

# **HEARING LOSS REHABILITATION FOR ACOUSTIC NEUROMA PATIENTS**

In the modern era of acoustic neuroma treatment, numerous hurdles have been cleared. In the early part of the 20th century, patients were at high risk of death from either the surgical treatment of these tumors or from the tumors themselves. After reducing the risk of death or mortality to <1%, the next goal was to avoid neurovascular injuries that could cause strokes or other significant morbidities, including facial palsy. Subsequently, research and increased expertise have improved the preservation of the facial nerve. In large volume centers, preservation of facial nerve function at normal or near normal levels is in the 90%+ range, depending on the size of tumor and treatment paradigm chosen (whether radiation or surgery).

One of the last great obstacles in the management of acoustic neuromas is hearing preservation and/or rehabilitation after hearing loss. Hearing loss is the most common disability in patients with acoustic neuromas and affects all age groups. Hearing loss is a symptom that affects one's quality of life, whether one chooses observation or monitoring of a tumor with frequent (serial) MRI scans, radiation therapy, or microsurgical excision. It can vary from no or mild hearing loss to complete deafness (also known as profound hearing loss or single-sided deafness – SSD).

Hearing loss can disrupt one's social and vocational life; it can contribute to depression and a sense of isolation. Hearing rehabilitation, through the use of hearing aids and assistive listening devices, can enhance one's ability to communicate with others (by phone or in person) and significantly improves a patient's quality of life.

## **HEARING REHABILITATION TAILORED TO THE INDIVIDUAL PATIENT**

As the degree of hearing loss from an acoustic neuroma varies among patients, the type of hearing aid best suited for each individual depends on many factors, especially the hearing level of both the ear affected by the tumor and the unaffected ear. Surgical removal of an acoustic neuroma can affect any remaining hearing, possibly causing partial or complete hearing loss. Surgery does not affect the hearing in a patient's good ear, and most patients can still hear adequately in most situations. However, with single-sided deafness, it can be hard to locate sounds as well as hear in situations with much background noise.

For small tumors, it may be possible for the surgeon to preserve the hearing in the affected ear (e.g., via a middle fossa or retrosigmoid approach). For larger tumors, the likelihood of preserving hearing is lower. If the surgeon has been able to preserve some hearing in the affected ear, it may be feasible to try a conventional type of hearing aid. If the unaffected ear has some hearing loss, a conventional hearing aid in that ear may also be helpful.

In the case of total hearing loss/deafness in one ear, called unilateral profound hearing loss or single-sided deafness (SSD), a conventional hearing aid will not be helpful. In the case of bilateral hearing loss, bilateral amplification is recommended to allow better understanding in noise and allow directionality of sound. This issue is faced by patients with bilateral acoustic neuromas, as is found in Neurofibromatosis Type II (NF-2), which accounts for about 5% of all acoustic neuromas. Although many options exist for both unilateral and bilateral hearing loss in patients with acoustic neuromas, this booklet will give a concise overview of the options available. Please discuss further with your neurotologist, or find an audiologist near you at <http://www.audiology.org>. (An audiologist is a professional who assesses and manages hearing and balance related disorders.)

### **TYPES OF HEARING LOSS**

There are four types of hearing impairment. First, **conductive hearing loss** occurs when sound is not conducted efficiently through the outer and middle ears. It usually involves a reduction in sound level; however, if sounds are amplified, speech understanding and hearing clarity are preserved.

Second, **sensorineural hearing loss** occurs when there is damage to the inner ear (cochlea) or nerve pathways from the inner ear to the brain. It involves a reduction in sound level, speech understanding and hearing clarity. Acoustic neuromas typically cause this type of hearing loss.

Third, **mixed hearing loss** occurs when a sensorineural hearing loss occurs in combination with a conductive hearing loss.

Lastly, **auditory processing disorders (APD)** occur when auditory portions of the brain are affected by aging, injury (trauma), dementias, tumors, or unknown causes. These disorders can disrupt sound localization, lateralization, discrimination, pattern recognition and the temporal aspects of sounds as well as the ability to deal with degraded and competing acoustic signals.

### **CONVENTIONAL HEARING AIDS PROVIDE WIDE RANGE OF FEATURES**

A conventional hearing aid can be a good choice if some hearing is preserved in the ear affected by the tumor. Several types and sizes of conventional hearing aids are available. The correct type depends on many factors; cost is important because most hearing aids are usually not covered by insurance. Although insurance companies may pay for the cost of audiologic tests, they rarely cover the cost of hearing aids. Other options available to qualifying patients include the Sertoma Hearing Aid Bank which provides hearing aids for individuals 65 years and older, based on income. To find a club near you visit <http://www.sertoma.org>. Also, the Lions Club (The Lions Affordable Hearing Aid Project) provides hearing aids for low-income individuals. To find a club near you visit <http://www.lionsclubs.org>. The patient should discuss in detail which of these options is best with an audiologist and the treating physician, since each case is highly individualized.

