

Dear Families,

Welcome to the Cochlear Implant Program at Texas Children's Hospital! Our goal is to provide comprehensive, specialized care for children with significant hearing loss and help them achieve the best possible communication outcomes.

Your child may be a candidate for a cochlear implant in one or both ears. Cochlear implants, or Cls, are innovative devices that directly stimulate the auditory (hearing) nerve and provide access to sound in children with severe or profound hearing loss. The decision about whether to recommend a Cl is highly individualized and requires the input of specialists in multiple areas, including Otolaryngology, Audiology, Speech Pathology and Neuropsychology, among others. These specialists are the members of the Cochlear Implant Team. They work in close collaboration to determine who is a candidate for a Cl, perform the implantation surgery and help patients through the rehabilitation afterwards.

This guide will help you understand the entire process, who may be considered a CI candidate, the basics of how the device works, how the surgery is done and the necessary rehabilitation to achieve a successful outcome.

Thank you for the opportunity to help your family through this journey. All of our team members are dedicated to helping children with hearing loss achieve their full potential. If you have any questions or concerns, please do not hesitate to contact us.

Sincerely,

The Cochlear Implant Program Team at Texas Children's Hospital



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Section 1 How We Hear

The auditory system, or hearing system, keeps us connected to our world 24 hours a day. **This system never turns off!** Even when we are asleep, the auditory system is working. The auditory system consists of the ears, auditory pathways, and the **brain**. It is the **brain** that gives those sounds meaning.

Outer Ear • Pinna

Middle Ear

• Eardrum

• Auditory canal

Ossicles – 3 little bones

Inner Ear

- Cochlea responsible for hearing
 - Fluid filled, snail-shaped structure
 - Contains sensitive hair cells, which are sensitive to specific frequencies
 - High frequency sounds are stimulated at the base of the cochlea
 - Low frequency sounds are stimulated at the tip
- Vestibular organs responsible for balance
- Auditory nerve

Central Audiory System

- Auditory pathway
- Brain

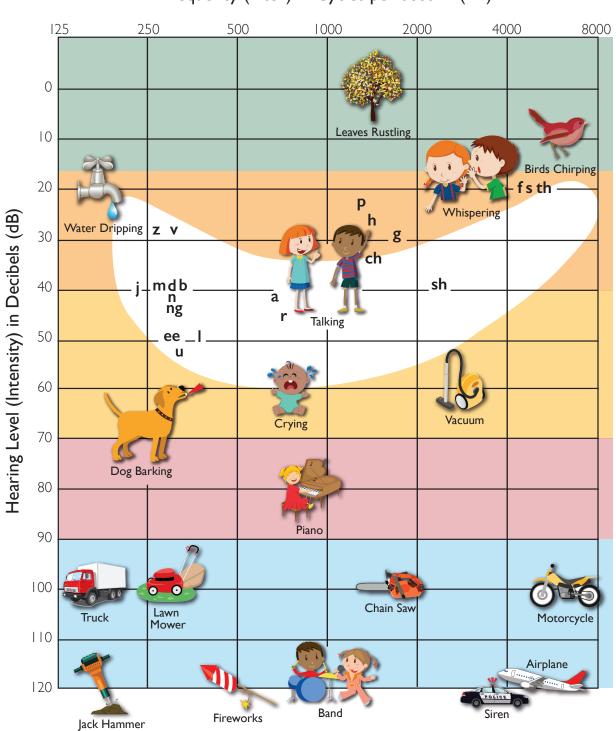
Outer Middle Inner Pinna Vestibular organs Auditory nerve sends signal to brain Cochlea contains hir cells

HOW HEARING WORKS

- The pinna collects sound waves and funnels them into the auditory canal.
- This sound makes the eardrum vibrate.
- The ossicles connect to the eardrum. As the eardrum vibrates, the ossicles move.
- The ossicles' movement creates enough force to move the fluid inside the cochlea.
- The movement of the fluid creates movement of the hair cells.
- The movement of the hair cells creates impulses that stimulate the auditory nerve.
- The auditory nerve passes sound to the auditory cortex of the brain.
- The brain processes the sound for meaning.

Section 2 Audiogram of Familiar Sounds

An audiogram is a graph that shows at what intensity (decibel, dB) you can hear different sound pitches (Hertz, Hz). Audiometric testing will compare your child's results to the results of other children with normal hearing.





