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Early Hearing and Communication Development

Canadian Working Group on Childhood Hearing (CWGCH) Resource Document

Canada

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Foreword

In response to growing interest in the field of early hearing and communication development (EHCD), the Health Surveillance and Epidemiology Division in Health Canada (now part of the Public Health Agency of Canada) established the Canadian Working Group on Childhood Hearing in September 2000.

The Working Group was multidisciplinary and included representatives of national professional associations, a parent representative and experts in otolaryngology, audiology, speech-language pathology, deaf education, nursing, child health and public health from across Canada. Its mandate was to review and evaluate scientific evidence in areas essential to the development of EHCD programs, and to develop a report that could function as a resource document.

The Working Group produced a draft report in early 2003, and then held consultations across Canada with representatives from provincial/territorial governments, health professionals, researchers, educators and consumer groups to get feedback on the draft report. The cooperation and participation of these individuals is gratefully acknowledged.

This resulting report constitutes a part of the information base from which EHCD programs and policies may be developed. It should facilitate the development of a framework of Canada-wide common goals, practices and procedures in the area of EHCD. The development and implementation of EHCD programs will vary provincially and regionally, depending on human, financial and material resources, and on the policies and priorities of those provinces and regions.

I wish to thank the members of the Canadian Working Group on Childhood Hearing for the many hours they dedicated to this project. Thanks also to the staff of the Public Health Agency of Canada for their hard work. We all hope that this resource document will be useful to our colleagues across the country who provide these important services to Canadian children and their families.

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Executive Summary

In the past decade, identification of hearing impairment in early infancy has emerged as an important public health issue. This has been spurred primarily by significant technological advances in hearing screening tests. Population-wide screening of newborns for hearing impairment has been frequently advocated and is now being widely implemented — for example, in Ontario, the United States and the United Kingdom. However, in the health services community, there remains a lack of full consensus about the appropriateness of universal newborn hearing screening.

Given the importance of this issue, as well as evidence of diverse patterns of early identification of hearing impairment throughout Canada, Health Canada established the Canadian Working Group on Childhood Hearing (CWGCH) in 2000. The Working Group was a multidisciplinary expert body that included relevant health professionals and public representatives.

The goal of the Working Group was to provide information that will assist individuals or agencies considering the development of programs for early hearing and communication development (EHCD). The approach selected was to develop and disseminate a summary of the latest scientific information on key aspects of the rationale and methods for EHCD. The Working Group adopted an evidence-based method in order to go beyond clinical opinion and withstand scientific scrutiny. The World Health Organization principles of screening and the conceptual framework of the International Classification of Functioning, Disability and Health guided the evidence reviews and terminology used.

The Working Group addressed the following areas:

- the burden of the disorder, including the number of children affected by hearing impairment (prevalence) and patterns of detection
- hearing screening tests
- audiologic assessment
- medical evaluation and management
- amplification
- effectiveness of different approaches to communication development

For these topics, the Working Group conducted formal evidence reviews by standard scientific methods, to the fullest extent possible within resource and timeline constraints. Working Group members also considered program infrastructure, evaluation and quality improvement, and cost-effectiveness, but did not formally review these topics.

The following are the main findings of this process:

The burden of the disorder is substantial. The prevalence of permanent childhood hearing impairment is about 1 per 1,000 live births in infancy for impairment greater than 40 dBHL in the better ear. In infants with documented risk factors such as extreme prematurity, congenital facial auricular defects or severe jaundice, this rate is up to 10 per 1,000.

In the absence of systematic screening, the detection, confirmation, diagnosis and management of hearing impairment are significantly delayed. With universal newborn hearing screening, the median age of diagnosis is less than 3 months.

Hearing screening is currently based on automated otoacoustic emission testing and automated auditory brainstem response testing. These tests perform well when appropriate protocols are used. Loss to follow-up is the largest single factor limiting the effectiveness of screening.

Complete audiologic assessment, which can be achieved in healthy children under 6 months of age, is essential to appropriate hearing aid fitting and to family decisions about communication development options. The existing literature is unclear with respect to the audiologic assessment and management of auditory neuropathy.

