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Microsurgical revascularisation of the hand in scleroderma

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Summary—Raynaud's phenomenon is a prominent manifestation of systemic sclerosis (scleroderma) affecting the hand. The resulting digital ischaemia may progress to digital tip ulceration or gangrene. Four patients with scleroderma, presenting with severe unremitting unilateral pain in the hand, were evaluated by arteriography and plethysmography. In addition to the usual changes of narrowing and occlusion of the digital arteries themselves, arteriography revealed more proximal occlusion of the radial and ulnar arteries at the wrist and the superficial palmar arch. Plethysmography confirmed virtual absence of pulsatile digital blood flow. Two patients underwent microsurgical reconstruction of the radial and ulnar arterial inflow into the hand and the superficial palmar arch using reversed interposition vein grafts, with immediate subjective resolution of their severe pain and rapid healing of the digital ulcers. Both remain pain free 1 year post-operatively, and pulse-volume recordings have confirmed objectively the restoration of pulsatile blood flow to the fingers.

In addition to visceral involvement of the gastrointestinal tract, kidneys, lungs and heart, systemic sclerosis (scleroderma) commonly affects the hands. Raynaud's phenomenon is usually the presenting symptom and may antedate other manifestations of this disease by several years. These paroxysmal attacks of blanching and cyanosis of the digits, followed by reactive hyperaemia, are usually induced by cold exposure or emotional upset. Progressive digital ischaemia may result in digital tip ulceration and occasionally frank gangrene. These vascular lesions are more frequent in the CREST syndrome variant of systemic sclerosis—calcinosis, Raynaud's phenomenon, (o)esophageal dysfunction, sclerodactyly and telangiectasia—than in other subtypes (Medsger, 1985).

An exaggerated sympathetic vasoconstrictive response to cold was initially proposed to explain the pathophysiology of Raynaud's phenomenon in scleroderma. This theory provided the rationale for medical management of Raynaud's phenomenon by various drugs which reduce sympathetic activity and the surgical operations of cervical sympathectomy and digital artery sympathectomy (Baddley, 1965; Flatt, 1980). According to the alternative theory, Raynaud's phenomenon is due to a normal vasoconstrictive response to a cold stimulus acting upon structurally abnormal arteries in the hand. This concept is supported by angiographic and histologic evidence of multiple small areas of narrowing and occlusion of the proper digital arteries.

Four patients with the CREST variant of scleroderma presented with unilateral increasing severity of Raynaud's phenomenon culminating in incapacitating pain in the hand. Digital plethysmography revealed absence of pulsatile flow in the fingers of the involved hand and arteriography confirmed proximal occlusion of the arterial inflow into the hand through the radial and ulnar arteries and superficial palmar arch. Since Kristensen (1982) has shown that blood flow within the fingers is directly related to the arterial inflow perfusion pressure, it seemed reasonable to attempt to alleviate these patients' symptoms by microsurgical reconstruction of the radial and ulnar arteries and superfical palmar arch to restore a more normal inflow pressure to the common digital arteries of the fingers. This was performed in two patients, one patient declined operative intervention and the medical condition of a fourth patient was considered a contraindication to operation.

Case 1

This 65-year-old woman had a 4-year history of the CREST variant of scleroderma with her predominant symptom being Raynaud's phenomenon. She was a non-smoker and her symptoms initially responded to nifedipine. However, 3½ months prior to admission she

developed constant excruciating pain in her right index and middle fingers and small tip ulcers in the same two fingers. Electromyography and nerve conduction studies of the median nerve were normal. Intra-arterial phentolamine provided temporary relief from the pain which was controlled only by 4 mg Dilaudid every 4 hours. Examination revealed that the right hand was cooler than the left hand and that the right ulnar artery was not palpable. The Allen test suggested occlusion of both the radial and the ulnar arteries. Pulse volume recordings showed complete absence of pulsatile flow in the fingers of the right hand compared with satisfactory pulsatile flow in the fingers of the left hand (Fig. 1). Arteriography

