Surgery of the Hand in Patients with Systemic Sclerosis: Outcomes and Considerations

EARL R. BOGOCH and DAGMAR K. GROSS

ABSTRACT. ***Objective.*** To assess the current status of hand surgery in patients who have systemic sclerosis (SSc) and to elucidate special issues of surgery in this patient group.

Methods. A systematic review of English language original studies of surgical procedures of the hand in patients with SSc was performed using Medline, PreMedline, Embase, and Web of Science, from 1975 to March 15, 2004.

Results. Thirty-four studies were reviewed: 5 describing surgical procedures on joints, 13 on calcinosis removal, and 20 on digital sympathectomy. When the hand is affected by advanced contracture and deformity due to scleroderma, a nominal measured improvement in position and function may lead to a substantial improvement in the patient's adaptive ability to perform certain activities of daily living. A major concern is the potential for postoperative digital ischemia secondary to vascular involvement, as most of these patients exhibit blood vessel wall changes and Raynaud's phenomenon. Surgical wounds generally heal well following fusion of the proximal interphalangeal (PIP) or distal interphalangeal joint. Correction of severe flexion contractures of the PIP joint improves function and may reduce the frequency of dorsal skin ulceration. Recurrent digital tip ulceration occurs in 31.8–71.4% (median 45.2%) of scleroderma patients, reported to progress to gangrene and autoamputation in 14–29% of cases. Microsurgical revascularization of the hand, digital arterial reconstruction, and peripheral sympathectomy may improve digital vascular perfusion, heal digital ulcers, and relieve pain. Subcutaneous calcifications occur in 8.9–73.1% (median 44.1%) of SSc patients, most commonly at the fingertip, causing pain, functional impairment, and ulceration. Calcinosis can be partially removed with a high-speed burr or carbon dioxide laser.

Conclusion. The goals of surgery for advanced SSc affecting the hand are limited and include pain relief through sympathectomy and increased perfusion, repositioning the digit, providing a functional position of fusion, and modest mobilization through resection arthroplasty. (J Rheumatol 2005;32:642–8)

Key Indexing Terms:
CALCINOSIS HAND JOINT SURGERY SYMPATHECTOMY SYSTEMIC SCLEROSIS

Systemic sclerosis (SSc), or scleroderma, encompasses both the diffuse (widespread) and limited (restricted) skin thickening subsets. The latter includes the CREST syndrome (calcinosis, Raynaud's esophageal dysmotility, sclerodactyly, and telangiectasias), a term that is infrequently used today. Scleroderma is an infrequent connective tissue disease of unknown cause, with an incidence ranging from 1.5 to 19.1 new cases per million population[1]. It is a systemic disorder that in addition to cutaneous involvement may affect the gastrointestinal tract, kidney, lung, heart, peripheral joints, and small blood vessels. The important manifestations of SSc in the hand include sclerodactyly, changes in blood vessel walls, Raynaud's phenomenon with digital ischemia, which may progress to ulceration or gangrene, flexion contracture of the interphalangeal (IP) joints, extension contracture of the metacarpophalangeal (MCP) joints, soft tissue atrophy over the distal phalanges, bony resorption of distal phalangeal tufts, nail deformities, and calcinosis[2-8]. These deformities are painful, may severely limit function, and are also cosmetically unsatisfactory.

The role of surgery in the management of hand manifestations of SSc remains unclear and literature published in this area is limited. The usefulness of reconstructive surgery of the hands in patients with advanced deformity in SSc has been questioned due to the frequent global stiffness of the hand and concerns that wound healing could be impaired by digital ischemia and sclerodactyly. We conducted a systematic review of the literature to assess reports of surgery for hand deformity in patients who have progressive SSc, and to elucidate the special issues of surgery in this patient group.

MATERIALS AND METHODS

A literature search was performed using Medline, PreMedline, Embase, and Web of Science to identify English language citations for studies of surgical procedures of the hand in patients with SSc, from 1975 to March 15, 2004. Keywords used included various combinations of scleroderma, sys-

From the Department of Surgery, Division of Orthopaedic Surgery, Martin Family Centre for Arthritis Care and Research, Mobility Program, St. Michael's Hospital, University of Toronto; and MedSci Communications & Consulting Co., Toronto, Ontario, Canada.

E.R. Bogoch, MD, Professor, Department of Surgery, University of Toronto, Director, Mobility Program, St. Michael's Hospital; D.K. Gross, MSc, President, MedSci Communications & Consulting Co.

Address reprint requests to Dr. E.R. Bogoch, 55 Queen St. East, Suite 800, Toronto, Ontario M5C 1R6, Canada. E-mail: bogoche@smh.toronto.on.ca

Submitted June 1, 2004; revision accepted October 12, 2004.

temic sclerosis, CREST, hand, finger, and surgery. Reference lists of articles obtained in the search were reviewed to identify additional articles as well as any studies published prior to 1975.