Patients may have concerns about cost and appearance of conventional hearing aids. However, these devices are used widely to help with hearing loss in patients with and without acoustic neuromas with excellent results. Many states do offer a 30-day return policy on the hearing aid sold; patients are expected to pay for the fitting fee and consultation. If the patient decides to return the hearing aid within the 30-day trial period, he/she will be refunded all of the money less a professional fee (typically around \$150 to \$200).

Hearing aids can vary in cost from hundreds to thousands of dollars, depending on size, complexity and extra features. When choosing a hearing aid, the patient should work with a reputable facility; service and a proper fit are as important as the device itself. Each device has benefits and limitations. It can take repeated fittings, fine-tuning of the device and patience to achieve optimal results.

The measure of effectiveness varies with each individual. For instance, even if one has poor speech discrimination (also known as the word recognition score), the hearing aid may enhance communication in combination with visual cues (e.g., improved lip reading) or improve awareness of sounds in the environment for greater safety (e.g., doorbell, car horn, police siren, fire alarm, dog barking, etc.).

#### **TYPES OF CONVENTIONAL HEARING AIDS**

Conventional hearing aids come in two basic types: analog and digital. Analog hearing aids, the oldest type, have a microphone that picks up sound and converts the sound into small electrical signals. These signals are then made louder, or amplified, and sent to the speaker on the hearing aid. Programmable analog aids can amplify quiet sounds until they are loud enough to be heard; these give less amplification to sounds that are already loud, protecting the user against uncomfortably loud sound levels. Analog hearing aids are simple to use and often inexpensive. However, they have been largely replaced by aids that use digital signal processing.

Digital aids work in a different way than analog aids. Digital hearing aids take the signal from the microphone and convert it into 'bits' of data that can be manipulated by a tiny microprocessor, or computer, in the hearing aid. This technology makes it possible to finely adjust sounds to suit individual needs and different listening conditions (e.g., quiet environments versus louder settings such as restaurants and large group gatherings). Digital processing requires less space, which allows manufacturers to include more programmable features in a smaller package.

#### **AVAILABLE STYLES OF CONVENTIONAL HEARING AIDS**

Both analog and digital hearing aids vary in size (behind-the-ear, in-the-ear, in-the-canal and completely-in-the-canal) and in circuitry features (see Table 1). The cost of a hearing aid generally increases with a decrease in size and an increase in advanced features.

### ***Behind-the-ear hearing aid (BTE)***

**Overview:** The BTE style hearing aid is housed in a small curved case which fits behind the ear and is attached to a custom earpiece or earmold, which is molded to the shape of your outer ear. Some BTE models do not use a custom earmold; instead, the rubber tubing is inserted directly into the ear. The case is typically flesh colored but can be obtained in many colorful patterns (for children) and shaded colors (to blend in with the patient's hair).

BTE hearing aids are also available with an "open fitting," which is a slim tube that goes down into the ear canal with an open dome or highly vented ear piece at the end. This allows for a reduction in the occlusion effect (hollow or echo sounding voice) and leads to a more natural sounding voice for the patient. They are less noticeable than hearing aids with earmolds and are typically more appropriate for individuals with higher pitched hearing loss; however, they may not be suitable for severe to profound hearing losses.

Receiver-in-the-canal (RIC) hearing aids refer to BTE aids with the loudspeaker or receiver component inserted in the ear canal instead of behind the ear. This modification allows for a smaller hearing aid; the receiver can be housed in a number of different ways: a dome, micro mold, or custom earmold. This style of BTE is cosmetically appealing; however, like the open fit models, it may not be appropriate for severe to profound losses.

#### **Benefits:**

- Typically, a BTE hearing aid is the most powerful hearing aid style available; it may be the best option for persons with severe to profound hearing loss.
- Non-occluding earmolds may be used with BTE hearing aids, if a medical condition exists that prohibits the use of in-the-ear styles, or if the patient reports a "plugged" sensation when wearing other hearing aid styles.
- Larger battery sizes used in BTE aids may be easier to handle than smaller styles for those with limited manual dexterity or vision deficits.
- BTE aids may be the most appropriate choice for young children, as only the earmold needs to be replaced periodically as the child grows and the ear changes in dimension.
- Special features such as FM, direct auditory input and directional microphone technology are routinely available with most BTE styles and models.
- Telecoil circuitry (which allows the hearing aid to pick up magnetic signals in telephones and assistive listening devices) is often more powerful than is seen with in-the-ear hearing aids.

#### **Limitations:**

- Fit and appearance may be less acceptable to some patients, although modern BTE designs have become smaller and less noticeable. This style is the most susceptible to moisture damage from sweat.

### ***Custom hearing aids***

**Overview:** No two ear canals (the tunnel running from the outer ear to the ear drum) are alike. Custom hearing aids provide patients with hearing technology molded to his/her ear canal shape and size. An earmold impression is the first step in the design phase of a custom hearing aid. An audiologist will take the mold of the patient's ear and help determine the most appropriate size and features for the hearing aid based on the individual's hearing loss.

