Review Article

Universal newborn hearing screening

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ABSTRACT

Hearing loss is an "invisible" handicap. If detection is delayed or denied, significant speech, language, social and emotional delay may result. Delayed identification of hearing loss in children will lead to missing a lot of sound input which can in turn affect their communication, learning, behavior and emotional development as well as future performance in school and work. The world literature on programs for neonatal hearing screening comes mainly from countries such as the United States of America where most births take place in hospitals. Nowadays in the Kingdom of Saudi Arabia, most births take place in birthing hospitals. It is therefore possible to apply the program in these hospitals, provided training of the staff and the necessary equipment is available.

Saudi Med J 2003; Vol. 24 (3): 245-247

Hearing loss is an "invisible" handicap. It is a frequently occurring abnormality present at birth or acquired later. It is often not suspected or diagnosed until a child fails to develop speech around the age of 2 years. If detection is delayed or denied, significant speech, language, social and emotional delay may result.[1] Delayed identification of hearing loss in children will lead to missing a lot of sound input which can in turn affect their communication, learning, behavior and emotional development as well as future performance in school and work.[2,3] In an epidemiological study of hearing impairment (HI) in children in Riyadh, Kingdom of Saudi Arabia (KSA) the prevalence of HI was found to be 7.5% of the children surveyed; 2.6% of them with sensorineural hearing loss which is not amenable to treatment.[4] In another survey completed in the year 2000 taking samples from randomly selected areas of KSA, approximately 10,000 children aged up to 15 years were screened. The prevalence of HI was found to be 13% and most causes were treatable and could have been prevented. The survey also disclosed that 1.5% of the children in KSA suffered from sensorineural permanent hearing loss which was either unilateral or bilateral.[5] Most of the affected children were of a young age. Fifty percent of the affected children had a hereditary risk factor; the rest were caused by infection that could be treated before damage of the hearing organs occurred. In around 30% of the hearing impaired children, no cause could be identified. Of those with unknown cause for their hearing problem, some etiological factors could be identified, using recent laboratory investigations, clinical examination and examination of the other organs namely, the eyes.[6] Most of the affected children were diagnosed late, and therefore little or no intervention was carried out. Early intervention, using hearing aids, hearing education, cochlear implants and speech training are unfortunately scarcely available in this country. Despite this, early identification is very important to provide the children with the sound environment, make parents and educators aware of the need of these hearing impaired children and direct the responsible people to provide support, financial and moral for those people and their families. The universal newborn hearing screening (UNHS) program is now compulsory in some developed countries for early detection of hearing loss.

In 1993, a consensus of statement on the early identification of hearing impairment in infants and young children was published in the United States of America (USA) by the National Institute of Health.[7] The panel concluded that all infants admitted to intensive

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care units should be screened for hearing loss prior to discharge and universal screening should be implemented for all infants within the first 3 months of life. In 1994, the joint committee on infant hearing in the USA stated that all infants with hearing loss should be identified before 3 months of age and receive intervention by 6 months.[8] The World Health Organization (WHO)[9] recommended that a policy of universal neonatal screening be adopted in all countries and communities with available rehabilitation services and that the policy be extended to other countries as rehabilitation services are established. Children born in hospitals with a universal newborn screening program for hearing loss performed much better than their peers who were born in hospitals that did not have the screening program; on parent reports of expressive language, receptive language, and expressive vocabulary; and on measures obtained form videotapes of the child interacting with their caregiver (total number of words produced and number of different words produced). The children in the screened group also did significantly better than their unscreened peers in speech measures of number of consonant types, number of initial consonant blends, and intelligibility.[10] The world literature on programs for neonatal hearing screening comes mainly from countries such as USA where most births take place in hospitals. Nowadays in KSA, most births take place in birthing hospitals. It is therefore possible to apply the program in these hospitals provided training of the staff and the necessary equipment is available. Technological advances have made newborn hearing screening simple and effective for example, using electrophysiologic procedures to screen newborn singly or in combination namely, automated auditory brainstem response, and otoacoustic emissions. This screening equipment allows clinicians and non-clinical personnel to conduct screening provided these staff are given instructional courses and training. The cost of identifying a newborn with hearing loss is less than one-tenth of the cost of identifying a newborn with phenylketonuria, hypothyroidism or sickle cell anemia.[11] The previously used methods namely, behavioral hearing screening, distraction testing are simple and give an idea of hearing problems as does Ewing's method or its modification using recorded sound produced via loud speakers, called the Compact Amsterdam Pediatric Audiometric Screener (CAPAS). However, these methods are usually applied with infants and children above 9 months of age and approximately 30% of those tested need to be re-tested or to be referred for further investigations.[12]

