Sonographic Findings of Hands and Wrists in Systemic Lupus Erythematosus Patients With Jaccoud Arthropathy

Carolina Freitas Lins, MD, Daniel Lima de Sá Ribeiro, MD,* Willer Gonçalves Dourado Santos,* Genevievi Rosa, MD,* Viviane Machicado, MD,* Ana Luiza Pedreira, MD,* Emanuela Pimenta da Fonseca, MD,* Anna Paula Mota Duque Souza, MD,* Carla Baleeiro Rodrigues Silva,* Luana Grazielle dos Santos Ferreira,* Verena Galvão, PhD,* Isabela Silva de Oliveira, PhD,† Ajax Merces Atta, PhD,† and Mittermayer Barreto Santiago, PhD*‡*

Introduction: Jaccoud arthropathy (JA) is a deforming nonerosive arthropathy, characterized by the presence of "reversible" joint deformity. The study aims at describing the main musculoskeletal ultrasound (US) findings of a series of systemic lupus erythematosus (SLE) patients with JA.

Methods: Consecutive patients diagnosed as having JA and SLE were included in the study. All patients underwent a whole clinical evaluation and US of the hands and wrists. The US scan evaluated the presence of synovial hypertrophy, tenosynovitis, and bone erosions.

Results: Thirty-nine of 40 patients were female. The US examination was performed on 560 joints of the hands and wrists. At least 1 change was observed in the US examination of 20 patients (50.0%), but there was not a statistically significant association with disease activity ($P = 0.33$). Nineteen patients (47.5%) had synovial hypertrophy, 9 (22.5%) had tenosynovitis, and 7 (17.5%) had both. Isolated small bone erosions were identified in 2 patients (5.0%).

Conclusions: Ultrasound examination is a reliable and noninvasive imaging method for the evaluation of joint involvement in SLE. Half of the patients with JA have ultrasonographic signs of joint inflammation, and these abnormalities may be found even in the absence of disease activity.

Key Words: Jaccoud arthropathy, joint, musculoskeletal, systemic lupus erythematosus, ultrasound

(*J Clin Rheumatol* 2018;24: 70–74)

Jaccoud arthropathy (JA) is a deforming nonerosive arthropathy, primarily described in patients with rheumatic fever after recurrent episodes of arthritis. In addition, it has been described in many other rheumatic conditions, particularly in systemic lupus erythematosus (SLE) with a prevalence of approximately 5%.[1] Clinically, JA is characterized by the presence of "reversible" joint deformities; it mainly affects the hands but is also observed in feet, knees, and shoulder joints.[2] The most common deformities seen in JA include "swan neck," ulnar deviation, "z"-thumb, and hallux valgus. It should be noted that, although the deformities in JA are "correctable," they may cause functional limitations and affect the quality of life of the patients.[1]

*Escola Bahiana de Medicina e Saúde Pública; †Departamento de Análises Clínicas e Toxicológicas, Faculdade de Farmácia, Universidade Federal da Bahia; and ‡SER da Bahia: Serviços Especializados em Reumatologia da Bahia, Salvador, Bahia, Brazil.

M.B.S. and A.M.A. have received a scholarship from Conselho Nacional de Desenvolvimento Científico e Tecnológico (CNPq).

The authors declare no conflict of interest.

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

ISSN: 1076-1608
DOI: 10.1097/RHU.0000000000000622

Characteristically, the plain radiographs of joints in patients with JA, even in the presence of deformities and subluxation, do not reveal erosions as those seen in patients with rheumatoid arthritis (RA). However, the more-refined imaging methods such as ultrasound (US) and magnetic resonance imaging (MRI) have illustrated erosions in the joints of these patients.[3]

Thus, the aim of this study was to describe the main musculoskeletal US findings in SLE patients with JA.

MATERIALS AND METHODS

Patients

This study included patients diagnosed as having SLE based on the American College of Rheumatology criteria[4] and JA as defined by Santiago.[5] All patients underwent a whole clinical evaluation including demographic data, disease duration, clinical manifestations, and disease activity. The disease activity was assessed by Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) score,[6] and the patients were arbitrarily considered as active when the SLEDAI score was greater than or equal to 6. However, visual assessment was excluded because ophthalmologic evaluation was not performed. Blood samples of all the patients were taken for laboratory tests. All patients signed an informed consent. The study was approved by the research ethics committee of our institution.