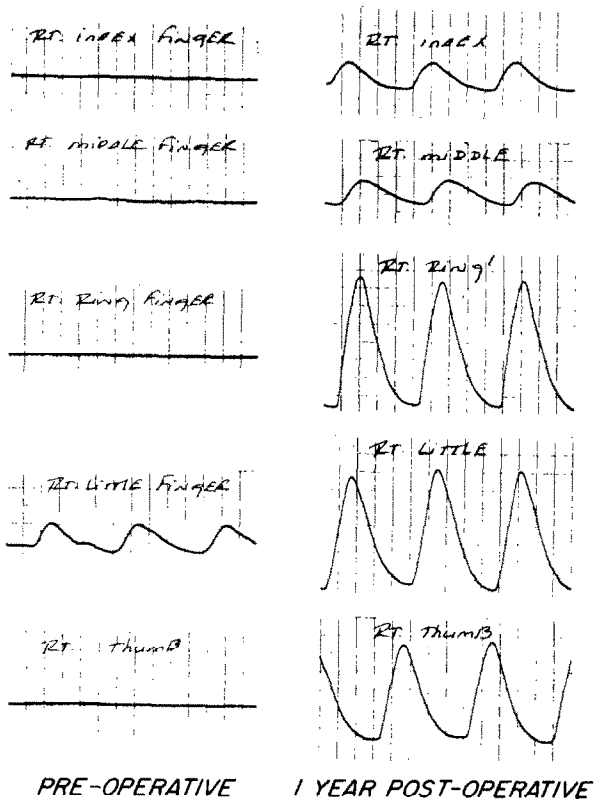

Fig. 1

Figure 1—Preoperative and 1 year postoperative digital plethysmography of the right hand of patient 1.

demonstrated occlusion of the radial artery at the level of the wrist and an extensive occlusion of the distal ulnar artery and superficial palmar arch (Fig. 2). There were characteristic small areas of narrowing and occlusion of the proper digital arteries and absent radial digital arteries to the right index and middle fingers.

At operation the occluded segment of the radial artery (Fig. 3) was replaced with a 6 cm vein graft from the distal radial artery to the bifurcation into the radial digital artery to the index finger and the princeps pollicis artery. A 13.5 cm vein graft was used to reconstruct the distal ulnar artery and the superficial palmar arch. The common digital arteries to the third and fourth web spaces were anastomosed end-to-side and the digital artery to the second web space anastomosed end-to-end to the distal end of the vein graft, thereby reconstituting the superficial palmar arch (Fig. 4). Histological examination of the superficial palmar arch revealed almost complete occlusion due to intimal hyperplasia characteristic of scleroderma (Fig. 5). Postoperatively the patient was treated with low molecular weight dextran for 5 days. Her severe pain was relieved almost immediately and the ulcers over the tips of the index and middle fingers healed within 4 weeks. Pulse volume recordings obtained 2 weeks postoperatively showed restoration of pulsatile flow in all the digits of the right hand. Symptomatic relief has been maintained 1 year postoperatively and pulse volume recordings at 1 year continue to demonstrate pulsatile flow in all digits (Fig. 1).

Case 2

This 68-year-old woman had suffered frequent episodes of Raynaud's phenomenon in both her hands for 5 years and had been diagnosed as having the CREST variant of scleroderma. Over the previous year, the Raynaud's phenomenon had become more frequent and severe in her right hand and two months prior to admission she began to complain of severe pain in the right index finger and ulceration of the tip of the finger. She was initially treated with reserpine 0.25 mg daily, nifedipine 30 mg daily and prednisone 20 mg daily. Despite this management her pain increased in severity and now involved the middle finger and the hypothenar eminence. Examination revealed an absent ulnar artery pulse at the right wrist and very sluggish filling of the hand from the radial artery on Allen testing. Plethysmography demonstrated absence of pulsatile flow and arteriography showed occlusion of the distal ulnar artery and superficial palmar arch with no filling of the radial digital artery to the index finger (Fig. 6). Nifedipine was increased to 90 mg daily and two cervical sympathetic blocks were performed, producing only a 2° F temperature rise in the index finger. Intra-arterial phentolamine and intravenous Bier block reserpine provided temporary symptomatic relief. Since the pain in her hand was uncontrollable even with frequent intramuscular morphine surgical intervention was felt to be indicated.

