DIAGNOSTIC TESTS

Newborn hearing screening

Harvey Coates, Senior Ear, Nose and Throat Surgeon, Princess Margaret Hospital for Children and Clinical Associate Professor, The University of Western Australia, and Kim Gifkins, Speech Pathologist, Telethon Institute for Child Health Research, Perth

SYNOPSIS

Advances in audiological testing equipment and techniques allow accurate hearing screening of the newborn, using either otoacoustic emission screeners or automated auditory brainstem evoked response audiometry. Hearing screening lowers the age of diagnosis of permanent hearing loss. Evidence also indicates that early detection and management of hearing loss leads to improved speech, language and educational outcomes.

In Australia, newborn hearing screening is not widely available. Screening is available to babies 'at risk' of hearing loss, to all babies born in hospitals where the West Australian screening program is implemented, and is either being trialled or developed in other states. Awareness of the benefits and limitations of newborn hearing screening will enable the healthcare professional to support children with a hearing loss, and their families, so that they are able to maximise their potential.

Index words: audiology, deafness.

(Aust Prescr 2003;26:82–4)

Introduction

Hearing screening for congenital sensorineural hearing loss has been called 'the great omission'.[1] The incidence of congenital sensorineural hearing loss in the newborn population is greater than the combined incidence of all the metabolic conditions that we currently screen for with blood tests.[2] The prevalence of congenital bilateral permanent hearing loss is approximately 1 per 1000 live births.[2,3]

In the USA, the Joint Committee on Infant Hearing has recommended that every newborn infant should be screened.[4] Most American states have introduced routine newborn hearing screening, and screening programs are also being implemented in Europe and throughout the UK. However, a review of critical studies on newborn hearing screening could not make a recommendation for or against screening because of insufficient evidence.[5,6]

A review of the evidence for universal newborn hearing screening shows that the technologies used (otoacoustic emission (OAE) and automated auditory brainstem response (AABR) testing) are accurate tests for detecting congenital hearing loss. In Australia, the average age of detection of sensorineural hearing loss remains beyond two years. The age of diagnosis can be reduced by universal screening of the newborn.[6,7]

This is important because early intervention results in significantly better speech and language outcomes than delayed intervention.[8,9] The critical age to commence intervention may be as early as six months.[8]

Newborn hearing screening in Australia

Throughout Australia, to a varying degree, babies 'at risk' are screened for sensorineural hearing loss. Babies who are 'at risk' have one or more of the established risk factors for hearing loss.[4] However, as studies have indicated that approximately 40% of all children ultimately identified with sensorineural hearing loss do not have an established risk factor[7], the efficacy of 'at risk' screening is limited.

The first large-scale newborn hearing screening program in Australia was established in 2000. This program screens all babies born at five of the major birthing hospitals in Perth, usually before they are discharged. To date, over 25 000 babies have been screened and the results for the first 12 708 babies were published recently.[10] In this group, 99% had a pass response in both ears at either the initial or follow-up screen. Only 23 babies were referred for audiologic assessment, with nine babies being diagnosed with bilateral permanent hearing loss. Results suggest that in the well baby population, 2702 babies need to be screened to detect one additional case requiring intervention.

Implementation of universal newborn hearing screening in other states of Australia is either under consideration[11] (Queensland, Northern Territory and Tasmania) or has recently commenced (New South Wales, Victoria, South Australia and the Australian Capital Territory).

Screening technology

Recent technological advances allow the detection of possible hearing loss within the first days of life. The increasing simplicity of operating the equipment enables non-specialist staff to screen for hearing loss. It is difficult to determine the cost of screening due to the high capital costs of establishing a screening program. In Western Australia the cost is approximately $35 per test.

Otoacoustic emission testing

In the healthy cochlea, vibration of the hair cells in response to noise generates acoustic energy, known as otoacoustic

emissions. Otoacoustic emission testing therefore measures the integrity of the inner ear. A lightweight probe is placed in the ear canal and generates wide-band 'clicks' (see Fig. 1). Acoustic energy produced in response to the clicks is detected by a microphone within the probe. Automated OAE screeners display the results of the test as either 'pass' or 'refer', requiring no test interpretation by screening personnel.

The test takes between one and five minutes in ideal conditions, with optimal test techniques. In practice, the average total time for testing, including discussion of the procedure with the parents, settling the baby, performing the test and recording the results, may be between 15 and 20 minutes.

Automated auditory brainstem response testing

This measures not only the integrity of the inner ear, but also the auditory pathway. It can therefore detect the rare condition of auditory neuropathy, in children who are deaf but have normal otoacoustic emissions (because the cochlea is normal).

