

# Inclusion Body Myositis (IBM)

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### A). Five Takeaways

- 1). IBM is a poorly understood and very complicated disease that attacks muscle. It has no known cause, is chronic and progressive. It appears that IBM may be a type of autoimmune disorder.
- 2). No effective treatment has been developed.
- 3). Potential weakness in the throat should be monitored as it may lead to weakness swallowing and choking.
- 4). Potential weakness in the diaphragm should be monitored as it may lead to respiratory problems.
- 5). As leg muscles weaken, a major risk becomes injuries from falling.

### B). Key Facts

#### IBM overview:

This disease is usually called sporadic inclusion body myositis (IBM). It is called sporadic because it develops unexpectedly. You will usually be the only one in your family who has this disease. It is called inclusion body because under the microscope the muscle cells display what they call inclusion bodies. It is called a myositis because it appears as an inflammation of the muscle. It is sometimes called an idiopathic disease – this means its cause is unknown.

IBM is a type of muscular dystrophy (there are also many other types of muscular dystrophy that are quite different). IBM attacks the long-striated muscles in the body, usually affecting the arms and legs first. It can also affect the diaphragm (impacting breathing) but does not appear to affect the heart. Nerves are not affected, and, although you may lose strength in your limbs, you should not lose sensation. IBM leads to progressive disability—as it gradually progresses, more and more muscles are impacted, and the patient’s level of overall disability increases—over a period of years, usually ending in severe (total) disability.

In the past, it has been reported that IBM has no effect on one's lifespan. However, one study (see Barghout, et al, 2014 [below]) identified a shorter lifespan in patients with IBM compared with the general population. There may be several reasons for this, including the poor management of respiratory involvement, complications arising from falls, and pneumonia related to choking on food.

People often confuse muscle disorders (muscular dystrophy) with multiple sclerosis, which is a neuron/nerve disease.

IBM is often initially confused with another type of muscle disease called polymyositis. Polymyositis has similar symptoms and is more common in the population, so doctors often assume that you have polymyositis. Patients are often treated with medications and do not respond as doctors would expect if it was polymyositis. Further investigation reveals that the patient actually has IBM.

A substantial percentage of IBM cases are initially incorrectly diagnosed with one or another disease. On average, it takes about five years to diagnose IBM.

IBM is very rare in the overall population; about 46 people per million. But, it is a disease related to aging: As people get older, IBM becomes much more common, and, in people over 50, the estimates of prevalence are about 139 per million (Tan et al, 2013, p. 334). With the approaching age bubble in our population, the burden of IBM as a disease on society will likely increase. Although the disease is most common after 45-50, about 20% of cases present symptoms before 50. Average age of onset is about 61-68.

Slightly more males are affected than females. The common form is the sporadic form, and it is not considered inherited—that is, it is not passed on from parents to children.

IBM symptoms come on extremely slowly, over months or years (whereas the symptoms of polymyositis generally come on over weeks or months). IBM is a progressive disease; it does not appear to go into remission periods. It just slowly gets worse and worse as muscles become weaker and weaker.

The rate and progression of the disease varies widely from person to person. As well, the initial symptom presentation (the exact pattern of muscles affected) can also vary widely from person to person. Most cases affect one side of the body more than the other (usually the non-dominant side).

#### Causes:

There is no recognized cause for IBM.

#### What IBM does:

IBM causes problems within the cells that make up the muscles. (It does not affect nerves). Somehow, in each muscle cell, the internal chemistry is disrupted, and the

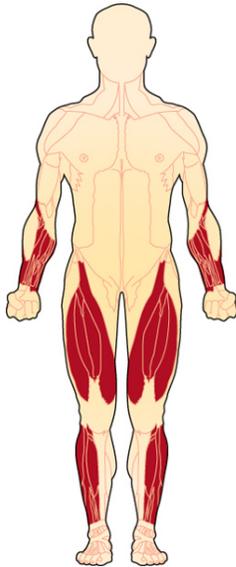
muscle cell dies. The problem continues, and over time, fewer and fewer muscle cells are left functioning. Eventually the muscle becomes very weak and ultimately turns into a brittle fiber.

