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Dual Sensory Loss: Overview of Problems, Visual Assessment, and Rehabilitation

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This article provides an overview of some of the problems and possible solutions surrounding the neglected issue of combined vision and hearing deficits. The subject is treated by considering each subpopulation, ranging from those who have no residual vision or hearing to those with mild coexisting vision and hearing losses. An attempt is made to relate the different types of visual deficit to the likely problems encountered in real-life activities, such as communication and travel, among individuals who also have a hearing impairment. The assessment and appropriate referral of patients with these combined deficits is discussed, including the interpretation of visual test results and the importance of factors other than standard visual acuity. Speculation is offered on potential strategies and solutions for rehabilitation as well as the need for future research and improvements in service delivery.

Keywords: visual impairment; contrast sensitivity; dual sensory loss; sign reading; speechreading

Introduction

Dual sensory loss is an increasing problem that is in urgent need of more attention by both vision and hearing rehabilitation researchers and professionals.[1] For the purposes of this article, the terms *deaf* and *blind* are taken to mean having no functional ability in the affected sense, whereas *impaired* vision or *hard of hearing* is taken to mean partial (less severe) loss. By this definition, very few people are deaf-blind—less than one hundredth of 1% of the population. Other definitions of deaf-blindness include individuals with functional hearing and/or vision. For example, legally, children in the United States are called deaf-blind if they have

> such severe communication and other developmental and learning needs that the persons cannot be appropriately educated in special education programs solely for children and youth with hearing impairments, visual impairments or severe disabilities, without supplementary assistance to address their educational needs due to these dual, concurrent disabilities.[2(p46756)]

This is not the definition used here.

Programs exist to assist those who are functionally deaf and blind. However, a vastly larger population of individuals with varying degrees of coexisting visual and auditory impairment has been slowly emerging; as our overall population ages, the number of older adults with both impaired vision and impaired hearing is growing rapidly, and many more "fragile" premature infants are surviving but often have vision and/or hearing impairments.

The characteristics of this larger population are far less easily defined than those who are deaf-blind by our strict definition. Their problems are less well understood and often overlooked. This is partly a result of the division of the professions serving the population; in medicine as well as in rehabilitation, there are separate and different professionals, institutions, and

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systems for serving the visually impaired and the hearing impaired. A low-vision practitioner may more often than not neglect even to ask a patient whether he or she has hearing or communication problems. An audiologist may similarly neglect to inquire into the patient's visual status. In either case, the different professionals often lack expertise in the other type of sensory loss.

For all these reasons, the problems caused by combined visual and auditory deficits have not been well addressed. In the remainder of this article, an attempt is made to better identify and define the population characteristics, the problems caused by such dual sensory loss, and the implications for rehabilitation.

Characteristics and Problems of Dual Sensory Loss Subpopulations

Overview

According to the Eye Diseases Prevalence Research Group, approximately 1 million people are legally blind (corrected acuity in the better eye 20/200 or worse, or visual field less than 20°).*[3] The same source estimates that 3 million have low vision, usually defined as best corrected visual acuity less than 20/70, whereas almost 14 million have a visual impairment that hampers performance and enjoyment of everyday activities. Approximately 260 000 (0.09%) individuals in the United States have light perception or less, including 130 000 (0.04%) who are totally blind (no light perception).[4]

Visual impairments and blindness are increasingly frequently associated with coexisting sensory, physical, and/or cognitive impairment.[5] One study reported that 68% of visually impaired children in Atlanta had multiple disabilities.[6] Among older people in the United States, 21% have both vision and hearing loss by age 70,[7] and the numbers are swelling as baby boomers reach that age group. A recent analysis[8] estimates that there are 45 000 to 50 000 deaf-blind (as defined by IDEA [Individuals with Disabilities Education Act]) individuals in the United States. All population estimates are heavily dependent on the definitions of impairment or disability used. The following sections examine the individual subgroups and problems in more detail.

Deaf-Blind

As noted earlier, few individuals are functionally blind and functionally deaf.† Historically, perhaps because of the severity of the problems caused by total or almost total deafness and blindness, this subpopulation has attracted at least some attention from researchers and has been served by distinct rehabilitation programs such as the Helen Keller National Center. In this population, communication is made possible using the sense of touch—Braille for reading, tactile sign, tadoma, and so on. Deaf-blind individuals cannot use the regular telephone devices for the deaf (TDDs) because of their visual displays, but special "Braille TDDs" with Braille output are available to enable deaf-blind individuals who know the Braille code to use the telephone. Similarly, Braille computer access systems can enable the use of computers. Variants of these devices can be used to enable face-to-face communication between a deaf-blind Braille user and a sighted individual via a keyboard and display interface.