Inclusion and exclusion criteria. Only studies on the surgical management of SSc that provided data specific to patients with a diagnosis of SSc were included. Original studies reporting new data, such as prospective studies, cross-sectional studies, retrospective chart reviews, and case reports, were included. Studies on the medical management of SSc that did not report or discuss the surgical management of SSc patients were excluded. Two review articles limited to analysis of microsurgical revascularization and digital sympathectomy studies were excluded, although their reference lists were examined to identify any original reports that were not identified through the literature search. Since surgery of the hand in patients wtih SSc is relatively uncommon, the majority of studies were retrospective chart reviews or case reports, without controls, precluding metaanalysis or quantitative comparison of results.

RESULTS

Thirty-four studies met the inclusion criteria, including 2 prospective studies, 11 cross-sectional studies, 7 retrospective chart reviews, and 14 case reports. Of the 34 studies, 5 studies describe the results of surgical procedures of the joints, 13 report on calcinosis removal, and 20 report the results for digital sympathectomy (Table 1).

Hand manifestations of SSc. Baron, *et al*[2] studied the articular manifestations of SSc in 38 patients. Of these, 66% experienced joint pain and 45% had limitation of joint movement. Radiological changes included periarticular osteoporosis (42%), erosions (40%), and joint space narrowing (34%). Other investigators have documented the presence of an erosive polyarthritis involving the small joints of the hand, most frequently in the distal interphalangeal (DIP) and MCP joints[3,5,40].

An additional consideration in the development of fixed flexion deformities of the hand is peritendinous sclerosis with subsequent tendon shortening[41]. Skin becomes inelastic and fixed with disease progression, ultimately becoming tightly stretched over the dorsal aspect of joints that are contracted in flexion. At these sites, the dermal capillary bed may be reduced by up to 80%. The resulting ischemia, along with the flexed position of the joints, renders the dorsal aspects susceptible to frequent minor trauma, infections, and ulceration.

Anesthesia. Local or regional anesthesia is preferred for patients who present with cardiac or pulmonary manifestations of SSc. Severe perioral involvement can result in difficulty with orotracheal intubation[7,20]. Normally, distal surgery may be performed with distal regional anesthesia. However, distal ring block is problematic in these patients, as even slow infiltration of fluid into the dense soft tissue of the digits is painful, and the density of the tissue inhibits adequate diffusion of the local anesthetic. Axillary or wrist blocks may provide a vasodilatory effect[8,25]. Gilbart, *et al*[22] used regional anesthetic at the wrist and recommended consideration of a more proximal scalene or brachial plexus regional anesthesia if wrist block is not possible. Establishing an intravenous line in the opposite extremity is difficult and should be performed in a proximal location where skin involvement is less severe. A tourniquet may be applied to the upper arm, but for no longer than 1 to 1.5 hours, and it should be carefully monitored because of the risk of further compromise of the vascular system[8,20]. In our experience, it is not usually necessary to inflate the tourniquet that has been applied.

Wound healing and joint fusion. When the hand is affected by advanced contracture and deformity due to SSc, a nominal measured improvement in position and function may lead to a substantial improvement in the patient's adaptive ability to perform certain activities of daily living. However, some physicians and surgeons are reluctant to recommend surgery to patients with SSc who have hand manifestations, including calcinosis and advanced joint contractures, associated with severe functional limitations. One major concern is the potential for postoperative digital ischemia secondary to vascular involvement, as most of these patients have blood vessel wall thickening and luminal narrowing secondary to connective tissue matrix production/deposition with superimposed Raynaud's phenomenon[42]. Nevertheless, in a retrospective study of 272 surgical procedures in patients with SSc, wound healing was reported to be uncomplicated for all procedures[8].

The most common hand deformity in SSc is progressive fixed flexion contracture of the proximal interphalangeal (PIP) joint[3,8], which is both disabling and cosmetically unsatisfactory. Reduction of severe flexion contractures not only improves function, but may also reduce the frequency of skin ulceration[22]. Two procedures previously described to address flexion deformities of the PIP joints in SSc are arthrodesis[7,8,20,22] and Swanson flexible implant arthroplasty[21]. The reported results of Swanson arthroplasty were poor. The postoperative range of motion reported was low (mean 13°, range 0°–28°), and complications included slow wound healing in 4 of 20 implants, of which 2 required extraction[21]. In addition, substantial skeletal shortening of the digit was required to overcome extensive soft tissue contracture and flexion deformity.