Hand and Wrist Ultrasound

Bilateral US examination of the hands and wrists of the patients 7was carried out by 2 radiologists specialized in the musculoskeletal system with more than 10 years of experience, using Esaote MyLab 25Gold machine with a 14-MHz transducer, according to the European League Against Rheumatism guidelines for musculoskeletal US in rheumatology diseases.[7] The radiologists were blinded to clinical findings, and they performed the US examination independently. Moreover, the agreement was assessed using the data in terms of presence or absence of US changes.

The images were obtained in the transverse and longitudinal planes of each point: wrist (dorsal side of radiocarpal and ulnocarpal joints), dorsal and volar sides of the second to fourth metacarpophalangeal (MCP) joints and proximal interphalangeal (PIP) joints, and flexor and extensor tendons from the second to fourth fingers on both hands. These anatomical sites were chosen because they are pathologically representative sites in accordance with previous studies on SLE patients.[8]

The present study evaluated the presence of synovial hypertrophy, tenosynovitis, and bone erosions in the patients. Synovial hypertrophy is defined as the presence of abnormal hypoechoic material (sometimes isoechoic or hyperechoic, comparable with subdermal fat) at the joint recess, which is also nondisplaceable

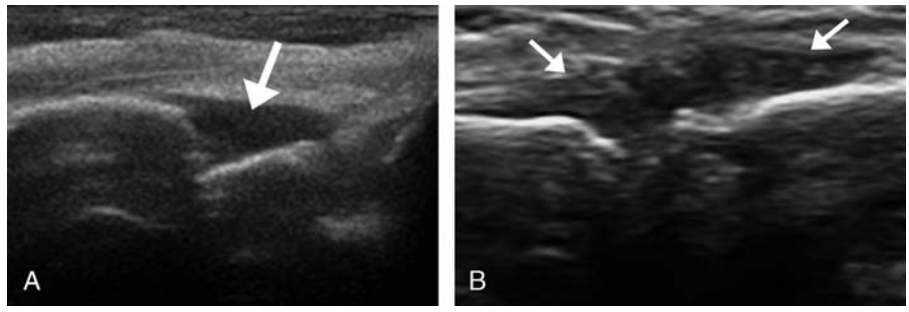

FIGURE 1. A, Ultrasonographic example of mild synovial hypertrophy in the third metacarpophalangeal joint (arrow). B, Ultrasonographic example of extensive synovial hypertrophy in the second MCP joint (arrows).

and poorly compressible, in accordance with the Outcome Measures in Rheumatology Clinical Trials.[9] Tenosynovitis is defined as thickened and hypoechoic tendon with or without synovial fluid in their sheath. In accordance with previous studies, a semiquantitative scale from 0 to 3 was used to evaluate synovitis and tenosynovitis through gray-scale study.[10–12] Thus, the degree of synovial hypertrophy was classified as follows: grade 0 = no synovial thickening, grade 1 = minimal synovial thickening (filling the angle between the periarticular bones, without crossing the line connecting tops of the bones), grade 2 = synovial thickening crossing the line connecting tops of the periarticular bones but without extension along the bone diaphysis, and grade 3 = synovial thickening crossing the line connecting tops of the periarticular bones with extension to at least 1 of the bone diaphyses (Fig. 1). The degree of tenosynovitis was also graded from 0 to 3: grade 0 = no signs of tenosynovitis (diameter of synovial tendon sheath ≤0.3 mm, on the maximal diameter detectable in transverse view); grade 1 = mild tenosynovitis (diameter of the synovial tendon sheath ≤2 mm, on the maximal diameter detectable in transverse view); grade 2 = moderate tenosynovitis (diameter of the synovial tendon sheath ≤4 mm, on the maximal diameter detectable in transverse view); and grade 3 = severe tenosynovitis (synovial sheath of the tendon diameter >4 mm, on the maximal diameter detectable in transverse view) (Fig. 2). Bone erosions are defined as greater than 2-mm defects in width and depth superficial cortical bone as identified in both the longitudinal and transversal planes (Fig. 3).[9]

Statistical analysis

Statistical analysis was performed using the Statistical Package for the Social Sciences (version 20; SPSS Inc, Chicago, IL). Certain results were expressed as mean ± SD. The χ^2 test was used to compare categorical variables. The interobserver agreement was assessed by κ index. For all statistical tests, $P < 0.05$ was considered significant.

