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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 this risk to <1%, the next goal was to avoid neurovascular injuries that could cause strokes or other serious injuries. Subsequently, research and 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.

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 MRI scans, radiation therapy, or surgical excision. It can vary from none or mild hearing loss to complete deafness (also known as profound hearing loss).

Hearing loss can disrupt social and work life; it can contribute to depression and a sense of isolation. Hearing rehabilitation through the use of hearing aids and/or assistive listening devices can enhance one's ability to communicate with others, whether via 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 hear adequately in most situations. However, it can be hard to locate sounds, as well as hear speech in situations with a lot of 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 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 in one ear, called unilateral hearing loss, 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 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. Many options are available for conventional hearing aids and for advanced technology hearing rehabilitation that exist for both unilateral and bilateral hearing loss.

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**, central *auditory processing disorders* (CAPD) occur when auditory portions of the brain are affected by injury, disease, tumor, heredity, or unknown causes. These disorders can disrupt sound localization, lateralization, discrimination, pattern recognition, the temporal aspects of sounds, and 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 or the treatment of the tumor. Several types and sizes of conventional hearing aids are available. The right type depends on many factors; cost is important because most hearing aids are usually not covered by insurance. The patient should discuss in detail which of these options is best with an audiologist (a professional who assesses and manages hearing and balance related disorders), 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. Some insurance companies cover the cost of hearing aids, some provinces have coverage for hearing aids through their health plans. Be sure to ask about 30 day trial periods.

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 a poor word understanding or discrimination, the hearing aid may enhance communication in combination with visual cues (i.e., 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 earphone 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 (i.e. quiet environments versus louder settings such as restaurants and large group gathering). 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 the outer ear. Some BTE models do not use a custom earmold; instead, the plastic 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).

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.
- BTE hearing aids are also available with an 'open fitting,' which is a small, soft earpiece at the tip of the tubing instead of an earmold. They are less noticeable than hearing aids with earmolds but are typically only suitable if hearing loss is mild. They can provide a very natural sound.
- 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. Open fitting BTEs may not be suitable for severe to profound hearing losses.

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 at times are 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; smaller battery size may be difficult to handle for those with manual dexterity problems and/or visual deficits, as can occur with the elderly.
- 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, these styles have characteristically been considered to be more modern and cosmetically appealing.

Benefits:
- Improved cosmetic benefits due to smaller styles (CIC, ITC); the smallest ITC and CIC aids fit right inside the ear canal and cannot usually be seen.
- Deep microphone and receiver placement with CICs may result in increased battery life (i.e., 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 repairing more often 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: Benefits and Limitations of Conventional Hearing Aids

Conventional Hearing Aids: Styles, Benefits, Limitations		
Type of Aid	**Benefits**	**Limitations**
Behind-the-ear (BTE)	•Fits a wide range of losses •More room for advanced feature •Appropriate option for severe-to-profound hearing loss •Non-occluding earmolds provide natural sound •Often less expensive •Very Durable	•May be large in size •May be more noticeable than smaller styles, depending on hair style, etc.
In-the-ear (ITE)	•Microphone is located in the ear •Comfortable, custom fit •Large enough for user controls (volume, etc) •Large enough for advance features (directional microphones)	•Feedback is more likely than with a BTE •Slightly less durable than BTE •More maintenance and cleaning required •May be more noticeable than smaller styles
In-the-canal (ITC) & Completely-in-the-canal (CIC)	•Cosmetics •Microphone location in the ear •Some large enough for user volume control •Comfortable, custom fit •Some provide less "occlusion effect" •Less gain required	•Not suitable for severe losses •No room for directional microphone (CIC) •No room for volume control (some) •Shorter battery life •More expensive •Not compatible with direct audio input systems (i.e. loop systems) (most)

Specialized Hearing Aids

If a conventional hearing aid is not feasible, such as in unilateral deafness, other types of specialized hearing aids are available. These options include the CROS (contralateral routing of signal) and BiCROS aids, referring to whether one (CROS) or both ears (BiCROS) has significant hearing loss. 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 is worn on the ear with the complete hearing loss. Sound signals presented to the deaf ear are picked up by the microphone and transmitted to an aid in the unaffected ear. Initially, the transmission was done across a wired system. More recently, the signal is sent via a wireless system using radio waves. Often, a limitation with the CROS system is the need to wear a receiving device in the unaffected ear. BiCROS aids, 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).

Bone-Anchored Hearing Aid (Baha®)
More recently, CROS and BiCROS aids have become supplanted by the use of the Bone-Anchored Hearing Aid (or Baha®, Cochlear Corporation). After being used in Europe for several decades, the popularity of this device has grown in North America.

Indications for Baha®:
The most common indication for the Baha in patients with acoustic neuromas is for unilateral hearing loss.