Most authors recommend fusion of the PIP joint, with the position of fusion ranging from 30° for the index finger to 55° in the little finger. In a study of moderate to severe hand manifestations of SSc by Jones, *et al*[7], 53 PIP joint contractures with secondary hyperextension at the MCP joints were treated successfully with PIP fusion in 45° to 55° of flexion, with radiographic union in 94% of joints within 8 weeks of surgery. In all cases, wounds healed well, and in some patients dorsal PIP ulcers healed with this technique alone. Lipscomb, *et al*[20] performed 16 PIP joint fusions at 30° to 60° of flexion in patients with SSc with very limited flexion through the MCP joints, thus allowing thumb-finger opposition for pinch and grasp function. In all cases, fusions were obtained. All wounds healed, although wound healing was

Table 1. Studies of hand surgery in patients with SSc, sorted by focus of study.

Study	Study Design	Patients (with SSc), n	Focus of Study	Followup	Procedures Performed	Results	Complications
Posner[9] (1980)	Case report	1 (1)	Amputation	Unknown	Partial amputation of digits (n = 8)	Vascular status of one of 2 digits remaining on left hand is precarious	
Berggren[10] (1965)	Case report	1 (1)	Calcinosis	Unknown	Calcinosis excision	Relief of pain, improved function (grasp)	Slow wound healing; wound flap tips did not survive
Bottomley[11] (1996)	Cross-sectional	6 (6)	Calcinosis	1 yr	Calcinosis treated with carbon dioxide laser (n = 21)	12/21 (57%) complete resolution of pain; 5/21 (24%) partial resolution of pain; 2/21 (10%) no improvement in pain; 2/21 (10%) recurrence of calcinosis within 3-4 mo	2/21 (10%) postoperative infection
Chamberlain[12] (2003)	Case report	1 (1)	Calcinosis	3 yrs	Calcinosis treated with carbon dioxide laser (n = 15)	Significant remission lasting at least 3 yrs	
Fahmy[13] (1998)	Cross-sectional	10 (6)	Calcinosis	12 mo	Calcinosis removal using a high-speed dental burr (n = 15)	12/15 showed symptomatic improvement	4/15 (27%) experienced dysesthesia around the stab incision, lasting up to 6 mo
Hussman[14] (1995)	Case report	6 (1)	Calcinosis	Unknown	Calcinosis excision (n = 8)	Resolution of pain, healing of recurrent skin ulcers, improved function	
MacDowell[15] (1969)	Case report	1 (1)	Calcinosis	4 yrs	Calcinosis excision (n = 9)	Relief of pain, improved function	None when using dental burr
Mendelson[16] (1977)	Cross-sectional	11 (7)	Calcinosis	8–63 mo	Calcinosis excision (n = 9)	Relief of pain, improved function	6/9 (67%) slow wound healing
Polio[17] (1989)	Case report	1 (1)	Calcinosis	8 mo	Calcinosis excision (n = 1)	Relief of pain and tenderness	
Schlenker[18] 1973	Retrospective chart review	11 (11)	Calcinosis	10–12 mo	Calcinosis excision (n = 2)	2/2 improved hand function	1/2 (50%) mild skin necrosis at wound margin
Thurman[19] (1991)	Case report	1 (1)	Calcinosis	6 mo	Calcinosis excision (n = 1)	Regained good function of hand	2/8 (25%) delayed wound healing
Lipscomb[20] (1969)	Case report	6 (6)	Joint procedure	8 wks to 4 yrs	IP arthrodeses (n = 18); MP joint arthroplasty (n = 4) MP joint capsulotomy (n = 4)	18/18 (100%) joint fusion, improved function	3/26 (12%) secondary infection of wound; 2/26 (8%) superficial cellulitis
Norris[21] (1985)	Cross-sectional	6 (6)	Joint procedure	1 yr	PIP joint arthroplasty (n = 20)	6/8 patients (75%) improved hand function	4/20 (20%) slow wound healing; 2/20 (10%) required prosthesis removal
Gilbart[22] (2004)	Cross-sectional	7 (7)	Joint procedure, calcinosis	1.5–9 yrs	MP excision (n = 6); PIP joint fusion (n = 13); DIP joint fusion (n = 10); Thumb IP joint fusion (n = 1); Calcinosis removal (n = 4)	100% fixation in IP joint fusion; 6/7 (86%) satisfactory wound healing	1/7 (14%) postoperative ischemia and fingertip autoamputation; 4/24 (17%) removal of figure-of-eight tension-band wires in second procedure
Jones[17] (1987)	Cross-sectional	31 (31)	Joint procedure, calcinosis, sympathectomy	1 yr	Digital amputation (n = 9); Digital sympathectomy (n = 5); Microsurgical revascularization (n = 2); PIP joint arthrodeses (n = 12); MP joint capsulotomies (n = 4); Calcinosis excision (n = 7)	Microsurgical reconstruction (n = 2): immediate resolution of pain, rapid healing of digital ulcers, asymptomatic at 1 yr followup	

Table 1. Continued next page.

