“Why is my skin this color?”

Disorders of pigmentation

Scott A. Norton, MD, MPH
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Disorders of pigmentation

No financial conflicts of interest.
Will discuss off-label uses.

“Why is my skin this color?”

Terminology of Pigmentation

Disorders of Hypopigmentation

Disorders of Hyperpigmentation

Treatment of Common Conditions

When to Refer

Causes of Macular (Flat) Hypo- & Hyperpigmentation

- Psoriasis vulgaris
- Psoriasis pustulosa basis
- Psoriasis etioietica
- Solar lentigo
- Lupus erythematosus
- Physical agents
- Infections
- Malignant neoplasms
- Chemotherapeutic agents
- Lithium dates
- Alcohol
- Other medications
- Other dermatologic diseases
- Certain systemic diseases
- Infections
- Disorders of pigmentation
- Melanocytic dysplasia
- Verruca vulgaris
- Congenital nevi
- Café-au-lait macules
- Neurofibromatosis type I
- Neurofibromatosis type II
- Maffucci syndrome
- McCune-Albright syndrome
- Incontinentia pigmenti
- Nevus anemicus
- Bier’s spots
- Nevus of Ota
- Nevus of Ito
- Mongolian spots

4 components of normal skin pigmentation

- Epidermal melanin (brown) -- most important pigment.
- Oxygenated hemoglobin (red).
- Deoxygenated hemoglobin (blue).
- Carotenoids (yellow).

Disorders that are too light

Is it Hypo- vs Depigmented?

Is it Congenital or Acquired?

Is it Generalized or Focal?
Hypopigmented

Depigmented or nonpigmented

Albinism (various types)

Albinism variants
Various genetic disorders: Phenylketonuria (PKU), Chediak-Higashi, Griscelli, Hermansky-Pudlak, etc.
Paternity issues

Always refer. Requires annual examination.

http://albinism.med.umn

Albinism

The point is - enzymatic defect in the melanin synthesis pathway can lead to albinism.

Always refer for diagnostic evaluation.

Congenital focal lack of pigment

Piebaldism
Tuberous sclerosis (ashleaf macules & confetti spots)
Nevus depigmentosus

Refer if syndromic.

Congenital focal lack of pigment

Piebaldism

White forelock, autosomal dominant, ventral > dorsal
Waardenburg’s syndrome

- Piebaldism
  - white forelock
  - other ventral midline sites
- Sensorineural deafness
- Pseudohypertelorism (dystopia canthorum)

Ashleaf macules & confetti spots of tuberous sclerosis

Sensorineural deafness

Pseudohypertelorism (dystopia canthorum)

Tuberous sclerosis complex

- Tumors of brain, heart, lungs, kidneys, eyes.
- Seizures, mental retardation.
- Autosomal dominant.

Angiofibromas (adenoma sebaceum)

Shagreen patch (lumbar or forehead)

Periungual fibromas

Nevus Depigmentosus

“Depigmentosus” is misnomer.
Usually hypopigmented, not nonpigmented.
Usually not ventral midline.
No white forelock or deafness. Nonsyndromic.

Nevus Depigmentosus

May be subtle and unnoticed at birth.
“Delayed birthmark”
No treatment. Fixed and stable – this is not vitiligo.

Congenital focal lack of pigment

- Piebaldism
- Tuberous sclerosis (ashleaf macules & confetti spots)
- Nevus depigmentosus

Refer if syndromic.
Our approach to hypopigmentation

Too light
Is it Hypo- vs De-pigmented?
Is it Congenital or Acquired?
Is it Generalized or Focal?

Acquired focal depigmentation

Could this be vitiligo?

Vitiligo
Jawaharlal Nehru, first Prime Minister of India, said that the three medical scourges of India are malaria, leprosy, and vitiligo.

Parsad D.
Quality of life in patients with vitiligo.
Health Qual Life Outcomes 2003;1:58-60

Acquired focal depigmentation

Vitiligo
- Acquired disorder with depigmented macules, often symmetric.
- Eyes and mouth, elbows, knees, digits, genital area.

Vitiligo
- Postinflammatory depigmentation
- Lichen sclerosus (LS&A)
- Chemical leukoderma (Scleroderma)
Vitiligo
- Well-circumscribed milky-white macules devoid of melanocytes.
- Risk for thyroiditis, alopecia areata, Addison’s disease, diabetes, & pernicious anemia.
- Can be a psychosocial disaster!

Segmental Vitiligo

Treatment

Topical steroids:
- Work best on sun-exposed surfaces; worst on acral skin.
- Usual risks; no guidelines on duration.
- Triamcinolone 0.1% on body daily, desonide on face daily.

Topical calcineurin inhibitors
- Pimecrolimus (Elidel) or tacrolimus (Protopic) twice daily.
- Black box warning; insurance difficulties.

Phototherapy (narrow band UVB) – probably most effective.

Camouflage (Lydia O’Leary, DermaBlend, CoverMark)

Perianal and vulvar vitiligo
Often difficult to distinguish from lichen sclerosus (LS&A). Refer.

Acquired focal depigmentation
Refer to Peds Derm if on genitals, accompanied by other autoimmune disorders, connective tissue disorders, or eye pain & photophobia.
**Acquired focal hypopigmentation**

Tinea versicolor
Pityriasis alba
Post-inflammatory hypopigmentation

Don’t refer – or send to a general dermatologist.

