# Critical ischaemia of the upper limb

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## Summary

Fifty-seven patients who presented to the Royal Surrey County Hospital, Guildford, with critical upper limb ischaemia between 1980 and 1989 were studied. Only 13 patients had emboli, while 23 presented with arteritis, seven involving large vessels and 16 with small vessel disease. Other causes included nine patients with trauma, six with atherosclerosis (of whom five were women), and four with vascular complications of thoracic outlet obstruction. Single examples of ischaemia due to radiation fibrosis and disseminated intravascular coagulation were also seen. Critical ischaemia of the upper limb remains an uncommon yet challenging problem. The review demonstrates that total limb arteriography should be performed in all patients, except the minority who present with direct arterial trauma or classical emboli.

## Introduction

Critical ischaemia presents in the upper limb much less commonly than in the lower limb and affects females more than males. Disability is common but limb loss is rare[1]. In this district, less than 2% of patients who required admission for symptoms of rest pain, gangrene or ulcer sepsis, were suffering from arm ischaemia. While atherosclerosis is the predominant pathology in the leg, this condition is responsible for only a minority of those with ischaemic arms, in whom arteritis, systemic embolism and trauma are more frequent. The differential diagnosis requires careful clinical evaluation and radiological assessment[2]. Since 1980, we have followed the policy of complete upper limb arteriography in all patients with critical ischaemia of the arm who do not also have the classical acute symptoms and cardiac signs of systemic embolism or direct arterial trauma. The management repertoire for the treatment of these patients is wide and the indication for vasoactive drugs, steroids, bypass, embolectomy, endarterectomy, thrombolysis, decompression of the thoracic outlet and sympathectomy by open or endoscopic methods are not well defined. This review was undertaken to audit current practice, to define more closely the indications for arteriography and surgery, and to examine the potential for the replacement of arteriography by non-invasive diagnostic methods.

## Patients and methods

Details of the presentation, investigation and management of 57 patients who required admission to the

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Royal Surrey County Hospital, Guildford, due to critical ischaemia of the upper limb, (defined as the presence of rest pain, ulcer sepsis or gangrene), were obtained from the admission summaries, copies of which were retained by the Vascular Firm. These were checked against Hospital Activity Analysis records, theatre and radiological records. Details are shown in Tables 1-6.

Over 9 years from 1980, 57 patients presented between the ages of 17 and 85 years; 43 were female. They were divided into diagnostic groups, arteritis (23), systemic embolism (13), atherosclerosis (6), trauma (9), thoracic outlet syndrome (4), and miscellaneous (2). In 12, the diagnosis of classical embolism from the heart was made clinically and no arteriography was performed. Similarly, with the patients who had obvious direct arterial trauma, operation was undertaken without previous arteriography. In all other patients, arteriography proved essential in diagnosis.

## Results

### Systemic embolism (Table 1)

Thirteen patients had emboli originating from the heart. They were aged between 57 and 85 years and 10 were female. Ten were in atrial fibrillation, one in congestive heart failure and one had a dilated ventricle with mural thrombus shown by echocardiography. None of these 12 underwent arteriography. The 13th patient was a 73-year-old woman in whom arteriography was undertaken because, in addition to her ischaemic right hand, she developed weakness in her well-perfused left arm. Arteriography showed extensive embolic occlusions in her right subclavian and common carotid arteries and she was managed by anticoagulation. Her right arm improved but the neurological weakness in her left arm remained unchanged.