Study	Study Design	Patients (with SSc), n	Focus of Study	Followup	Procedures Performed	Results	Complications
Melone[8] (1999)	Cross-sectional	Unknown	Joint procedure, calcinosis, sympathectomy	1.5–15 yrs	IP arthrodeses (n = 211); MP implant arthroplasty (n = 28); Thumb basal joint arthroplasty (n = 2); Calcinosis excision (n = 12); Digital sympathectomy (n = 10)	Uncomplicated wound healing; radiographic union of arthrodesis within 8 wks; improved vascularity, decreased pain, healing of ulcers following digital sympathectomy	
El-Gammal[23] (1991)	Case report	3 (1)	Sympathectomy	1–15 mo	Digital sympathectomy (n = 3)	Marked to complete relief of pain; healing of ulcers; cessation of subungual discharge	
Flatt[24] (1980)	Cross-sectional	8 (1)	Sympathectomy	2 yrs	Digital sympathectomy	Marked relief of pain; some healing of ulcers	
Gahhos[6] (1984)	Cross-sectional	59 (59)	Sympathectomy	up to 10 yrs	Surgical management of finger ulcers: Debridement, skin grafts, cervical sympathectomy, fingertip amputation	8/59 had cervical sympathectomy*: 1/8 (12.5%) pain relief for 10 years; 4/8 (50%) considerable pain reduction for 1–2 yrs; 3/8 (37.5%) no pain relief	
Greengrass[25] (2003)	Case report	1 (1)	Sympathectomy	6 mo	Digital sympathectomy	Complete resolution of pain and healing of digital ulcers	
Hafner[26] (1997)	Case report	2 (2)	Sympathectomy	2 yrs	Digital sympathectomy (n = 2)	Complete resolution of pain and healing of digital ulcers	
Jones[27] (1987)	Case report	2 (2)	Sympathectomy	1 yr	Microsurgical reconstruction (n = 2)	Immediate resolution of pain, healing of digital ulcers within 3–4 wks, asymptomatic at followup	
Koman[28] 1995	Cross-sectional	6 (5)	Sympathectomy	6 mo	Peripheral sympathectomy (n = 7)	Decreased pain and healing of digital ulcers	
McCall[29] (1999)	Retrospective chart review	7 (4)	Sympathectomy	1–76 mo	Digital sympathectomy (n = 16 digits)	Relief of pain, healing of ulcers	1/4 (25%) patients slow wound healing (16–24 wks); 1/4 (25%) patients recurrent ulcers after 2 yrs
O'Brien[30] (1992)	Prospective	13 (11)	Sympathectomy	1–5 yrs	Digital sympathectomy	Pain resolved (9/11) or improved (2/11); healing of ulcers	3/11 (27%) minor recurrence of ulcers
Ruch[31] (2002)	Cross-sectional	22 (22)	Sympathectomy	46 mo	Digital sympathectomy (n = 29)	24/29 (82%) hands decreased pain, improved ulcer healing, reduction in occurrence of digital ulcers	4 distal fingertip amputations performed in 3 patients (14%)
Stratton[32] (1997)	Retrospective chart review	13 (13)	Sympathectomy	19.3 mo	Digital sympathectomy	Mean pain score reduced from 3.9 to 3.2; mean ulcer score reduced from 0.92 to 0.54	2/13 (15%) minor wound sepsis; 1/13 (8%) required fingertip amputation
Taylor[33] (2002)	Retrospective chart review	15 (15)	Sympathectomy	1 yr	Microsurgical reconstruction (n = 8); periarterial sympathectomy (n = 2)	7/8 (88%) microsurgical reconstruction showed healing of ulcers, improved severity of Raynaud's phenomenon attacks; 2/2 digital sympathectomy had no healing of ulcers	1/8 (13%) postoperative healing complications
Tham[34] (1997)	Retrospective chart review	7 (6)	Sympathectomy	1–3 yrs	Digital sympathectomy (n = 7)	19/22 digits complete resolution of pain; 3/22 digits mild residual pain with cold exposure; all fingertip ulcers healed completely in 20–40 days	2/22 (10%) digits delayed wound healing
Tomaino[35] (2000)	Case report	1 (1)	Sympathectomy	1.5 yrs	Palmar sympathectomy; Digital arteriolysis	Complete resolution of pain	
Tomaino[36] (2001)	Retrospective chart review	6 (6)	Sympathectomy	2.5 yrs	Palmar sympathectomy (n = 8)	Significant pain reduction, complete resolution of ulcers	

Table 1. Continued.