Screening program. The objectives of the proposed program are: 1. To screen newborn babies in birthing (maternity) hospitals. 2. Compulsory hearing screening of infants and children attending well-baby clinic and health centers for immunization. 3. Early diagnosis during the first 3 months and reduction of the prevalence rate of severe hearing impairment in older children and early intervention not later than 6 months. This can be achieved by taking care of the children diagnosed with hearing impairment, and to improve the quality of life for them in addition to prevention of any complications that may arise (medical treatment of any disease, auditory training, provision of hearing aids and rehabilitation). 4. To have a registry of diagnosed cases of hearing impairment. 5. Training of staff of birthing hospitals and health care providers in well-baby clinics to be able to use equipment for screening.

Goal and benefits. The achieved goals and benefits of the programs are. 1. Early detection of hearing impairment during the first 3 months. Clinical studies indicate that early detection of hearing loss followed with appropriate intervention minimizes the need for extensive rehabilitation during the school years.[13] 2. To start early intervention not later than 6 months of age with amplification. Those with hearing loss can be mainstreamed in regular elementary and secondary education classroom provided they are fitted by as young as 4 weeks of age.[14] 3. To identify barriers to parents acceptance of hearing loss. 4. To stress the need for more audiologists and speech pathologists. 5. To have centers for hearing and ear care fully equipped and staffed at least in and around major cities in KSA.

Alternative detection program. Screening of infants with risk indicators and those in the intensive care units is an alternative way to screen for hearing impairment. But it was reported that 40%-50% of the infants with hearing loss would have been missed if the identification of risk indicators had been the screening method rather than direct testing.[15,16]

Data from other studies indicate that asymptomatic congenital human cytomegalovirus (HCMV) infection could account for hearing loss in children lacking risk factors. So screening newborns for HCMV rather than screening for hearing loss could be carried out, as children at risk for delayed onset hearing loss and visual problems could be identified.[16] The same studies show that a positive family history is the most likely missed risk factor. As reported before, approximately 50% of the cause of hearing impairment of children is genetic in origin.[3] Therefore, efforts should be directed at educating the primary health or care providers on the signs and symptoms of hearing loss in infancy when universal hearing screening is adopted. Those who need referral or re-testing may not report back to the same hospital, but they will report to health centers for immunization. A survey carried out by the same author[16] of primary care practitioners in the state of Michigan indicates a perceived knowledge deficit and a wide spread desire to learn about hearing loss and many respondents indicated that such information was missing in their training program.

In a study in KSA for training of health care workers[17] the authors conclude that: "We recommend that all general practitioners and pediatricians should have training in otoscopy by spending some of their training period in the Ear, Nose and Throat Department. They should also be trained in how to remove wax in order to

be able to visualize the drum. Nurses or health care providers especially those working with pediatric populations, should be trained and supervised in dealing with simple ear problems and referral of those who need further management. This will reduce the burden on hospital referrals".

The babies delivered normally (vaginal delivery) should be tested on the first day as most mothers have a short stay in hospital while those delivered by cesarean section tend to stay longer, so babies can be tested on the second or third day. As there is high estimated rate of babies lost to follow-up (40%-70%),[18] further screening should be carried out in well baby clinics (health centers). False positive cases should be referred for further evaluation in a hospital with audiological facilities.

In conclusion, the outcomes associated with early identification of hearing impairment are many: 1. Infants and children identified with hearing loss can be fitted with amplification as early as 4 weeks of age. They demonstrate significantly better language, speech and social emotional development after 6 months than children identified later.[14] 2. Early detection of hearing loss followed with appropriate intervention minimizes the need for extensive rehabilitation during the school years. 3. The better the language development, the less parental stress there is, and the better personal-social development of children. 4. Early identification is the initial step that leads to early and appropriated intervention. 5. Hearing aid (HA) fitting is always delayed due to the ignorance of the physicians, poor income of family who cannot afford to buy the HA or even repair it, failure of the health authority to provide HA free of charge and unawareness of the families.

Difficulties in applying universal newborn hearing screening. 1. Parental acceptance of the program and their cooperation especially for follow-up. 2. Shortage of qualified audiologists especially pediatric oriented ones. 3. Primary care and well baby clinic physicians do not know enough about screening, diagnosis, signs and symptoms of hearing loss and intervention. Training and courses should be organized. 4. To develop evaluation protocols to overcome these difficulties a) more audiologists should be encouraged to work in these programs. Their role should be recognized and given incentives; b) public awareness and parental education. 5. There will be an increased demand for qualified audiologists and personnel to provide identification and intervention.

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