T cells, myeloid dendritic cells, macrophages and plasma cells all invade IBM muscle, but it is the infiltration of muscle by T cells that is the most obvious microscopic feature of IBM muscle. This implies that it is an autoimmune based disease.

#### The effect of IBM on the body:

IBM does not affect all muscles at the same time. You may initially find some muscles are affected, some muscles won't change, and other muscles will try to compensate and get stronger. For example, as muscles in the front of the leg are weakened, enlargement of the calf muscles may be seen.

As you can see from the diagram below, the large quadriceps muscles in the front of the leg are often affected, causing weakness felt in climbing stairs, in getting up from chairs, etc. It often leads to frequent falls. "Toe drop" (the leg muscles don't raise the toe high enough when taking a step) commonly causes tripping. As well, weakness of the muscles in the arm is a common, early symptom. This causes loss of wrist strength, finger dexterity and weak grip strength (for example, weakness making a fist, opening a car door, or pushing down on the top of a spray deodorant bottle).



#### Diagnosis of IBM:

The diagnosis of muscle diseases is complicated. The diagnosis of IBM is even more complicated because it is rare and because the symptoms vary in each case. Generally speaking, as mentioned above, it is not unusual to take up to five years to achieve a diagnosis.

Usually diagnosis is made by a specialist in muscles—a neurologist—or sometimes by a rheumatologist. It is not uncommon to need to have a second opinion in

the diagnosis. As well, it is not uncommon to have a biopsy result that is reported as ambiguous. Patients have to be diligent and patient during the diagnosis process. There are various indicators used in making a diagnosis. The main ones appear below. As research goes on, new tests should become available that are more specific to IBM.

1). Clinical signs: what the doctor can see when you are examined. The doctor looks for a characteristic pattern of muscle weakness. IBM has unique physical examination features (such as finger flexor and knee extensor weakness) that sets it apart from most other muscle diseases.

2). A blood test is done for creatine kinase (CK), an enzyme in the blood showing muscle damage. CK is at most about 10X normal, although this may vary widely.

3). Anti-cN1A antibodies are present in about 50 % of patients with sIBM, thus, if you have these identified, it is a significant red flag for IBM. Unfortunately, anti-cN1A antibodies do not appear to correlate with any studied clinical or laboratory parameter and, therefore, were of limited value in the patient's assessment (Paul et al., 2021). However, the presence of anti-CN1A antibodies identified patients with a greater risk of more severe dysphagia (Lucchini et al., 2021).

3). Electromyography (EMG): Electrical studies of the muscle done with a computer (an electromyograph) will display abnormalities in IBM.

4). Muscle Biopsy: Surgical removal of a piece of muscle and its study in the laboratory. Most cases usually require at least one muscle biopsy for diagnosis.

5). MRI shows abnormalities, which were more severe within the lower limbs and the distal segments. The most prevalent MRI finding was fat infiltration. (Guimaraes et al., 2017).

### Direct treatment of IBM:

Unfortunately, there is no direct treatment at this time. The cause of IBM is unknown, and the exact way that IBM affects the muscle cells is not well understood. Until this is better understood, it will be difficult to develop a treatment directly aimed at IBM. See: Snedden et al., 2021.

In the past, a number of different medications have been tried, primarily prednisone and IVIG. No medication has been reliably shown to be effective. All of the medications used have serious and significant side effects. About half of IBM patients were traditionally given some sort of immunosuppressive treatment. It unfortunately appears that the group receiving treatment may sometimes develop greater weakness than those who did not receive treatment (Benveniste et al, 2011).

