OPEN ACCESS TEXTBOOK OF GENERAL SURGERY

UPPER LIMB ISCHAEMIA

J van Marle

INTRODUCTION

Upper limb ischaemia differs from lower limb ischaemia in various aspects:

- It is less common — only 5% of all limb ischaemia occur in the upper limb
- It affects mostly the smaller, more distal arteries. In the majority of patients with upper limb ischaemia the arteries distal to the elbow are affected. Small vessel disease is often associated with underlying systemic conditions which may pose a diagnostic challenge.
- Symptoms of progressive ischaemia usually present later due to excellent collateral circulation in the neck and around the shoulder as well as decreased metabolic requirements due to a smaller muscle mass.

Acute embolic occlusion of a proximal vessel, however, may cause limb threatening ischaemia whereas chronic occlusion of a large, proximal vessel may cause incapacitating symptoms with severe functional impairment.

ACUTE UPPER LIMB ISCHAEMIA

Etiology

Embolic arterial occlusion is responsible for 90% of acute upper limb ischaemia. The heart is the most important source of emboli: atrial fibrillation," mural thrombus, valvular lesions or ventricular aneurysm. 70% of upper limb emboli originate in the heart and 20% of all cardiac emboli lodge in the upper limb. Other sources of emboli include atherosclerotic plaque from the aortic arch or proximal large vessels, and aneurysms in the proximal 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 in that area.

Acute in situ thrombosis may occur secondary to atherosclerosis in the larger proximal vessels (acute on chronic occlusion of atherosclerotic plaque), thrombosis of a proximal vessel aneurysm or secondary to arterial thoracic outlet compression.
Trauma Blunt or penetrating trauma to the upper limb may cause acute arterial occlusion. Invasive monitoring (inra-arterial lines), diagnostic procedures, (arteriograms) as well as therapheutic procedures (percutaneous transluminal angioplasty and stenting) have become important causes for penetrating injury to the arteries.

Pathology in the aortic arch such as Takayasu's arteritis and acute dissection may cause acute occlusion of the origin of the arch vessels (brachiocephalic and left carotid and subclavian arteries) with subsequent acute upper limb ischaemia. Hypercoagulable states such as deficiencies of Anti- thrombin m, Proteins C and S, the presence of Antiphospholipid syndrome or paraneopiastic hypercoagulability, may cause acute thrombotics.

Clinical features

Typical symptoms of acute ischaemia include sudden onset of pain, pallor, poikilonthermia, paresthesia and paralysis. On examination the peripheral pulses are absent and the limb cold and pale. Muscle paralysis en paresthesia are manifestasions of muscle and nerve ischaemia and an indication for prompt intervention. As ischaemic time progresses, the skin may appear mottled which initinially blanches on digital pressure. Fixed blue staining of the skin, and tender, tense muscle compartments are signs of irreversible ischaemia and an unsalvageable limb.

TABLE 1 : CONDITIONS CAUSING ACUTE DIGITAL ISCAEMIA
• Embolism
• Vasospasm
• Vibration trauma
• Hypothermic injury (”frostbite“)
• Ergotism
• Intra-arterial injections
• Trauma
• Iatrogenic injury
• Fibromuscular dysplasia
• Radiation arteritis
• Congenital arteriopathies — Ehlers Danlos, Ivlarfan syndromes
• Bencet syndrome

Treatment of acute upper limb ischaemia

The patient who presents with acute upper limb ischaemia should be immediately anticoagulated using 70 — 100 units of unfractionated Heparin/kg body weight given intra venously. Appropriate analgesia should be given as required (titrated IV morphine). The patient is referred to a specialist vascular unit where the final treatment will depend on the exact cause of the ischaemia. Most emboli lodge at the site of the brachial bifurcation. Embolectomy can be performed under local anaesthesia. An S-shaped incision is made in the antecubital fossa, the brachial artery is exposed and a transverse arteriotomy made. The embolus is retrieved by passing an embolectomy catheter distally into the fore arm vessels and proximally into the brachial artery. Proximal patency is confirmed by the presence of strong pulsatile antegrade flow whereas distal patency is confirmed by intra-operative arteriography. The distal vessels are flushed with a heparin—saline solution and the
arteriotomy closed with interrupted 7/0 sutures.

ln patients who present with acute on chronic occlusion, catheter directed, intra- thrombus administration of a thrombolytic agent is of value for dissolution of thrombus to identify underlying lesions. Final treatment e.g. endovascular management or bypass surgery depends on the nature and location of the pathology.

