Newborn Hearing Screening Programs: Overview

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Abstract

Implementation of systematic programs for early identification of hearing impairment in the newborn and infant is increasing in Canada and worldwide. This article outlines the rationale for these programs, methods of screening, audiologic assessment and intervention, program outcomes and the crucial role of physicians. Sources of high-quality, current evidence on key aspects of these programs are identified. There is an emerging, evidence-based consensus that a systematic approach based on universal newborn hearing screening (UNHS) and timely, appropriate follow-up services is practicable and will yield substantial net benefit for many affected children and families. Early identification programs lead to physicians being faced with infants under six months of age who already have detailed and accurate audiometry. Important challenges include a systematic approach to etiologic evaluation of the young infant with permanent hearing impairment and the facilitation of prompt, non-medical interventions.

Sommaire

Les programmes de dépistage systématique de la surdité chez les enfants et les nouveau-nés gagnent en popularité au Canada et dans le reste du monde. Cet article revoit la logique de ces programmes, les méthodes de dépistage, l'évaluation et les interventions audiologiques, les résultats des programmes et le rôle crucial des médecins. Nous avons identifié des sources fiables sur les aspects clés de ces programmes. Un consensus basé sur des données probantes émerge à l'effet que le dépistage systématique et universel des nouveau-nés et un suivi approprié est une stratégie praticable qui génère des bénéfices importants pour les enfants et leurs familles. Ces enfants consulteront un médecin avant l'âge de six mois avec un dossier audiologique complet et précis. Les défis seront donc d'investiguer systématiquement les étiologies possibles de ce problème et de faciliter la prise en charge rapide par les intervenants non-médicaux.

Key words: early hearing detection and intervention, permanent childhood hearing impairment, universal newborn hearing screening

The Clinical Problem

Population screening and early intervention programs for hearing impairment in the newborn and infant are justifiable because there is now clear evidence that (1) in their absence, substantial harm to many children and families will occur; (2) they yield significant benefit in terms of earlier, improved hearing; (3) they yield significant benefit for long-term language development; (4) there are several ancillary benefits; and (5) there is negligible collateral harm from programs that are well designed and well executed.

Kennedy and McCann provided an excellent overview of the rationale, methods, and some new evidence of outcomes of universal neonatal hearing screening (UNHS).[1] One rationale for changing patterns of health services is that existing services are not effective. The overall harm accrued under the traditional system that was driven by the index of suspicion about hearing loss has three main components: number of cases; age at detection, diagnosis, and intervention; and impact on children and families.

Prevalence

The prevalence of hearing impairment depends strongly on what is included in the "target disorder." The modern focus is on permanent childhood hearing impairment (PCHI), which includes sensorineural impairment and "structural" conductive impairment. The latter is due to anatomic abnormality of the external or middle ears and is included because it may cause significant hearing loss for months or even years before surgical intervention. However, most PCHI is sensorineural and irreversible.

In an excellent review of PCHI prevalence, Fortnum focussed on PCHI averaging 40 dB HL (hearing level) or worse in the better ear.[2] Such impairment has an overall prevalence of about 1 in 1000 congenitally in Western, industrialized societies. A nationwide UK study suggested that this prevalence may double over

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the first decade of life, but the exact increase remains debatable.[3] Such increases are attributable to acquired, late-onset, and progressive impairment, the prevalence and time course of which are still unclear.

If audiometric criteria for "significant" PCHI are broadened to include lesser severities (eg, down to > 25 dB HL) and unilateral losses, the prevalence in early infancy increases to 2 to 3 in 1000. It is reasonable to include unilateral cases because of the risk of progression to bilateral impairment, the risk of temporary, conductive impairment in the normal ear, and a clear need for specific communication strategies by the family. One reason to include "mild" hearing losses as targets is the likelihood of severity progression. Moreover, the term "mild" is very misleading; anyone who doubts that a mild hearing loss confers significant disability should put themselves closer to the infant's predicament and try to cope with foreign language classes wearing well-fitted foam earplugs in the dark.

