CLINICAL MICROARTERIOLYSIS IN THE TREATMENT OF SPASTIC DISORDERS OF THE HAND, ESPECIALLY SCLERODERMA

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Arterial spasm due to exaggerated sympathetic response is an important mechanism for Raynaud's phenomenon in scleroderma associated often with periartiditial scarring. The results of cervical sympathectomy have been unsatisfactory in the upper limb because of additional sympathetic pathways. Flatt therefore devised a distal sympathectomy by stripping the vessels of their adventitia over a short length of artery. The results of this operation were found by Wilgis in a large series to be poor in patients with scleroderma. A radical distal microarteriolyis including adventitia and surrounding scar is described and the results in 13 patients, 11 with scleroderma, are reported. Minimum follow-up is one year. All patients had relief from pain at rest and healing of painful ulceration. Mild recurrence of small ulcers was seen in only four patients.

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... to the lower limbs the results of truncal sympathectomy are unsatisfactory in the upper limb (Brown, 1925). Flatt (1980) therefore devised distal sympathectomy in which the adventitia is circumferentially over a length of 3 to 4 mm. Flatt et al. (1981, 1983) removed adventitia from the common and proximal proper digital arteries in 13 patients with Raynaud's disease with excellent results with early return to work. Flatt et al. (1983) treated 13 patients with "superselective sympathectomy", removing adventitia from the common and proximal proper digital arteries in 13 patients with Raynaud's disease with excellent results with early return to work. Flatt et al. (1983) treated 13 patients with Raynaud's disease with excellent results with early return to work.

8. (1981, 1983 modified the technique described by Flatt et al. (1983) by removing the adventitia from the common and proper digital arteries distally to the proximal interphalangeal joint, usually at least 2 cm. Again excellent results were achieved by this technique using digital arteries in 13 patients with post-frostbite patients; but patients with the poorest response. Young patients with Raynaud's disease have had the best results.

... and mixed connective tissue disease (MCTD) and mixed connective tissue disease (MCTD) and mixed connective tissue disease (MCTD) and mixed connective tissue disease (MCTD) and mixed connective tissue disease (MCTD). It may progress to digital tip ulceration or gangrene, severe contracture of the interphalangeal and metacarpophalangeal joints, and subcutaneous calcinosis. In addition to the hand, visceral involvement of the alimentary tract, kidneys, lungs and heart commonly occurs in small vessel disease. Arterial spasm due to an exaggerated sympathetic vasomotor response to emotional upset; Kontos and Wasserman, the most important mechanism in the pathogenesis of the disease.

Cerebral arteries have been shown on angiography in almost all patients with scleroderma (Dabich et al., 1972). This has affected the ulnar artery at the wrist in almost 50% of patients, and the superficial palmar arch in the majority of patients. The arterial perfusion pressure is also reduced in the fingers of patients with scleroderma, and the pressure drop from wrist to finger suggests resistance to flow within the superficial palmar arch (Dabich et al., 1972; Kristensen, 1982).

Surgery is indicated in those patients with severe pain and ulceration. Unlike the lower limb, the results of truncal sympathectomy in the upper limb are temporary and unsatisfactory (Adson and Brown, 1925) because there are sympathetic pathways to the brachial plexus from the cervicothoracic sympathetic trunk, and these are left untouched in a cervical sympathectomy (Adson and Brown, 1925; Baddeley, 1965; Flatt, 1980). This prompted Flatt (1980) to devise a distal local sympathectomy, whereby the sympathetic nerves which are concentrated in the adventitia of the proper digital arteries are removed over 3 to 4 mm from the bifurcation of the common digital artery. More recently Jones (1989) described sympathectomy of the superficial palmar arch and common digital arteries in the ischaemic hands of scleroderma patients.