**Tinea versicolor**

- Ubiquitous fungus *Malassezia furfur* (*Pityrosporum* spp.)
- Clinically:
  - Versicolor: white, brown, red
  - Sharply demarcated hypo/hyperpigmented
  - Fine dusty scale
  - Trunk, neck, proximal arms

**Diagnosis:**
KOH prep. Don’t get fungal culture.
Gentle scraping.

**Treatment**

New diagnosis:
Topical antifungals for two weeks.
- clotrimazole & miconazole are least expensive.
- terbinafine (*Lamisil*) & nystatin are ineffective.

Selenium sulfide (*Selsun*) or ketoconazole shampoo weekly
- get wet, lather up, wait 10 minutes, rinse
- ketoconazole 1% is OTC; 2% is prescription
- ciclopirox (*Loprox*) is too expensive
- goal to reduce (inevitable) re-“infection”

**Pityriasis alba**

- Occasional feature of atopic dermatitis in children.
- Most common if medium pigmentation (Latinos, South Asians).
- Usually involves face.
- Post-inflammatory hypopigmentation.
- Resolves spontaneously.

**Treatment**

Inform patient:
- Pigmentation may take 2-4 months to restore.
  Continue using body shampoo monthly.

If condition persists or relapses:
- Oral agents: ketoconazole (dozens of regimens).
  Griseofulvin & terbinafine (*Lamisil*) don’t work.
**Treatment**
- Reassure.
- Explain that this is not vitiligo and it is not a fungus.
- Moisturize twice daily.
- No scrubbing of face.
- Hydrocortisone 1% or 2.5% daily for 2-3 weeks.
- Sunscreen to prevent accentuation of color difference

**Post-inflamatory hypopigmentation**
- Lip licker’s dermatitis
  - Hypopigmentation extends beyond corners of mouth
  - Treat with:
    - Education & reassurance.
    - Hydrocortisone 1% oint twice daily for 2 weeks then daily for 2 weeks then prn.
    - Barrier cream (zinc oxide) at bedtime.
    - Sunscreen.

**Post-steroid hypopigmentation?**
“That steroid cream bleached my kid.”
- Not likely if cortisones are lower strength.
- Often a no-win situation.
- Consider calcineurin inhibitors, eg tacrolimus/Protopic.

**Acquired focal hypopigmentation**
- Tinea versicolor
- Pityriasis alba
- Post-inflammatory hypopigmentation

**Subtle hypopigmentation in the tropics**
- Pityriasis alba
- Tinea versicolor
- Leprosy, BL

*Don't refer – or send to a general dermatologist.*
The most common presentation of early leprosy is a hypopigmented patch or plaque with ↓ sensation.

End-stage leprosy
- Madurosis
- Lagophthalmos
- Saddlenose deformity
- Resorption of digits
- Claw-hand deformity
- Ape-hand deformity

Acquired focal hypopigmentation
- Post-inflammatory hypopigmentation
- Tinea versicolor
- Pityriasis alba
- Leprosy (Hansen's disease)

Disorders that are “Too dark”
1. Too Dark
2. Congenital or Acquired
3. Generalized or Focal

Congenital generalized hyperpigmentation
- Congenital melanosis “carbon baby”
- Paternity issues

Congenital focal hyperpigmentation
- Mongolian spot & dermal melanosis
- Congenital melanocytic nevi
- Café-au-lait macules
- Physiologic variants

Don’t refer – or send to a general dermatologist.
If anesthetic, refer to me.

Whorls and swirls – refer ASAP.
**Mongolian Spots**
- Solitary or multiple.
- Few mm to many cm in diameter.
- Locations: sacrum, buttocks, back.
- Very common in Asians & African ethnicities.
- Many resolve over several years.
- May be mistaken for bruising (abuse).

**Congenital dermal focal melanosis**

**Nevus of Ota**
Melanocytes deep in the dermis.
May involve sclera & underlying mucosa.
Most common in Asian women.

**Giant congenital melanocytic nevus**

**Acquired generalized hyperpigmentation**
- Physiologic
- Suntanning
- Addisonian hyperpigmentation
- Genetic/metabolic disorders
- Diffuse melanosis (from melanoma)
- Drug-induced pigmentation
Addison's Syndromic

Acanthosis nigricans
- Velvety, grey-brown thickening.
- Posterior & lateral neckline, axillae, groin, inframammary.
- Obesity, metabolic syndrome

Obese teens with netlike discoloration of chest & back, resembling both acanthosis nigricans & tinea versicolor.

Minocycline 100mg twice daily for 6 weeks.

Acquired focal hyperpigmentation
- Acanthosis nigricans
- C.A.R.P.
- Post-inflammatory hyperpigmentation
- Becker's nevus
- Medication-induced hyperpigmentation
- Phytophotodermatitis
- Melasma
- Exogenous causes (tattoo etc.)

Control underlying condition. Time.

Confluent & Reticulated Papillomatosis of Gougerot & Carteaud

Post-inflammatory hyperpigmentation
**Post-inflammatory hyperpigmentation**

- Control acne.
- Apply tretinoin nightly.
- Apply hydroquinone 2% (Ambi) daily.
- Time. Sunscreen.

**Becker’s Nevus**

- Unilateral hyperpigmented and often hairy patch.
- Pectoral, scapular regions.
- Males > females.
- Onset in adolescence as a “delayed birthmark”

**Normal Pigmentary Variants**

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*Tuaregs. The legendary “Blue People of the Desert”*