Identifying congenital hearing impairment. Personal experience based on selective hearing screening

La diagnosi della sordità congenita. Esperienza personale basata sullo screening audiologico selettivo

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Summary

If all degrees of permanent uni- or bilateral hypoacusis are taken into consideration, hearing impairment is the most common congenital disease. Early detection of permanent infantile hearing impairment has become extremely important in preventive medicine, since steps can be taken with hearing aids and rehabilitation to ensure better development of language and higher cognitive functions. Aim of this study is to provide a critical review of the time of diagnosis of hypoacusis at our audiology laboratory, where two methods were used to screen hearing of children with/without risk indicators. Results of approximately 10 years' work were re-examined during which time outpatient screening was conducted on children referred by colleagues in neonatology and paediatrics. All were carriers of congenital risk indicators associated with sensorineural and/or conductive hearing loss, based on the Joint Committee on Infant Hearing findings, or were suspected of being hypoacusic even if they had no known congenital risk factors. Hearing screening was conducted in hospital on newborns with no risk factors, within the first few days of birth. Results of the present study showed that when selective hearing screening was performed, the mean age of high-risk patients diagnosed with hypoacusis was slightly higher than that in international guidelines. Moreover, these patients represent approximately half the hypoacusic population identified in the study period. The other half of congenital hypoacusic subjects identified had no risk indicators and there was a significant delay in diagnosis due to later manifestation of symptoms indicating hypoacusis, and thus, in turn, delayed referral for hearing tests. In contrast, subjects without risk indicators who underwent in-hospital hearing screening and proved to be hypoacusic, were diagnosed early. In our experience, however, universal screening has considerable disadvantages, such as difficulty in covering the entire population, difficulty in follow-up after discharge from hospital, and last, but by no means least, significant organisational and professional commitments, making it impossible to perform in all hospitals. In order to ensure effective hearing screening for congenital hearing loss and, thus permit prompt identification of hypoacusic children, use of hearing aids and rehabilitation

Riassunto

La sordità presenta la maggiore incidenza fra le malattie congenite qualora si consideri ogni grado di ipoacusia permanente mono o bilaterale. La diagnosi precoce della sordità permanente infantile riveste oggi grande importanza nell'ambito della medicina preventiva poiché consente, per mezzo di opportune procedure di protesizzazione e di riabilitazione un migliore sviluppo del linguaggio e delle funzioni cognitive superiori. Lo scopo di questo lavoro è stato quello di effettuare una revisione critica dell'epoca della diagnosi di ipoacusia nel nostro laboratorio di audiologia ove sono state eseguite due modalità di screening audiologico su bambini con e senza indicatori di rischio. Sono stati riesaminati i risultati di circa dieci anni di attività in cui sono stati sottoposti a screening audiologico ambulatoriale i soggetti inviati dai colleghi neonatologi e pediatri in quanto portatori di indicatori di rischio congeniti associati ad ipoacusia neurosensoriale e/o trasmissiva secondo il Joint Committee on Infant Hearing o perché, pur in assenza di fattori di rischio congeniti noti, sospettati di essere ipoacusici. Lo screening audiologico su soggetti esenti da indicatori di rischio è stato condotto in regime ospedaliero sui neonati privi di fattori di rischio, entro i primi giorni di vita. I risultati del nostro studio hanno evidenziato che qualora si esegua uno screening audiologico selettivo l'età media di identificazione dell'ipoacusia nei soggetti a rischio è moderatamente superiore a quella auspicata dalle linee guida internazionali e che questi ultimi rappresentano circa la metà della popolazione ipoacusica identificata nel periodo di studio. L'altra metà dei soggetti ipoacusici congeniti identificati era priva di indicatori di rischio ed è stata riconosciuta con un ritardo diagnostico significativo. Tale ritardo è da attribuire alla manifestazione tardiva dei sintomi che possono fare sospettare l'ipoacusia ed al ritardo quindi nell'invio alla valutazione audiologica. I soggetti risultati ipoacusici sottoposti a screening audiologico neonatale ospedaliero esenti da indicatori di rischio, al contrario, vengono diagnosticati tempestivamente ma nella nostra esperienza lo screening con intento universale presenta alcuni seri svantaggi come la difficoltà di copertura di tutta la popolazione, la difficoltà del follow-up dopo la dimissione ospedaliera ed infine il notevole impegno organizzativo e professionale che lo rendono non applicabile in tutte le realtà ospedaliere. Affinché lo screening audiologico delle ipoacusie conge-

screening should incorporate two aspects. First, selection should be compulsory, thereby reducing waiting time between collecting case histories and performing outpatient tests; second, hospital screening of children without risk factors should be performed whenever possible. Integrating these two aspects would make it possible to approach the "utopia" of universal hearing screening.

