Antisynthetase syndrome

If your doctor thinks you may have antisynthetase syndrome, or if you have received a diagnosis, it is important that you understand as much as you can about the condition and how you can best manage it.

Centre of Research Excellence in **Pulmonary Fibrosis**

Lung Foundation Australia

Contents

What is antisynthetase syndrome?

Antisynthetase syndrome is a rare chronic condition that can affect multiple parts of your body including your muscles, joints, lungs, skin or blood vessels. It is classified as a type of autoimmune disease, which means that your immune system attacks your own body, causing inflammation.

Antisynthetase syndrome can result in muscle weakness, joint pain, skin rashes, and breathing difficulties, but the impact of the condition on your body can vary greatly from one person to another.

Link between antisynthetase syndrome and interstitial lung disease

People with antisynthetase syndrome also have a higher risk of developing interstitial lung disease.

69–100%

of people with antisynthetase syndrome will develop interstitial lung disease at some point

What are the signs and symptoms of antisynthetase syndrome?

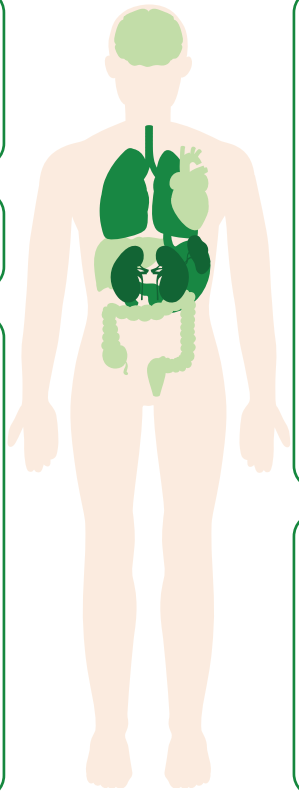
Every person is unique, and so the signs, symptoms and severity associated with antisynthetase syndrome may vary from one person to another.

Some of the possible signs and symptoms include:

Lungs (interstitial lung disease)
- Shortness of breath
- Coughing

Joints (polyarthritis)
- Pain and stiffness

Hands
- Thickened, cracked skin on your palms and the sides of your fingers (mechanic's hands)
- Colour changes and numbness in your fingertips and toes. Particularly in cold conditions (Raynaud's phenomenon)



General
- Fever
- Fatigue
- Loss of appetite
- Unintended weight loss
- Difficulty swallowing
- Skin rash, often spreads over the face or eyelids, or across the shoulder and upper back

Muscles
- Muscle inflammation (myositis)
- Muscle pain (myalgia)
- Muscle stiffness
- Muscle weakness

You may not experience all of these symptoms at once, however, they may develop at different times.

What causes antisynthetase syndrome?

The exact causes of antisynthetase syndrome are unknown, but it is thought to be related to production of a specific autoantibody (such as Anti-Jo-1) that attacks healthy tissues in the body because of an over-active immune system. People with antisynthetase syndrome have autoantibodies that target and attack certain enzymes in the body called aminoacyl-tRNA synthetase.

Did you know?
- Antibodies are proteins that are part of the body's immune system. Autoantibodies are antibodies that mistakenly attack healthy tissue in the body
- In antisynthetase syndrome, autoantibodies may attack aminoacyl-tRNA synthetase, an enzyme that helps to regulate the production of other proteins.

Who is affected by antisynthetase syndrome?

Anyone can develop antisynthetase syndrome.

Women are twice as likely to be affected than men

It can develop at any time from the teenage years to the elderly. Average age at diagnosis is around 50 years.

How is antisynthetase syndrome diagnosed?

Antisynthetase syndrome is usually diagnosed based on the symptoms you are experiencing, your medical history and the results of some tests.

Tests to help make a diagnosis may include:

Blood tests

Check for autoantibodies associated with antisynthetase syndrome such as anti-Jo-1, PL-7, PL-12, OJ, EJ, Ku or MDA5 antibodies.

The anti-Jo-1 antibody is the most common antisynthetase antibody.

High resolution CT scan (HRCT)

Look to see if your lungs are affected

Lung function tests

Determine how well your lungs are working

Electromyography, muscle MRI and/or muscle biopsy

Help determine the health of your muscles

Nailfold capillaroscopy

Investigates how well the circulation in your fingers works

There are specific medical criteria to help diagnose antisynthetase syndrome, but your doctor will consider all your symptoms and test results to confirm the diagnosis.

How is antisynthetase syndrome treated?

Although there is no cure for antisynthetase syndrome, you and your doctor will work together to find a treatment that works for you. Your treatment will aim to reduce inflammation, manage your symptoms, minimise the impact on your daily life and help prevent further damage to your body.

There is no single specific treatment for antisynthetase syndrome, so you may receive one or a combination of treatments depending on your symptoms.

