WHEN THE PHONE RINGS MY BED SHAKES

A Primary Care Physician’s Perspective on Hearing Loss

Philip Zazove, M.D.
Clinical Professor of Family Medicine

I. Consider how you use the word deaf or deafness. It has different implications for different people.
   A. Various uses of the word "deaf" and various definitions
   B. WHO definition of "deaf" - persons whose hearing impairment is so severe, they can not benefit from amplification.
   C. The American Speech and Hearing Association (ASHA) scale of deafness:

<table>
<thead>
<tr>
<th>Degree of Loss</th>
<th>dB loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>&lt;27</td>
</tr>
<tr>
<td>Mild</td>
<td>27-40</td>
</tr>
<tr>
<td>Moderate</td>
<td>41-55</td>
</tr>
<tr>
<td>Moderately severe</td>
<td>56-70</td>
</tr>
<tr>
<td>Severe</td>
<td>71-90</td>
</tr>
<tr>
<td>Profound</td>
<td>&gt;90</td>
</tr>
</tbody>
</table>

   Problem here is that hearing loss in different areas of the speech range have different impact on communication.

II. Factors that go into determining how much a hearing loss affects someone:
   A. Type of loss
      1. sensorineural
      2. conductive
      3. mixed (both sensorineural and conductive)
   B. Frequencies involved
Speech range is 500 to 4,000 cps: speech sounds mostly 1500 dB and under
Hearing losses in speech range have much more impact than in other areas of the
hearing spectrum

C. Intensity of loss (measured in decibels)
   Up to 25 decibels is considered within normal range of hearing

D. Age of loss
   • Pre-lingual (11%) – has biggest impact on communication and
     life (e.g., more likely to have lower education and income
     levels)
   • Post-lingual but pre-educational
   • Post-educational but prevocational (25%)
   • Post-vocational - most common in elderly, but still has a huge
     impact on life if not diagnosed and addressed.

E. Whether one or both ears affected

F. Idiosyncratic factor
   "Some individuals with mild hearing loss may experience
   substantial disability and handicap, whereas others with moderate
   hearing loss may not exhibit any form disability or handicap."
   Bess et al, 1989

III. Scope/Epidemiology of hearing loss
A. 1 in 10 people have it; 90% of these are oral and prefer English
   rather than sign language.
B. Perhaps 1 in 1000 have profound hearing loss
C. More common in:
   1. Elderly persons (up to 50% of those over 85 years of age)
   2. Male/female ratio = 118/100
   3. North Central and Western United States
   4. White race
   5. NBICU survivors
D. Second most common disability in the United States but most
   common single disability (orthopedic deformities is greatest
   category but that includes multiple types of conditions)

   The following quote remains true today:

   “Total number of people who have a hearing loss is greater than the
   combined total of individuals who have blindness, cancer, heart
disease, kidney disease, multiple sclerosis, tuberculosis and venereal disease. Tunley, 1974

E. Decreasing in younger ages due to
1. rubella immunizations
2. decreased ear infection complications
   May change due to increased hearing loss in NICU survivors, for reasons that remain unclear.

F. Burgeoning geriatric population is increasing overall importance of hearing loss to society

IV. Causes of hearing loss
A. Sensorineural - about 50% overall, more in adults
   1. Hereditary (15-40%)
      - different syndromes, including other organ involvement
      - sometimes other d/Deaf in family but not often not
      - 70% of those not associated with a syndrome and which are autosomally recessive, are due to abnormalities of Connexin 26 gene on chromosome 13
   2. Congenital
      - toxicity
         - Rubella (3%), CMV, syphilis
         - Drugs
         - Anoxia
      - trauma
         - Delivery causes intracranial bleeding or eighth
         - Nerve or cochlear (inner ear) damage
   3. Trauma
      - Skull fractures
      - bleeding
         a) concussion
   4. Infections
      - Mumps, scarlet fever, meningitis (12%), mastoiditis
   5. High fever in childhood
   6. Drugs
      - Various, especially aminoglycosides, furosemide
   7. Presbycusis
      - Probably cochlear ganglion cell atrophy
      - Starts with high frequency loss
   8. Hypothyroidism
      - Congenital and acquired
   9. Occupational/Environmental
      - Especially over 85-90 decibels for months or years
      - Temporary threshold shift and permanent threshold
      - Millions of dollars involved
   10. Meniere’s
   11. Neurological diseases
      - Acoustic neuroma
      - Tumors
      - Multiple sclerosis
      - Otoclerosis
      - Idiopathic
   12. Sudden deafness
      Vascular vs. viral (mumps)

