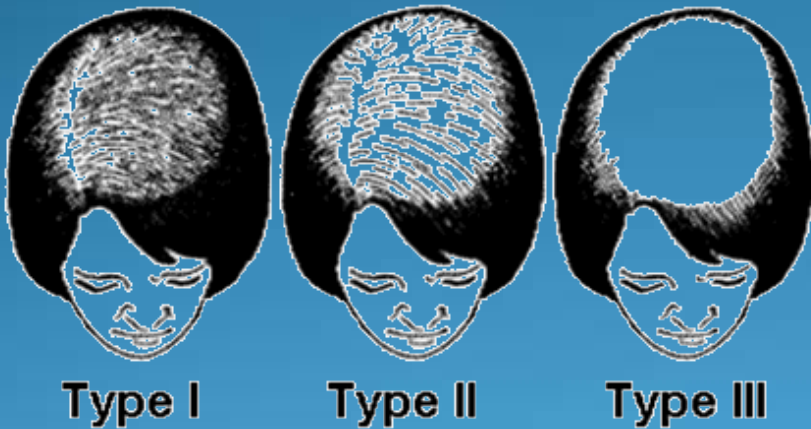
# Female Androgenetic Alopecia (Female Pattern Hair Loss)

- History
  - Gradual, may begin anytime after puberty with peaks in the 20s and 40s, perimenopausally, or at times of hormone change
- Clinical
  - Diffuse thinning over frontoparietal scalp with preserved frontal hairline
  - Christmas tree pattern midline part - widest anteriorly
  - Subset with male type pattern of bitemporal recession more common in postmenopausal women
  - Part width on crown > occiput
  - Hair pull test may be negative or positive
  - Trichodynia common symptom



# Female Androgenetic Alopecia

Ludwig's classification



# Female Androgenetic Alopecia

- Workup
  - Determined by signs and symptoms
  - Consider TSH, T<sub>3</sub>, T<sub>4</sub>, ferritin, DHEA-S, free testosterone and total testosterone
  - Most have normal androgen levels

# Female Androgenetic Alopecia

- Treatment
  - Topical minoxidil
  - Oral antiandrogens
    - spironolactone
    - cyproterone acetate
      - more effective than minoxidil when when signs of hyperandrogenism, hypersoberhhea, menstrual abnormalities and high body mass index
  - Finasteride in subset with temporal recession
  - Hair transplantation, wigs, interwoven hair

# Telogen Effluvium

- Clinical

- Increased shedding of club hairs with diffuse hair loss “from the roots”
- May lose 150-400 per day
- Trichodynia common symptom and often coexist with signs of depression, obsessive personality disorder and anxiety
- Acute TE-
  - Often triggering event 2-5 months before onset i.e. illness, parturition, operation, hospitalization, loss of loved one, divorce, medications change, crash diet
  - self limited usually within 6-12 months
- Chronic TE-
  - insidious onset or acute TE that never resolves
  - lasts 6 months to years
  - continuous or fluctuating hair loss

# Telogen Effluvium

- Exam

- Normal scalp, diffuse thinning, part width of crown = occiput
- Positive hair pull test-
  - grasp 40 hairs firmly and pull slowly
  - >4-6 club hairs is positive
- Other tests if hair pull test not positive
  - daily hair shed counts
  - collect all hairs lost during pre shampoo 1 minute combing session (back to front) x 3 consecutive days (10-15 normal, >50 common in TE)
  - clip test
  - trichogram evaluation
- Normal telogen hairs-depigmented club shaped bulb, no sheath
- Hair often appears to be of normal thickness (perceptibly thin hair more common with preexisting pattern alopecia)
- Shorter regrowing hairs in frontal and bitemporal areas



# Telogen Effluvium

- Genetics
  - no known genetic cause
- Histology
  - Normal follicles without inflammation
  - Active phase- >12-15% of terminal follicles in telogen
- Associations
  - Iron deficiency, thyroid disease, and medications (beta blockers, some antihyperlipemics, NSAIDS, HRT, anticoagulants)

# Telogen Effluvium

- Types

- 1 Immediate anagen release- Premature conversion of many anagen hairs through catagen to telogen induced by stressor (e.g. drugs, high fever, starvation/dieting)
- 2 Delayed anagen release- Prolonged anagen phase secondary to pregnancy hormones with large shift to telogen once hormonal drive removed
- 3 Short anagen- shorter hair which traverses the hair cycle more quickly and is shed more often- chronic TE and androgenetic alopecia
- 4 Immediate telogen release- can occur with topical minoxidil
- 5 Delayed telogen release- no known clinical correlate
- 6 Chronic