Acute ischaemia clue to inadvertent intra-arterial injections

Intra-arterial injection, depending on the type of drug, may 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) with severe limb ischaemia and tissue loss.

Management: When the diagnosis is made immediately with the needle or intra-arterial line still in situ, irrigate with a heparin-saline solution for dilution and anti- coagulant effect. This is followed by injection of a vasodilator (nitroglycerine / papaverine / lignocaine). The patient is fully anti-coagulated with unfractionated heparin: a bolus of 100 U/kg is given intravenously stat, with a maintenance dosage to maintain an aPTT 2 ½ - 3 times the normal control value.

Low molecular weight dextran is given as a continuous infusion of 20ml/hr together with Dexamethasone 4mg IVl 6 hourly.Prostacyclin (Ilioprost) is a very potent vaso-dilator and may be of value. Appropriate analgesia is given and the limb is elevated to prevent further swelling. Brachial plexus block may be of value. Thoracic sympathectomy sounds logical but is not proven. Arterial bypass and fasciotomy is only indicated with large vessel occlusion where thrombosis occurred at the injection site

CHRONIC UPPER LIMB ISCHAEMIA: PROXIMAL L LARGE VESSEL DISEASE

Patients may present with activity induced muscle fatigue ("c|audication"), digital ischaemia secondary to distal embolization or Raynaud's phenomenon. Subclavian steal syndrome is seen with occlusion of the subclavian artery proximal to the origin of the verteberal artery. The vertebral artery becomes a major collateral to the arm with reversal of blood flow in the vertebral artery thereby "stealing" blood from the posterior cerebral circulation. Neurological symptoms of vertebro-bassilar insufficiency.are provoked by exercise of of ipsi-lateral arm.

Etiology

Atherosclerosis is an important cause for upper limb ischaemia in elderly patients. Atherosclerotic plaque causes stenosis or occlusion of the vessels and maybe a source for distal embolization. The origin of the brachiooephalic and subclavian arteries or more distal parts of the subclavian, axillaiy and brachial arteries are involved. The origin of the left subclavian artery is more commonly involved than the right.

Aneurysms of the proximal vessels (brachiocephalic, subclavian, axillary arteries) cause ischaemia due to thrombosis or distal emboli. Chronic embolisation leads to progressive occlusion of the distal vessels.

Arteritis: Takayasu's arteritis and giant cell arteritis are auto-immune inflammatory conditions of the aortic arch and outflow vessels to the head, neck and arms. It is characterised by long segmental stenoses, occlusions or aneurysms. (Fig 1a+b)

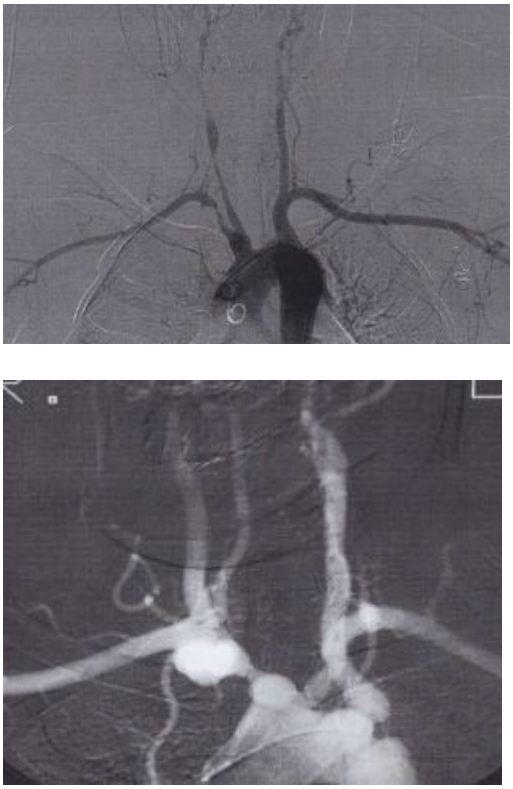

Fig 1a + 1b Takayashu's arteritis causing segmental stenosis (a) or aneurysm (b) in the aortic arch branch vessels

Arterial thoracic outlet compression syndrome: Arterial thoracic outlet syndrome is almost always associated with a cervical rib or anomalous 1st rib. The subclavian artery is compressed between the scalene muscles, cervical rib and 1st rib (Fig.2).