nite sia efficace, ovvero consenta di identificare, protesizzare e riabilitare tempestivamente i bambini ipoacusici, dovrebbe integrare a nostro avviso due aspetti: essere obbligatoriamente selettivo, abbreviando i tempi di attesa tra la raccolta dell'anamnesi e l'esecuzione degli esami ambulatoriali, ed essere completato nella maggior misura possibile dallo screening ospedaliero sui bambini esenti da fattori di rischio. L'integrazione tra questi due aspetti potrà consentire di raggiungere l'apparente utopia dello screening audiologico universale.

Introduction

Hearing impairment is the most common congenital disease (3-5.9‰) if all degrees of permanent uni- or bilateral hypoacusis are taken into consideration [1-5]. Early diagnosis of permanent hearing loss in infants is of great importance in terms of preventive medicine. It has been proven that timely prosthetic treatment and rehabilitation influence the future of the hypoacusic subject in terms of language development, psychological balance, as well as school and social integration [6-13]. With the use of a suitable hearing aid, within the first 6 months, these infants can acquire language and verbal communication skills comparable to those of normal-hearing children of the same age [14]. Together with early rehabilitation, these children can develop capabilities superior to those of children of the same age with the same degree of hypoacusis, but who were diagnosed and rehabilitated at a later stage [15 16]. It has also been shown that, in addition to early diagnosis and rehabilitation, positive involvement of the family of the hypoacusic child contributes to better language acquisition. Inversely, inadequate family participation always leads to a significant delay in language acquisition [9]. Therefore, one must assume that these four combined factors – early detection, hearing aids, rehabilitation and family support – can prevent hearing impairment from becoming a disability or handicap [17-19].

Currently, in areas without universal hospital screening programmes, the detection of permanent congenital infantile hearing loss occurs in subjects with no risk factors at an average age of 20 to 58 months [20]; hearing tests begin at the age of 4 to 10 months in profound hearing loss, 6 to 17 months in severe cases, and 9 to 42 months for moderate degrees of hypoacusis [19-21].

Aim of this study is to provide a critical review of the time of diagnosis of hypoacusis at our audiology laboratory.

Materials and methods

Results of infant audiology diagnoses made in our Institute between 1st January 1990 and 31st December 2001 were examined. Selective hearing screening was conducted on a group of subjects, referred by colleagues in neonatology and paediatrics, showing audiological risk indicators that were either congenital or associated with sensorineural and/or conductive hearing loss based on the classification drawn up by the Joint Committee on Infant Hearing [22-24].

We simplified and modified this classification to make the procedure easier. Therefore, the group of subjects with congenital risk indicators included those presenting these characteristics at birth and from birth to 28 days, and those with syndromic forms detectable between the ages of 29 days to 2 years [22 23].

We then identified a second group of subjects, defined as carriers of acquired risk indicators, i.e., children ranging in age from 29 days and 2 years with risk indicators associated with sensorineural and/or conductive hypoacusis, based on the JCIH classification [23]. This group differs from that proposed by the above-mentioned source [23] since it excludes two risk indicators. The first (syndromic lesions), as previously mentioned, was included in the group of subjects with congenital risk indicators, whereas the second was considered as a separate group, since it comprised subjects with no risk indicators referred to the audiology laboratory with suspected hypoacusis, delayed speech, and/or delayed psychophysical development. In all these patients, both the case history and clinical examination made it possible *a posteriori* to exclude the presence of any congenital or acquired risk indicators, thus eliminating any doubts vis-à-vis possible mistaken inclusion of these patients in the group. Therefore, the cause of hypoacusis in these patients remained unknown.