### Arteritis (Table 2)

Of 23 patients, aged 23-75 years, with arteritis, all seven with large vessel disease were female, while the 16 digital vessel disease patients included four males. Of the seven with large vessel arteritis, five showed changes in the subclavian vessels, three bilaterally (one metachronous). The diagnosis was usually delayed and the condition mistaken for musculoskeletal pain in the early stages. Symptoms were usually aching in the shoulder girdle, particularly at night. Three of the five, however, also had arm claudication and one presented with cyanosis and digital ulceration. Only four of the seven had an ESR greater than 30 mm/h and one had hypochromic anaemia. In all patients, upper limb arteriography was diagnostic[3] and all were initially managed with steroids. Their systemic symptoms were improved

*Table 1. Systemic embolization*

| Diagnosis | | Arteriography | Treatment | |
|---|---|---|---|---|
| **Classical emboli** (3 male, 9 female) | | | | |
| Atrial fibrillation | 10 | None | Anticoagulation | 3 |
| Congestive cardiac failure | 1 | | Embolectomy | 10 |
| Ventricular aneurysm | 1 | | Brachial | 9 |
| | | | Brachial and subclavian | 1 |
| | | | Deaths | 2 |
| **Non-classical embolus** (1 female) | | | | |
| Stroke-in-evolution | | Embolic occlusion of subclavian and common carotid arteries | Anticoagulation Completed stroke | |

*Table 2. Arteritis*

| Diagnosis | | Arteriography | Treatment | |
|---|---|---|---|---|
| **Large vessel** (7 female) | | | | |
| Subclavian/axillary (bilateral 3) | 5 | Diagnostic | Steroids | 7 |
| Brachial | 1 | | Thrombectomy and biopsy | 2 |
| Ulnar | 1 | | Transaxillary sympathectomy | 1 |
| **Small vessel** (4 male, 12 female) | | | | |
| Idiopathic | 6 | Diagnostic of spastic and/or fixed obstruction | Transaxillary sympathectomy and debridement | 4 |
| Connective tissue disorders (scleroderma 4) | 10 | | Debridement | 4 |
| | | | Digital amputation | 1 |

within a few days (loss of shoulder girdle pain being dramatic), but in two patients continued distal ischaemic symptoms led to surgical exploration, thrombectomy and biopsy. Histology of the biopsies confirmed the diagnosis of arteritis. One of these patients subsequently also underwent transaxillary sympathectomy, the remaining six being controlled by steroids. None required surgical bypass.

*Case report*
A 43-year-old woman presented with claudication in her left arm. Her brachial pulse was absent and arteriography demonstrated multiple arteritic strictures of her left subclavian artery. Treatment with steroids improved her symptoms but, several weeks later as the dose was reduced, she developed identical symptoms in the right arm. Repeat arteriography then showed progressive arteritic stricturing of the right subclavian vessel. Her symptoms again responded to high-dose prednisolone (40 mg/day), and did not recur when the dose was reduced to a maintenance level of 5 mg/day.

Of the 16 patients who presented with digital vessel arteritis, six had severe Raynaud's syndrome without

systemic disease, and four of these were critical, thus justifying transaxillary sympathectomy; all four experienced some symptomatic benefit and, on prolonged follow-up, did not require any further treatment. Ten patients had connective tissue disorders, of which four had scleroderma and, of these, three needed surgical debridement. The remaining patients with connective tissue disorders were treated with steroids and two also with azathioprine. Five had a good response but the sixth required cyclophosphamide and transaxillary sympathectomy. Despite these treatments, gangrene progressed and digital amputation was undertaken in this patient.

*Trauma (Table 3)*
Only in this group of nine patients do males predominate (6 : 3). They are divided into two categories, direct arterial injury, indicating a lesion at the site of trauma, and remote arterial injury, where traction on a blood vessel induces an avulsion injury to the vessel or its branches at a site remote from the obvious

*Table 3. Trauma*

| Diagnosis | | Arteriography | Treatment |
|---|---|---|---|
| **Direct arterial trauma** (5 male, 2 female) | | | |
| Penetrating | 2 | None | Local repair |
| Blunt trauma | 1 | None | Vein graft |
| Sharp trauma | 1 | None | Local repair |
| Fractured clavicle | 2 | None | Local repair Conservative |
| Brachial catheter | 1 | Long occlusion | Thrombectomy, vein patch |
| **Remote arterial trauma** (1 male, 1 female) | | | |
| Supracondylar fracture | 1 | Traction injury at shoulder, good collaterals | Bony fixation, nerve repair, conservative |
| Dislocated shoulder | 1 | Traction injury, good collaterals | Conservative |