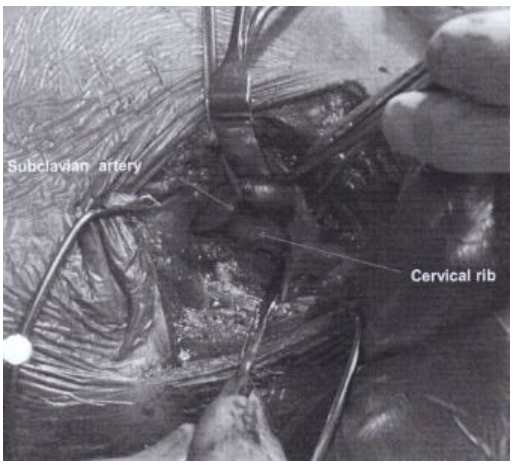

Fig 2 Arterial thoracic outlet compression syndrome

Chronic arterial trauma leads to fibrosis, focal stenosis, post stenotic dilatation and eventually aneurysm formation with thrombosis and/or distal embolization. (Fig. 3) Continuous embolization may lead to occlusion of the distal circulation and eventually an unsalvageable situation. The condition may present with symptoms of acute or chronic arterial insufficiency with activity induced fatigue (claudication), distal emboli (painful blue finger), ulceration and gangrene of the fingertips or unilateral Raynaud's. Clinical findings include a discrepancy in blood pressure between the two arms, a palpable cervical rib, a palpable subclavian artery in the supraclavicular space and a bruit and thrill over the subclavian artery.

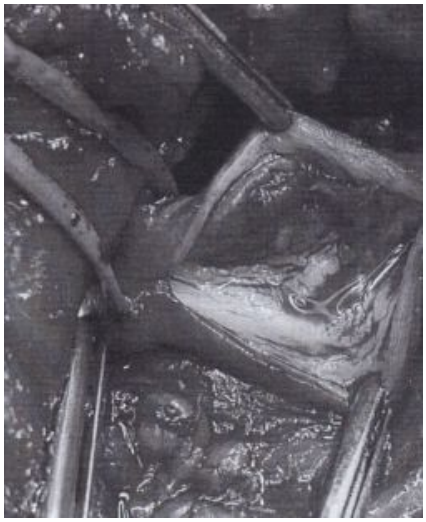

Fig 3 Structural damage to the subclavian artery caused by cervical rib compression

Clinical features

Patients may present with activity induced muscle fatigue ("claudication"), digital ischaemia, ulceration or gangrene secondary to distal embolization or Raynaud's phenomenon. (Fig 4+5) Clinical findings include difference in pulse status and blood pressure between the two arms, bruits and thrill in the supra-clavicular fossa indicating a proximal obstruction. A palpable cervical rib or subclavian artery ("high riding subclavian artery") is indicative of arterial thoracic outlet compression syndrome.

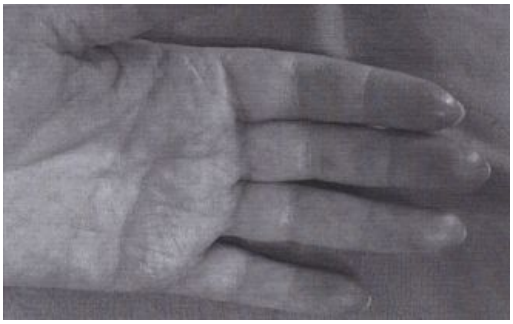

Fig 4 Cyanosis of fingers caused by occlusion og the subclavian artery

Fig 5 Peripheral embolisation from thoracic outlet compression (cervical rib) causing digital gangrene

Special Investigations

Standard x-rays of the thoracic outlet will demonstrate a cervical rib or other bony abnormalities causing thoracic outlet compression. (Fig 6) Arterial Duplex Doppler study will show abnormal flow and/or structural changes e.g. stenotic, post-stenotic dilatation and aneurysms. Reversal of blood flow in the stenotic vertebral artery (subclavian steal phenomenon) is demonstrated on an arterial Duplex Doppler or arteriography. (Fig 7).