RESULTS

Clinical and Laboratory Findings

The study included 40 patients (39 female patients), which corresponded to approximately two-thirds of the SLE cases with JA presented at our institution. Our present SLE cohort is composed of 1090 patients. The most frequent joint deformities observed in the hands were "swan neck," ulnar deviation, "z"-thumb, and hallux valgus. Thirteen patients (32.5%) had disease activity; that is, SLEDAI score was greater than or equal to 6. Demographic data, deformities of the hands, and the main clinical and laboratory findings of all the patients are shown in Table 1.

Ultrasound Findings

The US examination was performed on 560 joints of the hands and wrists and 480 tendons (including flexor and extensor of both hands). The interobserver analysis of radiocarpal synovial hypertrophy was approximately 0.4, with moderate concordance and statistical significance ($P = 0.025$). The other anatomic sites and variables analyzed did not reach statistical significance on radiologists' agreement.

A minimum of 1 US change was found in 20 patients (50.0%), but there was no statistically significant association between the presence of at least 1 US abnormality and disease activity ($P = 0.33$). From the entire cohort of this study, 19 patients (47.5%) had synovial hypertrophy, 9 (22.5%) had tenosynovitis, and 7 (17.5%) had both. Eighteen patients had mild synovial hypertrophy, 5 had moderate synovial hypertrophy, and only 1 patient showed extensive synovial hypertrophy.

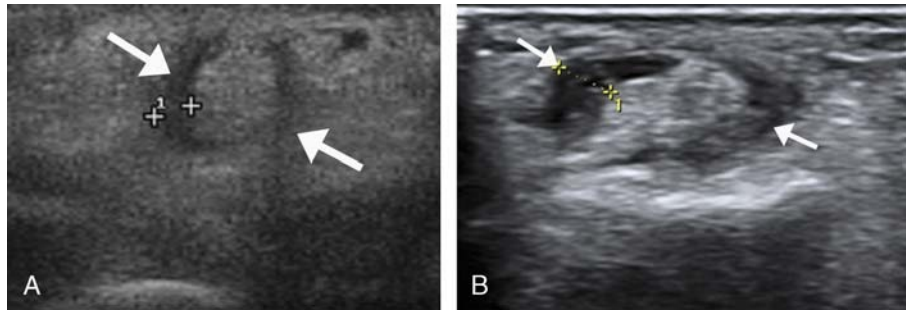

FIGURE 2. A, Ultrasonographic example of mild tenosynovitis at the third finger's flexor, in the transverse plane (arrows). B, Ultrasonographic example of moderate tenosynovitis at the second fingers' flexor, in the transverse plane (arrows). Color online-figure is available at http://www.jclinrheum.com.

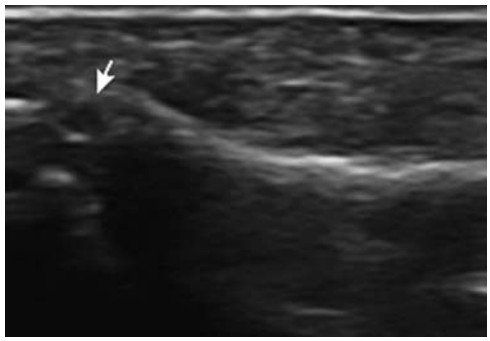

FIGURE 3. Ultrasonographic example of bone erosion in the third metacarpal bone, in the longitudinal plane (arrow).

Tenosynovitis of the flexor compartment was observed in 9 patients (22.5%) and extensor compartment in 3 patients (7.5%). Nine patients had mild and 2 patients had moderate tenosynovitis. Five isolated small bone erosions were identified in 2 patients (5.0%), which was located in the third MCP (3 erosions) and radiocarpal joints (2 erosions). Isolated wrist involvement (synovial hypertrophy) was detected in 11 patients (27.5%), whereas hand involvement alone was seen in 9 patients (22.5%), mainly in the third and fourth MCP joints. Seven patients (17.5%) showed the simultaneous involvement of both wrists and hands (Tables 2 and 3).