However, most deaf-blind individuals are those Usher's syndrome patients who suffer from loss of functional vision in adulthood. They learn to cope in the deafness world before losing their vision later on. Therefore, they are usually not familiar with the Braille code and learning it is often not attractive to them later in life when they lose their vision. For this population, prototype robotic finger-spelling hands have been produced,[9] designed to replicate the finger-spelling code used by this population. Efforts are ongoing to produce

*Understanding visual acuity and visual field values: 20/20 acuity (Snellen notation) means that the individual can see at 20 feet what a normal observer can see at 20 feet. Larger denominators reflect poorer acuity. For example, if an individual has 20/200 visual acuity, that means they can resolve at 20 feet what a normal individual can resolve at 200 feet; that is, objects have to be 10 times larger for that individual to see them than normal individuals require. The visual field is the portion of the environment an individual can see at a single look—that is, without moving their eyes around. The normal binocular visual field (ie, both eyes open) is about 180° wide (a semicircle) and 130° high. Certain diseases, for example, glaucoma and retinitis pigmentosa, cause a constriction of the visual field. This inability to see things in side vision is often termed *tunnel vision*. Twenty degrees diameter of visual field, a criterion for legal blindness, is slightly larger than a basketball held at arms length. Individuals with fields less than 20 degrees in diameter often have mobility problems, with the likelihood increasing as field size decreases.

†A list of major causes of deaf-blindness can be found at the Web site of the Oklahoma Deaf-Blind Technical Assistance Project: http://www.ou.edu/okdbp/mcauses.htm

a workable commercial version of such a device, which would facilitate face-to-face communication with those who are not familiar with the finger-spelling code as well as TDD and computer access.

Although communication is perhaps the main problem faced by the deaf-blind population, many other difficulties arise in travel, education, employment, and daily living activities for which solutions are (so far) few and far between.

Blind and Hard of Hearing

A population that has so far received less attention consists of blind individuals who also have a partial hearing loss. The size of this population is not known. A blind person who develops mild to moderate hearing loss (eg, through the natural aging process) may have amplification needs that differ from the needs of a person with normal eyesight and the same degree of hearing loss. Orientation and mobility skills taught to blind travelers depend heavily on the use of audition, and many of the cues are extremely subtle. For example, the tapping of the cane tip on the ground produces echoes reflected back from the environment which are analyzed by the blind traveler to inform him or her about nearby structures and obstacles. Reflections and shadows caused by ambient sounds are another subtle cue. Traffic sounds and their directionality are not only vital in street crossing but are a primary cue for maintaining a straight course, using the direction of receding traffic in the distance. Reverberation, or the reception of sounds after multiple reflections, is another important environmental cue used by blind persons in travel and many other situations. When the ability to receive and process these subtle cues is reduced by a hearing impairment, there is an impact on travel ease, confidence, and safety. There are techniques available for teaching orientation and mobility skills to blind or severely visually impaired travelers who also have a hearing loss. For example, Lolli and Sauerburger[10] emphasize making maximum use of residual vision as well as hearing, along with such strategies as soliciting aid, using dog guides, and following crowds when crossing streets.

Audiologists serving this population need to be aware of the importance of spatial as well as communication abilities when considering the alternatives for hearing aid fitting. Hearing aids are generally designed to optimize speech communication and may not be optimal for processing the types of spatial cues used by blind pedestrians. More work is needed in defining and solving the problems in this area, which would perhaps eventually result in hearing aids with user-selectable settings for optimizing communication and spatial cues. See the article by Simon and Levitt in this issue for a review of considerations of amplification needs of this population.