The stimulus (either clicks or tones) is presented using either earphones or an ear canal probe, and the electrophysiological response from the brainstem is detected by scalp electrodes (see Fig. 2). Automated devices allow screening to be performed by non-specialists. Responses from a large number of stimulus presentations are averaged and the automated screener uses a response algorithm to produce a 'pass' or 'refer' result. The 'pass' level is set at about 35 decibels.

This test takes 15–20 minutes, but once again this time may be longer if a child is restless, and does not include time for discussion and preparation before the test.

Screening protocols

The protocols of established newborn hearing screening programs throughout the world may use OAE only, AABR only, or a combination of technologies. For example, in the West Australian program, in well babies, an OAE test is performed initially, followed by an AABR test if a 'pass' response is not obtained in both ears. Babies who fail the AABR test are followed up and tested again with either OAE or AABR 3–4 weeks later. Children failing the follow-up screen are then referred for full audiological diagnostic testing.[10]

In the neonatal intensive care unit, protocols differ from those in the well baby nursery. Screening may be delayed until the baby is well enough. Although the condition of auditory neuropathy is rare in the well baby population, it can account for approximately 10% of hearing loss in the neonatal intensive care unit so virtually all neonatal intensive care unit screening programs use AABR.

Sensitivity and specificity

Sensitivity and specificity rates are affected by the screening protocol used, the population screened (well babies or neonatal intensive care unit infants), and other test variables. In general, all methods of newborn hearing screening show a screen specificity of greater than 90%. Most of the infants who screen positive for hearing loss are found to have normal hearing on further diagnostic testing.[7] Estimates of sensitivity for OAE range from 80–98% and for AABR from 84–90%.[6,7]

Test limitations

Both the OAE and the AABR screen require a quiet baby and a quiet testing environment. Restlessness can affect the time taken for the test, or may result in the test being discontinued. OAE relies on a functional outer, middle and inner ear, and AABR a functional outer, middle and inner ear, and lower auditory pathway. These screening tests are not designed to detect central hearing impairment (where there is hearing loss secondary to the dysfunction of the pathways from brainstem to the auditory cortex).

As the stimuli for both tests are introduced via the external ear canal, debris in the canal or middle ear fluid can affect the accuracy of the test. In particular, OAE testing may be affected by amniotic fluid in the ear canal when testing is conducted in the first 48 hours following birth. This may account for some false positive results.

Fig. 1

Neonatal hearing screening utilising otoacoustic emission screener

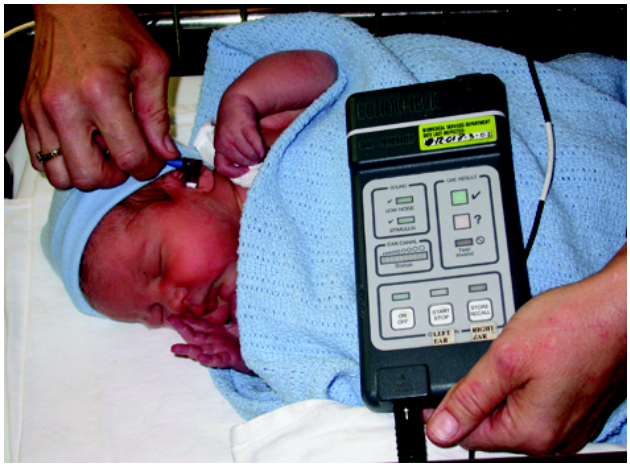

Fig. 2

Neonatal hearing screening utilising automated auditory brainstem response screener

Photographs reproduced with permission from the Public Relations Department, King Edward Memorial Hospital for Women, Perth

Parents' views of screening

Screening is acceptable to parents even if it may result in increased parental anxiety. In the West Australian program screening was well accepted by parents, with only 0.4% refusing screening.[10] Many tests resulting in a 'refer' outcome are ultimately false positives. This potentially can lead to increased levels of anxiety until diagnostic tests are performed, although some mild anxiety may remain even after a normal result.[12] To allay anxiety parents must be provided with accurate information regarding the screening, effective counselling and rapid follow-up.

Follow-up services for hearing loss

It is important that children and families are able to access 'habilitation' and intervention services as soon as possible after the diagnosis of permanent hearing loss. This process usually involves referral to the following specialists:

- ear, nose and throat surgeons
- geneticist
- Australian Hearing (provides audiology services, supplies hearing aids at minimal cost and provides monitoring of the child's hearing throughout childhood)
- early intervention services (in the larger cities, there are education and intervention centres for the hearing impaired, as well as a number of community support groups; in rural areas, a visiting teacher of the deaf service is often available).