The bottom line is that, in our society, overall, 2 to 3 in 1000 infants will have congenital hearing impairment that merits early detection. This implies up to 1100 new cases annually across Canada.

Pattern of Detection, Diagnosis, Intervention

The best study to date of ages at detection, diagnosis, and intervention in Canada is due to Durieux-Smith and Whittingham.[4] They examined the records of 613 children fitted with hearing aids at the Children's Hospital of Eastern Ontario. The ages at diagnosis ranged from about 2 to 4 years in a referred group with risk indicators and from 2 to 7 years in a group without risk indicators, the lower age limits relating to "profound" impairment and the upper age limits to "mild" impairment. The median age at diagnosis was 6 months or less in a screened group, irrespective of impairment severity. Reports from other jurisdictions internationally are generally consistent with these findings.

Impact on Children and Families

Under the traditional system, diagnosis of hearing impairment was often not achieved under 2 years, even for severe and profound impairment in the presence of a risk indicator. Many children experienced 2 to 7 years of undiagnosed impairment. This sensory deprivation in a period of rapid neurologic and cognitive development is largely avoidable and might be considered unacceptable per se in a developed, affluent society.

Undetected hearing impairment will compromise the child's speech and language development by an amount that increases with the severity of the loss and the delay in diagnosis; important processes underlying normal language development begin under 6 months of age.

In early infancy, affected families will be unaware of any hearing impairment and are unlikely to use appropriate communication strategies. They will lack the information needed to make important decisions about lifestyle and about intervention to improve hearing and/or language development. Also, families may experience anxiety and frustration because failure to attain language development milestones may be attributed wrongly to "natural" developmental variation, even by health professionals. Beyond infancy, aberrant responsiveness to speech may be incorrectly attributed to developmental, psychological, or behavioural abnormalities.

Additionally, in some cases, the broader medical care of the child is compromised by a lack of timely information about hearing; an identified PCHI may contribute to earlier identification of certain syndromes or to etiologic diagnosis of other pathologic conditions.

Overall, this array of harms from late identification and diagnosis of PCHI is substantive and diverse. These harms accrue continually, in the absence of systematic, population-wide programs for early identification, diagnosis, and intervention.

A delay in detection has arisen because the behavioural manifestations of hearing impairment are such that families cannot make a valid assessment of responsiveness to sound, especially in the first year. Even if they were to seek medical advice, traditional behavioural screening tests for hearing impairment done in physicians' offices are notoriously unreliable. This is not a matter of individual physician skills. Rather, it is an inevitable result of intrinsic properties of auditory perception and responsiveness of infants. See, for example, Robertson and colleagues and Hayes.[5,6] The only practicable way to detect the required range of hearing impairments reliably in an infant under about 6 months developmental age is by modern, physiologic screening tests, coupled with modern, objective, diagnostic hearing assessment in infants who do not pass the screening. For older infants, sophisticated operant conditioning procedures are required to measure hearing sensitivity reliably, but no behavioural technique will work well in infants in the presence of visual or motor comorbidities or neurodevelopmental delay.

Early Hearing Detection and Intervention Programs

Systematic screening may be universal or may be targeted at the 8 to 10% of newborns who are likely to be at specific risk of PCHI. The main problem with targeted screening is that only about 50 to 60% of infants with PCHI manifest any of the current risk indicators, so the population sensitivity of targeted screening is, at best, about 50%; it is also manifestly inequitable to restrict access to necessary interventions on the basis of risk. Other problems are that comprehensive and accurate determination of risk is very difficult and time consuming; it is quicker and more accurate to use one of the modern screening tests than it is to assess risk prop-

erly. Also, many risk indicators are interrelated, their individual predictive values are not well understood, and their evidence base is often less than adequate. Some major risk indicators, such as asymptomatic cytomegalovirus infection, are not even routinely determined. For these reasons, modern approaches generally emphasize UNHS. UNHS is properly implemented as an integral component of what are widely known as early hearing detection and intervention (EHDI, pronounced "eddy") programs.