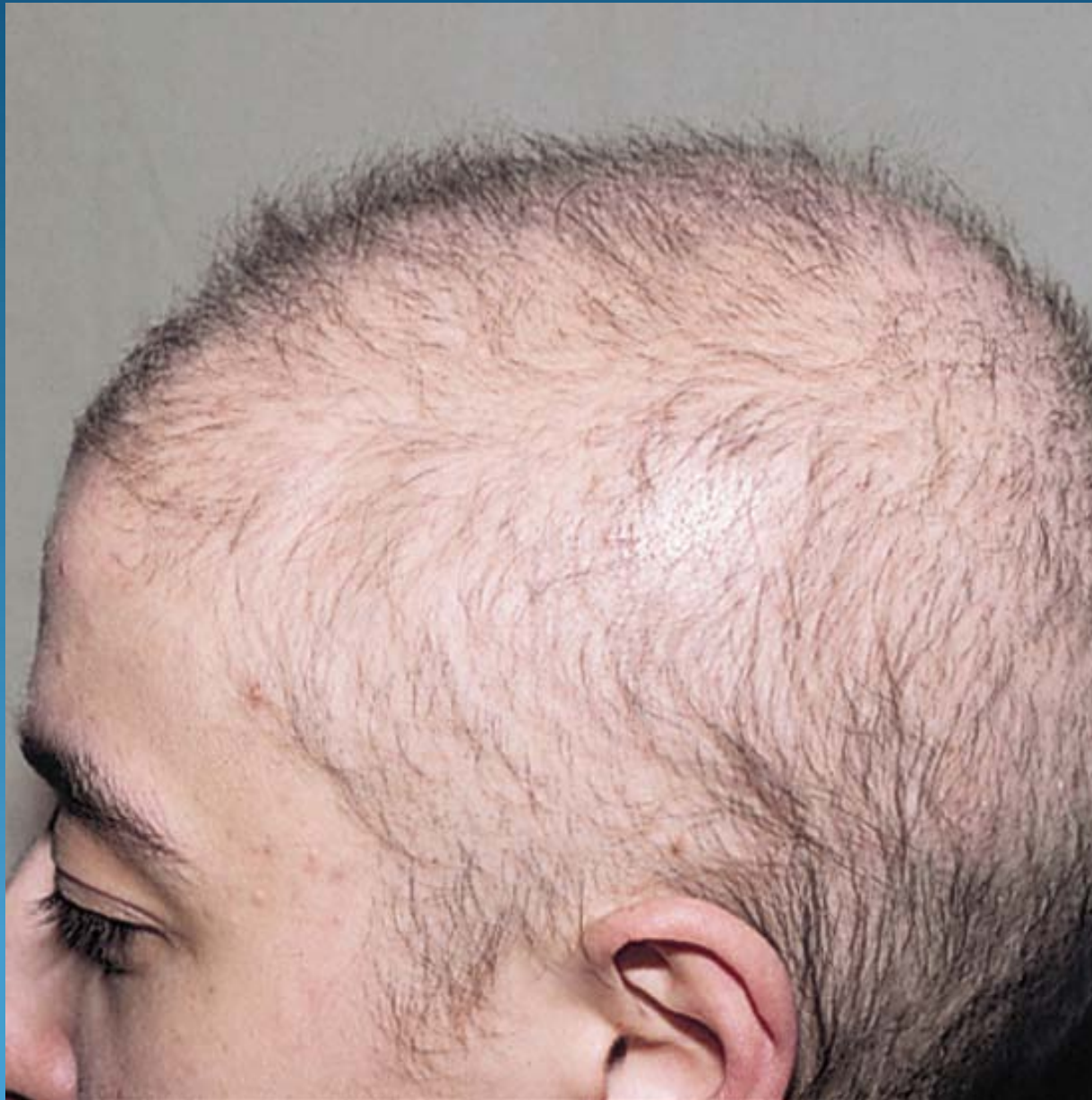
# Telogen Effluvium

- Evaluation
  - Iron and thyroid labs if history suggestive or if course prolonged
  - scalp biopsy if diagnosis unclear
- Treatment
  - Address any coexisting scalp disease
  - Iron replacement
  - 5% Minoxidil



# Anagen Effluvium

- History
  - Abrupt loss of all growing hairs (90%) secondary to hair shaft fracture
  - 1-2 months following chemotherapy/trigger
- Exam
  - Diffuse alopecia, no scalp irritation
- Histology
  - Normal



# Anagen Effluvium

- Pathogenesis
  - It is always abnormal to shed anagen hairs
  - Mitotic inhibition temporarily shuts down the rapidly dividing hair matrix cells resulting in tapering of the shaft (Pohl-Pinkus constrictions) and breakage of the hair when this thin portion reaches the surface
- Treatment
  - Drug cessation
  - Re-growth begins a few weeks after insult
  - May have different color or texture
  - Pressure cuff around scalp during chemo and scalp hypothermia may prevent
  - Minoxidil may shorten period of baldness by 50 days

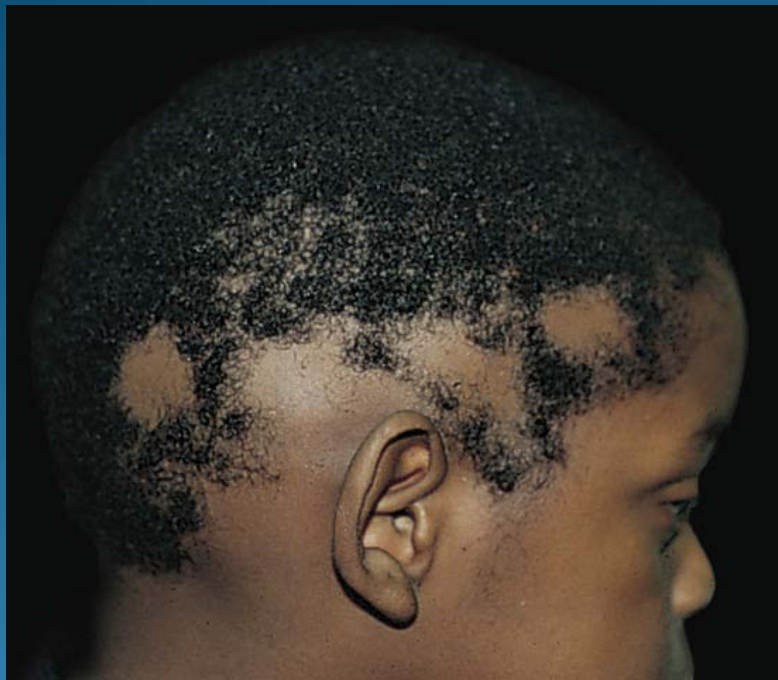
# Alopecia Areata

- Clinical
  - Rapid, complete loss of hair in patches
  - Types
    - diffuse (least common)- widespread thinning
    - totalis
    - universalis
    - ophiasis (bandlike along peripheral temporal and occipital scalp)
    - sisaipho (loss of hair on entire scalp except for temporal and occipital)
  - generally presents as an anagen effluvium, with an inflammatory insult to the hair matrix resulting in tapering of the hair shaft and fracture of anagen hairs
  - migratory poliosis

# Alopecia Areata

- Exam
  - Bald, circumscribed patches
  - Exclamation point hairs at periphery (distal end broader than proximal end)
  - White re-growth
  - If active, positive hair pull at periphery
  - associated nail findings
    - uniform pitting, trachyonychia, onychomadesis, red or spotted lunulae





