

# General physician information about Inclusion Body Myositis (IBM)

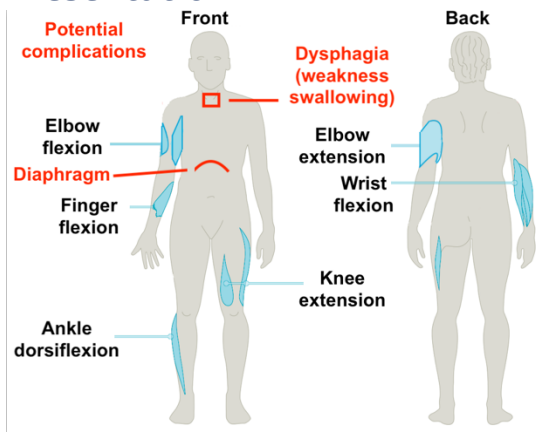
## Overview

Inclusion body myositis (IBM) is a chronic, sporadic, adult-onset myopathy characterised by a slowly progressive course over years, ultimately leading to significant functional impairment. Weakness usually presents asymmetrically, most notably affecting the quadriceps and finger flexors. It is the most common acquired muscle disease in individuals over 50 (prevalence approximately 180 per million). There are no approved drug treatments; however, early diagnosis and proactive management—including fall risk assessment, management of dysphagia and comorbidities, and a personalised, supervised exercise programme—can minimise complications and help maintain quality of life.

## Incidence

IBM is strongly age-related, with a typical onset in the mid-60s and a wide range at presentation. Around one in five patients reports symptom onset before age 50.

## Presentation



IBM affects individuals at different ages with varying patterns and rates of muscle involvement, but a hallmark is marked weakness of the quadriceps and deep finger flexors. Weakness often begins in the quadriceps, leading to toe or foot drop and unexplained falls due to knee buckling or tripping from reduced ankle dorsiflexion; in others, it starts in the forearm and hand flexors, causing difficulty pinching, buttoning, making a fist, and gripping objects. Oropharyngeal weakness is common, and dysphagia may be an early or presenting symptom in many patients. Respiratory involvement usually results from diaphragm weakness rather than primary cardiac disease; reduced vital capacity is frequent, but IBM is not typically

linked with primary cardiomyopathy. IBM occurs twice as often in men as in women. Phenotypic differences related to sex, age at onset, and race (including between Black and White patients) have been described. Many patients report significant muscle pain and bothersome buildup of thick saliva or bronchial mucus. Malnutrition from fear of choking, depression, and anxiety is common.

## Diagnosis

Typically diagnosed by a neurologist through clinical examination, EMG, biopsy, MRI, and serology (cN1A) antibodies, which are present in 50% of patients. Creatine kinase (CK) is usually normal or only slightly elevated, generally up to about 10 times the upper normal limit. Most patients need a biopsy. The average duration from symptom onset to accurate diagnosis is five years. It is often initially misdiagnosed as polymyositis, which has a faster onset, higher CK levels, and responds to corticosteroids. “PM-Mito” is a precursor of IBM. Other muscular dystrophies (e.g., limb-girdle, Becker) and ALS should be excluded.

## Progression

Progression rates are variable but slow, relentless, and cumulative, leading to irreversible loss of muscle function. On average, five years after onset, many patients require aids to walk; within ten years, a power wheelchair and home modifications are generally necessary (bathroom/bedroom lifts, etc.).

## Pathology/Causes

IBM is a complex inflammatory-degenerative myopathy characterised by endomysial inflammation involving CD8+ T-cells and macrophages infiltrating non-necrotic muscle fibres (without identified antigen), abnormal protein aggregation with rimmed vacuoles (including mislocalised TDP-43 and other proteins), and notable mitochondrial abnormalities. Evidence indicates a multifactorial pathogenesis involving genetic susceptibility, age-related changes, and immunosenescence; no definitive initiating cause has been found.

## Diagnosis

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## Complications

Falls and fractures are common due to gait instability. Dysphagia is very frequent and increases the risk of aspiration (50% will require a PEG feeding tube). Diaphragmatic weakness presents as nocturnal hypoventilation that resembles sleep-disordered breathing, with significant daytime fatigue and hypercapnia. Poor oral intake, chronic dysphagia, and feeding difficulties can also result in malnutrition. IBM-related causes of death are typically respiratory complications (such as aspiration pneumonia) or malnutrition.

## Comorbidities

Overlap with other autoimmune conditions is frequent in IBM, especially with rheumatoid arthritis and Sjögren's syndrome. Patients may experience higher incidences of peripheral neuropathy, hematologic malignancies (notably T-LGLL), hypertension, hyperlipidaemia, diabetes, and anaemia. Depression, anxiety, chronic pain, and fatigue are common and can significantly impair function.

## Treatment/Management

The standard of care is non-pharmacological, focusing on maintaining function, safety, and quality of life, with emotional and psychological support as well as fall prevention. Input from physical, speech, and occupational therapy is recommended. An individualised exercise programme is advised. Baseline and periodic assessments of dysphagia (e.g., videofluoroscopic or endoscopic swallow studies) and of respiratory involvement (spirometry, overnight oximetry, or sleep study) are recommended. Palliative care is beneficial in the late stages.

## More Information

An overview with practical information for patients and physicians, and curated research highlights:  
<https://www.ibmmyositis.com/>

Recent reviews:  
(Open Source). <https://doi.org/10.1097/bor.0000000000000958>  
<https://doi.org/10.3390/ijms25052742>  
<https://pubmed.ncbi.nlm.nih.gov/40471678/>  
<https://doi.org/10.1212/cont.0000000000001616>

## Credits

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