Study	Study Design	Patients (with SSc), n	Focus of Study	Followup	Procedures Performed	Results	Complications
Tomaino[37] (2002)	Case report	2 (2)	Sympathectomy	1.5 yrs	Digital arterial reconstruction (n = 2)	2/2 complete resolution of pain and healing of digital ulcers	
Ward[38] (1995)	Prospective	7 (7)	Sympathectomy	50.7 mo	Digital sympathectomy (n = 9)	Digital ulcers healed an average of 3.7 wks after surgery	3/9 (33%) hands recurrence of ulcers; 1/9 (11%) reflex sympathetic dystrophy; 1/9 (11%) delayed wound healing; 2/9 (22%) fingernail detachment
Yee[39] (1998)	Retrospective chart review	9 (6)	Sympathectomy	10–47 mo	Digital sympathectomy (n = 10)	100% healing of ulcers	2/10 (20%) spontaneous loss of distal tip; 2/10 (20%) necrotic distal tip amputation

IP: interphalangeal; DIP: distal interphalangeal; PIP: proximal interphalangeal; MP: metacarpophalangeal. * None prevented finger ulcers. Fingertip amputation was the most successful surgical procedure for management of ulcers.

considered to be slow in some patients (more than 4 weeks), and 5 procedures developed secondary infections, which were successfully and quickly treated. One patient was reported to have necrosis of skin edges and a secondary infection. More recently, a retrospective study of 211 IP joint arthrodeses in 70 patients with SSc over 15 years reported uncomplicated wound healing and radiographic union within 8 weeks of surgery[8]. Finally, Gilbart, *et al*[22] reported 13 PIP fusions at 30° to 45° of flexion, all of which healed without delay. In cases where one IP joint is corrected for deformity and the neighboring joint is becoming deformed, correction of both deformities in one procedure was recommended to obviate a subsequent procedure[22].

Correction of DIP contractures generally requires fusion[20,22]. Amputation for ulcers has been reported[6]. In the Gilbart study[22], all 10 DIP fusions united. Wounds healed without complications in 86% of patients, but one patient experienced postoperative ischemia and subsequent autoamputation of a fingertip.

Contracture. Some rigid, deformed digits benefit from MCP joint resection to overcome contracture, reposition the digit, and introduce a small range of mobility. Bone resection must be extensive because the soft tissues are stiff and stable. Risk of postoperative instability is low, and risk of persistent contracture is high. Some studies recommended capsulotomy or arthroplasty to correct for MCP hyperextension deformity, which provided a modest improvement in MCP range of motion, from less than 20° to an average of 50°[7,8,20]. Although range of motion may not be substantially improved through the MCP joint, the better position of the digit improves overall hand function[20,22]. In order to restore joint mobility, substantial shortening of the metacarpal is required[8,22].

Severe fixed finger-in-palm deformities can be treated with a combination of MCP joint excisional arthroplasty and PIP joint fusion, which involves resection of bone from both the distal metacarpal and the proximal phalanges[22]. This procedure substantially improved finger position and function, but produced axial shortening of the hand and a loss of the normal MCP joint contour.

Wire exposure. In a recent study of patients with complex hand manifestations of SSc, Kirschner wires utilized for IP joint fusion were easily removed 8 weeks after surgery in clinic, whereas figure-of-eight tension band wiring using steel suture sometimes required a more invasive procedure to remove the implanted hardware[22]. The steel suture loops became exposed as the skin in these patients gradually contracted 6 to 24 months after surgery, and the steel sutures are not as easily explantable in clinic. Gilbart, *et al*[22] reported that phalangeal bone stock was sufficient in most cases to anchor Kirschner wires, and figure-of-eight wires were not generally necessary. Nevertheless, they recommend adding a figure-of-eight wire if stability of fixation is uncertain. Melone, *et al*[8] also recommend the use of crossed Kirschner wires for surgical fixation, as well as tension-free wound closures.