At operation a 9 cm vein graft was used to replace the distal ulnar artery and superficial palmar arch. The distal end of the vein graft was anastomosed end-to-end to the ulnar digital artery of the index finger and the common digital arteries to the third and fourth web spaces were anastomosed end-to-side to the vein graft. Low molecular weight dextran was administered postoperatively for 5 days. Her pain was relieved immediately postoperatively,

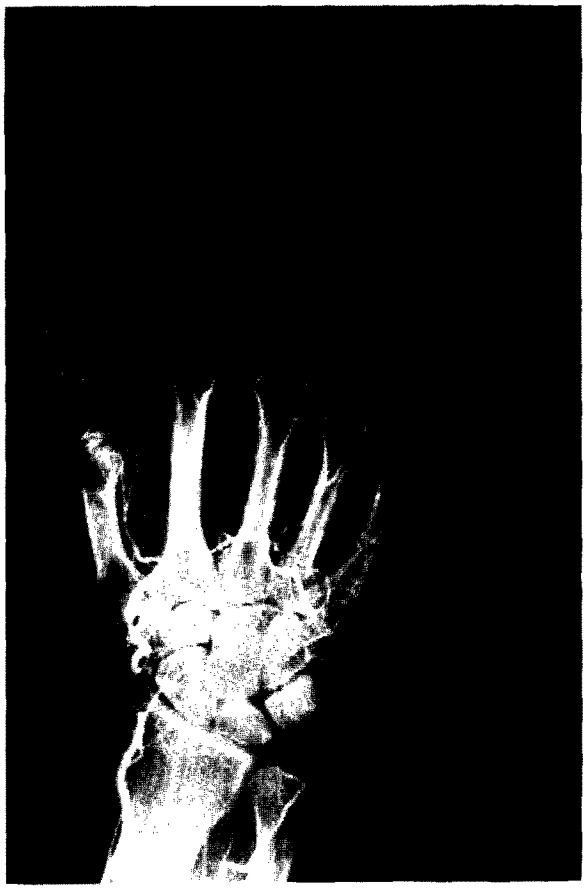

Fig. 2

Fig. 3

Figure 2—Arteriogram showing occlusion of the radial artery at the level of the wrist and extensive occlusion of the distal ulnar artery and superficial palmar arch. Figure 3—Intraoperative photograph showing occlusion of the distal radial and ulnar arteries (arrows indicate the extent of the occlusion).

the hand become warmer and the ulcer over the index finger healed within 3 weeks. Pulse volume recordings obtained one year postoperatively have confirmed pulsatile flow in all the fingers of the right hand with blood pressures comparable to those in her left hand.

Discussion

Patients with systemic sclerosis (scleroderma) may be divided into two groups—those with diffuse scleroderma in which there is bilateral symmetrical diffuse involvement of the skin of the trunk, face and proximal as well as distal parts of the extremities, and those with the CREST variant—calcinosis, Raynaud's phenomenon, (o)esophageal dysfunction, sclerodactyly and telangiectasia (Medsger, 1985). Raynaud's phenomenon is usually the presenting symptom and uniformly occurs in patients with the CREST syndrome. There is usually a prolonged delay from the onset of Raynaud's phenomenon to the appearance of visceral involvement (Porter *et al.*, 1976).