All patients underwent clinical evaluation and testing of auditory brainstem response (ABR) using Amplaid Mk 15 equipment to mark the threshold. In cases presenting an altered pattern compared to the intensity-latency function of J5 in our reference data, we performed complete hearing tests according to NIH indications [25]. Variation from normative data was considered as such when the recorded value differed from the expected value by more than one standard deviation from the mean, or when a latency value of J5 showed an increased threshold, even though it was within the normal range at the higher intensities. The ABR threshold was defined as normal when the re-

sponse was clearly identifiable up to the acoustic stimulation intensity of 20 dB nHL (where nHL stands for the normalisation of the click threshold in a group of adults with normal hearing). The absence of J5 at the maximum intensities of acoustic stimuli (130 dB SPL) showed severe/profound hearing loss in the range of frequencies examined by the method (2-4 kHz) [26].

We then assessed the results of universal hearing screening performed on a second group of subjects with no risk factors, using transient evoked otoacoustic emissions (TEOAE). In this case, we followed a two-stage procedure: TEOAE at birth and, in cases showing no response, ABR testing within the age of 3 months; complete hearing testing followed if the suspicion of hypoacusis was confirmed. Between January 1999 and March 2001, 2,425 full-term newborns were tested at the Newborn Centre. From 1st January 1999 to 8th May 2000, 1,543 newborns with no risk factors underwent TEOAE testing using quickscreen Otodynamic ILO88-92 equipment within the 4 days of birth. Subsequently, between 9th May 2000 and 23rd March 2001, 882 newborns were tested using automatic equipment (Madsen Echoscreen). TEOAE, in one ear only, was considered sufficient as a "pass" criterion [27 28], and the test was stopped after unilateral presence was detected.

ABR tests were performed using an Amplaid MK 15 auditory evoked potential system. The signals were recorded using miniaturised Beckman-type silver chloride electrodes applied to the crown (active electrode), the mastoid processes (detector electrode) and the contralateral mastoid (earthing electrode). First, the infant's skin was thoroughly cleansed with alcohol to reduce impedance of the electrodes to at least 3 kOhms. The stimuli used were 2,048 alternating polarity clicks generated by rectangular electrical impulses with a duration of 0.1 msec, sent through a TDH 49 cap at a rate of 21 stimuli per second at an initial intensity of 60 dB SPL p.e. This intensity was increased or decreased in 10 dB steps, depending on the presence or absence of a response. The signal was filtered using an analogue bandpass filter (20-2000 Hz-3 dB; 6 dB per octave band). The time of analysis was 15 msec. The patterns were viewed on an oscilloscope and recorded on an analogue plotter. The test was repeated at least three times in order to assess intra-individual variability.

The electrical impulse used by the ILO88-92 is rectangular, lasting 80 μs and sent at a frequency of 50 pps (80 pps in quickscreen) via a Knowles 1712 transducer inserted in a newborn-size probe together with a Knowles 1843 microphone, which can record the signal in the external auditory canal. Each click is separated from the following by an interval of 12 ms and has a spectral response of 600 to 5000 Hz. The amplitude of the electrical click that reaches the transducer in the probe is reduced by about 20 dB in the neonatal type. The stimulation paradigm used is the non-linear differential type (multiplexing), in which a train of stimuli is composed of 3 impulses in compression of equal amplitude, followed by a rarefaction impulse 3 times the intensity of the 3 previous impulses (1, 1, 1, -3) [29]. The total number of stimuli sent is calculated considering that the sample used is made up of two trains of 4 clicks and that the responses to the first train are recorded on pattern A, and the responses to the second on pattern B. The final result is the mean of 520 trains of alternating stimuli accepted in two sub-means. The two patterns, A and B, are projected onto the screen with a 2.5-12 ms analysis window. The extent of overlap of the two patterns is an index of test reproducibility, expressed as a percentage.

At our laboratory, 70% [30] is the minimum required overall correlation rate for considering the presence of a response, because this value has been shown to increase the effectiveness of the method [31].

At the standard setting, the ILO88 supplies an acoustic stimulus intensity of 75 to 85 dB p.e. SPL [32]. In accordance with the response acceptance criteria of the National Consortium, a TEOAE response is considered present if it can be reproduced or has an acceptable margin for at least 3 of the 4 frequency bands centred at 1600, 2400, 3200 and 4000 Hz [33].

