

THE NERVOUS SYSTEM IN MARFAN SYNDROME

The nervous system—the brain and spinal column—are surrounded by fluid contained in a membrane called the dura, which is primarily made up of connective tissue. The enlargement of this membrane (and sometimes the presence of cysts) is referred to as dural ectasia. Dural ectasia is present in more than 60 percent of people who have Marfan syndrome.

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In 99 percent of people with dural ectasia, it occurs in the lowest part of the spine because this is where the fluid pressure is greatest when standing. It can also occur in the neck or in the upper torso, but this is very rare. Dural ectasia can develop following trauma, such as a car accident, in the general population, but it doesn't commonly occur this way in people with Marfan syndrome.

What are the common symptoms of dural ectasia in people with Marfan syndrome?

The presence of dural ectasia does not always cause problems, although in some people it causes aching in the very low back, almost in the tailbone, as well as abdominal pain, headaches, and leg pain. Pain and numbness in the perineum (the area between the legs in front of the anus) can also occur. Sitting for long periods of time may cause pain in people with dural ectasia.

Dural ectasia also thins the spinal vertebrae, which can have implications for someone having surgery on the spine for any reason.

The need for an evaluation depends on the degree of symptoms and disability. If you can tolerate the symptoms, there is no urgency for an evaluation. Dural ectasia is best identified through MRI imaging, particularly of the lower spine with a person standing upright. A myelogram, CT scan, or plain spinal films might also reveal the presence of dural ectasia.

How is dural ectasia treated in people in Marfan syndrome?

Currently, dural ectasia is treated primarily with pain medications. Unfortunately, it is not possible to replace the dura like it is the aorta. In extreme cases, surgical treatment of "sacs" of dural ectasia may be needed. Consult with your orthopedist and pain specialist on the best way to manage your symptoms.

Do you have questions? Would you like more information?

- Call our help center, 800-862-7326, ext. 126 to speak with a nurse who can answer your questions and send you additional information.
- Visit our website at marfan.org. You can print information that interests you and ask questions online.