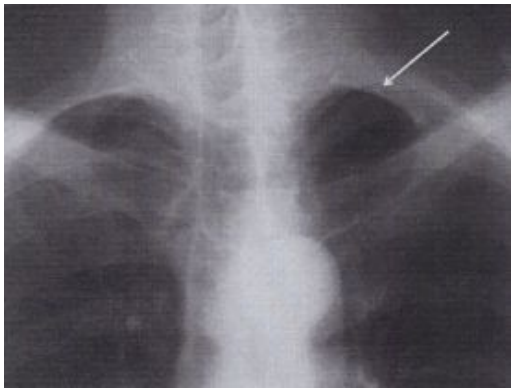

Fig 6 Cervical rib

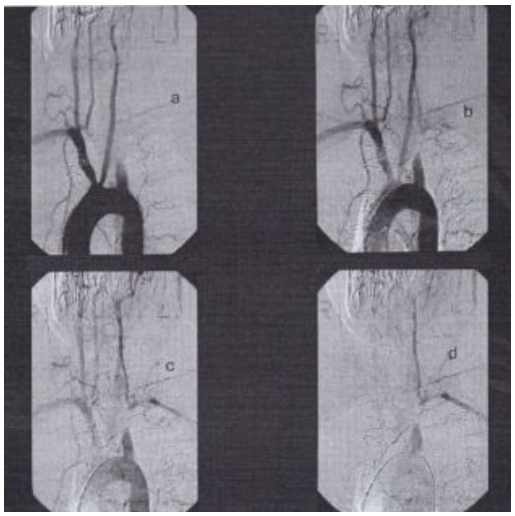

Fig 7 Subclavian steal phenomenon caused by obstruction of the proximal left subclavian artery (A) Reverasl of blood flow in the vertebral artery (b) provides collateral low to the distal sbclavian artrey (c+d)

Treatment of chronic upper limb ischaemia

Medical management consists of optimal management of risk factors including cessation of smoking, treatment of dysplipidemia, diabetes and hypertension and the use of anti-platelet therapy. Intervention is indicated in patients with severe repetitive symptoms that effect lifestyle and is determined .by the pathology and extent of the disease. Management varies from percutaneous transluminal angioplasty with or without stenting to major bypass procedures e.g. bypass from the aortic arch in the case of Takayasu's arteritisis (Fig 8), carotid —subclavian transposition or bypasses from the carotid artery to the subclavian, axillary or brachial arteries.

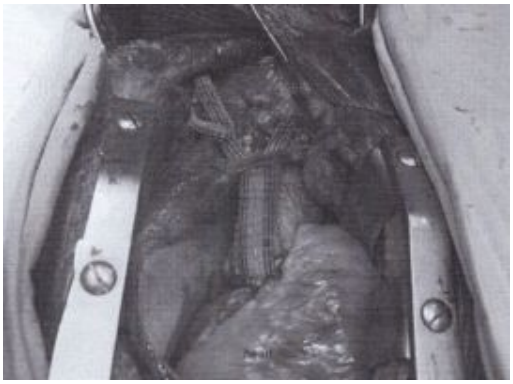

Fig 8 Graft placed on the ascending aorta (a) with brances leading to the right sbclavian (b), right common carotid (c) and left common carotid arteries (d) in a patient with Takayashu's arteritis

Treatment of arterial thoracic outlet compression consists of resection of the cervical rib and/or an anomalous 1st rib. The damaged segment of the subclavian artery is resected and replaced with a vein or prosthetic graft (Fig 9).

