UNIVERSITY OF MICHIGAN HEALTH
MICHIGAN MEDICINE

Mondini Dysplasia

What is Mondini dysplasia?

The **cochlea** is an organ in your inner ear. It is normally shaped like a snail shell with 2.5 turns or spirals. The cochlea sends sound signals to your hearing nerve and up to your brain. **Mondini dysplasia** (or Mondini malformation) is a type of inner ear **malformation** (when a body part develops in a way that is not normal). With Mondini dysplasia, the cochlea has less than the usual 2.5 turns.

Mondini dysplasia can happen in one ear or both ears, and how severe the malformation is may be different for different people. Because the inner ear codes sound signals and sends them to the brain, having a malformed inner ear sends a distorted (changed or different) signal to the brain. This causes hearing loss and problems with understanding speech.

How is Mondini dysplasia diagnosed?

Mondini dysplasia is diagnosed by a medical professional, typically an **otolaryngologist** (an ear, nose, and throat doctor, or ENT). They will use imaging tests, such as an MRI or CT scan, of the inner ear to figure out the cause of your hearing loss.

- 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 Mondini dysplasia?

Mondini dysplasia is a type of **congenital** (present at birth) cause of hearing loss. It can happen in one or both ears.

Mondini dysplasia commonly happens in people who have Pendred syndrome. Some syndromes can cause other significant health issues, and it is important to follow up with the appropriate specialist. Some other conditions that happen with Mondini dysplasia include:

- DiGeorge syndrome
- CHARGE syndrome
- Klippel-Feil syndrome
- Autosomal dominant and recessive inheritance

Mondini dysplasia also commonly occurs with another inner ear malformation called **enlarged vestibular aqueduct (EVA)**. An EVA is a condition where a narrow, fluid-filled tube in the inner ear is wider than normal.

Children with Mondini dysplasia may also have other inner ear malformations that affect the **vestibular** (balance) system, so a vestibular audiologist or physical therapist can be helpful to treat balance issues, if needed.

How is Mondini dysplasia treated?

- Depending on the how much hearing loss you have, hearing aids or cochlear implants may help you hear better. This will help you to develop speech (talking) and understand others.
- If you have balance issues, you may get a recommendation for **vestibular therapy**. Vestibular therapy is a type of physical therapy, involving exercises that improve dizziness and problems with balance caused by inner ear disorders.

What are other resources for Mondini dysplasia or hearing loss?

National Organization for Rare Disorders • Overview of Mondini dysplasia: rarediseases.org/gard-rare-disease/mondini-dysplasia	
American Speech-Language-Hearing Association • List of hearing loss organizations: www.ASHA.org/public/hearing/hearing-loss-organizations-and-associations	

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