

# ERS clinical practice guidelines on treatment of sarcoidosis.

Guideline ID: 2288

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## European Respiratory Society (ERS)

*Baughman RP, Valeyre D, Korsten P, et al. ERS clinical practice guidelines on treatment of sarcoidosis. Eur Respir J. 2021 Dec 16;58(6):2004079. [62 references] [PubMed](#)*

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## Overview

### Guideline Objective

To provide guidance to physicians treating the very heterogenous sarcoidosis manifestations

### Patient Population

Patients with pulmonary, cutaneous, cardiac, or neurosarcoidosis

## Recommendations

### Recommendation Statements

### Pulmonary Sarcoidosis

## **Patients, Intervention, Comparator, Outcomes (PICO) 1: In patients with pulmonary sarcoidosis, should glucocorticoid treatment be used *versus* no immunosuppressive treatment?**

For untreated patients with major involvement from pulmonary sarcoidosis believed to be at higher risk of future mortality or permanent disability from sarcoidosis, we recommend the introduction of glucocorticoid treatment to improve and/or preserve forced vital capacity (FVC) and quality of life (QoL). (**Strong recommendation; Low quality of evidence**)

## **PICO 2: In patients with pulmonary sarcoidosis, should one add immunosuppressive treatment or remain on glucocorticoid treatment alone?**

For patients with symptomatic pulmonary sarcoidosis believed to be at higher risk of future mortality or permanent disability from sarcoidosis who have been treated with glucocorticoids and have continued disease or unacceptable side-effects from glucocorticoids, we suggest the addition of methotrexate to improve and/or preserve FVC and QoL. (**Conditional recommendation; Very low quality of evidence**)

For patients with symptomatic pulmonary sarcoidosis believed to be at higher risk of future mortality or permanent disability from sarcoidosis who have been treated with glucocorticoids or other immunosuppressive agents and have continued disease, we suggest the addition of infliximab to improve and/or preserve FVC and QoL. (**Conditional recommendation; Low quality of evidence**)

## **Cutaneous Sarcoidosis**

### **PICO 3: In patients with cutaneous sarcoidosis, should glucocorticoid treatment be used *versus* no immunosuppressive treatment?**

For patients with cutaneous sarcoidosis and cosmetically important active skin lesions which cannot be controlled by local treatment, we suggest oral glucocorticoids be considered to reduce skin lesions. (**Conditional recommendation; Very low quality of evidence**)

### **PICO 4: In patients with cutaneous sarcoidosis, should one add other immunosuppressive treatment when treatment with glucocorticoids has not**

## been effective?

For patients with cutaneous sarcoidosis who have been treated with glucocorticoids and/or other immunosuppressive agents and have continued cosmetically important active skin disease, we suggest the addition of infliximab compared to no additional treatment to reduce skin lesions. (**Conditional recommendation; Low quality of evidence**)

## Cardiac Sarcoidosis

**PICO 5: In patients with clinically relevant cardiac sarcoidosis, should glucocorticoids with or without other immunosuppressives *versus* no immunosuppression be used?**

For patients with evidence of functional cardiac abnormalities, including heart block, dysrhythmias or cardiomyopathy, we recommend the use of glucocorticoids (with or without other immunosuppressives). (**Strong recommendation; Very low quality of evidence**)

## Neurologic Disease

**PICO 6: In patients with neurosarcoidosis, should immunosuppressive treatment be used *versus* no immunosuppressive treatment?**

For patients with clinically significant neurosarcoidosis, we recommend treatment with glucocorticoids. (**Strong recommendation; Very low quality of evidence**)

For patients with neurosarcoidosis that have been treated with glucocorticoids and have continued disease, we suggest the addition of methotrexate. (**Conditional recommendation; Very low quality of evidence**)

For patients with neurosarcoidosis that have been treated with glucocorticoids and a second-line agent (methotrexate, azathioprine, mycophenolate mofetil) and have continued disease, we suggest the addition of infliximab. (**Conditional recommendation; Very low quality of evidence**)

## Fatigue

## **PICO 7: In patients with sarcoidosis-associated fatigue, should immunosuppressants, neurostimulants, exercise or other treatments be used *versus* no treatment for fatigue?**

In patients with sarcoidosis who have troublesome fatigue, we suggest a pulmonary rehabilitation programme and/or inspiratory muscle strength training for 6–12 weeks to improve fatigue. (**Conditional recommendation; Low quality of evidence**)

In patients with sarcoidosis who have troublesome fatigue that is not related to disease activity, and after consideration of a pulmonary exercise or rehabilitation programme, we suggest the use of D-methylphenidate or armodafinil for 8 weeks to test its effect on fatigue and tolerability. (**Conditional recommendation; Low quality of evidence**)

## **Small-fibre Neuropathy**

### **PICO 8: In sarcoidosis patients with small-fibre neuropathy, should immunosuppressants or intravenous immunoglobulin be prescribed *versus* no treatment?**

No recommendations were made for this PICO question due to a lack of sufficient evidence.

## **Evidence Rating Scheme**

Certainty in the body of evidence was assessed using Grading of Recommendations, Assessment, Development and Evaluation (GRADE) methodology.

## **Recommendation Rating Scheme**

Recommendations were graded as **strong** or **conditional** after considering the quality of the evidence, the balance of desirable and undesirable consequences of compared management options, the assumptions about the relative importance of outcomes, the implications for resource use, and the acceptability and feasibility of implementation. The panel formulated the clinical recommendations and decided on their strength first by consensus and then by voting for final recommendations. Following the GRADE approach, strong recommendations were worded as "we recommend," while conditional recommendations were worded as "we suggest."

A **strong** recommendation was made for an intervention when the panel was certain that the desirable consequences of the intervention outweighed the undesirable consequences and a strong recommendation against an intervention was made when the opposite was true. A strong recommendation indicates that most patients and healthcare providers would choose to have, or not to have, the intervention.

A **conditional** recommendation for an intervention was made when the panel was uncertain that the desirable consequences of an intervention outweighed the undesirable consequences in most patients and a conditional recommendation against an intervention was made when uncertainty existed that undesirable consequences of an intervention outweighed the desirable consequences in most patients. Reasons for uncertainty included low quality of evidence, a close balance between desirable and undesirable effects or patients' values and preferences. A conditional recommendation indicates that different patients and healthcare providers may make different choices regarding an intervention.

## Related Content

### Supporting Documents

- [Supplement 1: Individual Therapies.](#)
- [Supplement 2: Evidence Summaries and Evidence to Decision Tables for All PICOs.](#)
- [Using the GRADE Approach for Guideline Development: A Step-by-Step Guide.](#)

### Implementation Tools

- [Shareable Guideline](#); 2021.

### Patient Education

No patient education materials available.

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# TRUST Scorecard

## Composition of Guideline Development Group (GDG)

Multidisciplinary GDG Members

Yes

Methodologist Involvement

Yes

Incorporation of Patient and Public Perspective



## Systematic Review of Evidence

Literature Search



Study Selection



Evidence Synthesis



## Foundations for Recommendations

Strength of Evidence Grade



Description of Benefits and Harms of Recommendations



Summary of Evidence Supporting Recommendations



Strength of Recommendations Rating



Clear Articulation of Recommendations



## Funding Source

Yes

## Disclosure and Management of Financial Conflicts of Interests



## External Review



## Updating

