

HOW WE HEAR

The 3 parts of the ear

The ear consists of three main parts:

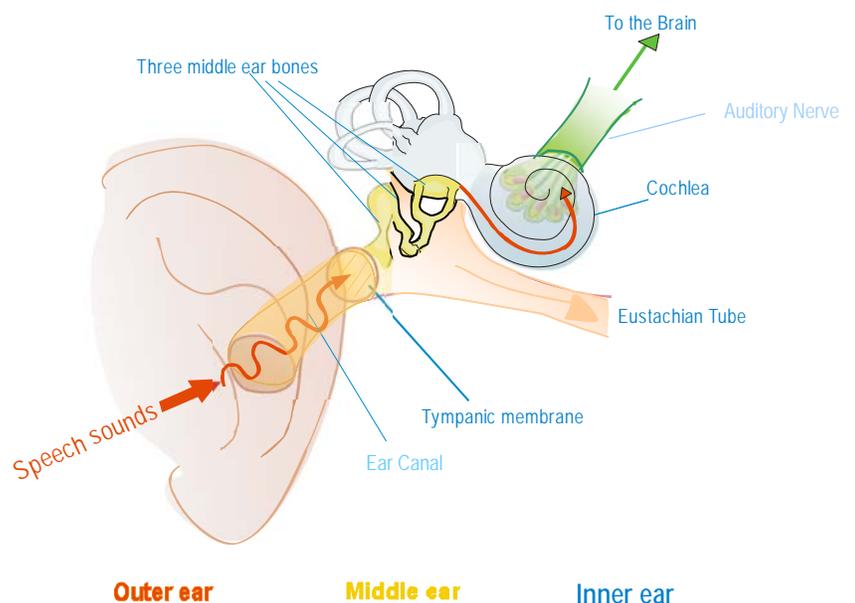
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|-------------------|---|
| 1. The outer ear | The part you see, which is called the auricle (ohr-a-kal). |
| 2. The middle ear | The middle ear is a sealed area, with the eardrum on one end and the cochlea (ko-klee-uh) on the other. Inside this area are the three bones of the middle ear, also called the ossicles (ah-sih-kulz), and the muscles attached to them. |
| 3. The inner ear | The part that is deep within the ear, encased in the temporal bone of the skull, is called the cochlea. The cochlea contains the tiny hair cells that trigger a signal to the auditory nerve, which takes the information to the brain.

The inner ear also contains the vestibular (ve-stib-ew-lar) system, which helps us maintain our sense of balance. |

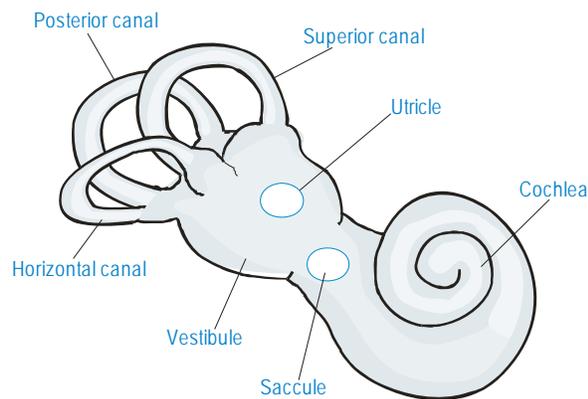
The Ear in Action

The Outer and Middle Ear

The outer and middle ear serve to capture sound and send it to the cochlea, where the mechanical vibrations are turned into electrical impulses that travel along the hearing nerve to the brain. The most common problems to occur here are outer or middle ear infections, both of which are easy to treat with antibiotics or minor surgery and rarely cause permanent hearing loss.



PERIPHERAL AUDITORY SYSTEM



The Cochlear and Vestibular Systems

The Inner Ear

The inner ear contains the cochlea and the vestibule. The cochlea is the organ of hearing. The vestibular system is attached and controls our sense of balance. These two are connected and affect each other. When one is experiencing difficulty it is not uncommon for the other to experience difficulty.

The cochlea has tiny hair cells that respond to sounds coming into the ear. These hair cells move in the fluid of the cochlea, and stimulate the auditory nerve. The auditory nerve carries the information to the brain, where it is interpreted.

The cochlea has two types of hair cells. Each plays a role in hearing. The outer hair cells serve to help make sounds that come into the ear louder. They are a natural amplifier for the ear. The inner hair cells' job is to trigger impulses to send this signal to the auditory nerve.

In normal hearing individuals these two types of hair cells work together to create a signal that is picked up by the auditory nerve and carried to the brain for interpretation. In the case of PMD, it seems that the cochlea is working fine as well as the first part of the auditory nerve. However, the later waves of the auditory brainstem responses (ABR) are absent which suggests that part of the brainstem (and may be also some more central parts of the central auditory system) are not functioning properly. Most individuals with PMD are not deaf but have an issue with the clarity of the signal that is being send and processed by the brain.

Because of this damage, the signal that arrives at the brain for interpretation is not received in an organized fashion. In some cases, it may be that most sounds seem almost the same to the brain. They all sound like static or simply noise. Such individuals can sometimes detect sounds, but cannot tell what the sound is. A voice

may sound the same as water running, a dog barking may sound the same as a car horn, a bird chirping may sound the same as a pan banging.

