

Hearing loss in CdLS

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Hearing loss has been widely reported in children with CdLS and can be conductive (trouble with sound getting to the hearing nerve) or sensorineural (a decrease in nerve hearing) in nature. Most published reports suggest that sensorineural hearing loss is most common in children with CdLS; however, the degree of loss is unclear.

Many families report that their young children have hearing loss, along with hearing test results that confirm the finding. Despite this, many families say that their child's hearing is better than the test results suggest and that results actually improve as the child gets older. The mechanism behind this improvement is unclear and is an area that needs exploration. While it has not been studied, a diagnosis of auditory neuropathy may be something to consider in children with these findings. To confirm this diagnosis, children need to be tested with otoacoustic emissions in addition to an auditory brainstem response test.

A common cause for hearing difficulties are ear infections. A recent study looking at the incidence of ear infections and conductive hearing loss in children ages 1 to 18 with CdLS found that 94 percent had ear infections with fluid behind the ear drum and associated conductive hearing loss. The study also found that the likelihood of having an ear infection was the same across the age range, suggesting that fluid behind the ear drums may persist in children with CdLS far longer than in typical children.

A number of treatment options for ear infections exist, ranging from no treatment to recurrent antibiotic therapy to ear tube placement. The presence of fluid without infection may be left to resolve on its own, or a nasal steroid may assist in opening the eustachian tube (the tube behind the nose that drains fluid from the middle ear).

Because of hearing concerns in children with CdLS, it's recommended that a newborn hearing screening be completed. In addition, repeat testing is needed when there is suspicion of hearing loss, recurrent ear infections or speech and language delays.

For children with known sensorineural hearing loss, it's recommended that they undergo regular hearing tests and evaluation by an otolaryngologist (also known as an ear, nose and throat surgeon). The frequency of hearing testing is based on the presence of hearing trouble and the changes seen in hearing over time, although yearly evaluation is common. It's also important to consider that children with sensorineural



hearing loss have an even harder time hearing when they have fluid behind the ear drum or an ear infection. For these children, some doctors recommend a more aggressive treatment for ear infections.

For children with moderate or severe nerve hearing loss, a trial with a hearing aid may assist in language development and/or understanding.

In order to learn more about hearing difficulties in children with CdLS, a number of children and adults have undergone computed tomography (CT) scans to look at the hearing bones and the middle and inner ear structure. Analysis of these films is ongoing but researchers are hopeful the information can provide insight into the nature and degree of hearing loss seen in individuals with CdLS.

Collection of hearing testing (behavioral audiograms, brainstem hearing testing and otoacoustic emissions) in children and adults with CdLS will help set up future studies to follow hearing over time and determine what the natural history of hearing loss is in CdLS.

