



# TREATING SARCOIDOSIS

*Understanding the professional guidelines*



**ERS** EUROPEAN  
RESPIRATORY  
SOCIETY  
every breath counts



**ELF** EUROPEAN  
LUNG  
FOUNDATION



**European  
Reference  
Network**  
for rare or low prevalence  
complex diseases  
**Network**  
Respiratory Diseases  
(ERN-LUNG)



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## Introduction

### Who is this document for and what is it about?

This document explains the recommendations in the European Respiratory Society (ERS) clinical guideline for the treatment of sarcoidosis. It is aimed at people living with sarcoidosis, or their care givers.

This document covers cases of sarcoidosis that affects the lungs, the skin, the heart and brain as well as fatigue (extreme tiredness) that is caused by sarcoidosis. It does not include treatments related to other issues, such as eye issues or pain associated with the condition. This is because there is not yet enough research into these areas to draw conclusions about the best treatments.

### What are clinical practice guidelines?

Clinical guidelines are produced by experts, patient representatives and methodologists. First medical librarians conduct a thorough review of all research studies. The available evidence is then reviewed by a panel. These guidelines also consider the opinions of leading experts and the priorities of patients and carers who have experience of a condition. Clinical guidelines are aimed at healthcare professionals. They use them as a 'best practice' document to manage and treat specific conditions.

### What does this document include?

This document summarises the key points from the clinical guideline on treating sarcoidosis. It explains them in a way that is more easily understood by people who do not work in a medical field. It will cover what sarcoidosis is and how it should be treated.



By providing this information in an accessible way, this document aims to help people living with sarcoidosis and their carers understand more about the treatment process and feel informed when making decisions about care.

The American Thoracic Society (ATS) has published a guideline for the diagnosis of sarcoidosis – there is a link in the further reading section at the end of this document.

## What is sarcoidosis?

Sarcoidosis is an inflammatory condition without a known cause. It causes patches of red, swollen tissue (granulomas) to form in the body. It is thought this happens when the immune system has overreacted. Usually, when the immune system fights off infections, it causes swelling and redness (inflammation) in the area of the body where it is fighting off germs. This dies down once the infection has cleared. In conditions, such as sarcoidosis, the immune system is thought to overreact and attack its own tissues and organs, resulting in the inflammation and granulomas.

Sarcoidosis can affect many organs in the body, but the lungs are affected most often. It can also affect the skin, eyes, heart, muscles, joints, bones, liver, kidneys and brain (and more). Chest physicians (pulmonologists) are most often involved in diagnosing and managing this disease.



## Who should receive sarcoidosis treatment?

The way that sarcoidosis develops, and how severe it is, varies from each person. Many people with sarcoidosis do not need treatment, and the disease may go away on its own.

The decision on when and who should receive treatment for sarcoidosis is based on two factors – how much the disease is impacting on a person's quality of life and whether a person is at a risk of death or disability from the condition.

Treatments are available that can help to slow the disease and improve symptoms. There is Currently no cure for this disease.

## How should sarcoidosis be treated?

This guideline covers the different treatments that are used to reduce the symptoms of sarcoidosis. It focuses on treatments that aim to control the granulomas that are caused by sarcoidosis.

It does not cover the wider range of treatments, including those that may be beneficial but not related to granulomas, such as transplantation and oxygen therapy.

The two main types of treatment that should be considered for managing sarcoidosis are:

- Glucocorticoids – these are a type of steroid that work to reduce inflammation in the body.
- Immunosuppressants – these drugs work by calming/controlling the immune system's response when it has overreacted



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These drugs can improve symptoms of sarcoidosis. Although the disease can also get worse once steroid treatment has stopped. There are side effects to consider as steroids can cause weight gain, diabetes, hypertension, weakening of the bones and mood swings.

Immunosuppressants are often used to help people come off steroid drugs without their symptoms getting worse. This is known as steroid sparing.

Healthcare professionals should discuss options with you to help find a balance between the right dosage to manage any symptoms and reduce the risk of side effects.

The full guideline includes recommendations for when and how these two types of treatments should be used for sarcoidosis that affects different parts of the body.

## **Pulmonary sarcoidosis – affecting the lungs**

People at a high risk of long-term disability or death from sarcoidosis affecting the lungs, should be given glucocorticoid treatment. The aim of this treatment is to improve and/or preserve a person's quality of life and their lung function.