The second piece of equipment used, the Madsen Echoscreen, is automatic and does not require an operator to interpret the pattern. A binomial statistical calculation is applied to each sample point of the response in the time interval of 6 to 12 ms. Four consecutive positive and negative peaks exceeding the value of 3Σ, in a random distribution are considered a "pass" (99.9% significance for each peak).

Results

During the above-mentioned period, 1,059 preschool children were assessed as outpatients; 717 (67.7%) were from Group 1 (congenital audiological risk indicators), 56 (5.7%) from Group 2 (acquired risk indicators), and 286 (27%) from Group 3 (no risk indicators). Of the 717 with congenital risk indicators, 68 (9.5%) showed hypoacusis. Of these, 26 (3.6%) had severe-profound bilateral hearing loss, 20 (2.8%) had unilateral hearing loss [profound in 6 (0.8%) and moderate-severe in 14 (1.9%)], and 22 (3.1%) had moderate-severe bilateral hypoacusis. The mean age at diagnosis of the 68 affected children was 10.1 ± 6.9 months (min. 1 month – max. 3.5 years).

Of the 56 children in Group 2, 4 (7.1%) had profound bilateral hearing loss, whereas the others had normal hearing. For these children, it was not considered necessary to calculate the mean age at diagnosis of

hypoacusis, since the time at which the tests were carried out was obviously determined by the onset of the acquired risk indicator.

Table I shows the number of children audiologically at risk for each factor identified, next to the number of children who were effectively found to be hypoacusic and the cause of hypoacusis.

Group 3 – children with no known audiological risk

indicators, either congenital or acquired – comprised 286 individuals, of whom 35 (12.2%) were found to be hypoacusic: 8 (2.8%) had profound bilateral hearing loss, 12 (4.2%) had unilateral hearing loss (profound in 8 cases and moderate-severe in 4), and 15 (5.2%) had moderate-severe bilateral sensorineural hypoacusis. For this group, the mean age at diagnosis was 32.3 ± 13.5 months, with a minimum of 14

Table I. Number of children audiologically at risk for each factor identified.

Newborn audiological risk factors (0-28 days) (JCIH, 1990, 1994, 2000)		
	Number of children	Hearing loss
Intra-uterine infections		
Cytomegalovirus	15	2A
Toxoplasmosis	11	1B
German measles	5	1A
Cranio-facial abnormalities		
Cleft lip and palate	13	1A+2B
Auricular malformations	25	1C
Aural atresia	4	4D
Hydrocephalus	5	1A+1D
Various	26	1A+1B+1C
Family history of infantile sensorineural hypoacusis	51	3A
Low birth weight (< 1500 g)	83	7A+1B+3D
Disorders usually associated with sensorineural and/or conductive hypoacusis		
Trisomy 8	1	1A
Trisomy 21	61	3A+4B
Hypothyroidism	4	1C+1D
Severe dysfunction at birth		
Hypotonia	2	1A
Hypertonia	5	1B
Foetal alcohol syndrome	1	1B
Jaundice	26	4B
Hospitalisation NICU >48 hours		
Premature birth	299	4A+7B+3C+5D
Respiratory distress	73	1A
Dystocia	7	
Postnatal audiological risk factors (29 days-2 years) (JCIH, 1990, 1994, 2000)		
Postnatal infections associated with sensorineural hypoacusis		
Bacterial meningitis	45	3A
Chicken pox	1	
Recurrent or persistent otitis media	5	
Cranial trauma	5	1A

Legend. A: profound bilateral hearing loss; B: moderate-severe bilateral hearing loss; C: profound unilateral hearing loss; D: moderate-severe unilateral hearing loss.

months and a maximum of 5 years. Mean age at diagnosis of profound bilateral hearing loss was 28.7 ± 15.3 months (min. 16 months – max. 5 years), for unilateral hearing loss 36.5 ± 15.2 months (min. 14 months – max. 5 years), and for moderate-severe bilateral hearing loss 31.1 ± 10.9 months (min. 16 months – max. 4 years).

With regard to the application of newborn screening criteria in infants with no risk factors in the first period (ILO88-92 quickscreen), 1543 (94.2%) newborns were assessed out of a total of 1637 births: 1405 (91%) were classed as "pass" (monoaural criteria) and 138 (8.9%) were "refer" (absence of binaural response). When checking the 138 "refer" infants 3 months later, 107 (77.5%) were found to be "pass", 7 (5%) were classified as "fail", and 24 (17.4%) were classified as "missed".

