

## **Hand Surgery in the Context of Scleroderma**

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### **ABSTRACT**

Patients with systemic sclerosis (SSc, scleroderma) offer challenges inherent in surgical interventions involving the hands, including poor circulation, compromised and inelastic dermis, constantly ulceration, and immunocompromised systemic status, constantly aggravated by immunosuppressive drugs. Despite these challenges, certain drawbacks that are affecting the hands of patients with CSS are best treated by surgery. Surgical methods in the hands of patients with SSc mainly aim to address the next complications: Digital ischemia, deformities of the interphalangeal (IP) and metacarpophalangeal joints (MCP), and Calcinosis cutis.

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### **INTRODUCTION**

Systemic sclerosis (SSc, scleroderma) is a connective tissue disorder with unclear aetiology. Patients with CSS have the possibility of experiencing significant pain and functional restrictions in damaged hands. The usual protests of the hand of SSc integrate joint contractures, ulcerations of the dermis, ischemia of the digital tip, Raynaud's phenomenon and calcinosis cutis. Surgery is usually reserved for patients in whom the conservative procedure has failed.<sup>1</sup>

### **INDICATIONS FOR SURGERY**

Patients with systemic sclerosis (SSc) and complications involving the hand are best managed by a team that includes a rheumatologist, an occupational therapist, and a hand surgeon. At first, everything feasible is done to preserve functionality, optimize perfusion and recover displacement without surgery. This is done with medication, using exercises to maintain the range of displacement and using appropriate splints. However, the non-surgical procedure to increase displacement is usually unsuccessful. Fertilization in the maximum corrected posture without undue pressure can delay the progression of the deformity. Impaired circulation, compromised dermis and immunosuppressed condition, properties of patients with SSc, show significant challenges for the surgical procedure.<sup>1-3</sup>

Digital ischemia: Almost all patients with SSc experience Raynaud's phenomenon, and nearly half of patients possess fingertip ulceration at some point in their clinical course. The aetiology of digital ischemia in CSS that impairs the hand is

possibly multifactorial and may involve Raynaud's phenomenon with elements of vasospasm, as well as fibrosis and narrowing of the intima, thrombosis and occlusion, and vascular compression by connective tissue. Early Raynaud's phenomenon may present as paroxysmal pallor of the finger, especially in states of frost or stress, followed by hyperemia. As the pathology progresses, patients have the possibility of experiencing painful ischemia and ulceration at their fingertips, which have the possibility of taking time to heal due to the compromised blood supply.<sup>1-3</sup>

Joint deformities: 3 hand deformities are commonly found in patients with reduced or diffuse EFS. The most recurrent head of deformity is a flexion contracture of the proximal interphalangeal joint (PIP) with compensatory hyperextension of the MCF joint. Hyperextension of the PIP joint (gooseneck deformity) and contracture of the dermis and muscle of the first web space are less frequently observed.<sup>1-3</sup>

Patients with systemic sclerosis (SSc) offer challenges inherent in surgical interventions involving the hands, including poor circulation, compromised and inelastic dermis commonly with ulceration, and immunocompromised systemic status commonly aggravated by immunosuppressive drugs. Despite these challenges, certain drawbacks that are affecting the hands of patients with CSS are best treated by surgery. These drawbacks integrate digital ischemia, joint deformities that limit the capacity of the hand or cause ulceration of the dermis and calcified deposits.<sup>1-3</sup>

## Hand Surgery in the Context of Scleroderma

Refractory or progressive ischemia procedure: The rules for surgical mediation integrate severe and disabling Raynaud's phenomenon, ischemic digital pain refractory to the medical procedure, and persistent ulcerations of the fingertips.<sup>1-3</sup>