*Table 4. Atherosclerosis (one male, five female)*

| Diagnosis | | Arteriography | Treatment | |
|---|---|---|---|---|
| Subclavian | 3 | Occlusion | Axillo-axillary bypass | 2 |
| | | | Conservative | 1 |
| Axillary and subclavian | 1 | Stricture and ulceration | Endarterectomy (failed) and bypass | |
| Axillary | 1 | Stenosis | Endarterectomy and sympathectomy | |
| Generalized | 1 | Multiple blocks | None | |

bony or soft tissue damage. Remote arterial injury requires arteriographic diagnosis[4].

*Atherosclerosis (Table 4)*

Six patients had the history of acute-on-chronic ischaemia and the arteriographic signs were those of atherosclerosis. The patients were aged between 42 and 82 years, but only one was male. In three patients there was an occlusion of the subclavian artery, while in one the axillary artery only was involved. The sixth patient had diffuse changes throughout the upper limb.

*Thoracic outlet syndrome (Table 5)*

Four patients suffered chronic ischaemia as a result of thoracic outlet compression. All were women, from 20 to 71 years old. Two underwent thoracic outlet decompression alone, but the other two required multiple procedures. One had progressive ischaemia of her dominant right hand due to micro-emboli which had arisen on an ulcerated stricture in the subclavian artery. This was treated by excision of the compressing rib, endarterectomy and subsequent transaxillary sympathectomy. The fourth patient presented with acute arm ischaemia and atrial fibrillation: initially, a diagnosis of systemic embolism was made and embolectomy undertaken by a supra-condylar approach, without arteriography of the proximal vessels. The early result was satisfactory but the occlusion recurred one week later. Arteriography then showed arching of the artery around a cervical rib, with an intimal irregularity in that area. Thrombolysis, using a standard protocol[5], was unsuccessful and we would not generally advocate this management of occlusive disease in the upper limb, because of the risks of proximal pericatheter clot formation and cerebral embolization. Removal of the cervical rib and embolectomy brought little benefit and a third arteriogram was required to confirm that the proximal artery was now normal but there was persistent clot below the elbow. Finally, an embolectomy from the elbow gave a good result.

*Miscellaneous*

This group consisted of two female patients. The first underwent axillo-axillary bypass to treat fibrotic

ischaemia of the subclavian artery following radio-therapy for carcinoma of the breast, and the second developed widespread arterial thrombosis in her arm due to disseminated intravascular coagulation (DIC), secondary to post-cholecystectomy sepsis. In her case also, thrombolysis was attempted, without success, and due to occlusion of all potential collaterals, gangrene and amputation followed. Her rehabilitation, however, was excellent.

**Discussion**

Critical ischaemia is much less common in the upper limb than in the lower limb, accounting for only 5% of peripheral vascular cases[1], yet there are many causes. Diagnosis requires careful clinical and radiological assessment[1,2,6-10] but, unlike the lower limb, the role of non-invasive methods in diagnosis is not well established.

Clinical evaluation should include a history of accidental or occupational trauma; sudden ischaemia in the presence of atrial fibrillation suggests embolization. A significant number of patients with a connective tissue disorder will present with systemic symptoms. These may be local, for example aching in the shoulder girdle due to subclavian arteritis, or distal, due to the effects of ischaemia. Non-invasive laboratory studies are a useful adjunct to confirm clinical assessment, but arteriography[7] remains the 'gold standard'. Doppler ultrasound and digital arterial pressure recordings confirm the degree of ischaemia and are useful to monitor change, but they do not demonstrate the cause of the ischaemia. For this, total extremity arteriography, sometimes including magnification, subtraction and cryodynamic techniques[11], may all be useful. Porter and his colleagues have reported the use of digital plethysmography to replace arteriography 'unless a surgically correctable lesion is suspected'[12], but this begs the question of how that suspicion would be aroused unless the vascular anatomy can be shown.