Screening is not defensible ethically in the absence of appropriate procedures and resources to provide prompt confirmation of impairment, diagnosis, and access to interventions that are reasonably effective. These requirements were originally specified by the World Health Organization[7] and have been refined by various groups. A modern, high-quality EHDI program comprises the following components: (1) UNHS; (2) a surveillance subprogram for emergent impairment that is not present neonatally; (3) comprehensive audiologic assessment to confirm and quantify the impairment; (4) psychological and informational support for families of children diagnosed with PCHI; (5) medical referral for etiologic investigation and management as indicated; (6) assistive technologies, including hearing aids, FM systems, and cochlear implants; (7) adjunctive counselling, information, and training to support the technologies; and (8) communication development options, including auditory-verbal therapy, manual language development, and other, related interventions.

The precise designs and implementations of these programs are context specific, but the goals and desired performance benchmarks are generally similar across programs and contexts. Programs must include sophisticated information systems to track and facilitate timely delivery of services and record events and outcomes. Provider personnel training and quality improvement subprograms are essential. The usual, critical indices of program quality are high population screening coverage, the timeliness and accuracy of confirmation and diagnosis of impairment, and strong family involvement with recommended interventions, all of which contribute substantially to overall program effectiveness.

In the United States, UNHS is currently legislated in 38 states, and the population hearing screening coverage is now about 90% of all newborns. See the Web site of the National Centre for Hearing Assessment and Management at <infanthearing.org> for an enormous amount of information about EHDI programs and many useful links. For an international perspective, see the UK national program Web site at <www.nhsp.info> and google on "International Working Group on Childhood Hearing."

In Canada, to my knowledge, EHDI programs are established in Ontario, New Brunswick, and the Northwest Territories. British Columbia has very recently announced a comprehensive program. Quebec is evaluating program feasibility, as may be the situation for other provinces. For an outline of the Ontario Infant Hearing Program, which was fully implemented in 2002, see the article by Hyde and colleagues.[8]

Universal Newborn Hearing Screening

Modern hearing screening tests are objective, physiologic, reasonably accurate, noninvasive, quick, and inexpensive. The most widely used methods are automated auditory brainstem response (AABR) and automated otoacoustic emissions (AOAEs). AABR screening devices deliver a rapid series of low-intensity clicks through an insert or supra-aural earphone and record electrical activity from the scalp via sensors. Averaged electrical waveforms are computed, and automated, statistical response detection algorithms evaluate the presence or absence of the auditory brainstem response (ABR). The test outcome for any ear is a "pass" or a "refer" result. The term "fail" is not used because of its negative connotations and because many infants who do not pass the screen may have normal hearing. This is so despite the fact that the AABR has very good sensitivity (probability of a refer result when PCHI is present, at least 90%) and specificity (probability of a pass result when the disorder is absent, about 95 to 98%) because PCHI is relatively rare. For example, for every 1000 children screened, 3 may refer with the target impairment (prevalence about 3 in 1000), but 20 false refer results (false-positive screens) are expected, so the predictive value of a refer outcome is about 15%.

A screening test is not a hearing test, and a positive screen means simply that the child must receive a definitive audiologic assessment to determine the true hearing status because the risk of a genuine impairment is now very substantial.

Otoacoustic emissions (OAEs) are faint sounds generated in the outer hair cells of the normal cochlea and radiated back through the middle ear to the external canal, where they can be recorded by a miniature microphone. Automated OAE screening devices measure either transient-evoked OAEs or distortion-product OAEs. Both types reflect outer hair cell functioning and have an accuracy similar to that of screening tests. Low-level sounds (either transient or tonal) are delivered by an insert earphone, and any OAE developed is recorded by a canal microphone. Computer processing and automated detection algorithms yield the pass or refer result. Most normal ears will yield an OAE, but the likelihood of obtaining a response decreases rapidly to almost zero in the presence of a PCHI of about 35 dB HL or greater. Essentially, OAE presence or absence, respectively, reflects normal or abnormal hearing sensitivity up to and including the outer hair cells.

