

Let's Talk About...

Schwannomatosis: Tumors that affect the nervous system

Schwannomatosis (sh-WAHN-no-muh-TOH-siss) is a genetic disorder that causes noncancerous (benign) tumors, called schwannomas, to grow on the peripheral and spinal nerves. These tumors can cause pain that are hard to control.

One in 40,000 people worldwide develop schwannomatosis, a rare form of neurofibromatosis (new-roh-FIBE-row-muh-TOH-siss) each year. Neurofibromatosis is a group of genetic disorders that affect the nervous system.

What causes schwannomatosis?

Because schwannomatosis is a genetic disorder, your child either:

- Inherited an abnormal gene from a parent, or
- Inherited a gene mutation

Many children who have schwannomatosis are the first in their family to have symptoms of the disorder. However, these children can then pass schwannomatosis to their own children.



What are the signs of schwannomatosis?

Signs of schwannomatosis include:

- Chronic pain anywhere in the body (caused by schwannomas pushing on the nerves)
- Numbness or tingling
- Headaches
- Vision changes
- Weakness (including facial weakness)
- Problems with bowel movements
- Trouble urinating

Your child may have mild or severe pain, and they may not have other symptoms of schwannomatosis until later. This makes it hard to diagnose the disorder.

How is schwannomatosis diagnosed?

Your child's healthcare provider will look at your child and ask questions about their pain and symptoms. Sometimes the provider may be able to feel a tumor during a physical exam, but the schwannomas are usually found deeper in the body.

To diagnose schwannomatosis, your child may have:

- An MRI (a machine that uses magnets to take detailed pictures of the inside of the body)
- Genetic testing (blood or tumor samples with DNA)

If your child is diagnosed with schwannomatosis, they must have one of the following:

- Two or more schwannomas that are not in the skin (and one must be confirmed by a blood test)

- One schwannoma or meningioma (noncancerous tumor near the brain) and a parent or sibling who has schwannomatosis
- Two or more tumors that haven't been confirmed as schwannomas but will have chronic pain that makes schwannomatosis likely

How is schwannomatosis treated?

Treatment for schwannomatosis requires a multi-disciplinary team to manage your child's treatment. Although schwannomatosis has no cure, and there is no medicine to treat schwannomas, your child's care team will help manage the disorder by focusing on relieving your child's symptoms.

Treatments may include:

- Medicine for nerve pain
- Tricyclic antidepressants for depression and nerve pain
- Epilepsy medicine
- Surgery to remove schwannomas

Removing all tumors can reduce your child's pain and may be considered if the tumors are causing problems with other organs or the nervous system. However, not every child can have surgery to remove the tumors.

It's important for your child to see a specialist with experience treating schwannomatosis. Your child's healthcare provider should examine your child each year to see if their symptoms have improved and determine if any new tumors have formed.

What if I have questions about schwannomatosis?

If you have questions about schwannomatosis or your child's symptoms, contact your child's healthcare provider. You can also find more information about schwannomatosis on the Children's Tumor Foundation website, ctf.org.

Notes

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