BC Early Hearing Program

A service of the Provincial Health Services Authority

Screening Training Manual

Section 4:
The Auditory System

4.1 Anatomy and Physiology of the Ear
4.2 Types, Degrees and Implications of Hearing Loss
4.3 Risk Factors for Late onset Hearing Loss
4.1 Anatomy and Physiology of the Ear

The components of the auditory system can be categorized as peripheral or central. The 3 major components of the peripheral auditory system are:

- Outer ear
- Middle ear
- Inner ear

A summary of the anatomy of the peripheral auditory system is provided in this section.

Figure 1 - Diagram of the peripheral auditory system
The Outer Ear

The outer ear consists of the auricle (pinna) and the ear canal. The pinna is the visible part of the ear and consists of cartilage covered with skin. It has a number of characteristic folds and indentations unique to each person (Flexer, 1999; Preves and Curran, 1986). The outer ear ends at the tympanic membrane (eardrum), which separates the outer ear from the middle ear (Flexer, 1999), [please see Figure 2 for a diagram of the outer ear]. Sound travels through the ear canal and moves, or vibrates, the eardrum.

![Diagram of the pinna with key landmarks](image)

**Figure 2 - Diagram of the pinna with key landmarks (reprinted with permission from OTO Hearing Products)**

The Middle Ear

The middle ear is between the eardrum and the inner ear. The middle ear contains 3 tiny bones (the ossicles) and the eustachian tube. The eardrum vibrates, causing the middle ear bones to move and send the sound through the middle ear to the inner ear. There is also a tube that runs from the middle ear space to the back of the throat, called the eustachian tube.
The Inner Ear

The inner ear includes the cochlea (a snail-shaped organ), the semi-circular canals (the balance mechanism), and the auditory nerve. Please see Figure 3 for a diagram of the inner ear. The cochlea is the part of the inner ear that is associated with hearing. When the sound vibrations enter the inner ear from the middle ear, the cochlea sends nerve impulses to the brain through the auditory nerve. Once the brain receives the nerve impulses, there is a sensation of hearing.

The function of the outer hair cells is the basis of the audiometric test called Otoacoustic Emissions (OAE).

The function of the cochlea and the auditory nerve is the basis of the audiometric test called Auditory Brainstem Response (ABR).

Figure 3 - The Inner Ear
4.2 Types, Degrees and Implications of Hearing Loss

4.2.1 Types of Hearing Loss

Hearing loss can happen in any part of the ear, including the outer ear, the middle ear, and the inner ear. Hearing loss can happen in any one of these places, or in more than one place. Each type of hearing loss has a different name and has different possible treatments or therapeutic strategies.

Conductive Hearing Loss:

This type of hearing loss occurs in the outer ear and/or in the middle ear. Something prevents sound waves from reaching the inner ear. This blockage can be partial or complete. Conductive hearing losses don’t cause total hearing loss but do cause a loss of loudness. When the sound is made louder, it is heard clearly.

Most conductive hearing losses can be treated with medication, surgery, or environmental change. Sometimes hearing aids or other amplification devices can help by making sound louder.

Common causes of conductive hearing loss are:

✓ Faulty development of the outer ear and/or middle ear
✓ Blockage of the ear canal (wax or a foreign object)
✓ Damage to the eardrum
✓ Damage to the 3 bones in the middle ear
✓ Failure of the eustachian tube to let air into the middle ear space
✓ Infections of the middle ear
✓ Growth of extra bony material around the three bones in the middle ear
✓ Cysts

Sensorineural Hearing Loss:

This type of hearing loss occurs in the inner ear: the auditory nerve or the parts of the brain that receive the nerve impulses from the auditory nerve. This type of hearing loss is sometimes called nerve deafness. Hearing loss due to trouble with the auditory nerve is called auditory neuropathy or auditory dysynchrony syndrome.

Sensorineural hearing loss can range from mild to profound. Not only is there a loss of loudness but there is also a loss of clarity. Almost all children with this type of loss will have some degree of loudness and/or clarity left. The remaining hearing is called residual hearing, and it can be amplified; therefore, the child may be able to be taught to pick up sounds and/or speech. This type of hearing loss cannot be treated by medication or surgery. It is considered permanent because nerve cells cannot grow back or be replaced.

Common causes of sensorineural hearing loss are:

✓ Faulty development of the inner ear
✓ Family history of hearing loss
✓ Damage to the inner ear and/or hearing nerve from an illness before birth
Mixed Hearing Loss:

A mixed hearing loss occurs in both the outer/middle ear and in the inner ear. It is a combination of conductive and sensorineural hearing loss.

Unilateral Hearing Loss:

A unilateral hearing loss occurs in only one ear, and it can be conductive, sensorineural or mixed. Although a child with this loss has good hearing in one ear, he/she will have difficulty knowing where sound is coming from, hearing in noisy environments, and hearing on the affected side.