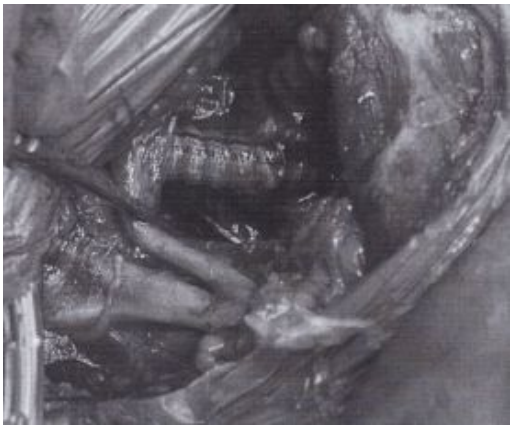

Fig 9 Segmental graft placement after removal of the cervical rib

SMALL VESSEL / DISTAL ARTERIAL DISEASE

The vast majority of upper limb ischaemic symptoms are caused by conditions affecting the the smaller

vessels in the hands and fingers. The symptoms in distal disease are mainly caused by vasospasm and/or occlusion of the smaller vessels.
There is often an underlying inflammatory condition present.

Etiology

Vasospasm is characterized by Raynaud's phenomenon (RP) which consists of episodic digital vasoconstriction (ischaemia) followed by reflex vasodilatation producing the characteristic tri-color response of white (vasoconstriction), blue (sluggish flow), and red (reflex hyperaemia). .Attacks are precipitated by exposure to cold or emotional stress. When vasospasm occurs as a primary condition and there is no identifiable cause or associated disorder, it is referred to as primary Raynaud's or Raynaud's disease (RD). In secondary Raynaud's or Raynaud's Syndrome (RS) there is a positive etiological agent or association with another disorder. Raynaud's is discussed in more detail in a separate section

Connective Tissue Disease (CTD): Occlusion of the digital arteries due to vasculitis occurs in a wide range of CTD's. Systemic sclerosis /scleroderma is the most common, but it is also seen in systemic lupus erythematosus, rheumatoid arthritis, polimiocytis / dermatomiocytis, mixed and undifferentiated connective tissue diseases.

Although these are systemic conditions, often with organ involvement, these patients may initially present with localized symptoms in the hands ranging from RP to digital gangrene. (Fig 10 a + b) Crest syndrome is an example of wide spread systemic involvement and consists of **c**alcinosis, **R**aynaud's phenomenon, **e**sophageal dismotility, **s**clerodactyly and **t**elangiectasis.

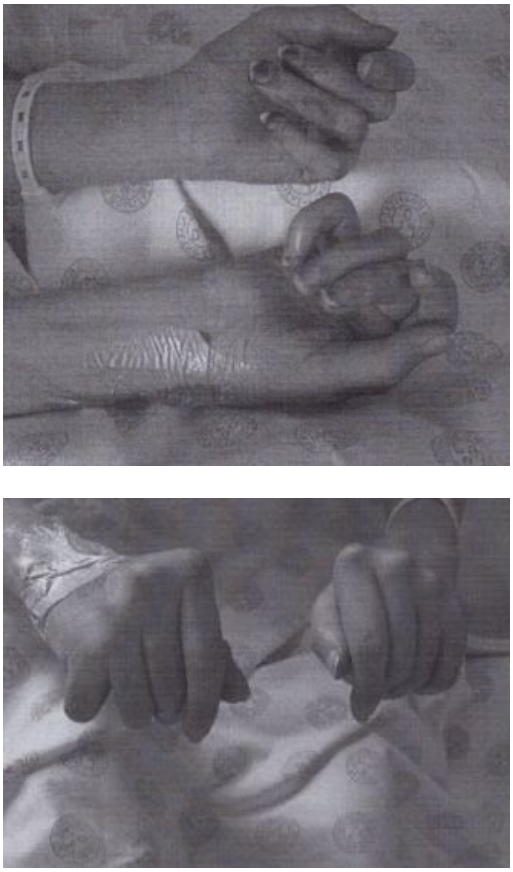

Fig 10 Digital gangrene (a) in a patient with Rheumatoid arteritis (b)

Buerger's disease is an inflammatory condition of the intermediate and small arteries which causes segmental thrombosis and occlusions. It usually involves the distal lower limb, but 30-40 % of patients may have upper and lower limb involvement and 10% of patients may present with isolated upper limb involvement. There is often associated venous involvement with migrating thrombophlebitis. It is seen almost exclusively in males and there is a strong association with smoking. Patients present with gangrene or non-healing ulcers of the fingers and toes. There are absent or reduced distal pulses and RP is often present.

