

Surgery for Jaccoud Arthropathy

A Systematic Review

Willer D. Santos, MS,* Carla Baleeiro, MS,* and Mittermayer Barreto Santiago, MD, PhD*†

METHODS

Introduction: Jaccoud arthropathy (JA) is characterized by the presence of joint deformities similar to those seen in rheumatoid arthritis but generally with a “reversible” pattern. The etiopathogenic mechanisms are not known, and its therapeutical approach has been regarded as disappointing. The aim of the present study was to perform a systematic review of the literature on the scientific evidence of the surgical procedures for JA.

Methods: The MEDLINE, LILACS, and Scielo databases were searched using the following keywords: “systemic lupus erythematosus,” “rheumatic fever,” “Jaccoud arthropathy,” “deforming arthropathy,” “surgery,” and their corresponding terms in Portuguese and Spanish. The search period was between 1966 and 2014.

Results: Only 7 articles fulfilled the inclusion criteria, and a total of 58 patients underwent surgical procedures for JA. Such studies were limited to small case series, there was no uniform definition of the outcome, and the follow-up time varied largely.

Conclusions: There is no consensus on the best approach for the surgical procedures in patients with JA, who are the best candidates to undergo this, and when to indicate the procedure.

Key Words: Jaccoud arthropathy, surgery, systematic review

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Jaccoud arthropathy (JA) was first described in the 19th century by S. Jaccoud¹ in a patient who had had several attacks of rheumatic fever (RF). This arthropathy has also been described in patients with systemic lupus erythematosus (SLE), an estimated prevalence of 5%, and less frequently in other clinical conditions.^{2–5} The classic manifestation of JA is the presence of “reversible deformities,” that is, reducible to passive movement with preservation of the joint structure when evaluated by plain radiography.⁶ Jaccoud arthropathy generally affects the hands, but can also be observed in feet,⁷ knees,^{3,4} and shoulders.⁸ The most common joint deformities are ulnar deviation, “swan neck,” “boutonniere,” thumb in Z and hallux valgus.⁶ Curiously, in older deformities, a limitation of the joint motion can be observed, probably because of fibrosis secondary contractures of the soft tissues of the joint.⁹ In addition, it was observed that the JA can be associated with an increased risk of tendon rupture.¹⁰

At present, the therapeutic approach for JA is similar to the patients with SLE and joint manifestations, regardless of the presence of this complication. The objective of this review was to examine the evidence with respect to the surgical procedures utilized in the corrections of deformities of JA.

From the *Escola Bahiana de Medicina e Saúde Pública and †Serviços Especializados em Reumatologia da Bahia, Salvador, Bahia, Brazil. M.B.S. is currently receiving a scholarship from Conselho Nacional de Desenvolvimento Científico e Tecnológico (CNPq).

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Correspondence: Mittermayer Barreto Santiago, MD, PhD, Serviços Especializados em Reumatologia da Bahia, Rua Conde Filho 117, Graça, Salvador, Bahia, Brazil CEP 40150-150.
E-mail: mbsantiago2014@gmail.com.

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(a) Inclusion/exclusion criteria of the studies: This review included all articles describing the surgeries in patients with JA. There were no restrictions with regard to the language and the year of publication. Review articles were excluded from the study.

(b) Search strategy: The MEDLINE, LILACS, and Scielo databases were searched using the following keywords: “systemic lupus erythematosus,” “rheumatic fever,” “Jaccoud’s arthropathy,” “deforming arthropathy,” “surgery,” and the corresponding terms in Portuguese and Spanish. The search period was between 1966 and 2014. Additional secondary references were also obtained.

Methodology for Reviewing

The data collected included study design, number of patients described, and the procedure performed.

RESULTS

Only 8 articles that fulfilled the previously established criteria were obtained from the aforementioned databases and secondary references. All the articles were case reports or case series that described several surgical procedures performed on a total of 62 patients. One article was excluded from the analysis because the authors did not describe details of the surgery performed in 4 of 7 SLEs with deforming arthropathy.¹¹ Thus, a total of 7 articles and 58 patients were included.

The details of the surgical procedures are presented in the following section and have been summarized in the Table 1.