Degree of Hearing Loss	Decibles (dB HL)	Sound Examples	Communication Concerns
Normal Hearing	0-15 dB HL	Rustling of leaves	Will hear speech and language clearly.
Mild	16-40 dB HL	Quiet/whispered speech	Difficulty hearing soft speech especially in noisy areas.
Moderate	41-55 dB HL	Normal conversational speech	Difficulty hearing speech clearly especially at a distance, in background noise, and when speaker is not facing the child.
Moderately- Severe	56-70 dB HL	Normal to loud speech, vacuum cleaner, baby crying	Difficulty hearing speech clearly especially at a distance, in background noise, and when speaker is not facing the child.
Severe	71-90 dB HL	Loud shouting, dog barking	May hear loud voices up close but continue to have difficulty understanding speech even with hearing aids. May need a cochlear implant to understand speech.
Profound	≥9I dB HL	Airplane, chainsaw, fireworks	Difficulty understanding speech with hearing aids. May need a cochlear implant to understand speech.

dB HL = Decibels Hearing Level

Section 3 What is a Cochlear Implant? The Basics

When hearing loss is caused by dysfunction of the inner ear (damaged hair cells or auditory nerve) it is called sensorineural hearing loss (SNHL). SNHL cannot be reversed because hair cells cannot be repaired once damaged.

A cochlear implant is an amazing hearing device that bypasses the impaired hair cells. A cochlear implant is different from a hearing aid.

HOW HEARING AIDS WORK

- Make sounds louder
- If hearing loss is severe, the volume and the quality of the sound with the hearing aid may not be good enough for a child to learn to speak and communicate.
- Sounds still have to travel through an impaired inner ear, specifically the hair cells of the cochlea



HOW COCHLEAR IMPLANTS WORK

- Cls bypass the damaged hair cells
- They stimulate the auditory nerve directly

A cochlear implant may be a good choice for your child:

- If hearing aids provide little benefit
- If listening and talking are the desired communication outcome
- If the inner ear anatomy supports this technology

Parts of a Cochlear Implant: All manufacturers have similar parts. A. External Processor

- A. External Processor
- B. Coil and Cable
- C. Internal Processor and Electrode Array

Use the QR codes for links to videos about how Cls work. Each manufacturer has a video.



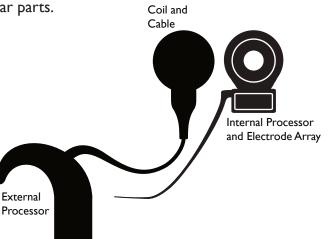
Advanced Bionics

Cochlear Americas



MEDEL





Section 4 The Doctors, Specialists and Tests that are part of the Cochlear Implant Process

WHY DOES MY CHILD NEED ALL OF THESE APPOINTMENTS?

- Evaluation for a CI requires input from several specialized professionals working as a team.
- These visits and tests give important information to understand if a CI can benefit your child.
- Not all children need all of these visits/tests. Your doctor will tell you the ones needed.

WHAT ARE THE APPOINTMENTS? WHAT WILL I LEARN?

ENT (Ear, Nose and Throat) Appointments – An ENT will:

- Review all relevant medical testing and results of prior evaluations
- Check the structures of the ears at an office exam and reviewing imaging results, such as MRI and/or CT, to determine if the inner ear structures are able to support a CI
- Recommend other testing as needed
- Discuss risks, benefits and expectations of surgery

Audiological Evaluations – An Audiologist will:

- Diagnose hearing loss, specifying the type and severity
- Perform proper fitting of hearing aids to amplify sound
- Test and monitor performance of the child's hearing with and without hearing aids with follow-up testing

IMPORTANT – It may take several visits to get all of the information needed. **The child must wear hearing aids to all appointments.**

Before we can determine if a child is a candidate for Cl, a hearing aid trial of 3 months may be required.

Speech-Language Evaluation – A Speech-Language Pathologist, also called an Audiology-Verbal Therapist (AVT), who specializes in children with hearing loss, will:

- Assess your child's current speech, language and listening skills
- Provide education and counseling to understand how a CI could benefit your child
- Make recommendations and develop a short and long-term plan of care
- Provide highly specialized therapy services before and after cochlear implantation

IMPORTANT – It may take several visits to get all of the information needed. **The child must wear hearing aids to all appointments.**







ADDITIONAL EVALUATIONS

Program Coordinator: Main point of contact for the CI program, assists with the scheduling and coordination of appointments, gathers records from other facilities, and makes sure that all evaluations are completed in a timely manner, among other responsibilities.