Ischaemia related to occupational injury: Repetitive trauma to the arteries in the hand and fingers may initially cause arterial spasm and later structural damage to the arteries with thrombosis and occlusion. In the hypothenar hammer syndrome

repetitive trauma to the distal ulnar artery, as it lies anterior to the hamate bone, causes aneurysm formation with thrombosis and distal embolization into the fourth and fifth fingers. (Fig 11). Professional athletes involved in ball catching such as cricket, baseball, handball, netball, etc. may develop hand and digital ischaemia.

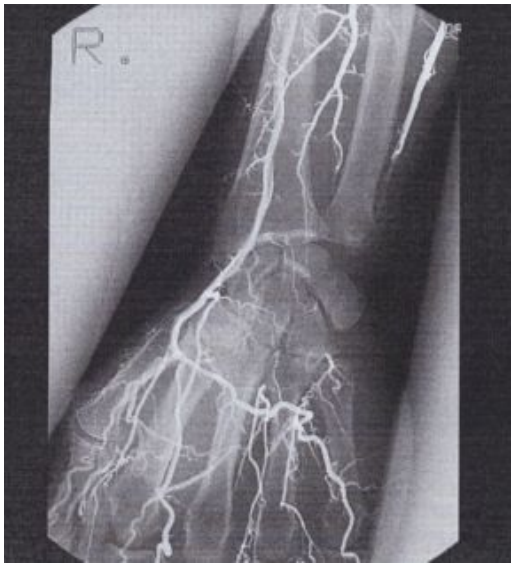

Fig 11 Thrombosis of the ulnar artery in a patient with hypotenar hammar syndrome

People working with pneumatic tools such as jackhammers, drills, chainsaws etc, may present with the so called "vibration-induced white finger". Constant cold exposure causes vasomotor changes in the hands, presenting as RP. Workers exposed to polyvinyl chloride may develop multiple stenosis and occlusions in the digital arteries.

Vasculitis is characterized by inflammation in the wall of small muscular arteries, capillaries and venules. Pathology ranges from partial obliteration of the lumen to necrosis of the blood vessel wall. There is usually an underlying systemic disease of which the manifestations of skin iscaemia include digital and nail fold infarcts, skin ulceration, vesicles and livido reticularis. This seen with conditions such as poly arteritis nodosa, Churg-Strauss disease, Behcet's disease and HIV vasculitis. Patients usually have intact peripheral pulses, and there are features of the underlying systemic disease. The diagnosis is confirmed with biopsy of the skin lesions.

Miscellaneous:Various hematological conditions may cause local thrombosis with digital ischaemia and gangrene. This includes cold agglutinins, cryoglobulins, Polycythaemia vera and antiphopholipid antibodies. Diabetics with renal failure may present with digital gangrene due to calciphylaxis

Diagnosis

History should be obtained about the typical clinical features, symptoms of possible underlying CTD's or other systemic disease, occupational exposure and medication. Patients usually present with bilateral involvement of the hands and multiple digits.

(Fig. 11a) A complete vascular examination should be performed with special attention to the hands and digits looking for signs of underlying CTD's. Special investigations are directed towards establishing or excluding underlying systemic conditions. This includes blood tests for CTD's. Enlarged nail fold capillaries can be seen with an ophthalmoscope and are present in patients with systemic sclerosis.

Management of small vessel disease

Contributing and causative factors such as cold exposure, vibration injury, smoking, etc should be avoided. The treatment of vasospastic conditions is discussed in the relevant section. For the majority of patients the emphasis of treatment is directed at the underlying systemic condition. A significant number of elderly patients will have concomitant atherosclerotic arterial disease which will require optimal medical treatment of diabetes, dyslipidemia, hypertension, etc. The

treatment of digital ischaemia and ulceration is given in table 2.

