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Definition: A Conflict of Interest may occur in situations where the personal and professional interests of individuals may have actual, potential or apparent influence over their judgment and actions.

I have received fees/honoraria (local PI of Clinical Trials) from the following sources: {Merck, GSK, BMS, Astra, others too over 30yrs}

Jean Cusson Acrosyndrome WS, Oct 29 2016
I intend to make therapeutic recommendations for medications that have not received regulatory approval for that matter, and I will make it clear.
ACROSYNDROMES

(*BLUE AND OTHER COLORS*)

CSIM WORKSHOP Oct 29th 2016

Jean Cusson MD PhD FRCPC
Internal Medicine
WHY

• CLINICAL CHALLENGE

• VERY CLINICAL, NOT RARE, SOMEWHAT OMITTED

• UNCONFORTABLE FOR THE CONSULTANT, LITTLE HELP FROM LAB AND USUAL SOURCES
WHY I INITIALLY SAID NO......

• MORE CLINICAL EXPERIENCE THAN DATA

• WILL NOT BE REVEALING SECRETLY KEPT HARD DATA

• NO INTELLIGENT DATA BASE NOR RELEVANT COHORT STUDY TO SHARE
HOW I PREPARED FOR THIS WORKSHOP

- TRIED TO PRESENT A CLINICAL APPROACH WHICH MADE SOME SENSE BASED ON HOW PATIENTS ARE ADDRESSED

- REVIEWED MY CONFIDENTIAL ANONYMOUS PHOTO DATABASE, AGREEING PATIENTS SEEN IN VASCULAR LAB OR ON WARDS

- REVIEWED MEDICAL LITERATURE FOR NEW INSIGHTS
Clinical Challenges

- Finding the right diagnosis WO help of lab/imaging
  - You wish to get support with lab testing etc, but you also want to avoid asking for unnecessary tests

- Not missing the occasional serious problem

- Prescribing the most appropriate empirical/symptomatic treatment
Additional difficulties

• Mechanism often speculative

• Natural evolution over time often unpredictable

• Treatment modalities supported by soft data
Objectives

1. Differentiate clinical conditions presenting as acrosyndromes
2. Recommend the investigations necessary for the pertinent acrosyndromes
3. Select appropriate therapeutic options and assure follow-up for the different acrosyndromes

....Not to review each disease included in the Diff Dx and their management
Otherwise healthy person with intermittent whitening of fingers
Most likely Raynaud's Disease

- starts around 15-25 yo usually, more often women
- Other cases in the family
- Livedo reticulosa on thighs
Most likely Raynaud's Disease

- starts around 15-25 yo usually, more often women
- Other cases in the family
- Livedo reticulosa on thighs

- No obvious sign of systemic disease
- No fingertip wounds-ulcers
- No sclerodactyilia
Fig. 3. The six capillaroscopic parameters: a) Irregularly enlarged capillary, b) Giant capillary, c) Micro hemorrhage, d) Loss of capillaries, e) Disorganization of the vascular array, f) Capillary ramifications.

Usefulness of nailfold videocapillaroscopy for systemic sclerosis

Satoshi Kubo* and Yoshiya Tanaka
Management, Raynaud's Disease

- Conservative approach if typical and mild-moderate
  - Reassurance + Explanations on cold-induced arterial spasm
  - Smoking counseling...
  - Lab unlikely to change management, baseline labs ok
  - Think twice before ordering immune tests (ANA...)
  - Consider capillaroscopy
  - Schedule next appointment early winter to reassess severity
  - Discuss the potential use of CCBs to alleviate symptoms

- Dihydropyridines (nifedipine, amlodipine)
  - Explain symptomatic treatment
  - Start with low dose, reassess in winter and as needed
  - Consider seasonal use before recommending yearly Tx
  - Offer annual follow-up in early winter or late fall
Consider alternative Dx vs Raynaud's Disease