Deaf and Visually Impaired

Usher's syndrome affects approximately 3% to 6% of all deaf and hard-of-hearing children. Broughman et al[11] estimated the prevalence of Usher's in the United States at 4.4 per 100 000 or 12 000 cases. In this congenital condition, the hearing problem is combined with retinitis pigmentosa, a progressive visual disorder that causes degeneration of the sensory cells (photoreceptors) in the retina. As the disease progresses, patients generally lose their peripheral vision, leaving what is known as "tunnel vision." It is by the criterion of reduced field size, rather than visual acuity, that these individuals are classified as legally blind at least initially. The size of the residual visual field in Usher's patients is dependent on light level, with the residual visual field decreasing at lower light levels. Thus, it is crucial that these individuals be tested and function under good lighting. Furthermore, given the small size of their residual fields, only a small region of space can be seen at a time; communication may be improved when the speaker or signer is some distance away, for example, across the room. Only a fraction of these patients become totally blind in adulthood. If a deaf individual develops a sufficiently severe visual impairment, difficulties in independent travel may also occur. Deafness in combination with tunnel vision may lead to the need for orientation and mobility training to ensure that, for example, the individual scans the environment sufficiently to detect approaching vehicles when attempting street crossings.

A deaf individual who is accustomed to heavy reliance on vision for information processing and communication can experience significant difficulties in communicating when a visual impairment occurs. For example, sign language reading, which involves watching both face and hands, is likely to be considerably disrupted by central field deficits of the type that occur in the most common age-related visually disabling disease: age-related macular degeneration (AMD). This condition is characterized by a scotoma or "blind spot" at or near the center of vision, the fovea and macula, the region that

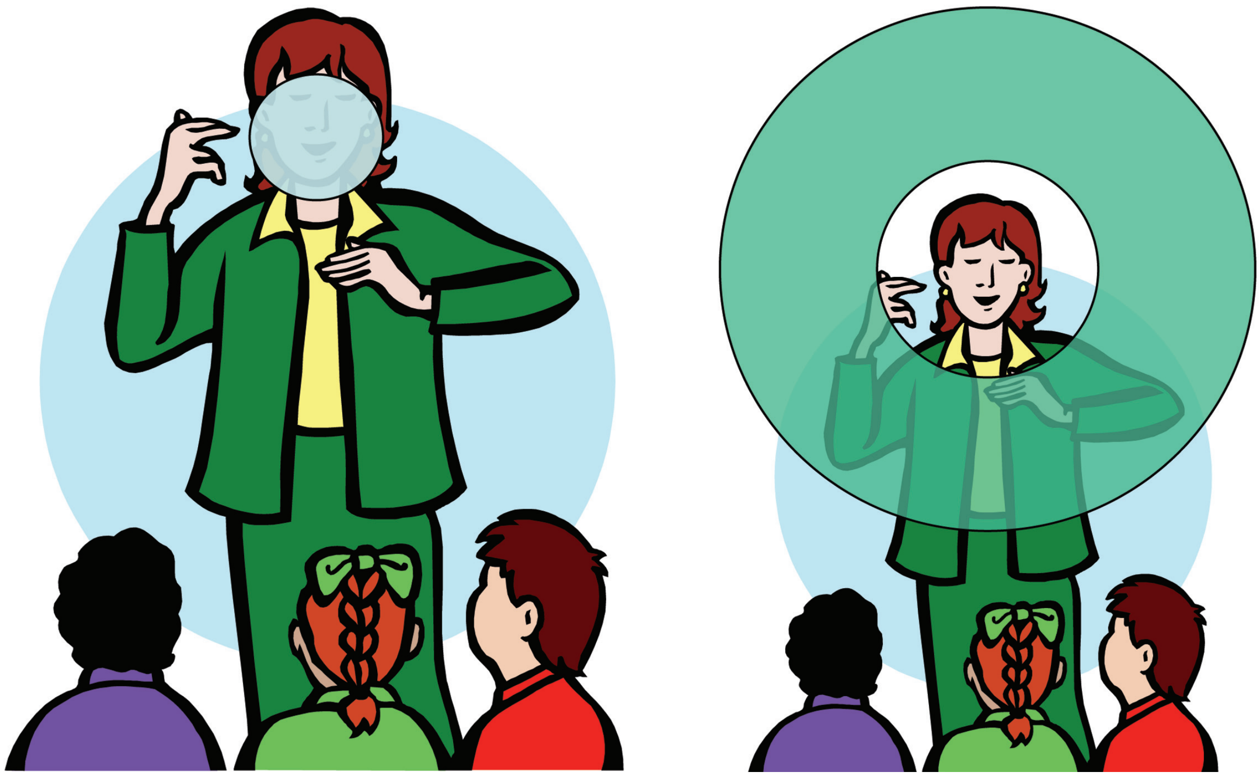

Figure 1. Left: Sign reading with a central scotoma. Right: Sign reading with restricted visual field (tunnel vision).

gives us fine-detail vision. This can obliterate the individual's high-resolution central vision and force him or her to use the peripheral areas of the visual field, which have lower resolution and contrast sensitivity. No research has been done on the impact of this problem on sign language reading, but the ability to concentrate both on the sender's face and on the movements of the hands is likely to be considerably disrupted, at least in severe cases (Figure 1).