The main focus of treatment is on the prevention of complications and on maximizing quality of life as long as possible. This is often done in consultation with a physical and/or occupational therapist. When necessary, speech therapy may be part of the treatment as well (swallowing issues are monitored). Consultation with specialists in respiration is recommended to detect and monitor respiratory weakness. The planning of appropriate palliative care during the end stages of IBM is strongly recommended.

### Indirect treatments:

Until a specific treatment can be developed for IBM, one strategy is to try to develop treatments that will counterbalance the impact of the disease, for example, to try to increase the amount of muscle with the idea that if there is an increase in overall mass, there may be gains in function. These treatments are not directly focused on IBM and would not alter the IBM disease process itself.

Several treatments along these lines are in experimental stages. Bimagrumab (BYM338) (a myostatin inhibitor) achieved preliminary positive results but has initially failed in advanced studies. Another experimental approach uses gene therapy (using follistatin) to inhibit myostatin.

### Other forms of “treatments:”

Many different types of treatments are often promoted for diseases like IBM (and cancer). Unfortunately, companies often prey upon people with such illnesses and make emotional and exaggerated claims for various “cures.” None of these different diets or potions or supplements have been shown to have any effect whatsoever on IBM. I often hear patients say, “there must be something I can do,” or, “I just can’t sit here, I have to do something.” Being an IBM patient myself, I understand these feelings, but I must emphasize that when no treatment is recognized to improve symptoms, we are risking side effects unnecessarily, and we are simply throwing our money away in false hope.

## C). Coping

### Adaptations:

Unfortunately, IBM is a progressive disease and presents somewhat of a moving target as weakness slowly develops. An adaptation that works today may not work in 6 months or in a year from now. This is important in planning ahead. For example, house renovations should not be based on your condition today but rather should look at where you will likely end up in the future.

### Falls:

Because the muscles in the legs are affected early in the disease, falls are one of the early symptoms. People often experience what is called “foot drop” this means that when you take a step your toes do not fully lift up, and you tend to drag your toes and trip, falling forward on your face.

I will describe two other characteristic types of falls. The first “the slow-motion tree falling in the forest.” You end up losing your balance and slowly tip over and fall. You can feel yourself falling, often in a feeling of almost slow-motion, but you cannot do anything about it. This kind of fall is dangerous because you can hit your head.

The second kind of fall I will call, “straight down in a crash.” In this fall, usually you are taking a step forward and your knee gives out, and you literally fall straight down. These falls tend to be instantaneous, and you are on the ground before you even realize you are falling. These falls can be dangerous because all of your weight is going straight down on your ankles and knees.

One thing for certain is that you cannot “catch yourself” by grabbing onto something when you fall. Prevention is everything. Often, the use of a cane can be helpful in keeping one’s balance. In my case, I progressed from a cane to a walker. Then I eventually ended up falling onto my walker and had to get a wheelchair. The advantage of a wheelchair is that it is far safer to get around when compared to risking a fall, and it also preserves your energy compared to using a walker.

### Swelling of the ankles and feet:

Edema (that’s what the swelling is called) is a common problem that can best be addressed by using compression socks on a daily basis. Prevention is the key. Usually swelling of the ankles and feet is caused by inactivity, especially in people who are in wheelchairs. The only downside of using compression stockings is that they are difficult to put on. You will usually need help putting them on. You can get these in different compressions (strengths), and it’s best to talk to your doctor about what is recommended for you.

Edema can become a serious problem if it becomes excessive; it can cut off the circulation, and the skin in ankles and feet may turn a purple color. If your skin becomes discolored, you need to see a physician.