Digital ulceration and hand vascularization. Roughly 31.8–71.4% (median 45.2%) of all patients with SSc experience recurrent digital tip ulceration at some stage of the disease[6,7,16,34,43]. Digital ulcerations are usually slow healing and, without early and aggressive medical intervention with vasodilators, may progress to gangrene and autoamputation. Tham and Grossman[34] reported that dry gangrene of the fingertip was present in one of 7 digits (14%) with chronic nonhealing ulceration, while Jones, *et al*[7] reported that 9 of 31 patients (29%) developed dry gangrene that required formal

surgical amputation of the digit, and another study noted "frequent" superficial gangrene[43]. Nevertheless, Gilbart, *et al*[22] reported that wounds healed without delay in 6 of 7 patients, and reduction of severe PIP joint flexion contractures helped reduce the frequency of skin ulceration. The seventh patient experienced ischemia of the fingertip following calcinosis removal and DIP fusion with correction from an extreme position of 120° of flexion, which led to necrosis and subsequent autoamputation of the fingertip. The possibility of tissue loss always exists when correcting severe digital deformities in patients with SSc, particularly in the more distal joints. In general, wet gangrene or osteomyelitis of a phalanx is an indication for amputation, whereas autoamputation is preferred in cases of dry gangrene to maximize the amount of residual viable tissue.

Vascular obstruction of the hand in SSc most commonly occurs in the ulnar artery and the proper digital arteries[44]. Microsurgical revascularization of the hand, digital arterial reconstruction, and peripheral or digital sympathectomy have been reported to improve digital vascular perfusion and heal digital ulcers and substantially relieve or eliminate pain from one to 46 months postoperatively in cases of severe distal and proximal arterial occlusion and digital vasospasm[25,27,28,30-32,34,37]. Healing of digital ulcerations generally occurs within 4 to 6 weeks[27,34,37,38]. However, some studies reported partial recurrence of ulceration, in 25% to 33% of patients[29,30,36,38] and a few cases of distal fingertip amputations[31,32,39]. In those cases where wound healing was reported to be slow, it was usually associated with large incisions; modification of the surgical technique to reduce the incision size in later procedures resulted in improved wound healing[29,36]. There was also one reported complication of reflex sympathetic dystrophy[26]. A comprehensive review of the outcomes of digital sympathectomy for chronic digital ischemia was recently completed by Kotsis and Chung[45]. Revascularization of ulnar artery occlusive disease has also been shown to dramatically improve Raynaud's phenomenon and healing of digital ulcers[33].

A trial of continuous regional anesthesia that restores blood flow and initiates healing of digital ulcers is reported to be an indicator for the effectiveness of peripheral sympathectomy; it also provides an effective treatment bridge until surgery[25]. Arteriography may be helpful in determining the status of digital vascularization when less invasive techniques such as Doppler examination or sympathetic blockade are not conclusive[6,9,27,31,33,34].

Profound arterial occlusion, especially of the ulnar artery, has been described in some patients with severe hand manifestations of SSc[7,9,27,33,34] and can be a cause of failure of surgery and extensive digit necrosis requiring amputation. In an unusual case of severe vascular deterioration, multiple partial amputations of 8 digits over a 10 year period were necessary due to gangrene, despite little other visceral involvement of SSc[9]. A case report of large vessel arterial thrombosis in SSc has suggested that antiphospholipid antibody syndrome may be a contributing factor in digital arterial insufficiency[46].

Medical therapies for vascular obstruction of the hand in SSc exist, but are beyond the scope of this article and have not been reviewed.

Calcinosis. Subcutaneous calcifications occur in the hands of 8.9% to 73.1% (median 44.1%) of patients with SSc[2-7,18,22,43,47-49]. They are associated with more severe Raynaud's phenomenon and frequent digital necrosis[5,48]. Calcinosis is more common in patients with SSc of more than 10 years' duration and with limited cutaneous disease[5,7,43]. Calcinosis is more frequently seen in patients with articular erosions than in those without (67% vs 39%, respectively)[2], particularly when the calcinosis is proximal to the MCP joints[5]. Calcification commonly occurs on the palmar side of the distal phalanges, causing pain, functional impairment, and in some cases ulceration[4,5,7,11,14,18].

Surgical excision of calcinosis provides moderate results with respect to pain relief and function. However, the procedure requires extensive incisions and carries a risk of slower wound healing, which may lead to skin necrosis and a possible reduction in range of motion[10,15-18]. Polio and Stern[17] report a case of intraneural calcification of the radial digital nerve of the index finger, resulting in 2-point discrimination greater than 20 mm on the radial side of the digit. Microsurgical excision of the calcinosis provided dramatic pain relief with no wound complications, although 2-point discrimination on the radial side of the digit remained greater than 20 mm.

Calcinosis can be effectively treated using a high-speed dental (micro-point) burr to break up calcium deposits that are then flushed out with saline[13,15,22]. This procedure requires only a small stab incision, which permits rapid wound healing (4–14 days), provides relief from pain and tenderness, and consequently improves function. Prolonged drainage of calcium deposits may occur[13].

A carbon dioxide laser has also been used to vaporize calcium deposits, with minimal bleeding and an average healing time of 4 to 10 weeks, resulting in a small scar[11,12]. Laser therapy provided complete or moderate resolution of pain in 81% of treated areas[11], with a concomitant improvement in function, as well as significant remission lasting at least 20 months[11] to 3 years[12].

