The Scleroderma Education Project™

Understanding the New 2013 Formal Diagnostic Criteria for Systemic Scleroderma

In late 2013, the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) approved a new set of diagnostic criteria for systemic sclerosis (SSc), replacing the older 1980 diagnostic criteria. These new standards will improve clinical diagnosis of systemic scleroderma, but it is very important to understand that the reason for developing these new diagnostic standards was "**to develop a set of criteria that would enable identification of individuals with SSc for *inclusion in clinical studies,***" not for the purpose of normal diagnosis of patients in a clinical setting. The authors of the special report that formally introduces the new criteria note that many symptoms that are used for clinical diagnosis are not included in these formal research criteria, including common symptoms such as tendon friction rubs, calcinosis, difficulty swallowing, as well as less common but more serious complications, such as renal crisis.

Note: The table below is a slightly simplified version of the new classification criteria:

2013 ACR/EULAR Classification Criteria for Systemic scleroderma

Item	Sub-Item(s)	Weight
Skin thickening of the fingers of both hands that extends at least up to the joint at the base of the fingers (third joint on fingers, second joint on thumb) **(sufficient criterion)**		9
Skin thickening of the fingers (only count the higher score)	Puffy fingers	2
	Thickening of the fingers up to the second finger joint	4
Fingertip lesions (only count the higher score)	Digital tip ulcers	2
	Fingertip pitting scars	3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or interstitial lung disease (maximum score is 2)	Pulmonary arterial hypertension	2
	Interstitial lung disease	2
Raynaud's phenomenon (can be self-reported)		3

Scleroderma-related autoantibodies (maximum score is 3)	Anti-centromere Anti-Scl-70 (Anti–topoisomerase I) Anti–RNA polymerase III	3

Source: Van den Hoogen et al. 2013 Classification Criteria for Systemic Sclerosis. *Arthritis and Rheumatism* Vol. 65, No. 11, November 2013, pp 2737-2747.

The total score is determined by adding the maximum weight (score) in each category. Patients with a total score of 9 or greater are classified as having definite systemic scleroderma. For example, a patient with definite skin thickening on both hands all the way to the base of the fingers receives a score of 9 just for that single symptom and is automatically classified as having definite systemic scleroderma. For the other categories, you receive points based on the highest scoring symptom in that category. To illustrate, a patient who has Raynaud's (3), fingertip lesions with pitting scars (3), anti-centromere antibodies (3), and abnormal nailfold capillaries (2) would receive a total weighted score of 11 and would also be diagnosed with systemic scleroderma. Note that within a general category, e.g., "Skin thickening of the fingers," you would "earn" 4 points for skin thickening up to the second finger joint OR 2 points if you just had puffy fingers, but not 6 points for both.

There is no question that these new diagnostic criteria will be helpful to clinicians as well as researchers, but there are a number of issues that will arise in clinical diagnosis because of the way these criteria were developed. For example, you will note that there is nothing in these criteria that includes any GI involvement, which is very common with all forms of systemic scleroderma. There is also no mention of renal (kidney) problems, which are rare but a strong clinical complication that occurs with some forms of systemic scleroderma.

These were excluded from these research criteria for different reasons. In the case of GI symptoms such as GERD (reflux), from a research classification perspective they are not specific enough to only systemic scleroderma to be useful in patient classification, since they can occur with many other different diseases, e.g., lupus. On the other hand, while renal crisis associated with some of the other symptoms is very specific to systemic scleroderma, it is actually so rare that it didn't reach the level of significance in doing the classification research. so there was no benefit to including it in the classification criteria.

It is also very noteworthy that the "scleroderma-related autoantibodies" category adds anti-RNA polymerase III to the standard anti-centromere and anti-Scl-70 antibodies that have been associated with systemic scleroderma for many years. As mentioned in another article on this website, the anti-RNA polymerase III antibody is associated with one of the diffuse variants of scleroderma and has a different typical clinical symptom profile than diffuse patients with the anti-Scl70 antibody. Also, the paper introducing these new criteria discussed additional antibodies, indicating that they are likely to be added to the table in the future as more research is done to allow better understanding of the clinical significance of these less common antibodies. However, it is worth noting that the new criteria only result in a diagnosis of systemic scleroderma, but do not directly indicate which form of scleroderma, even at the general level of limited or diffuse, in spite of directly including three specific antibodies in the table.

Scleroderma diagnosis will remain a clinical challenge in many cases, notwithstanding the new diagnostic criteria. For example, clinicians still need to consider clinical symptoms that support a diagnosis of systemic scleroderma but which are not included in the new 2013 ACR criteria, e.g., GI symptoms such as GERD (reflux), difficulty swallowing, etc. An additional

challenge for clinicians has been the switch to the new ICD-10 diagnostic coding system on October 1, 2015, which required more specific diagnosis than was previously required.

The reality is that in most cases, when patients start developing symptoms, such as Raynaud's, heartburn, puffy fingers, muscle pain and weakness, their first visit will be to their primary care clinician, who is likely to be an internist, family medicine doctor, or a nurse practitioner. In most cases, these clinicians will have rarely, if ever, encountered a patient with scleroderma and may not have read anything about the disease since they were formally trained , which might have been 20 years earlier! Because of the rarity of systemic scleroderma, many primary care clinicians may not initially think of autoimmune diseases. However, once the patient or clinician starts to consider a potential autoimmune disease as the cause of the patient's symptoms, it is almost always the best course of action to bring a rheumatologist into the diagnostic loop since s/he will be trained in diagnosing and treating autoimmune diseases. It is still important to realize that, especially in a small community, most rheumatologists may have never seen a patient with scleroderma, but at least they are much more likely to have the training needed to diagnosis scleroderma correctly and work with patients to determine the best treatment options for their particular situation.