There are important differences between AABR and AOAE screening tests. AABR devices are more

expensive, and screening of two ears takes up to 10 minutes versus about 2 minutes for AOAE. AABR requires more expertise, but both tests can be done properly by a wide range of persons, given brief training and reasonable aptitude. AABR has an advantage in that it will detect auditory neuropathy (AN), whereas AOAE will not. As currently defined, AN is not a single pathophysiologic entity. Rather, it is a cluster of pathologies that may involve inner hair cells, synapses with primary nerve fibres, and/or the cochlear nerve itself. Not all AN cases are genuine neuropathies, but some are associated with other peripheral neuropathy. AN was recognized relatively recently,[9] and it may comprise up to 5% or even 10% of all PCHI in infants (see, for example, Madden and colleagues[10]). There appears to be an association with a variety of perinatal insults, including hyperbilirubinemia and severe hypoxia, but there are also several genetic varieties. AN is also referred to as auditory dyssynchrony (AD), a term that is not etiologically specific and that reflects a presumed reduction in synchrony of the neural events, such as action, potentials that underlie audition.

AOAE screening refer results that are false-positive for PCHI can be caused by any mechanism that interferes with sound transmission from the earphone to the cochlea and back to the recording microphone. Common problems include debris or fluid in the middle ear or in the external canal, and the latter is more common when AOAE screening is done within 24 hours of birth. AABR is less affected by these factors.

UNHS protocols typically involve more than one screening test, often an immediate repetition, and, possibly, subsequent rescreening. A common approach is AOAE followed by AABR in AOAE refer results. Different approaches may be taken in the well-baby and intensive care nurseries. In the Ontario Infant Hearing Program, for example, babies at high risk of PCHI are screened with AABR only, whereas babies not at risk are screened by AOAE and then by AABR in AOAE refer outcomes.

These screens are usually done before hospital discharge because of ready access to the babies at that time and in the light of possible difficulties with tracking and attendance after discharge. There may also be another screen by AABR in babies who refer from hospital screening. These multistage protocols are designed to achieve very low overall screening false-positive rates to reduce the overall impact on families and the resources required for follow-up tracking and testing.

The sensitivity and specificity of a screening protocol improve as the target disorder criterion is made more strict. For the broader criterion defined earlier, a well-designed and well-executed multistage screening protocol can achieve a sensitivity (overall refer outcome when PCHI is present) of at least 90% and an overall specificity of at least 98% (2% false-positive rate). For the better-ear 40 dB criterion, the sensitivity of AABR may exceed about 95% and the specificity may exceed 99%. For more detailed information on screening test protocols and performance, see Watkin.[11]

Audiologic Assessment

Babies who do not pass screening protocols must receive definitive audiometry to confirm and quantify any PCHI. A common benchmark is complete audiometry by about 3 months corrected age or within a month of discharge after an extended postnatal hospital stay. Ideally, the audiometry will be done at about 2 to 3 months of age and not immediately postdischarge. The brief delay facilitates resolution of neonatal middle ear fluid and ear canal debris, as well as some prospect of resolution of minor, neonatal middle ear pathologies, any of which may have caused the refer result on newborn screening. Also, rapid maturational changes in the anatomic and acoustic properties of the middle ear and external ear occur especially in the first few months of life, and these can give rise to audiometric errors. Furthermore, the usual target for hearing aid fitting is by 6 months of age.[12] Earlier fitting is commonly associated with greater practical difficulties, and, to date, there is no evidence of incremental benefit from conventional hearing aids by fitting at, say, 1 or 2 months as opposed to at 4 to 6 months.

High-quality, definitive audiometry in a child under 6 months of age requires a very carefully designed protocol and substantial skill. In the Ontario program, for example, only registered audiologists with pediatric experience who have received additional, specialized training provided by the program may conduct these tests with program funding.