### ***In-the-ear hearing aid (ITE)***

**Overview:** The ITE style hearing aid fits directly into the external ear. The circuitry is housed primarily in the concha (external) portion of the ear. ITE aids have their working parts in a custom made shell so the whole aid fits into the ear. This style has typically been considered to be more cosmetically appealing. (However, modern BTE hearing aids have become smaller and are often less noticeable than some ITE hearing aids.)

#### **Benefits:**

- More secure fit and easier insertion and removal than with BTEs.
- Less wind noise in the smaller styles than with BTEs.
- Directional microphone technology available for most styles.
- All components are integrated into a one-piece shell, which may be easier to handle and operate than for BTE styles.

#### **Limitations:**

- Tend to need repairing more often than BTE aids.
- Some ITE aids can be seen from the side.
- Battery size in ITE aids is typically smaller than BTE aids; this may be difficult for those with manual dexterity problems and/or visual deficits.
- This style is not as powerful as BTE hearing aids and may not be suitable for severe to profound hearing losses.

### ***In-the-canal (ITC) and Completely-in-the-canal (CIC) hearing aids***

**Overview:** Due to the miniaturization of the component parts (including the microphone, receiver and battery), it is possible to make hearing aids small enough to fill only a portion of the concha (ITC) or fit deeply into the ear canal (CIC). Along with ITE aids, these styles have characteristically been considered to be more modern and cosmetically appealing.

#### **Benefits:**

- Improved cosmetic benefits due to smaller styles; the smallest ITC and CIC aids fit right inside or deep in the ear canal and cannot usually be seen.
- Less wind noise than with BTEs.
- Deep microphone and receiver placement with CICs may result in increased battery life (e.g., reduced power consumption) and high frequency amplification compared with other styles.
- Directional microphone technology available for most ITC styles.
- Some people find them easier to put in and take out than BTE styles.

**Limitations:**

- If a patient has severe hearing loss or small ear canals, this type of aid may not suit him/her.
- These aids tend to need cleaning and repairing more often than BTE aids; slightly less durable than BTE aids.
- Very small models, though less noticeable, may be more difficult to operate, particularly for patients with dexterity or vision problems.
- They may not be compatible with assistive listening devices such as a loop system. (A loop helps one hear sound from a television, stereo, or radio. A loop may be set up with a microphone to help hear conversations in noisy places or a show in a theater.)
- Directional microphone technology is not generally available for CICs.
- Some limitations with fitting range for severe to profound hearing losses.

**Table 1: Conventional Hearing Aids: Styles, Benefits, Limitations**

| Type of Aid  | Benefits  | Limitations  |
|--|---|--|
| <p><b>Behind-the-ear (BTE)</b></p>   | <ul style="list-style-type: none"> <li>• Fits a wide range of losses</li> <li>• More room for advanced features</li> <li>• Appropriate option for severe to profound hearing loss</li> <li>• More opportunities for non-occluding earmolds which provide more natural sound</li> <li>• Often more durable</li> <li>• Larger batteries offer longer life</li> <li>• More flexibility with changing hearing losses</li> <li>• Appropriate for all ages</li> </ul> | <ul style="list-style-type: none"> <li>• May be large in size</li> <li>• May be more noticeable than smaller styles, depending on hairstyle, etc.</li> <li>• Susceptible to moisture damage from sweat</li> </ul>  |
| <p><b><u>Custom Hearing Aids</u></b><br/> <b>In-the-ear (ITE)</b><br/> <b>In-the-canal (ITC)</b><br/> <b>Completely-in-the-canal (CIC)</b></p> | <ul style="list-style-type: none"> <li>• Microphone is located in the ear, offering better directionality</li> <li>• Comfortable, custom fit</li> <li>• Large enough for user controls (volume, etc.)</li> <li>• Cosmetics</li> <li>• Decreased wind noise</li> </ul>   | <ul style="list-style-type: none"> <li>• Feedback is more likely than with a BTE</li> <li>• Slightly less durable than BTE</li> <li>• More maintenance and cleaning required</li> <li>• Limited advanced features (depending on size)</li> <li>• Not suitable for severe to profound losses</li> <li>• Shorter battery life</li> <li>• May not be compatible with direct audio input systems (e.g., loop systems)</li> <li>• May not offer telecoil</li> <li>• Increased occlusion without deep insertion depth</li> </ul> |

## **SPECIALIZED HEARING DEVICES**

If a conventional hearing aid is not feasible, such as in unilateral deafness, then other types of specialized hearing devices are available. These options include the CROS (contralateral routing of signal) and BiCROS aids, referring to whether one (CROS) or both ears (BiCROS) have significant hearing loss warranting rehabilitation. Typically, these two options are used in patients with complete hearing loss in one ear, such as in patients with acoustic neuromas.

