1. **Differs from lower limb ischaemia:**
   - Less common – only 5% of all limb ischaemia occur in the upper limb
   - Affects mostly the smaller more distal arteries – more than 90% small arterial disease vs. 10% large artery disease
   - Symptoms of progressive ischaemia usually present later due to excellent proximal collateral circulation in the neck and shoulder girdle as well as decreased metabolic requirements due to a smaller muscle mass

2. **Chronic upper limb ischaemia**

2.1. **Small vessel disease / distal arterial disease:** responsible for 90% of patients with upper limb ischaemia. In contrast with proximal large vessel disease where the pathology is mainly arterial obstruction or embolism, the symptoms in small vessel disease (distal disease) are caused by vasospasm and/or occlusion of the palmar and digital arteries and arterioles.
   - Raynaud’s syndrome (see chapter)
   - Connective tissue disease:
     - Scleroderma is the most common. Also SLE, RA, PAN, mixed and undifferentiated connective tissue diseases. Occlusion of the digital arteries are due to vasculitis. Present with symptoms that vary from Raynaud’s phenomenon to digital gangrene. These are systemic diseases with organ involvement (kidneys) and systemic symptoms.
     - CREST syndrome: Calcinosis, Raynaud’s, Esophageal dismotility, Sclerodactyly, Telangiectosis
   - Buerger’s disease:
     - Usually involves the lower limbs. Only 10% have isolated upper limb involvement (See section on chronic arterial insufficiency of the lower limbs)
   - Ischaemia related to occupational injury:
     - Repeated trauma to the digital arteries- initially spasm, later thrombosis and occlusion
       - Vibration injury – pneumatic tools (jack hammers, drills, chain saws, etc)
       - Hypothenar hammer syndrome: mechanics, carpenters. Injury to the ulnar artery in the hypothenar space → aneurysm, thrombosis and digital emboli
   - Various haematological conditions cause local thrombosis with digital ischaemia:
     - Cold agglutinins
     - Cryoglobulins
     - Polycythaemia vera
     - APLA
   - Calciphylaxis – renal failure, diabetes
2.2. **Large vessel involvement / proximal disease:** is the cause of 10% of chronic upper limb ischaemia

- **Atherosclerosis**
  - Most important cause for upper limb ischaemia in elderly patients
  - Involves the origin of the large vessels (a.brachiocephalic, a.subclavia) or more distal (distal a.subclavia, a.axillaris, a.brachialis)
  - Left a.subclavia more commonly involved than the right a.subclavia.
  - Presents with ischaemia (“claudication”) or embolic episodes

- **Aneurysms:**
  - Brachiocephalic, subclavian, axillary arteries
  - Causes ischaemia via thrombosis or distal emboli
  - Chronic embolisation leads to progressive occlusion of the distal vessels

- **Arteritis:**
  - Takayasu’s arteritis
  - Giantcell arteritis:
    - These are auto-immune inflammatory conditions of the aortic arch and outflow vessels to the head, neck and arms. Characterised by long segmental stenoses, occlusions or aneurysms (see later)

- **Arterial thoracic outlet syndrome:**
  - Compression of the subclavian artery usually in the presence of a cervical rib
  - Chronic trauma leads to:
    - Local injury $\rightarrow$ stenosis $\rightarrow$ thrombosis
    - Stenosis $\rightarrow$ post stenotic dilatation $\rightarrow$ aneurysm with distal embolisation (see chapter on thoracic outlet syndrome)

3. **Acute upper limb ischaemia**

Caused by:

3.1. **Embolic occlusion:**

- Responsible for 90% of acute upper limb ischaemia
- Source of emboli:
  - Cardiac:
    - 70% Of upper limb emboli originate in the heart
    - 20% Of all cardiac emboli lodge in the upper limb
  - Other sources of emboli:
    - Atherosclerotic plaque from the aortic arch or proximal large vessels
    - Aneurysms in the proximal larger vessels

Embolic occlusion of the brachial artery proximal to the origin of the arteria profunda brachii leads to limb threatening ischaemia due to insufficient collaterals.
3.2. Acute in situ thrombosis (acute on chronic occlusion)
- Secondary to atherosclerosis in the larger proximal vessels: arteria subclavia, axillaris
- Thrombosis of a subclavian artery aneurysm
- Arterial thoracic outlet compression, cervical rib:
  - Chronic injury of the subclavian artery → thrombosis

3.3. Trauma:

3.4. Secondary to pathology in the aortic arch
- Takayasu’s arteritis
- Dissection of the thoracic aorta:
  - Congenital arteriopathies (Marfan, Ehlers Danlos syndromes). Abnormal collagen and elastin cause weakening of the arterial wall with dissection of the thoracic aorta with subsequent occlusion of the arch vessels.

3.5. Acute digital ischaemia:
- Embolism
- Vasospasm
- Vibration trauma
- Hypothermic injury (“frostbite”)
- Ergotism
- Intra-arterial injections
- Cardiac emboli
- Trauma
- Iatrogenic injury
- Fibromuscular dysplasia
- Radiation arteritis
- Congenital arteriopathies – Ehlers Danlos, Marfan syndromes
- Behçet syndrome

4. Clinical appearance

4.1. Symptoms:
1. Acute ischaemia:
   - Typical symptoms of pain, pallor, poikilothermia, paresthesia, paralysis, etc

2. Chronic ischaemia:
   - Activity induced muscle exertion = “claudication“
   - Subclavian steal syndrome:
     - Seen with proximal subclavian artery occlusion
     - Collateral supply via vertebral artery
     - Neurological symptoms elicited by arm movements (see subclavian steal syndrome)
   - Digital ischaemia:
     - Raynaud’s syndrome and
     - Embolic phenomena (painful blue finger)
4.2. Clinical evaluation:

- **Examination of the hand:**
  - Signs of digital and palmar ischaemia
    - Pallor, cyanosis, digital embolic phenomena, ulcers, gangrene, splinter haemorrhages
  - Signs of underlying connective tissue disease e.g. scleroderma, rheumatoid arthritis

- **Examination of the pulses:**
  - Presence /absence of radial, ulnar, brachial and axillary pulses
  - Atrial fibrillation
  - Allan’s test: radial and ulnar arteries

- **Compare blood pressure in both arms:**
  - A difference of more than 20mmHg is significant
  - Indicative of stenosis or occlusion of the subclavian or axillary arteries
  - Left subclavian artery more often involved than right

- **Examination of the base of the neck/supraventricular fossa:**
  - Subclavian artery is displaced upwards via a cervical rib and is therefore palpable above the clavícula
  - Bruit/thrill due to compression or stenosis of the subclavian artery
  - Palpable cervical rib

- **Examination for thoracic outlet compression syndrome (see later)**

5. Special investigations

5.1. Laboratory tests:

- Tests with regards to underlying connective tissue disease or haematological conditions:
  - ESR, CRP, RF, ANA, cardiolipin, cryoglobulins, etc

- Atherosclerotic risk profile:
  - Lipogram, glucose, homocystein

5.2. X-rays:

- Soft tissue: hands
  - Calcinosis (CREST)
  - Diffuse calcification (calciphylaxis)

- Chest X-rays (CXR)
  - Abnormalities associated with thoracic outlet syndrome:
    - Cervical ribs, fractured clavicle, 1st rib anomalies
  - Lung fibrosis indicative of systemic sclerosis

5.3. Vascular examinations:

- Duplex doppler:
  - Flow and structural abnormalities from subclavian to distal arteries

- Arteriography:
  - Important for planning of treatment
  - Vascular tree should be seen from the aortic arch into the hand
  - Examination done with positional changes of the arm for thoracic outlet compression syndrome
• CTA and MRI:
  o Vascular imaging
  o Information regarding soft tissue abnormalities in the thoracic outlet

6. **Treatment**

6.1. **Treatment of acute upper limb ischaemia:**

6.1.1. Anticoagulation:
- Heparin: 70-100 units/kg IVI stat
- Antiplatelet therapy: 300mg Aspirin p.o. stat
- Analgesia
- Refer for vascularisation

6.1.2. Revascularisation
- Embolectomy:
  o More than 90% of cases of acute upper limb ischaemia are caused by embolic occlusion. Embolectomy can be performed under local anaesthetic

- Acute on chronic occlusion:
  o Thrombolytic therapy of value for the dissolution of thrombus in the peripheral outflow tract and can also identify underlying lesions for final treatment e.g. stenoses for PTA, bypass, etc

- Bypass surgery:
  o Depending on the location of the pathology (treatment the same as chronic upper limb ischaemia – see 6.4

6.2. **Treatment of acute vasospastic conditions:**
- Calcium channel blocker
- Nitroglycerine intra-arterial at the time of arteriography
- Intravenous prostaglandin I₂ (Prostacyclin/iloprost)

6.3. **Treatment of acute ischaemia due to inadvertant intra-arterial injections**
- Intra-arterial line or direct intra-arterial injection:
  Intra-arterial injection cause particle embolisation and changes in pH with severe vasospasm, platelet aggregation, secretion of thromboxane and other inflammatory mediators with eventual damage to arterial and venous endothelium. The end result is thrombosis which affects mostly the micro-circulation (pulses may be intact). Causes severe limb ischaemia and tissue loss.

Treatment:
1. Where the diagnosis is made immediately with the needle or intra-arterial line still in situ:
   o Irrigate with heparin-saline solution for dilution and anti-coagulant effect. Followed by an injection of a vasodilator (nitroglycerine/papaverine/lignocaine)
2. Systemic anticoagulation:
   Heparin bolus 100 U/kg intravenously stat, maintenance dosage to
   maintain a PTT 2-2.5 the control value
3. Low molecular weight dextran (dextran 40) continued infusion 16-20ml/hr
4. Dexamethasone 4mg IVI 6 hourly
5. Analgesia
6. Elevate the limb
7. Vasodilators:
   Prostacycline (iloprost)
8. Brachial plexus block may be of value
9. Thoracic sympathectomy sounds logical but not proven
10. Arterial bypass and fasciotomy only indicated with larger vessel occlusion
   where thrombosis occured at the injection site

6.4. Treatment of chronic upper limb ischaemia:
6.4.1. Large vessel disease:
   • Treat risk factors
   • Antiplatelet therapy
   • Revascularisation procedures:
     o The type of the procedure is determined by the pathology and
       varies from percutaneous transluminal angioplasty to major bypass
       procedures e.g. bypass from the aortic arch to the outflow vessels
       of the neck in case of Takayasu’s arteritis. The most common
       procedures however are:
         ▪ PTA and stenting
           • Indicated for stenoses at the origin of the subclavian
             artery
         ▪ Carotid-subclavian transposition or carotid-subclavian
           bypass
         ▪ Carotid-axillary bypass
         ▪ Carotid-brachial bypass

6.4.2. Small vessel disease:
   • Avoid causative factors e.g. cold exposure, vibration injury, etc
   • Treatment vasospastic conditions (see Raynaud’s disease)
   • Treatment of underlying connective tissue and haematological conditions
   • Local treatment of digital ischaemia and necrosis:
     o Conservative debridement of necrotic and infected tissue
     o Retain normal viable tissue and skin as far as possible
     o Avoid damaging blood supply
     o Appropriate wound cover (moist wound dressings)
     o Appropriate systemic antibiotics
     o Protect against further injury