Currently, high-quality assessment of babies under about 6 months corrected age is usually based on frequency-specific, ear-specific, nonautomated ABR threshold measurement by air conduction, with insert or special supra-aural earphones, and with bone-conduction ABR threshold measurement, if indicated. The testing is done using standard, diagnostic ABR equipment but requires very specialized protocols. Behavioural testing is not considered reliable in this age group. Accurate estimates of perceptual threshold can be derived from tone-pip ABR thresholds. The ABR testing is augmented by diagnostic OAE analysis, plus tympanometry and acoustic reflex testing using high-frequency probe tones and specific, normative data for neonates and young infants.[13] Conventional tympanometry is done with a probe tone of low frequency and is not reliable in infants under about 6 months of age. An additional protocol is necessary for evaluation of suspected auditory dyssynchrony. Such protocols involve click ABRs, OAEs, and cochlear microphonic potentials (electrocochleography).

A well-designed audiologic assessment protocol can provide an audiogram that is sufficiently accurate

and detailed for all medical and audiologic management decisions in almost all infants referred from UNHS. Most infants under about 6 months can be tested by frequency-specific ABR in natural sleep. Infants over 6 months are likely to require sedation, typically with oral chloral hydrate, to obtain satisfactory electroencephalographic conditions in sedated sleep. Sedation may also be indicated in special situations, such as the family having travelled a great distance for the assessment, in which case, a successful and complete test is especially important.

If PCHI is detected by the audiologic assessment, referral to an otolaryngologist is usually considered appropriate, with the expectation that difficult cases will be referred to a pediatric otolaryngologist or otologist, where available. The medical assessment required is discussed later in this article. In most cases, there is no medical or surgical consideration that would contraindicate audiologic intervention, and the desired course is to proceed to audiologic habilitation as speedily as possible.

If the audiometry reveals a conductive impairment or overlay and suggests a middle ear disorder, medical referral for management of the conductive disorder is usually indicated. Definitive audiometry may be deferred for a month or two, anticipating resolution of the conductive components.

If AN(AD) is detected, audiologic management can be complicated. ABR thresholds are rendered invalid by the neuronal dyssynchrony, which usually causes absence or gross abnormality of the ABR at high stimulus levels, even if the true perceptual thresholds are near-normal. Estimation of accurate hearing levels may have to be deferred until behavioural thresholds can be obtained, and the thresholds may change over time. Sometimes such thresholds may not be obtainable, in which case, interventions must be considered in the absence of clear information about hearing sensitivity.

For infants with a developmental age of at least 6 months who have normal vision and motor control, the preferred technique is visual reinforcement audiometry (VRA). This is an operant conditioning technique that pairs auditory stimuli with visual response reward. The method requires a well-designed test protocol and considerable clinical skills. Given these, it is usually possible to obtain ear-specific and frequency-specific hearing thresholds that are accurate within about 10 dB of true perceptual thresholds provided that the child has a developmental age of at least 6 months and is free of visual or motor disorders. VRA may be combined with OAE measurements, standard tympanometry, and acoustic reflex tests to increase the diagnostic information. If, for any reason, the older infant cannot give reliable behavioural thresholds, then ABR under sedation may be required.

A new technique based on auditory steady-state responses or ASSR is a variant of ABR measurement that has the potential for faster and more objective threshold measurements. There are as yet insufficient normative data to determine the proper role of this method as a replacement or as a complementary test to frequency-specific ABR.

Surveillance for Emergent Impairment

Some hearing impairment in infancy is not detectable at the neonatal screening. This includes impairment that is not yet present, such as late-onset or acquired impairment, as well as impairment that may be present congenitally but is initially of insufficient severity to be detected by the screening. Some perinatal risk indicators, such as cytomegalovirus infection and certain syndromes, are clearly associated with late-onset or progressive hearing impairment. Some postnatal events, such as bacterial meningitis, carry a very high risk of auditory sequelae and indicate prompt referral for audiologic assessment. For many risk indicators, however, their predictive value is poorly understood, as is the time course of impairment expression. The overall prevalence of emergent impairment is often reported to be more than 10% of all impairment present in early childhood, but the true figure may be much greater. For a more detailed review, see Fortnum.[2] This issue of emergent impairment does not diminish the intrinsic merit of UNHS. What it does mean is that additional actions and systems above and beyond UNHS are necessary to ensure that all young children with significant PCHI are detected promptly.