With the CROS hearing aid, a microphone worn behind-the-ear is placed on the ear with the complete hearing loss. Sound signals presented to the deaf ear are picked up by the microphone and transmitted to a behind-the-ear (BTE) aid in the unaffected ear. Initially, the transmission was done across a wired system involving a headband. More recently, the signal is sent via a wireless system using radio waves, which is more cosmetically appealing. Often, a limitation with the CROS system is the need to wear a receiving device in the unaffected ear.

BiCROS devices, which are suitable if some hearing loss exists in the unaffected ear, amplify sound from both sides and feed it into the ear with hearing. By localizing the sound source and ensuring that one does not miss sounds on the deaf side, this technology helps to restore a sense of hearing in the deaf ear (although the result is not true bilateral hearing because all the sound is sent to only one ear).

### ***Phonak CROS System***

The new Phonak CROS is the smallest and arguably most stylish wireless CROS/BiCROS option available. Phonak CROS consists of only two parts – a transmitter microphone for the ear that cannot benefit from a hearing instrument, and a receiver hearing instrument on the better hearing ear. This means sound from your poorer hearing side is wirelessly sent to your better ear.

### **CROS:**

- A small transmitter, placed behind or inside the ear that cannot benefit from a hearing aid, picks up sound and transfers it wirelessly to a receiver on the normal ear. This allows hearing even when sounds come from the non-hearing side.



### **BiCROS:**

- A small transmitter, placed behind or inside the ear that cannot benefit from a hearing aid, picks up sound and transfers it wirelessly to your ear fitted with a hearing aid. At the same time this hearing aid amplifies sounds coming directly to this ear. This improves hearing, understanding and communication capabilities.

**Accessory Options:**

- **Phonak ComPilot** – provides easy wireless access to TVs, MP3 players and phones to name a few.
- **Phonak RemoteMic** – lightweight wireless microphone combined with ComPilot transfers partner’s voice conversation directly to the hearing aids over a distance of up to 60 feet.
- **Phonak PilotOne** – focuses on the essentials when it comes to discreet control, simplicity and user-friendly ergonomic design.
- **myPilot** – a remote control with a color LCD display provides users access to a multitude of functions, including status information.

For more information go to [www.phonak-us.com](http://www.phonak-us.com).

**BONE CONDUCTION HEARING DEVICES**

Currently, there are four bone conduction hearing devices available in the U.S. for acoustic neuroma patients with unilateral hearing loss—the Cochlear™ Baha® BP100 (Cochlear Americas), the Ponto Pro (Oticon Medical), TransEar® (Ear Technology Corporation) and SoundBite (Sonitus Medical). The Baha BP100 and the Ponto Pro are bone-anchored hearing devices requiring surgery. Baha hearing devices have been available for more than 10 years in the U.S., and the BP100 replaced the Baha Divino, which is no longer manufactured. The Ponto Pro is made by Oticon, which is a well known manufacturer of air-conduction hearing devices. The TransEar is a bone conduction hearing device that is similar in appearance to a conventional hearing aid, but without the need for surgery. The SoundBite is a dental prosthetic that became available in 2012.

**Bone Conduction – How It Works**

Each of the four devices above transmits sound through bone conduction. Bone conduction hearing is a natural way to hear. For example, whenever someone brushes their teeth and hears the sound of brushing, that sound is conducted through bone to the inner ear. Since the inner ear is encased in the bone of the skull, the inner ear picks up sound when the skull vibrates. Sound travels through bone at nearly the same speed it travels through air. One difference between sounds traveling through air versus bone is that high-frequency sounds are attenuated, or “absorbed,” when traveling through bone. Each of the bone conduction devices is able to amplify or “boost” specific frequencies that would normally be attenuated through bone conduction. In this way, hearing is made to sound even more natural.

Bone conduction hearing devices are an option for those patients who have no hearing in one ear (e.g., single-sided deafness) and normal hearing in the opposite ear. The bone conduction device is worn on the deaf side. In this way, sound is picked up by the bone conduction device and then transmitted to the inner ear of the other, normal hearing ear. Since the hearing device is on the deaf side, the perception is that hearing is coming from the side with no hearing. However, hearing in the deaf ear is not restored; the hearing loss is rehabilitated.



For those with no hearing in one ear *and* with a moderate hearing loss in the other hearing ear, a special type of bone conduction hearing device is an option. This option is known as the Cochlear Baha Intenso, a slightly larger and more powerful device than the BP100. In this case, the patient is fitted on the deaf side with an Intenso and the opposite side with a hearing aid. The more powerful Intenso is necessary to adequately amplify sound to the opposite side.

Each of the bone conduction devices is helpful for two of the three hearing difficulties that patients experience with single-sided deafness—inability to hear on the deaf side, difficulty hearing when there is background noise, and an inability to localize the direction of sound. Most patients that use any one of the bone conduction hearing solutions will report that listening on the deaf side is improved considerably, while hearing when there is background noise is improved, but the difficulty is not eliminated.

