58 Articular Surgery for the Scleroderma Hand: Arthrodesis and Arthroplasty

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Abstract
Classic features of systemic scleroderma are disabling and disfiguring proximal interphalangeal (PIP) and metacarpophalangeal (MCP) joint contractures of the hands. Although surgical correction has often been recommended, seldom has it been employed due to concerns for devitalized wound and compromised healing capacity inherent in the disease process. Nonetheless, despite characteristically ischemic and precarious wound conditions, carefully planned and executed arthrodesis and arthroplasty can prove considerably beneficial to medically stable patients with systemic scleroderma. In the authors' experience, techniques of PIP arthrodesis based on dorsal cutaneous arterial (DCA) network and MCP silicone arthroplasty perfused by the dorsal metacarpal collateral artery (DMCA) plexus have consistently demonstrated uncomplicated wound healing and considerably improved function.

Keywords: arthrodesis, arthroplasty, arthropathy, diffuse cutaneous systemic sclerosis, scleroderma, systemic sclerosis

58.1 Introduction

Systemic scleroderma is a disabling autoimmune disease characterized by an intrinsic fibro-occlusive vascular disorder resulting in extensive tissue ischemia and sclerosis that typically afflict the hands. Raynaud's vasospastic syndrome, invariably associated with the disease, further contributes to the vascular disturbance. In its most frequent and debilitating form, termed diffuse systemic scleroderma (dSSc), classic features are progressively severe proximal interphalangeal (PIP) joint and metacarpophalangeal (MCP) joint contractures, apt to be the major source of painful dysfunction (▶ Fig. 58.1).

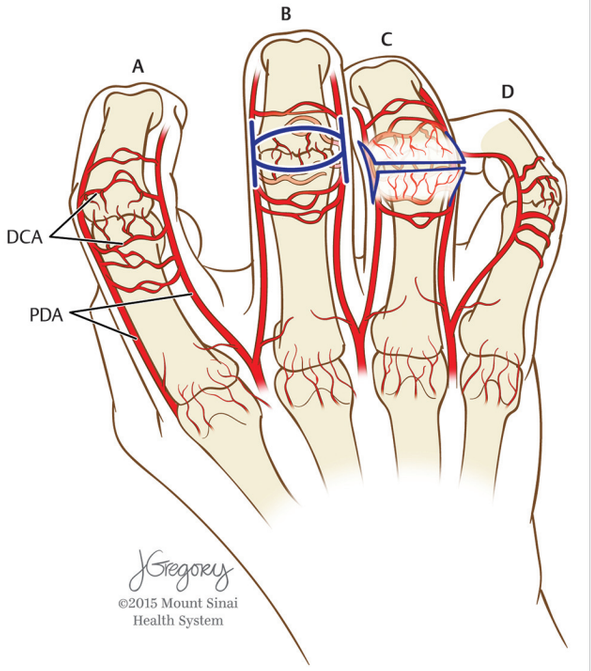
Fig. 58.1 Scleroderma hand deformity classically involves extension contractures of the metacarpophalangeal (MCP) joints and flexion contractures of the proximal interphalangeal joints with vascular compromise of adjacent soft tissues.

Although articular reconstruction is generally recommended for these deformities, surgical techniques are infrequently reported and the literature provides minimal guidance for optimal soft tissue and skeletal management. This chapter describes our experience with specific techniques designed to preserve wound vascularity, promote uncomplicated wound healing, and enhance outcomes for PIP and MCP articular reconstruction.

58.2 Indications

For patients with this high-risk diffuse disease process, only those in optimal medical condition are considered for articular surgery. Elective surgery for poorly managed patients with deteriorating health is prone to failure and ill advised. Progressive flexion contracture of the PIP joint, often associated with painful and infected dorsal ulceration, is the hallmark deformity of scleroderma, and the principal indication for reconstructive surgery. The PIP flexion contracture often exceeds 90 degrees, creating excessive tension with severe ischemia of the overlaying dorsal soft tissue that results in a predictable sequence of attenuation, atrophy, and painful ulceration. Previously the deformed PIP joints have been managed by a variety of techniques, including capsulotomy[1], implant arthroplasty,[1] and arthrodesis.[1,2,3,4,5,6,7,8] Because of the deficient, sclerotic soft tissues, and insufficient bone stock, attempts to preserve PIP joint mobility are prone to failure and thus a consensus exists that arthrodesis is the optimal treatment for the dysfunctional PIP joint.[2,3,4,6,7,8,9] The PIP arthrodesis performed by the authors employs specific flap designs for unobstructed exposure, preservation of wound vascularity, and tension-free closure, and is based on consistent proximal and distal dorsal cutaneous arterial branches (DCAB) of the radial and ulnar proper digital arteries (▶ Fig. 58.2).[10] Consistently high fusion rates resulting in a more extended joint posture markedly improve digital function and alleviate dorsal soft tissue tension with its painful ulceration. Moreover, once solid fusion has occurred, the reconstructed joint is relatively protected from osteolysis, secondary infection, pathological fracture, or other detrimental effects of this progressive disease.

