Let's Talk About...

Neurofibromatosis type 2: A genetic disorder causing noncancerous tumors on cranial nerves

Neurofibromatosis (NEW-row-FIBE-row-muh-TOH-siss) type 2, or NF2, is a genetic disorder that causes schwannomas (shwah-NO-muhs), or noncancerous tumors, to grow on the eighth cranial nerves. These nerves bring sound and balance information from the ear to the brain. A child who has neurofibromatosis may have partial or complete hearing loss.

NF2 affects 1 in 25,000 people worldwide. It can also cause:

- Meningiomas (men-IN-gee-OH-muhs), or brain membrane tumors
- Ependymomas (ee-PEND-ih-MOH-muhs), or tumors of the brain ventricles
- Juvenile cataracts, or blurry lenses in the eyes

What causes neurofibromatosis type 2?

Neurofibromatosis type 2 is a genetic disorder, which means your child either inherited the disorder from a parent or was born with the disorder. Some children who have NF2 are the first in their family to have the disorder.

What are the signs of neurofibromatosis type 2?

The signs of NF2 include:

- Ringing in the ears (tinnitus)
- Balance problems
- Gradual hearing loss in one or both ears
- Brain and cranial nerve damage
- Facial weakness
- Seizures
- Trouble swallowing
- Vision problems

Many of these symptoms are caused by schwannomas growing on the nerves that go to the ears. Nerve damage can affect your child's muscles, senses, and even some organs.

How is neurofibromatosis type 2 diagnosed?

Your child's healthcare provider will look at your child and ask questions about their pain and symptoms. They may ask about their medical history and whether any relatives have NF1.

Your child may need x-rays or an MRI (more detailed photos of the body) to see if they have brain and spinal cord tumors. They may also need genetic testing (blood or tumor samples with DNA).

Children diagnosed with NF2 have:

- Vestibular (inner-ear) schwannomas that affect both ears

Or

- A parent or sibling with NF2
- A vestibular schwannoma that affects one ear or two of the following:
  - Ependymoma
  - Neurofibroma
  - Schwannoma
  - Juvenile cataracts
  - Meningioma

Or

- A vestibular schwannoma that affects one ear
- Two of the following:
  - Ependymoma
  - Neurofibroma
  - Schwannoma
  - Juvenile cataracts
  - Meningioma

Or

- 2 or more meningiomas
- A vestibular schwannoma that affects one ear
- Juvenile cataracts or two of the following:
  - Neurofibroma
  - Ependymoma
  - Schwannoma

How is neurofibromatosis type 2 treated?

If your child has NF2, they will be treated at a neurofibromatosis clinic with providers who have experience treating the disorder. Your child may also be referred to healthcare providers who work in:

- **Genetics:** Help with genetic disorders
- **Ophthalmology** (OP-thall-MALL-oh-gee)**:** Help with eye and vision problems

- **Neuro-ophthalmology:** Help with vision problems caused by the nervous system
- **Otolaryngology** (oh-to-LAIR-in-joll-oh-gee)**:** Help with ear, nose and throat problems
- **Dermatology:** Help with skin, nail, and hair problems
- **Orthopedics:** Help with muscle and bone conditions
- **Neurology:** Help with brain and nerve conditions
- **Oncology:** Help with benign and cancerous tumors
- **Neurosurgery:** Help with surgery for the brain and spinal cord

What if I have questions about neurofibromatosis type 2?

If you have questions about NF2 or your child's symptoms, contact your child's healthcare provider. You can also find more information about NF2 on the Children's Tumor Foundation website, ctf.org.

Intermountain
Primary Children's Hospital
The Child First and Always®