To improve the results this approach has been extended. The aim is to increase the blood flow to the hand by performing a distal adventitiotomy and removal of periarterial scar (arteriolyis) of the ulnar artery, superficial palmar arch, the common digital arteries and the proper digital arteries down to the distal interphalangeal joint level. In those cases in which the ulnar artery or the superficial arch were occluded a vein graft was used if proximal flow and distal run-off could be established. The ulnar artery and the superficial palmar and digital systems are commonly affected, but the radial artery, deep palmar arch and metacarpal arteries are rarely involved.
Evaluation of patients

Only patients with severe scleroderma who have had severe pain and ulceration refractory to conventional treatment were referred for radical microsympathectomy. Digital subtraction angiography was employed to evaluate proximal vessel disease, especially in the ulnar artery and superficial palmar arch.

A modified isolated cold stress test (Koman et al., 1984) of both hands is done pre-operatively and at three-monthly intervals post-operatively. The room temperature should be constant and the patient adjusted to this. Both hands were immersed in water at a temperature of 15°C for 15 minutes. Resting digital temperatures were recorded before the immersion, and at five-minute intervals for 20 minutes after the immersion. In the normal hand the digital pulp temperature returns to normal within 20 minutes.

Koman et al. (1984) have shown that Raynaud's disease is characterized by a low baseline temperature, a rapid cooling phase, and very slow rewarming compared to a normal digit.

Surgical technique

The patients were operated on under supraclavicular brachial regional anaesthesia plus light general anaesthesia. The supraclavicular anaesthesia gives a sympathetic block and leads to increased pinkness of the digits. Prophylactic antibiotics (appropriate to pre-operative cultures if ulcers are present) are given prior to application and inflation of a pneumatic arm tourniquet. Incisions were marked out as shown in Figure 1.

The angiogram indicates the level at which the ulnar artery becomes abnormal. The proximal incision commences over the ulnar artery in the distal forearm and passes directly over the superficial palmar arch in the palm, to the radial digital artery of the index finger. This allows access to the ulnar artery and the superficial palmar arch. Three intermediate short longitudinal incisions pass from the second, third and fourth interdigital webs towards the palmar proximal incision leaving a 1 cm skin bridge between the two incisions. These incisions overlie the common digital arteries, their bifurcations, and communications with the deep palmar arterial system and the proximal segments of the proper digital arteries. A curved incision is used in the fingers, allowing access to the proper digital arteries in the fingers, but not in continuity with the longitudinal incision over each common digital artery. The palmar incisions are made with care without undermining any flaps. In the digits, the flaps are elevated from near the mid-lateral line, making them thick, to prevent skin necrosis. With the aid of loupé magnification (3–4 x) the ulnar artery, superficial palmar arch, and digital vessels are dissected carefully away from adjacent veins and nerves, ensuring that all branches are preserved. The tourniquet is released and haemostasis achieved using bipolar diathermy. The digital vessels often require dissection under the operating microscope.

The adventitiotomy is also performed using an operating microscope. The adventitia containing sympathetic fibres is excised circumferentially from the artery, superficial palmar arch and digital arteries using fine jeweller’s forceps and microsurgical scissors taken not to damage the intima or media, facilitated by releasing the tourniquet pre-operatively. Any occasional perforators may be repaired with 10/0 nylon sutures. In advance of the plane of cleavage between adventitia and media, a fusiform incision is made with scissors to proceed with removal of the adventitia.

The occlusion of the ulnar artery and superficial palmar arch that is usually shown on angiography is not confirmed at operation, and a lumen flow is usually observed. The wall may appear thickened, with intimal proliferation. If the artery is occluded it is bypassed with reversed veins from the ipsilateral forearm or foot, using an end-to-end anastomosis proximally into the ulnar or radial. The distal anastomosis is either end-to-side superficial palmar arch or end-to-end between branches of the graft and the patent common...
ADRENERGIC IN SCLERODERMA

previously described (Jones et al, 1987b). If
tion of the superficial arch and no obvious
ulnar artery, no vein graft is inserted. It is
ther or not a vein graft augments the blood
and.
the vessels have had circumferential adventi-
ting with or without microvascular reconstruc-
tor palmar vessels, dilatation of the
is often evident, although this is better
wards off the end of the operation.Digits are
pink than pre-operatively.
ends are closed with interrupted 4/0 and 5/0
without any drainage. Low molecular weight
par or other antithrombotic agents have not
Operating time has been reduced to five and
and patients are discharged from hospital in
adal 10 mg orally twice daily has been
ator. A volar plaster slab is used for
to prevent flexion deformity, then gentle
n is commenced with physiotherapy. Sutures
el of three weeks. Appropriate early splintage
ain flexion and extension of the digits. This
portant aspect of management.
series, each hand was operated on in two
ly from the ulnar artery to the termination of
al vessels in the web spaces, and later
vessels proper were operated on. We now
plete the operation in one stage. Early in
incisions were continuous, but this led to
traction in a few cases so now the incisions
n. The follow-up is continued at three-month intervals, with
ing being performed.