Surgery:
The Baha is surgically implanted about 2 inches behind the ear canal. The implantation is performed under local or general anesthesia and takes about 1 hour. The implanted portion of the device, which is made of titanium, undergoes osseointegration (i.e., bone grows around a titanium screw and fixes it in position). After completion of the osseointegration of the device (about 12 weeks), the external hearing aid processor is then fitted; 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; Baha's bypass the normal sound transduction anatomy. (In normal hearing, sound waves go through the ear canal, hit the ear drum, and move the 3 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 unilateral hearing loss, the Baha is placed behind the deaf ear and stimulates the other (hearing ear's) cochlea.

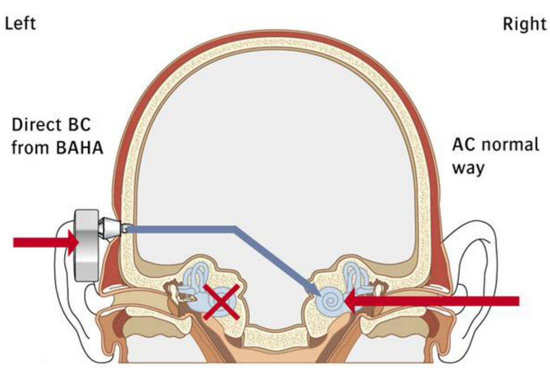

Illustration from Cochlear Americas

Essentially, the Baha device reroutes auditory information sent to the deaf ear to the hearing ear via bone conduction. Although this device won't replace one's hearing completely, it provides a sense of awareness of hearing on the deaf side and some direction-sensing ability. (A common complaint in patients with only one hearing ear is that they can't tell from which direction sounds are coming.) To date, Baha is the most commonly used implantable hearing aid. Others exist but are expensive and not commonly used at this time.

Cochlear Implants
Patients with acoustic neuromas and hearing loss, before or after treatment of their tumors, often ask about cochlear implants. Cochlear implants have been around for about 25 years. Almost 100,000 of them have been surgically implanted worldwide during that time frame.

Indications for Cochlear Implants:
Cochlear implants are typically covered by provincial health plans only when the patient has significant hearing loss in both ears. In addition, the ear to be implanted must still have 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 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 NF2, 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 ability to talk on the telephone); however, they do allow an ability to hear environmental sounds and assist with lip-reading.

TransEar®
One of the newer options available, TransEar® (Ear Technology Corporation), is a bone-conduction hearing aid designed for individuals with unilateral deafness, however TransEar is NOT available in Canada yet.

How the TransEar works:
TransEar looks like a conventional behind-the ear hearing aid, but instead of simply amplifying sound, it relies on bone conduction to transmit the sound to the better ear, without the need for a surgically-implanted device in the skull bone. TransEar is made of three parts: a modified behind-the-ear hearing aid, which is joined by a small connector wire to a transfer unit – an acrylic shell that contains a small oscillator. These components are fit on and in the deaf ear. Using bone conduction, the oscillator conducts vibrations through the skull from the deaf side to the working cochlea on the other side.

Indications for TransEar:
If a patient has a normal ear canal on the deaf side and good hearing on the other side, TransEar may be a good option.

Insurance Coverage for Specialized Hearing Aids:
Insurance coverage for the specialized devices varies. One should determine ahead of time what will be covered. As of 2008 they hope to start to regulatory process within the next year or so.

Assistive Listening Devices
Additional options for dealing with hearing loss in patients with acoustic neuromas include Assistive Listening Devices (ALDs) (e.g., FM sound systems) that can be found in some public places (e.g., churches, theaters, etc.).

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 for patients with hearing loss due to acoustic neuromas. Conventional hearing aids, implantable devices, and bone conduction hearing systems, such as Baha 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.

***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 telephone 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 broad-band noise, 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 theatre. Telecoils are not available in some smaller models due to space limitations.

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WHAT IS THE ACOUSTIC NEUROMA ASSOCIATION OF CANADA?

In early 1983, Virginia Garossino, Velma Campbell and Linda Gray met for the first time. The three Edmonton women had had acoustic neuroma surgery within a year of each other, and had encountered individual sets of difficulties and recoveries. They discovered early in their search that there wasn't a support or information system in place for Canadians dealing with this unique tumor. Along with husbands Dick Garossino and George Campbell, the three agreed to establish a charitable organization to meet those needs and allow others across the country to share experiences and information.

The vision was of an organization of national scope with a distinguished medical advisory board. The mission was to decrease diagnostic time and improve treatment. The focus was simple and succinct: give others what had not been available for them—relieve the pain, the anxiety, and the tumultuous search for answers and information. The founders realized that success would be totally dependent on enthusiastic, knowledgeable volunteer work from recovering ANs and their families and health professionals. In March 1984, their dream was realized with the incorporation of the Acoustic Neuroma Association of Canada, a national charitable organization with the vision statement, "The Hope is Recognition and Treatment." The ANAC National Office resided in Edmonton, Alberta until October, 2007 when the office was relocated to Ontario. For further information please visit our website www.anac.ca or call 1-800-561-2622.

ANAC recommends treatment from a medical team with substantial acoustic neuroma experience.

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