Surgery for Hand Deformities of JA

Between 1965 and 1978, Dray et al¹⁷ performed 51 operations on 10 SLE patients with deformities in their hands, with a mean arthritis duration of 14.3 years. The goal of this treatment was to improve the flexion of the metacarpophalangeal (MCP) joints. For those cases with “reversible” deformities, the surgical methods included relocation of the extensor tendons to the dorsal aspect of the MCP joint capsule, imbrication of the radial side of the joint capsule, and release of the ulnar intrinsic tendon. There was “treatment failure” in 70% of the corrections, and some cases had worsening of the original deformity or even led to an irreversibility of the movements. For fixed deformities and for those cases that experienced “treatment failure,” an MCP joint arthroplasty was performed in 33 digits (17 for “treatment failure” and 16 for fixed deformities). Thirteen of 17 of the arthroplasty procedures in the “treatment failure” group yielded good results, that is, mild deformity and an increase of 50 degrees in the arc of flexion in up to 73 months of follow-up. On the other hand, arthroplasty performed on those joints with fixed flexion deformity provided only “fair” results in half of the operated joints. Surgeries were also performed for deformities of the thumb. A joint fixation with the Kirschner wire (2 joints operated both had recurrence of the deformities) or ligamentous reconstruction (success in 3 of 4 joints

TABLE 1. Main Characteristics of the Published Studies on Surgery for Patients With JA

First Author (Reference)	Year	Origin	Studied Patients	Gender	Mean Age, y	Mean MDD	Deformity	Surgery	Results
Schumacher et al ⁹	1976	USA	One SLE	F	33	14	Ulnar deviation at MCP joints and swan neck deformities	MCP and PIP joints soft tissue procedure MCP arthroplasty and thumb arthrodesis	Recurrence after soft tissue procedure, but restoration of ability to pinch and grip after arthroplasty and arthrodesis
Evans et al ¹²	1977	Canada	Three SLE	F	35	13.3	Ulnar deviation at MCP joints; swan neck and boutonniere deformities; thumb subluxation	MCP soft tissue procedure Metacarpal osteotomy	All joints recovered the extension and recovered 70 to 90 degrees of flexion. Metacarpal shaft rotation was controlled and all 6 hands had improvement in function
Dray et al ¹³	1981	USA	10 SLE	NA	NA	NA	MCP joints of the fingers Passively correctable flexion deformity (after failure of extensor tendon relocation) Fixed flexion deformity CMC joint of the thumb Passively correctable instability without degenerative changes of the thumb Fixed dislocation	ETR MCP arthroplasty MCP arthroplasty Reduction CMC joint with K-wire fixation Reconstruction of CMC joint ligaments CMC fusions Modified trapezium arthroplasty	70% recurrent deformity 76.4% average further arc of 50-degree flexion 50% average further arc of 30-degree flexion 100% failure 75% painless and located, 25% failure 75% painless, 25% failure 100% painless and located
Joseph and Chacko ⁷	1984	India	One RF	M	18	NA	Medial deviation at MTP joints ^a	MTP soft tissue procedure Metatarsal osteotomy	100% correction of the deformity
Wood et al ¹⁴	1989	USA	Two SLE	F	51.5	43	Ulnar deviation at MCP joints	MCP soft tissue procedure	Ulnar deviation correction and reasonable results on strength
Alnot et al ¹⁵	2004	France	16 IJA 10 SLE 5 MCTD 2 RF 2 SS 2 PSO 3 CCA	NA	NA	NA	Reversible ulnar deviation Irreversible ulnar deviation Reversible ulnar deviation + swan neck and/or boutonniere Irreversible ulnar deviation + swan neck and/or boutonniere	ETR MCP arthrodesis MCP arthroplasty with Swanson implant Metacarpal osteotomy MCP arthrodesis Extensor tendon relocation MCP arthrodesis ETR + MCP arthroplasty Metacarpal osteotomy	79% efficacy 100% efficacy 66.6% efficacy Reasonable results 100% efficacy 80% efficacy 100% efficacy 50% efficacy Reasonable results
Waguri-Nagaya et al ¹⁶	2009	Japan	One SLE	F	43	12	Medial deviation at MTP joints and hallux valgus ^a	Metatarsal osteotomy MTP joints soft tissue procedure Hallux arthroplasty	Good foot alignment, but foreign body reaction and return of hallux deformity after 2 y

^aPresence of deformities in feet.

