

ANTIYSNTHETASE SYNDROME (ASYS)

What is ASyS?

Antisynthetase syndrome (ASyS) is one of the inflammatory myopathies (IIM) and is sometimes called Anti-Jo1 syndrome. ASyS is an autoimmune condition characterized by having autoantibodies to aminoacyl transfer RNA synthetases (anti-ARS). ASyS includes symptoms and clinical features like myositis, polyarthralgia, Raynaud's phenomenon, Mechanic's hands, and interstitial lung disease (ILD).

How is ASyS diagnosed?

Diagnosing ASyS can be difficult and for some it can take many months or years. ASyS is a syndrome, so diagnosis is made based on specific criteria. A diagnosis is made when there is the presence of antisynthetase antibodies (including Anti-Jo-1 and others), plus two major criteria or one major and two minor criteria:

- **Major Criteria.** Myositis symptoms (eg. weakness in arms and legs, difficulty swallowing, and/or skin lesions) as well as ILD.
- **Minor Criteria.** Diagnosis of polyarthrititis, Raynaud's phenomenon, and Mechanic's hands.

Diagnosis may include exams and several tests:

- First, doctors usually get a patient history and conduct a physical exam.
- Then, they may order blood tests to check for myositis-specific antibodies, muscle enzymes (CK), and other inflammatory, autoimmune, and cancer markers.
- Often, diagnosis is confirmed after an MRI, nerve conduction test, EMG, and muscle biopsy.

What are the symptoms of ASyS?

Because ASyS is a syndrome, there are a variety of possible symptoms and not everyone with ASyS will experience every symptom. Certain antibodies are associated with specific symptoms:

- Anti-Jo-1: myopathy, severe arthritis, and Mechanic's hands.
- Anti-PL-12: higher rates of Raynaud's phenomenon.
- Anti-PL-7, anti-PL-12, anti-KS, and anti-OJ: higher rates of ILD.
- Anti-PL-7: presence of heliotrope rash.

Often, patients with non-Jo-1 autoantibodies, such as PL7 or PL12, are more likely to be Black *and* have more severe disease. ASyS symptoms depend on what other conditions you are diagnosed with. Those with DM or PM experience muscle inflammation, leading to symmetrical weakness in the muscles closest to the body's core, such as hips, neck, shoulders, upper arms, and thighs. Those with DM may experience skin symptoms like rashes and itching.

Those living with ASyS may look healthy while struggling internally. Others may use oxygen and require assistive devices for mobility. Everyone with ASyS is different.

How is ASyS managed?

While there is no cure, there are a variety of medications that can be used to treat ASyS, depending on your diagnosis criteria. Continuing to remain active and using assistive devices can also help manage symptoms. Most people with ASyS manage with a combination of the following:

- **Medication.** ASyS treatments often start with topical, oral, and/or IV steroids. Off-label use of other medications may be successful, including antimalarials, immunosuppressive agents, chemotherapy medications, and infusion therapies, like IVIG, and monoclonal antibodies, like rituximab.
- **Assistive devices.** A cane, walker, rollator, or wheelchair can help improve balance or weakness. Assistive devices can be used for short periods, such as during flares, or for long-term muscle weakness or damage.
- **Movement.** Physical therapy and exercise are usually recommended for all myositis patients. Talk with your doctor before starting any exercise routine.

Where can I find more information?

Myositis Support and Understanding has a robust website full of resources including information about IIM, how to manage these difficult conditions, and where to find support. Go to www.understandingmyositis.org or use this QR code to learn more!

