

Progressive skin fibrosis is associated with decline in lung function and worse survival in people with dcSSc



Worsening skin disease could be used as a marker of internal organ damage in people with diffuse cutaneous subtype of systemic sclerosis (dcSSc)

INTRODUCTION

Systemic sclerosis is a rare but serious disease of the connective tissues, characterised by thickening of skin and by triggering the immune system to attack itself (autoimmunity). This can cause hardening (fibrosis) and swelling of the skin and damage of different organs such as lungs, heart, kidneys and gastrointestinal problems. It is more common in women than in men.

The diffuse cutaneous subtype of systemic sclerosis (often shortened to dcSSc) affects around 30% of people with systemic sclerosis. It is linked with early damage to a person's internal organs including the lungs and kidneys, as well as painful skin thickening that quickly gets worse.

WHAT DID THE AUTHORS HOPE TO FIND?

The authors wanted to find out whether progressive skin fibrosis, measured by the modified Rodnan skin score (mRSS) is linked to worsening of organ damage and death in people with dcSSc.

WHO WAS STUDIED?

The study looked at just over 1000 people with dcSSc. Everyone included in the study visited their doctor at least once a year, and had a measured mRSS score of 7 or more at the first visit, as well as repeated mRSS 9–15 months later.

HOW WAS THE STUDY CONDUCTED?

This was an observational study from the prospective EUSTAR (European Scleroderma Trials and Research group) database. The EUSTAR database is a multi-centre online database that follows more than 16,000 people with systemic sclerosis in more than 200 international centres (www.eustar.org). This means that the authors analysed the existing database, with yearly collected, predefined data assessments from people in a real-life clinical situation. There was no interventional treatment given as part of this study.

The authors analysed the EUSTAR database to find the records of people with dcSSc whose mRSS score had worsened over 9–15 months, suggesting that they had progression of skin fibrosis. They then looked to see if there was a link between the mRSS measurement and progression of disease in people's organs, including their lung function and causes of death.

WHAT WERE THE MAIN FINDINGS OF THE STUDY?

The study found that worsening skin fibrosis over a period of 1 year was associated with a decline in lung function and worse survival in people with dcSSc at long-term follow up. The results suggest that the mRSS measurement can be used as a marker in people with dcSSc to identify those at risk of organ damage or death at later disease stages. These results will be helpful in choosing SSc patients for future trials of hopefully more effective new therapies, as well as in working out who is at most risk.

ARE THESE FINDINGS NEW?

Yes. This is the first time that this link has been investigated.

WHAT ARE THE LIMITATIONS OF THE STUDY?

There are some weaknesses of observational studies. Firstly, the outcome for each person cannot be disentangled from their baseline characteristics or the treatment that they received. Additionally, there can be information missing from patient records, because it was not collected or recorded properly, or because people did not return to the clinic for assessment. In this study, for those people who died, it was not always clear what the cause of death had been. However, the authors are confident that the way they did their statistical analysis means that this is not a problem for the overall results.

WHAT DO THE AUTHORS PLAN ON DOING WITH THIS INFORMATION?

The authors expect that this information will be used to design future clinical trials in people with dcSSc, using mRSS as an outcome measure, for example as part of a combined measure, to develop better therapies.

WHAT DOES THIS MEAN FOR ME?

If you have rapidly progressive dcSSc, there are limited treatment options at the moment. Better treatments are needed for people with this disease, but it is hoped that in the future there will be more options. If you have skin fibrosis that gets worse over a short period of time, your doctor should monitor you for progression in your organs – especially your lungs.

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

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