Visually Impaired and Hard of Hearing

The largest segment of the dual sensory loss population consists of those with mild to moderate impairments in both vision and hearing. The 2 impairments increasingly occur together, especially in older people.

Brennan et al[12] reported that one fifth of a sample of individuals more than 70 years old had dual sensory loss, and the double loss was associated with increased difficulty in activities of daily living tasks. In the Blue Mountains Eye Study,[13] approximately 17% of older people were found to be visually impaired (better-eye acuity less than 20/40), and the same percentage was hard of hearing (average better-ear, pure-tone air conduction threshold greater than 25 dB at 500-4000 Hz). There is a strong relationship between the 2 impairments: for each 1-line (5-letter) reduction in best-corrected visual acuity (eg, 20/40 to 20/50), hearing loss prevalence increased by 18%.[14]

In normal aging, changes in the eye and visual system result in a degradation of vision. These changes include a loss of clarity of the optical system (lens, vitreous, and cornea), including cataracts, causing light to be scattered in the eye, a reduction in the ability of the retina to adapt quickly to different light levels, and the loss of the pupil's ability to dilate in dim light, which therefore admits less light. One result is reduced ability to see in low- or changing light conditions, in the presence of glare, or in "low-contrast" situations where the background is not much darker or brighter than the object of regard.[15-17]

In the Smith Kettlewell Institute (SKI) vision study,[15,16] older people were tested on a broad battery of vision tests and also underwent a screening test for hearing. The hearing test stimuli were four 40 dB HTL (hearing threshold level) test tones at 500, 1000, 2000, and 4000 Hz presented to each ear. We found that 14% of the sample of 828 older people (average age 75, range 58-104 years) had some visual impairment, defined as acuity worse than 20/40 (using both eyes and their normal glasses), and 12% had at least a moderate hearing loss (unable to hear any tone in either ear). About 5% of the sample had both vision and hearing loss. Only 8.6% with acuity better than 20/40 had hearing loss; however, this rose to nearly 29% among those with acuity between 20/40 and 20/70 and 37% among those with 20/70 or worse acuity. This association between hearing and vision losses reflects the decline of both with age. Indeed, when age is taken into account, the association between acuity loss and hearing loss is not statistically significant. However, even when age is accounted for, an association between vision and hearing is evident when vision is assessed using low-contrast letter targets, particularly at slightly reduced light levels (tested using the SKILL [Smith-Kettlewell Institute Low Luminance] Card,[18] an acuity test with black letters on a dark gray background designed to simulate vision under typical rather dim "living room" light levels). For each 0.3 log unit loss (ie, doubling of threshold letter size or 3 lines lost on the chart) of low-contrast acuity, low-contrast low-luminance acuity, or low-contrast acuity in glare, the odds of failing the hearing test increase by 30% to 50%. A strong association with hearing loss is also seen when low-contrast targets are presented in the presence of glare.

Clearly, the combination of mild to moderate hearing and vision deficits can have a synergistically negative effect. For example, Capella-McDonnall[1] analyzed 2001 National Health Interview Survey Data and found that persons aged 55 and older with dual sensory loss were significantly more likely than those with hearing loss alone to experience depressive symptoms. Hearing impairment also increases the dependence on the use of visual cues such as lip and facial movement as well as gesture, but as mentioned earlier, questions about vision are not often asked in hearing clinics, and vice versa. Many older people are not aware of how dependent they are on speechreading until their vision becomes impaired. As noted in the "Deaf and Visually Impaired" section above, when the most common of visually disabling diseases, namely AMD, becomes advanced enough to produce central blind spots or scotomata, it forces the patient to adjust his or her gaze above, below, or to one side of the object of interest, so that its image falls on a still-working portion of the retina. Unfortunately, these more peripheral areas of the retina have much less resolution and contrast sensitivity than the center or fovea. Thus, in lipreading and speechreading, the scotoma would obscure the view of the speaker's lips in normal gaze (Figure 1), so the viewer must shift his gaze to see them, resulting in lower resolution and a diminished ability to interpret the lip movements. To date, these and other increasingly common problems have been largely neglected by both researchers and practitioners.

