

## Dr. Lisa Christopher-Stine: Polymyositis? It's more likely something else

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### EXPERT ANALYSIS FROM THE WINTER RHEUMATOLOGY SYMPOSIUM

True polymyositis is rare, so it's important to carefully consider other likely diagnoses, according to Lisa Christopher-Stine, MD.



Dr. Lisa Christopher-Stine

“When someone refers you [a patient with suspected] polymyositis, I want you to do a checklist in your head and say, ‘Have I thought about these five things?’ ” [Dr. Christopher-Stine](https://www.hopkinsmyositis.org/doctors/dr-lisa-christopher-stine/) <<https://www.hopkinsmyositis.org/doctors/dr-lisa-christopher-stine/>> , director of the Johns Hopkins Myositis Center, Baltimore, said at the Winter Rheumatology Symposium sponsored by the American College of Rheumatology.

The five most common diagnoses in patients labeled as having polymyositis are immune-mediated necrotizing myopathy (IMNM), overlap with other rheumatologic conditions, antisynthetase syndrome, inclusion body myositis (IBM), and muscular dystrophy, she explained.

“You may say, ‘look, it’s all what you call it,’ but I think we need to be a little bit more careful in what we call it,” she said.

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#### IMNM

Patients with IMNM present with clinical symptoms similar to those seen in polymyositis and dermatomyositis – mainly proximal muscle weakness.

However, there are some important differences, both clinically and histologically, Dr. Christopher-Stine said.

“Look for higher [creatinine kinase (CK)] levels,” she said. “In the thousands, usually multiple thousands ... like 5,000, 10,000, 2,000 ... that’s when you’re thinking about a necrotizing phenotype before you even look at the biopsy.”

CK levels will usually be under 30,000 U/L in IMNM, she noted, adding that data increasingly suggest that the extensive muscle necrosis in IMNM explains the elevated CK levels versus those seen in other myopathies.

Myalgias also tend to be more prominent in IMNM than in polymyositis.

“These folks hurt,” she said, noting that IMNM patients tend to have more extensive muscle atrophy and functional disability. “Many will be wheelchair bound within 9 months of diagnosis; it’s not subtle.”

The most important tool for making an IMNM diagnosis is muscle biopsy; look for prominent myocyte necrosis and a relative paucity of lymphocytes, she advised.

## Overlap

Sometimes patients with polymyositis also have other rheumatologic conditions that shouldn’t be overlooked, therefore “overlap is its own category,” she said.

“In our experience, the most common overlap is scleroderma,” she noted, adding that the scleroderma is often, but not always, subtle, and that there may be overlapping autoantibodies.

Overt sclerodactyly is rarely seen, although a small amount may be present, but significant Raynaud’s phenomenon is common in these patients, and tiny telangiectasias across the neck are a tell-tale sign.

“Why does that matter? It’s not an esoteric argument; those are the folks that go on to have pulmonary hypertension,” she said. “They can have the same [interstitial lung disease] and all of the other internal scleroderma manifestations.”

Think about overlap and “look close phenotypically and with antibodies,” she advised.

There is also “the typical RA seropositive overlap,” she said, but lupus only rarely overlaps with myositis.

“However, the next diagnosis on the list – antisynthetase syndrome – can be a *forme fruste* where you first see a seronegative RA-like picture, and it’s important to think about that as well,” she said.

## **Antisynthetase syndrome**

In patients referred for polymyositis, it’s also important to evaluate for antisynthetase syndrome, Dr. Christopher-Stine said.

The arthritis seen in the extramuscular phenotype of the syndrome is rarely deforming, but despite what many physicians were taught, “it absolutely can be erosive,” she said.

In fact, 40% of people with this syndrome present with an isolated *forme fruste* seronegative rheumatoid arthritis, she said.

Roughening and desquamation of the skin on the radial surface of fingers or palms – a sign known as mechanic’s hands – that doesn’t have another identifiable cause suggests this diagnosis in patients with this type of arthritis, as does interstitial lung disease and Raynaud’s phenomenon.

The Raynaud’s can be “fairly significant in the sense that it is bothersome,” but it usually doesn’t lead to ulceration or digital necrosis.

This is different from what is seen with the scleroderma phenotype, she said, adding that “if you’re starting to see gangrene and digital loss, think of something else.”

## **IBM**

IBM is “probably the No. 1 most-missed diagnosis” among patients referred for what is initially believed to be polymyositis, Dr. Christopher-Stine said.

“I used to think that this was missed at entry, that everybody [with IBM] had all of these criteria and that rheumatologists really didn’t understand this phenotype ... but some people morph into this,” she said, explaining that they often start out looking like they have polymyositis with proximal muscle weakness.

“They may even initially respond to steroids. And then they get this phenotype,” she said.

Older men are more likely to present with the phenotype from the beginning; women, in her experience, tend to present with what appears to be polymyositis, and then develop the phenotype over time, she noted.

An IBM diagnosis requires age over 30 years, but most patients are over 50, she said.

“This is the only one of the myopathies that is preferential to men,” she added, noting that it affects men twice as often as it does women.

The syndrome is characterized by proximal strength loss and muscle atrophy. Also, a finding that a patient’s knee extensors are weaker than their hip flexors is “a fantastic bedside sign” differentiating IBM from polymyositis, she said.

That’s not to say IBM patients don’t have hip flexor weakness, but their knee extensors usually are “considerably weaker by a grade strength or more” versus their hip flexors, she explained.

“It’s a very easy bedside test. In typical other myopathies we have this, but the knee extensors aren’t that weak in general, or they’re not as weak as the hip flexors,” she added.

Another sign is distal strength loss, particularly in the forearm and finger flexors.

“I was taught to have them make a fist; don’t have them make a fist,” she said, explaining that this recruits intrinsic muscles which basically allows cheating that may mask weakness.

Instead, ask them to flex just their distal interphalangeal joints by making a claw and using the fingers to pull against your fingers, she suggested.

Mixed myopathic and neuropathic features on electromyography also indicate IBM, she said.

Muscle biopsy may be helpful, but inclusions are seen in less than one-third of IBM patients.

“At times, we have had to biopsy three times to see them at all, and some people never show them, so you have to rely on your clinical acumen if you don’t see them,” she said.

Also, keep in mind that these patients are often labeled as having treatment-resistant polymyositis.

“Please, when somebody refers to you somebody that’s treatment resistant, that may be the case, but I want you to think maybe they’re treatment resistant because they don’t have that disease.”

## Muscular dystrophy

Some cases of myositis mimic certain types of muscular dystrophy, Dr. Christopher-Stine said, providing a checklist of muscular dystrophies that can look “clinically completely indistinguishable from a typical inflammatory myopathy,” and should therefore be considered in these patients.

The checklist includes Duchenne’s manifesting carrier, limb girdle muscular dystrophy type 2b, myotonic dystrophy (usually type 2), and facioscapulohumeral muscular dystrophy.

Dr. Christopher-Stine reported having intellectual property interest in a novel Inova Diagnostics autoantibody assay detection for anti-HMGCR. She was also the safety officer for the JBT-101 Trial sponsored by Corbus and funded by the National Institutes of Health.

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