Acral vascular disorders

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Hôpital Erasme
Acral vascular disorders

- Any condition either primary or secondary, either vasospastic or obstructive, that induces disturbances in the cutaneous microcirculatory network of the extremities
- Overall prevalence $\geq 10\%$
- Related conditions
  - Chilblains (Pernio)
  - Frostbite and non-freezing cold injuries
  - Paroxysmal finger hematoma
## Acral vascular disorders

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- **Permanent**
- **Paroxystical**
Acral vascular disorders

**Vasoconstriction**
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  - Livedo
- Paroxystical
  - Raynaud’s phenomenon

**Vasodilation**
- Permanent
  - Achrocholose
  - Red palms
    - *(Lane’s syndrome)*
- Paroxystical
  - Erythromelalgia
Acrotriglhoe

- Functional symptoms manifested by a feeling of cold extremities without evidence of clinical disturbances and prevailing in young women
- Related to a sympathetic hypertonia without organic substrate
- Benign, sometimes associated with conditions such as hypothyroidism
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Acrocyanosis

**Definition**
- Unlike Raynaud phenomenon, patients with acrocyanosis do not experience a return to baseline circulation between vasospastic episodes: persistent cyanosis and coldness of the extremities, exacerbated by cold and reduced by warming.
- No pain
- No skin lesions
- Often associated with hyperhidrosis (hands, feet)

**Physiopathology**
- Vasomotor sympathetic hypertonia with capillary-venular stasis
Acrocyanosis

**Forms**
- **primary**: more likely to occur in females < 30 years
- **secondary**: number of conditions such as cryoglobulinemia, cold agglutinins, antiphospholipid antibodies...

**Incidence**: unknown
  - M ~ F

**Diagnosis**: history, clinical examination, blood sample, nailfold capillaroscopy

**Treatment**: ? - sympathectomy
  - SC Botulinum toxin
Acrocyanosis
Primary acrocyanosis

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Red palms

(Lane’s syndrome)
Livedo

- **Definition**
  Relatively common physical finding consisting of macular, violaceous, connecting rings that form a netlike pattern

- **Physiopathology**
  - Secondary to organic or functional disorders of the dermal arteries or arterioles: deoxygenation in the venous plexus (ex: vasospasm caused by cold or autonomic output, arterial embolism, increased blood viscosity)
  - Venodilation of vessels can also result in a livedo appearance (ex: altered autonomic nervous system function, circulating venodilators, local hypoxia)
Livedo: anatomy and physiology of the cutaneous microvascular system.

- Ascending dermal arterioles (center of capillary beds)
- Subpapillary venous plexus (periphery of capillary beds)
- Capillary beds

Anything that increases the visibility of the venous plexus can result in a livedo appearance.
Livedo reticularis, racemosa ... ?

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

Livedo

Forms

1. **Physiologic livedo (cutis marmorata)**
   - appears in response to cold exposure and resolves completely with rewarming
   - most commonly found in neonates and fair-skinned girls and women
   - more common in preterm infants, may fluctuate throughout life in Down syndrome and other genetic anomalies

2. **Primary livedo**
   - diagnosis of exclusion: appearance and resolution independent of ambient temperature and in the absence of underlying disease

3. **Idiopathic livedo**
   - persistent livedo without an underlying cause
   - most typically seen in women between ages 20 and 60

Livedo

4. Secondary livedo

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**Table 1. Causes of Livedo Reticularis**

<table>
<thead>
<tr>
<th>Arteriolar spasm</th>
<th>Response to cold</th>
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<tbody>
<tr>
<td></td>
<td>Drug-induced (ergotamine, cocaine)</td>
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**Vessel Inflammation**

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<thead>
<tr>
<th>Noninfectious (vasculitis)</th>
<th>Systemic polyarteritis nodosa</th>
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<tr>
<td></td>
<td>Cutaneous polyarteritis nodosa</td>
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<tr>
<td></td>
<td>Wegener granulomatosis</td>
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<tr>
<td></td>
<td>Churg–Strauss syndrome</td>
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<td></td>
<td>Microscopic polyangiitis</td>
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<tr>
<td></td>
<td>Drug-induced vasculitis (thiouracil)</td>
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<td></td>
<td>Vasculitis associated with SLE or RA</td>
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**Infectious**

