

Loeys-Dietz syndrome

Caring for Cardiovascular Problems

How To Care For Cardiovascular Problems

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 some ways to take care of the heart and blood vessels in a person with LDS.

Take Medications

- Medications (drugs) that lower heart rate and/or blood pressure might help prevent bulging or tearing of blood vessels. Doctors often use medications called angiotensin receptor blockers and/or beta blockers to treat LDS in this way.
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Monitor 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 (aortic root) at least once a year.
- Either a CTA or MRA 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. It is recommended that an individual with LDS does not go more than two years without imaging of any portion of the body.

Some people with LDS need to be checked more often than once a year. The frequency of an echocardiogram and other vascular imaging depends on 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.

Get Regular But Gentle Exercise Continued

- Try walking and gentle hiking, bike riding or swimming as safe ways to stay physically active.
- Avoid exercises such as weight lifting, push-ups, chin-ups and sit-ups or other exercises which strain your muscles.
- 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 A 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 (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 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

Read the other fact sheets on LDS. Find them online at the Loeys-Dietz Syndrome Foundation Web site at www.loeysdietz.org/resources.php.