Most emboli to the upper limb present with classical symptoms[13]. They account for only 10-15% of all peripheral emboli[14,15], but the outcome is better than in the lower limb[16]. Success in treatment depends on early diagnosis and anticoagulation[13]. Embolectomy

*Table 5. Thoracic outlet syndrome (four females)*

| Diagnosis | Arteriography | Treatment |
|---|---|---|
| Fibrous band | Compression | Division of band |
| Cervical rib, subclavian ulcer, distal emboli | Proximal lesion and distal occlusions | Excision of rib, endarterectomy and sympathectomy |
| Bilateral cervical ribs | Bilateral compression | Excision of ribs |
| Atrial fibrillation and cervical rib | Embolus, cervical rib stenosis and debris at elbow | Embolectomy, thrombolysis, resection of rib, subclavian and brachial embolectomy |

from an antecubital approach can be expected to produce a good result within the first 12 hours, as may be seen in these 12 patients, and there is a place for delayed surgery (up to five days) in those with adequate collateral vessels[17]. Anticoagulation is usually recommended to counter the risks of further emboli which may threaten up to a third of the patients[6] and is associated with a high mortality. Both early and late surgery have been shown to be superior to anticoagulation alone[18]. Arteriography was not required in these 12 classically-presenting patients but was undertaken in the patient who presented with a 'stroke in evolution', as well as an ischaemic arm; when clot had been demonstrated in both subclavian and common carotid arteries, she was managed by anticoagulation alone. While arteriography is not essential for the management of classical emboli, other symptoms require arteriographic diagnosis.

Several names have been given to vasculitis affecting large vessels[19], 'pulseless disease', Takayasu's disease or brachio-cephalic arteritis[20]. Both the aorta and its branches may be affected, including the renal artery[21]. The cause is unknown but an association with HLA-BW52 in Japan[22], and MB3-DR4 histocompatibility antigens in the USA[23], has been described. Large peripheral arteries may also be involved in patients with giant cell arteritis[24], which results in thickening due to proliferation of connective tissue, and an inflammatory cell infiltrate with multinucleate giant cells. The internal elastic lamina is disrupted and medial fibrosis and intimal damage lead to thrombosis, aneurysm and dissection. Symptoms result both from the local effects of inflammation, such as an aching shoulder, or from regional effects due to arm ischaemia. Angiographic signs include multiple long strictures[3] which may improve with steroids. Surgery is reserved for the relief of severe obstruction[25,26]. Surgical procedures for unrelieved obstruction have been reviewed recently[19], but the results are poor[20] when compared to those for occlusive disease due to atheroma[25]. The value of complete arteriography from the thoracic outlet to the finger-tips is emphasized, since lesions may be shown at several sites[20].

The poor long-term results of sympathectomy for vasospastic symptoms, with a relapse rate of 77% from 1 to 7 years[27,28], have restricted its use here to severe digital ischaemia in those in whom the obstruction has been fixed despite intra-arterial tolazoline. Porter[12] has suggested that all of these patients have occult systemic disease. Of the 16 patients in this series who suffered small vessel arteritis, ten were shown to have an underlying connective tissue disorder. Porter and his colleagues have also reviewed the medical treatment of Raynaud's syndrome, confirming that treatment is less effective when fixed digital occlusions are shown, for example, by cryodynamic imaging. Treatment with prostaglandins[29], the fibrinolytic-stimulating drug, stanazolol[30], and plasmaphoresis[31], have also been tried, but long-term benefit has not been proved. In this series, four of the six patients with idiopathic Raynaud's syndrome and fixed blocks responded well to transaxillary sympathectomy. Those with known underlying connective tissue disorders were also treated with steroids and azathioprine (2), with some improvement. One patient required digital amputation.