DISCUSSION

Joint involvement in SLE is commonly found in approximately 90% of the patients.[13] In most of the cases, it is mild and intermittent, but approximately 5% of the cases develop a chronic and deforming arthropathy known as JA. This complication is characterized by deformities that are correctable by passive movement and have less morbidity than the fixed deformities seen in RA.[1]

The US findings of joints in SLE patients are less studied. A systematic review reported that most studies demonstrated US to be a more efficient way of detecting musculoskeletal changes than physical examination.[14] These findings were also confirmed by our group.[15] However, only a few studies exist on US findings in patients diagnosed as having JA. Hence, to the best of our knowledge, the present study is the largest case series of patients diagnosed as having JA that were evaluated using US. Recently, we performed a study where a group of 32 SLE patients with JA (part of the 40 patients included in the present study) was compared with 32 SLE patients without JA matched by age and disease duration. In that study, considering the physical examination itself, there was a positive association between swollen and/or tender joints and synovitis or tenosynovitis by US in the JA group.[16]

Gabba et al.[17] studied 108 cases of SLE using US examination and found that 6 patients had JA and 4 had US abnormalities; tendon involvement was seen in 3 patients, effusion in 2 (wrist, second and third MCP joints), and erosion in 1 patient (second MCP joint). Wright et al.[18] studied 17 patients with SLE, and "some of them" were also diagnosed as having JA. As they had not specified the number of patients with JA, it was not possible to conclude from the US findings in those patients. Similarly, Dreyer et al.[19] and Mosca et al.[20] also studied 2 and 10 patients diagnosed as having JA, respectively, and failed to describe the details of the US findings.

Buosi et al.[21] evaluated 62 patients with SLE, among which 7 patients had JA, and 10 erosions were identified by the US examination, but they did not mention how many participants

TABLE 1. Clinical and Laboratory Findings[a] of 40 SLE Patients With JA

Feature	n (%)
Age, mean ± SD, y	44 ± 12
Sex (female), n	39/40 (97.5)
Disease duration, mean ± SD, y	16.3 ± 7.4
Duration of arthritis, mean ± SD, y	16.3 ± 7.5
"Swan neck" deformity	40 (100)
Ulnar deviation	27 (67.5)
"Z"-thumb	20 (50)
Hallux valgus	7 (17.5)
SLEDAI >6	13 (32.5)
Malar rash	21 (52.5)
Photosensitivity	30 (75)
Discoid lesion	4 (10)
Oral ulcers	15 (37.5)
Leukopenia (<4000/μL)	23 (57.5)
Lymphopenia (<1500/μL)	15 (37.5)
Thrombocytopenia (<100,000/μL)	7 (17.5)
Autoimmune hemolytic anemia	12 (30)
Pericarditis	8 (20)
Pleural effusion	12 (30.)
Lupus nephritis[b]	17 (42.5)
Psychosis	6 (15)
Seizures	8 (20)
Raynaud phenomenon	18 (45)
Venous thrombosis	3 (7.5)
ANA positive	40 (100)
Anti-dsDNA	36 (90)
Anti-SSA/Ro	15 (37.5)
Anti-SSB/La	6 (15)
Anti-Sm	13 (32.5)
Anti-RNP	15 (37.5)
Immunoglobulin G anticardiolipin	4 (10)
Immunoglobulin M anticardiolipin	3 (7.5)

[a]Except for SLEDAI, findings refer to present or past.

[b]Lupus nephritis = creatinine greater than reference range and/or proteinuria greater than 1 g/24 h.

ANA indicates antinuclear antibody.

TABLE 2. Ultrasound Findings of the Hands and Wrists of 40 Lupus Patients With JA

Findings	n (%)
Synovial hypertrophy	19 (47.5)
Tenosynovitis	9 (22.5)
Bone erosions	2 (5.0)
Isolated wrist involvement (synovial hypertrophy)	11 (27.5)
Hand involvement alone	9 (22.5)
Hand and wrist involvement at the same time	7 (17.5)
At least 1 US alteration	20 (50.0)

n Indicates number of patients.