Surgery of the hand for SSc is generally considered for pain reduction, severe fixed deformity with functional limitations, ulceration, and calcinosis. Both surgeon and patient must have a realistic expectation of the modest benefits of surgical procedures. The goals of surgery are limited and include pain relief through sympathectomy and increased perfusion, repositioning the digit, providing a functional position of fusion, and in some cases modest mobilization through resection arthroplasty, to marginally improve finger function for patients with marked preexisting limitations.

REFERENCES

1. Medsger TA Jr. Epidemiology of systemic sclerosis. Clin Dermatol 1994;12:207-16.
2. Baron M, Lee P, Keystone EC. The articular manifestations of progressive systemic sclerosis (scleroderma). Ann Rheum Dis 1982;41:147-52.
3. Bassett LW, Blocka KLN, Furst DE, Clements PJ, Gold RH. Skeletal findings in progressive systemic sclerosis (scleroderma). AJR Am J Roentgenolo 1981;136:1121-6.
4. Brun B, Serup J, Hagdrup H. Radiological changes of the hands in systemic sclerosis. Acta Derm Venereol 1983;63:349-52.
5. Doyle T, Littlejohn G, Miller M, Barnett A. The radiographic changes of scleroderma in the hands. Australas Radiol 1990;34:53-8.
6. Gahhos F, Ariyan S, Frazier WH, Cuono CB. Management of sclerodermal finger ulcers. J Hand Surg Am 1984;9:320-7.
7. Jones NF, Imbriglia JE, Steen VD, Medsger TA. Surgery for scleroderma of the hand. J Hand Surg Am 1987;12:391-400.
8. Melone CP Jr, McLoughlin JC, Beldner S. Surgical management of the hand in scleroderma. Curr Opin Rheumatol 1999;11:514-20.
9. Posner MA, Herness D, Green S. Severe peripheral vascular deterioration in scleroderma. A case report. Acta Orthop Scand 1980;51:239-41.
10. Berggren RB, Long PM, Trevaskis AE, Randall P. Calcinosis circumscripta: report of a case involving the hands. Plast Reconstr Surg 1965;36:609-18.
11. Bottomley WW, Goodfield MJ, Sheehan-Dare RA. Digital calcification in systemic sclerosis: effective treatment with good tissue preservation using the carbon dioxide laser. Br J Dermatol 1996;135:302-4.
12. Chamberlain AJ, Walker NPJ. Successful palliation and significant remission of cutaneous calcinosis in CREST syndrome with carbon dioxide laser. Dermatol Surg 2003;29:968-70.
13. Fahmy FS, Evans DM, Devaraj VS. Microdrilling of digital calcinosis. Eur J Plast Surg 1998;21:387-90.
14. Hussman J, Russell RC, Kucan JO, Khardori R, Steinau HU. Soft-tissue calcifications: differential diagnosis and therapeutic approaches. Ann Plast Surg 1995;34:138-47.
15. MacDowell F Jr. Digital involvement of extremities in scleroderma: method of treatment. NY State J Med 1969;69:935-7.
16. Mendelson BC, Linscheid RL, Dobyns JH, Muller SA. Surgical treatment of calcinosis cutis in the upper extremity. J Hand Surg Am 1977;2:318-24.
17. Polio JL, Stern PJ. Digital nerve calcification in CREST syndrome. J Hand Surg Am 1989;14:201-3.
18. Schlenker JD, Clark DD, Weckesser EC. Calcinosis circumscripta of the hand in scleroderma. J Bone Joint Surg Am 1973;55:1051-6.
19. Thurman RT, Jindal P, Wolff TW. Ulnar nerve compression in Guyon's canal caused by calcinosis in scleroderma. J Hand Surg Am 1991;16:379-81.
20. Lipscomb PR, Simons GW, Winkelmann RK. Surgery for sclerodactylia of the hand. Experience with 6 cases. J Bone Joint Surg Am 1969;51:1112-7.
21. Norris RW, Brown HG. The proximal interphalangeal joint in systemic sclerosis and its surgical management. Br J Plast Surg 1985;38:526-31.
22. Gilbart MK, Jolles BM, Lee P, Bogoch ER. Surgery of the hand in severe systemic sclerosis. J Hand Surg Br 2004;29:599-603.
23. El-Gammal TA, Blair WF. Digital periarterial sympathectomy for ischaemic digital pain and ulcers. J Hand Surg Br 1991;16:382-5.
24. Flatt AE. Digital artery sympathectomy. J Hand Surg Am 1980;5:550-6.