# Alopecia Areata

- Epidemiology
  - M=F
  - increased risk with
    - other autoimmune conditions (vitiligo, DM, lupus erythematosus, RA, thyroiditis, myasthenia gravis)
    - atopic dermatitis, Down syndrome, lichen planus
- Genetics
  - 25% with positive family history
  - Family history strongest in early onset disease (<30 y/o)
  - early onset severe familial disease associated with HLA DR4, DR11, and DQ7

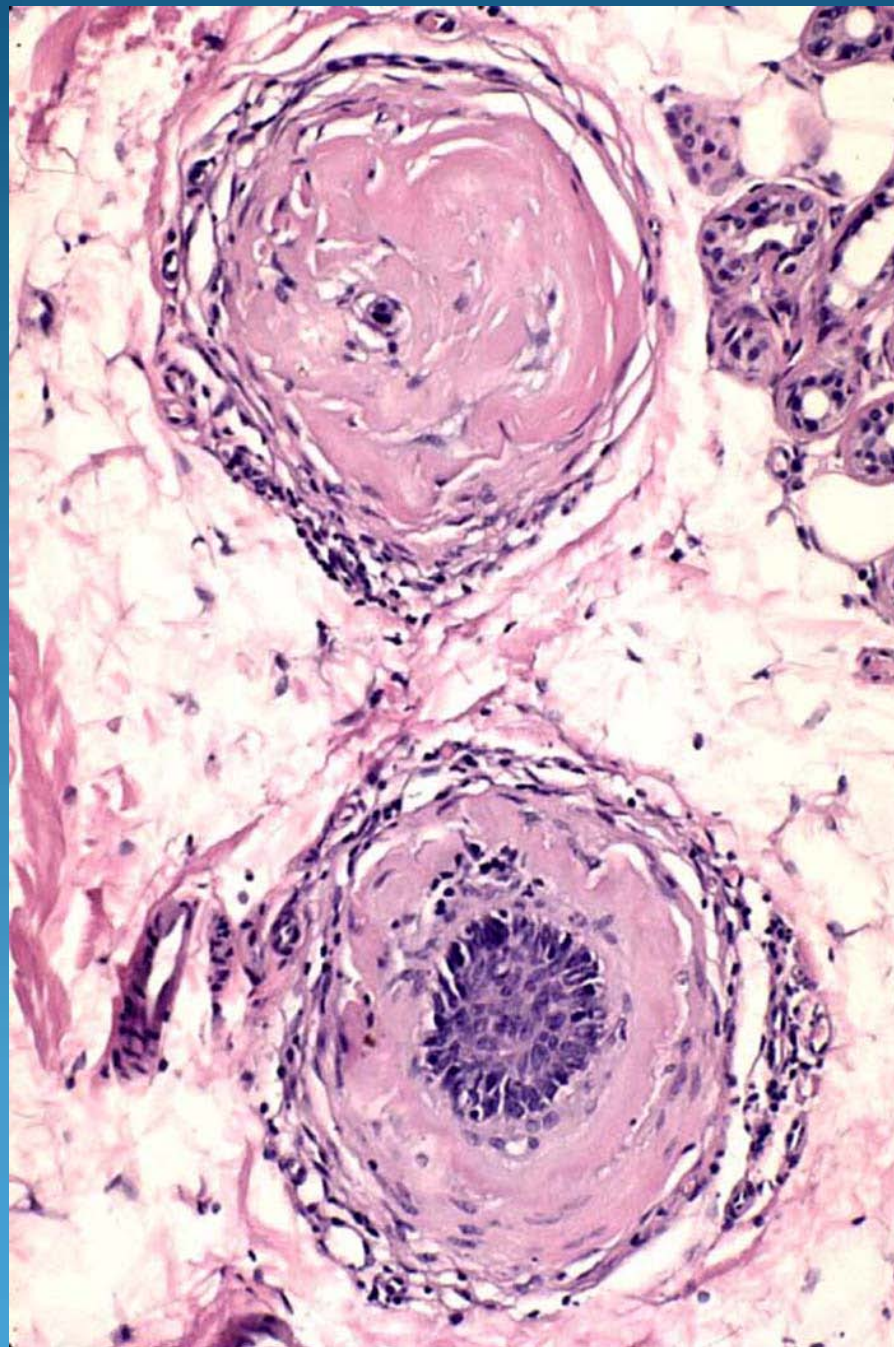


# Alopecia Areata

- Pathogenesis
  - T cell directed autoimmune attack on hair follicle
  - target= follicular melanocytes (white hair rarely affected, regrowing hair often depigmented)
  - collapse of immune privilege?
    - normal anagen hair bulb lacks MHC I and II
    - HLA expressed in AA allowing interaction of cytotoxic T cells with matrix cells

# Alopecia Areata

- Histology
  - “swarm of bees”: T-cell infiltrate around anagen hair bulbs
  - miniaturization of follicles
  - fibrous tract remnants beneath miniaturized bulbs contain lymphoid cells, eos, and melanin pigment
- Work up
  - thyroid function tests if suggestive signs or symptoms or if strong family history of thyroid disease
- Prognosis
  - high rate of spontaneous recovery in postpubertal onset
  - poor prognosis with atopic dermatitis, childhood onset, widespread involvement, ophiasis, duration longer than 5 years, onychodystrophy



# Alopecia Areata

- Treatment
  - IL steroids treatment of choice for localized cosmetically conspicuous patches
  - high strength topical steroids
  - Immunomodulators
  - Squaric acid dibutyl ester, DNCB, diphencyprone in refractory cases (blockade of leukocyte trafficking and extravasation)
  - PUVA
  - SCAT
  - topical minoxidil

# Trichotillomania

- History:
  - Often presents in childhood, F>M
  - Patient unaware of own behavior; presents with “hair loss”
  - May elicit history of stress/anxiety when hair loss began
  - may relate history of localized follicular pain relieved with plucking
- Exam:
  - irregularly shaped patches
  - hairs of various lengths
  - Normal hair pull test
  - Hair re-growth in shaved “windows”
- Pathogenesis:
  - Self-induced plucking or breaking of hair
  - Manifestation of an obsessive-compulsive disorder/ anxiety/ depression