- Raynaud phenomenon is unilateral
- Only 1-2 fingers involved and always the same 1-2 fingers
- Starts later in life (older than 35-40)
- Alteration of the skin of the finger-toes-tips (papules, ulcers)
- Sclerodactyilia
- Known rheumatological immune disease
- Positive capillaroscopy, Anticentromer AB
Otherwise healthy person with persistent redening of fingers
Otherwise healthy person with persistent redening of fingers
Benign Acrocyanosis

- Young patient, more females
- Patient sent for "Raynauds" but can be without Raynaud's
- Less prevalent than Raynaud's, but not rare
- Not painful
- Purple/dark pink/red color of dorsum and palm of fingers
Benign Acrocyanosis

• Can be worst with cold temperature, not a necessary element

• Increased sweating = not rare, sometimes the main problem

• No fingertip wounds

• Otherwise, person looks healthy
Benign Acrocyanosis: Management

• Conservative approach, reassure

• Difficult to discuss physiopathology as largely unknown

• Not a prelude to hand-finger damage

• No specific investigation

• No specific treatment
The painless sweathy red fingers (and toes)

- Analyse and treat as hyperhydrosis
- Good luck..., might have to refer for botulinum toxin
- Later... my (scientically unvalidated) trick.....
The painful red fingertips or toetips

- Cold-sensitive finger-toe tips w/wo Raynaud's
- Redish/purplish papules or/and inflamed look
Pernio
(Chilblain)
Pernio : traitement

- Pentoxifylline
- Hydroxychloroquine
- Mycophenolate mofetil
- Corticostéroïdes
- Nifedipine
- Diltiazem
- Amlodipine
- Prazosin
- Acide nicotinique
- Thymoxamine
- Vitamine D

- Vasodilatateur topiques
- Corticostéroïdes topiques
- Heparin ointment
- Calcium IV
- Vitamine K IV
- Ginger separated moxibustion
- Iontophorèse
- Photothérapie
- Sympathectomie
Chilblain

- Manage like Raynaud's
- Perhaps 1/6 will develop SLE
The burning/painful red-and-sweathy fingers/hands + toes/feet
The burning/painful red-and-sweathy fingers/hands + toes/feet

- Fortunately rare
- Erythromelalgia must be considered
- Myeloproliferative disorder could be underlying: check!
- Already long-standing when first internal medicine consultation, patient discouraged with high expectations and sought internet help
- Previous chronic use of cold/ice exposure to alleviate pain and chronic cold/ice exposure can possibly induce secondary vascular damage
Fig. 1 Schematic of voltage-gated sodium channel alpha subunit and localization of reported PN mutations. The alpha subunit consists of four domains (I-IV), each domain contains six helical transmembrane segments (S1-S6). The S4 segment of each domain contains positively charged amino acid residues.
The burning/painful red-and-sweathy fingers/hands + toes/feet

- Little help from pharmacological approaches
  - ASA for MPD-related erythromelagia
  - Many anecdotal reports, difficult to conduct trial...
  - Underlying pathophysiology speculative
  - Topical xylocaine has been recommended
White/Blue 3rd to 5th fingertips on only one side
White/Blue 3rd to 5th fingertips on only one side

- Suspect hypothenar hammer syndrome / occupational trauma of arteries when only 3rd, 4th, 5th fingers are showing discoloration, pain, cold-sensitivity, and a normal radial pulse, in a person otherwise in good health and history of potential repeated microtrauma to the hand.

- Consider angiogram +/- in situ arterial thrombolysis followed by ASA + dihydropyridine (+ empirical 4-week antico-LMWH) to reduce the subacute and chronic acral ischemia.

- Together with smoking cessation, Tx for DLPD, etc.
Anastomoses des artères du poignet, vue antérieure (gelatinatique).

The cold and painful blue hand
TIME TO WORRY
The benign pseudo-ischemic toe
Acrosyndromes, worry if:

- Fingertip ulcers-necrosis
- Sclerodactyilia
- Blue toes
- Livedo racemosa
- Systemic symptoms