B. Conductive - about 50% overall, more in children
   1. Obstruction external ear canal
- Earwax - common
- Other such as congenital syndromes that have no ear canal

2. Eardrum problems
- Perforation of eardrum
- Thickening due to scarring or other cause

3. Middle ear - most common cause of conductive hearing loss
- Otosclerosis
- Adhesions
- Tumors
- Ossicle problems

4. Bone disorders

V. Prelingual deafness
a. Average age at diagnosis remains 2-3 years old
1. This has been slowly decreasing over time
2. Traditionally, 75% are detected by family, not by physicians
3. The delay in identifying moderate or profound hearing loss results in a delay in teaching these babies language
4. Thus, some advocate screening high risk babies routinely

b. High-risk infants
1. High risk criteria pick up approximately 50% of people with significant hearing losses
2. Of all high risk infants screened, 2-10% have a significant hearing loss
3. Background rate of hearing loss in general population is at most 1%
4. The high risk criteria suggesting the need for screening are:
   1. Admission to NBICU
   2. Family history of childhood hearing impairment
   3. Congenital infection associated with hearing loss, e.g. TORCH
   4. Craniofacial anomalies including cleft palate
   5. Birth weight under 1500 grams
   6. Hyperbilirubinemia requiring exchange transfusion
   7. Ototoxic drugs in more than usual amounts
   8. Bacterial meningitis
   9. Severe asphyxia at birth
   10. Prolonged mechanical ventilation >9 days
   11. Stigmata associated with hearing impairment
   12. Extracorporeal membrane oxygenation

c. Universal Newborn Screening
1. This is being increasing advocated and there’s a push to make this a national law.
2. It is currently mandated by law in at least 40 states – but NOT Michigan.
3. It is usually done by utilizing Acoustic Brainstem Responses to sound reception or Otoacoustic Testing. If positive, more comprehensive follow-up testing is needed to see if this is truly a hearing loss or just a false positive reading.
4. It has significantly increased the identification of infants with moderate to severe hearing loss by age 10 months (57% vs. 14%).
5. Joint National Commission goal is to screen all infants by 3 months of age and, where abnormal, begin intervention by 6 months
6. The major limitation/unknown, however, is whether identification of these infants at a very early age does indeed improve long-term language outcomes; this has not been proven.

d. Genetic Causes of pre-lingual hearing loss
1. 50% of non-syndromic, congenital, autosomal recessive hearing loss is due to polymorphisms in the Connexin gene
   a. Especially Connexin 26 and 30
   b. Pendred, Usher’s syndrome are other common genetic conditions though all are not necessarily presenting at birth
   c. MT-RNR1 (A1555G) and MT-TS1 – mitochondrial genetic conditions causing hearing loss
2. May be other significant polymorphisms we haven’t yet identified