There are some anecdotal reports of patients able to localize certain sounds with any one of the bone conduction devices, although this is not common. Sound localization is dependent on two, functioning ears. If the sound source is to one side of a person, the sound is delayed as it enters the ear compared to the other side. The brain uses this sound delay as a means to localize sound. Since current treatments for single-sided deafness use only one functioning ear, sound localization is not routinely reported. For those who can localize certain sounds with their bone conduction device, sound localization may occur as a result of the slight time delay for sound to be transmitted through the skull.

***Bone-Anchored Hearing Device - Baha® - Cochlear Americas***

Now in addition to the CROS and BiCROS aid options, there are several bone-anchored hearing devices that are available. After being used in Europe for several decades, the popularity of this device has grown in the U.S. during the past 10 years, particularly as most insurance companies and Medicare cover its use.



**Baha BP100**

**Surgery:**

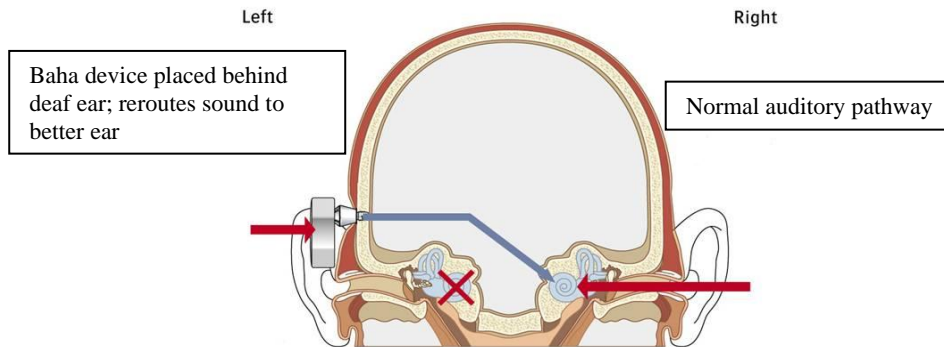
The Baha is surgically implanted into the bone of the skull, about one inch behind the ear canal. The implantation is performed under local or general anesthesia and takes about one hour. The implanted portion of the device is a titanium fixture that is surgically attached to the skull behind the deaf ear. An abutment or “snap” is screwed into the titanium fixture. The abutment sits above the level of skin so the sound processor can be snapped onto the abutment when the patient wishes to use the device. This system was designed so that if the abutment is changed in the

future, the abutment can be unscrewed and replaced in the physician's office without the need for further surgery.

Several weeks are allowed to pass after surgery to allow for bone growth (known as osseointegration) to occur into the fixture. Osseointegration allows the fixture to be firmly attached to the skull and results in the direct transfer of sound to the skull. Both the fixture and abutment are MRI safe. The processor comes in different colors to match the patient's hair color. (Occasionally, the Baha is implanted during acoustic neuroma surgery.)

**How the Baha works:**

The Baha processor works via bone conduction; the device will bypass the normal sound conduction anatomy. (In normal hearing, sound waves go through the ear canal, hit the ear drum, and move the three ear bones in the middle ear, which ultimately displaces the inner ear fluid. This movement causes hair cells and nerve fibers of the inner ear to discharge, sending a signal to the brain that allows us to hear sound.) The Baha processor and titanium screw stimulate the inner ear (cochlea) by conducting sound vibration through the bone. In patients with single-sided or unilateral deafness, the Baha is placed behind the deaf ear and stimulates the other (hearing ear's) cochlea. New programmable sound processors offer automatic, optimum sound quality for Baha users.



*Illustration from Cochlear Americas*

The BP100 has a Europin, which is a universal connection for FM and phone adaptors. The Divino required proprietary adaptors for these applications. The BP100 has status indicators with beeps and LEDs. There are multiple programs that can be set for various listening situations. The programs are controlled with a single button on top of the sound processor. Volume is controlled with up and down buttons alongside the program button.

The BP100 has multiple channels that the audiologist can tune depending on the thickness of the person's skull and hearing capabilities of the opposite ear. The Divino did not allow such fine-tuning.

For more information go to [www.cochlear.com](http://www.cochlear.com).

### ***Ponto Pro - Oticon Medical***

The Pronto Pro functions in the same manner as the Baha for single-sided deafness patients; the device reroutes sound from the deaf ear to the hearing ear via bone conduction. Similar to the Baha device, the Ponto Pro sound processor also sits on the skin and attaches to an abutment that is adhered to a titanium fixture in the skull.

The Ponto Pro is shaped differently than the BP100 in that the Ponto Pro has a slight curve to its design. The program button is on the side. Features of the Ponto Pro include multiple programming capabilities, a self-learning volume control and a Europin connector. In addition, the device is water-resistant. The Ponto Pro fits on the Cochlear Baha abutment if the abutment was placed before November 2009. In this way, someone may upgrade from the Divino to the Ponto Pro without difficulty. Oticon released an additional, more powerful device similar to the Intenso in the Spring of 2011. The device is digital and programmable.