CCA indicates chondrocalcinosis; CMC, carpometacarpal; DIP, distal interphalangeal; ETR, extensor tendon relocation; IP, interphalangeal; F, female; IJA, idiopathic JA; M, male; MCTD, mixed connective tissue disease; MDD, mean disease duration; NA, information not available; PIP, proximal interphalangeal; PSO, psoriasis; SS, Sjögren syndrome.

operated) was performed when the deformity was passively correctable and without degenerative changes. When joint dislocation was irreducible or when the subluxation was passively correctable but accompanied by degenerative changes, either joint fusion or arthroplasty with modified trapezium prosthesis was performed. Joint fusion was effective in 3 of 4 operations, with pain relief and correction of the deformity. The arthroplasty was satisfactory in the 2 cases for which they were used. In both cases, the pain was alleviated, and motion was restored to a very functional level. In 6 of their 10 patients, there was a severe carpal collapse, and they excised the distal ulna in 4 and inserted a Silastic prosthesis in one with "satisfactory alleviation of the pain." Steinmann pin fixation was performed on 2 wrists, but there was recurrence in one of them after 3 months only.

A Canadian group^{12,13} studied 3 patients with SLE who developed severe, persistent, and deforming hand arthritis. They had MCP joint subluxation and/or dislocation. In addition, 2 had thumb subluxation. One also had swan neck deformity, and another showed boutonniere deformity. For the correction of the MCP joint deformities, the surgery was divided in the following steps: (1) synovectomy of the residual synovial membrane, (2) release of the ulnar intrinsic tendon, (3) release of the volar plate and the collateral ligaments, (4) metacarpal osteotomy and shortening of the bone with stabilization through intramedullary pins, wires, or screws, (5) centralization of the extensor tendons, and (6) transfer of the intrinsic tendons to inhibit the ulnar deviation. All 24 joints (6 hands) recovered the extension and achieved 70- to 90-degree flexion. The authors concluded that there is no justification for performing arthroplasty in JA of the hands, because there is no bone erosion, and the bone and joint architecture is intact enough for soft tissue reconstruction.

Wood et al¹⁸ performed hand surgeries in 2 SLE patients. The patients were 39 and 67 years old, and the disease duration was 18 and 25 years, respectively. These patients were submitted to the MCP soft tissue reconstruction for ulnar deviation and correction of the subluxation. The surgical procedure was similar to that described by the Canadian group, but osteotomy was not performed. The surgery was divided into the following steps: (1) incision of the tendon junction, (2) opening the capsule for removal of the residual synovial membrane, (3) release of the volar plate, (4) release of the intrinsic ulnar tendon and transfer to the adjacent radial collateral ligament insertion, (5) centralization of the extensor tendons, and (6) fixation of the MCP joints with Kirschner wire in extension and its removal after 3 weeks. The follow-up time was up to 3 years. Ulnar inclination was partially corrected, but the grip strength and pinch did not improve significantly after surgery. The patients scored the result as "good" or "excellent," but one of the patients presented with finger radial deviation after 1 year of follow-up.

Alnot et al¹⁴ performed surgery on 41 hands, of which of 40 patients were with JA, 16 with idiopathic JA, 24 with JA secondary to rheumatic diseases (10 SLE, 5 Sharp syndrome, 3 chondrocalcinosis, and 2 RF, 2 Sjogren syndrome, and 2 psoriasis). The surgical techniques were chosen in accordance to the deformities presented by the patients and were divided into 4 groups. Groups I and II had reversible and irreversible ulnar deviation, respectively. Groups III and IV were more complex cases with ulnar deviation, swan neck, or boutonniere deformities. Group III fell under the "reversible" deformities category, and group IV under "fixed" deformities. They classified the results of the surgical procedures as "very good," "good," "reasonable," and "failure" based on the deformity, strength, and discomfort. The patients were followed up to 12 years. In 19 hands of group I, the surgery included "stabilization" of the MCP joints and realignment of the extensor apparatus, and the initial results were

considered positive in 15 of them. Two reoperations were required. Arthrodesis was performed in 3 MCP joints with similar positive results. In 5 of 11 operated hands from group III, the stabilization of the MCP joints and realignment of the extensor apparatus yielded 80% efficacy. In groups II and IV with irreversible deformities, the MCP joint arthroplasty with the Swanson implant had 66% efficacy. Three metacarpal osteotomies were performed with reasonable results.

