Fabry disease is a rare disease caused by gene mutations. People with Fabry have trouble breaking down and getting rid of certain fatty waste substances. The disease can affect many parts of the body, including the kidneys, eyes, heart, skin, and vascular system.\textsuperscript{1,3}

The rate of hearing loss in the Fabry community is much higher than that in the general population and tends to start earlier and progress faster.\textsuperscript{4}

Hearing loss in Fabry is believed to be caused by an accumulation of these waste substances in the blood vessels of the inner ear. This blocks and narrows the blood vessels, reducing blood flow and damaging the auditory nerve.\textsuperscript{2,3,5}
YOUR EAR

The human ear is designed to capture sound and convey its signals to the brain. The ability to hear sound helps us understand our environment, communicate with others, learn, and develop.

THE EAR IS MADE UP OF 3 DIFFERENT PARTS:

THE OUTER EAR includes the visible part called the pinna, which catches sound and sends it to the ear canal inside.

THE MIDDLE EAR contains the eardrum and three tiny bones called ossicles that vibrate to the sound.

THE INNER EAR houses the spiral-shaped cochlea, which converts sound vibrations into nerve impulses that are sent to the brain.

Each part of the ear is essential to good hearing and any impairment or damage along the way can affect your ability to hear.

Hearing loss is quite common. According to the World Health Organization, over 5% of the world’s population has disabling hearing loss. The rate of hearing loss in the Fabry community is much higher than that in the general population and tends to start earlier and progress faster.

The most common type of hearing loss associated with Fabry is called sensorineural hearing loss, which is caused by damage to the nerve fibers within the inner ear. This reduces both the volume and clarity of perceived sound. While this type of hearing loss is permanent, progressive, and irreversible, there are many ways it can be treated—from hearing aids to cochlear implants.

HOW DO I KNOW IF I HAVE HEARING LOSS?

This symptom checklist may help you determine if your hearing may be impaired.

DO YOU:

- Find yourself constantly turning up the volume on the phone, television, or other media devices?
- Hear a constant ringing in your ears (tinnitus)?
- Have difficulty understanding or keeping up with conversations?
- Need to be face-to-face with someone in order to carry on a conversation?
- Frequently ask people to repeat themselves?
- Often think people are mumbling?
- Have trouble understanding others when there is background noise?
- Feel you are constantly straining to hear?
- Avoid encountering new places, new people, groups, or social situations?

If you have answered yes to any of these questions, you may have hearing loss and should consider discussing a full hearing evaluation with your healthcare provider.
How is hearing tested?¹²

Hearing or audiology testing is simple, painless, and noninvasive. It is performed by a hearing healthcare professional and usually involves:

- A thorough hearing health history
- Audiometry testing for tone and speech. During these tests you will wear headphones and:
  - Listen for tones at different volumes and pitches
  - Repeat words heard at different loudness levels
- Tympanometry testing to gauge your eardrum’s movements. During this test a soft plug that changes sounds and pressure is placed in your ear
- Reviewing a graph depicting the results of your tests, called an audiogram, with your healthcare professional

Why should I seek treatment for hearing loss?¹³

Hearing loss can impact your quality of life in a variety of ways—from talking with friends and colleagues, to enjoying media and cultural events, to engaging in social activities. The sooner you treat hearing loss, the sooner you can expand or enrich all kinds of communications and experiences. And your friends and loved ones will feel the impact, too.

If left untreated, hearing loss can also have more serious effects on your overall health and well-being.¹³

In adults, untreated hearing loss may:

- Put a strain on interpersonal relationships
- Lead to withdrawal from social activities, contributing to loneliness and isolation¹³⁻¹⁵
- Be linked to emotional issues, such as irritability, frustration, negativity, and anger, and lead to psychological problems including depression, stress, and anxiety¹³⁻¹⁵
- Result in reduced alertness and awareness, increasing the risk to personal safety¹⁶,¹⁷
- Contribute to mental decline including impaired memory, thinking, and judgment¹⁸,¹⁹
- Have a negative economic effect. People with untreated hearing loss are more likely to be unemployed—and those who find work tend to earn less, and have more difficulty functioning on the job than those with normal hearing.²⁰,²¹
  Their medical costs tend to be higher as well, which can also take its toll on finances²²,²³

Hearing loss in children can have even more devastating effects and should be addressed early to ensure normal development.²⁴

Why? The ability to hear is key to learning how to listen and to talk. It’s fundamental to developing language skills and spoken communication. If left untreated, childhood hearing loss can have a lasting impact, even beyond speech development. Difficulties in communicating can:

- Impede the development of social skills
- Lead to social isolation and low self-esteem
- Interfere with the development of the learning and attention skills needed to succeed in school and life²⁴
What types of treatments are available?