bilateral and results

Traumatic ischaemia as described above has been
11 patients to date. 13 of these patients (17
post-operative follow-up of greater than one
constitute the basis for this report (Table 1).
consists of 8 female and 5 male patients with
age of 54 years (range 22–73). Scleroderma
may 41 patients with mixed connective tissue
Raynaud’s disease in the other 2
mean duration of the disease was 12 years
of. Six of these patients had previous cervical
omy and three had a chemical sympathectomy
urban amputations of necrotic digits. In
hands the wrist and palm were treated in the
nd the digits in a second stage. The remaining
treated by a single stage procedure. In five
procedures were required to bypass occluded
ed, with.
ends were followed up for between one and
, ulceration and intolerance to cold were
features in all these patients. Pre-operative
exquisite and very disabling. The most
improvement following surgery has been the
relief of pain and ulcer-healing which occurred in all
patients. All patients also had improved tolerance to cold
although there was still evidence of circulatory changes,
especially in cold weather. There was recurrence of mild
superficial ulceration in four patients. Two other patients
developed ulceration in an unoperated thumb, which
healed following digital microangiectomy. One of these
two also had ulceration of the feet. Freedom from pain
has continued during the period of the follow-up.

Delayed wound healing was observed in six hands
early in this series. This probably relates to the severity
of involvement of the microcirculation, but inadequate
flare design may have contributed. Drains are no longer
inserted in the palm and sutures are retained for three
weeks. These measures have eliminated delays in wound
healing.

The modified incisions, volar plaster slabs, more
intense early physiotherapy and sometimes the use of a
night extension splint have eliminated permanent stiff-
ness and contracture.

The results of the cold stress were analysed in 14 of the
17 hands treated. In seven of these hands the resting
atmosphere temperature was increased by 2.1°C (range 1.2–3.9°C)
and rewarmed to 2.4°C higher (range 0.7–3.2°C) 20
minutes after the cold stress compared with the
pre-operative temperatures. A representative graph of pre-
and post-operative temperature testing is shown in Figure
2. In two hands there was no difference between pre- and
post-operative temperature. In five the resting tempera-
ture was 2.6°C lower but in two the hands rewarmed
more quickly following the cold stress test. All these five
patients were relieved of pain and had their ulcers healed.

Other features of improved blood supply have been
better and more normal nail growth and the growth of the
hair on the dorsum of the proximal phalanges. The plu-
ces of arterial scanning in the assessment of blood flow,
and skin potential tests to assess the sympathetic
innervation of sweat glands are under investigation.

Discussion

The use of vasodilators, collagen inhibitors, immunosup-
pressants and rheological agents provide temporary
improvement in the course of scleroderma (Gahhos et al,
1984).

Significant clinical improvement has been noted with
daily administration of reserpine, guanethidine, methyl-
dopa, tolazine, phenoxybenzamine and prazosin. Intra-
artrial reserpine, though beneficial, can be hazardous in
terms of arterial damage (Kristensen, 1979; Porter et al,
reserpine as a Bier’s block is much less hazardous (Taylor
et al, 1982). Its action is short lived and lasts only a few
weeks. Ketanserin (a serotonin receptor blocker) and
nifedipine (a calcium channel blocking agent which
produces vasodilatation) have been reported to increase
blood flow and heal digital ulcers (Roald and Seem, 1984;
Winston et al, 1983). Intravenous prostaglandin E₁, a
### Table 1—Sympathectomy patients