Your child may need to undergo the following evaluations.

MRI and/or CT Scans – These tests will:

• Show the physician the inner ear, auditory nerve and related structures

ECG – This test will:

• Examine your child's heart activity

Genetics Evaluation – A Genetics Health Provider may:

• Investigate genetic reasons for hearing loss

Neuropsychology – A Neuropsychologist will:

- Assess your child's overall development.
- Evaluate your child's ability to learn
- Include caregivers in the testing. Support is **critical** to the child's success with a cochlear implant

Ophthalmology Consult – An Ophthalmologist will:

• Check your child's vision

Lab work – This is to:

• Investigate a possible reason for hearing loss

Vestibular Testing:

• Observe and test your child's balance

Social Work – A Social Worker will:

- Give emotional support and community resource guidance for caregivers during the process
- Provide resources for lodging and transportation assistance (based on insurance benefits)

An Insurance Specialist will:

• Verify insurance coverage for CI surgery

VACCINATIONS

Children with hearing loss and cochlear implants are at a higher risk of contracting meningitis. Meningitis is an infection that causes serious and possibly life-threatening inflamation of the brain and spinal cord. It is caused by either a virus or bacteria. Vaccinations against *Streptococcus pneumoniae* reduce that risk. You will be asked to provide vaccination records.

If your child is younger than 24 months, please check with your pediatrician or health care provider to make sure the child is up to date on all routine vaccinations, particularly **Prevnar 13 (PCV 13).**

If your child is 24 months or older, the **Pneumovax 23 (PPSV 23) vaccine** is recommended by the Centers for Disease Control and Prevention (CDC). Visit your pediatrician or health care provider to receive the vaccine.

Section 5 Cochlear Implant Candidacy Criteria

TO GET A COCHLEAR IMPLANT, YOUR CHILD MUST:

- I. Be diagnosed with significant
 - a. Sensorineural Hearing Loss (SNHL) or Auditory Neuropathy Spectrum Disorder (ANSD)
 - b. Inadequate speech and language development
- 2. Have regular appointments scheduled with
 - a. An auditory-verbal therapist who focuses on the development of listening (or auditory) skills, which enable the production of speech and language
 - b. Deaf and hard of hearing specialist (DHHS) who will assist the family in creating an academic plan, which is tailored to the child's on-going listening, speech, language and developmental needs
- 3. Be evaluated by the following Texas Children's CI team members
 - a. Otolaryngologist (Ear, Nose and Throat, or ENT surgeon)
 - b. Auditory-Verbal Therapist
 - c. Cochlear Implant Audiologist
 - d. Others as recommended
- 4. Have limited success with consistent hearing aid use The team uses hearing (audiological) testing and listening performance in daily activities to determine if your child can hear better with with hearing aids.
- 5. Have an inner ear (cochlea) and auditory nerve that allows the CI to work correctly.
- 6. Have caregiver support and realistic expectations All of the evaluations will help the CI team estimate what the device can do for your child. Every child's expected benefit with a CI is different. For some, the expected benefit is to learn, listen and speak as well as their hearing children of the same age. For others, the expected outcome may be improved quality of life through improved awareness of sound.
- 7. The team also considers the child's
 - a. Educational placement
 - b. Additional medical issues
 - c. Current speech therapy services
 - d. Level of caregiver support and involvement

Section 6 Cochlear Implant Pathway

- I. Identify hearing loss in your child.
- 2. Visit with an ENT doctor to discuss treatment options.
- 3. Meet your child's audiologist for a hearing aid fitting and trial.
- 4. Have an auditory-verbal speech evaluation and enroll in audio-verbal therapy specialized to your child's needs.
- 5. Complete any additional imaging and consultations as recommended by your child's care team (for example, MRI/CT, neuropsychology, genetics or others).
- 6. Visit with an audiologist specialized in cochlear implantation to conduct an aided audiometric evaluation. (We will help connect you with the right audiologist if your current provider does not specialize in cochlear implantation.)
- 7. After these evaluations have been completed, the Texas Children's Hospital multi-disciplinary cochlear implant review board will discuss your child's hearing. The team will determine the best way to help based on their results and progress. We will notify you, if your child is approved or denied a Cl.
- 8. If approved, the CI coordinator calls you to set dates for the surgery to implant the CI and other appointments needed before and after surgery.
- 9. Two to four weeks after surgery, your child's ENT will check the healing at the surgical site and the audiologist will activate the Cl.
- Attend activation appointments and audiology check-ups to optimize the CI and your child's hearing. Continue to participate in auditory-verbal therapy services as scheduled.





