



Heart-Healthy: How to Control Cardiac Risks in Neuromuscular Disease

Cardiac disease is a common and potentially serious component of many neuromuscular diseases. For neuromuscular disease providers, understanding how the heart can be affected, when those complications may occur, and how to intervene early are critical considerations for comprehensive care.

Cardiac risks in neuromuscular disease

In neuromuscular conditions, two heart issues most commonly occur: cardiomyopathy and arrhythmia.

Cardiomyopathy

“The heart is made of muscle called myocardium,” says Gavin Hickey, MD, advanced heart failure and transplant cardiologist at the University of Pittsburgh Medical Center. “For certain neuromuscular diseases, such as Duchenne muscular dystrophy (DMD), the heart can become weak, just like other skeletal muscles in the body. We diagnose this as cardiomyopathy.”

The degree of weakness depends on multiple factors, but it is not always directly correlated with the weakness of other muscles.

To measure cardiac weakness, physicians use tests such as echocardiograms and cardiac MRIs. “Both tests report left ventricular ejection fraction (LVEF), which is a measure of how well the left side of the heart is pumping,” Dr. Hickey says.

He provides the following guide:

- Normal LVEF is >50%–70%.
- Treatment is typically needed when LVEF is <40%.

In some neuromuscular diseases, such as DMD or Becker muscular dystrophy (BMD), treatment to prevent cardiomyopathy may be initiated even at normal ejection fraction values.

Arrhythmia

The second prominent cardiac risk is arrhythmia, or irregular heartbeat. “Patients who develop cardiomyopathy, scarring, or fibrosis in the heart, or who have certain types of neuromuscular disease such as myotonic dystrophy (DM), are at risk for either atrial or ventricular arrhythmias, which can become common later in life,” Dr. Hickey says.

There are two types of arrhythmias:

- Atrial arrhythmias are irregular heartbeats that can cause the heart to beat too fast or too slow, causing poor circulation. These arrhythmias occur when the upper chambers of the heart beat out of rhythm with the lower chambers.
- Ventricular arrhythmias cause the heart to beat too fast, too slow, or erratically, and can cause sudden cardiac death. They originate in the heart’s lower chambers.

Other risk factors

In addition to any heart risks associated with their disease, people with neuromuscular diseases face the typical cardiovascular risks related to age.

“As more patients with neuromuscular disorders enter adulthood and live into their fifth decade or longer, other risk factors for cardiovascular disease can lead to coronary artery disease, heart attacks, and strokes,” he says. “Therefore, maintaining good overall preventive cardiovascular care is important.”

These risk factors include:

- Hypertension
- Hyperlipidemia
- Diabetes
- Smoking
- Weight gain

Treating and managing cardiac disease

Depending on the specific neuromuscular disease and cardiac testing results, treatment protocols and management strategies vary.

For treating cardiomyopathy in a patient with an LVEF less than 40%, Dr. Hickey recommends following the four pillars of “[Guideline-Directed Medical Therapy in Heart Failure](#).”



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The pillars consist of four medication classes:

- Beta-blockers
- Angiotensin receptor blockers
- Sodium-glucose transport 2 inhibitors (SGLT2i)
- Mineralocorticoid receptor antagonists (MRA)

“These medications have been well validated,” Dr. Hickey says. “For specific populations, these four medications in combination can reduce mortality by as much as 76%.”

Dr. Hickey’s clinical approach is early intervention using the four pillars to prevent cardiomyopathy, slow its progression, and stop remodeling that can lead to scarring, fibrosis, or other changes that worsen LVEF or lead to arrhythmias.

For atrial arrhythmias, physicians use medications to help the heartbeat stay in rhythm and/or medications such as blood thinners to prevent stroke.

Treating ventricular arrhythmias often requires defibrillator placement if it aligns with the patient’s goals.

More advanced testing and imaging in the last decade have improved treatment protocols.

“We continue to learn more about how new medications and therapies affect long-term cardiac involvement of neuromuscular disease. Improved imaging techniques, such as cardiac MRI, allow us to see changes in LVEF over time as well as scarring or fibrosis in the heart,” Dr. Hickey says.

He adds that for some patients at high risk of developing cardiomyopathy, such as those with DMD or BMD, physicians can initiate some or all of these therapies to prevent the progression of muscular weakness. This preventive treatment approach would be a promising area for future research, as it has not been well studied.

Strong partnerships for heart health

Effective cardiac care in neuromuscular disease depends on strong partnerships among providers, patients, and families. For neuromuscular disease specialists, this means having a cardiologist to whom they can refer patients for heart health.

“For patients, the most important thing is to have regular follow-ups with a cardiologist who will screen for cardiovascular risk factors, obtain imaging of the heart, and work with you to initiate available therapies at the earliest time possible,” he explains. “As a cardiologist, my most important role is working with key team members in neurology and pulmonology to inform them of any cardiac progression or changes.”

He says this coordinated approach offers a full spectrum of care for the potential complications of neuromuscular diseases.

“Partnering with other specialists and relying on their experience has resulted in improved patient care and also patient and caregiver satisfaction,” Dr. Hickey says.

Resources

- Ready to test your knowledge? E-mail MedEd@mdausa.org to be invited to our new virtual Case Challenge series, including a session on cardiomyopathies.
- Watch the [MDA webinar: Cardiac Care for Neuromuscular Disease.](#)
- Download MDA’s clinician flashcard: [Cardiac Care in Neuromuscular Disease: Considerations & Resources.](#)
- Learn about Management of Management of Cardiac Involvement Associated With Neuromuscular Diseases
 - Read the full article published in Circulation: [Management of Cardiac Involvement Associated With Neuromuscular Diseases: A Scientific Statement From the American Heart Association.](#)
 - Read [Top Things to Know: Management of Cardiac Involvement Associated With Neuromuscular Diseases.](#)