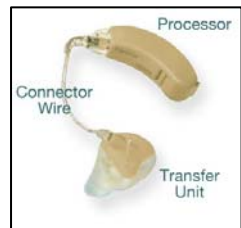
For more information go to [www.oticon.com](http://www.oticon.com).

### ***TransEar® - Ear Technology Corporation***

TransEar is a bone conduction hearing aid designed for individuals with unilateral deafness and looks like a conventional behind-the-ear hearing aid but does not require surgery.

#### **How the TransEar works:**

TransEar relies on bone conduction to transmit the sound to the better ear, without the need for a surgically-implanted device in the skull bone. The TransEar consists of a speech processor that rests behind the ear and is connected through a thin tube to an ear mold that fits in the ear canal. One of the unique features of the TransEar compared to a conventional hearing aid is that the mold of the TransEar fits very deeply inside the ear canal. In this way, the mold can directly stimulate the bone underneath the thin skin of the ear canal. Using bone conduction, the oscillator conducts vibrations through the skull from the deaf side to the working cochlea on the other side.



The current TransEar device, the 380-HF, became available in October 2008. The 380-HF consists of a high frequency bone vibrator, which has its peak energy at 2,100-2,300 Hz. As a result of this peak energy, the 380-HF is able to deliver better high frequency gain compared to its predecessor, the 270, whose peak energy was between 650-700 Hz. High frequency gain is important, since bone will attenuate or “slow down” sound in the region of 2,000 Hz.

Another new feature of the 380-HF compared to the 270, is that the 380-HF has a four channel digital sound processor with two memories or programs. These two programs are typically set for everyday listening situations and the other program for situations involving background noise. The 380-HF also has digital feedback reduction, which helps prevent feedback when in background noise.

For more information go to [www.transear.com](http://www.transear.com).

### ***SoundBite™ - Sonitus Medical***

SoundBite (Sonitus Medical) is the newest addition to bone-conduction hearing devices and was FDA approved in January 2011. It is a prosthetic device that is a non-surgical and removable hearing solution that relies on bone conduction to imperceptibly transmit sound via the teeth for patients with single-sided deafness.



#### **How the SoundBite works:**

Nearly invisible when worn, the SoundBite system consists of two components: an easy to insert and remove ITM (in-the-mouth) hearing device and a small BTE (behind-the-ear) microphone unit worn on the impaired ear. No dental work or modifications to the teeth are required.

#### **BTE (Behind-the-Ear) Microphone Unit**

- To take advantage of the natural acoustic benefits provided by the patient's own ear, sound in the environment is picked up by a tiny microphone placed within an open fit dome worn in the canal of the impaired ear and connected to the BTE by a small translucent tube. The BTE uses a digital signal processor to process the sound and a wireless chip to transmit the signals to the ITM hearing device, which in turn sends the imperceptible vibrations via the teeth that are near – simultaneously to both cochlea. In this way, sound is rerouted from the impaired ear directly to the good cochlea – bypassing the middle and outer ear entirely – to effectively restore the perception of hearing from the impaired ear. SoundBite Hearing System delivers sound across a broad frequency range of 500 Hz to more than 12 kHz.

#### **ITM (In-the-Mouth) Hearing Device**

- The ITM device is custom made to fit around either the upper left or right back teeth, does not require any alteration of the teeth and is easily inserted and removed by the wearer. It contains electronics, a sealed, flat rechargeable battery, wireless capability that picks up sound transmissions from the BTE and a small actuator that converts those signals into vibrator energy. All of these miniaturized components are hermetically sealed inside a dental grade acrylic that has been safely used for making dental appliances for many years.

For more information go to [www.sonitusmedical.com](http://www.sonitusmedical.com).

***Sophono™ Alpha 1 – Sophono, Inc.***

Alpha 1 is unique to traditional bone-anchored hearing systems as it is an abutment-free bone anchored hearing device, yet offers the same features as other bone-conduction systems.

**How the Alpha 1 works:**

An external processor connects to an implantable magnet (Alpha 1 M), eliminating hair removal around an abutment placement site and complications with cleaning this site. Another option is placement of an external processor on a soft band or headband (Alpha 1 S). Similar to other bone-conduction devices, the Alpha 1 is placed behind the poorer ear, stimulating the other inner ear (hearing ear's) cochlea by conducting sound vibration through the bone.

***Cochlear Implants***

Patients with acoustic neuromas and hearing loss, before or after treatment of their tumors, often ask about cochlear implants. The cochlear implant bypasses the damaged parts of the inner ear and stimulates the nerve endings for hearing in the inner ear. For this reason, a cochlear implant requires an intact and functioning hearing nerve to be successful. However, new information indicates that in certain circumstances the hearing nerve can be stimulated, making this an option for AN patients.