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Med-El	www.medel.com	888-633-3524	Sonnet 2	Rondo 3	12	4	10 years	5 years	1.5 and 3.0 Tesla without surgery or magnet removal	With waterproof sleeve	>
Cochlear	www.cochlear.com	800-523-5798	Cochlear Nucleus 8	KANSO 2	22	4	10 years	5 years	1.5 and 3.0 Tesla without surgery or magnet removal	Aqua +	2
Advanced Bionics	www.advancedbionics.com	877-829-0026	Sky CI M		16	m	10 years	5 years	1.5 and 3.0 Tesla without surgery or magnet removal	Waterproof battery	Š
Manufacturer	Website	Customer Service	Sound Processors		Number of Channels	Number of Program Slots	Internal Device Warranty	Sound Processor Warranty	MRI Compatibility	Water-Wear Options	Streaming, FM Compatible,

Section 8 Yearly Maintenance Appointments

After your child gets the CI, the following appointments are needed in addition to regular Auditory-Verbal Therapy. These appointments help your child to make the expected progress.

APPOINTMENTS INCLUDE:

I. Audiology

To make an appointment, call: Texas Medical Center: 832-822-3249 The Woodlands: 936-267-7350 West Campus: 832-227-1030 Sugar Land: 281-494-7010

2. ENT Surgeon

Physician requests are necessary to get replacement CI equipment and batteries. Your CI surgeon must see your child in order to write prescriptions. Your child should see the ENT at least I time every 2 years. See your ENT sooner if requested by your physician or if there is a medical concern.

To make an appointment, call: Texas Medical Center: 832-822-3250 The Woodlands: 936-267-7400 West Campus: 832-227-1420 Bellaire: 713-839-0164 Eagle Springs: 281-666-5006 Sugar Land: 281-494-7010 Texas Hearing Institute: 281-661-4858

3. Speech and Language Re-evaluation

To ensure your child is using the CI as expected, a speech and language re-evaluation is recommended 6 months after CI activation.

- When you call, please tell the scheduler this is an evaluation specific to cochlear implants.
- You will need a physician's referral.
- To make an appointment call: The Texas Medical Center Campus: 832-822-3280 or West Campus: 832-227-1900

Section 9 **Resources**

INFORMATION ABOUT HEARING LOSS IN CHILDREN

- Alexander Graham Bell Association for the Deaf and Hard of Hearing www.agbell.org
- All Ears: 281-615-5475 www.allearscenter.org
- Hearing First, Powering Potential www.hearingfirst.org
- Hearing Loss Association of America www.hearingloss.org
- John Tracy Clinic www.jtc.org
- Laurent Clerc National Deaf Education Center, Gallaudet University
 www.clerccenter.gallaudet.edu
- Navigating a Forest of Information; One tree at a time www.gallaudet.edu/documents/clerc/ci.pdf
- Success for Children with Hearing Loss www.successforkidswithhearingloss.com
- Texas Hands and Voices www.txhandsandvoices.org
- Texas Hearing Institute and the Melinda Webb School in Houston: 713-523-3633 www.texashearing.org
- The Listening Room Hearing Journey www.hearingjourney.com
- UpWords: 832-227-1828
 www.texaschildrens.org/upwords-program

Section 10 Glossary

Activation: The appointment when the cochlear implant (CI) processor is "turned on" for the first time. Your child will not have improvement in hearing between the surgery and this appointment as the implant is not receiving information.

Auditory Brainstem Response (ABR): A hearing test that measures brainwaves in response to sounds. The test does not require the patient to respond. An ABR is used for very young children and infants.

Auditory Steady State Response Test (ASSR): A hearing test similar to an ABR that gives frequencyspecific information. This test gives more information for diagnosing a profound hearing loss.