The role of the general practitioner

General practitioners may play a role in educating and supporting parents and families about newborn hearing screening, both in the antenatal period and after birth. It is crucial that the general practitioner is kept fully informed of the results of screening. In the West Australian program, the newborn screening results are recorded in the child's personal health record.

The general practitioner has an important role in dealing with the implications of the diagnosis of hearing loss and the ongoing management issues for both the child and the family. Children with a sensorineural hearing loss should be monitored closely for middle ear conditions throughout childhood, so that conductive hearing loss resulting from otitis media does not further compromise hearing levels. Hearing aids may also predispose the child to otitis externa. The general practitioner may also play a role in promoting acceptance of hearing aids, encouraging consistent wearing of aids, and providing information regarding early intervention services.

Neonatal hearing screening will not detect all cases of congenital hearing loss – it only provides an indication of the baby's hearing at the time of the screening. Mild hearing losses and hearing losses outside the main speech frequencies may not be detected. Hearing impairment may develop after the neonatal period[3], and therefore it is crucial for the general practitioner to encourage parents to continue to have their child's hearing checked. The general practitioner should maintain a high index of suspicion if there are manifestations of hearing loss such as speech and language delay. Any parental concerns regarding children's hearing should also be thoroughly investigated.

Conclusion

The technology and expertise for implementation of neonatal hearing screening is available, accurate and acceptable. Australia also has excellent hearing services, including Australian Hearing, cochlear implant technology and early intervention programs. Early identification of children with a hearing loss, so that access to services can be commenced as soon as possible, will enable improved speech, language and educational outcomes.

E-mail: harveyc@cyllene.uwa.edu.au

ACKNOWLEDGEMENTS

We would like to acknowledge the excellent help of Ms Helen Bailey, Coordinator of the WA Newborn Hearing Screening Program. The support of the WA Department of Health, the Telethon Institute for Child Health Research, King Edward Memorial Hospital, Princess Margaret Hospital for Children, and the Garnett Passe and Rodney Williams Memorial Foundation in the establishment of the WA Newborn Hearing Screening Program is gratefully acknowledged.

REFERENCES

1. Mehl AL, Thomson V. Newborn hearing screening: the great omission. Pediatrics 1998;101:E4.
2. Mehl AL, Thomson V. The Colorado newborn hearing screening project, 1992-1999: On the threshold of effective population-based universal newborn hearing screening. Pediatrics 2002;109:E7.
3. Fortnum HM, Summerfield AQ, Marshall DH, Davis AC, Bamford JM. Prevalence of permanent childhood hearing impairment in the United Kingdom and implications for universal neonatal hearing screening: questionnaire based ascertainment study. Br Med J 2001;323:536-40.
4. Joint Committee on Infant Hearing. Year 2000 position statement: principles and guidelines for early hearing detection and intervention programs. Am J Audiol 2000;9:9-29.
5. Newborn hearing screening: recommendations and rationale. U.S. Preventive Services Task Force. Rockville, MD: Agency for Healthcare Research and Quality; 2001. http://www.ahrq.gov/clinic/3rduspstf/newbornscreen/newhearrr.htm
6. Thompson DC, McPhillips H, Davis RL, Lieu TL, Homer CJ, Helfand M. Universal newborn hearing screening: summary of evidence. JAMA 2001;286:2000-10.
7. Davis A, Bamford J, Wilson I, Ramkalawan T, Forshaw M, Wright S. A critical review of the role of neonatal hearing screening in the detection of congenital hearing impairment. Health Technology Assessment 1997;1. http://www.hta.nhsweb.nhs.uk (cited 2002 Sept 19)
8. Yoshinaga-Itano C, Sedey AL, Coulter DK, Mehl AL. Language of early- and later-identified children with hearing loss. Pediatrics 1998;102:1161-71.
9. Moeller MP. Early intervention and language development in children who are deaf and hard of hearing. Pediatrics 2000;106:E43.
10. Bailey HD, Bower C, Krishnaswamy J, Coates HL. Newborn hearing screening in Western Australia. Med J Aust 2002;177:180-5.
11. Wake MA. Newborn hearing screening: decision time for Australia. Med J Aust 2002;177:172-3.
12. Poulakis Z, Barker M, Wake M. Six month impact of false positives in an Australian infant hearing screening programme. Arch Dis Child 2003;88:20-4.

Patient support organisations

Deafness Forum and Australian Hearing

See page 87