

## Immunosuppressant drugs may help in systemic sclerosis (dcSSc), but new treatments are needed



Immunosuppressants show some benefit in early dcSSc, but better treatments are still needed.

### INTRODUCTION

Systemic sclerosis is a rare but serious disease of the connective tissues, where the tissue grows abnormally and triggers the immune system to attack itself (autoimmunity). This can cause hardening (fibrosis) and swelling of the skin, as well as joint pain, digestive problems and lung disease caused by fibrosis and scarring. It can also affect the heart and kidneys. It usually starts in people aged 30–50 years of age, and is more common in women than men.

The diffuse cutaneous subtype of systemic sclerosis (often shortened to dcSSc) is even rarer, affecting just one in every four people with the disease. It is linked with early damage to a person's internal organs, as well as painful skin thickening that quickly gets worse. Only half of people diagnosed with dcSSc will survive for 10 years or more.

Medicines for dcSSc are very limited, and there are no options at the moment that have very good results. Immunosuppressant medicines work by lowering the body's immune response. These sorts of medicines are sometimes used in dcSSc, although there have not been trials to see how well they do in people with this specific form of the disease. Because the disease is so rare, it is difficult to research.

### WHAT DID THE AUTHORS HOPE TO FIND?

The authors wanted to see how well people with early dcSSc did when they received one of four different standard treatments.

### WHO WAS STUDIED?

The study looked at 326 people from 50 clinics in 19 countries in Europe, North America, the Middle East and Australia. Everyone had skin thickening caused by dcSSc that had started less than 3 years ago. Everyone was over the age of 18.

### HOW WAS THE STUDY CONDUCTED?

This was a prospective observational study. This means that the authors looked at information collected in a set of patients, but made no interventions and there was no medicine under investigation. The doctors taking part in the study chose the treatment most appropriate for each person in their care from their list of four normal standard treatments. The standard treatments were mainly immunosuppressant drugs (methotrexate, mycophenolate mofetil, or cyclophosphamide) but also included a 'no immunosuppressant' option.

### WHAT WERE THE MAIN FINDINGS OF THE STUDY?

The study found that people's skin got better in all four groups. People who did not receive an immunosuppressant drug survived for a shorter time than people who did. Although there were no significant or statistical differences between the four treatment groups, the authors thought that there was some evidence to support using immunosuppressant drugs in people with early dcSSc.

### ARE THESE FINDINGS NEW?

Yes. This is the first time that these standard treatments have been compared in this sort of study.

### WHAT ARE THE LIMITATIONS OF THE STUDY?

The weakness of observational studies is that the treatment outcome for each person cannot be disentangled from their baseline characteristics. However, the authors are confident that the way they did their statistical analysis means that this is not a problem.

### WHAT DO THE AUTHORS PLAN ON DOING WITH THIS INFORMATION?

The information collected is being used to answer more questions. For example about how disability affects people who have early dcSSc, and if there are patterns that can be used to predict how people's skin symptoms might change.

### WHAT DOES THIS MEAN FOR ME?

If you have dcSSc, there are very limited treatment options at the moment. Better treatments are needed for people with this disease, but because it is a rare disease it is hard to do trials of new drugs. It is hoped that in the future there will be more options. In the meantime, your doctor might prescribe you an immunosuppressant medicine, but it is likely that you might get only modest improvements from it.

If you have concerns about your disease or the medicine you are taking, you should talk to your doctor.

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