TABLE 3. Distribution of Analyzed Joints and Their Sonographic Features

	Ultrasonography Exam		
Joint	Synovitis, n (%)	Tenosynovitis, n (%)	Bone Erosions, n (%)
PIP (n = 240)	12 (5.0)	77 (32.1)	1 (0.4)
MCP (n = 240)	76 (31.6)	77 (32.1)	6 (2.5)
Wrist (n = 80)	23 (28.7)	30 (37.5)	0 (0)
Total (n = 560)	111 (19.8)	42 (4.7)	7 (1.2)

n Indicates number of joints; PIP, proximal interphalangeal.

had these findings. This figure seems to be overestimated as compared with our series and can be interpreted as a selection bias probably due to the inclusion of the most aggressive cases with longstanding deformities. In a recent report,[22] 1 patient with SLE underwent an US examination of wrists and hands only because she had JA in her hands and positive C-reactive protein and high erythrocyte sedimentation in the absence of joint pain or swelling. The examination revealed synovial hypertrophy in the radiocarpal joints and bilateral tenosynovitis in the carpal flexor, demonstrating the subclinical nature of disease in some patients. Furthermore, a recent systematic review mentioned the poor understanding of the formation and evolution of bone erosion in SLE patients, especially those with JA.[23] Of note, power Doppler associated with the B-mode analysis of joints was not used in the present study. Indeed, the cited systematic review observed that power Doppler has unknown significance as a marker of disease activity in SLE, because of the lack of standardization of its criteria of evaluation.[23]

There are only 2 studies that report detailed imaging findings by MRI in patients with JA. Ostendorf et al.[24] studied both hands of 14 patients with SLE, in which only 4 were diagnosed as having JA, as defined by Spronk et al.[25] They found edematous tenosynovitis and capsular swelling in all of them and subchondral cysts in only 2 of 4 patients. Another recent publication presented 20 patients with SLE and JA (19 females and 1 male) who were examined using MRI.[26] It reported some degree of synovitis in 202 (67.3%) and small areas of erosion in 16 (5.3%) of 300 examined joints. Tenosynovitis was detected in 77 (38.5%) of 200 compartment tendons evaluated. Twelve of 40 patients of the present series have participated in the MRI study, but as the time frame between these 2 studies was more than 4 years, the comparison of the prevalence of the findings is not reliable. On the other hand, there was the agreement between the 2 methods regarding the percentage of bone erosions. Thus, this suggests that US examination is a reliable, cheaper, and noninvasive imaging method for the evaluation of joint involvement in SLE patients with or without JA.

Half of the patients with JA had ultrasonographic signs of joint inflammation, but these findings were not necessarily associated with the presence of systemic disease activity. It may suggest that the deformities in JA can occur progressively secondary to a subclinical joint inflammation in patients with the disease apparently under control. This phenomenon has also been observed in JA secondary to rheumatic fever.[27]