Severe Raynaud's phenomenon with progression to digital ulceration may be disabling. Multiple small areas of narrowing or occlusion of the proper digital arteries are seen on angiography of patients with scleroderma (Dabich *et al*, 1972). The common digital arteries are less frequently involved and the radial artery relatively spared. However, narrowing or occlusion of the ulnar artery at the wrist has been reported in almost 50% of patients and total obstruction of the superficial palmar arch in 10% of

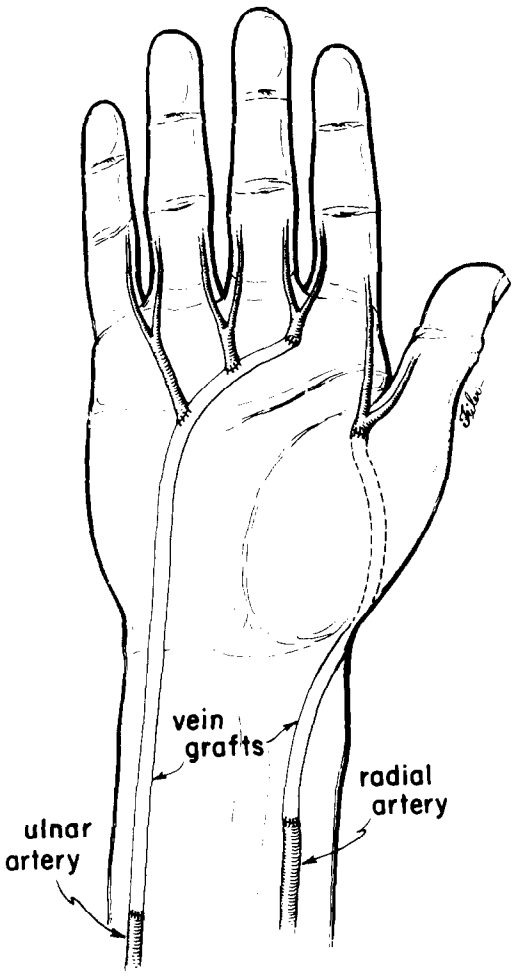

Fig. 4

Figure 4 –Diagrammatic representation of the microvascular anastomoses of the digital arteries to the interposition vein grafts.

patients. Rodnan *et al.* (1980) examined the digital arteries obtained at post-mortem from patients with scleroderma and Raynaud's phenomenon. Most specimens showed marked narrowing of the arterial lumen due to intimal hyperplasia and fibrosis. Fibrosis of the adventitia and telangiectasis of the vasa-vasorum were also documented. Since thickening of the intima produces a reduction in the lumen of the artery and because blood flow is inversely proportional to the fourth power of the radius of the lumen, such arterial narrowing would cause a significant reduction in blood flow to the fingers. Kristensen (1982) has confirmed that blood flow is markedly diminished in the fingers of patients with scleroderma, and because the pressure decrement is greatest between the wrist and the proximal phalanges, he postulated an increased resistance to flow within the palmar arch. Consequently even a minimal increase in the vasoconstrictive response to a cold stimulus would produce further significant functional narrowing in an artery already partially occluded by structural changes.

In the cases presented here, increasing severity of Raynaud's phenomenon and progression to incapacitating ischaemic pain may be attributable to markedly diminished arterial inflow into the palmar arch. Restoration of arterial inflow into the palmar arch was performed in the hope that this would then increase the perfusion pressure into the

Fig. 5

Figure 5 –Cross-section of the superficial palmar arch stained with haematoxylin and eosin showing marked intimal hyperplasia with virtual occlusion of the lumen.

