Progressive systemic sclerosis is a slowly spreading disorder of unknown aetiology, frequently grouped with the collagen diseases (systemic lupus erythematosus and polyarteritis) because there are features in common.

The early signs of progressive systemic sclerosis include Raynaud's phenomenon, the development of thin atrophic skin over the extremities, widening of the periodontal membrane and shrinkage of soft tissue around the mouth. As the disease progresses, the atrophic skin of the extremities may ulcerate. The response to sympathectomy is disappointing.

Over a period of 3 years an adult male presented on six occasions for amputation of a digit severely involved by a process considered to be a severe form of progressive systemic sclerosis. None of these procedures was extensive or prolonged (average duration 25 min) yet they all caused postoperative pain requiring large doses of analgesic drugs which, however, had minimal effect.

General anaesthesia was used on all six occasions; in four instances the anaesthetic sequence included a narcotic analgesic either in premedication or during the operative procedure; in two instances regional anaesthesia of the brachial plexus was combined with light general anaesthesia (table I).

The requirements for analgesia after operation were similar following each anaesthetic, apart from a time lag of 15-18 h on the two occasions on which regional anaesthesia was employed. The first analgesic after operation was required within 30 min of emergence from anaesthesia, followed by one or more additional doses within the next 2 h.

Anaesthetics nos 4 and 5 are illustrative of the severity of the problem. Following anaesthetic 4, control of postoperative pain was achieved only after a total dosage of papaveretum 30 mg, morphine 5 mg and diazepam 10 mg had been given over a period of 100 min. Following anaesthetic 5, the pain was so severe following sequestrectomy of a metatarsal that a sciatic nerve block was performed at 90 min after total failure to produce analgesia with pentazocine and morphine. Relief following this was instantaneous and lasted for 16 h.

DISCUSSION

Two features of this series of anaesthetics are of interest—the very prolonged period of sensory anaesthesia following regional block, and the reduced response to systemic analgesics.

Prolonged sensory anaesthesia in patients with progressive systemic sclerosis has been reported previously. Eisele and Reitan (1971) attributed the effect to severely impaired perfusion with a possible reduction in tissue pH. Lewis (1974) found that the prolongation of anaesthesia was unaffected by an infusion of low molecular weight dextran, but was decreased significantly following an infusion of sodium bicarbonate. As tissue pH was not measured it was not possible to determine whether this decrease was the result of an alteration in tissue pH or improved perfusion.

The sympathetic nervous system and the motor and sensory components of the peripheral nervous system are all involved in the disease process. Henriksen, Kristensen and Wadskov (1977) found a decreased or absent vasoconstrictor response to an
TABLE I. Details of surgical procedure, premedication, anaesthesia and analgesia used on six occasions for a patient with progressive systemic sclerosis

<table>
<thead>
<tr>
<th>Operation</th>
<th>Premedication</th>
<th>Anaesthesia</th>
<th>Analgesia after operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amputation (L) ring, little (R) middle fingers (30 min)</td>
<td>Pethidine 50 mg, Promethazine 50 mg, Hyoscine 0.4 mg</td>
<td>Althesin, halothane, DF 118 50 mg</td>
<td>Pethidine 100 mg, 30 min, Pethidine 100 mg, 240 min</td>
</tr>
<tr>
<td>Amputation (R) index finger (25 min)</td>
<td>Pethidine 50 mg, Promethazine 50 mg</td>
<td>Thiopentone, halothane</td>
<td>Pentazocine 50 mg, immediately Pentazocine 50 mg, 40 min, Pentazocine 50 mg, 100 min</td>
</tr>
<tr>
<td>Amputation (L) thumb (15 min)</td>
<td>None</td>
<td>Thiopentone, halothane, supraventricular brachial plexus block</td>
<td>Pentazocine 50 mg, 920 min, Pethidine 50 mg, 950 min</td>
</tr>
<tr>
<td>Amputation (R) ring finger (45 min)</td>
<td>Pethidine 50 mg, Promethazine 50 mg</td>
<td>Thiopentone, halothane, fentanyl 0.1 mg</td>
<td>Papaveretum 20 mg, 10 min, Diazepam 10 mg, papaveretum 10 mg, 60 min, Morphine 5 mg i.v., 100 min Papaveretum 20 mg, 240 min</td>
</tr>
<tr>
<td>Sequestrectomy 4th (L) metatarsal (15 min)</td>
<td>None</td>
<td>Thiopentone, halothane, pentazocine 60 mg</td>
<td>Pentazocine 50 mg, 15 min, Morphine 10 mg i.v., 75 min Sciatic nerve block, 90 min Morphine 10 mg, 960 min</td>
</tr>
<tr>
<td>Amputation (R) thumb (25 min)</td>
<td>None</td>
<td>Thiopentone, halothane, supraventricular brachial plexus block</td>
<td>DF 118, 1000 min Physeptone 5 mg, 1120 min</td>
</tr>
</tbody>
</table>

increase in venous transmural pressure and attributed this to a sympathetic neuropathy.

Christopher and Robinson (1972) found a decrease in conduction velocity in both sensory and motor peripheral nerves. These authors suggested that the peripheral neuropathy was the result of impairment of the blood supply to the nerves, a conclusion which seems to be borne out by a recent report by Cormane, Hamerlinck and Nunzie (1978) which postulates that the primary event in progressive systemic sclerosis is an immunological attack on the peripheral parts of the vascular system.

Histological studies of the skin of patients with progressive systemic sclerosis have shown an increased number of nerve endings per unit area (Milne, 1972). The possibility that stimulation of a disproportionate number of nerve endings following surgery might explain the decrease in the response to systemic analgesics was considered, and that this might be reflected in an alteration in two-point discrimination. This was tested and was found to be within the normal range (Gellis and Pool, 1977).

Advanced degenerative arterial disease is frequently associated with necrosis of the extremities requiring amputation of digits or portions of limbs: after surgery the patient's pain can be controlled with simple analgesics. However, patients with severe rest pain do not respond to large doses of systemic analgesics, yet show an immediate response to paravertebral nerve block with local anaesthetics or ablative agents (M. E. Coull, 1979, personal communication).

It is concluded that the intense pain following surgery in this patient with progressive systemic sclerosis was ischaemic in nature—the lack of response to systemic analgesics and immediate response to sciatic nerve block mirroring the effects of rest pain in advanced degenerative arterial disease.

The prolonged period of sensory anaesthesia in this patient and in others with progressive systemic sclerosis is likely to be a result of impaired blood supply to the peripheral nerve—the explanation previously advanced to account for the peripheral neuropathy.

The patient in this report was considered to suffer from a very severe and atypical form of progressive systemic sclerosis and his response to pain may not be the same as that of other patients with the disease. However, it has not been possible to trace any patients requiring similar surgery to compare their response to analgesics or regional nerve block.

The condition is extremely rare. An average of one new case per year is referred to the Dermatology Units of the major teaching hospitals in Glasgow, each having approximately 4000 new referrals per year.
CONCLUSION
The inclusion, when possible, of a regional nerve block in the anaesthetic regime for patients with progressive systemic sclerosis would seem to be advantageous, providing a prolonged period of analgesia after surgery. The author (Neill, 1978) has found previously that simple analgesics were adequate following regional block for hand surgery, but this would not appear to be the case in patients with progressive systemic sclerosis.

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REFERENCES