CEDARS-SINAI

“Newborn Hearing Screening”

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Significant hearing loss is one of the most common major abnormalities present at birth and, if undetected, will impede speech, language, and cognitive development. Early detection is of enormous benefit, because language development begins while the infant is still preverbal (not yet talking). Significant bilateral hearing loss is present in l to 3 per 1000 newborn infants in the well baby nursery population, and in 2-11 per 100 infants in the intensive care unit population. The loss is not total in most cases, but approximately 1 in 3000 children does have a hearing impairment significant enough to impact speech and language development, education, and social development. The incidence of approximately 3 of every 1000 births having a hearing loss makes this the most frequently occurring birth defect. Not surprisingly, therefore, congenital hearing loss is much more common (between 4 and 100 times) than other disorders for which babies are screened during the newborn period. The American Academy of Pediatrics supports the statement of the Joint Committee on Infant Hearing (1994), which endorsed the goal of universal detection of hearing loss in infants before 3 months of age, with appropriate intervention no later than 6 months of age.

The early detection of hearing loss (HL) greatly improves the quality of life of the individuals who are screened, diagnosed with hearing loss and then started promptly in early intervention. Infants whose HL is identified by six months of age demonstrate significantly better receptive and expressive language skill than infants whose HL is identified after six months.

Universal Newborn Hearing Screening programs target identifying permanent bilateral or unilateral, sensory or conductive hearing loss, averaging 30-40 dB or more in the frequency region important for speech recognition. This encompasses the frequency range of approximately 500- 4000 Hz.

TYPES OF HEARING LOSS

There are three types of hearing loss, distinguished by the anatomic basis of the deficiency.

Sensorineural HL is the most common cause of HL in the newborn period. It is caused by disease or defect in the sensory cells of the inner ear or the pathway of hearing along the acoustic branch of the VIII cranial nerve.

Conductive HL is due to disease or defects in the structure of the external or middle ear. In the newborn, this could include blood, mucous, or vernix caseosa that remains from birth.

Auditory neuropathy is a recently described disorder in which patients demonstrate a loss, absent or abnormal auditory brainstem response, with normal outer hair cell function.

CAUSES OF HEARING LOSS IN THE NEWBORN PERIOD

The era of molecular biology and genetics has led to a much greater degree of understanding about the physiology and pathophysiology of many disorders. Hearing and hearing loss are no exceptions. Particularly over the last five years, it has become clear that genetic causes of deafness occur more frequently than was ever previously imagined. Genetic causes of hearing impairment in the newborn period are at least as common as acquired forms of the disorder. It is worthwhile to consider each of these categories.

Genetic Causes of HL

The majority of early childhood hearing loss is now thought to be genetic in nature. Genetic research in the last five years has identified many genetic connections to hearing loss. 50% of prelingual hearing loss in the United States is genetic and only 30% of genetic hearing loss is syndromic (that is, there are other clinical features which are associated with the hearing loss). The net result is that close to one-third of all newborns with hearing loss will have no relevant infectious or environmental causes, no associated syndromic stigmata, and many cases will have no family history of hearing loss. Thus, to significantly increase the likelihood of early diagnosis and ensure appropriate treatment of significant hearing impairment, these infants need to be part of a universal screening program.

Acquired Hearing Loss:

It has been estimated that 50% of early childhood hearing loss in the United States is caused by infectious and environmental factors. Such factors include prenatal infections (toxoplasmosis, rubella, CMV, herpes simplex, syphilis), meningitis, low birth weight (<1500g), prematurity, hyperbilirubinemia (at serum levels requiring exchange transfusion), some medications, and mechanical ventilation with prolonged hyperventilation, ECMO, and admission to a NICU. In recently published data describing neurodevelopmental and functional outcomes of 1151 extremely low birth weight infants (400-1000g), the overall incidence of hearing impairment (defined as the need for the use of an amplification device to assist hearing) was 11%.

It is known that certain medications used to treat infections in infants pose a risk for causing hearing loss. When needed, medications are selected with great care, and always with consideration of "risk versus benefit." This includes weighing the risk of a side effect (such as a potential hearing loss) against the benefit of avoiding prolonged illness, serious impairment, or death which might occur without treatment. In addition to considering the risks of using or not using a medication, doctors undertake monitoring of the levels of medications in the body, to minimize the side effects which could occur. This is done by drawing blood periodically to ensure that the amount of medicine in the body is at a level expected to be effective for treatment and not a risk for side effects. Individuals can vary in their tolerance of medications and the ways in which their bodies metabolize them. This can, at times, lead to side effects, even with careful monitoring.

Hearing Loss Later In Infancy

While newborn screening is extremely effective, there are some patients who may develop hearing loss later in infancy, so complete reliance on a newborn screen is not a wise idea, if an infant or child appears to have difficulty hearing, or if other conditions are identified.

A particular viral infection which a fetus could acquire in utero, provides one such example. The major cause of sensorineural hearing loss (SNHL) in childhood is congenital infection with cytomegalovirus (CMV). The incidence of congenital CMV is as high as 1% in the United States. While the majority of cases are asymptomatic, as many as 30 to 65% of the symptomatic cases will have SNHL. This is a worrisome group of babies in terms of overall outcome, but they are, obviously, recognized at or around birth, and are evaluated. The more concerning group is that comprised of the babies without symptoms at birth, who are at a lower, but still significant risk of hearing loss (8 to 15%) during childhood.

Another system of screening is needed to help identify these babies. As techniques improve, it may be warranted to add CMV screening to routine newborn screening, so that these babies can be identified early after birth, and routine periodic hearing screening performed on this high-risk group.

To learn more about newborn hearing screening, causes of hearing loss in infants, or aminoglycoside (Gentamicin) hearing loss, go to the Harvard Medical School continuing medical education website www.theanswerpage.com

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