In the visual perception of sign language, the situation is further complicated by the requirement to view the speaker's moving hands at the same time as the lips and face. For an individual with a central scotoma, it is not clear what the ideal gaze strategy under these conditions should be, and studies of eye movements under these conditions may help identify optimal eye gaze strategies that could be used in training and rehabilitation. In the future, technology that performs lipreading and reception of sign in an automated manner could be valuable in this situation.

Visual Assessment and Referral

Observations on Assessment

With the increase in the incidence of dual sensory loss, it is becoming important that more cross-disciplinary collaboration and referral is conducted between experts in vision and hearing research, practice, and rehabilitation. Although it may not be practical for clinicians in either of the 2 disciplines to become experts in the other as well, it is certainly possible to develop an increased awareness of the problems the patient may be facing as a result of his or her visual impairment in combination with the hearing deficit and to make appropriate referrals (eg, to a low-vision clinic). If no timely report is available from a vision care practitioner or low-vision clinic, it may even be possible to administer simple vision screening tests or questionnaires such as the National Eye Institute Vision Functioning Questionnaire.[19] This instrument, available online, consists of 25 items, with an appendix of an additional 13 optional items). The Vision Functioning Questionnaire asks how much difficulty the individual has doing a broad variety of tasks because of their vision. The questionnaire may provide the audiologist with a general sense of the visual status of the client.

The following information on vision assessment is in no way meant to take the place of appropriate referrals but is provided as a reflection of the authors' opinions on the importance of including nonstandard vision measures in assessing the likely impact of dual sensory loss.

The familiar letter acuity chart, the "gold standard" of visual function, only measures high-contrast

Figure 2. Left: Speaker against window. Right: Simulation of view of speaker against window by an individual with low-contrast vision equivalent to an average 80 to 85 year old (for simulations of other scenes see Brabyn et al[17]).

acuity, that is, the ability to identify black letters on a white background in good lighting. However, the standard acuity test often underestimates the problems actually faced by patients, because the real world does not consist of high-contrast targets in favorable lighting conditions. Rather, most of the visual world is low contrast, meaning that the differences between dark and light components are small. Low-contrast, poor lighting, and background glare have a disproportionate effect on visual performance in older people or younger individuals with eye disease, who often have excellent acuity. Even in normal aging, vision in low-contrast, low-light situations and glare declines much faster than regular acuity.[16,17] Thus, it is quite common for individuals to have good vision (20/20) on the standard acuity test as measured in the clinic but be significantly impaired when tested with low-contrast acuity charts (gray letters on white backgrounds)[20] or contrast sensitivity charts, such as the Pelli-Robson test.[21]

As an example, the task of speechreading inherently involves viewing a target with fairly low contrast (see Figure 2), unless the speaker is wearing bright (or dark) lipstick. Therefore, in screening for likely difficulty in speechreading, it may be beneficial to measure low-contrast vision as well as administering the standard high-contrast test. Several variations of low-contrast vision tests are available. A commonly used, commercially available test using large letters varying in contrast is the large wall-hung Pelli-Robson chart.[21] On this chart, letters at the top of the chart are of high contrast and decrease in contrast in small steps as one reads down the chart. The score is expressed as "log contrast sensitivity." Normal values are ≥ 1.85, and because the scale is logarithmic, a score of 0.85 would correspond to a reduction by a factor of 10 in one's ability to detect contrast. (For comparison, the legal 20/200 definition of blindness corresponds to a reduction in acuity by a similar factor of 10 from the normal value of 20/20.) It can be expected that any score ≤ 1.30 would contribute to difficulty with speechreading and many other tasks.[22] This level of contrast loss is common among older people. In our population,[16] 60% of those who were more than 85 years old performed at this level or worse on the Pelli-Robson Chart. Of those with significant contrast sensitivity loss, only 30% would be classified as visually impaired (20/70 or worse) by standard acuity measures.