25. Greengrass RA, Feinglass NG, Murray PM, Trigg SD. Continuous regional anesthesia before surgical peripheral sympathectomy in a patient with severe digital necrosis associated with Raynaud's phenomenon and scleroderma. Reg Anesth Pain Med 2003;28:354-8.
26. Hafner J, Della Santa D, Zuber C, Christen Y, Bounameaux H. Digital sympathectomy (microarteriolysis) in the treatment of severe Raynaud's phenomenon secondary to systemic sclerosis. Br J Dermatol 1997;137:1011-31.
27. Jones NF, Raynor SC, Medsger TA. Microsurgical revascularization of the hand in scleroderma. Br J Plast Surg 1987;40:264-9.
28. Koman LA, Smith BP, Pollock FE Jr, Smith TL, Pollock D, Russell GB. The microcirculatory effects of peripheral sympathectomy. J Hand Surg Am 1995;20:709-17.
29. McCall TE, Petersen DP, Wong LB. The use of digital artery sympathectomy as a salvage procedure for severe ischaemia of Raynaud's disease and phenomenon. J Hand Surg Am 1999;24:173-7.
30. O'Brien BM, Kumar PA, Mellow CG, Oliver TV. Radical microarteriolysis in the treatment of vasospastic disorder of the hand, especially scleroderma. J Hand Surg Br 1992;17:447-52.
31. Ruch DS, Holden M, Smith BP, Smith TL, Koman LA. Periarterial sympathectomy in scleroderma patients: intermediate-term follow-up. J Hand Surg Am 2002;27:258-64.
32. Stratton R, Howell K, Goddard N, Black C. Digital sympathectomy for ischaemia in scleroderma. Br J Rheumatol 1997;36:1338-9.
33. Taylor MH, McFadden JA, Bolster MB, Silver RM. Ulnar artery involvement in systemic sclerosis (scleroderma). J Rheumatol 2002;29:102-6.
34. Tham S, Grossman JAI. Limited microsurgical arteriolysis for complications of digital vasospasm. J Hand Surg Br 1997;22:359-61.
35. Tomaino MM. Digital arterial occlusion in scleroderma: is there a role for digital arterial reconstruction? J Hand Surg Br 2000;25:611-3.
36. Tomaino MM, Goitz RJ, Medsger TA. Surgery for ischaemic pain and Raynaud's phenomenon in scleroderma: a description of treatment protocol and evaluation of results. Microsurgery 2001;21:75-9.
37. Tomaino MM, King J, Medsger T. Rationale for and efficacy of digital arterial reconstruction in scleroderma: report of two cases. J Reconstr Microsurg 2002;18:263-8.
38. Ward WA, Van Moore A. Management of finger ulcers in scleroderma. J Hand Surg Am 1995;20:868-72.
39. Yee AM, Hotchkiss RN, Paget SA. Adventitial stripping: a digit saving procedure in refractory Raynaud's phenomenon. J Rheumatol 1998;25:269-76.
40. Lovell CR, Jayson MIV. Joint involvement in systemic sclerosis. Scand J Rheumatol 1979;8:154-60.
41. Nalebuff EA. Surgery in patients with systemic sclerosis of the hand. Clin Orthop Rel Res 1999;366:91-7.
42. Rodnan GP, Myerowitz RL, Justh GO. Morphological changes in the digital arteries of patients with progressive systemic sclerosis (scleroderma) and Raynaud phenomenon. Medicine 1980;59:393-408.
43. Tuffanelli DL, Winkelmann RK. Systemic scleroderma. A clinical study of 727 cases. Arch Dermatol 1961;84:359-71.
44. Dabich L, Bookstein JJ, Zwifler A, Zarafonetis CJD. Digital arteries in patients with scleroderma: Arteriographic and plethysmographic study. Arch Intern Med 1972;130:708-14.
45. Kotsis SV, Chung KC. A systematic review of the outcome of digital sympathectomy for treatment of chronic digital ischemia. J Rheumatol 2003;30:1788-92.
46. Shapiro LS. Large vessel arterial thrombosis in systemic sclerosis associated with antiphospholipid antibodies. J Rheumatol 1990;17:685-8.
47. Misra R, Darton K, Jewkes RF, Black CM, Maini RN. Arthritis in scleroderma. Br J Rheumatol 1995;34:831-7.
48. Vayssairat M, Hidouche D, Abdoucheli-Baudot N, Gaitz JP. Clinical significance of subcutaneous calcinosis in patients with systemic sclerosis. Does diltiazem induce its regression? Ann Rheum Dis 1998;57:252-4.
49. Yune HY, Vix VA, Klatte EC. Early fingertip changes in scleroderma. JAMA 1971;215:1113-6.