On the other hand, Schumacher et al⁹ failed to demonstrate the benefit of the soft tissue reconstructive surgery for correction of joint deformities (ulnar deviation and swan neck) on their patient who was a 33-year-old housewife who has lupus. Although an initial improvement was noticed, there was recurrence of the deformities a few months after the first soft tissue surgery. She was submitted to another corrective soft tissue surgery, but the deformities recurred. The first soft tissue procedure was as follows: "The swan neck deformities of the fingers were corrected by the volar placement of a part of the lateral bands." The subluxation in the MCP joints of the thumb and index finger was reduced and fixed with a "K" wire, which was removed later. "The dorsal joint capsules were plicated, and the extensor tendons advanced so that the corrected joint position would be maintained after the K-wire removal." The second procedure was the relocation of the extensor tendons of all digits on the dorsum of the MCP joints from their ulnar positions and the plicated dorsal capsules. Metacarpophalangeal joint arthroplasty with Silastic implant-type Swanson was then indicated, and the patient reported significant improvement in the joint appearance and function with return of her ability to pinch and grip. The follow-up time was 3 years.

Surgery for Foot Deformities in JA

Waguri-Nagaya et al¹⁵ studied a 43-year-old patient with SLE for 12 years, who developed JA in the feet characterized by hallux valgus and medial dislocation of the metatarsophalangeal (MTP) joints. The patient was treated by arthroplasty of the first joint and stabilization of the second to the fifth MTP joints utilizing the intramedullary Kirschner wires that were removed after 2 weeks. The patient was satisfied with the results of the foot alignment. However, after 2 years of the surgery, a bone erosive lesion around the silicone implant resembling a foreign body reaction and subluxation of the first metatarsal-cuneiform joint, without bone erosion, were observed through plain radiography.

Joseph and Chacko⁷ described an 18-year-old man with JA secondary to RF, who had symmetrical and fixed deformities of the feet with medial dislocation of the MTP joints. The surgery consisted of an osteotomy of the metatarsal heads, tenotomy of the long flexor tendons, and stabilization of first MTP with Kirschner wires. They obtained total correction of the deformity.

DISCUSSION

As the etiopathogenic mechanisms for JA are not known, it is difficult to establish a strategy to avoid its development or progression. At present, in the case of SLE with established deformities typical of JA, the management does not differentiate from the cases with arthritis without JA, which includes physical therapy, anti-inflammatory drugs, corticosteroids, hydroxychloroquine, and methotrexate. An area open for future research deals with the identification of a specific genetic background or a plasma biomarker that could allow preventive measures to be implemented before the development of such deformities. Recent observations indicate a higher serum level of interleukin 6 in patients

with JA secondary to SLE, but there is no study demonstrating the benefit of therapy targeting this cytokine in JA.¹⁶

One should be aware that the spectrum of JA is broader than the typical scenario of mild “reversible” deformities as it also encompasses more aggressive cases with fixed deformities and important limitation of the functional capacity and quality of life, which seems to make room for surgical corrective procedures. However, the present review revealed that only a few studies addressed this issue. Moreover, such studies were limited to small case series; there was no uniform definition of the outcomes, and the critical issue of follow-up time varied largely. Thus, they did not allow for definite conclusions about the appropriateness of surgery or a best surgical procedure for JA. However, despite the limited scientific evidence indicated previously, there are some expert opinions on this subject. According to Nalebuff,¹⁹ arthroplasty of the MCP joints may be considered in JA. However, based on his experience (unpublished) of this type of procedure, it is not as successful as in RA, because patients with JA tend to recover joint movement in an excessive way leading to fracture of the prosthesis and return of the deformity.

Generally speaking, in JA, there is no bone erosion, although clinically the joint deformities can be very similar to those seen in RA. Thus, in theory, regardless of the mechanism responsible for the development of JA, the same procedures utilized for correction of deformities in RA could be applied for joints with JA. The integrity of the bone structure in joints with JA might be regarded as a positive point in favor of the indication of surgery in said joints. On the other hand, the multisystemic nature of SLE, which is at present the leading disorder associated with JA, should be taken into consideration when such procedures are being considered.

