

### Useful Reading and Web Links

1. Harper, PS, *Myotonic Dystrophy the facts* 2002: Oxford University Press
2. International Myotonic Dystrophy Organization [www.myotonicdystrophy.org](http://www.myotonicdystrophy.org)
3. Muscular Dystrophy Association [www.mdausa.org](http://www.mdausa.org)
4. Washington University in St. Louis <http://neuromuscular.wustl.edu/>
5. Athena Diagnostics [www.athenadiagnostics.com](http://www.athenadiagnostics.com)
6. National Institute of Neurological Disorders and Stroke <http://www.ninds.nih.gov/>

## For more information

Please visit our website  
[www.myotonicdystrophy.org](http://www.myotonicdystrophy.org)

Testing and carrying an alert card is important for everyone diagnosed with Myotonic Dystrophy. You can request a free medical alert card by emailing your name and address to [info@myotonicdystrophy.org](mailto:info@myotonicdystrophy.org).

**International Myotonic Dystrophy Organization (IMDO)** promotes and advances the welfare, care, treatment, and research of persons with Myotonic Muscular Dystrophy or Congenital Myotonic Dystrophy. It educates the persons with Myotonic Dystrophy or Congenital Myotonic Dystrophy on their disease, as well as educates the general public, and expedites the discovery of a cure for Myotonic Muscular Dystrophy and Congenital Myotonic Dystrophy by funding projects that will assist IMDO in meeting its goals.



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**International  
Myotonic Dystrophy  
Organization, Inc.**

**Heart Problems  
in  
Myotonic Dystrophy**

[www.myotonicdystrophy.org](http://www.myotonicdystrophy.org)

*Leading the way to management and treatment*

## What is Myotonic Dystrophy or DM?

Myotonic Dystrophy, also known as Steinert's Disease or Dystrophia Myotonica (DM), is the most common inherited muscle disease, affecting about one in every 8,000 people. It is the most common form of Muscular Dystrophy in adults.

In general, DM consists of muscle weakness and myotonia (slow relaxation of muscles after contraction), which gets more severe over time. Specific problems in other systems of the body can also occur. Since DM can affect many tissues and organs, it is called a "multi-systemic" disorder.

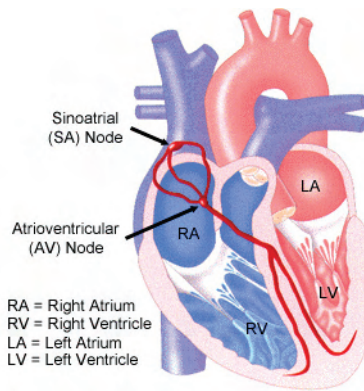
Myotonic Dystrophy is an extremely variable condition. In fact, it is one of the most variable disorders known, which can lead to difficulties in getting an accurate diagnosis.

DM can vary in severity in the systems of the body it affects and in the age of onset. DM has equally severe effects on most other organs of the body and the symptoms include: cataracts, heart defects, breathing difficulties, mental impairment, testicular atrophy, diabetes and premature balding.

Myotonic Muscular Dystrophy is known as DM1 and Congenital Myotonic Dystrophy is classified as DM1. The second variety of Myotonic Muscular Dystrophy is known as DM2 and also known as Proximal Myotonic Muscular Dystrophy or PROMM.

The discovery of the gene alteration, which causes DM, helps explain this conditions' incredible variation. The discovery was commercialized by Athena Diagnostics in 1992.

## Myotonic Dystrophy and the Heart



Myotonic Dystrophy is a multi-systemic disorder (affecting many tissues and organs) DM1 and DM2 affects the heart muscles.

However, there are also many cases where heart muscle and vessels are not affected. For example, coronary

heart disease (the most common cause of death in many populations) is not increased, nor is high blood pressure or stroke. In fact, blood pressure is often low in myotonic dystrophy.

Although coronary heart disease is not increased in DM patients, there is a greater than average occurrence of disturbed conduction issues among DM1 and DM2.

The cardiac conduction defects are usually characterized by palpitations, fainting or near fainting spells can occur and should never be ignored. These can be fatal. These arrhythmias can occur in an otherwise healthy heart.

An arrhythmia is a disturbed conduction of the heartbeat. Cardiac arrhythmias and sudden death are a major cause of mortality in Myotonic Dystrophy patients, even in young patients with limited muscle problems. Acute arrhythmic disorders should be treated by a cardiac specialist who has experience with DM patients.

Patients must always let their attending physician know that they have Myotonic Dystrophy. In many cases the physician may not be familiar with DM so the patient should be prepared to present materials to the physician.

## Cardiac Management of Myotonic Dystrophy

It is generally agreed within the medical community that careful monitoring of cardiac issues of Myotonic Dystrophy patients is necessary, but there remains debate as to the method and frequency.

The treatment required will depend on the type of rhythm disturbance and is best determined by a heart specialist. If drugs do not restore or maintain regular rhythm, electrical treatment may be needed for this, while for slow conduction ('heart block') an artificial pacemaker or defibrillator may be needed. There is a good deal of debate as to when an artificial pacemaker may be needed.

In asymptomatic patients, recommended screening for cardiac follow-up is one ECG each year, Holter monitor every 2 years, and echocardiography every 2 years, as long as there are no clinical cardiac symptoms, and the ECG remains normal.

Extra caution should be used when undergoing procedures requiring anesthesia. Continuous ECG monitoring is recommended. It is critical that physicians be informed of the increased anesthesia risks associated with DM patients. Some informal information indicates that surgery or anesthesia may trigger a cardiac event.

**For a complete list of practical recommendations for surgery and anesthesia in DM patients please visit our website at [www.myotonicdystrophy.org](http://www.myotonicdystrophy.org).**