# Trichotillomania

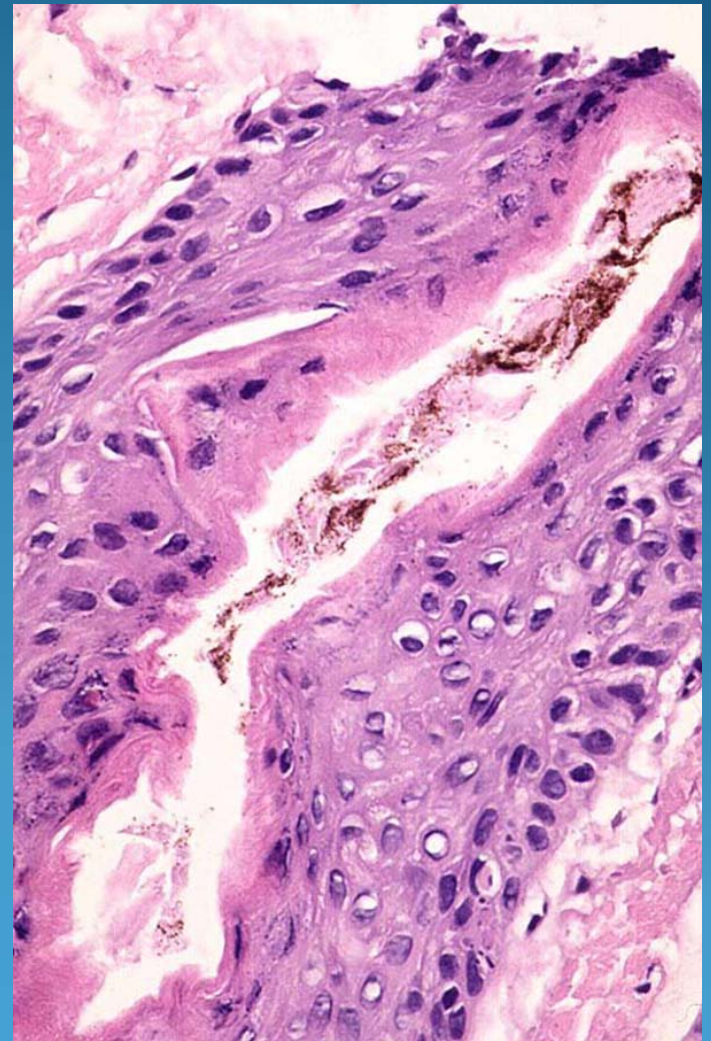




# Trichotillomania

- Histology:
  - empty anagen follicles
  - increased catagen hairs
  - pigment casts within infundibulum
  - trichomalacia
  - hemorrhage
- Treatment:
  - Psychiatric evaluation
  - SSRIs

# Trichotillomania





# Traction Alopecia

- Clinical

- African Americans, Japanese women, and Sikh men in India
- Children and young adults
- Hair “won’t grow” along the periphery of scalp
- Usually complain of itching and dandruff

- Exam

- Bilateral hair loss along periphery, especially temples and above the ears
- Initially, pruritus, perifollicular erythema, and hyperkeratosis may be present, creating a seborrheic picture
- abundance of broken hairs
- scale and pustules
- may have positive hair pull test
- persistent traction--> follicular atrophy --> fine, short hair

# Traction Alopecia



# Traction Alopecia

- Pathogenesis

- Prolonged tension on hair causes increased release from roots and can lead to secondary inflammation
- Excessive traction for prolonged periods leads to conversion of the anagen phase to the telogen phase

- Histology

- Increased telogen count
- Late development of fibrous tracts

- Treatment

- Prevention- process is reversible early

# Scarring Alopecias

# Scarring Alopecias

- Diagnostic hallmarks are visible loss of follicular ostia and destruction of the hair follicle on histopathologic examination
- Primary
  - Target of destruction is the hair follicle
- Secondary
  - Nonfollicular disease indirectly causes follicular destruction e.g. sarcoid, morphea, leprosy
- many scarring alopecias begin as nonscarring
- inflammation typically affects upper portion of follicle

# Scarring alopecias

- Treatment
  - should be managed as a “trichologic emergency” during initial nonscarring phase
  - Topical minoxidil if coexistent AGA- may improve cosmesis by enlarging miniaturized hairs (1 mL of 5% sol'n bid x at least 1 year)
  - wig, hair color-matched powder
  - hair transplantation and scalp reduction once completely burned out
  - sun protection

# Proposed NAHRS working classification of primary cicatricial alopecia

- **Lymphocytic**
  - Chronic cutaneous lupus erythematosus
  - Lichen planopilaris
    - Classic
    - Frontal fibrosing alopecia
    - Graham- Little syndrome
  - Central centrifugal cicatricial alopecia
  - Alopecia mucinosa
  - Keratosis follicularis spinulosa decalvans
- **Neutrophilic**
  - Folliculitis decalvans
  - Dissecting cellulitis
- **Mixed**
  - Acne keloidalis
  - Acne necrotica
  - Erosive pustular dermatosis
- **Nonspecific**

# Lymphocytic Cicatricial Alopecias



# Discoid Lupus

- Clinical
  - females > males
  - onset between 20 and 40 years of age
  - Patients complain of hair loss, pruritus, stinging, burning, scalp tenderness
- Exam
  - Well-circumscribed, coin shaped erythematous, atrophic plaque with mottled hyper- and hypopigmentation
  - Plugged follicular ostia (“carpet tacking”)
  - Pull test often yields anagen hairs
  - May show other rash of DLE
- Pathogenesis
  - Autoimmune
  - UV light possible antigenic stimulus