Progressive Hearing Loss:

A progressive hearing loss occurs when a child loses the sense of hearing over time. A baby may be able to hear at birth and gradually lose the ability. It is important to note that, just because a baby passes newborn hearing screening, it does not mean that normal hearing will always be present. Progressive loss can be conductive, sensorineural or mixed.

Fluctuating Hearing Loss:

This type of loss is one that changes frequently by improving or worsening. Some conductive hearing losses are fluctuating.

An example of this is hearing that worsens when a child has fluid in the middle ear, caused by an ear infection, and the hearing improves when the infection is resolved.

Syndromic Hearing Loss:

Sometimes an infant or young child who is deaf or hard-of-hearing may have other signs or symptoms, too. When multiple congenital malformations appear together, they may be described as a syndrome. This is important because, if hearing loss is detected early, specialists such as genetic professionals may be able to test for certain syndromes that may not be physically identifiable by only appearance. Two examples of such syndromes are:

- Usher Syndrome which is associated with progressive loss of vision
- Jervell Lange-Nielsen Syndrome which is associated with heart defects
4.2.2 Degrees of Hearing Loss and Potential Effects

The following chart helps to explain what sounds a child may or may not hear without amplification, based on the degree of hearing loss. It identifies how amplification helps a child and the potential effects the hearing loss might have on their ability to hear and recognize spoken conversation and environmental sounds. This is, however, only a guide; each child has unique potential and uses that potential differently.

<table>
<thead>
<tr>
<th>Degree of Loss</th>
<th>Decibels</th>
<th>Potential Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Slight Hearing Loss</td>
<td>16 - 25 dB</td>
<td>A minimal loss of some sounds. May have difficulty hearing quiet or distant conversations, especially in noisy environments.</td>
</tr>
<tr>
<td>Mild Hearing Loss</td>
<td>26 - 40 dB</td>
<td>Without amplification, the child can hear most conversations up close and in quiet environments but is likely to miss parts of words. The child may appear to be hearing when she/he wants to. Amplification and lip reading may supplement understanding of what is said.</td>
</tr>
<tr>
<td>Moderate Hearing Loss</td>
<td>41 - 55 dB</td>
<td>Without amplification, the child will have difficulty hearing spoken conversation. 50 - 100% of spoken conversations may be missed. Proper amplification and intervention should enable the child to hear and recognize all sounds.</td>
</tr>
<tr>
<td>Moderate to Severe</td>
<td>56 - 70 dB</td>
<td>Conversation must be very loud to be heard without amplification. Proper amplification will help the child to develop awareness of spoken language. Age of amplification, consistent use of hearing aids, and intervention are important to help the child learn to use his/her hearing.</td>
</tr>
<tr>
<td>Severe Hearing Loss</td>
<td>71 - 90 dB</td>
<td>Without amplification, the child may hear loud voices and sounds close to the ear. With early and consistent use of hearing aids, many children will be able to detect sounds such as speech. Most children will use vision in addition to, or in place of, hearing.</td>
</tr>
<tr>
<td>Profound Hearing Loss</td>
<td>91 dB or &gt;</td>
<td>Without amplification, the child will be only aware of loud sounds/vibrations. The child may rely on vision rather than hearing as the primary means for communication and learning. Amplification may or may not be useful in hearing spoken conversation.</td>
</tr>
</tbody>
</table>
Unilateral Hearing Loss

The child may have difficulty hearing faint or distant spoken conversations. The child will usually have difficulty understanding spoken conversations coming from the side of the head with the hearing loss, and difficulties understanding when the environment is noisy.

4.3 Risk Factors for Late Onset Hearing Loss

Late onset hearing risk indicators are factors which predispose the infant to have or to develop permanent hearing loss in later childhood. They can also be called late onset and, sometimes, late onset/progressive indicators. Please see Appendix 9 – Late Onset/Progressive Hearing Loss Risk Factors.

Late Onset Risk Factors Present at Birth:

Screening personnel will obtain this information at the time of screening by asking the baby’s nurse or family, as appropriate. Information about the first three risk factors (A–C) needs to be determined for all babies, including infants screened in the Neonatal Intensive Care Unit (NICU). The remaining factors are applicable only to babies in the NICU for more than 48 hours.

A. Family History: Is there a family member with permanent childhood hearing loss (baby’s parent, brother, sister, grandparent, first cousin, aunt or uncle)? If so, then the following information about each affected family member needs to be collected.

   Family relationship: baby’s parent, brother, sister, grandparent, first cousin, aunt or uncle.

   Age of onset if known – must be before age 12 years. Adult late onset hearing losses due to noise, aging etc. are NOT risk factors for childhood hearing loss.

