# Inclusion-Body Myositis

## *What is Inclusion-Body Myositis (IBM)?*

- Inclusion-body myositis (IBM) is a disease that causes inflammation of the muscles and unlike other myopathies, IBM also causes muscle degeneration creating a mass of degenerated cells to form within the muscle tissue.
- IBM symptoms include progressive weakness of wrist and finger muscles, the muscles of the front of the thigh, and the muscles that lift the front of the foot.*
- The pathophysiology of IBM involves a T cell mediated autoimmune response potentially along with abnormal proteins leading to an inflammatory and degenerative response.
- Women are about twice as likely as men to develop IBM.

**Key facts about IBM:**

- There are an estimated 22,000 cases of IBM in the US.**
- IBM is the most common acquired myopathy in patients older than 50.
- Patients with IBM will often require a cane or be bound to a wheelchair 5-10 years after initial diagnosis.***

## *Patient Impact*

- Muscle degeneration in the legs leads to difficulty walking and patients frequently falling.
- Wrist and hand muscles degenerate leading to patients being significantly disabled as the disease progresses.
- Muscles around the throat and neck can become weak and often lead to dysphagia and increased potential for aspiration pneumonia.
- The most common cause of death in patients with IBM is from respiratory disorders.

## *Treatment Options*

- Currently, there is no cure or any therapies indicated specifically for IBM.
- Traditional therapies for autoimmune diseases and other inflammatory myopathies such as steroids, methotrexate, and cyclophosphamide are generally ineffective in IBM patients.
- Some novel therapies, including biologics, have been tested in the disease, but have not proven effective.

Inclusion-body myositis patients lack any cures or therapies for their disease.

### *About KPI Therapeutics™:*

KPI Therapeutics is a clinical stage biotechnology company, which develops first in class therapies for unmet medical needs in autoimmunity using its novel Kv1.3 channel blocker based platform. Its lead drug, dalazatide is being clinically advanced for Inclusion Body Myositis, (IBM) an orphan disease, and for lupus. Our autoimmune platform molecules are also being developed for new therapies to address atopic dermatitis and uveitis.

Our autoimmune program is developing a systemic biologic Kv1.3 inhibitor targeting the immune cells that cause Inclusion-Body Myositis.

www.kpitherapeutics.com | 206-788-4600

*https://www.mda.org/disease/inclusion-body-myositis
**http://emedicine.medscape.com/article/1172746-overview
***https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3535450/