| Lucio’s phenomenon |

**Vascular Obstruction Without Inflammation**

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<th>Thrombosis</th>
<th>Antiphospholipid syndrome</th>
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<td>Sneddon syndrome</td>
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<tr>
<td></td>
<td>Livedoid vasculopathy</td>
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<tr>
<td></td>
<td>SLE, RA</td>
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<td></td>
<td>Coumarin-induced necrosis</td>
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<td></td>
<td>Disseminated intravascular coagulation</td>
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<td></td>
<td>Dysproteinemia (type I cryoglobulinemia)</td>
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<td></td>
<td>Procoagulant genetic factors (factor V)</td>
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<td></td>
<td>Sickle cell anemia</td>
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<td>Drugs</td>
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<th>Atrial myxoma</th>
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<td>Cholesterol</td>
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<th>Vessel wall disorders</th>
<th>Calciniphaxis</th>
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<tr>
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<td>Hypercalciuria</td>
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**Table 2. Clinical Associations of Livedo Reticularis With Purpura, Necrosis, and/or Nodules**

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<tr>
<th>Isolated Livedo Reticularis (Caused by Spasm)</th>
<th>Livedo Reticularis with Retiform Purpura, Necrosis, Ulcers, Nodules (Caused by Vascular Obstruction)</th>
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Abbreviations: RA, rheumatoid arthritis; SLE, systemic lupus erythematosus.
Livedo (reticularis)
Cholesterol emboli
Livedoid vasculopathy
Calciphylaxis

Polyarteritis nodosa
Acral vascular disorders

**Vasoconstriction**

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- Paroxystical
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**Vasodilation**

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  - Achrocholose
  - Red palms

- Paroxystical
  - Erytromelalgia

*(Lane’s syndrome)*
Raynaud phenomenon
Raynaud phenomenon

1862 Maurice Raynaud

Associated complaints:
sensations of pins and needles
numbness and/or clumsiness of the hand
finger aching
pain or ulceration of the skin: only mildly severe RP

White attack
and/or
Blue attack

Erythema of reperfusion
Diagnosis of Raynaud phenomenon
Ask three screening questions

1. Are your fingers unusually sensitive to cold?
2. Do your fingers change color when they are exposed to cold temperatures?
3. Do they turn white, blue, or both?

Diagnosis of RP is confirmed by a positive response at all three questions

Idiopathic Raynaud Disease (Primary RP)
Primary RP (Idiopathic Raynaud Disease) and Secondary RP (Raynaud Syndrome)

- Current criteria for the diagnosis of Idiopathic RP
  - More common in women (4W/1M)
  - Onset generally between 15 and 30 years
  - Symmetric episodic attacks (thumb often spared)
  - No evidence of peripheral vascular disease
  - No tissue gangrene, digital pitting, or tissue injury
  - Normal nailfold capillaroscopy
  - Negative antinuclear antibody test and normal ESR
IIary Raynaud phenomenon
Most common associated disorders
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<th>Connective tissue diseases</th>
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<td>Vibration-induced (pneumatic drills…) Hypothenar hammer syndrome</td>
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<td>Neurological compression</td>
<td>Carpal tunnel syndrome</td>
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<td>Cold agglutinin disease, cryofibrinogenemia, cryoglobulinemia…</td>
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<tr>
<td>Drug-induced</td>
<td>Beta blockers, ergot, bleomycin, cisplatin, vinblastine, interferon α…</td>
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Environment

Jackson Tamper
Cryofibrinogenemia?

= a person whose plasma, but not serum, forms a cryoprecipitate
Raynaud - Diagnosis

- History
- Clinical examination
- ANA, ESR
- Nailfold capillaroscopy
- Cold water challenge: NOT recommended (inconsistent responses)

Allen test
Measurement of BP on both arms
Compression techniques for TOS
Phallen and Tinel tests
Allen test
Raynaud’s Disease

- **Anamnèse**
- **Examen clinique**
- **Biologie**
- **Capillaroscopie**
- **Tests au froid**

**Diagram:**
- Capillary microscopy
- LDF
- tcpO₂

**Arteriovenous shunt**
- 2–10%
- 90–98%
Raynaud - Treatment

- Protection against cold (heaters, adapted gloves...)
- Causal treatment if any
- Stop smoking
- Limited surgical treatment
- Drugs if necessary: calcium channel blockers
Acral vascular disorders

Vasoconstriction
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Vasodilation
- Achrocholose
- Red palms
  \((\text{Lane’s syndrome})\)