Digital sympathectomy: The mainstay of the surgical procedure to improve digital circulation in SSc is the ablation of the sympathetic innervation of the digital vessels. This is achieved by stripping the adventitia of the target vessels, where the sympathetic nerves reside. Some techniques and locations for adventitious subtraction have been described. In what certain surgeons recommend the subtraction of adventitia as distally as it is at ease, to the degree of the own digital arteries in the finger and the usual digital arteries in the palm, other surgeons recommend operating more proximally, to the degree of the radial and ulnar arteries in the wrist and the palmar arch and the usual digital arteries in the palm. The expansion of the adventitious detachment ranges from 3 mm to 2 centimetres. Most surgeons remove at least 1 centimetre of the adventitia in the palm involving the usual digital arteries. The use of the surgical microscope is essential to do digital sympathectomy to ensure complete circumferential ablation of the adventitia over a contiguous length of the vessel without affecting the middle layer or the intima layer, which could predispose to thrombosis.<sup>1-3</sup>

Digital sympathectomy optimizes digital blood flow, relieves ischemic pain and optimizes the healing of ulcers on the fingertips. However, the effects of digital sympathectomy are usually short-lived and sympathetic reinnervation is generated with time. Adverse outcomes in patients who have undergone digital sympathectomy integrate delays in wound healing and development of infection. Progression or recurrence of pathology can lead to mutilation. However, at both 4 years of follow-up, more than 80% of patients report better pain control, fewer ulcerations, and a shorter healing time for ulcerations.<sup>1-3</sup>

There has been interesting in the use of botulinum toxin A for chemical sympathectomy as opposed to classical surgical periarterial sympathectomy. Botulinum toxin A is implied to inhibit sympathetic nerves, however, its precise mechanism of action is unknown. A total of 50 to 100 reconstituted units of botulinum toxin A are injected by hand, using a dorsal approach, in divided doses in the web spaces. The role of botulinum toxin A in the procedure of refractory or progressive ischemia secondary to Raynaud's phenomenon is examined in detail separately.<sup>1-3</sup>

Vascular recomposition: Patients with correctable vascular wounds, such as ulnar artery thrombosis, radial artery or palmar arch, have the possibility of benefiting from microvascular recomposition of the damaged vessel. Once a reconstructible lesion is assumed, preoperative angiography is effective in delineating arterial anatomy; however, if the diagnosis is the clear or once urgent correction of ischemia is warranted, it might be reasonable to abandon preoperative studies. Once angiography identifies a vessel susceptible to surgical recomposition, the damaged vessel is surgically

scanned, the segment of the artery that is thrombosed is identified and resected, and the artery is reconstructed with a microsurgical technique using an inverted vein interposition graft removed from the forearm. It should be noted that the resection of the damaged vessel grants an effective sympathectomy of that vessel in the advantage of the excision of the adventitious segment, which can also favour the procedure of ischemia. If constrictive fibrous tissue is identified around the vessel in question, arterial decompression can develop through the excision of constrictive connective tissue.<sup>3-6</sup>

Mutilation: Severe ischemia of the fingertips can lead to ulceration or gangrene, which may require mutilation. Despite the optimal medical procedure (e.g., local topical antimicrobials, measures to improve perfusion), ulcerations have the potential to be slow to heal or not cured at all. The tuft of the underlying distal phalanx may be reabsorbed, and if gangrene is eventually performed, self-amputation may occur.<sup>3-6</sup>

They have the possibility of passing secondary infections, more than 90 per cent of which are caused by *Staphylococcus aureus*. *Pseudomonas aeruginosa* is another viable culprit organism. Infections have the possibility of progressing to paronychia, panadizo, osteomyelitis of the distal or middle phalanx or pyogenic flexor tenosynovitis. In such scenarios, surgical mutilation to the degree of the distal interphalangeal joint (DIP) could be elementary. The existence of ischemia complicates the usual procedure of osteomyelitis (e.g., systemic antibiotics, debridement) and the conservative procedure is more likely to fail in patients with SSc.<sup>3-6</sup>

## CONCLUSION

Surgical treatment will be indicated in the cases described, to reduce the morbidity associated with scleroderma, the management of surgical techniques is the norm, preferring the referral of patients to specialized centres for the optimal management of them, as well as their complications.

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## Hand Surgery in the Context of Scleroderma

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