

Duchenne's Muscular Dystrophy and The Heart

How does Duchenne's Muscular Dystrophy (DMD) affect the heart muscle?

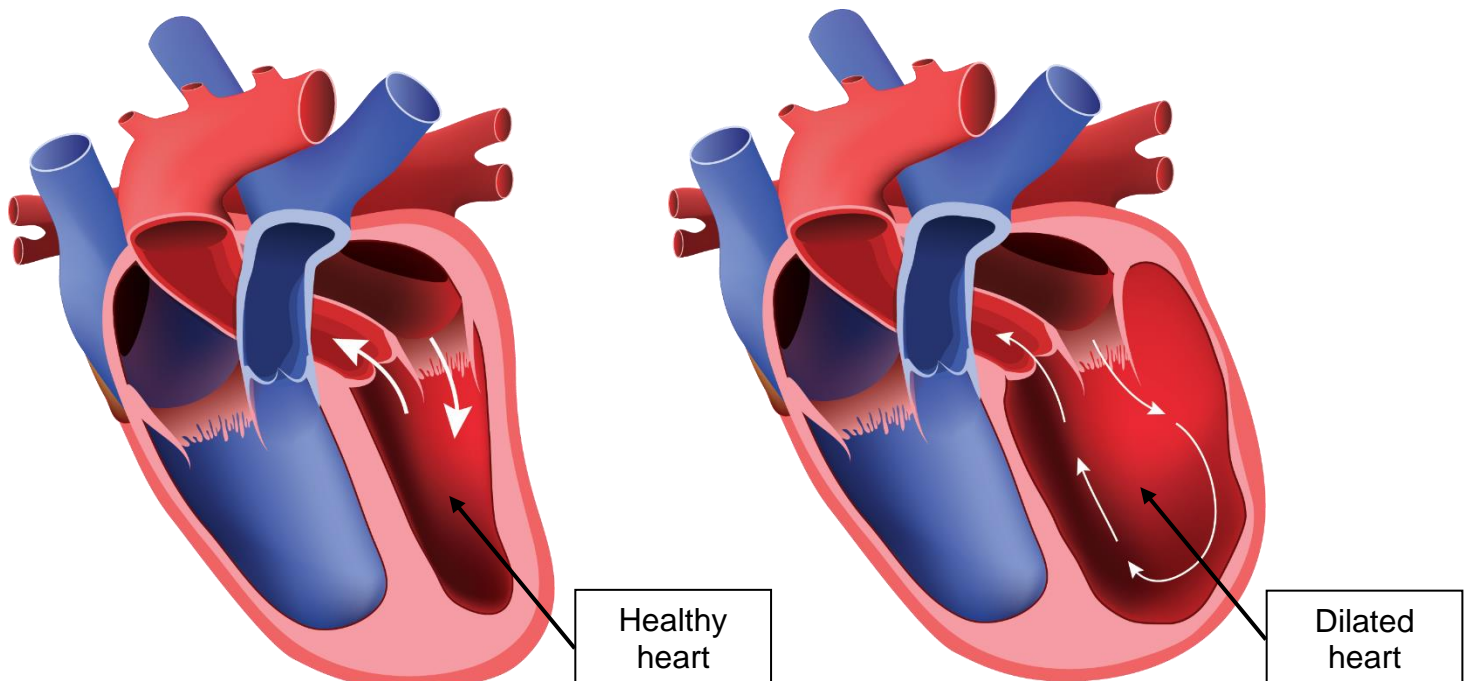
All people with Duchenne's Muscular Dystrophy have heart muscle disease. Heart muscle disease is also called **cardiomyopathy**. There are different types of cardiomyopathies.

What Causes Cardiomyopathy in DMD?

- People with DMD do not have a protein called dystrophin.
- Young kids with DMD do not have this protein even when they are very young.
- Over time, muscle cells without dystrophin become injured. Injured muscle cells are replaced with scar tissue and they become weak. The heart is a muscle and becomes weak too.
- As the heart muscle weakens, people with DMD eventually develop a heart condition called *Dilated Cardiomyopathy*.
- We can slow down the weakening of the heart by giving you certain medicines. You should discuss this with your child's doctor.

What is Dilated Cardiomyopathy (DCM)?

- In Dilated Cardiomyopathy, the heart muscle wall is weak and thin.
- The heart may not be able to pump enough blood to keep up with all of the body's needs.
- The heart's main pumping chamber (the left ventricle) becomes enlarged.



What are the stages of heart involvement in DMD?

- Preclinical stage (infancy/early childhood)
 - The heart exam is usually normal.
 - Children do not have heart symptoms.
- Walking stage (childhood/late childhood/early adolescent)
 - Mild scar tissue in the heart muscle begins to form.
 - Tests may start to show a mild heart weakening.
 - There may also be no signs at all.
 - A small number of people have heart symptoms. An example can be a change in how your heart beats (heart rhythm problem).
- Non-walking stage (late childhood/early adolescent/adolescent/young adult)
 - Heart testing by echocardiogram or MRI are often abnormal.
 - Many people will feel some symptoms of heart weakening or rhythm changes.
 - Breathing problems are common at this stage. They can make the heart work harder.
 - Even if breathing support is provided, heart failure can lead to hospitalization or death.