REFERENCES

1. Santiago MB, Galvao V. Jaccoud arthropathy in systemic lupus erythematosus: analysis of clinical characteristics and review of the literature. *Medicine*. 2008;87:37–44.
2. Ribeiro DS, Santiago M. Imaging of Jaccoud's arthropathy in systemic lupus erythematosus: not only hands but also knees and feet. *Rheumatol Int*. 2012;32:567–568.
3. Saketkoo LA, Quinet R. Revisiting Jaccoud arthropathy as an ultrasound diagnosed erosive arthropathy in systemic lupus erythematosus. *J Clin Rheumatol*. 2007;13:322–327.
4. Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum*. 1997;40:1725.
5. Santiago MB. Jaccoud's arthropathy: proper classification criteria and treatment are still needed. *Rheumatol Int*. 2013;33: 2953–2954.
6. Bombardier C, Gladman DD, Urowitz MB, et al. Derivation of the SLEDAI. A disease activity index for lupus patients. The Committee on Prognosis Studies in SLE. *Arthritis Rheum*. 1992;35:630–640.
7. Backhaus M, Burmester GR, Gerber T, et al. Guidelines for musculoskeletal ultrasound in rheumatology. *Ann Rheum Dis*. 2001;60: 641–649.
8. Yoon HS, Kim KJ, Baek IW, et al. Ultrasonography is useful to detect subclinical synovitis in SLE patients without musculoskeletal involvement before symptoms appear. *Clin Rheumatol*. 2014;33: 341–348.
9. Wakefield RJ, Balint PV, Szkudlarek M, et al. Musculoskeletal ultrasound including definitions for ultrasonographic pathology. *J Rheumatol*. 2005;32:2485–2487.
10. Delle Sedie A, Riente L, Bombardieri S. Limits and perspectives of ultrasound in the diagnosis and management of rheumatic diseases. *Mod Rheumatol*. 2008;18:125–131.
11. Filer A, de Pablo P, Allen G, et al. Utility of ultrasound joint counts in the prediction of rheumatoid arthritis in patients with very early synovitis. *Ann Rheum Dis*. 2011;70:500–507.
12. Szkudlarek M, Court-Payen M, Jacobsen S, et al. Interobserver agreement in ultrasonography of the finger and toe joints in rheumatoid arthritis. *Arthritis Rheum*. 2003;48:955–962.
13. Messuti LZ, A, Gremese E, Ferraccioli G. Joint involvement in SLE: the controversy of RHUPUS. *Int Trends Immunity*. 2014;2:155–161.
14. Lins CF, Santiago MB. Ultrasound evaluation of joints in systemic lupus erythematosus: a systematic review. *Eur Radiol*. 2015;25: 2688–2692.
15. Lins CF, Lima de Sa Ribeiro D, Dourado Santos WG, et al. Ultrasound findings on hands and wrists of patients with systemic lupus erythematosus: relationship with physical examination [published online ahead of print September 5, 2017]. *Ultrasound Med Biol*. 2017.
16. DS Ribeiro CL, Pimenta E, Galvão V, et al. OP0290 Correlation of CXCL13 and ultrassonographic findings of Jaccoud arthropathy in systemic lupus erythematosus. *Ann Rheum Dis*. 2017;76(suppl 2).
17. Gabba A, Piga M, Vacca A, et al. Joint and tendon involvement in systemic lupus erythematosus: an ultrasound study of hands and wrists in 108 patients. *Rheumatology*. 2012;51:2278–2285.
18. Wright S, Filippucci E, Grassi W, et al. Hand arthritis in systemic lupus erythematosus: an ultrasound pictorial essay. *Lupus*. 2006;15: 501–506.
19. Dreyer L, Jacobsen S, Juul L, et al. Ultrasonographic abnormalities and inter-reader reliability in Danish patients with systemic lupus erythematosus—a comparison with clinical examination of wrist and metacarpophalangeal joints. *Lupus*. 2015;24:712–719.
20. Mosca M, Tani C, Carli L, et al. The role of imaging in the evaluation of joint involvement in 102 consecutive patients with systemic lupus erythematosus. *Autoimmun Rev*. 2015;14:10–15.

21. Buosi ALP, Natour J, Machado FS, et al. Hand ultrasound: Comparative study between "no rhupus " lupus erythematosus and rheumatoid arthritis. *Mod Rheumatol*. 2014;24:599–605.
22. Feced Olmos CM, Ivorra Cortes J, Negueroles Albuixech R, et al. Usefulness of ultrasound in Jaccoud's arthropathy. A case report. *Reumatol Clin*. 2015;11:127–129.
23. Zayat AS, Md Yusof MY, Wakefield RJ, et al. The role of ultrasound in assessing musculoskeletal symptoms of systemic lupus erythematosus: a systematic literature review. *Rheumatology*. 2016;55:485–494.
24. Ostendorf B, Scherer A, Specker C, et al. Jaccoud's arthropathy in systemic lupus erythematosus: differentiation of deforming and erosive patterns by magnetic resonance imaging. *Arthritis Rheum*. 2003;48:157–165.
25. 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.
26. Sa Ribeiro D, Galvao V, Luiz Fernandes J, et al. Magnetic resonance imaging of Jaccoud's arthropathy in systemic lupus erythematosus. *Joint Bone Spine*. 2010;77:241–245.
27. Levin EB. Jaccoud's arthritis. Post-rheumatic fever complication—not rheumatoid arthritis. *Calif Med*. 1970;112:19–22.