TREATMENT FOR TINNITUS

There are a variety of treatments available for tinnitus—from medications to masking devices to various forms of therapy. Which one is right for you will be determined during a medical examination. If your tinnitus is associated with hearing loss, hearing aids may be helpful.

HEARING AIDS

Hearing aid technology has advanced dramatically in recent years. These noninvasive devices are designed to amplify sound—and they come in so many different types and configurations, there should be one to fit every ear and budget! Most hearing aids are wireless and battery operated. The two main types are:

In-the-ear (ITE)

These aids consist of a single piece designed to fit snugly in your ear canal. There are three types of ITEs:

- The smallest, most discreet ITEs sit deep in the ear canal and are called completely in the canal (CIC)
- In the canal (ITC) are a bit larger than the CIC and sit in the lower portion of the outer ear bowl. Their size makes them more visible, but that allows them to include more features, such as volume control
- Low profile (LP) are the largest, filling up to 100% of the ear bowl, and can accommodate even more components

Behind-the-ear (BTE)

This type of aid has two components: a piece that sits behind the ear that holds the electronics and an eartip inside the ear canal. Like the ITE aids they come in different sizes that provide a variety of features, including receiver in the ear (RITE) and receiver in the canal (RIC).

COCHLEAR IMPLANTS

For those with severe or profound hearing loss, the surgical implantation of a device that directly stimulates the auditory nerve may be a viable option. Rather than amplifying sound, the cochlear implant provides the sense of sound. It consists of two components. The external piece contains a microphone, processor, and transmitter. The internal part contains a receiver that converts signals from the transmitter to electrical pulses that are sent through the nerve to the brain, where they are perceived as sound.

ASSISTIVE LISTENING DEVICE (ALD)

ALDs provide amplification of sound or improved communication for specific hearing challenges. Some are designed to work alone and some with hearing aids. Categories include:

- **Telephone:** A wide variety of options are available to enhance telephonic communication—for both mobile and landline—from amplified phones to captioned telephone service, from Bluetooth ear pieces to hearing aid–compatible phones
- **Television:** There are many devices that can help you hear sound without ramping up the volume—from wireless headsets with personal volume control to Bluetooth-enabled accessories for your hearing aids
- **Alerts:** So you don’t miss important warnings, alerts, or appointments, there are a number of available systems that feature enhanced sound, visual cues, vibration, or some combination of these. They include vibrating alarm clocks that shake you awake in the morning; doorbells, smoke alarms and carbon monoxide detectors that use increased volume and flashing lights to get your attention; and wearable technology, such as bracelets, smart watches, and pagers that use vibration and light to alert you to incoming calls and alarms
- **Wireless microphones:** These small, light, portable devices are designed to boost your hearing in noisy situations, like restaurants, meeting rooms, or outdoors near traffic. From clip-ons to discs to pens, you can carry them with you wherever you go

SPEECH THERAPY

Speech therapy can help children with hearing loss catch up on delays in speech development once they have been fitted with hearing devices.

The cost of testing and treatment will depend on where you live, the type of healthcare available, and your health insurance coverage.
Here are some strategies and workarounds to promote better communication with hearing loss:

- Talk to people about your hearing loss. If they understand your situation, they can work with you to enhance communication.

- Don’t be embarrassed to ask for what you need in a given situation, whether it’s having someone repeat what they have said or requesting a seat where you can see everyone’s faces.

- Try to sit face-to-face when you are speaking with someone, so you can hear them more clearly and pick up on visual cues.

- Listening closely requires serious concentration and can use lots of energy. If you are tired under these weather don’t push yourself, take a break in a peaceful place.

- Connect with other people with hearing loss. They can be a great source of support and information.

Resources*
Check out these online sources for information on hearing loss, treatment and support.

Hearing loss in Fabry

Hearing loss in children

Hearing loss, treatment, financial assistance