Medical evaluation of an infant with hearing impairment should be initiated at less than 3 months of age and management (e.g., amplification) should be started by 6 months of age. Common causes of bilateral sensorineural hearing impairment include:

- nonsyndromic gene mutations, such as connexin 26 mutations
- genetic syndromes, such as Waardenburg syndrome
- nongenetic causes, including preterm birth, asphyxia, meningitis, kernicterus, intrauterine infection and auditory neuropathy

There is a need for evidence-based rational decision strategies, embracing history taking, physical examination, risk assessment and genetic testing and their interpretation, in children with bilateral sensorineural hearing impairment.

Between 70% and 90% of children will experience fluctuating conductive hearing impairment secondary to otitis media with effusion, with or without acute otitis media, in the first two years of life. Unilateral otitis media with effusion clears after an average of five weeks with or without a history of acute otitis media. Bilateral otitis media with effusion clears on average after eight to nine weeks. Bilateral myringotomy and ventilation tube placement reduces the mean duration of otitis media with effusion and improves hearing thresholds, as well as some behavioural problems and expressive language scores in some children. Antibiotic therapy and conjugate pneumococcal vaccine should be considered in relation to middle ear disease in children.

Accurate assessment of otitis media with effusion is important for the diagnosis and treatment of children with sensorineural hearing impairment. Many physicians advocate an aggressive treatment approach to children with otitis media with effusion and underlying sensorineural hearing impairment.

Hearing aids can improve auditory performance in children with auditory impairment who have some hearing bilaterally. To ensure the accurate fitting of amplification in young infants, highly qualified professionals and paediatric-specific hearing instrument fitting protocols are required.

Based on existing research, it is not possible to determine the effectiveness of the four most common communication development options for children with permanent congenital hearing impairment: aural-oral, auditory verbal therapy, American Sign Language and total communication.

Some studies have concluded that early identification and strong family involvement improve the development of speech and language in infants and young children with hearing impairment.

Public health system models and linkages seem more appropriate than traditional medical models for effective delivery of comprehensive EHCD programs.

Universal newborn hearing screening must be accompanied by appropriate, accessible services for confirmation, diagnosis and effective hearing and communication development options for all children referred through screening. EHCD programs should reflect demographic and cultural factors as well as existing systems, infrastructure and well-developed collaborative links with other health care, social support and educational systems.

The CWGCH has concluded that newborn hearing screening leads to early identification of hearing impairment. Early identification leads to improved hearing and facilitates communication development. The detailed reviews of evidence underlying this position are available on request.

This document also contains recommendations for further research. For example, the generation of evidence on a broad range of possible outcomes for affected children and families is a high priority.

The issues of ethics and societal values are beyond the scope of this report. Nevertheless, these questions are crucial and should be weighed, along with the available evidence, when considering whether to implement a new EHCD program.

Chapter I: Introduction

Purpose

Author: Sharon Bartholomew

The Canadian Working Group on Childhood Hearing (CWGCH) has prepared this document as an evidence-based resource on early hearing and communication development (EHCD). The evidence is reviewed and presented here for the reader by subject. The final chapter draws conclusions where possible and briefly addresses the broader context that needs to be taken into account in the development of programs.

The intention of the CWGCH is that this resource document will be useful for individuals in jurisdictions in Canada who are making EHCD policy and programming decisions. Overall, the desired outcome for this document is to enhance EHCD programs and services for children.

The document has several purposes. It will:

- lead to an increase in the use of evidence-based knowledge on newborn hearing screening, diagnostic testing and early intervention
- serve as a dissemination vehicle for current evidence on early hearing and communication development
- encourage evidence-based approaches in support of best practices
- make a statement on conclusions that will summarize the evidence
- lead to the lowering of the age of initiation of intervention and improve long-term outcomes for children with hearing impairment

The target audience for this document includes:

- advocates for children with hearing impairment
- decision makers at all levels (bureaucrats and politicians)
- health professionals (audiologists, speech and language pathologists, otolaryngologists, paediatricians, neonatologists, family practitioners, etc.)
- educators of health professionals
- researchers in the field of hearing impairments
- opinion leaders

Approach

The CWGCH was established in September 2000 by the Health Surveillance and Epidemiology Division in the Public Health Agency of Canada (formerly the Population and Public Health Branch of Health Canada). It was established in response to a growing interest in the field of EHCD.