Many EHDI programs include some kind of targeted rescreening or retesting of infants who pass UNHS but are at risk of late-onset or progressive impairment. However, prompt detection of acquired impairment requires a keen awareness of risk indicators on the part of the medical community. Some EHDI programs include education and information strategies for physicians, and several efforts at development of evidence-based guidelines to facilitate prompt detection throughout infancy and early childhood have occurred or are in progress. Other educational efforts are directed at families to promote awareness of language development milestones and other indicators of impaired hearing.

Family Support

The degree to which the family is positively engaged is very important throughout the entire process of screening, confirmation, diagnosis, and intervention. There is evidence that what many families want is involvement in decision making based on timely and unbiased information about the nature of procedures, the implications of test results, and decision options to optimize hearing and communication development.

In the screening and diagnostic phases of EHDI, the key objectives are to minimize needless family anxiety while at the same time promoting the highest possible

compliance with recommended program procedures. It is important, for example, that the real meaning of a refer result on screening is properly explained and understood. This can be initiated by pamphlets and video information made available before and during the hospital stay. It can be reinforced by well-trained screening personnel. Also, families may consult their child's physician(s), especially after hospital discharge with a screening refer result. It is important that physicians be in a position to endorse and reinforce the information given in the EHDI program, emphasizing the importance of attendance for recommended follow-up procedures, whether they are further screening, audiologic assessments, or interventions. For many programs, even those of the highest quality, loss to follow-up between program stages is a significant threat to overall success in delivering timely and effective interventions.

Many programs provide special support services following confirmation of PCHI. Families may need psychological support to cope with the diagnosis and to progress to the point of being willing and able to make informed decisions about options for hearing and communication development.

Assistive Technologies

Assistive technology includes conventional (air conduction) hearing aids and cochlear implants, as well as less commonly used devices, such as bone-conduction hearing aids and FM systems. Most infants with bilateral PCHI of a moderate degree or greater are candidates for hearing aids. Binaural fitting is the norm. The US Joint Committee on Infant Hearing recommends that such intervention should occur by 6 months of age wherever feasible. In well-established EHDI programs, this target is being met in the majority of cases. However, significant impediments to prompt provision of HAs include serious comorbidities, prolonged hospital stays, chronic middle ear disease, limited or delayed family compliance, and delays in medical assessment and management.

Successful hearing aid provision for a young infant is much more demanding than for most adults. Hearing aids must be selected, specified, and fitted with exquisite attention to the characteristics of the individual child and to the unique and evolving anatomic and acoustic characteristics of the individual infant's ears. It is especially important to use validated, evidence-based algorithms for hearing aid performance specification and to directly verify sound levels obtained in the ear canal. Periodic retesting of hearing is required to monitor for PCHI progression and for maturational changes in ear canal acoustics.

Cochlear implants are an option in many infants with bilateral PCHI of at least a severe degree whom do not appear to benefit adequately from a trial with a properly fitted hearing aid. Increasingly, cochlear implants are fitted as young as 1 year of age and in a wider range of PCHI. EHDI programs facilitate this trend by providing definitive audiometry in the first year of life and earlier access to hearing aid trials and cochlear implant candidacy assessments.

Communication Development Options

Fitting a device such as a hearing aid or cochlear implant may confer useful hearing almost immediately, but it is only the beginning of the communication development process. An individualized and lengthy program of information and training is needed to optimize device use, listening and communication strategies, and early acquisition of spoken language. A common approach is auditory verbal therapy, which seeks to maximize exclusive use of auditory information for language development. Other orally based approaches may blend elements of auditory verbal therapy with a range of visual information, including speechreading (lipreading) and a variety of manual signing systems; some of these approaches constitute what is called total communication. Yet other approaches may emphasize a manual language, such as American Sign Language or Langue des Signes Quebecoise, as the primary or initial language; this is more likely to occur if at least one parent is also deaf and espouses Deaf culture. In general, there are many strong opinions about what constitutes the best approach to language development for the individual child or even for large subgroups of children. However, such opinions appear not to be based on scientific evidence that would meet modern standards typically applied to health services research. An unpublished, systematic review of studies of effectiveness of approaches to language development, commissioned by Health Canada's Canadian Working Group[14] on Childhood Hearing, could find little substantive evidence of acceptable quality in support of any specific approach. Certainly, auditory verbal therapy has achieved remarkable success in the context of cochlear implants, but such success says little about whether an alternative approach might yield equivalent or superior outcomes.