Initial treatment

When you are first diagnosed, you may need to begin more intensive treatment straight away to reduce the inflammation and help prevent long-term damage to your lungs or other parts of your body. This may include:

Immunosuppressants help stop your immune system from attacking your body. Depending on the severity of your condition, you may be prescribed a combination of two or three different types, which can be given either orally or through an intravenous (IV) line:

- IV Cyclophosphamide or IV Rituximab
- Oral Mycophenolate, Tacrolimus, or Azathioprine

High doses of **corticosteroids** to help reduce inflammation

IV immunoglobulins to help reduce inflammatory effects

- Immunoglobulins are antibodies produced by the body's natural immune system to help fight infection and disease. IV immunoglobulin is a treatment that combines immunoglobulins donated by different people (through blood donations) into a treatment for fighting a range of autoimmune diseases

Plasma exchange, although rarely used, removes some antibodies found in your plasma that may be causing the disease and replaces them with donor plasma

- Plasma along with red blood cells, white blood cells and platelets combine to make up your blood

In severe cases, you may be admitted to hospital to receive urgent treatment under close medical supervision

Ongoing treatment

Antisynthetase syndrome is a chronic condition, meaning treatment may need to be continued for several years or even longer, depending on your individual situation:

Corticosteroids

Immunosuppressants including oral mycophenolate, azathioprine, tacrolimus or sometimes IV rituximab

Anti-fibrotic therapies such as nintedanib may be used if you have progressive scarring (fibrosis) on your lungs despite immunosuppressive treatment

IV immunoglobulin

It is important to look after yourself when living with antisynthetase syndrome to get the most out of your treatment and care. There are a range of ways to help manage your symptoms and maintain your health, including:

Participating in a pulmonary rehabilitation program or physiotherapy to help manage any muscle or breathing symptoms – to learn more, visit the **Exercise and Pulmonary Fibrosis** fact sheet

Maintaining a healthy diet and daily exercise – to learn more, visit the **Life with Pulmonary Fibrosis** booklet

Keeping up to date with vaccinations – to learn more, visit the **Preventing and managing respiratory infections** booklet

Oxygen therapy if prescribed – to learn more, visit the **How can oxygen therapy help?** booklet

Speaking with your healthcare team about whether **a referral to palliative care** would be beneficial as part of your ongoing care. To learn more about palliative care, see our **How can palliative care help you?** resource.

For more information on looking after yourself, see our **Living with PF: Non-pharmacological Treatments** resource.

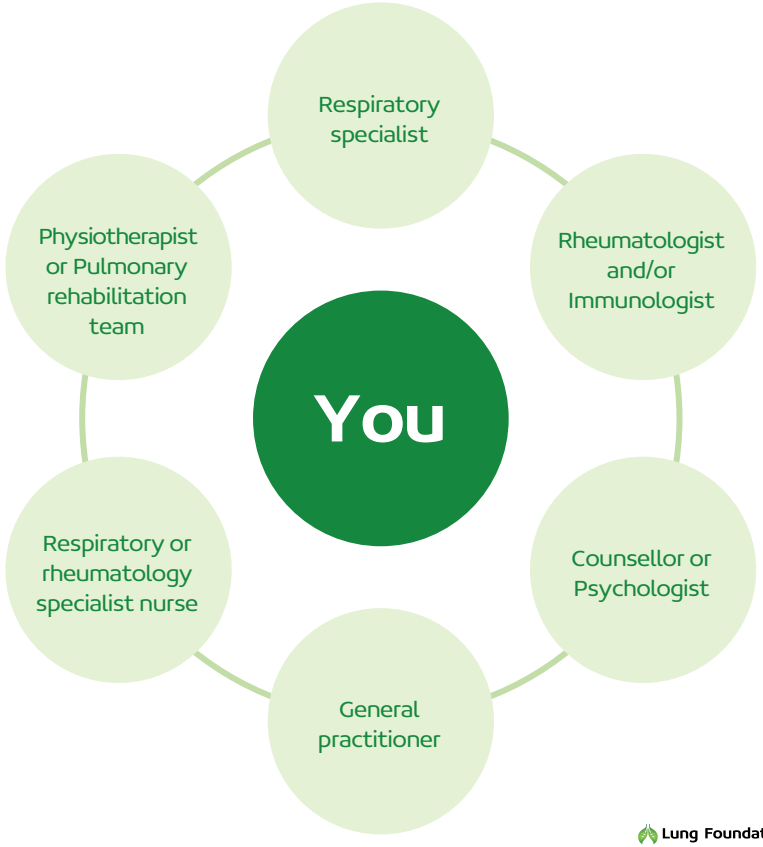
What support is available?

Receiving a diagnosis of antisynthetase syndrome can be difficult. It is a serious condition that can cause severe symptoms in some cases. This is why it is important to stay connected with your healthcare team and seek support when you need it.

As antisynthetase syndrome affects different parts of your body, you will have a team of health professionals around you to help manage the condition and ensure you get the best treatment possible.

If you have other health conditions, it is important that these are also managed and coordinated by your GP.

Some of your healthcare team members may include:



To connect with support services and additional information, contact Lung Foundation Australia's Information and Support Centre on **1800 654 301** (option 3) or learn more **here.**

Lung Foundation Australia Services

- Information and Support Team
- Lung disease information resources
- Education webinars
- Support groups
- Peer-to-peer connections
- Referral to pulmonary rehabilitation and Lungs in Action exercise programs
- E-newsletter

EXTERNAL LINKS

Myositis Association Australia	https://myositis.org.au/

We thank the individuals who contributed to the content and expert review of this fact sheet, in particular Dr Matthew Parker, Rheumatologist and Professor Tamera Corte, Respiratory Physician.

In collaboration with

Centre of Research Excellence in **Pulmonary Fibrosis**

Freecall 1800 654 301
enquiries@lungfoundation.com.au
lungfoundation.com.au
PO Box 1949 Milton, QLD 4064

Lung Foundation Australia