My Fingers Are Blue: Benign or Worrisome?

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Objectives
• By 1pm, the learner (that’s you) will be able to...
  – appropriately triage a teenager with discolored fingers, including work up and referral
  – distinguish between benign digital color changes, and those more concerning for a larger disease process
  – list at least three treatment strategies for Raynauds Phenomenon
  – shamelessly admit that you’ve always secretly wanted to be a rheumatologist

Episodic Cold Blue Fingers in Adolescents
• Differential Diagnoses (most common to least common)
  – Benign Acrocyanosis
  – Raynaud’s Phenomenon
  – Chillblains = Pernio
  – (Frostbite)
  – (Erythromyalgia)

“Classic” Raynauds
• Well demarcated triphasic color changes
  – White (Vasospasm)
  – Blue/Gray, Purple, Black (O2 extraction, ischemia)
  – Red, warm, swollen, stinging (Reperfusion)
• In the hand, only involves the fingers, never thumb, and never proximal to the MCPs
• Episodic, triggered by cold, stress
• May be accompanied by pain, numbness, parathesias

Disclosures
• I’m a rheumatologist
• “I have no actual or potential conflict of interest in relation to this program.”
Raynauds

Benign Acrocyanosis

- More generalized blue/purple discoloration
- May involve fingers, thumbs, hand, up into wrist
- Episodes less distinct, and often persistent
- Triggers less apparent, although usually exacerbated by cold.
- Associated with slow capillary refill and sometimes hyperhidrosis or livedo reticularis (mottling)
- Other than feeling cold, typically asymptomatic

Benign Acrocyanosis

Pathophysiology

- Two hits
  - Peripheral vasospasm in response to cold
  - Dysregulation of determinants of vascular tone
    - Autonomic stimuli
    - Circulating catecholamines
    - Response characteristics of vascular smooth muscle
    - Injury
    - Vasoactive drugs
    - Autoimmune disease?

"Most things in medicine are very black and white... Everything that is gray is referred to rheumatology clinic."

-Anonymous

Now…

Acrocyanosis
Raynauds
Raynauds Phenomenon

• A vasospastic phenomenon
• Described by Raynaud in 1862
• 15% of children (self-reported)
• 80% female
• Mean age of onset 12.3 +/- 4.3 years
• Mean age of diagnosis 13.4 +/- 4 years

Raynauds Phenomenon

• Triggered by cold, stress
• May be associated with CNS stimulants and malnutrition (anorexia, bulimia)
• 46% monophasic, 32% biphasic, 22% triphasic
  – No difference in this ratio between primary and secondary RP

Raynauds Phenomenon

Primary Raynauds

• No associated systemic disease
• Most common variant in kids (69%) and adults (80%)

Secondary Raynauds

• More likely to have ischemic damage or nailfold capillary changes
• More likely associated with positive ANA
• Mean time between RP and development of disease 2-6 years

Raynauds Phenomenon

• Physical Exam
  – Normal (most common in primary)
  – Periungual capillary changes
  – Cuticle staining
RP – Physical Exam

- Tapering of fingers due to loss of subcutaneous pulp
- Finger pitting

Associated Diseases

- Scleroderma**
  - Sclerodactyly with shiny, thick or atrophic skin and flexion contractures
  - SOB, reflux, dysphagia, sub cutaneous calcinosis
  - Associated with +ANA, anti-SCL70 ab (dcSSC), anti-centromere ab (lcSSC, aka CREST)
- Dermatomyositis
  - Muscle weakness, fatigue, Gottron’s papules
- Mixed Connective Tissue Disease
- Lupus
- JIA/RA

Benign Acrocyanosis

- Slowed blood flow
- Common in adolescent girls
- Episodic or persistent
- Exacerbated by cold or stress
- Thought to be due to an exaggerated vasomotor response
- Usually asymptomatic
  - Associated with
    - CNS stimulants
    - malnutrition
    - smoking
  - Other signs of autonomic dysfunction
    - Hyperhidrosis, livedo reticularis, orthostatic hypotension

Benign Acrocyanosis

Approach to Color Change

- All patients:
  - Thorough history and physical exam looking for red flags
  - Consider CBC, CK, ANA, TSH, T4
- If no color changes during evaluation, ask for pictures and follow-up
- If any Red Flags, then refer to Rheumatology!

Cold Blue Fingers

- Red Flags
  - Pain
  - Numbness
  - Tri-phasic color changes
  - Sores or pits on finger tips
  - Well demarcated color changes involving fingers only
  - Swelling
  - Nodules
  - Periungual capillary changes
  - Cuticle staining
  - Skin tightening
  - Loss of normal range of motion of the fingers
  - Positive ANA
Cold Blue Fingers

- Reassuring
  - Excessive sweating
  - No white color change
  - Involves more than just digits
  - Normal physical exam
  - Normal Labs

Approach to Suspected Benign Acrocyanosis

- Reduce cold exposure
- Increase water intake (and salt)
- Trial off ADHD meds (?)
- Address malnutrition concerns
- Educate about Raynauds phenomenon, and follow-up once in 1-3 months for repeat evaluation
- Pictures!

Treatment of Symptomatic Raynauds

- Cold precautions
  - Central and peripheral (layers, hats, hand warmers)
- Discontinue ADHD meds
- Medical Therapy:
  - Calcium channel blockers
  - Nitro-paste
  - SSRIs
  - Viagra, Cealis
- Surgical Therapy:
  - Surgical sympathectomy

Refer to Rheumatology

- Any red flags
  - Symptomatic
  - Abnormal physical exam
  - Abnormal labs...
- When in doubt, call or refer!

Pernio (aka Chilblains)

- Localized inflammatory skin lesion resulting from abnormal response to cold (non-freezing cold)
- Fingers (proximal, dorsal), toes
- Single or multiple erythematous or violaceous edematous lesions

Pernio (aka Chilblains)

- Young women (thin body habitus)
- Rare
- Intense pain, itching, burning
- Typically resolves in 1-2 weeks, but may be chronic
- Can be associated with autoimmune disease (lupus)
- Treatment supportive, although nifedipine has been shown to be helpful
Frost Bite

- Literal freezing of tissue with resultant necrosis

Summary

- The most common digital color changes in adolescents are benign acrocyanosis and Raynaud’s phenomenon
- These two entities appear to fall on the same spectrum
- The majority of cases are benign, but maybe associated with or the initial presentation of autoimmune disease (secondary RP)
- Abnormal physical exam findings and abnormal labs are predictive of autoimmune disease (particularly scleroderma)

Summary

- Work up should include labs (consider CBC, CK, ANA, TSH, T4)
- Treatment strategies include cold avoidance, drug withdrawal, vasodilatory medications, and surgery
- Referral to rheumatology should be considered, especially if “red flags” present

References