   Type or cause: congenital or progressive sensorineural loss, or conductive hearing loss due to congenital malformation. Hearing losses resulting from ear infections, head injury or childhood illness are NOT hereditary and are not risk factors for the infant. A hearing loss detected later in childhood may, in retrospect, be attributed to an illness; however, if there is no clear evidence the illness caused the hearing loss, it could be coincidental with an actual hereditary loss.

   Genetic hearing loss may be dominant progressive, recessive non-syndromic or non-syndromic X-linked. In each of these, the hearing loss may be progressive, with onset anywhere from birth to early adulthood. Some types are more likely to have early onset (at less than 2 years of age) than others. In some cases Large Vestibular Aqueduct Syndrome may be hereditary. This is associated with progressive SNHL, which is present in close to 50% of cases. The onset of hearing loss can range from infancy through to adulthood.
B. Craniofacial Defects of Head, Neck and Ears: Unusual appearance of head, face or ears, including cleft palate (NOT cleft lip in isolation) and microtia/atresia (NOT including ear tags or dimples around the ears).

Visual Inspection by Screener:

Screeners are to do a visual check of the outer ear prior to screening. Consult with the nurse or physician prior to classifying an abnormality.

Cleft Lip and Palate:

Cleft lip and cleft palate are birth abnormalities of the mouth and lip which affect about one in every 700 births and are more common among Asians and certain groups of First Nations than among Caucasians. They occur less frequently among African-Americans. Cleft lip and cleft palate occur early in pregnancy when the sides of the lip and the roof of the mouth do not fuse together as they should. A child can have cleft lip, cleft palate, or both. Cleft lip and cleft palate together are more common in boys. It is also important to know that most babies born with a cleft are otherwise healthy with no other birth abnormalities.

The eustachian tube does not function well, which can lead to recurrent ear infections. Middle ear disease is present at birth in most infants with cleft palate.
Atresia/Stenosis of External Ear Canal:

The Congenital Auditory Atresia (CAA) rate is 1 in 10,000-20,000 live births. A unilateral presentation of CAA is more common than a bilateral presentation, with an approximate ratio of 3:1. Atresia is a complete blockage of the external ear canal. Stenosis is a narrowing of the external ear canal. The outer portion of the ear canal may appear normal, with narrowing or closure occurring further down. This may be difficult or impossible for the screener to identify as an otoscopy is not performed. A complete absence of the ear canal, or a blockage which can be seen easily upon visual inspection should be noted to the nurse or physician for further evaluation.

![Atresia](image1.png)

![Microtia with normal ear canal](image2.png)

Unusually Shaped External Ears:

Note: Infant cartilage is very soft and the ears are quite malleable at birth. The birthing process or position of babies' heads when sleeping may cause temporary folding or an unusual appearance of the outer ear. Unusual ear shapes may also be hereditary.

Look for underdeveloped pinna (microtia) or very low set ears.

![Microtia with normal ear canal](image3.png)
C. Syndromes Associated with Progressive/Late Onset Hearing Loss:

There are a number of genetic syndromes which are associated with sensorineural/mixed/conductive hearing. In many cases, later onset or progressive hearing loss has also been documented. Some common syndromes which have hearing loss associated with them are:

- **Down Syndrome** – can result in higher risk for otitis media (middle ear fluid or infection) due to craniofacial structure, or higher risk for sensorineural hearing loss, which may be progressive. Requires long term monitoring of hearing.

- **Stickler Syndrome** – can result in higher risk for otitis media related to the cleft palate, but also may have progressive high frequency sensorineural hearing loss.

- **Fetal Alcohol Syndrome/Effects (FAS/FAE)** – Otitis media is present in 93% of cases, but sensorineural hearing loss is also present in approximately 29% of cases. It may be progressive. **NOTE:** Neonatal Abstinence Syndrome (NAS) is not a risk factor for late onset hearing loss.

- **Brancho-Oto-Renal Syndrome**
- **Waardenburg Syndrome**
- **Pendred**
- **CHARGE association**
- **Mitochondrial disorders**
- **Usher Syndrome**
- **Alport**
- **Large Vestibular Aqueduct (LVA)**

This is not an exhaustive list. Syndromes and associated risks must be researched individually. This will be done by the hearing clinic that receives the referral, not the screener.