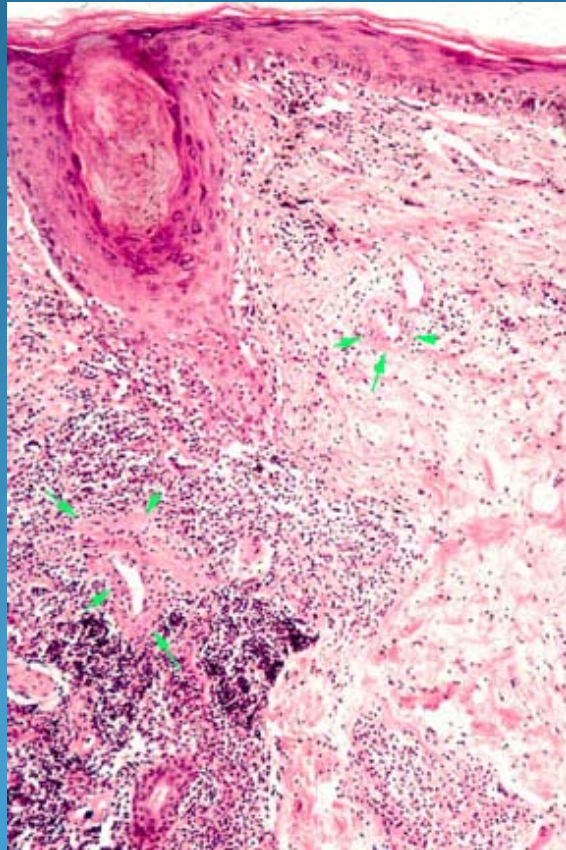
# Discoid Lupus

- Course
  - Spontaneous remission in 1/3 to 1/2 within 4 years
  - 5-10% of adults and 26-31% of children and adolescents --> SLE
    - course of systemic disease often severe with renal or neurologic involvement unless SLE predates DLE
- Complications
  - disfigurement
  - ulceration
  - SCC- 31% metastasis rate

# Discoid Lupus

- Histology
  - hyperkeratosis, follicular plugging, vacuolar interface dermatitis, BMZ thickening, dermal mucin, pigment incontinence
  - patchy perivascular and periadnexal inflammation
  - perifollicular lymphoid infiltrates involve the isthmus
  - inflammation of fibrous tract remnants creates dense vertical columns of lymphocytes
  - scarring of follicular units and intervening dermis
  - DIF- “full house”-continuous granular IgG , IgM, IgA, and C<sub>3</sub> at D-E junction

# Discoid Lupus



- Follicular plugging
- Perivascular hyalinization
- Follicular lichenoid infiltrate
- Thick, hyalinized BMZ
- Dermal mucinosis.

# Discoid Lupus

- Treatment
  - Sun avoidance
  - Limited active disease
    - class I or II steroids topically or intralesional steroids or both
    - Reports of calcineurin inhibitors, imiquimod, tazorac
  - Rapidly progressive or extensive disease
    - antimalarials
    - Oral prednisone as temporizing measure if severe
    - Oral retinoids if resistance to antimalarials
    - Dapsone, thalidomide, sulfasalazine, mycophenolate mofetil, and methotrexate also been reported
  - Low threshold to biopsy persistent treatment resistant hyperkeratotic or ulcerated foci

# Lichen Planopilaris

- A follicular variant of lichen planus
- Hair is a necessary cofactor
- 3 forms
  - Classic lichen planopilaris, Frontal fibrosing alopecia, and Graham- Little syndrome

# Lichen Planopilaris

- Pathogenesis

- Similar to LP
- Postulated that an antigenic trigger (i.e certain medications) initiates disease and ultimately elicits lesional expression of keratinocyte autoantigens with an ensuing CD 8+ T-cell mediated lichenoid response with destruction of follicular basilar epithelium
- Implicated medications include gold, atabrine, quinacrine, hepatitis B vaccine
- associated with Hepatitis C



# Lichen Planopilaris

## Classic

- Clinical
  - onset in middle age
  - females > males
  - 17%-28% with extracranial LP at presentation and 50% during course
  - Common symptoms include hair loss, pruritus, scaling, burning, and scalp tenderness

# Lichen Planopilaris

## Classic

- Exam
  - Active disease marked by follicular hyperkeratosis and perifollicular erythema
  - Burnt out disease marked by depigmented shiny patches
  - typically multifocal and central
  - hair loss and scarring after months
  - disease activity limited to hair bearing periphery of cicatrized alopecia (vs DLE, alopecia mucinosa)
  - pull test positive for anagen hairs