Membership on the Working Group included: representatives of national professional associations; consumers/parents; and individual experts in otolaryngology, audiology, speech-language pathology, nursing, child health and public health from across Canada. The multidisciplinary nature of the Working Group is critical to the success of this document. All of the relevant disciplines impacted by EHCD were included on the CWGCH, so that this resource document will be as relevant as possible to a broad range of individuals and professions.

In order to produce this report, critical and systematic reviews were conducted in the following areas:

1. Screening Justification (Burden of Target Disorder)
2. Diagnostic Methods/Technology
3. Amplification
4. Assessment Techniques
5. Medical Management
6. Communication Development Options

These reviews were not done as formal systematic reviews,[1] apart from that on communication development options. However, they were completed by experts in the field using common time periods (1966–2002) and a variety of electronic databases, gray literature and manual searching of relevant journals. The term "critical reviews" was used instead of either "narrative reviews" or "systematic reviews." The individual reviews were agreed upon by the group before being finalized. The full reviews were summarized for this document and the complete English versions can be obtained through the Maternal and Infant Health Section, Health Surveillance and Epidemiology Division, Public Health Agency of Canada.

Consultations

In order for this document to be as relevant as possible to its target audience, consultations were held on a draft version. These consultations were held across Canada over a period of six months during 2003. The invitees to the consultations included representatives from provincial/territorial governments, professionals, researchers, educators and consumer groups. The feedback from these consultations was very valuable and has been incorporated into this report.

Terminology Used in this Report

The readers should be advised that some of the terminology used in this document may be different than what has been in common use before. This was a conscious decision by the CWGCH to more accurately reflect the work currently being done in the field. Some examples of changes in terminology include:

New Term/Phrase	Replacing
• early hearing and communication development	• early hearing detection and intervention
• hearing and communication development options	• interventions

While the term "early hearing detection and intervention" (EHDI) is popular, especially in the United States, it has significant limitations. The term "intervention" is non-specific and may be seen as inappropriate in a context that emphasizes a family-centred process aimed at early and maximal development of hearing function and communication function.

The term "early hearing and communication development" (EHCD), on the other hand, emphasizes the primary goal of enhancing development of communication function, which may include oral language, visual language and speech. It also connotes accelerated development of hearing ability, which is a desired outcome of early detection of hearing impairment. The ability to hear is of value in itself as well as by virtue of its intermediary role in communication development. The term also accentuates the positive (as does the carefully chosen "early hearing detection" component of EHDI), and also emphasizes the ultimate purpose of the program activity, rather than the necessary but not sufficient components of it, such as detection of hearing impairment.

The CWGCH would also like to clearly define how the term "universal newborn hearing screening" (UNHS) is being used in this document. UNHS refers specifically to population-wide screening, and not to EHCD as a whole.

The term "hearing impairment" has been chosen for use in this document rather than "hearing loss." Although the term hearing loss has become more popular in North America, the CWGCH believes that for this document hearing impairment is more appropriate and more consistent with the World Health Organization (WHO) *International Classification of Functioning, Disability and Health* (ICF).[2]

Guiding Principles

The CWGCH is committed to the following guiding principles:

(i) National Role

The CWGCH will provide leadership in the development and dissemination of a resource document on early hearing and communication development (EHCD) in Canada. This report will be a reference for individuals in all provinces and territories in Canada who wish to develop their own EHCD program or policy.

(ii) Evidence-Based Approach

The CWGCH will take an evidence-based approach to the development of the resource document for EHCD. This approach involves the systematic and critical review of currently available research and program information on childhood hearing.