<table>
<thead>
<tr>
<th>Age/sex/disease</th>
<th>Date</th>
<th>Hand</th>
<th>Site</th>
<th>Ischaemic pain relief</th>
<th>Ulcer control</th>
<th>Cold stress test</th>
<th>Flexion contracture</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. 58/F/scleroderma</td>
<td>2/86</td>
<td>R</td>
<td>Palm</td>
<td>+</td>
<td>NA</td>
<td>NA</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td></td>
<td>5/86</td>
<td>R</td>
<td>Digits 2-5</td>
<td>+</td>
<td>NA</td>
<td>NA</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2/86</td>
<td>L</td>
<td>Palm</td>
<td>NA</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>2. 23/F/scleroderma</td>
<td>10/87</td>
<td>L</td>
<td>Palm</td>
<td>NA</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>3. 51/M/Raynauds</td>
<td>11/89</td>
<td>R</td>
<td>Palm/Digits 2-5</td>
<td>++</td>
<td>++</td>
<td>+</td>
<td>++</td>
<td>Also underwent sympathectomy of left planar vessels for ischaemic toes</td>
</tr>
<tr>
<td></td>
<td>24/8</td>
<td>R</td>
<td>Thumb</td>
<td>++</td>
<td>+</td>
<td>+</td>
<td>++</td>
<td>Vein graft from ulnar artery to first common digital artery</td>
</tr>
<tr>
<td></td>
<td>5/88</td>
<td>L</td>
<td>Palm</td>
<td>NA</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Vein graft ulnar artery in forearm to superficial palmar arch</td>
</tr>
<tr>
<td>4. 43/M/scleroderma</td>
<td>8/87</td>
<td>R</td>
<td>Palm/index</td>
<td>NA</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Vein graft radial artery to superficial palmar arch</td>
</tr>
<tr>
<td></td>
<td>10/89</td>
<td>L</td>
<td>Palm/index</td>
<td>NA</td>
<td>++</td>
<td>+</td>
<td>0</td>
<td>Vein grafts from ulnar artery to 3 common digital vessels</td>
</tr>
<tr>
<td>5. 66/F/scleroderma</td>
<td>5/88</td>
<td>R</td>
<td>Palm</td>
<td>++</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>Vein graft from superficial palmar arch to first common digital artery</td>
</tr>
<tr>
<td>6. 19/M/mixed connective tissue disease</td>
<td>10/88</td>
<td>R</td>
<td>Palm</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td></td>
<td>11/88</td>
<td>R</td>
<td>Digits 2-5</td>
<td>++</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2/89</td>
<td>L</td>
<td>Palm/digits 2-5</td>
<td>++</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>7. 69/F/scleroderma</td>
<td>11/88</td>
<td>R</td>
<td>Palm/Digits 2-5</td>
<td>++</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>8. 69/F/scleroderma</td>
<td>12/88</td>
<td>R</td>
<td>Palm/Digits 2-5</td>
<td>++</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>9. 64/M/scleroderma</td>
<td>4/89</td>
<td>R</td>
<td>Palm/Digits 2-5</td>
<td>++</td>
<td>N.A.</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>10. 56/F/scleroderma</td>
<td>9/89</td>
<td>R</td>
<td>Palm/digits 2-5</td>
<td>++</td>
<td>+</td>
<td>N.A.</td>
<td>++</td>
<td></td>
</tr>
<tr>
<td>11. 72/F/scleroderma</td>
<td>3/90</td>
<td>R</td>
<td>Palm/thumb</td>
<td>N.A.</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>12. 40/M/scleroderma</td>
<td>5/90</td>
<td>R</td>
<td>Palm/Digits 2-5</td>
<td>++</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>13. 43/F/scleroderma</td>
<td>6/90</td>
<td>L</td>
<td>Palm/index, ring, little fingers</td>
<td>++</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td></td>
</tr>
</tbody>
</table>

**Ulcer control**
- NA— not present pre-op.
- 0—recurrence requiring reoperation or amputation
  +—minor recurrence
  ++—resolved

**Ischaemic pain relief**
- NA— not present pre-op.
- 0—unchanged or worse.
  +—improved
  ++—resolved

**Flexion contracture**
- 0—full ROM or unchanged from pre-op.
  +—decreased ROM from pre-op, not functionally limiting
  ++—limited ROM with limitation of function of wrist and/or flexion of digits
Some patients have high fibrinogen levels and the effect of the fibrinolytic stimulating agent stanozolol has been promising (Jarrett et al, 1978). Plasmapheresis has also been used with satisfactory results, attributed to defibrination and lysis of fibrin deposits (Talpos et al, 1978; O'Reilly et al, 1979). Not all patients have abnormal serum proteins or altered viscosity.