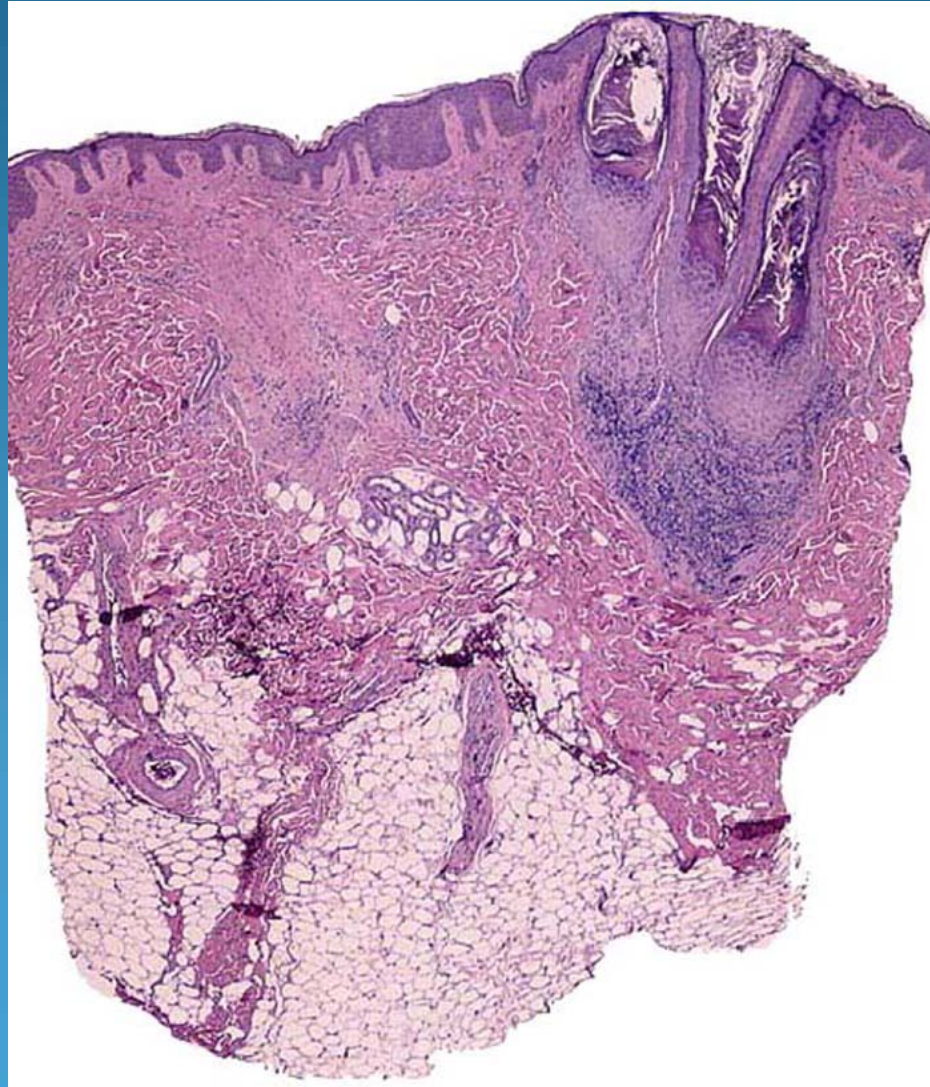
# Lichen Planopilaris

## Classic

- Pathology
  - D-E junction of the upper follicle (infundibulum in particular) obscured by lichenoid lymphocytic infiltrate
  - Follicular destruction and foreign body hair shaft granulomas with evolution
  - wedge shaped superficial dermal scar
  - DIF- patchy or shaggy fibrinogen and clumped IgM along the follicular BMZ



# Lichen Planopilaris



# Lichen Planopilaris

## Classic

- Evaluation
  - drug history
  - hep C status if eroded or ulcerated scalp lesions
- Treatment
  - local disease
    - topical moderate or high potency topical steroids, intralesional steroids, or both
  - steroid refractory, rapidly progressive, or extensive active or symptomatic disease
    - low dose oral retinoids
    - antimalarials
    - reports of griseofulvin, cyclosporine, and low molecular weight heparin

# Lichen Planopilaris

## Frontal fibrosing alopecia

- A patterned variant of LPP that primarily affects postmenopausal women
- Clinical
  - shiny, uniformly pale, bandlike zone of incomplete hair loss 1-8 cm in width affecting frontotemporal hairline
  - new hairline with perifollicular erythema and hyperkeratosis
  - thinned to absent eyebrows
  - nonscarring axillary and extremity hair loss
  - may also have classic LPP or extracranial LP

# Lichen Planopilaris

## Frontal fibrosing alopecia

- Pathology
  - Indistinguishable from classic LPP
- Treatment
  - twice daily midpotency topical steroids may lead to stabilization
  - oral prednisone or chloroquine may be tried for rapidly progressive disease



# Lichen Planopilaris

Frontal fibrosing alopecia



# Lichen Planopilaris

## Graham-Little syndrome

- AKA Graham- Little- Piccardi- Lassuer syndrome
- Clinical
  - adults
  - patchy cicatricial alopecia of the scalp with follicular hyperkeratosis or erythema
  - nonscarring alopecia of axillae and pubis
  - Grouped spinous follicular papules that resemble lichen spinulosus or KP on the trunk and extremities
- Pathology
  - features of LPP or keratosis pilaris atrophicans
- Treatment
  - reports of topical, intralesional, or oral steroids, cyclosporine

# Lichen Planopilaris

## Graham- Little syndrome



# Pseudopelade of Brocq

- Named for its likeness to alopecia areata (*la pelade*)
- AKA alopecia cicatrisata
- distinct entity vs variant of certain primary cicatricial alopecias (LPP and DLE) vs common final stage of several cicatricial alopecias
- Clinical
  - chronic, insidious onset
  - typically asymptomatic

# Pseudopelade of Brocq

- Exam
  - confetti like, coin sized, hypopigmented or flesh colored, atrophic oval to round plaques of noninflamed cicatricial alopecia (“footprints in the snow”)
  - commonly involves the vertex
  - no pustules, crusts or broken off hairs
  - never has an inflammatory stage
  - pull test positive for anagen hairs

# Pseudopelade of Brocq

- Histology
  - no pathognomonic features described
  - absent epidermal atrophy and interface changes
  - DIF usually negative
- Treatment
  - reported success with topical steroids, intralesional steroids, prednisone, hydroxychloroquine, isotretinoin
  - the disease usually reaches an inactive endpoint after many years





# Central Centrifugal Cicatricial Alopecia

- Encompasses the terms *hot comb alopecia*, *follicular degeneration syndrome*, *pseudopelade in African Americans*, and *central elliptical pseudopelade in Caucasians*
- Unique entity vs common morphologic pattern shared by different disorders
- Clinical
  - Slowly progressive, symmetric, noninflammatory cicatricial alopecia of the central scalp
  - Most commonly seen in African American females
  - May have islands of unaffected hair within areas of scar
  - Frequently asymptomatic, but pins and needles sensation, pruritus and tenderness also common



# Central Centrifugal Cicatricial Alopecia

- Pathophysiology
  - Unclear whether due to intrinsic defect in inner root sheath desquamation or exogenous factors such as various hair styling procedures
- Pathology
  - Premature inner root sheath desquamation, perifollicular lymphocytic infiltrate surrounding upper follicle, follicular fibrosis in end stage disease
- Treatment
  - Discontinue traumatic hair grooming practices
  - Success reported with daily potent topical steroid and tetracycline 500 mg bid



