CD Summary

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CENTER FOR DISEASE PREVENTION & EPIDEMIOLOGY • OREGON HEALTH DIVISION

NEWBORN SCREENING IN OREGON POISED TO BREAK THE SOUND BARRIER

NEWBORN HEARING SCREENING will start July 1, 2000 for most of Oregon's newborns. Oregon's 71st Legislative Assembly adopted House Bill 3246 in August 1999. With the passage of House Bill 3246, Oregon joined twenty-four other states with legislation requiring newborn hearing screening.

Hospitals with more than 200 live births per year are required to provide hearing screening to all babies born in their facility, preferably before discharge and at least within one month of age. Thirty-nine hospitals, delivering over 90% of Oregon's babies, fall into this category. No newborn may be denied screening because of inability to pay. Parents may request a religious waiver.

This *CD Summary* discusses the rationale for screening newborns for hearing loss and the implications of implementing this program, both for Oregon's children and health care providers. Listen up, because Oregon's primary care providers will play a critical role in ensuring the success of this legislative initiative.

WHY NEWBORN SCREENING?

Hearing loss is among the most common congenital disorders and far more common than the other disorders screened for at birth.[1] The prevalence of severe newborn and infant hearing loss is estimated to range from 1.5-3 per 1000 live births, depending on the criteria used.[2,-4] A hearing loss of 30 dB HL and greater in the frequency region important for speech recognition (approximately 500-4000 Hz) will interfere with the development of speech and language.[5]

The prevalence of permanent unilateral hearing loss may be 35-48% that of bilateral loss.[6,7,8] Children with unilateral hearing loss are ten times as likely to be held back one grade compared to a matched control group of children. Combining severe and moderate, bilateral and unilateral hearing loss in newborns, rates may approach 4.5 per 1000 live births.

With approximately 46,000 babies born in Oregon each year, between 80-200 children are born deaf or hard of hearing. Unfortunately, we hear that fewer than 15 children under the age of 6 months were enrolled in early intervention programs for deaf or hard of hearing children during 1999.

The age of identification and intervention is the second most important determinant of the extent of language development in deaf or hard-of-hearing children; cognitive ability is first. Those who are identified before 6 months of age demonstrate significantly better receptive and expressive language skills than children whose hearing losses are identified later, irrespective of method of intervention .[9]

Behavioral methods of identification, including use of developmental milestones or gross confrontational testing (loud noises) are insensitive and inaccurate, especially at these age levels. Infants may have developmentally and educationally significant hearing loss without being completely deaf. Parental and provider uncertainty have led to unfortunate delays in diagnosis. Without newborn hearing screening, the average age of identification has been 2.5 years.

Earlier identification and intervention is now possible with newer technologies. Newborns can be screened using one of two physiologic methods designed to detect hearing loss of 30 dB HL and greater in the frequency region important for speech recognition.

HOW TESTING WORKS

There are two technologies used to screen newborns for hearing loss. Otoacoustic emission (OAE) testing detects a response to stimuli from the cochlear hair cells. Automated acoustic brainstem responses (AABR) relies on an intact central auditory neurological system to detect a response to a sound stimuli. The sensitivity of these tests is near 100% and false negative test results are rare. Both methods out-perform behavioral methods, which are unreliable in infants less than 6 months of age.

Results of physiologic testing are reported as "pass" if no abnormalities are detected or as "needs further testing." The false-positive rate depends on a number of variables, including age (in hours) at which the testing occurs, the technology chosen and screener experience. Accuracy is improved if the screening test is repeated at least once.

The positive predictive value of the test (the number of true positive results divided by the number of total positive results — or how often a child with a non-passing result turns out to have a hearing loss) is low (from 5-19%, depending on screener training and technology chosen) but compares very favorably with other newborn screening tests.[1]

Any newborn who does not pass the screening test should be referred for definitive diagnostic testing, performed before 3 months of age by a licensed audiologist or neurologist with experience in testing infants.

It is the Health Division's responsibility to prepare one list of sites capable of providing screening services and another list of diagnostic testing sites; parents should be given the appropriate list before discharge. Both lists will be available on our website, *http://www.ohd.hr.state.or.us/ccfh/*, no later than July 1.

To establish criteria for placement on the diagnostic list, the Health Division has established a protocol for diagnostic testing; the protocol includes tympanometry (to rule out middle ear effusion) and otoacoustic emissions re-screening, followed by threshold-search and frequency-specific ABR testing.

Parents and primary care providers must be provided with the results of the screening within ten days of the test. Most hospitals will inform the parents of the results before discharge. The method for notification of primary care providers may vary by hospital. If you have any questions about hospital procedures, speak to the nurse manager of the perinatal unit.

Hospitals or birthing centers with fewer than 200 live births per year are exempt from screening. These hospitals, however, must provide to the parents information (developed by the Health Division) on the importance of newborn hearing screening and a list

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of sites able to perform the test. Twenty hospitals and birthing centers fall into this category; some have already chosen to provide screening services.