**The remaining risk factors are found in infants who have spent at least 48 hours in the NICU. The screener should check with the nurse for these factors and not ask the family.**

D. Birth weight less than 1200 grams:

A low birth weight is not typically the risk factor, in and of itself. These infants experience multiple complications, which may place them at risk for hearing loss. They may be at greater risk for progressive hearing loss as well.
E. Breathing Problems at Birth:

i. APGAR score: a five minute APGAR score of less than or equal to 3. Appearance, pulse, grimace/grip, activity and respiration are scored at one and five minutes after birth.

ii. Hypoxic-Ischemic Encephalopathy (HIE), moderate/severe Sarnat II or III: HIE is a term for brain damage caused by lack of oxygen and lack of blood flow to the brain. Damage can occur within minutes. Once brain damage occurs, it is irreversible.

iii. Congenital Diaphragmatic Hernia (CDH): Diaphragmatic hernias may be congenital or acquired. Congenital diaphragmatic hernias are sometimes present at birth. They occur because of abnormal development of the embryo’s bowel, due to the abdomen herniating upward through the diaphragm into the chest (thoracic) cavity.

iv. Extra-Corporeal Membrane Oxygenation (ECMO) or inhaled Nitrous Oxide (iNO) or High-Frequency Oscillatory (HFO) or Jet (HFJ) ventilation: These are types of ventilation used for babies with severe breathing problems.

Sensorineural hearing loss is diagnosed in about 20-40% of these infants, and progressive hearing loss is common, especially in the early years. The length of time that mechanical ventilation was needed, seems to be a major factor in whether there is a hearing loss or not. The babies who were diagnosed with hearing loss (initially or progressive) were also the ones who had more severe cases and required ECMO while in the NICU. The incidence of sensorineural hearing loss in this population is 25 times higher than in the general NICU population.

F. Brain Dysfunction:

i. Intra-Ventricular Hemorrhage (IVH), Grade III or IV (Grade IV is seen by neonatal follow-up at Children’s Hospital): bleeding into the normal fluid spaces (ventricles) within the brain. IVH is also used to refer to bleeding in areas near the ventricles even if the blood is not within them. The extent of IVH is graded from I to IV. Complications, of which hydrocephalus is the most common, occur primarily with grades III and IV.

ii. Peri-Ventricular Leukomalacia (PVL): is damage and softening of the whitematter, the inner part of the brain that transmits information between nerve cells and the spinal cord, as well as from one part of the brain to another. Babies with PVL have a higher risk of developing cerebral palsy.

G. Hyperbilirubinemia \( \geq 400 \, \mu M \) OR meeting any standard criteria for exchange transfusion.

Hyperbilirubinemia (Jaundice): is reflected in the colour of the infant’s skin. Most babies with jaundice are not at risk for hearing loss, unless the infant has had a very high level, which may necessitate a blood transfusion. These babies are at risk for sensorineural hearing loss which may be progressive or late onset. They are also at higher risk for neurological issues such as Central Auditory Processing or Auditory Dysynchrony.
H. Lab-Proven Infection:

i. Perinatal (in the baby) TORCHES infection:

Toxoplasmosis – is contracted by contact with cat feces or undercooked meat. In the U.S., 6/1000 women contract it during pregnancy, and 10-15% of infants born to these women, are born with evidence of the infection. Others may be born with subclinical infection. Hearing loss may be congenital or later onset.

Rubella (German Measles) – is very rare, ever since development of a vaccine in 1969. If prenatal rubella infection occurs in the first 20 weeks of pregnancy, heart, eye and auditory damage can result. Hearing loss may be congenital or progressive.

Cytomegalovirus – is believed to be the leading cause of non-hereditary hearing impairment. It is a member of the herpes virus group. Women who work with children carry an 8-12% chance of contracting CMV infection (non-symptomatic). Approximately 1-2% of newborns are infected at birth. Hearing loss can occur with symptomatic infants as well as those who are asymptomatic. It occurs in 30-62% of symptomatic infants and in 8-17% of asymptomatic infants. In both groups, the hearing loss may be later onset, and progressive – it is progressive in 80% of cases. Hearing loss can develop months or years after birth (onset in a few cases has been found as late as even 7 years of age).

Herpes – is more likely to result in hearing loss in cases of prenatal infection. Hearing loss may be unilateral or bilateral, congenital or progressive.

Syphilis (Congenital) – is an infection which can be passed from mother to child during fetal development or birth. Congenital syphilis is a severe, disabling, and often life-threatening condition for the infant. Nearly half of all children infected with syphilis during gestation die shortly before or after birth. These infants are at greater risk for developing late onset hearing loss.

ii. Meningitis – is a viral or bacterial infection of the tissues (meninges) and sometimes the Cerebral Spinal Fluid (CSF) that surrounds the brain and spinal cord. Literature reports that the incidence of hearing loss after this infection varies between 10-20%.

I. Overdose of Gentamycin or other Aminoglycosides

Gentamycin is one of a group of antibiotics, called aminoglycosides, that are effective in treating certain types of bacteria. They are used extensively in the NICU. When infants receive an accidental overdose, fivefold or greater, they are at an increased risk for hearing loss.