Hearing Loss and Down Syndrome

Due to certain anatomical differences, people with Down syndrome are predisposed to experiencing difficulties in the middle ear and Eustachian tube, which often lead to hearing deficits. Auditory testing is strongly recommended for these individuals because up to 70 percent may present with hearing loss.1

Early diagnosis and treatment of hearing loss is critical to ensure that this condition does not contribute to or cause delays in speech and language, which could mistakenly be labeled as cognitive.

Causes
Hearing loss in the Down syndrome population primarily is associated with Eustachian tube dysfunction due to alterations in the skull base.2 The Eustachian tube leads from the middle ear to the back of the mouth and tends to be much smaller in people with Down syndrome. It is structured in a manner that can lead to an accumulation of fluid in the middle ear, causing infection and recurrent otitis media with effusion (OME).3 Conductive hearing loss can result from this condition.

Ear canals of people with Down syndrome, especially children, tend to be much smaller and narrower. Their canals often have blockage caused by cerumen, or ear wax. These blockages of the external ear canal also can cause hearing deficits.

In addition, a significant minority may have permanent, sensorineural deafness. Others experience processing difficulties that can affect auditory memory. Assessment for interventional purposes is critical.

Treatment
Management of hearing loss in patients with Down syndrome may involve one or a combination of hearing tests, medical intervention, amplification, auditory-verbal therapy and caregiver education.4

Current literature recommends that intervention occur early. Children should be tested for hearing loss every six to 12 months for the first 10 years of life.4 Once a child’s middle ear function is assessed, medical management may be suggested. This often includes surgical treatment with pressure equalization (PE) tubes, a procedure found to be very successful in restoring normal hearing levels.3 Procedures to clean and examine the outer ear often are necessary.2

If medical management cannot resolve the hearing loss, amplification options should be explored. These may include hearing aids—even for minor loss—as well as FM listening devices, sound field amplification units or other devices.

Education
Children may need to be taught how to hear after experiencing hearing loss. This involves lessons in incidental listening, auditory discrimination, and strategies for overcoming other problems related to background noise or attention.

Caregivers should know the signs and symptoms of OME, as well as other risks, and be aware of the potential for hearing deficits and speech-language difficulties.

References

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