Audiogram: A graph showing the range of a person's hearing. Hearing threshold levels (in decibels) are plotted on a chart to show the softest sound a person can detect at frequencies ranging from 250 Hz (lowest pitch) and 8,000 Hz (highest pitch).

Audiologist: A person trained to identify and measure hearing impairments and related disorders using a variety of tests and procedures, and recommend different rehabilitation options.

Auditory Nerve: The eighth cranial nerve that connects the inner ear to the brainstem.

Auditory Neuropathy Spectrum Disorder (ANSD): A type of hearing loss that occurs when there is damage in the inner hair cells in the cochlea, in the connections between the inner hair cells and the auditory nerve, or the auditory nerve itself. The ear can hear sounds but due to a defect along the pathway from the ear to the brain, the sounds are disorganized. The child may be able to hear, but speech is typically hard to understand. The effect ANSD has on a child's ability to understand and develop speech varies from mild to severe. It is usually diagnosed when ABR testing shows a significant hearing loss, but other tests show the cochlea works as it should.

Auditory Perception: Ability to identify, interpret and attach meaning to sound.

Auditory-Verbal Therapy (AVT): This form of therapy helps children develop speech and language skills through listening and using spoken language. AVT is parent-centered meaning that parents or caregivers are trained to become the child's primary language model. AVT concentrates on hearing as the primary means of communication (rather than a visual language such as sign language.) Children must use hearing technology that provides access to all of the frequencies of speech for AVT.

Auditory-Verbal Therapist (AVT): A health care professional who specializes in teaching children with hearing loss to learn how to listen and talk.

Aural Rehabilitation: Techniques used for people with hearing loss to help improve their ability to understand speech through listening with or without lip reading.

Balance: The body system that helps a person know where their body is in space and maintain stability. Normal balance depends on information from the labyrinth, part of the inner ear, as well as information from other senses such as sight, touch and muscle movements.

Bilateral Implantation: Cls in both ears.

Bimodal Rehabilitation: A CI in one ear and hearing aid in the other ear.

Channel: CI electrodes have multiple channels, or pairs of electrodes, that help stimulate the auditory nerve at different positions inside the cochlea. The channels are numbered in order. Not all channels are used at the same time and may be turned off individually for various reasons by the CI Audiologist.

Cochlea: The snail-shaped part of the inner ear. Nerve endings inside the cochlea turn the sounds you hear into messages that go to the brain.

Cochlear Implant (CI): A medical device implanted in the inner ear. The implant bypasses the damaged part of the inner ear and directly stimulates the auditory nerve.

Cochlear Implant Candidate: Someone for whom a Cl is a good option. Candidacy is determined after evaluation by the Cl Team. The decision to implant or not to implant is based on the series of test results from different specialists (see candidacy evaluations handout).

Coding Strategy: A programming system inside the CI. The program uses a series of calculations to measure and analyze the sound entering the microphone on the CI. It determines which CI electrodes to stimulate and how they should be stimulated to best represent the original sound. The program then makes a code that tells the CI which channel to stimulate, when to stimulate it, and how loud the stimulation should be. This happens many thousands of times per second.

Coil: The part of the CI that is placed on the head. It uses a magnet to attach to the part of the CI inside the body. The coil sends the coded information from the speech processor to the internal part. Radio signals send the message through the skin. The radio signals produced by the coil are a special frequency that only the implant receiver understands. The coil can also be called a "transmitter" or "headpiece."

Coil Cable: The cable or wire that connects the coil to the speech processor.

Communication Mode: The way a person understands and uses language. For people with hearing impairment, the communication mode may be through spoken language, sign language, or a combination of both.

Conductive Hearing Loss: Hearing loss caused by dysfunction of the outer or middle ear. Examples include ear infections and problems with the outer and/or middle ear structures.

Congenital: Present at birth.

Decibel: The unit of measurement that describes the intensity, or loudness, of a sound.

Detection: An auditory response that indicates a sound was heard. Detection is simply sensing the presence or absence of a sound.

Discrimination: The ability to tell the difference between two sounds.

Ear Hook: The part of the CI that hangs over the top of the outer ear to hold the speech processor on the ear. It may contain the microphone depending on the CI manufacturer.