REFERENCES

- Jaccoud S. *Sur une forme de rhumatisme chronique: leçons de clinique médicale faites à l'Hôpital de la Charité*. Paris, France: Delahaye; 1869:598–616.
- Babini SM, Cocco JA, de la Sota M, et al. Tendinous laxity and Jaccoud's syndrome in patients with systemic lupus erythematosus. Possible role of secondary hyperparathyroidism. *J Rheumatol*. 1989;16:494–498.
- de la Sota M, García-Morteo O, Maldonado-Cocco JA. Jaccoud's arthropathy of the knees in systemic lupus erythematosus. *Arthritis Rheum*. 1985;28:825–827.
- Spronk PE, ter Borg EJ, Kallenberg CG. Patients with systemic lupus erythematosus and Jaccoud's arthropathy: a clinical subset with an increased C reactive protein response? *Ann Rheum Dis*. 1992;51:358–361.
- Szczepanski L, Targonska B, Piotrowski M. Deforming arthropathy and Jaccoud's syndrome in patients with systemic lupus erythematosus. *Scand J Rheumatol*. 1992;21:308–309.
- Santiago MB. Miscellaneous non-inflammatory musculoskeletal conditions. Jaccoud's arthropathy. *Best Pract Res Clin Rheumatol*. 2011;25:715–725.
- Joseph B, Chacko V. Chronic post-rheumatic-fever arthritis (Jaccoud's arthritis) involving the feet. A case report. *J Bone Joint Surg Am*. 1984;66:1124–1125.
- Siam AR, Hammoudeh M. Jaccoud's arthropathy of the shoulders in systemic lupus erythematosus [published online ahead of print June 1, 1992]. *J Rheumatol*. 1992;19:980–981.
- Schumacher HR, Zweiman B, Bora FW Jr. Corrective surgery for the deforming hand arthropathy of systemic lupus erythematosus [published online ahead of print June 11, 1976]. *Clin Orthop Relat Res*. 1976;292–295.
- Alves EM, Macieira JC, Borba E, et al. Spontaneous tendon rupture in systemic lupus erythematosus: association with Jaccoud's arthropathy [published online ahead of print December 3, 2009]. *Lupus*. 2010;19:247–254.
- Russell A, Percy J, Rigal W, et al. Deforming arthropathy in systemic lupus erythematosus. *Ann Rheum Dis*. 1974;33:204–209.
- Evans JA, Hastings DE, Urowitz MB. The fixed lupus hand deformity and its surgical correction. *J Rheumatol*. 1977;4:170–175.
- Hastings DE, Evans JA. The lupus hand: a new surgical approach. *J Hand Surg Am*. 1978;3:179–183.
- Alhot JY, Livermeux P, Welby F. Jaccoud's arthropathy. Surgical results of 41 hands [published online ahead of print December 3, 2004]. *Chir Main*. 2004;23:229–236.
- Waguri-Nagaya Y, Yamagami T, Tsuchiya A, et al. Silicone-induced foreign-body reaction after first metatarsophalangeal joint arthroplasty for Jaccoud's arthropathy. *Rheumatol Int*. 2009;29:1367–1371.
- Atta AM, Oliveira RC, Oliveira IS, et al. Higher level of IL-6 in Jaccoud's arthropathy secondary to systemic lupus erythematosus: a perspective for its treatment? [published online ahead of print June 19, 2014]. *Rheumatol Int*. 2015;35:167–170.
- Dray GJ, Millender LH, Nalebuff EA, et al. The surgical treatment of hand deformities in systemic lupus erythematosus. *J Hand Surg Am*. 1981;6:339–345.
- Wood VE, Ichtertz DR, Yahiku H. Soft tissue metacarpophalangeal reconstruction for treatment of rheumatoid hand deformity. *J Hand Surg Am*. 1989;14(2 pt 1):163–174.
- Nalebuff EA. Surgery of systemic lupus erythematosus arthritis of the hand. *Hand Clin*. 1996;12:591–602.