Permanent

Paroxystical
- Raynaud’s phenomenon
- Erythromelalgia
Acrocholose

- Sensation of burning of the extremities
- Substratum?
- Sometimes associated with hyperthyroidism
- Exclude an underlying peripheral neuropathy
## Acral vascular disorders

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Lane’s syndrome

Constitutional microcirculatory anomaly performing an array of palmar erythema
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Erythromelalgia

- Red, edematous, burning extremities that are warm to touch generally provoked by heat exposure (« reverse RP »)
- Improvement by cold or by elevating the affected part
- May be related to abnormal arachidonic acid metabolism in polycythemia vera platelets

Erythromelalgia

Forms

**Primary** : rare, often symmetrical, sometimes hereditary
usually < 40 y

**Secondary** : often > 40 y, sometimes asymmetrical

- Erythromelalgia and the associated symptom of acral paresthesias are considered to be pathognomonic microvascular thrombotic complications in *polycythemia vera* and *essential thrombocythemia*, and are associated with platelet count usually > 400,000/ mm$^3$.

- Symptoms respond dramatically to aspirin in low doses
Erythromelalgia - treatment

- Causal: hydroxyurea (PV, essential thrombocythemia)
- Aspirin
- NSAIDS
- Beta blockers
- Prophylaxis of risk situations (avoid heat...
Acral vascular disorders
Related conditions
Chilblains (Pernio)

- Inflammatory cutaneous lesions in patients exposed to nonfreezing weather (and damp conditions) during late winter or early spring (Nov-Apr)
- Pathogenesis unknown: vasculopathy?
- The lesions typically present as painful erythematous or purple lesions with associated swelling or itching (sometimes with cutaneous necrosis, ulceration or blistering) of the fingers or toes (or both): frequently misdiagnosed as vasculitis or embolic events

Chilblains (Pernio)

- Self-limiting process that usually resolves within 1-3 weeks
- Predominantly seen in females and associated with low BMI
- It is a clinical diagnosis and can be treated conservatively with excellent prognosis

Digital chilblains
Typical chilblains
Chilblains – treatment

- Nothing is really effective
- Protecting the affected area and avoiding further cold exposure
- Calcium channel blockers (felodipine) but there are no controlled studies to show if they are effective
Chilblain lupus

- Similar lesions as pernio, for which it is often mistaken, that may be found in systemic and cutaneous lupus erythematosus
- Initiated in cold, damp environments, but is less likely to regress during warmer months

Fisher DA, Everett MA. Arch Dermatol 1996;132:459-462
Chilblain lupus
Cold damages to the extremities

- **Frostbite**
  - True tissue freezing caused by heat loss sufficient to cause ice crystal formation in superficial or deep tissues: evidence of the role of thromboxanes and prostaglandins.
    
  
  - Enormous spectrum of injury: from minimal tissue loss with mild long term sequelae, to major necrosis of the distal limbs with subsequent major amputations and phantom limb pain.
    
Cold damages to the extremities

- Non-freezing cold injuries (NFCI)
  - Occur when tissue fluids do not freeze (usually at about – 0.5°C), but local temperatures remain low for several hours or days.
  - Probably often unreported and under-diagnosed: there is often a history of having been cold and wet for a sustained period and having been unable to dry out correctly. On rewarming, it becomes apparent that the limb has developed a localised sensory neuropathy.
  - Generally few objective clinical signs.
  - Severe cases: cold sensitisation so that individuals are unable to work outside with sometimes oedema, hyperhydrosis and/or chronic pain resembling causalgia.
Cold damages to the extremities

- **Treatment**
  - **Frostbite**
    - warm *quickly* (drink warm fluids, remove boots, remove wet gloves and socks...)
    - Aspirin 75 mg/d for antiplatelet effect
    - Ibuprofen 800 mg/d for anti-prostaglandin effect
    - Iloprost, Hyperbaric oxygen therapy, Surgery
  - **NFCI**
    - Unlike freezing cold injuries re-warm *slowly*
    - Standard conservative treatment as in frostbite
Frosbite after an Himalaya’s trekking (D 15)
Paroxysmal finger hematoma
(Achenbach syndrome)

- Sudden digital painful hematoma generally isolated to the volar aspect of the digit
- Pathogenesis unknown
  - Rupture of local superficial veins?
  - Reduction in digital blood flow found on angiography may play a role
- Spontaneous healing within a few days

Paroxysmal finger hematoma (Achenbach syndrome)