In the second study period (Madsen Echoscreen), 882 infants (72.4%) were assessed out of a total of 1218 births: 793 (89.9%) were found to be "pass", whereas 89 (10.1%) were found to be "refer". Three months later, 18 infants (20.2%) were found to be "missed"; of the remaining 71 who were reassessed, 66 (74.1%) were classified as "pass" and 5 (5.6%) as "fail".

Lastly, complete hearing testing revealed 12 "fails" at the age of 3 months, out of the total of the two periods: bilateral sensorineural hypoacusis requiring a hearing aid was identified in 2 cases, whereas the other infants had unilateral hypoacusis that did not require prosthetic treatment.

Discussion and conclusions

The review of our case histories has highlighted a topic worthy of consideration: although hearing screening seems to be accepted unanimously and is strongly recommended in the international literature [24 25 34 35], it is still not widely used in daily clinical practice. It has been confirmed that certain newborns with a high risk of hypoacusis can be diagnosed on the basis of easily detectable preset criteria by examining their auditory threshold [3.] However, diagnostic procedures can also be applied to the entire newborn population for early identification of hypoacusic infants with no risk factors [36].

Our results clearly suggest that selective hearing screening allows us to confirm hypoacusis in carriers of known congenital audiological risk indicators (Group 1) at a slightly higher average age than that recommended in international guidelines [24]. Even though the majority of these infants are promptly detected, the significant delay with which some infants are assessed has considerably shifted the mean age at diagnosis. The possible reasons for this delay can be attributed in part to concomitant diseases that overshadowed the problem of possible hypoacusis, and in part to incorrect information given to parents or to the lack of support shown by them [37]. Unlike Group 3, for Group 1, it was not deemed unnecessary to divide the mean age at diagnosis into sub-groups according to the degree of hypoacusis. This is because the infants were referred for screening since the risk factor was already known, as it was present at birth - and not because hypoacusis was suspected. Therefore, it is clear that, in this case, there is no correlation between the extent of hearing impairment and the time at which it was diagnosed.

In Group 2 (acquired risk indicators), detection was rapid in cases brought to our attention on account of the triggering event. In fact, all the patients in our case history were assessed within a short time of occurrence of the acquired risk factor. We have no data concerning the number of patients who may potentially have been exposed to the acquired risk and were not brought in for assessment.

In patients without risk indicators (Group 3), the delay in diagnosis was significantly greater compared to Group 1. On average, suspected hypoacusis was confirmed at about 2 years of age in the profound bilateral forms, or even at the age of 3 in the unilateral forms, whereas intermediate time spans were recorded for moderate-severe forms, similar to those reported in the literature [18]. This confirms that the age at which hypoacusis is diagnosed is correlated with the degree of hearing impairment [38 39].

The development of language skills is negatively affected by mild and, above all, moderate-severe hypoacusis [40 41]. In particular, the sensorineural forms appear to be important from an epidemiological standpoint due to their insidious nature and the delayed suspicion of a problem within the family setting. Aside from the sense of security created by the absence of risk indicators, this delay is also encouraged by the condition described as amodal perception. This is an innate characteristic in children whereby they grasp information in a sensory form and transform it into another one [42], creating parent-child interaction during the first 12 months that is identical in infants with or without a sensory deficit. However, amodal perception alone cannot explain the serious delay in diagnosis observed in some cases, for which other causal factors must thus be cited (disinformation, socio-economic factors).

Considering that unilateral hypoacusis has far less impact than the bilateral form, that no early intervention is planned for this type of hypoacusis, and that the costs involved in detection of the condition, it is almost the same as that in diagnosing a bilateral form, the cost-benefit ratio is debatable in this case [43]. However, the fact that unilateral hearing impairment, in a school-age child, causes problems for his/her hearing, language and behavioural abilities, cannot be ignored [44]. Furthermore, awareness of this