People who are experiencing a gradual worsening of their quality of life, but who are not at risk of death, should discuss the risks and benefits of steroid treatment with their healthcare professional. It is best to start off with a low to medium dose of treatment.



Glucocorticoids should not be used for people not at risk of death or those who do not have a worsened quality of life. This is because of the side effects they cause.

The guideline recommends adding an immunosuppressive drug to treatment if a person sees no improvement in their symptoms or suffers negative side effects of glucocorticoids. The aim of this is to improve their symptoms and/or preserve their quality of life. The guideline recommends first using a drug called methotrexate. If no positive change is seen from this, then it recommends trying a drug called infliximab.

There are other options that but there is less evidence supporting them. They are mentioned in the full clinical guideline as alternatives that have been used successfully in some people with sarcoidosis and are included in the Appendix.

### **Cutaneous sarcoidosis –affecting the skin**

Steroid treatment is recommended for people who have painful and potentially disfiguring bumps on the skin (lesions) that cannot be managed by skin creams. The treatment should be used sparingly due to the long-term side effects of glucocorticoid treatment.

If people do not see any improvement from their steroid treatment, the guideline suggest adding the immunosuppressive medication, infliximab.

Other options for treatment do exist, and may be proposed by healthcare professionals, but there is less evidence to support their use.



## **Cardiac sarcoidosis –affecting the heart**

For people with any heart abnormalities, including when the heart beats more slowly or at an unusual rhythm, the guideline recommends glucocorticoids. There was not enough evidence to recommend whether an immunosuppressive drug was also needed. However, it was the opinion of the experts working on the guideline that immunosuppressive drugs can help reduce the side effects associated with taking glucocorticoids on their own.

## **Neurosarcoidosis – mainly affecting the nervous system**

For patients who have sarcoidosis that affects the nervous system – including numbness and weakness in parts of the body, the guideline suggests treatment with glucocorticoids. If symptoms continue, an immunosuppressive treatment should be added: methotrexate should be tried first and if needed, infliximab can also be added.

## **Fatigue caused by sarcoidosis**

For people who experience severe fatigue (tiredness) that affects their quality of life, the guideline suggests a pulmonary rehabilitation programme and muscle strength training for 6-12 weeks. These programmes focus on increasing levels of activity and learning more about how improved activity levels and muscle strength can be beneficial to improve fatigue.

For some people who have debilitating, persistent fatigue, a type of drug called a neurostimulator could be used. The guideline recommends two types of neurostimulator, d-methylphenidate or armodafinil, which should be used for 8 weeks to test whether they improve symptoms.



## Established methods of treating sarcoidosis

During the process of producing the clinical guidelines, experts working on the document found a lack of evidence in several areas. Unfortunately, gaps in research are common for rare diseases, such as sarcoidosis.

To help provide information to cover this gap in the research, the professional guideline also covers the established methods of treating sarcoidosis. These are widely used by healthcare professionals and considered to be an established approach, despite the lack of research into these areas.

The diagrams below show the suggested approach for healthcare professionals treating different types of sarcoidosis:

### Diagram key

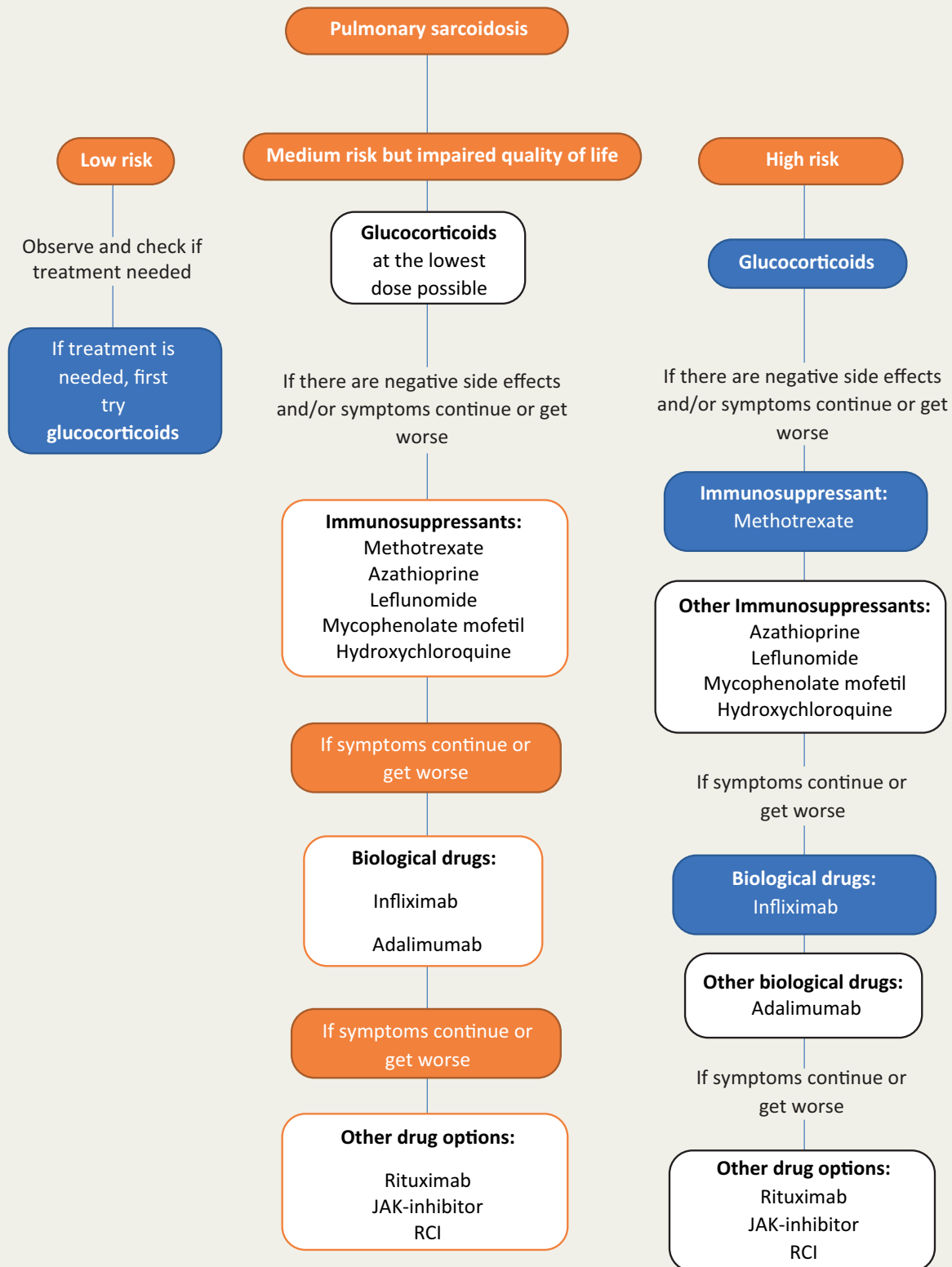
Type of disease and level of symptoms

Recommended treatment based on evidence

Established methods with lack of supporting evidence

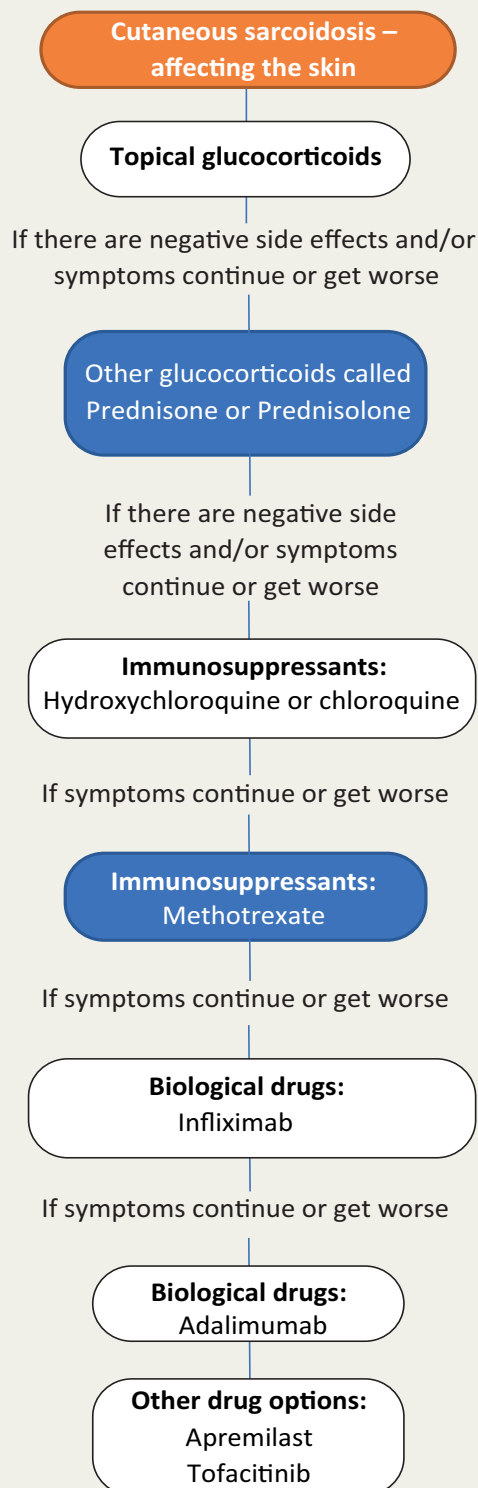


## Pulmonary sarcoidosis – affecting the lungs



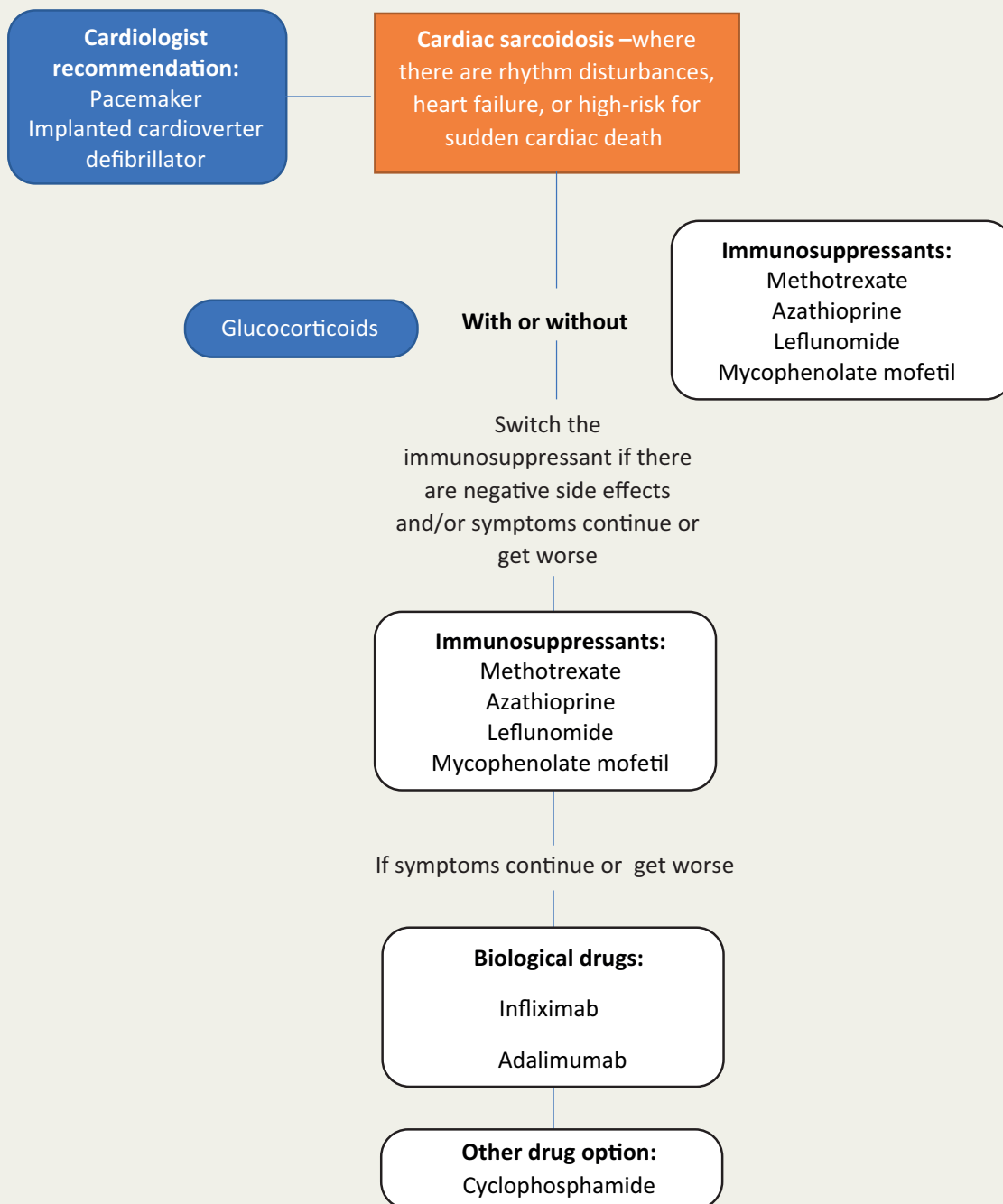


## Cutaneous sarcoidosis –affecting the skin



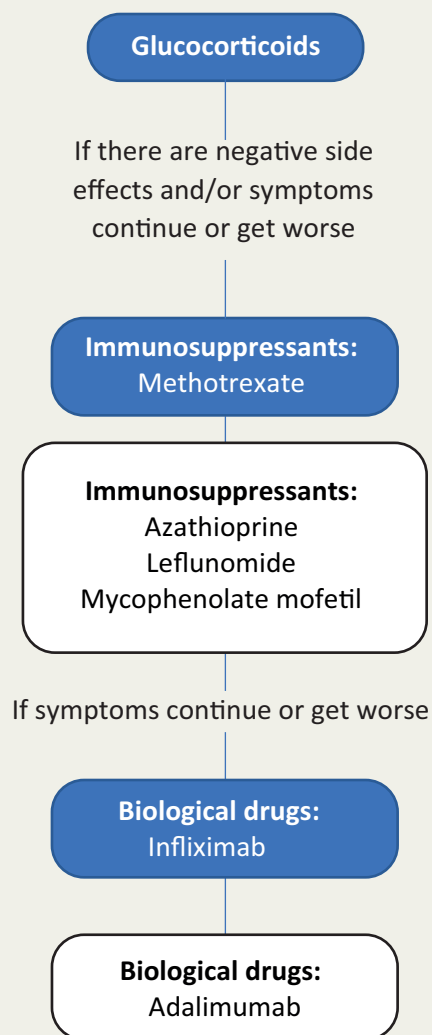


## Cardiac sarcoidosis –affecting the heart





## Neurosarcoidosis – mainly affecting the nervous system





## Fatigue caused by sarcoidosis

### Fatigue caused by sarcoidosis

No response to treating  
ongoing sarcoidosis  
symptoms

Exercise training  
Inspiratory muscle training

Continued symptoms

Neurostimulators  
Armodafinil  
D-Methylphenidate

Continued symptoms

Low dose of glucocorticoids  
and/or methotrexate



## Future research

The recommendations in this guideline are based on a review of the existing evidence and on the experience of the experts working on the document. The authors of the study noted that there was a lack of research in many areas of sarcoidosis. In particular, there was a lack of evidence covering forms of sarcoidosis that affects the liver, bones or eyes. There was also a lack of