Interactive Workshops

- **Practical Follow-up of DVT and PE patients**
- **Vascular acrosyndromes**
- **Unusual site thrombosis**
- **Women’s health issues**
WHAT IS IT?
WHAT IS IT?

1. Acrocyanosis
2. Raynaud’s phenomenon
3. Livedo
4. Chilblain
5. Acrorhigose
WHAT IS IT?

1. Acrocyanosis
2. Raynaud’s phenomenon
3. Livedo
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VASCULAR ACROSYNDROMES

- Any condition either primary or secondary, either vasospastic or obstructive, that induces disturbances in the cutaneous microcirculatory network of the extremities.

- In many ways it is a Flying Dutchman: every physician has heard about it, many have seen it, yet nobody knows for sure what it is... These acrosyndromes may overlap, making clinical discernment challenging...

- Many synonyms
  - acral vascular disorders, vasospastic disorders, Raynaud’s phenomenon and related syndromes...
### VASCULAR ACROSYNDROMES

<table>
<thead>
<tr>
<th></th>
<th>Permanent</th>
<th>Paroxysmal</th>
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<tr>
<td><strong>Vasoconstriction</strong></td>
<td>Acrorhigose</td>
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<td><strong>Vasodilatation</strong></td>
<td>Acrocholose</td>
<td>Erythromelalgia</td>
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<td>Red palms</td>
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<td>(Lane’s syndrome)</td>
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An easy checklist to help you categorize ...
Acrocyanosis

Hyperhidrosis

Blue toes
LIVEDO

Cholesterol emboli  Livedoid vasculitis  Buttock livedo CTD
CREST SYNDROME (LIMITED SCLERODERMA)
CREST SYNDROME (LIMITED SCLERODERMA)

Telangiectasia
VASCULAR ACROSYNDROMES

- Overall prevalence $\geq 10\%$
- Related conditions
  - Chilblains (Pernio)
  - Frostbite and non-freezing cold injuries
  - Paroxysmal finger hematoma
CHILBLAINS
DB 34-year old man

- Smoking a pack of cigarettes/day
- History
  - Unremarkable
- Problem
  - Since 2010, periodic sudden onset of ankle edema with purpura and inflammatory syndrome (about 4 episodes)
  - Spontaneous recovery with scarring brownish
  - Consultations in Dermatology, Internal Medicine, Rheumatology did not allow to identify the origin of the problem
  - Treatment with methylprednisolone 32 mg/d for 15 days was unsuccessful
  - The patient is referred for further advice
DB 34-year old man

- **Physical examination**
  - Good condition
  - 93 kg – 183 cm (BMI 27.8)
  - BP nl
  - Lower legs
    - Arterial and venous examination normal
    - Irregular brown dermatitis on both ankles
    - Light pink patches on back feet and hyperkeratosis in front of the knees
    - No true edema
DB 34-year old man
WHAT IS IT?

1. Chilblain
2. Frostbite
3. Livedo reticularis
4. Retiform Purpura
5. Stasis dermatitis
WHAT IS IT?

1. Chilblain
2. Frostbite
3. Livedo reticularis
4. Retiform Purpura
5. Stasis dermatitis
WHAT WILL YOU SUGGEST TO DO?

1 - Complete autoimmune biology

2 - Skin biopsy

3 - Antiphospholipid antibodies

4 - HIV

5 - IgA, IgG, IgM
Livedo: anatomy and physiology of the cutaneous microvascular system

Anything that increases the visibility of the venous plexus can result in a livedo appearance...

Capillary beds

Subpapillary venous plexus (periphery of capillary beds)

Ascending dermal arterioles (center of capillary beds)

Livedo: anatomy and physiology of the cutaneous microvascular system
LIVEDO RETICULARIS, RACEMOSA … ?

Depending on the authors: probably preferable to speak of «livedo»

**WHAT WILL YOU SUGGEST TO DO?**

1. Complete autoimmune biology | 0%
2. Skin biopsy | 0%
3. Antiphospholipid antibodies | 0%
4. HIV | 0%
5. IgA, IgG, IgM | 0%
DB  34-year old man

- **Investigations**
  - ANA, ANCA, APL, HIV –
  - IgA, IgG, IgM : nl
  - IgG 4  0.367 g/l (nl)
  - No thrombophilia
  - Homocystein 8.7 µM/l (nl)
  - CMV (IgG/IgM), EBV (IgG/IgM), Wright, Borrelia (IgG/IgM) : -
  - Toxoplasma –
  - Skin biopsy : pseudo-vasculitis (extravasation of red blood cells and capillary microthrombi in superficial dermis) with IF IgA, IgG and IgM
WHAT DOES THIS SUGGEST?