TABLE 2 : TREATMENT OF DIGITAL ULCERS
• Conservative debridement of necrotic and infected tissue • Retain normal, viable tissue and skin as far as possible • Avoid damaging blood supply • Appropriate wound cover: moist wound dressings • Treatment of vasospastic conditions • Appropriate systemic antibiotics where required • Protection against further injury

RAYNAUD'S DISEASE

Primary Raynaud's / Raynaud's disease: This is a common condition and typically occurs in young women. There is often a familial predisposition. lt is a reversible vasospastic condition provoked by cold exposure and/or emotional stress. The digital blood pressure is normal in between attacks. Ulceration and gangrene are rare. Histologically no obstructive arterial lesions are found.

Secondary Raynaud's / Raynaud's Syndrome: This is associated with a variety of underlying disorders or causative agents. See Table 3. lt should be considered where there is an early onset in children younger than 10 years of age or late onset in adults older than 30 years. Recurrent chill blains, digital ulcers or gangrene as well as perennial symptoms also suggest RS. These patients may have structural / irreversible arterial lesions with decreased digital blood pressure.

TABLE 3 : CONDITIONS ASSOCIATED WITH RAYNAULD'S SYMDROME	
Connective tissue disease	Systemic sclerosis, SLE ,RA ,mixed connective tissue diseases, dermatomysis
Arterial occlusive disease	Atherosclerosis, Buerger's disease, thoracic outlet compression syndrome (cervical rib)
Occupational trauma	Vibrating tools, cold environment expore to vinyl chloride
Drug therapy	Beta blockers, ergot preparations
Neurological conditions	Carpal tunnel syndrome, reflex sympathectic dystrophy
Blood dyscrasia	Hyper viscosity syndrome, cold agglutinins, myelo prolipherative disorders
Diverse conditions	Hypothyroidism, malignancy

Management

General measures: avoid cold exposure by wearing appropriate gloves, socks and insulated boots. Environmental temperature should be regulated. Smoking is contra-indicated and all causative agents and drugs should be withdrawn.

Appropriate occupational adjustments should be made.

Drug therapy: the calcium channel blocker, nifedipine, is the drug of choice. lt is a vasodilator and may cause hypotension, flushing and peripheral oedema. Initiate therapy with 5mg tds and increase dosage according to clinical response. Once the adequate dosage has been established a, slow release, long acting substitute may be given. Other vasodilators such as naftidrofuryl and cilostazol, are not available in the RSA. Prostaglandin PGI2 (prostacyclin) is a very potent vasodilator which is indicated for severe symptoms. lt is administered intra-venously in a high care setting

with with monitoring the blood pressure. Surgery: Sympathectomy has a limited effect in RS, but lumbar sympathectomy may be considered in patients with severe symptoms of RD of the feet. Thoracic sympathectomy has a high recurrence rate and is no longer considered for upper limb RD. Selective digital sympathectomy is under investigation.

ASSESSMENT OF THE PATIENT WITH UPPER LIIVIB ISCHAEMIA

HISTORY

Local symptoms: effort fatigue, rest pain, skin lesions, effect of cold exposure.

Systemic symptoms: underlying systemic disease, cardiac, renal, CTD's

Risk Factors: for atherosclerotic disease

Occupational environmental factors

EXAMINATION

- Examination of the hand:
  - Signs of digital and palmar ischaemia
    - Pallor, cyanosis, digital embolic phenomena, ulcers gangrene, splinter hemorrhages
  - 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
- Examination of the supraclavicular fossa:
  - Subclavian artery is displaced upwards by a cervical rib and is therefore palpable above the clavicle
  - Bruit/thrill due to compression or stenosis of the subclavian artery
  - Palpable cervical rib

The upper limb is examined in the neutral position as well as with arm elevated and shoulder abducted.

SPECIAL INVESTIGATIONS

Blood 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

X-rays:

- Soft tissue: hands
  - Calcinosis (CREST)
  - Diffuse calcification (calciphylaxis)
- Chest X-rays (CXR)
  - Abnormalities associated with thoracic outlet syndrome:
    - Cervical ribs, fractured clavicle, lst rib anomalies
  - Lung fibrosis indicative of systemic sclerosis

Vascular examinations:

- Arterial Duplex doppler:
  - Flow and structural abnormalities.
- Vascular Imaging:
  - 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

Digital subtraction angiography, CTA, MRA