What is more complex is the assessment of visual field deficits, particularly the central scotomata or blind spots that occur in AMD. The Amsler grid is an inexpensive test for mapping scotomata but is critically dependent on eye position being held steady at the center of the chart and thus requires a vigilant and skilled administrator. Recently, it has been suggested that a screening test for field loss can be carried out by instructing the patient to stare at the examiner's nose and asking if all other facial features look clear, distorted, or "missing."[23] This test has the advantage of allowing the examiner to monitor the patient's fixation during the task. In general, if field deficits are suspected, it is doubly important to ensure that the client is referred to a low-vision service for assessment and advice. In the presence of central scotomata, low-contrast vision in other parts of the field of view are usually also affected.

Cross-Disciplinary Collaboration and Referral

As noted earlier, it is of the utmost importance that referrals to appropriate optometric, ophthalmic, or low-vision services be made if a visual impairment appears to be present. Assessment of vision impairments that include losses in the central or peripheral field is especially difficult to perform, and if these are suspected, it is doubly important to ensure that the client is referred to a low-vision service for assessment and advice. Many individuals may need to be referred for professional orientation and mobility training to ensure that their travel remains safe. Agencies such as the American Foundation for the Blind maintain listings of low-vision services and orientation and mobility programs that can be searched by state from their Web site.

Similarly, vision care practitioners should also be aware of the combined impact of hearing loss. For example, as mentioned earlier, many older people unconsciously lipread as their hearing ability decreases and then find difficulty in this task when their vision function is reduced. Low-vision practitioners need to be sensitive to the needs of such tasks requiring both vision and hearing when assessing and prescribing aids and rehabilitation strategies for their patients.

Implications for Rehabilitation

In the earlier sections on the various subpopulations, a number of specific tasks were mentioned that are normally performed visually by hard-of-hearing persons and that may be affected by specific types of visual deficits. By analyzing the nature of the different types of common visual deficits, it is possible to gain a better understanding of which tasks may be affected and in what way. For example, the most common type of dual impairment is that which occurs in an older individual who may be losing partial vision and hearing ability. It may be very helpful to ask such clients questions about whether they are having trouble seeing details of lip movement or facial expressions. For those clients who use sign language and finger spelling, it would be important to ask whether they are having increased difficulty seeing finger and hand movement. One might also inquire whether clients are having increased difficulty accomplishing independent travel or other tasks, which normally involve both vision and hearing, because of changes in the ability to see or hear. Such questions may help point the way toward possible rehabilitation interventions (and more often to the need for more research!).

Many of the problems caused by dual sensory loss might be ameliorated by appropriate rehabilitation methods, aids, and techniques. Even though we run the risk of being too simplistic, we could consider the following examples:

- If practical, speakers and signers addressing hard-of-hearing or deaf individuals or audiences ought to stand against backgrounds and wear clothing to maximize the contrast of the hands.
- Wearing lipstick and brow pencil might increase the contrast of a speaker's or signer's face to aid communication for many individuals with impaired vision.
- Good lighting can work wonders for most low-vision conditions and may widen the visual field in retinitis pigmentosa and even make scotomata become partial or "disappear" in AMD. Try a strong light on the speaker or signer with no backlighting or glare.
- Some problems may be amenable to the use of optical aids such as a telescope; for some individuals, magnifying the speaker's face (or simply sitting closer) may help speechreading.
- Individuals with poor low-contrast vision and who are likely to be subject to glare should avoid facing a window while communicating in the daytime—they will find speechreading far easier if they are seated with the window behind them.
- Individuals with tunnel vision will find sign reading easier if they are seated further away from the signer so their remaining visual field includes the signer's hands, but not so far as to lose the ability to use face cues.
- Individuals with central scotomata will need to fixate eccentrically to view a speaker's lips and may need to practice different eye movement and scanning strategies to optimally receive sign language.
- Similarly, individuals with tunnel vision need to practice scanning around (eg, when traveling) to make sure they do not miss hazards they would otherwise detect auditorily (eg, approaching cars).

Conclusions

The combination of vision and hearing deficits can cause complex interactions with task performance because of the manifold dimensions of both types of deficit. Little formal study has been done of the practical problems resulting from these increasingly common dual sensory losses, but the rapid increase in the affected population makes it important to devote more attention to them.

There are a number of practical steps the audiologist can take when confronted with a client with dual sensory loss. Perhaps most important is asking all incoming patients whether they also have a problem with their vision and making sure they are referred to appropriate low-vision services.

Practitioners in both vision and hearing service provision can benefit their clients and their professions by devoting greater attention to the interactions between their patients' vision and hearing problems, including collaboration and mutual referrals.

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