MCP joint contractures, usually fixed in hyperextension but occasionally in flexion, are also typical of progressive dSSc, and severely limit both grasping and releasing functions of the hand. In an attempt to improve both functional posture and mobility of the MCP joints, and since soft tissue and skeletal structures are more substantive at this level, the senior author has exclusively employed silicone implant arthroplasty for these deformities. This method has consistently corrected the deformities, alleviated pain, and restored functional mobility of the MCP joints. The characteristic presence of sclerotic collateral soft tissues and the typical absence of recurrent synovitis seemingly enhance both stability and durability of the implants.



PDA: Proper digital artery; **DCA**: Dorsal cutaneous artery

Fig. 58.2 Flap design for proximal interphalangeal (PIP) arthrodesis with apposing dorsal pedicles based on the consistent proximal and distal dorsal cutaneous arterial branches of the radial and ulnar proper digital arteries. Judicious flap elevation provides thorough joint exposure and preserves the delicate cutaneous vasculature. Printed with permission from ©Mount Sinai Health System.

Fig. 58.3 Markings for elliptical excision of devitalized soft tissues and for the apposing bipedicle advancement flaps to expose and resurface the affected proximal interphalangeal (PIP) joints.

58.3 Operative Techniques

For all cases of articular surgery, basic tenants for success are:

Tourniquet control with a bloodless field; balanced regional anesthesia with its vasodilatory effect and its avoidance of intubation; a warm operating environment to limit Raynaud's syndrome; and a tension-free wound closure.

58.3.1 PIP Arthrodesis

The dorsal soft tissues over the contracted PIP joint are commonly attenuated and ulcerated. This devitalized soft tissue is carefully delineated and excised in a transverse elliptical fashion. For thorough joint exposure, opposing dorsal bipedicle flaps are designed in an H-shaped configuration with the central transverse limb directly over the joint and bilateral vertical limbs extending axially along the radial and ulnar aspects of the joint from the proximal phalanx to the middle phalanx. This design creates two pedicles, each with a 2:0.5–1 width to length ratio that facilitates wide exposure with minimal flap undermining, ease of flap advancement, tension-free closure, and preservation of the proximal and distal dorsal cutaneous arterial vasculature (▶ Fig. 58.3). In contrast to the other types of incisions, this DCAB based bipedicle flap has been designed specifically for exposure and coverage of the reconstructed PIP joints in scleroderma. The symmetrical vascularity of the dual pedicles has proved consistently sufficient to offset the characteristic digital ischemia and promote uncomplicated soft tissue healing. For patients afflicted with the inherent vascular deficiency of scleroderma, the dorsal bipedicle flap has provided a reliable method of exposing and resurfacing the devitalized arthrodesis site.

Elevation of the dorsal flaps invariably reveals severe soft tissue fibrosis with an atrophied extensor mechanism firmly adherent to the underlying bone. The joint is characterized by deformation and subluxation of contiguous articular surfaces. The phalangeal head at the proximal aspect of the joint demonstrates resorption and displacement into a prominent dorsal position whereas the adjacent base of the middle phalanx is abnormally flat, wide, and displaced volarly and proximally into a position of hyperflexion (▶ Fig. 58.4). This continuous joint flexion contracture contributes to progressive bony deformity, resorption, and loss of subchondral cancellous bone. Notably, and in contrast to other arthropathies (i.e., rheumatoid arthritis), these cases of advanced joint destruction do not have synovitis as a prominent finding.

Correction of the deformity requires excision of the phalangeal head as well as adjacent and severely contracted collateral ligaments. Judicious skeletal shortening preserves the critical subchondral cancellous bone and permits precise coaptation of the cancellous surfaces at the fusion site, while alleviating tension on the dorsal skin (▶ Fig. 58.5). Position of the fusion is determined by assessing the functional requirements of individual digits, the pre-existing deformity, and the extent of critical cancellous bone.

In order to prevent excessive volar tension and serious vascular compromise overcorrection of the flexion deformity is avoided by joint positioning in functionally and cosmetically improved flexion postures of 40 to 50 degrees.