deficit can aid educators in their task and help the child learn, since simple procedures can be adopted. The advantage of universal hearing screening criteria is that they allow profound hypoacusis to be identified in patients with no risk factors who could potentially be diagnosed with considerable delay. Good outcome of the programme depends on when assessment is made and whether it is judged to be a uni- or bilateral "pass". Likewise, it depends on the validation criteria of the response, and on the ability to organise and plan the follow-up of those patients according to the size of the population requiring reassessment [37]. In fact, as can be seen from our data and as shown in previous school investigations [45], the percentage of failure in recording TEOAE and, therefore, the number of newborns who are not assessed during their hospital stay, varies considerably. In the present series, this ranged from 5.8% in the first period to 27.6% in the second; these figures are in keeping with those reported in the literature [45]. This variability in the results is reflected in screening coverage, which is one of the key quality indexes. Screening coverage figures are often downplayed in the literature, but in known cases they vary from 56-63% (lowest reported values) to 80-90% (intermediate values) and to more than 90%, these figures, nonetheless, never reach 100% [46]. In some hospitals at which universal screening is performed, the minimum coverage figure has been set by law at 85% [47]. Well-organised follow-up of children in whom hypoacusis is suspected is also crucial. The loss of children to follow-up testing affects screening efficiency: from a statistical standpoint, there is a higher proportion of true positives in the unexamined population with respect to the population that has been examined [48] [49]. Therefore, in the second stage of the procedure, the children who have not been assessed should not exceed 10% [43]. Nevertheless, as shown by our own experience (17.4% in the first period and 20.2% in the second) and by that of other authors [1], this is difficult to achieve.

Selective hearing screening, performed by our audiology laboratory, is the reason why approximately 50% of children with congenital hypoacusis go unidentified. In fact, in accordance with data reported in the literature [50-54], there is a 30.7% rate of failure to diagnose profound bilateral hypoacusis (8 cases untested out of 26 patients with risk indicators in the same period of time). However, if we also consider moderate-severe bilateral sensorineural forms, the percentage rises to 47.9% (23 infants without indicators out of 48 with indicators).

On first analysis, screening based on the selection of audiological at-risk subjects seems simpler and less expensive to perform. It has been shown that it is now possible to identify all newborns with congenital hypoacusis for the same amount – or less – than what it cost 10 years ago to assess and identify hypoacusic subjects at risk. Consequently, universal screening could be underwritten for the very same amount [55]. Based on our experience with selective screening, the assessment period must be moved up. This could be achieved by improving information and collaboration with neonatologists, paediatricians and parents.

To ensure that hearing screening of congenital hypoacusis cases will be effective, it should be universal, and it should be carried out in a hospital setting in the newborn period [24] [34] [35]. Our results confirm that recognising only subjects who are carriers of audiological risk indicators exposes subjects without risk indicators to a severe delay in diagnosis. On the other hand, universal screening inevitably means there is still a possibility that a certain number of subjects will not be assessed. It also entails an enormous amount of work, as well as significant professional and organisational commitments.

Based on our experience, selective hearing screening should always be performed at all health facilities by assessing the infants on an outpatient basis within the shortest possible time. However, it should also be supplemented by hospital screening of subjects without any risk factors. Achieving the aim of prompt identification also of the hypoacusic subjects, in this group, may be partial or total, depending largely upon quality of the screening.

References

[1] Colorado Program Report. *Summary statistics reported at the Annual Meeting of the Consortium for Universal Newborn Hearing Screening*, Washington DC. Sept. 30. 1994. Data available from National Center for Hearing Assessment and Management, Utah State University, Logan, UT 1994.

[2] Grosse S. *Cost comparison of screening newborns for hearing impairment and biochemical disorders.* Newborn Screening and Genetic Conference, May 2001.

[3] Parving A. *Prevalence of congenital hearing impairment and risk factors. Neonatal hearing screening.* The European Consensus Development Conference on Neonatal Hearing Screening. Grandori F, Lutman M, editors; Milan, 1998. p. 6-10.

[4] Utah Program Report. *Summary statistics reported at the Annual Meeting of the Consortium for Universal Newborn Hearing Screening*. Washington, DC: 1994. Data available from National Center for Hearing Assessment and Management, Utah State University, Logan, UT. 1994.

[5] White KR. *Universal newborn hearing screening using transient evoked otoacoustic emissions. Past, present, and future*. Seminars in hearing 1, 1996;7:171-83.

[6] Downs MP, Yoshinaga-Itano C. *The efficacy of early identification and intervention for children with hearing impairment.* Pediatric Clin North Am 1999;46;79-87.

