



Loeys-Dietz Syndrome: How to Care for Cardiovascular Problems



The NMF gratefully acknowledges the Loeys-Dietz Syndrome Foundation for their partnership in creating this fact sheet. For more information about the Loeys-Dietz Syndrome Foundation, visit www.loeysdietz.org.

People with Loeys-Dietz syndrome (LDS) need special medical care and management. The most important care involves the cardiovascular system—the heart, the aorta (the main blood vessel that carries blood from the heart) and other blood vessels. Here are ways to take care of the heart and blood vessels in a person with LDS:

TAKE MEDICATION

- Medications (drugs) that lower heart rate and/or blood pressure might help prevent bulging or tearing of blood vessels. Doctors often use medications called beta blockers to treat LDS in this way. They can use medications called calcium channel blockers when a person cannot take a beta blocker.
- Some blood pressure medications called angiotension receptor blockers (ARB's) may also help treat LDS in other ways. We need more research to learn more about how and whether ARB's can treat LDS. While we are waiting for more information, some doctors are using an ARB called losartan to treat LDS. Losartan is FDA-approved for lowering blood pressure, and the risks of using it appear low while the potential benefit appears high. You should talk with your doctor about using losartan to treat LDS.

MONITOR (CHECK ON) YOUR AORTA AND OTHER ARTERIES WITH SPECIAL TESTS

People with LDS should have:

- An echocardiogram to check the valves of the heart and the part of the aorta closest to the heart at least once a year.
- Either a CT or MRI angiogram—with contrast—of the head, neck, chest, abdomen (stomach area) and pelvis (lower stomach area just above the legs) on a regular basis. How often to have these scans depends upon the initial findings (results of your first scan.) The scans are looking for aneurysms and/or dissections (tears) in any of the arteries in the head, neck, chest, abdomen and pelvis.

Some people with LDS need to be checked more often than once a year. How often to have an echocardiogram and the scans depends upon the size of any aneurysms and how fast they are growing. Larger and faster growing aneurysms need more frequent monitoring.

GET REGULAR, BUT GENTLE, EXERCISE

- Most people with LDS can and should be physically active, but should not exercise to the point of becoming exhausted. As a general rule, while exercising, you should be able to comfortably talk with another person without needing to take breaths in the middle of short sentences.
- Try walking and gentle hiking, bike riding or swimming as safe ways to stay physically active.



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- You should not do exercises such as weight lifting, push-ups, chin-ups and sit-ups or other exercises that strain your muscles.
- You should avoid “contact sports” such as football and basketball or any other activity where there is a high risk of a sharp blow to the head or chest.
- Based upon the size of blood vessels or instability of the neck, some people with LDS need to be even more careful and gentle when exercising.

HAVE VASCULAR SURGERY (SURGERY OF THE BLOOD VESSELS)

Most people with LDS can be helped by “planned vascular surgery.” This means having surgery to remove aneurysms before they cause a life-threatening dissection (tear) or rupture. This is a very important part of LDS care.

- The most common vascular surgery is replacement of the aortic root (the part of the aorta closest to the heart).
- Doctors can use the size of an aneurysm, how fast the aneurysm is growing, where the aneurysm is located, and/or family history of early dissection or rupture to decide when it is time to perform vascular surgery.
- Vascular surgery is generally very successful in people with LDS.

Keep in mind that replacement of the aortic root is also done in people with other connective tissue disorders, especially for Marfan syndrome. Using Marfan guidelines to decide when to do surgery in a person with LDS can be dangerous, however, because aneurysms in people with LDS can tear or rupture at smaller sizes and at younger ages.

TALK WITH YOUR DOCTOR

LDS is not the same for everyone with the disorder. Some people have more medical problems than others. Therefore it is very important to talk with your doctor about what care is right for you.

TO LEARN MORE ABOUT LOEYS-DIETZ SYNDROME

- Call the National Marfan Foundation (NMF) Resource Center at 1-800-862-7326 ext. 26. When you call, you will speak with a nurse who can answer questions and also suggest ways to find a doctor who knows about LDS.
- Read the other NMF fact sheets on LDS. They have more information on LDS features, how LDS is different from Marfan syndrome and how LDS is diagnosed. They are available on-line at www.marfan.org or from the NMF Resource Center.
- Go to the Loeys-Dietz Foundation website at www.loeysdietz.org