(iii) Family-Centred Approach

The CWGCH endorses an integrated approach to families, reflecting an understanding of the physical, emotional, mental and psychosocial aspects of hearing and communication development (HCD) options for children with hearing impairment and their families.

(iv) Partnership and Collaboration

The CWGCH is a partnership with various stakeholders, including federal, provincial and territorial governments; professional associations; consumers/parents; and national and international experts in otolaryngology, audiology, speech-language pathology, nursing, child health and public health. This report on EHCD is a result of collaboration among all stakeholders to build on experiences, create linkages and provide opportunities for further capacity building and promotion of best practices in HCD.

(v) International Classification of Functioning

The CWGCH believes that it is important that the WHO's ICF be used to guide the terminology in clinical practice, research and programming. The ICF offers a conceptual framework for information that is applicable to personal health care. This includes prevention, health promotion and the improvement of participation by removing or mitigating societal hindrances and encouraging the provision of social supports and facilitators.

The ICF, which was published in 2001, succeeds the *International Classification of Impairments, Disabilities and Handicaps* (ICIDH). It has progressed from describing the consequences or outcomes of chronic disease to a classification of human functioning and disability.

The four main aims of the ICF are to:

1. Provide a scientific basis for understanding and studying health and health-related states, outcomes and determinants.
2. Establish a common language for describing health and health-related states in order to improve communication between different users, such as health care workers, researchers, policy makers and the public, including people with disabilities.
3. Permit comparison of data across countries, health care disciplines, services and time.
4. Provide a systematic coding scheme for health information systems.[2]

Principles and Practices of Screening

The basic purpose of screening is to detect individuals who are at risk for a condition by applying a test to all asymptomatic persons in a population. The goal of screening is to improve the outcome in affected individuals.

In order for a screening program to be justified it should meet certain criteria. A classic example of these criteria is presented in the WHO document *Principles and Practices of Screening for Disease*, 1968.[3]

The definition of screening used by the WHO comes from a conference on chronic illness held in 1951 where screening was defined as "the presumptive identification of unrecognized disease or defect by the application of tests, examinations, or other procedures which can be applied rapidly." The document goes on to point out that screening tests sort out apparently well persons who probably have a condition from those who probably do not. A screening test is not intended to be diagnostic. Persons with positive or suspicious findings must be referred to physicians or other specialists for diagnosis and necessary treatment.[4]

Principles and Practices of Screening for Disease[3] describes the aims of early detection as: discovering and managing conditions which occur in the presence of pathological change, but which have not so far reached a stage at which medical aid is sought spontaneously; and achieving more per unit expenditure by saving the time of highly trained professionals, since part of the work can be performed by less highly trained personnel who are able to carry out screening tests.

The 10 principles of early disease detection discussed in the WHO document are:

1. The condition sought should be an important health problem.
2. There should be an accepted treatment for patients with recognized disease.
3. Facilities for diagnosis and treatment should be available.
4. There should be a recognizable latent or early symptomatic stage.
5. There should be a suitable test or examination.
6. The test should be acceptable to the population.

7. The natural history of the condition, including development from latent to declared disease, should be adequately understood.
8. There should be an agreed upon policy on whom to treat as patients.
9. The cost of case finding (including diagnosis and treatment of patients diagnosed) should be economically balanced in relation to possible expenditure on medical care as a whole.
10. Case finding should be a continuing process and not a "once and for all project."

The WHO principles have been used, in whole or in part, for justifying screening programs for many years. However, these principles have been seen to fall short in a few areas for today's health care situation. The National Screening Committee of the British National Health Service found it could improve on the WHO principles by including criteria around three issues: the adverse effects of screening; the opportunity costs associated with screening; and, instead of suggesting that there be an acceptable treatment, adding a statement about the strength of the evidence regarding the treatment (see Table 1).