Auditory Neuropathy (AN)

Patients with PMD usually present with:

- Normal outer hair cell function.
- Abnormal neural function at the level of the brainstem.
- A problem with auditory timing.
- Hearing levels on the audiogram are variable.
- The clarity of sounds is usually impaired

The exact site in the auditory system where the timing break down occurs is still not known. What we do know is that the *outer hair cells* and the *inner hair cells* usually work well, but there is a problem with the auditory pathways in the brainstem and may be also at a more central level. Most patients have a wave I (and sometimes II) on the ABR which seems to indicate that the inner ear is working normally.

Most (if not all) individuals will have difficulty hearing in noisy environments. Hearing in quiet environment can also be impaired. This is due to the fact that the firing of the nerve fibers in the brainstem is abnormal.

Unlike with the more common form of permanent hearing loss that is due to *outer hair cell* loss, in cases of AN the auditory brainstem response (ABR) and *audiogram* do not provide us with information as to the severity of hearing impairment.

The *central auditory evoked potentials* are usually present in PMD patients despite the lack of later waves on the ABR, which indicate that sounds are reaching the level of the auditory cortex. Although it is not possible to precisely determine which sounds are really reaching the cortex and how they are processed by the brain, it seems that this technique has more potential than the ABR to assess the hearing abilities of patients with PMD. It can also be used to monitor the hearing of individuals over time.

Because auditory input is distorted for individuals with PMD, supplementing the auditory signal with visual information can dramatically aid in understanding

language. With PMD, a child may not require much in the way of amplification with hearing aids because the natural process of amplification in the inner ear (the role of the outer hair cells) usually works well. Hearing aids allow sounds to become louder but do not improve the clarity. In PMD it is suspected that sounds can become quite unclear to the listener, so making sounds louder may not help the child understand them. An FM system (described below) might however provide help to the listener by reducing the amount of noise he/she is exposed to, hence increasing the clarity of what is being said. This is important when we're trying to listen in a noise environment, which represents a natural listening situation for most of us.

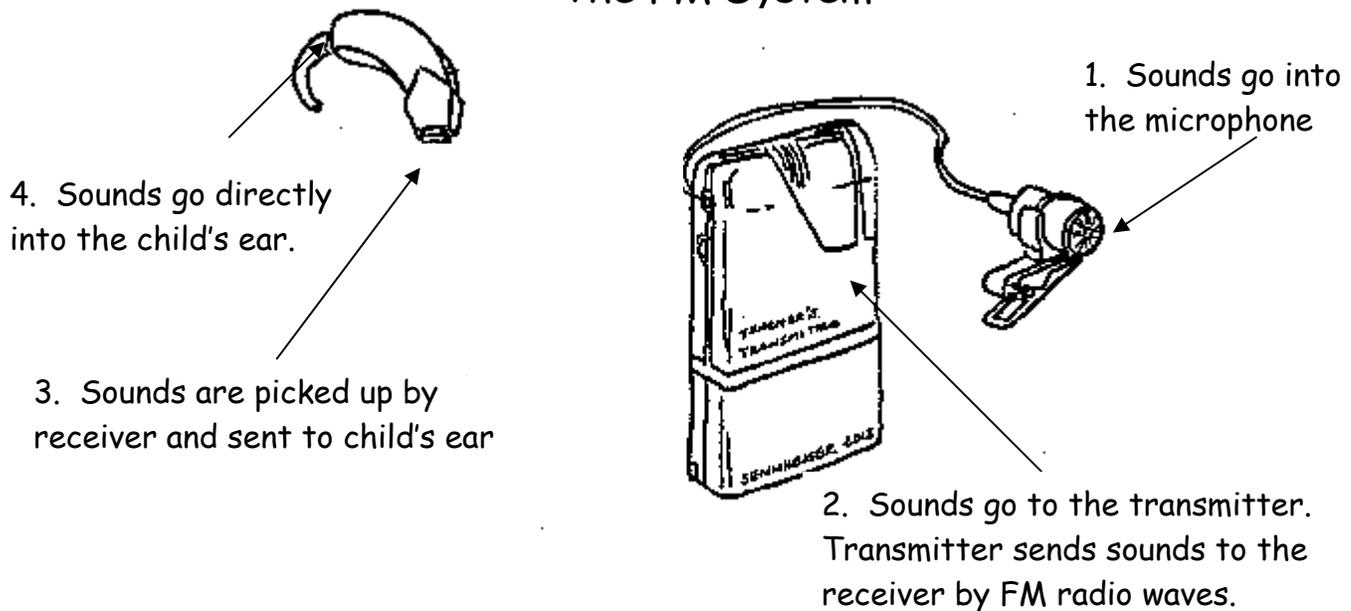
The FM System

The team may recommend an FM system. The FM system is similar to a personal radio system. The FM system does two things:

1. It will reduce background noise
2. It will reduce the distance from the speaker's voice.

These are both important for children with ANSD who need sound clarification, more than sound amplification.

The FM System



An FM system will help to make the speaker's voice louder than the background noise so that your child can hear and understand the speech signal.

Other Recommendations

A quiet environment

Individuals with PMD struggle to isolate and focus on the signal of the teacher's voice. A room that is noisy makes this much more difficult for them. This student will benefit from a room that is as quiet as possible. This can be facilitated by using carpet in the room, having the walls covered with sound-absorbing materials such as paper, using some shades over the windows, and putting tennis balls on noisy chair and desk legs.

Students with PMD need to sit away from noise producers such as heating systems or water fountains. Additionally, they need to sit in the front and they need to be able to see the teacher at all times.