The cochlear implant is a two-part system. There is an outer or external part with a headpiece and, most commonly, a behind-the-ear sound processor. The second part is the internal device that is surgically implanted completely under the skin. Cochlear implant surgery is approximately 2 hours in length, under general anesthesia, and is a same day surgical procedure.

Cochlear implants have been around for about 25 years. Almost 200,000 of them have been surgically implanted worldwide during that time frame.

**Indications for Cochlear Implants:**

Cochlear implants are indicated when the patient has significant hearing loss in both ears. In addition, the ear to be implanted must still have the cochlear nerve and blood supply intact. Unfortunately, this is often not the case in patients with acoustic neuromas because of the size of the tumor or treatment with radiation and/or surgery. However, some patients after tumor removal have some remaining hearing, and then they lose hearing in the other ear. In these patients, placement of a cochlear implant is a reasonable option in one or both ears.

A device related to the cochlear implant is the auditory brainstem implant (ABI). These devices, which are typically used in patients with NF-2, can be placed in one or both ears. The ABI does not require the presence of the cochlear nerve or blood supply. This device requires surgical implantation at the brainstem. The ABI does not work as well as cochlear implants (e.g., the ABI does not typically give one the ability to talk on the telephone); however, it does allow an ability to hear environmental sounds and assist with lip-reading.

## **INSURANCE COVERAGE FOR SPECIALIZED HEARING DEVICES**

Insurance coverage for the above devices varies. For devices that require surgery (e.g., the Baha), some companies will cover the surgery but not the hardware. One should determine ahead of time what will be covered. Insurance companies will likely require a letter of medical necessity from your physician. Cochlear implants are typically covered by insurance or Medicare only when the patient has significant hearing loss in both ears. Medicare started to cover bone-anchored hearing devices in 2006.

## **ASSISTIVE LIVING DEVICES**

Additional options for dealing with hearing loss in patients with acoustic neuromas include Assistive Living Devices (ALDs) (e.g., FM sound systems) that can be found in some public places (e.g., churches, theaters, etc.). Similar to this are products such as TV Ears, that assist AN patients with hearing in the home.

Improved hearing in everyday listening situations is of great importance for patients with acoustic neuromas who strive to communicate with family and friends. Fortunately, numerous hearing amplification options do exist. Conventional hearing aids, implantable devices and bone conduction hearing systems, such as the Baha, Ponto Pro and TransEar, enhance the patient's ability to communicate with others. While preservation of hearing is still a great hurdle in acoustic neuroma management, today's hearing rehabilitation options offer a sense of hope for patients and improve overall quality of life.

## **MODERN HEARING AID TECHNOLOGY PROVIDES ADVANCED FEATURES**

In addition to the basic features of hearing aids, many other features are available in modern digital hearing aids (some for convenience and ease of use) while others are designed to improve speech understanding or listening comfort. Following is a brief description of some of these features and the manufacturers of current hearing aid models that provide them.

### **Wireless Hearing Aid Technology:**

In the past two years, we have seen a lot of new "wireless" hearing aids on the market. The term "wireless" can mean different things for different hearing aids. Some patients use certain wireless features that others do not. Here are some common applications for wireless hearing aids.

1. **Hearing television:** A hearing aid system that allows the hearing aid user to receive the TV signal right into their hearing aids. This is done by a small box that plugs into the TV. It like watching TV with headphones but without the wires.
2. **Hands-free Bluetooth:** This allows the patient to stream wirelessly to your cellphone via a small wireless accessory. This allows for hands-free phone use as well as binaural hearing (hearing related to two ears).

3. **One-on-one communication in noise:** Some of the hearing aid manufacturers have a “mini-microphone” that another person can speak into. The microphone is discreet and can be clipped onto a collar or lapel. This allows that person’s voice to be sent to the hearing aids. The result is that the voice can be heard over and above any noise in the environment. This can be especially useful for conversations in the car or one-on-one at a noisy restaurant.
4. **Remote controls:** This gives the patient control of the hearing aid via a remote rather than pushing the buttons on the hearing aid itself. With a remote control, you have access to hearing aid volume and can change to various programs or settings within the hearing aid.
5. **Hearing aid to hearing aid communication:** Many of today’s wireless hearing aids actually talk to each other. With directional microphone equipped hearing aids, this can mean that the hearing aids will actually adjust themselves to focus in on a speech signal, regardless of where it is coming from.

**Adaptive Feedback Cancellation:** Many of today's hearing aids have an automatic feature that quickly detects acoustic feedback (whistling from the hearing aid sometimes caused by placing one's hand or a phone next to the ear) and cancels it.

**Automatic Gain Control-Output (AGCo):** AGCo, or output compression, puts a "ceiling" on loud sounds and can be adjusted to match the patient's threshold of discomfort by maintaining sounds below this level.

**Automatic Gain Control-Input (AGCi):** AGCi, or input compression, also called wide dynamic range compression (WDRC), compresses speech or other incoming sound signals to fit into the reduced range of the hearing aid user, which provides more gain for soft sounds than for average and more gain for average sounds than for loud. With this feature, many hearing aid users have little need for a volume control.