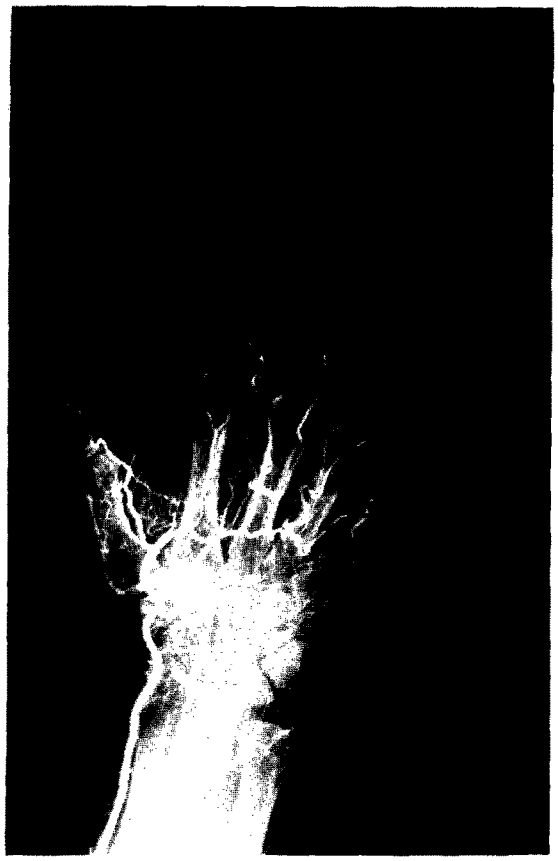

Fig. 6

Figure 6—Arteriogram from patient 2 demonstrating occlusion of the distal ulnar artery and superficial palmar arch with an absent radial digital artery to the index finger.

digits, even though the multiple small areas of narrowing and occlusion of the proper digital arteries remain. Improved digital blood flow was confirmed on pulse volume recordings obtained immediately postoperatively and maintained one year later.

Persuading patients to stop smoking and minimise cold exposure by wearing gloves, hats and scarves is important in the conservative management of Raynaud's phenomenon and scleroderma. Several drugs which reduce sympathetic activity have been advocated. The clinical effectiveness of intra-arterial injections of reserpine remains controversial (Siegel and Fries, 1974) but Taylor *et al.* (1982) have documented angiographic and plethysmographic improvement in patients with Raynaud's phenomenon following administration of reserpine intravenously in a Bier block. Nifedipine, a calcium channel blocking agent that produces vasodilatation, and more recently, ketanserin, a serotonin receptor blocker, have both been reported to increase digital blood flow and heal digital ulcers in patients with Raynaud's phenomenon (Winston *et al.*, 1983; Roald and Seem, 1984).

Cervical sympathectomy has been proposed for the surgical management of Raynaud's phenomenon, but the results have been equivocal (Baddeley, 1965). Additional contributions to the sympathetic trunks of the upper limb from the sinuvertebral nerve, the carotid plexus and the nerve of Kuntz may explain these disappointing results. Flatt (1980) therefore advocated a distal sympathectomy in which the adventitia is removed circumferentially from the proper digital arteries. Wilgis (1981) reported the effectiveness of a modification of this technique—removing adventitia from both the common and the proper digital arteries, in healing digital ulcers in patients with chronic digital ischaemia. Microsurgical reconstruction of the radial and ulnar arteries and superficial palmar arch to restore arterial inflow into the hand of patients with chronic digital ischaemia was also introduced by Wilgis (1981) in his paper.

The conclusion to be drawn from these patients with scleroderma and progressively severe unilateral Raynaud's phenomenon is that such patients should be investigated by plethysmography and arteriography to exclude more proximal occclusion of the arterial inflow into the hand. Should large vessel occlusion be documented, it may be possible for the microsurgeon to improve dramatically the digital blood flow by revascularisation of the superficial and deep palmar arches. But since skin grafts placed on a sclerodermatous bed eventually develop signs of scleroderma themselves, a similar transformation may affect these interposition vein grafts (Fries *et al.*, 1971). It therefore remains uncertain as to whether these improvements in digital blood flow will be maintained long-term. However, because both patients remain asymptomatic with objective evidence of improved pulsatile digital blood flow one year postoperatively, it may be concluded that microsurgical revascularisation may be indicated for salvage of severe hand or digital ischaemia in Raynaud's phenomenon and scleroderma.

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