Overall Program Performance Issues, Benefits, and Harms

System Performance

An EHDI program is only as strong as its weakest link. Each program component (eg, newborn screening, audiologic assessment) may be individually optimized, but the linkages between the elements are crucial for overall success. For example, screening test protocol sensitivity may be 90%, but suppose that only 85% of the newborn population were screened, 80% of those with screening refer results attended audiologic assessment, and 75% of those with significant PCHI actually received an appropriate and timely intervention. Then

the effective sensitivity of the program is $0.9 \times 0.85 \times 0.80 \times 0.75 = 0.46$ or 46%! Losses accumulate at every stage, but the overall performance can be no better than the lowest probability value.

Most good EHDI programs are now achieving over 90% coverage, but losses to follow-up are a serious limitation in many US state programs, for example. Losses between predischarge screening and rescreening or audiologic assessment may be very sensitive to the way in which the meaning of the screening refer result is explained. In an attempt to minimize anxiety, such as by dwelling on probable transient, middle ear conditions or the generally low positive predictive value of a refer result on initial screening, personnel may actually convince families that there is no need to attend for follow-up. This may be reinforced by informal and completely invalid "tests" of hearing by the family. A careful balance must be struck with families between undue anxiety and an inappropriate lack of concern, especially prior to the definitive audiologic assessment.

Once PCHI is confirmed, it is usually appropriate to start audiologic intervention promptly. Significant delays in delivery of, say, a hearing aid compromise the whole point of systematic early detection. If a PCHI is shown to be absent, it is important to make families aware that a normal hearing test does not guarantee that hearing will remain normal and that ongoing attention to responsiveness and communication development milestones is needed.

Benefits of EHDI

Traditionally, arguments for EHDI have emphasized long-term language development as the key outcome. This is a peculiar and flawed position. The primary disorder is hearing impairment, and negation or amelioration of that impairment is obviously the primary health outcome. Improved hearing is not a proxy for language development or quality of life. Rather, improved language or quality of life is an important secondary outcome, for which hearing ability is a causal intermediate factor. To emphasize the point, if an adult were afflicted with a sudden, bilateral PCHI, what would be sought is remedy of the impairment, not an examination of possible impact on quality of life. It can be argued that the child's ability to hear is at least as important as that of the adult, probably more so during a period of rapid neurologic, cognitive, and social development.

Improved hearing ability is achievable for many infants by appropriate fitting of hearing aids or cochlear implants. The evidence of improved audibility by cochlear implants is overwhelming, and for conventional hearing aids, it is in large measure a matter of acoustics. It does not require a lengthy clinical trial to prove that 35 dB of amplifier gain will render a 50 dB signal audible in the presence of a 60 dB hearing loss. The overall amount of audibility benefit of this kind (hearing months/years) accruing from EHDI programs depends on the number of cases who receive effective audibility intervention under EHDI, together with the reduction in age at which the intervention is delivered, relative to pre-EHDI performance, such as was reported by Durieux-Smith and Whittingham.[4]

Proof that EHDI improves long-term language has been difficult to establish. This is no surprise because measures of early language development are imperfect and there is a host of confounding variables that affect language development. Validating EHDI by language measures at age 5 years is as oblique as would be validating newborn vision screening by reading ability at school entry. An important report by Yoshinaga-Itano and colleagues indicated that identification of PCHI under 6 months of age led to favourable language outcomes.[15] This was widely heralded by the clinical community as proof of effectiveness but did not convince the US Preventive Services Task Force, which maintained that there were significant methodologic limitations of the available evidence and that the evidence base at that time for EHDI effectiveness was "inconclusive."[16,17] Very recently, convincing evidence from the only large controlled trial of UNHS to date has been presented that UNHS is associated with substantial long-term gains in receptive and expressive language.[18,19] Preliminary information about that evidence was given by Kennedy and McCann.[1]