### Exercise:

This has been a controversial topic in IBM. In the past, exercise was not generally recommended. However, as more research accumulates I believe it is accurate to say that exercise is viewed more positively and may play a role in slowing down loss of function. Some research suggests that individuals with IBM may respond differently to exercise: “A few patients seem to respond very well with larger improvements in function and tolerating a very high exercise frequency and intensity, while yet a smaller group of patients might not tolerate even mild exercise. There is an urgent need for more, larger studies looking into exercise in inclusion body myositis” (Alexanderson, 2018, p. 295). Here is a study that found different responses in different subgroups. “In clinical practice, it is evident that not all patients with IBM follow a similar trajectory of disease progression; our study attempted to define whether there is a continuous spectrum of severity or whether patients congregate into discrete subgroups. The variation of progression among trajectory subgroups was marked” (p. 80). . . . “Our identification of IBM subgroups, according to longitudinal strength and functional status change, may allow for more focused eligibility criteria for clinical trials reducing baseline heterogeneity among treatment and placebo groups . . . We have identified clinical subgroups within the IBM cohort, according to strength and functional status change over time. These findings may lead to enhanced clinical prognostication and potential stratification of trial participants” (Oldroyd et al., 2020, p. 81).

There is no clear consensus in the recent literature: “Despite the promising results reported in case studies and even clinical experience, the evidence sustaining physical training in sIBM is still fragile. . . . Currently, there is limited evidence to funding the safety of physical exercise in sIBM. Based on the conflicting and low-quality evidence,

physical exercise might slightly improve muscle strength or, at least, reduce the speed of decline” (de Souza et al., 2020, p. 5).

Here is an example of a recent positive endorsement: “Physical therapy plays a significant role in the treatment of IBM, since it leads to improvement of the functional capacity of patients in daily activities, thus reducing their disability” (Stevanović et al., 2020, p. 1220).

Because there is a significant risk of overdoing exercise, I believe the best advice is to carefully discuss this with your medical team to determine what exercises are recommended in your particular case.

#### Diet:

Because IBM impairs normal movement, and because you cannot exercise as a normal person would, the single most important thing is to eat well and to eat less. When you are inactive and you eat a normal diet, you will gain weight. Naturally, the more you weigh, the more problems you will have in general, because you don't have the muscle structure to support normal body weight. So, the best advice is to try to cutback the amount you eat.

#### Attitude:

This is a serious and chronic disease. You will have IBM for the rest of your life. Attitude becomes a critical factor in being able to cope and not become bitter, cynical or depressed. Don't give up—find ways to adapt. Our lives will change—for example, I had to give up my job, but now I have different things in my life to keep me busy. Attitude is critically important when you have a long-term disease that has no treatment.

#### Using a cane or walker:

A cane can help you with balance in the initial stages of the illness. Likewise, the use of a walker may be beneficial at some point. As your disease progresses, you will need to continue to adapt and change your strategy. For example, you may find yourself falling down on top of your walker (as I did)—this shows it is time to move to a wheelchair. It makes no sense to have “false pride” and be in the hospital with a broken hip. One of our IBM friends fell and broke his hip. He never came out of hospital.

#### Wheelchair:

Understandably, there are a number of drawbacks, problems and stigma associated with being in a wheelchair. However, a wheelchair is far safer than risking falls and broken bones. I have met many people who realize they *should* be in a wheelchair. However, they stubbornly refuse to consider the possibility. In my opinion, if you are using a cane or a walker and are still falling, you should seriously consider that your safety is more important than pride. Recovery from a broken bone or concussion presents a serious prospect for the patient with IBM.

Note: Many IBM patients will never require a wheelchair. Many factors enter into this, including: when symptom onset occurred; how rapid the progress is; and the

patient's overall health. Many patients will pass away from other causes before reaching the stage where they need a wheelchair due to IBM. Eventually, many people do end up with what they call total disability and, in general, the mean/median time to wheelchair use is considered about 10 to 15 years after symptom onset.

### Swallowing:

Many IBM patients develop weakened swallowing (called dysphagia [DIS-phage-EE-ah]). This is a major complication leading to potential choking. As soon as a person is diagnosed with IBM, he or she should be assessed for swallowing weakness, and this should be monitored as the disease progresses.

Sometimes you try to swallow, and nothing will happen—food “will not go down,” or food will get stuck “halfway down.” This can lead to choking that can be life threatening. As well, if this happens, food can go into the lungs (called aspiration), resulting in a type of pneumonia that is often fatal in older people.