**Multiple Channels:** The majority of today's hearing aids have multiple channels. Each channel represents a portion of the frequency range important for understanding speech. Gain and compression can be programmed differently in each channel to reflect changes in the patient's hearing across frequencies.

**Digital Noise Reduction:** This feature allows the hearing aid to analyze an incoming signal and differentiate speech from a broadband noise (which presents simultaneously in several channels) and reducing gain if a signal is believed to be noise in a given channel.

**Directional Microphone Technology:** Using special microphones or phase cancellation signal processing, it is possible to configure a hearing aid so that sounds from the side and back of the user are amplified less than sounds originating from the front. Hearing aid users have reported this especially useful when background noise originates from behind the listener, the talker is in front of the listener, the listener is close to the talker and the room has low reverberation.

**Multiple Memories:** A memory is a location to store settings designed for a particular listening situation (for example listening in quiet, listening in noise, or for listening on the telephone). It is common for hearing aids to come with two or three memories that can be switched by using a button. In some digital hearing aids, it happens automatically.

**Telecoils:** The telecoil is a small component in the aid that picks up signals from a loop system or hearing aid compatible telephone. A loop system helps to hear sound from a television, stereo, or radio, or can be set up with a microphone to help hear conversations in noisy places or even in the theater. Telecoils are not available in some smaller models due to space limitations.

**Sound Recover:** Sound recover technology, from Phonak, compresses and shifts high frequency information into an area of greater audibility for the patient, resulting in increased audibility of high frequency sounds.

| <b>Table 2: Manufacturers and Models</b> |                                   |
|--|-----------------------------------|
| <b>Manufacturer*</b>                     | <b>Models*</b>                    |
| <b>Phonak</b>                            | Audeo S, Virto, Naida S, Bolero   |
| <b>Oticon</b>                            | Agil Pro, Acto, Chili             |
| <b>Starkey</b>                           | Wi Series, 3 Series, Tour, Ignite |
| <b>Widex</b>                             | Clear, Mind, Menu                 |
| <b>GN Resound</b>                        | Verso, Alera, Vea, Essence        |
| <b>Siemens</b>                           | Life, Pure, Motion, Nitro         |

**\* Note: Additional manufacturers and models of hearing aids are available. An audiologist will assist the patient in selecting the most appropriate hearing aid(s).**

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## WHAT IS THE ACOUSTIC NEUROMA ASSOCIATION (ANA)?

Acoustic Neuroma Association was founded in Carlisle, Pennsylvania, in 1981 by a recovered patient, Virginia Fickel Ehr. She found no patient information or patient support available when she had surgery for the removal of an acoustic neuroma in 1977. She resolved that future acoustic neuroma patients should have easy-to-read medical material about their condition, and support and comfort from each other. With the help of her physician, she contacted eight other patients and formed the organization.

The association is incorporated and is a 501(c)(3) non-profit organization. The patient-focused, member organization now serves close to 5,000 members, is governed by an all-patient Board of Directors and is operated by a small staff in metropolitan Atlanta, GA.

ANA membership benefits include receipt of a quarterly newsletter, patient information booklets, access to a network of local support groups, access to a list of acoustic neuroma patients willing to talk about their experience throughout the country, our website Member Section and an invitation to a biennial symposium on acoustic neuroma. Our exclusive website Member Section includes published medical journal articles on acoustic neuroma and all of our patient information booklets and newsletters and many symposium presentations. ANA also maintains an interactive website at [www.ANAUSA.org](http://www.ANAUSA.org) with an ANA Discussion Forum.

ANA is patient-founded, patient-focused and patient-funded. ANA recommends treatment from a medical team with substantial acoustic neuroma experience. Although the association cannot recommend specific doctors, medical centers or medical procedures, guidelines for selecting a qualified medical professional can be found at the ANA website, [www.ANAUSA.org](http://www.ANAUSA.org). Now available on our website is a listing of medical resources. The physicians and organizations listed have self-reported data to meet criteria established by ANA for having substantial experience in treating acoustic neuromas. The listings should NOT in any way be construed as an endorsement or recommendation by ANA. It is every individual's responsibility to verify the qualifications, education and experience of any healthcare professional.

|   |  |
|---|--|
|  | <p style="text-align: center;"><b>PIN FOR SINGLE-SIDED DEAFNESS</b></p> <ul style="list-style-type: none"><li>• Designed to be worn on the side of your non-hearing ear.</li><li>• Helps direct the speaker to your hearing ear</li><li>• The pin is approximately 1.75" wide by 2.75" tall and the printing is in red and blue colors. <b>Cost: \$5.00</b></li><li>• Order online at <a href="http://www.ANAUSA.org">www.ANAUSA.org</a>, "ANA Store" link, or by calling ANA National office at 1-877-200-8211.</li></ul> |
|  | <p><b>Remember to specify right or left.</b></p>   |

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