Harms from EHDI

The main harm that is speculated to arise from EHDI is family anxiety, especially that due to false-positive screening tests. Because the prevalence of PCHI is low, even though hearing screening tests have very good specificity, there may be 10 families who experience such a result for every 1 infant correctly identified with PCHI. The evidence to date indicates that the proportion of families who experience measurable and significant anxiety is very small and that most of whatever anxiety exists is not long-lasting. See Young and Andrews[20] and <www.childhearinggroup.isib.cnr.it> or google on "UNHS and family anxiety" for reviews of the evidence on anxiety and other speculative harms, such as the impact on parent-child bonding.

Role of Physicians in EHDI Programs

The support and active participation of physicians are absolutely crucial to successful EHDI. The precise nature of the involvement is context specific. In the United States, for example, pediatricians have assumed a very prominent role in EHDI program advocacy and implementation; see, for example, the American Academy of Pediatrics (AAP) Web site <www.aap.org> for further information and links to UNHS and EHDI.

Irrespective of the exact context, however, some key areas of physician involvement include the following:

1. Promoting family awareness of the nature and importance of EHDI
2. Strongly encouraging family compliance with EDHI program processes
3. Making prompt referrals into EHDI based on keen awareness of risk indicators
4. Providing prompt management of middle ear disorders to minimize their impact on audiologic interventions for PCHI
5. Providing timely and comprehensive etiologic assessment of all PCHI cases, including additional tests and referrals
6. Facilitating prompt audiologic intervention by any practicable means
7. Providing information and support for families that reinforces and synergizes with that provided by the EHDI program
8. Contributing leadership to program advocacy, quality improvement, and facilitation of change to a new pattern of hearing health care service provision

Medical Assessment and Management of Infants with PCHI

There is a need for definition of a systematic, evidence-based process for etiologic investigation of infants with proven PCHI. The purpose of etiologic assessment is to determine the potential cause(s) of the PCHI. This may involve additional examinations, tests, and referrals. The process may lead to identification of specific pathologies or to syndromes and may have implications for management and prognosis in relation to hearing or beyond. Genetic studies are increasingly common because the number of known genetic causes of specific auditory dysfunctions is now in the hundreds and growing rapidly. Nance and Roizen have provided useful reviews of genetic and nongenetic causes of hearing impairment, respectively.[21,22] See the risk indicator guidelines of the US Joint Committee on Infant Hearing,[12] the review by Fortnum of the epidemiology of PCHI,[2] and Doyle and Ray on the role of the otolaryngologist.[23] See Greinwald and colleagues, Preciado and colleagues, and Morzaria and colleagues for more detailed information on approaches to evaluation of the young child with PCHI.[24–26] See also the 2004 AAP/American Academy of Family Physicians guidelines on diagnosis and management of acute otitis media.

A detailed discussion of evaluation and management approaches is beyond the scope of this article, but Maki-Torkko has provided a very comprehensive review of current issues and recommendations.[27] Currently, it is not unusual for cohorts of infants with PCHI to manifest at least a 40% rate of unknown etiology. A recent systematic review reported a 38% rate of unknown etiology in children with PCHI of at least 40 dB HL bilaterally.[28]

It may be feasible to reduce the proportion of unknown etiology substantially through a more systematic approach to clinical evaluation. In addition to the core of careful physical examination, family history, birth history, and audiometric findings, the etiologic assessment may include computed tomography, urinalysis, electrocardiography, ophthalmologic assessment, viral serology, thyroid function tests, and genetic assessments and testing, including screening for common mutations, such as 35delG for connexin 26 and any others associated with potential syndromes. The relevance, timing, and utility of some of these components are matters of judgement and ongoing debate.

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