# Alopecia Mucinosa

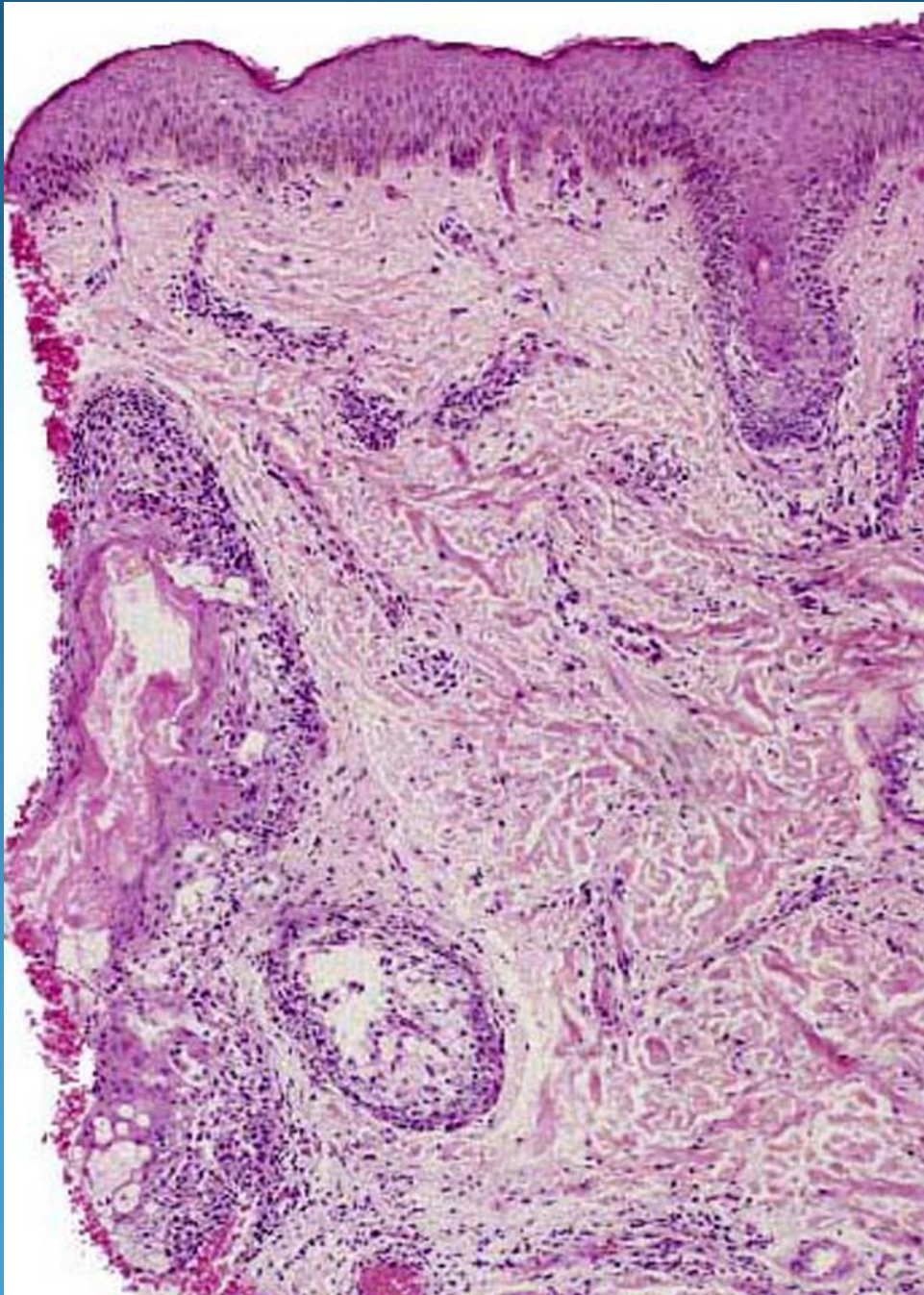
- Inflammatory condition of the pilosebaceous unit that can result in nonscarring and scarring alopecia
- Alopecia mucinosa = disease process
- Follicular mucinosis= histologic features
- Skin limited vs associated with follicular MF
- Clinical
  - All ages
  - Lesional pruritus, sensory dissociation, anhidrosis
- Exam
  - Classically involves head and neck
  - hypopigmented or erythematous, scaly or eczematous plaques, or plaques composed of flesh colored follicular papules
  - May see black dot sign because affected hair shafts prone to breakage



# Alopecia Mucinosa

- Associated malignancy
  - No reliable clinical criteria for differentiation of benign from malignancy associated disease
  - Adults- MF reported to occur in 9-60%
  - Children and young adults- Hodgkin's lymphoma
  - Can present as a paraneoplastic phenomenon
  - Follicular MF is more refractory to treatment and has a worse prognosis than classic CTCL
- Pathology
  - Lesional T cell clonality is common but doesn't connote malignancy or predict progression to lymphoma
  - mucin (hyaluronic acid) within cells of sebaceous gland and outer root sheath
  - No concentric lamellar fibrosis vs other primary cicatricial alopecias





# Alopecia Mucinososa

- Treatment
  - Regular examinations with lymph node palpation
  - Serial biopsies with disease progression
  - Topical, intralesional, and oral steroids, minocycline, anti staph antibiotics if staph culture positive, topical and oral retinoids, dapsona, topical and oral indomethacin, topical nitrogen mustard, phototherapy, excision, PUVA

# Keratosis Follicularis Spinulosa Decalvans

- AKA keratosis pilaris decalvans, Siemens-1 syndrome
- Characterized by cicatricial alopecia of the scalp, widespread KP followed by atrophy, and photophobia
- Morphologically similar to ulerythema ophryogenes and atrophoderma vermiculata
- defect on gene for spermidine/spermine N(1)-acetyltransferase in one X linked form



# Keratosi Folliculari Spinulosa Decalvans

- Clinical

- onset in infancy or early childhood with follicular hyperkeratosis first on the face with eventual widespread distribution
- then develop patchy scalp, eyebrow, and eyelash alopecia followed by scarring
- onset of photophobia coincides with cutaneous disease and is marked by corneal dystrophy with punctate defects