Electrode Array: The part of the CI that is surgically placed in the cochlea. The array contains individual electrode contacts that provide electrical stimulation.

Enlarged Vestibular Aqueduct (EVA): An enlarged space in the vestibular portion of the inner ear that is just past the cochlea. EVA is a cause of hearing loss and balance problems in some people. Hearing loss associated with EVA usually becomes worse over time.

Frequency: Measured in Hertz (Hz), frequency is the "pitch" of a sound from low (bass) to high (treble).

Hair Cells: Tiny cells inside the cochlea with hair (or cilia) at the top. These hair cells sense the pitch and intensity of sound waves that travel through the fluid of the cochlea. In most instances of hearing loss, the hair cells or part of the hearing system associated with them, do not function properly.

Hearing: The ability of the ears to take in and to notice sounds.

Hearing Aid Trial: A period of time a patient wears hearing aids. A trial is completed to determine if the hearing aids help a person detect sound. During a trial, the hearing aids must be worn all waking hours.

Hearing Loss: The loss of ability to take in or notice sounds. People with hearing loss have reduced sensitivity to sound in comparison to normal hearing. There are many types and causes of hearing loss. Hearing loss ranges from slight to profound.

Inner Ear: The part of the ear that contains both the cochlea (the organ of hearing) and the vestibular system (the organ of balance).

Labyrinth: Same as inner ear.

Localization: The ability to determine the direction of a sound source.

Magnet (CI): A CI device has 2 magnets. One magnet is in the external coil. The other is in the internal device. The magnet strength of the coil can be adjusted. This ensures the two are connected firmly so the internal device receives the programming information from the external processor.

Map: The program stored in the speech processor. It tells the CI how to process sound on each channel. Programming maximizes sound loudness while ensuring comfort. Each person uses different programming in their CI. Maps need to change over time as the brain adjusts to using the CI.

Mapping Sessions: An appointment with a CI audiologist. The purpose of this appointment is to program the CI and assess the responses to any program changes.

Middle Ear: The space behind the eardrum that holds three tiny hearing bones called ossicles. The ossicles send sound vibrations from the ear canal towards the inner ear.

Ossification: New bony growth that occurs inside the cochlea after an infection. It is important to have hearing tests soon after having meningitis or head trauma to identify severe hearing loss. A Cl can be used to keep hearing loss from becoming worse. It is important to act quickly as ossification can turn the cochlea to bone and a Cl cannot be used.

Otoacoustic Emissions (OAE): Low-intensity sounds produced by the inner ear hair cells. OAEs can be quickly measured with a microphone placed in the ear canal. The test for OAEs helps determine the type of hearing loss. Examples include sensory hearing loss or neural hearing loss.

Ototoxic Drugs: Drugs that can damage the hearing and balance structures in the inner ear.

Outer Ear: External part of the ear that we see. It includes the pinna, or auricle and the ear canal.

Progressive Hearing Loss: A hearing loss that gets worse over time.

Post-lingual Hearing Loss: Hearing loss starting after the child learned to speak.

Pre-lingual Hearing Loss: Hearing loss starting very early, before the child learned to speak.

Receiver/Stimulator: The internal part of the CI attached to the electrode array. This part is placed in the ear during surgery.

Residual Hearing: The ability to hear some sounds even though a hearing loss is present.

Round Window Membrane: Membrane separating the middle ear and inner ear.

Sensorineural Hearing Loss: Hearing loss caused by damage to the sensory cells of the inner ear and/or auditory nerve.

Sign Language: A true language with its own rules of grammar and syntax. It consists of hand shapes, facial expressions, and movements used as a form of communication. Examples of sign language are American Sign Language (ASL) or Signing Exact English (SEE).

Speech Processor: The external portion of a CI. The speech processor collects sound from the environment.

Spoken Language: A true language with its own vocabulary and rules for grammar and syntax. Examples of spoken language are English, Spanish and Vietnamese.

Telemetry: A feature of the CI that allows for testing of the internal part of the implant.

Threshold: The level at which a person hears a sound, 50% of the time.

Total Communication: Uses a variety of gestures, language of signs, speech reading, finger spelling, reading, writing and residual hearing to help with communication. It is not a true language like sign language or spoken language.

Vestibule: One of the organs of the inner ear responsible for balance.



Visit us online at Texaschildrens.org/ENT

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