Information for General Physicians about inclusion body myositis. (2024) By William Tillier.

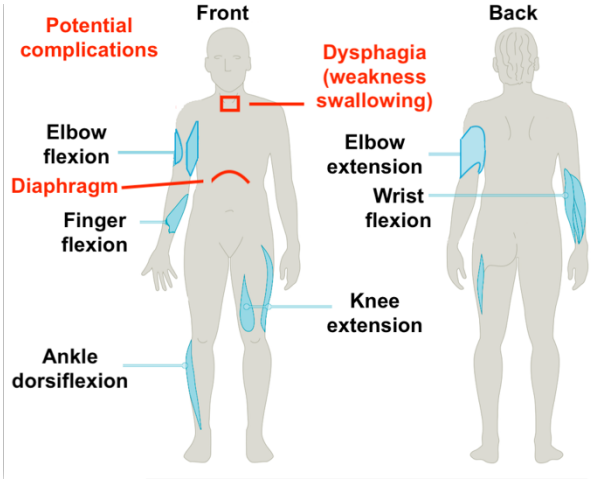
Overview: Inclusion body myositis (IBM) is a sporadic/rare muscle disease usually seen after 40. It is poorly understood, chronic, and progressive. A characteristic pattern of weakness of finger flexion, knee extension and ankle dorsiflexion develops over months or years, leading to profound disability (wheelchair). No treatment.

Cause: No cause or trigger event is known. Two major theories are autoimmune or myodegenerative disorder. The interplay of environment, genetic predisposition, and aging is implied. IBM is sporadic and acquired.

Incidence: IBM is age-related. The median onset is 55-65, with wide variation: 20% show symptoms before 50. There are estimated to be 20,000 - 26,000 cases in the US. The most common acquired muscle disease after 50.

Pathology: T cells, myeloid dendritic cells, macrophages, and plasma cells all invade IBM muscle, thought to arise from chronic stimulation by an unknown antigen. Inclusion bodies of abnormal proteins (amyloid/TDP-43) and rimmed vacuoles are also seen. Anti-cN1A antibodies are found in the blood in ~50% of patients.

Presentation: IBM has unique clinical features (elbow, wrist & finger flexors; elbow and knee extenders; ankle dorsiflexion). Presentation varies widely – people are affected at different ages, in different ways, to different degrees, and at various rates. Seen 2X as often in men. Progression may be more rapid in men and those with later onset. Quadriceps may be affected first; toe drop, unexpected falling, and tripping are common first symptoms. Falls from knee-buckling due to quadriceps weakness or tripping due to ankle dorsiflexion. For others, IBM begins with the finger flexors, causing weakness in wrists and fingers, difficulty pinching, buttoning, making a fist, and gripping objects. Weakness in the throat often causes dysphagia (~65%). The diaphragm weakens in about 20% of cases – it does not affect the heart. Muscle pain and excess, thick saliva, and mucus are common.



Diagnosis: Usually diagnosed by a neurologist/rheumatologist, clinical exam, EMG, biopsy, MRI, and serology. Average time from symptom onset to correct diagnosis is five years. Often initially misdiagnosed as ALS or polymyositis. IBM weakness occurs over months or years and slowly, steadily progresses (polymyositis – much faster onset, responds to steroids). Other muscular dystrophies (e.g., limb-girdle, Becker) and ALS must be ruled out. Many doctors are not familiar with its presentation.

Progression: 5 years from symptom onset, many patients require a cane, walker, or rollator; within ten years, a power wheelchair and house modifications (bathroom/bedroom, etc.).

Treatment: The standard of care is strictly nonpharmacological management, including emotional support, physical therapy, and fall prevention. In the early stages, an individualized exercise program is suggested.

Complications: 65% develop progressive, often severe dysphagia (half of them eventually require a feeding tube). Diaphragmatic weakness may present as sleep apnea – hypoventilation causes hypercapnia. IBM-related causes of death are usually respiratory complications from aspiration pneumonia or diaphragmatic involvement.

Comorbidities: Include higher rates of hypertension (66%), hyperlipidemia (48%), diabetes mellitus (34%), peripheral neuropathy (36%), anemia (31.6%), Coexisting autoimmune disorders are frequent (~50%) – most common, rheumatoid arthritis and Sjogren's syndrome.

Management: Evaluation and ongoing monitoring of potential complications (dysphagia, respiratory involvement, depression, fatigue, etc.). Social and mental health support. Isolation/depression are risks if support is poor. Integration of palliative care is helpful at the end stages.

More information: Please see http://www.ibmmyositis.com/ https://doi.org/10.1007/s40674-020-00169-4

https://doi.org/10.3390/ijms25052742 https://doi.org/10.1097/BOR.0000000000000958