The management of direct trauma has been well documented[7,32], and five of the seven cases in this series were successfully managed by local repair. Techniques used were resection and end-to-end anastomosis, thrombectomy, vein patch and repair of intimal dissection. Only one patient with direct arterial trauma needed arteriography, due to the late presentation of a long occlusion after cardiac catheterization. He underwent thrombectomy, with repair of the damaged vessel wall, because of disabling arm claudication, which is known to follow such injuries in up to 70% of patients, particularly if wrist pressure indices are reduced to half on exercise[33,34]. Conservative treatment alone will, however, usually be sufficient to prevent limb loss[34]. Paraesthesiae may persist, despite arterial repair, due to ischaemic nerve damage[33].

The problem of arterial injury remote from the site of trauma[4] may easily be overlooked, even in comprehensive reviews, and presents significant difficulty in diagnosis. Of the two patients described here, one had a dislocated shoulder with a closed traction injury to the root of the neck. Arteriography showed that the subclavian artery was occluded but there was no bleeding and a good collateral circulation was seen; he was, therefore, managed conservatively. Such lesions may present late[35], and successful conservative treatment depends on safe, early heparinization[4]. In the other case, arteriography demonstrated the lesion remote from the site of bony fracture, thus avoiding fruitless blind exploration.

Atherosclerosis is generally more common in men[36], but in the upper limb more women than men are affected; in this series, five of the six patients were female. The surgical treatment is well described[19], including endarterectomy and bypass. Extra-thoracic bypass is preferred, since this is safer than intra-thoracic procedures[37]. Sympathectomy may be helpful when reconstruction is not practicable[19].

Vascular complications may develop in 10-20% of patients with thoracic outlet syndrome[19]. Compression of the subclavian artery between the first rib, the scalenus anterior muscle and a cervical rib may result in medial degeneration, with intimal hyperplasia, stenosis and post-stenotic dilatation. Distal embolization causes progressive digital ischaemia. Full arteriography should be performed and, if negative, should be repeated. This was necessary in one of these patients before the source of the emboli could be identified. On-table arteriography has been used, but the results should be interpreted with caution due to the frequency of spasm in the upper limb vessels. When the arteriogram is negative and there is a high index of suspicion[9], blind exploration has been advised, but surgical access is difficult and every effort must be made to show the lesion preoperatively. Treatment of the proximal lesion and resection of the band is helpful in preventing further emboli[9], but distal blocks are more difficult, as anastomosis between digital vessels is very limited. Sympathectomy may help to improve viability and control pain.

Two patients were placed in the miscellaneous group. The first showed the value of axillo-axillary bypass for the ischaemic post-radiation stricture, when this simple procedure can be carried out while avoiding the irradiated field[38]. The second patient, who developed thrombosis of the vessels of her left arm, due to DIC, was diagnosed late due to the effects of shock on all her limbs. It was only on resuscitation that the diagnosis of local thrombosis in one limb was made.

This review has confirmed that, with the exception of classical emboli and direct open trauma, all patients with critical arm ischaemia require total limb arteriography. Duplex ultrasound scanning and pressure studies are useful and might even replace arteriographic diagnosis in the leg[39]. However, difficulties in Doppler examination of upper limb vessels and the wide spectrum of pathology responsible for ischaemia of the arm, suggest that arteriography is unlikely to be replaced by non-invasive methods in the foreseeable future.

Fifty-seven patients with critical upper limb ischaemia were treated in a community hospital, serving a population of 200 000 people, over a period of nine years. Although uncommon, critical upper limb ischaemia remains a challenging problem with a diverse aetiology. To identify the cause and to plan and execute treatment, total limb arteriography should be performed in all patients except those who present with obvious direct trauma and classical systemic embolism. Digital subtraction angiography, pressure studies and Duplex scanning may be helpful but cannot replace arteriography.

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