Some people with IBM eventually have to have tubes inserted into their stomachs because they can no longer swallow. Sadly, some people simply stop eating and die of malnutrition.

Pay close attention when you are eating. Do not be in a hurry. Do not have conversations while eating. Eat smaller portions and chew the food well. If you notice problems swallowing, you should have this evaluated and keep an eye on any potential progress.

In general, you should always have a noncarbonated liquid available when eating. I find that drinking with a straw is the most practical. If you are with others, and you begin to have problems you should try to draw their attention to your situation. Try to take a small sip of liquid to dislodge the blockage. I have discovered the most important thing in the situation is to try to remain calm. If you choke frequently, you should not eat alone, and those who are with you should be attentive and know what to do, for example, how to apply the Heimlich manoeuvre.

### Breathing:

In many cases, IBM weakens the diaphragm muscle. This can lead to reduced air volumes, especially during sleep—this means that you are not breathing in and out enough air to adequately remove carbon dioxide from the body. Over time, carbon dioxide can increase in the blood slowly poisoning you (most people misunderstand this as involving oxygen—generally there is enough air volume to supply sufficient oxygen; therefore, usually no supplemental oxygen is needed). The usual treatment is to use a BiPAP machine at night to give the necessary amount of airflow in order to exhale enough carbon dioxide. As well, a patient may require respiratory support during the daytime. For example, I have a machine on the back of my wheelchair that supplies me with extra room air (a mouthpiece respirator). Respiratory failure through weakness of the respiratory muscles is a recognized cause of death in IBM. As soon as a person is

diagnosed with IBM, he or she should be assessed for diaphragmatic weakness, and this should be monitored as the disease progresses.

#### Pain:

Generally speaking, when you read about IBM, it is not associated with pain. However, people with IBM often report having pain. Pain can result for several reasons. If a muscle is pressed or stretched, it can cause pain. Likewise, if you are in a position for a long time, the body can become stiff, and this can lead to pain in the muscles.

#### Mucus and phlegm:

Many patients with IBM report having excess mucus and phlegm in the throat and chest. This symptom is not normally reported in the research literature. The cause of these excess and thick secretions and its association with IBM is a mystery.

#### Skin:

People sitting in one position for a long time, whether in a wheelchair or in an ordinary chair, are often prone to developing soreness and breakdown of the skin on the buttocks. In IBM, the nerves are not affected, and you should have normal pain receptors. If you are sitting and begin to hurt, then you need to move, even if someone has to move you to a different position. Pain is a sign that pressure is becoming excessive. Pressure sores in the skin can be painful and can become a major problem if they become infected. As well, pressure sores take a long time to heal.

#### Beds:

One of the major challenges of IBM is that you generally have trouble turning over in bed. If your bed is too soft, you will end up “in a hole” unable to move. If your bed is too hard, you will have problems with pressure, usually felt on the hips. This can be painful. Generally speaking, a lot of experimentation is necessary to find the proper mattresses for your needs.

Most doctors recommend elevation of the head. This is helpful to prevent fluid buildup in the lungs. A hospital bed that is adjustable is ideal.

As the disease progresses, falling while getting in or out of bed is a major threat. Adaptive devices may be necessary, for example, the use of a ceiling lift for safe transfers.

#### Bathroom:

The bathroom presents many obstacles for the IBM patient. Early on, it may become difficult to get out of a bathtub. As well, rising from a toilet can be challenging as the symptoms progress. Adaptive devices may be necessary, for example, the use of a shower chair (both for showering and rolling over a toilet).

#### D). Other

#### Research on IBM:

IBM is a very complex and challenging disease to research. In approximately the last 45 years of research, much has been learned about the disease, but frustratingly, more remains unknown. The understanding of IBM and its description is an evolving phenomenon as more is learned over time.

For reviews of the research you can see the yearly breakdowns on my webpage (<http://www.ibmmyositis.com>).

