

Cochlear Nerve Deficiency (CND) and Narrow IAC

What is cochlear nerve deficiency (CND) and narrow IAC?

The **cochlea** is an organ in your inner ear, shaped like a snail shell, that sends sound information to your hearing nerve and up to your brain. **Cochlear nerve deficiency (CND)** is a term that includes 2 conditions: cochlear nerve aplasia and hypoplasia.

- **Cochlear nerve aplasia** is when a person does not have a hearing nerve.
- **Cochlear nerve hypoplasia** is when a person's hearing nerve is smaller than normal.

The **internal auditory canal (IAC)** is an opening in a bone on your skull around your ear (called your temporal bone). Your hearing, balance, and facial nerves run through the IAC where it connects to your brain. **Narrow IAC** is when this canal is smaller than normal. Usually someone with CND will also have a narrow IAC.

How severe the narrow IAC or hypoplasia is may be different for different people. These conditions can happen in one or both ears. Having a narrow IAC or hypoplasia will cause significant hearing loss.

How are CND and narrow IAC diagnosed?

CND and narrow IAC are diagnosed by a medical professional, typically an **otolaryngologist** (an ear, nose, and throat doctor). They will use imaging tests, such as an MRI and CT scan, to look at the structures of your ear.

- An **audiologist** (a doctor specializing in diagnosing and treating hearing loss) will do a hearing test with you to understand how well you can hear and how much hearing loss you may have.

What are other conditions that someone might have if they have CNND and narrow IAC?

CND and narrow IAC are types of **congenital** (present at birth) causes of hearing loss. They can happen together or with other ear **malformations** (when a body part develops in a way that is not normal) or **syndromes** (groups of symptoms related to a medical disorder), so genetic testing can be helpful. Some other conditions that happen with CNND and narrow IAC include auditory neuropathy spectrum disorder (ANSD) and CHARGE syndrome. If someone has a narrow IAC because of a syndrome, it is important to have this diagnosis so they can get appropriate treatments and talk to the right specialists.

How are CNND and narrow IAC treated?

- Cochlear implants may help people with CNND and narrow IAC if they have a cochlear nerve. However, cochlear implants usually don't work as well for people with CNND and narrow IAC compared to people who have a normal nerve structure. It may work better for these patients to use a combination of speech (talking) and sign language for communication. This is called **total communication**.
- Instead of a cochlear implant, an **auditory brainstem implant (ABI)** may help your hearing by skipping your cochlea and cochlear nerve and sending sound signals directly to your brain. People with an ABI usually use a combination of speech and sign language for communication. Talk with your doctor if you are interested in learning more about ABIs.

Where can I find more information and resources about hearing loss?

American Speech-Language-Hearing Association

- List of hearing loss organizations:
www.ASHA.org/public/hearing/hearing-loss-organizations-and-associations



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