Some hospitals may make arrangements before (or at the time of) discharge for diagnostic follow-up for babies who need further testing, but others will leave that to the parents to arrange voluntarily.

NOW HEAR THIS!

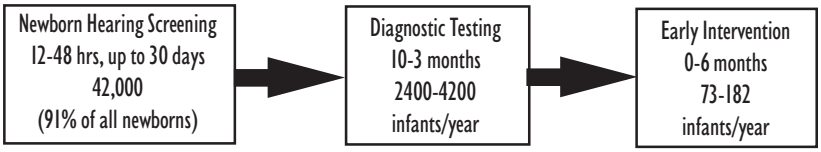
In Oregon, the baby's primary care provider has a critical role to play. Other states have found that a large proportion of babies referred for diagnostic testing do not get the necessary follow-up diagnostic testing and intervention. The Oregon legislature was silent on this issue. Unlike traditional newborn metabolic screening, there will be no statewide system for tracking and monitoring a child's progress through the process from birth to screening, to diagnosis and finally to enrollment in early intervention. It will fall to you.

If this sounds like a big task, it is. Screening is only of value if identified children get diagnosed and enrolled in early intervention.

PRIMARY CARE PROVIDERS' ROLE

Primary care providers will need to ensure that every baby in their care has had a newborn hearing screening test. If a baby has not, it will fall to the health care provider to explain the importance of identifying hearing loss and to encourage the parent to get screening.

If the baby has been tested and needs further testing, you must walk a careful line between generating unnecessary fear – as a screening test does not mean that a hearing loss exists – and complacency. The health care provider must simultaneously reassure the parents and stimulate sufficient motivation to assure that the baby receives the diagnostic testing needed.



The health care provider should also examine any infant who did not pass his/her hearing screening test for signs of an underlying etiology, including genetic or acquired causes. It is estimated that half of all cases of childhood deafness are hereditary, primarily inherited as an autosomal recessive trait, and that definable syndromes account for approximately 20% of congenital hearing loss, although associated findings may be subtle (e.g preauricular pits or skin tags).[10,11] It is worthwhile to perform a complete history and physical exam on all infants diagnosed with significant hearing loss.

The American College of Medical Genetics issued a statement in January 2000 recommending that, given the large number of syndromic forms of hearing loss, the genetic complexity of heritable forms of hearing loss, and the importance of an accurate genetic diagnosis, all children with confirmed hearing loss be referred for evaluation and genetic counseling.[12]

Once a child has been identified as deaf or hard of hearing, the health care provider should encourage the family to apply for early intervention services — either through the eight Regional Programs for Deaf or Hard of Hearing Children or one of Portland's two private facilities — (also listed on our website), as soon as possible and then monitor the baby's progress in accessing and receiving such services.

If you have any questions of us, we're all ears! Please contact Martin Lahr, MD (503/731-4399 or Ken D. Rosenberg, MD (503/731-4507) at the Health Division.

REFERENCES

1. Mehl AL, Thomson V. Newborn hearing screening: the great omission. Pediatrics. 1998;101(1). URL: *http://www.pediatrics.org/cgi/content/full/101/1/e4.*
2. Watkin PM, Baldwin M, McEnery G. Neonatal at risk screening and the identification of deafness. Arch Dis Child. 1991;66:1130-1135.
3. Parving A. Congenital hearing disability: epidemiology and identification; a comparison between two health authority districts. Int J Pediatr Otorhinolaryngol. 1993;27:29-46.
4. White KR, Behrens TR. The Rhode Island hearing assessment project: implications for universal newborn hearing screening. Semin Hearing. 1993;14:1-119.
5. Joint Committee on Infant Hearing 1994 Position Statement. Pediatrics. 1995; 95:152-156.
6. Broohhouser PE, Worthington DW, Kelly WJ. Unilateral hearing loss in children. Laryngoscope. 1991;101:1264-1272.
7. Kinney C. Hearing impairments in children. Laryngoscope. 1953;63:220-226.
8. Watkin PM, Baldwin M, Laoide S. Parental suspicion and identification of hearing impairment. Arch Dis Child. 1990;65:846-850.
9. Yoshinaga-Itano C, Sedey AL, Coulter DK, Mehl AL. Language of early- and later-identified children with hearing loss. Pediatrics. 1998;102:1161-1171.
10. Tomaski SM, Grundfast KM. Hearing loss in children: a stepwise approach to the diagnosis and treatment of hereditary hearing loss. Pediatr Clin North Am. 1999;46:35-48.
11. Brookhouser PE. Pediatric Otolaryngology: Sensorineural hearing loss in children. Pediatr Clin North Am. 1996;43:1195-1216.
12. American College of Medical Genetics. Statement of the ACMG on Universal Newborn Hearing Screening. January 2000. URL:*http://www.faseb.org/genetics/acmg/pol-35.htm.*