### Polymyositis (PM) and IBM:

Patients with IBM are often initially diagnosed as having polymyositis (PM). IBM is quite rare and relatively unknown, whereas PM is more common and better known. PM has a different onset—it comes on over weeks, whereas IBM has a slow onset—over months or years. PM usually responds quickly to medication whereas IBM usually shows no response to medication. If you have been diagnosed with PM and given medication, and you have not responded, then clearly your diagnosis should be reviewed.

### Genetics and IBM:

IBM is not considered an inherited disorder: It is not passed on to the children of people with IBM. However, it appears that a predisposition to developing IBM may be linked to a group of genes commonly seen in Caucasians from Northern European ancestry. People with these genes appear to be predisposed to develop autoimmune disorders. IBM is associated with a small group of such genes (HLA-DR3) in many patients (see Britson, Yang, & Lloyd, 2018). If you happen to have this particular set of genes, you may be somehow predisposed to develop IBM (or one of several other autoimmune disorders).

It is also possible that some combination of genes may interact with each other and with environmental variables to increase the likelihood of IBM developing in a given individual.

### Familial Inclusion Body Myositis (fIBM):

This is a rare sub-type of IBM. fIBM occurs in two or more siblings in a family in the same generation (but it is not passed on to their kids). The symptoms and features of fIBM are very similar to those seen in the sporadic form of IBM. fIBM is also linked to the same genes as IBM, raising the possibility that fIBM and IBM share similar inherited predispositions. The familial occurrence of such a rare disorder likely highlights the importance of genetic predisposition in the causation of IBM (Needham, Mastaglia, & Garlepp, 2007).

### Hereditary Inclusion Body Myopathy (hIBM):

Hereditary inclusion body myopathies (HIBM) are a group of rare genetic disorders which have different symptoms. Generally, they are neuromuscular disorders characterized by muscle weakness developing in young adults. Hereditary inclusion body myopathies comprise both autosomal recessive and autosomal dominant muscle disorders that have a variable expression (phenotype) in individuals, but all share similar structural

features in the muscles. notice that this group of illnesses are referred to as myopathy because they do not show the inflammation seen in myositis. In other words, inclusion body myopathy is not the same disease as inclusion body myositis: the latter showing inflammation as a primary symptom. These various genetic disorders are exceedingly rare. For more information, see: Wikipedia contributors. (2021, February 16).

## E). Information checklist

- ✓ IBM has been known since the late 1960s, but it is quite rare. It is not unusual for physicians to be unfamiliar with the disease.
- ✓ Muscle diseases are complex, and many have similar symptoms. Diagnosis is always a long and sometimes frustrating process. Misdiagnosis is common. The best advice on diagnosis comes from your physician, not from the Internet. My best advice is to be patient and become a strong advocate for yourself throughout the process. As I've said before, this is a serious disease that you will have for the rest of your life, and my philosophy is that the more you know about it, and the more professional you can be in your approach as an IBM patient, the better.
- ✓ IBM has variations in symptom presentation, making it hard to compare two different patients. This also makes it hard to predict the rate of progression.
- ✓ IBM has no recognized medical treatment today. All treatments that have been tried have failed, and all have significant side effects that outweigh any short-term or marginal benefits. Generally, no supplements nor other diets have been shown to make a difference.
- ✓ At least four complications need to be considered and managed: unexpected falls; weakness swallowing; weakness in the diaphragm; and lower leg edema.
- ✓ Very careful management of the disease and its complications will lead to the highest quality of life and maximize one's lifespan.

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## G). Disclaimer

I am not a medical Doctor and this information is not intended to be read as medical advice nor is it a substitute for medical advice. Please consult your Physician

when you have medical concerns. I have done my best to offer a layman's interpretation of this material based upon my experience with the illness and my reading of the literature. I have tried to avoid references here although I have included a few where critical. Any opinions offered are personal and do not reflect those of my employer. Thank you.

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