

What is inclusion body myositis?

STORY BY CONNIE BRYSON / PHOTO BY VEER



WHEN CALGARY NEUROLOGIST DR. CHRIS WHITE sees an older patient who has a tendency to fall or trip or who can't make a fist, his mental checklist includes a disease called inclusion body myositis (IBM). "This disorder is relatively common and is always a consideration when an older patient presents with progressive weakness especially when it causes falls," says Dr. White.



IBM is one of a group of myopathies (muscle diseases) that also includes polymyositis and dermatomyositis. Each of these three diseases has a different prognosis and response to therapy. Whereas the other two are treatable, IBM is not. IBM usually appears after the age of 50 and progresses slowly. It can affect all muscle

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groups and results in substantial weakness and atrophy. The first muscles affected are often the quadriceps in the legs, which makes patients prone to falling, and the finger flexors, which are needed to make a fist. Up to 40 percent of patients also have some difficulty swallowing. In a few of the most

Quick Facts

IBM: inclusion body myositis; a slow progressing muscle disease that can result in weakness and atrophy

severe cases, patients must have a feeding tube.

The prevalence of IBM is estimated at approximately nine cases per million people; the disease is more common in men than women. Based on his experience, Dr. White suspects that this estimate is low. He notes that he has about six patients with IBM in his care, and he expects that his four colleagues in the neuromuscular clinic have a similar number of patients.

The cause of IBM is unknown, but it is a subject of research. Some symptoms suggest that it is an autoimmune disease like rheumatoid arthritis and the other inflammatory myopathies. Other clinical findings, such as abnormal proteins found in muscle biopsies, suggest that IBM is a degenerative condition like Alzheimer's. "The question is whether IBM is a degenerative condition with secondary inflammatory changes or an inflammatory condition with secondary degenerative changes," explains Dr. White. "The jury is still out."

Although research has not yet solved this fundamental puzzle, it has led to some discoveries that may help physicians make a diagnosis. For example, a recent scientific paper identified a certain degenerative protein called TDP-43 that is present almost all of the time in IBM but not in other inflammatory myopathies. "This is potentially very helpful," says Dr. White. "Unless you have some reason to suspect IBM or recognize the pattern of weakness, it can be tough to make a diagnosis. Plus, there are some patients who don't have the characteristic pattern of weakness, which makes diagnosis even more difficult. A definitive muscle biopsy test would be helpful."

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Many treatments have been tried unsuccessfully, including steroids and intravenous immunoglobulin. Experimental therapies for IBM are currently in clinical trials. They include a drug that blocks a protein called tissue necrosis factor, which is involved in inflammation, and an agent that induces cells to produce heat shock protein, which is known to protect cells. These trials are ongoing.

The diagnosis of IBM, while disheartening, should not be considered devastating, says Dr. White. There is no question that quality of life is affected

and that muscle weakness gets worse over time, yet most people are able to carry on walking and using their hands, which are crucial to maintaining independence. ✱

About this feature

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About the researcher

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