

# Myotonic Dystrophy Type 2

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## What is myotonic dystrophy type 2?

Myotonic dystrophy is a medical term for a condition that affects muscle function. The term myotonic comes from several words in Greek:

- ‘myo’ means muscles (muscles are affected)
- ‘tonia’ meaning tension (the tension of the muscle is affected)

Myotonic dystrophy type 2 (DM2) is one of several types of muscular dystrophy.

## What are the signs and symptoms of DM2?

### **Muscle weakness**

Each person responds differently to the disorder. Symptoms often do not appear until after 20 years of age. Individuals with this condition typically have a normal lifespan.

- DM2 commonly affects the muscles of the neck, shoulders, elbows and hips.
  - People with DM2 often have prolonged myotonia (sustained muscle contractions), and are not able to relax certain muscles after use. For example, difficulty releasing grip on a doorknob, or temporary locking of their jaw.
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**Other common signs or symptoms include:**

- heart irregularities (known as “heart block”)
- type II diabetes
- cataracts

**What causes DM2?**

DM2 is a genetic condition which is typically inherited from one parent. The abnormal gene can be inherited from a mother or a father. Having the abnormal gene means that you have the condition. This is called “autosomal dominant inheritance”.

**How do you know I have DM2?**

Your doctor or specialist can tell if you have DM2 from:

- talking with you about your symptoms and family history
- checking the strength of your muscles
- reviewing the results of the following tests

**Genetic tests**

- Individuals with DM2 have an expansion (extra information) in the non-coding region of the gene *CNBP*. The extra material is four genetic letters, CCTG, which are repeated over and over. This can be measured in your blood (genetic test).

**Electromyography (EMG)**

- This test measures the electrical activity in your muscles and nerves. A acupuncture needle is inserted into a muscle and listens for a distinct sound known as “myotonic discharges” which occur in the muscles of an individual with myotonic dystrophy.
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## How is DM2 treated?

DM2 is usually managed with a combination of methods. Your treatment/management plan will be designed to meet your needs.

**The goal of treatment is to prevent or lessen your symptoms and help you retain the strength in your muscles.**

### Heart

- Monitoring your heart on a yearly or bi-yearly basis is important to check for a heart irregularity known as “heart block”.
- An ECG is done to check for changes in the rhythm of the heart and other potential cardiac symptoms.

### Eyes

- Checking eyes for cataracts on a yearly or bi-yearly basis.

### Mobility

- It is important to have neurological and strength assessments regularly to monitor any progression of your DM2.
- Assistive devices may be considered such as, ankle-foot orthoses or other assistive devices.

### Exercise

- Exercise is always helpful to help build and maintain your muscle strength! We can provide strength monitoring and exercise advice with a DVD and exercise handouts.
  - We recommend slowly increasing exercise tolerance starting off slowly and gradually to eventually attain 3 to 4 sessions each week of mixed endurance (such as biking, rowing, walk, jog, elliptical) and strength (such as weights, elastic bands, pilates) exercise. However, the most important thing is to do a type of exercise you enjoy.
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## How will having DM2 affect my life?

Each person responds differently to DM2. Outcome depends on the severity of your condition.

You will visit the Neuromuscular and Neurometabolic Centre, where the health care team will plan your treatment and provide your care.

Living with a chronic condition can be challenging and emotional. Here are some ideas for living well with a chronic disease.

### Educate yourself

- Learn as much as you can about the disease and how to manage symptoms. If you have questions, ask your health care providers. This will help you make informed decisions about your care.

### Take part in your care

- Work closely with your health care providers, as a team. Follow your treatment plan. Keep track of your symptoms and your response to treatment. Tell your health care providers how you are doing.

### Take care of yourself

- Learn to listen to your body. Rest and conserve your energy when you feel tired. When you are active, go at your own pace.

## Notes and questions

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