6. Hearing Loss in Elderly Persons
   a. Significant increase prevalence with age
      | Age          | Prevalence |
      |--------------|------------|
      | 65-74 years  | 23%        |
      | 75-84 years  | 33%        |
      | 85 and older | 48%        |
   b. Many/most of these persons with hearing loss refuse to acknowledge it
      1. This is because people often ridicule or make fun of individuals who have hearing losses.
      2. Also, these persons find it increasingly difficult to understand others because they can’t hear enough, causing increasing isolation and depression
      3. Need to consider this and screen for hearing loss in elderly Persons
   C. Consider screening with the Hearing Handicap Inventory for the Elderly- Screening version (HHIE-S) and/or audioscope. Combination has a PV of .63. (Lichtenstein et al, 1988).
      Does a hearing problem cause you to feel embarrassed when you meet new people?
      Does a hearing problem cause you to feel frustrated when talking to members of your family?
      Do you have difficulty hearing when someone speaks in a whisper?
      Do you feel handicapped by a hearing problem?
      Does a hearing problem cause you difficulty when visiting friends, relatives or neighbors?
      Does a hearing problem cause you to attend religious services less often than you would like?
      Does a hearing problem cause you to have arguments with family members?
      Does a hearing problem cause you difficulty when listening to television or radio?
      Do you feel that any difficulty with your hearing limits or hampers your personal or social life?
      Does a hearing problem cause you difficulty when in a restaurant with relatives or friends?
      (Score 4 points for each "yes" answer, 2 points for each "sometimes" answer, and 0 points for each "no" answer. A score of 10 or more suggests a significant hearing loss.)

7. Four Basic Groups of D&HH persons
   A. Deaf - capitalized D in Deaf refers to members of the Deaf Community (at most 10% of all D&HH persons)
      - Are <= 10% of people with significant hearing loss
      - Most Deaf oppose cochlear implants or preconceptual genetic testing for deafness
      - Are proud of deafness and are public about it
      - Prefer to communicate in American Sign Language
B. deaf - small deaf refers to persons not part of Deaf community
   - Usually prefer oral communication (English, Spanish, etc)
   - Are the vast majority of people with hearing loss
   - **favor** cochlear implants and preconceptual genetic testing for deafness
   - Are embarrassed about hearing loss and keep quiet about it

C. Hard of Hearing (along with deaf, comprise 90% of all D&HH persons)
   - largest group
   - include those who lose hearing in old age (presbycusis)
   - also often embarrassed and keep quiet about problems
   - have wide range of hearing losses, thus difficult to concisely categorize

D. All others
   - Spanish Sign Language and other sign languages
   - Other oral languages

7. Communication - THE major issue of D&HH people
   A. Deaf learn language differently
      It is critical that a child learn a language before reaching 3 years of Age - otherwise lose ability for abstract thinking per findings of the RAND study
   B. Deaf persons often learn American Sign Language (ASL)
      Different from English
      Different from Signed English or Contact Signed English
      Unique syntax, grammar, idioms
   C. Deaf community recognized as distinct minority culture. Unique:
      1. Language
      2. History
      3. Traditions
      4. Beliefs
         • e.g., Deaf community against cochlear implants and prenatal genetic testing for deafness
      5. Self-contained group
   D. Interpreters
      1. Must meet certain specifications
         a. Includes code of ethics and confidentiality
            1. For example, they are obligated to tell the patient everything the physician says, even if the physician is giving an aside to a nurse
         b. Different levels of expertise
            - For medical interpreting, level 3 is ideal because of medical jargon and complex concepts
      2. Can be sign or oral interpreters
      3. Family members are NOT appropriate for interpreting
      4. Americans with Disabilities Act states that D&HH persons have a right to an interpreter at the physician's expense

8. Socioeconomic status of the Deaf community in Michigan
   A. Education
      1. Read at 6th grade level
      2. Only 13% graduate college
   B. Economic
      1. 45% with full-time job, usually lower paying ones
      2. 60% with pretax income <= $25,000

9. Psychosocial Aspects
   A. A strong identifiable subculture in the Deaf community
   B. Parents of deaf children - initial reactions on finding out are often
Like those of death and dying, i.e., shock, denial, grief

C. Hearing children of deaf parents issues -- similar in some ways to those of children of alcoholic parents

D. Depression - unclear if more common in young persons with hearing losses; studies currently being conducted. Definitely present in elderly persons with hearing losses (even mild ones).