# Keratosis Follicularis Spinulosa Decalvans

- Pathology
  - compact hyperkeratosis and hypergranulosis of the upper follicular epithelium early
  - sparse perivascular and perifollicular mononuclear cell infiltrate with mucin and loose connective tissue around the upper follicle with advancing disease
  - granulomatous inflammation with follicular destruction, concentric perifollicular and horizontal adventitial lamellar fibrosis, and scarred follicular tracts late
- Treatment
  - sustained topical and intralesional steroids
  - oral retinoids
  - baseline and routine ophthalmologic exams

# Neutrophilic Cicatricial Alopecias

# Folliculitis Decalvans

- A destructive suppurative folliculitis
- Clinical
  - young and middle aged adults
  - typically affects scalp
  - periodic crops of painful or pruritic follicular pustules or papules that coalesce and eventually form atrophic areas of scarring alopecia
  - active disease continues peripherally as a marginated perifollicular pustules and tufted folliculitis
  - chronic and slowly progressive

# Folliculitis Decalvans



# Folliculitis Decalvans

- Pathogenesis

- *S aureus* usually isolated from pustules and thought to be etiologic antigen by some
- Postulated that follicular destruction secondary to abnormal suppurative immune response likely incited by staph and other organisms vs chronic staph infection

# Folliculitis Decalvans

- Histology
  - intra- and peri- follicular neutrophilic infiltrate affecting upper and middle parts of follicle
  - wedge shaped superficial dermal scar
- Treatment
  - if other symptoms of immunodeficiency, evaluate accordingly
  - culture pustules
  - anti-staph antibiotic
  - the combination of rifampin + clindamycin, fusidic acid (alternatively topical mupirocin or topical erythromycin) and oral zinc sulfate have resulted in remission for months to years

# Tufted Folliculitis

- Pattern of scarring alopecia seen in a wide range of scarring alopecias
- presents with doll's hair like bundling of follicular units
- 5-20 hairs emerge from common dilated follicular orifice





# Dissecting Cellulitis

- AKA Perifolliculitis capitis abscedens et suffodiens
- Thought to be result of abnormal keratinization leading to obstruction, secondary bacterial infection, and follicular destruction
- Clinical
  - >80% black males 18 to 40 years old
  - vertex and occiput are sites of predilection
  - follicular inflammatory nodules suppurate and undermine to form intercommunicating sinuses
  - initially develop a nonscarring alopecia over nodules which eventually becomes scarring
  - ~ 1/3 have coexisting acne conglobata or hidradenitis suppurativa and at risk for HLA B27 negative spondyloarthropathy



# Dissecting Cellulitis

- Histology
  - intra- and peri- follicular neutrophilic infiltration
  - with follicular perforation, develop abscesses in perifollicular mid to deep dermis and superficial fat
  - Inflammation replaced by fibrosis
- Treatment
  - isotretinoin first line
    - 1 mg/kg/d x 4 months minimum followed by 0.75-1 mg/kg/d x 5-7 months
  - intralesional steroids
  - oral antibiotics
  - oral zinc
  - I and D or excisional CO<sub>2</sub> laser with secondary intention healing/ marsupialization

# Mixed Cicatricial Alopecias

# Acne Keloidalis

- Clinical

- predominantly affects the nuchal hairline of young black, Hispanic or Asian postpubertal males
- A unique race related property of the pilosebaceous unit, hair shaft, or scalp skin has been postulated to underlie disease evolution
- Flesh colored to reddish brown smooth firm follicular papules
- May enlarge and coalesce into a large keloidal plaque
- Proposed precipitants include mechanical trauma (shirt collar, excoriation) , infection (demodex, bacteria), and autoimmunity
- Drug induced reports in white males
  - diphenylhydantoin + carbamazepine (1)
  - cyclosporine (4)

# Acne Keloidalis

- Pathology
  - follicular dilatation and peri infundibular mixed infiltrate
  - eventual follicular rupture with granuloma or microabscess formation around extruded hair shaft fragments
  - no keloidal collagen
- Treatment
  - topical and intralesional steroids
  - topical retinoids
  - topical and oral antibiotics
  - surgical excision



# Acne Keloidalis



# Acne Necrotica

- 2 forms
  - acne necrotica miliaris
    - nonscarring superficial folliculitis that possibly represents variant of same disease process with individual host response responsible for clinical expression
  - acne necrotica varioliformis
    - necrotizing follicular disorder that heals with varioliform scars
    - proposed causes: abnormal response to *S aureus* or *P acnes*, excoriation of underlying folliculitis, and rosacea like genesis



# Acne Necrotica

- Clinical (varioliformis)
  - chronic relapsing disorder of adults
  - commonly involves anterior hairline, also seborrheic face and chest
  - crops of pruritic, tender red-brown papules or pustules that umbilicate and undergo central necrosis leaving varioliform scars
- Pathology
  - confluent necrosis of follicular epithelium and adjacent epidermis containing fragmented bits of hair
- Treatment
  - culture directed antibiotics
  - oral tetracyclines (if culture negative)
  - topical or intralesional steroids if refractory
  - isotretinoin in culture proven *P acnes* or intractable cases
  - doxepin

# Erosive pustular dermatosis

- Idiopathic, chronic sterile pustular dermatosis of scalp resulting in scarring alopecia
- Often history of trauma (sunburn, zoster, abrasion, radiation, topical 5-FU, cryotherapy, topical tretinoin, chronic actinic damage)
- Clinical
  - usually elderly
  - female predominance
  - large, asymptomatic, boggy, superficially crusted plaque that when unroofed reveals a beefy red exudative erosion with flaccid pustules
  - episodic pustular flares with slow enlargement

# Erosive pustular dermatosis

- Pathology
  - nonspecific
- Treatment
  - typically improves rapidly with class I and II topical steroids BID, requires maintenance
  - antibiotics
  - topical calcipotriol 0.005% cream BID x 2 months
  - zinc sulfate