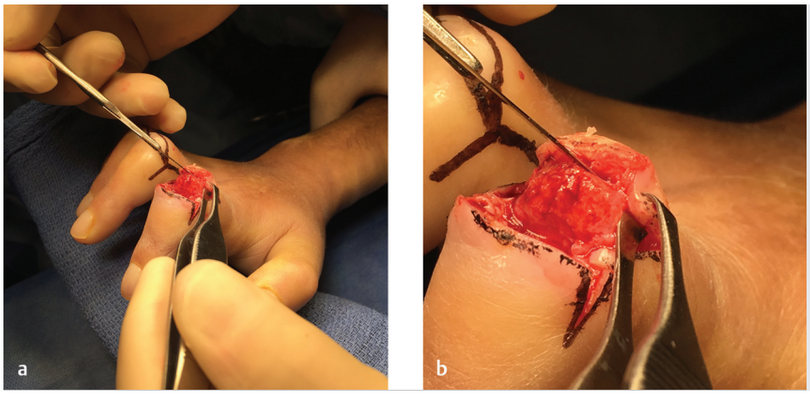


Fig. 58.4 (a, b) Elevation of advancement flaps for access to proximal interphalangeal (PIP) joint, bony shortening, correction of deformity, and preparation of cancellous fusion site.

Fig. 58.5 Proximal interphalangeal (PIP) arthrodeses secured with minimally invasive K-wire fixation demonstrating improved joint position with tension-free flap closures.

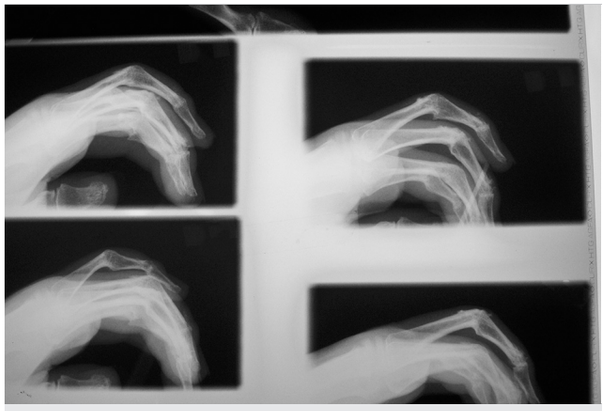


Fig. 58.6 Radiographic evidence of solid proximal interphalangeal (PIP) joint arthrodesis 8 weeks postoperatively.

In the early phase of the senior author's practice, arthrodeses were secured by a combination of intraosseous and K-wires. However, with increasing experience, the use of percutaneous K-wires alone proved a consistently reliable method of secure fixation. This allows for rapid and accurate insertion with minimal trauma, firm stabilization, and easy removal. Fine crossed K-wires are now the preferred method of fixation.[2] Postoperative wound care with continuous splinting is utilized until radiographic documentation of solid fusion is evident (▶ Fig. 58.6).

58.3.2 PIP Joint Arthrodesis Outcomes

In a review of over 100 PIP arthrodesis for scleroderma, our follow-up evaluation typically revealed uncomplicated primary wound healing and consistent radiographic union of the arthrodesis within 8 weeks of surgery. Patients also demonstrated complete eradication of painful ulcers, substantial functional and aesthetic improvement, and high levels of satisfaction.

58.3.3 MCP Implant Arthroplasty

For those hands requiring both PIP and MCP reconstruction, the PIP joints are fused in positions of improved function as a first stage, and MCP joints are reconstructed by silicone arthroplasties as a second stage. The destroyed MCP joints are characterized by excessive extension or fixed flexion with narrowed joint spaces and flattened as well as eburnated cartilaginous surfaces. Similar to the PIP joints and in contrast to other arthropathies, erosive changes are minimal and proliferative synovitis is not encountered.[2,11,12,13,14]

In each case of MCP arthroplasty a transverse incision just proximal to the metacarpal heads is used with minimal soft tissue reflection and preservation of the critical dorsal vasculature (▶ Fig. 58.7). The transverse incision is well vascularized by the DMA network and provides exposure of all four joints simultaneously. In contrast to MCP arthroplasty for other conditions, no attempt is made to preserve or reconstruct the collateral ligaments; rather, a wide resection of the metacarpal head and condyles, including the collateral ligaments, corrects the deformity and achieves relative lengthening of the chronically contracted extensor and flexor tendons. This wide resection achieves sufficient bony shortening to reconstruct a sizeable joint space for implant insertion for restoration of functional tendon excursion and joint mobility. This skeletal resection also facilitates a tension-free primary wound closure.