evidence showing whether treatments are effective at improving quality of life, rather than just improving clinical measurements. The authors estimate that an update of the guideline will be needed within 5 years as more information becomes available.

## Further reading

This guideline was produced by the European Respiratory Society and the European Lung Foundation. You can find out more about these organisations and access the full professional guideline using the links below:

### Full clinical guideline - published in the European Respiratory Journal:

<https://erj.ersjournals.com/content/early/2021/06/10/13993003.04079-2020>



## Further resources for patients and carers:

European Lung Foundation's Patient Priorities website:  
<https://europeanlunginfo.org/sarcoidosis/> This provides information for people with sarcoidosis and their family and friends. Developed with the help of people with experience of the condition and sarcoidosis healthcare experts across the world.

European Lung Foundation's Sarcoidosis factsheet: <https://europeanlung.org/en/information-hub/factsheets/sarcoidosis/>



## About ERS

The European Respiratory Society (ERS) is an international organisation that brings together physicians, healthcare professionals, scientists and other experts working in respiratory medicine. It is one of the leading medical organisations in the respiratory field, with a growing membership representing over 140 countries. The ERS mission is to promote lung health in order to alleviate suffering from disease and drive standards for respiratory medicine globally. Science, education and advocacy are at the core of everything it does. ERS is involved in promoting scientific research and providing access to high-quality educational resources. It also plays a key role in advocacy – raising awareness of lung disease amongst the public and politicians. [www.ersnet.org](http://www.ersnet.org)

## About ELF

The European Lung Foundation (ELF) was founded by ERS to bring together patients and the public with professionals. ELF produces public versions of ERS guideline to summarise the recommendations made to healthcare professionals in Europe, in a simple format for all to understand. These documents do not contain detailed information on each condition and should be used in conjunction with other patient information and discussions with your doctor. More information on lung conditions can be found on the ELF website: [www.europeanlung.org](http://www.europeanlung.org)