[7] Hall JW. *Screening for and assessment of infant hearing impairment.* J Perinatol 2000;20:S113-21.

[8] Hayes D. *State programs for universal newborn hearing screening.* Pediatric Clin North Am 1999;46;89-94.

[9] Moeller MP. *Early intervention and language development in children who are deaf and hard of hearing.* Pediatrics 2000;106:43-51.

[10] Schildroth A. *Hearing-impaired children under age 6. 1977-1984.* Am Ann Dea 1986;131:85-90.

[11] Sininger YS, Doyle KJ, Moore JK. *The case for early identification of hearing loss in children. Auditory system development, experimental auditory deprivation, and development of speech perception and hearing.* Pediatric Clin North Am 1999;46:1-14.

[12] Van Straaten HL. *Automated auditory brainstem response in neonatal hearing screening.* Acta Paediatr 1999;88(Suppl 432):76-9.

[13] Yoshinaga-Itano C, Apuzzo ML. *The development of deaf and hard of hearing children identified early through the high-risk registry.* Am Ann Deaf 1998;143:416-24.

[14] Robinshaw HM. *Early intervention for hearing impairment: differences in the timing of communicative and linguistic development.* Br J Audiol 1995;29:315-34.

[15] Moeller MP. *Personal communication on The Scientific Basis for Newborn Hearing Screening 2000.* NCHAM 1997.

[16] Yoshinaga-Itano C, Sedey A, Apuzzo M, Carey A, Day D, Coulter D. *The effect of early identification on the development of deaf and hard of hearing infants and toddlers.* Paper presented at the Joint Committee on Infant Hearing Meeting, Austin, Texas 1996.

[17] World Health Organization. *The international classification of impairments, disabilities, and handicaps: a manual relating to the consequences of a disease.* Geneva: WHO. 1980.

[18] NCHAM (National Center for Assessment and Management) Diagnostic Audiology. AAA Meeting. Utah State University. 2001.

[19] Türk R, Nekahm D. Breidert H. *Die früherkennung kindlicher Hörstörungen: eine aktuelle Analise.* Audio Akust 1992;31:166-73.

[20] Harrtmann H, Hartmann K. *Früherkennung? Memorandum zur Früherkennung und Frühförderung hörgeschädigter Kinder.* Bundesgemeinschaft der Eltern und Freunde hörgeschädigter Kinder, e V. 1993.

[21] Davis A, Banfort 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 Technol Assess Winch Eng 1997;I:1-176.

[22] Joint Committee on Infant Hearing. *1990 Position Statement.* American Speech-Language-Hearing Association 1991;33(Suppl.5):3-6.

[23] Joint Committee on Infant Hearing. *Position Statement.* American Speech-Language-Hearing Association 1994;36:38-41.

[24] Joint Committee on Infant Hearing. *Position Statement.* American Speech-Language-Hearing Association 2000;42:23-4.

[25] NIH Consensus Development Conference Statement. *Early identification of hearing impairment in infants and children.* Inter J Ped Otorhinol 1993;27:215-27.

[26] Weber BA. *Handbook of clinical audiology. 4th Ed.* Katz J, editor. 1994. p. 375-86.

[27] Salomon G, Groth J, Anthonisen B. *Preliminary results and considerations in hearing screening of newborns based on otoacoustc emissions.* Br J Audiol 1993;27:139-41.

[28] Welzl-Muller K. *Neonatal hearing screening: screening test for hearing disorders in newborn infants.* Report on the "European Consensus Development Conference on Neonatal Hearing Screening", Milan, 15-16 May. HNO 1998;46:704-7.

[29] Kemp DT, Ryan S, Bray P. *A guide to the effective use of otoacoustic emissions.* Ear Hear 1990;11:93-105.

[30] Van Dijk P, With HP. *The occurrence of click-evoked otoacoustic emissions ("Kemp echoes") in normal hearing ears.* Scand Audiol 1987;16:62-4.

[31] Hurley RM, Musiek FE. *Decision matrix analysis of evoked otoacoustic emissions.* American Speech-Language-Hearing Association 1991;33:145.

[32] Kemp DT, Ryan S. *The use of transient evoked otoacoustic emissions in neonatal hearing screening programs.* Seminars in Hearing 1993;14:30-45.