Arthroplasties are managed by early and intensive therapy initiated on postoperative day 3 to 4 (▶ Fig. 58.8). The program begins with active motion and dynamic extension splinting followed by passive flexion cuffs until implant encapsulation is complete and maximum mobility achieved, a period usually encompassing 4 to 6 months.

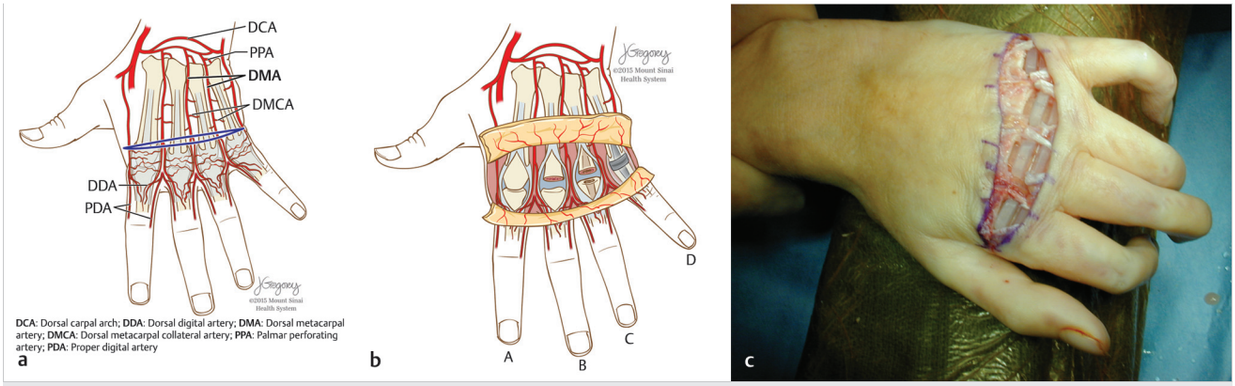


Fig. 58.7 (a, b) The transverse dorsal incision, well perfused by the dorsal metacarpal arterial system, affords wide exposure for implant arthroplasty of all four metacarpophalangeal (MCP) joints simultaneously. Printed with permission from ©Mount Sinai Health System. **(c)** Correction of joint deformity and restoration of functional MCP joints with silicone implant arthroplasties. The extensor tendons are preserved and relatively lengthened for repair.

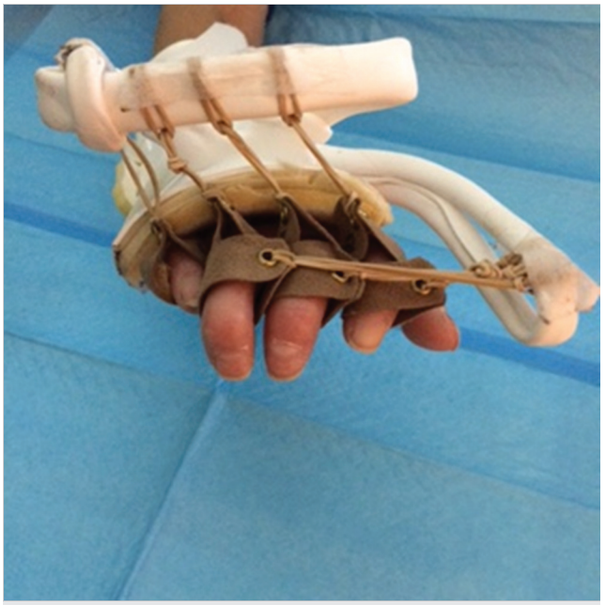


Fig. 58.8 Postoperative dynamic extension splint facilitates recovery of metacarpophalangeal (MCP) joint mobility.

Fig. 58.9 Maintenance of well-aligned and functional metacarpophalangeal (MCP) joints with silicone implants 10 years postoperatively.

58.3.4 MCP Implant Arthroplasty Outcomes

The silicone implant arthroplasties have proved highly functional, having maintained MCP joint stability and restored substantial joint mobility usually in the range of a 50-degree flexion extension arc. Moreover the implants have demonstrated durability with preservation of pain-free function and with no evidence of implant failure for as long as 10 years after insertion (▶ Fig. 58.9).

58.4 Conclusion

Scleroderma is a debilitating systemic disease that entails the classic hand deformity, including progressively severe PIP joint and MCP joint contractures. This chapter described our experience with specific techniques designed to preserve wound vascularity, promote uncomplicated wound healing, and enhance outcomes for PIP and MCP articular reconstruction.

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