

This Week's Citation Classic

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Tuffanelli D L & Winkelmann R K. Systemic scleroderma: a clinical study of 727 cases. *Arch. Dermatol.* 84:359-71, 1961.
[Mayo Clinic and Mayo Foundation, Rochester, MN]

Seven hundred twenty-seven patients with systemic scleroderma seen at the Mayo Clinic in the period from 1935 through 1958 were studied. The usual course and findings of the disease were determined and the unusual features were stressed. The prognosis of scleroderma was determined. [The *SCI*[®] indicates that this paper has been cited in over 275 publications since 1961.]

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"The cited article was largely a product of the Mayo Clinic system of medicine and biostatistics. Richard Winkelmann, an enthusiastic staff member, suggested that a clinical review of the Mayo Clinic experience with scleroderma patients would be valuable as a basis for future evaluations and study of the disease. Because of the clinic's status as a tertiary referral care center, and the specific interests of Paul O'Leary of the dermatology department, a vast clinical experience with scleroderma had accumulated. The prospect of doing a scholarly, in-depth study of a single, chronic, catastrophic illness appeared promising to me, and I accepted the project.

"The study was quite time-consuming, and occupied most of my spare time over the next two years. Current patients were examined, and all available charts were reviewed from cover to cover, in the precomputer era. The clinical and laboratory data on 727 patients with systemic scleroderma observed at the Mayo Clinic from 1935 through 1958 were correlated. Scleroderma is a capricious disease. Some patients had a fulminating course with death in a year or two. Others had a chronic, extremely debilitating illness. Rare patients improved spontaneously.

Many of the charts were poignant short stories. The tragedy overwhelming many of the patients was amply documented in the records. I enjoyed library research and read over 400 references on scleroderma. The superb Mayo Clinic library was a pleasant place to spend the cold Minnesota evenings.

"Finally, the literature review and clinical data were presented as a master's thesis, and with Winkelmann's collaboration three published papers resulted,^{1,2} the major one being the *Archives of Dermatology* article.

"The article was, and remains, a review of the largest number of patients with scleroderma. The incidence of Raynaud's phenomenon, and cutaneous features including sclerosis, telangiectasia, calcinosis, cutaneous ulcerations, and pigmentary changes, were documented. Similarly, the incidence of systemic features, particularly gastrointestinal, pulmonary, and renal, was determined. Prognostic studies furnished the valuable information that 29.7 percent of the patients died in less than five years (from time of diagnosis at Mayo) while 70.3 percent of patients survived five years without benefit of a specific therapy. The ten-year survival rate of 58.9 percent indicated that the prognosis was not as bleak as previously thought.

"The study focused my academic interest on connective tissue diseases and following dermatology residency I decided to further study this fascinating group of diseases. For the past 20 years, I have conducted a connective tissue disease clinic and immunopathology laboratory at the University of California in San Francisco. Over 150 publications have been generated by this experience.³

"The small coterie of physicians interested in scleroderma has provided international friendships and meetings.⁴ Medical directorship of the United Scleroderma Foundation, a national group of scleroderma patients, has provided further personal experience with the disease.

"So the paper has certainly been a personal hallmark. The paper is perhaps cited because it is a simply presented, statistically relevant documentation of the clinical features of a fascinating medical problem."

1. Tuffanelli D L & Winkelmann R K. Scleroderma and its relationship to the "collagenoses": dermatomyositis, lupus erythematosus, rheumatoid arthritis and Sjogren's syndrome. *Amer. J. Med. Sci.* 243:133-46, 1962.
2., Diffuse systemic scleroderma: a comparison with acrosclerosis. *Ann. Intern. Med.* 57:198-203, 1962.
3. Tuffanelli D L. Connective tissue diseases. (Malkinson F D & Pearson R W, eds.) *The year book of dermatology*. Chicago: Year Book Medical Publishers, 1978. p. 9-36.
4. Tan E M, Rodnan G P, Garcia J, Morol Y, Fritzier M J & Peebles C. Diversity of antinuclear antibodies in progressive systemic sclerosis: anti-centromere antibody and its relationship to CREST syndrome. *Arthritis Rheum.* 23:617-25, 1980.