1 - Livedoid vasculitis

2 - Klinefelter syndrome

3 - Sneddon syndrome

4 - Psoriasis

5 - Another vasculitis
WHAT DOES THIS SUGGEST?

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REVERSIBLE RETIFORM PURPURA: A SIGN OF COCAINE USE

- Cocaine contaminated by levamisole may be a new cause of retiform purpura, livedo reticularis and, ultimately cutaneous necrosis (legs, ears). Mechanism unclear.
- Levamisole has been used for its immunomodulatory properties in various autoimmune diseases, nephrotic syndrome and neoplasms.
- Currently, it has been increasingly found as a contaminant and possible addictive to the North American cocaine supply since 2007.

C Han, G Sreenivasan, JP Dutz. CMAJ 2011;183:E597
REVERSIBLE RETIFORM PURPURA: A SIGN OF COCAINE USE

Box 1: Differential diagnosis of retiform purpura*

Systemic coagulopathy
- Protein C- and S-related (e.g., coumadin necrosis, purpura fulminans)
- Antiphospholipid antibody

Vascular coagulopathy
- Livedoid vasculopathy

Disorders related to cold
- Cryoglobulinemia, cryofibrinogenemia

Disorders of platelet plugging
- Heparin necrosis
- Thrombocytosis secondary to myeloproliferative disorders
- Paroxysmal nocturnal hemoglobinuria

Embolization or crystal deposition
- Cholesterol emboli
- Oxalate crystal deposition
- Others: marantic endocarditis, atrial myxoma

Infectious agents causing vascular occlusion
- Vessel-invasive fungi (e.g., *Aspergillus*, usually in immunocompromised patients)
- Ecthyma gangrenosum (often *Pseudomonas*)
- Disseminated strongyloidiasis

Other
- Cutaneous calciphylaxis
- Sickle cell disease, severe hemolytic anemias

*Partial list.

- 48-year old man
- Peace officer walking all day in Brussels (mean 20 km per day)
- Progressive painful blue-red lesion on the right heel making walking impossible
- The patient consulted various specialists (vascular surgeon, internist and finally a dermatologist). The patient is sent at our surgery by the dermatologist to exclude a vascular malformation
WHAT DIAGNOSIS DO YOU SUGGEST?

1. Venous malformation
2. Frostbite
3. Chilblains
4. Non-freezing cold injuries
5. Something else
WHAT DIAGNOSIS DO YOU SUGGEST?

1. Venous malformation
2. Frostbite
3. Chilblains
4. Non-freezing cold injuries
5. Something else
Before the start of complaints, the patient had bought new shoes. As he did not feel comfortable right, he was accustomed to walk on the spur of his shoe...

The lesion was due to local chronic abnormal friction since the patient was walking about 20 km per day.

The patient was asked changing his shoes...
Apr 1st, 2011 (2 months later)  Oct 11, 2011 (9 months later)
Acrosyndrome

Clinical case
Acrosyndrome

- Woman 47 years old.
- Unilateral acrosyndrome
- Suspected cutaneous lupus
- Recurrent episodes of swelling fingers, with blue-red dyscoloration for one year.
- No history of digital ulcers
Acrosyndrome: clinical exam

Cold hand, swelling fingers with severe erytho-cyanosis
Doppler: normal radial and cubital flow. But no flow in the palmar arch
No digital flow
Capillaroscopy: impossible
Biology

- Normal blood exam
- No anti-nuclear antibody
- No cryoglobulin, no cryofibrinogenemia
Arteriography
Arteriography with papaverine injection

30mg PAPaverine in loco

Total 60 mg PAPaverine
After discharge, home treatment with Nifedipine 30 mg and Minitran 5
Recurrence one month later

- Identical clinical problem: cold hand, no digital flow (thumb ok), little scares like frostbites external side of the forefinger
- Arteriography: new full vasospasm
- Transthoracic Echocardiography: pulmonary hypertension > 40 mm Hg before the PG infusion
- Pulmonary scintigraphy: normal
Acrosyndrome
Recurrence one month later

- Infusion prostaglandins during < 96 hours
  Prostin VR (Pg E1) iv 3 ng/kg/min 2 h thus 6 ng/kg/min 12 h, thus 12 ng/kg /min for 72 h.
Clinical improvement: color, no more pain and normal temperature. Scares are disappearing.

Control of the Doppler after 96 h of treatment: good flow of palmar arch and every digital flow until the distal pads.

Discharge with Nifedipine 30 mg and Minitran 5

Evolution