E. School environment is often quite different for deaf children

10. Health Care Utilization and Health Status of D&HH persons

A. They have trouble communicating with physicians
   1. 44% trouble communicating (higher educated deaf)
   2. Over half write:
      • problem because ASL is different grammar than English
      • many not know common terms such as nausea, allergic
      • 59% not understand written notes how to take their Medications
   5. McEwen showed that D&HH persons are greater risk than other non-English speaking persons for miscommunication problems

B. National Health Interview Survey data reveals that they:
   1. Visit physicians roughly twice as often
      a. Deaf visit physicians less than hearing person
      b. deaf visit physicians more than hearing persons
   2. Have more bed days ill or disabled
   3. Assess themselves less healthy

C. Mortality studies
   1. Results contradictory; mostly poor-quality studies
   2. Probably similar mortality rates to hearing persons

D. Physician Attitudes towards D&HH patients. They:
   1. Feel less comfortable with them
   2. Understand them less
   3. Often don't use interpreters even though they agree that interpreters help significantly with communication
   4. Believe these patients are less likely to understand their diagnosis and treatment

E. D&HH patients have different attitudes and lifestyle behaviors
   1. Smoke, drink and use drugs less
   2. Different beliefs about and attitudes towards AIDS
   3. Specific cultural beliefs about physicians/health care, e.g.:
      • don't like going to doctors because of difficulties communicating
      • feel are not treated as a person by health care providers

F. Knowledge about health care issues is deficient
   1. Preventive Medicine
      a. D&HH patients know less about current recommendations
      b. Physicians perform fewer preventive interventions on D&HH patients
      c. If spouse is hearing, D&HH person has better knowledge
   2. Disease Understanding
      a. D&HH more likely to think can acquire AIDS from toilets and appear to be more intolerant of people with AIDS
   3. Education
      a. More difficult due to communication barriers; e.g., average Deaf person reads at 6th grade level.
b. Unclear how to improve as language appropriate interventions don’t necessarily improve health care knowledge

G. Occupational Hearing Loss
   1. Multi-billion dollar issue
   2. OSHA has strict rules to prevent hearing loss
   3. Many don’t comply
   4. People who develop hearing loss, have same issues as non-Deaf persons who lose their hearing in other ways
      a. Embarrassed
      b. Hide it from others
         - Even if tell others, supervisors often downplay hearing loss

11. Americans with Disabilities Act (ADA)
   A. Requires that physicians furnish and pay for reasonable auxiliary aids and services, including interpreters, necessary to ensure effective communication with the patient
   B. Patients make the decision what aids they need and they MUST notify the physician ahead of time if they expect the physician to pay for this.
   C. Lawsuits have been filed claiming physicians breached the ADA, and some d/Deaf patients have won monetary awards against physicians.

Know what your ADA responsibilities are - it’s the right thing for optimal patient care to allow you to get a good history and communicate with your patients.

12. What to DO and NOT DO when interacting with deaf persons
   A. DO
      1. Look directly at the deaf person when speaking to them
      2. Be aware of culture, communication issues
      3. Be aware of legal rights under the ADA to interpreter and other auxiliary aids at the physician’s expense
      4. Ask about the possibility of hearing loss in WCC visits
      5. Ensure there is good lighting
      6. Enunciate clearly but don’t over-enunciate
      7. Know the differences between Deaf, deaf and hard of hearing
      8. Be prepared to repeat information several times
      9. Consider possibility of hearing loss in young children if state doesn’t have mandatory newborn screening
     10. Understand that many D&HH have lower understanding of basic health care issues
   B. DO NOT
      1. Over enunciate
      2. Talk to the interpreter
      3. Assume patient understands without double-checking
      4. Stand before a window or other bright light that would glare in the patient’s eyes
      5. Whisper or speak asides to the nurse or interpreter
      6. Write with patients unless they request that mode of communication
      7. Assume the patient prefers a specific language

The test questions.

What will not be on the test:
   1. Etiologies of hearing loss
2. Occupational/Environmental information
3. Specific frequencies and decibels of various speech sounds
4. List of high risk criteria for newborn infants (though this could be on the board exam)
5. Hearing Handicap Inventory

What may be on the test:
1. Americans with disabilities requirements of physicians
2. Communication and socio-economic issues related to hearing loss
3. Health outcomes of hearing loss
4. Differences between deaf, Deaf, Hard of hearing and implications from that.