[33] National Consortium for Newborn Hearing Screening. *TEOAE-based universal newborn hearing screening.* Georgetown, University School of Medicine, Washington DC: November 16-18, 1995.

[34] American Academy of Pediatrics. *Newborn and infant hearing loss: Detection and intervention.* Pediatrics 1999;103:527-30.

[35] Grandori F, Lutman M. *The European Consensus Development Conference on Neonatal Hearing Screening (Milan, 15-16 May, 1998).* Eur Arch Otorhinolayngol 1998;255:521-2.

[36] Kemp DT. *The Past and Present of Hearing Screening Technology.* The European Consensus Development Conference on Neonatal Hearing Screening. Grandori F, Lutman M, editors; Milan, 1998. p. 21-24.

[37] Molini E, Ricci G, Alunni N, Simoncelli C, Brunelli B. *Risultati e considerazioni a proposito di uno screening audiologico neonatale basato sull'impiego delle otoemissioni acustiche evocate transienti.* Acta Otorhinolaryngol Ital 1997;17:1-8.

[38] Stein L, Jabaley T, Spitz R, Stoakley D, McGee T. *The hearing-impaired infant: Patterns of identification and habilitation revisited.* Ear Hear 1990;11:201-5.

[39] Harrison M, Roush J. *Age of suspicion, identification, and intervention for infants and young children with hearing loss. A national study.* Ear Hear 1996;17:55-62.

[40] Pillsbury HC, Grose JH, Hall JW III. *Otitis media with effusion in children. Binaural hearing before and after corrective surgery.* Arch Otoryngol Head Neck Surg 1991;117:718-23.

[41] Hall JW, Grose JH. *The effect of otitis media with effusion on the masking level difference and the auditory brainstem response.* J Speech Hear Res 1993;36:210-7.

[42] Preisler G. *The development of communication and language in deaf and severely hard of hearing children: implications for the future.* Int J Pediatr Otorhinolaryngol

1999;49(Suppl 1):S39-43.

43 Govaerts PJ. *Cost of screening programs*. In: Grandori F, Lutman M, editors. *The European Consensus Development Conference on Neonatal Hearing Screening*. Milan 1998. p. 25-9.

44 Bess FH, Tharpe AM. *Unilateral hearing impairment in children*. Pediatrics 1984;74:206-16.

45 Molini E, Ricci G, Chiarella G, Simoncelli C. *L'applicazione delle emissioni otoacustiche evocate transienti (TEOAE) nei programmi audiologici neonatali*. Otorinolaringol Pediatr 2001;XII:61-5.

46 Bamford J, Davis A. *Coverage and yield from neonatal and infant screening*. In: Grandori F, Lutman M, editors. *The European Consensus Development Conference on Neonatal Hearing Screening*. Milan 1998. p. 44-7.

47 Hayes D. *Newborn hearing screening: selected experience in the United States*. Scand Audiol 2001;30(Suppl 53):29-32.

48 Johnson A, Ashurst H. *Screening for sensorineural deafness by health visitors*. Arch Dis Child 1990;65:841-5.

49 Davis A. *Public health perspective of childhood deafness*. In: McCormick, ed. *Paediatric Audiology 0-5 years. 2nd Ed*. VI. London: Whurr; 1993. p. 1-41.

50 Elssmann SF, Matkin N, Sabo M. *Early identification of congenital sensorineural hearing impairment*. Hearing J 1987;40:13-7.

51 Feinmesser M, Tell L, Levi H. *Follow-up of 40,000 infants screened for hearing defect*. Audiology 1982;21:197-203.

52 Mauk GW, White KR, Mortensen LB, Behrens TR. *The effectiveness of screening programs based on high-risk characteristics in early identification of hearing impairment*. Ear Hearing 1991;12:312-9.

53 Pappas DG, Schaibly M. *A two-year diagnostic report on bilateral sensorineural hearing loss in infants and children*. Am J Otol 1984;5:339-43.

54 Watkin PM, Baldwin M, McEnery G. *Neonatal at risk screening and the identification of deafness*. Arch Dis Child 1991;66(10 Spec No):1130-5.

55 Robinette MS. *Methods of Infant Screening. Neonatal Hearing Screening*. In: Grandori F, Lutman M, editors. *The European Consensus Development Conference on Neonatal Hearing Screening*. Milan 1998. p. 11-20.

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