Only 20% of Baddeley's (1965) 64 cases of primary Raynaud's disease were cured by truncal sympathectomy, with a further 34% improved but not cured. He therefore suggests that cervical sympathectomy should be reserved for occasional severe progressive cases where the survival of digits is in jeopardy. Although serotonin or catecholamine sensitivity may contribute, it is the additional sympathetic pathways that play a significant role in the high relapse rate. In addition to the contribution of the cervicothoracic sympathetic trunk to the brachial plexus, there are also communications from the sinusvertebral nerve, carotid plexus, and the nerve of Kuntz. These latter pathways remain untouched during cervical sympathectomy. Furthermore, the sympathetic fibres do not travel as distinct bundles but are widely dispersed, and there are many branches from cutaneous nerves to the vascular tree at multiple levels in the limb; these run from the radial nerve and lateral cutaneous nerve of the forearm to the radial artery, from the ulnar nerve and medial cutaneous nerve of the forearm to the ulnar artery, and the common digital nerves to the superficial palmar arch (Adson and Brown, 1925; Baddeley, 1965; Flatt, 1980).

In the digits, sympathetic fibres travel with the digital nerves; there are approximately four large sympathetic branches which pass to the digital artery and form a sympathetic nerve network within the adventitia of the arteries (Flatt, 1980; Morgan et al, 1983).

Jones et al (1987a; 1987b; 1989) have noted an improvement in the symptoms of patients with scleroderma whose disabling Raynaud's phenomenon and digital ulceration has been refractory to medical management when treated with digital sympathectomy. Two patients shown to have proximal occlusion of the radial and ulnar arteries due to intimal hyperplasia rather than pure sympathetic overactivity had dramatic immediate resolution of their severe pain and healing of digital ulcers following vein grafting. The results were sustained after one year. The concept of palmar arch adventitiotectomy was also described.

Our treatment is aimed at producing vasodilatation by stripping the vessels of their sympathetic supply and separating them from surrounding fibrotic tissue, starting from relatively normal artery (ulnar above the wrist) and extending it to the rest of the superficial vasculature of the hand. By increasing the blood flow to the hand, occlusion of organically narrowed vessels can be prevented or delayed. Where total occlusion was present (five cases) the involved segment was bypassed with a vein graft. All patients were relieved of their severe pain and had their ulceration healed. Improved tolerance to
cold as demonstrated by the cold stress test occurred in seven of 14 hands. Temperature recordings are not an infallible guide to improved blood flow, since pain and ulceration may disappear while temperature recordings are not improved.

In conclusion, a more radical approach to interruption of the peripheral sympathetic pathway has improved the blood supply to the hand and led to healing of ulcers, relief of pain, and improved tolerance to cold in these patients with scleroderma. Good results were achieved in patients with advanced scleroderma in whom the response has previously been poor (Wilgis, 1981; 1985). The period of follow-up has now been as long as five years and there has been little deterioration. This radical surgical approach is now carried out safely in one stage and is recommended as the surgical method of choice. Cervical sympathectomy is no longer an alternative procedure in the vasospastic disorders. Prostaglandin infusions may be of greater benefit in the post-operative phase than pre-operatively.

Acknowledgements
The assistance of a number of previous Fellows in the Research Centre is gratefully recorded. We have appreciated the referral of patients by rheumatologists.

Addendum
Since submission of this publication the total number of hands operated upon is now 42, the longest follow-up being seven years. Of 32 hands with complete data and follow-up longer than three months, five had recurrence of pain (three mild, two at pre-operative level) and four had recurrence of ulcers.

References

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