

Articular Surgery of the Ischemic Hand in Systemic Scleroderma: A Vascular Basis for Arthrodesis and Arthroplasty

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Hallmark deformities of systemic scleroderma are early onset and progressively disabling flexion contractures of the proximal interphalangeal (PIP) joints often in conjunction with extension or, less frequently, flexion contractures of the metacarpophalangeal (MCP) joints. Although surgical correction is generally recommended, a prevailing reluctance for operative treatment exists owing to the inherent ischemia of the disease with its potentially compromised healing capacity. Nonetheless, with recognition and preservation of the tenuous but well-defined and constant periarticular vascular networks of the PIP and MCP joints, articular reconstruction with uncomplicated wound healing can prove consistently successful for patients with scleroderma. This article describes the authors' preferred methods of PIP arthrodesis vascularized by the dorsal cutaneous arterial network and MCP silicone implant arthroplasty perfused by the dorsal metacarpal arterial plexus. (*J Hand Surg Am.* 2018; ■(■):1.e1-e9. Copyright © 2018 by the American Society for Surgery of the Hand. All rights reserved.)

Key words Scleroderma, diffuse systemic scleroderma, proximal interphalangeal joint arthrodesis, metacarpophalangeal joint silicone arthroplasty, intrinsic hand flaps.



INCREASINGLY ENCOUNTERED BY THE hand surgeon, scleroderma is a disabling autoimmune disease characterized by a diffuse fibro-occlusive vascular disorder, compounded by the usually coexistent vasospastic Raynaud syndrome. The resultant vascular deficiency leads to widespread ischemic fibrosis and soft tissue sclerosis that invariably afflicts the hands. In its most frequent and debilitating form,

termed diffuse systemic scleroderma (dSSc), classic features are progressive proximal interphalangeal (PIP) and metacarpophalangeal (MCP) joint contractures, apt to be the major source of patient pain and dysfunction (Fig. 1). Typically, the hand demonstrates severe PIP flexion deformities, often exceeding 90°, that lead to a predictable sequence of increased soft tissue tension over the dorsal aspect of the joints, diminished circulation, and ultimately, painful dorsal ulceration.¹⁻⁴ Simultaneously, the MCP joints are prone to reciprocal extension or, less frequently, flexion contractures that markedly limit digital mobility and contribute to further functional and aesthetic decline.

Despite the serious functional and psychological consequences of these deformities, and the usual recommendation for digital joint reconstruction, techniques of articular surgery for scleroderma are infrequently reported, and thus, there are few published guidelines for management of the ischemic

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tissues. This article describes the authors' techniques of articular reconstruction, specifically aimed at the recognition, usage, and preservation of consistently present digital arterial systems. We had success in treatment of 80 dSSc patients over a 10-year period from 2004 to 2014. For the PIP joint, the dorsal cutaneous artery (DCA) network provides a sound vascular basis for arthrodesis employing an arterial perfused dorsal bipedicle flap (Fig. 2),⁵ and for the MCP joints, the dorsal metacarpal artery (DMA) system supplies an ample source of vascular perfusion facilitating implant arthroplasty (Fig. 3).

INDICATIONS

Proximal interphalangeal joint arthrodesis

Considered by most surgeons as the keystone of surgical treatment for the hand in dSSc,^{3–10} arthrodesis is indicated for correction of disabling PIP joint flexion contractures, often associated with painful and infected dorsal ulcerations. In our experience, arthrodesis and soft tissue closure employing the vascularized dorsal bipedicle flap has consistently resulted in solid fusion of the afflicted joint in a more functional extended posture with elimination of the painful ulcers. The successful fusion also creates a barrier to osteolysis, recurrent infection, and other detrimental effects of the progressive disease process.

At present, the authors recommend arthrodesis for painful progressive flexion contractures exceeding 60°. At this stage of deformity, further progression is usually inevitable with resultant increase in soft tissue tension and ischemic attrition over the dorsal aspect of the joint. The continuous vascular compromise poses a serious threat to tissue viability and an increasing obstacle to wound healing. Timely arthrodesis facilitates operative treatment and lessens patient morbidity.

Silicone MCP implant arthroplasty

Silicone MCP joint arthroplasty is indicated for irreversibly contracted MCP joints that considerably contribute to disability by limiting the major arc of digital motion and severely impeding grip and pinch functions.⁸ The procedure usually is recommended as a second stage of articular reconstruction for those patients who previously benefited from PIP joint arthrodesis. In cases with pain-free stable PIP joints, MCP silicone arthroplasty substantially alleviates the discomforting taut dorsal soft tissue, consistently improves digital mobility, and considerably lessens the disfiguring claw hand deformity of dSSc (Video A; available on the *Journal's* Web site at

www.jhandsurg.org). Even a modest increase in mobility is likely to effect a major functional improvement, and a seemingly minimal cosmetic improvement in digital posture can markedly enhance patient self-esteem.^{8,9}

In contrast to the more traditional techniques of MCP reconstruction, such as resection arthroplasty^{2–4,6,11–13} and MCP capsulotomy,^{7,10,14} silicone arthroplasty has reliably maintained digital length with preservation of a flexible and durable joint space and a functional arc of motion.

CONTRAINDICATIONS

Foremost among contraindications to articular surgery in dSSc is a medically unstable patient. Patients with unremitting disease and deteriorating health, usually associated with vital organ dysfunction, require intensive medical management and are not candidates for elective procedures.

Another major deterrent to surgery for this immunodeficient group is active infection. Soft tissue or skeletal sepsis is apt to be progressive and result in a digital, limb, or occasionally, systemic life-threatening situation. Prompt and thorough infectious disease management is essential to eradicate or control the septic process prior to consideration for surgery. For the frequently encountered superficial dorsal ulcer infection of the deformed PIP joint, short-term antibiotics and wound care are usually sufficient preparation for elective arthrodesis coupled with excision of the offending ulcers. In these cases, a solid arthrodesis with thorough wound healing is the optimal means of preventing these painfully recurrent episodes of digital infection.

SURGICAL ANATOMY

Owing to the diffusely ischemic nature of systemic scleroderma, awareness and preservation of the vital periarticular vascular anatomy is critical to successful small joint reconstruction. Discreet, constantly present periarticular arterial networks have been described by numerous investigators at both the PIP^{15–17} and the MCP^{18–21} joint levels and serve as reliable vascular foundations for uncomplicated arthrodesis and arthroplasty.

The DCA network arising from the proper digital arteries (PDAs) is the basis for the dorsal bipedicle flap employed for exposure and arthrodesis of the PIP joints (Fig. 2). Originating from the radial and ulnar PDAs, 2 or 3 dorsal cutaneous branches emerge bilaterally at specific distances both proximal and distal to the joint and converge dorsally over the

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