What can I expect after a DMD diagnosis?

- See a heart specialist shortly after DMD is diagnosed.
- Have your heart checked at least every year and more often, if needed.
- Have tests done to monitor the heart.
- Start medicine called an ACE inhibitor before age 10. This will slow the progression of heart weakening even before an echocardiogram shows changes.
- Other heart medications may be recommended by your doctor as cardiomyopathy gets worse.
- You may be asked to see other doctors including a pulmonary (lung) doctor or a heart rhythm specialist.

What are some possible tests to monitor the heart?

- Echocardiogram (ECHO) is an ultrasound that shows a moving picture of your heart.
- Electrocardiogram (EKG) shows the electrical activity (rhythm of the heart).
- Holter Monitor is a device you wear that shows the electrical activity of the heart. You will need to wear this for up to two weeks.
- Cardiac MRI uses magnets to take detailed pictures of the heart muscle.

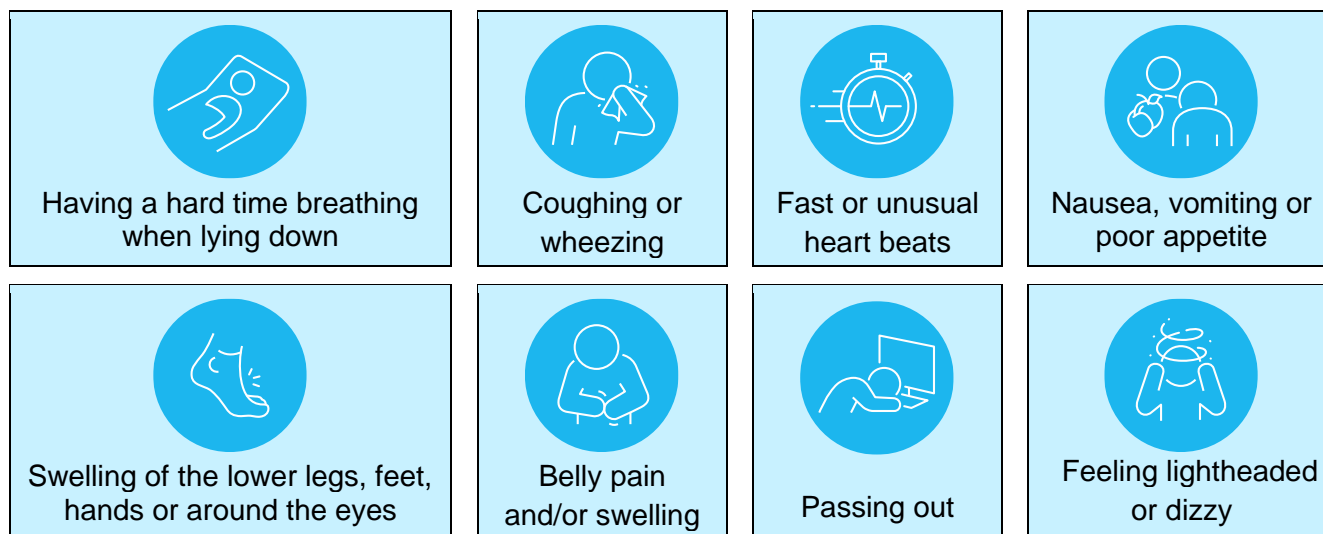
Heart Care for Carriers

- Women who are carriers of the Duchenne gene may develop heart problems, such as **dilated cardiomyopathy**.
- Recommend evaluation with a cardiologist when diagnosed.
- Recommend echocardiogram (heart ultrasound)
 - First in their late teens/early 20s.
 - Suggest repeating every 3-5 years.


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What are some signs and symptoms of heart weakening?

- People living with DMD may not have heart symptoms for many years.
- Some people may feel or have:



What do I do when there is an emergency or urgent health problem?

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| Call 911 if you experience the following:  | Having a hard time breathing even while resting |
| | Having chest pain that does not get better with rest |
| | Feeling confused |
| | Difficulty waking up or unable to wake up |
| | Passing out |

Call Your Nurse Care Manager or Cardiology Clinic when:

- you feel uncomfortable when breathing.
- feeling fast or unusual heart beats.
- having belly pain or swelling.
- you notice any new swelling in your eyes, feet, or lower legs.
- you are not eating well or not wanting to eat.

Who do I need to contact with questions or concerns?

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| Cardiology Clinic | 323-361-2461 | |
| Nurse Care Managers | Ani Yeremian 323-361-4622 ayeremian@chla.usc.edu | Lucy Dautrich 323-361-3355 lucyd@chla.usc.edu |
| Cardiologist On-Call | 323-660-2450 <ul style="list-style-type: none"> • Dial "0" for the Operator, ask for the Cardiologist On-Call • If after the clinic closes, on weekends and holidays | |

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4650 Sunset Blvd., Los Angeles, CA 90027 | CHLA.org

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