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Table 1: Criteria Developed by the National Screening Committee[5]

The Condition	1. The condition should be an important health problem. 2. The epidemiology and natural history of the condition, including development from latent to declared disease, should be adequately understood, and there should be a detectable risk factor, disease marker, latent period or early symptomatic stage. 3. All the cost-effective primary prevention interventions should have been implemented as far as practicable.
The Test	4. There should be a simple, safe, precise and validated screening test. 5. The distribution of test values in the target population should be known and a suitable cut-off level defined and agreed. 6. The test should be acceptable to the population. 7. There should be an agreed policy on the further diagnostic investigation of individuals with a positive test result and on the choices available to those individuals.
The Treatment	8. There should be an effective treatment or intervention for patients identified through early detection, with evidence of early treatment leading to better outcomes than late treatment. 9. There should be agreed evidence-based policies covering which individuals should be offered treatment and the appropriate treatment to be offered. 10. Clinical management of the condition and patient outcomes should be optimized by all health care providers prior to participation in a screening program.
The Screening Program	11. There should be evidence from high-quality randomized controlled trials that the screening program is effective in reducing mortality or morbidity. 12. There should be evidence that the complete screening program (test, diagnostic procedures, treatment/intervention) is clinically, socially and ethically acceptable to health professionals and the public. 13. The benefit from the screening program should outweigh the physical and psychological harm (caused by the test, diagnostic procedures and treatment). 14. The opportunity cost of the screening program (including testing, diagnosis and treatment) should be economically balanced in relation to expenditure on medical care as a whole. 15. There should be a plan for managing and monitoring the screening program and an agreed set of quality assurance standards. 16. Adequate staffing and facilities for testing, diagnosis, treatment and program management should be available prior to the commencement of the screening program. 17. All other options for managing the condition should have been considered (e.g., improving treatment, providing other services).

Chapter II: Background

The Canadian Historical Perspective on Early Hearing and Communication Development

Author: Dr. Andrée Durieux-Smith

In the past 35 years, the importance of the early identification and management of hearing impairment in children has been the subject of many conferences and task forces in Canada. During this time, recommendations have been formulated addressing the need to identify permanent childhood hearing impairment (PCHI) early in children, with three main themes recurring consistently. These include: the methods to identify hearing impairment accurately in newborns and infants; the population to be screened; and the need to educate physicians, other health care professionals and parents on the signs of hearing impairment in children.

In the 1960s, a conference on "The Young Deaf Child" took place in Toronto, bringing together more than 30 experts from North America, Great Britain, Scandinavia and the Netherlands.[1] The objective of the meeting was to find ways to alleviate the handicap to auditory communication imposed by early hearing impairment. The participants were already aware of the importance of early hearing and communication development (EHCD). Considerable discussion took place on "definitive tests of hearing." Systematic reviews of available tests were presented including new electrical techniques to detect cortical evoked responses to sound in young infants. The hope of identifying hearing impairment soon after birth was expressed, but the technology was not yet available. Neonatal hearing testing was seen as a goal; even then, the possibility of universal neonatal testing was discussed but seen as not achievable because of a lack of accurate methods.

Participants at this conference decided that a focus on infants at risk was a good starting point. However, consensus was not reached on the age at which the use of amplified sound should be initiated. The majority agreed that two years was the maximum delay that could be acceptable, even though some advocated for the use of amplification by 2 months of age. Participants felt that, although research on the most valid and reliable methods was necessary, programs needed to be developed to identify children by 6 months of age. Two steps were recommended — the use of a high-risk register, and the screening of healthy babies in well-baby clinics using simple, well-planned tests and questionnaires.

Ten years later in the 1970s, the Nova Scotia Conference on the Early Identification of Hearing[2] took place in Halifax. The focal point of discussion at this conference was the approval of methods for screening the hearing of newborns and for identifying children most likely to have a hearing impairment. The recommended procedures included the use of the high-risk register, together with behavioural screening. It is now well known that screening newborns and infants with behavioural tests is not sensitive, specific or reliable.[3] Discussions took place on the most accurate methods

to assess hearing impairment in infants and whether this was possible. The resulting queries became the subject of a second conference entitled the Early Diagnosis of Hearing Loss in Children,[4] which took place in Saskatoon. This conference dealt with methods for confirming the presence and degree of hearing impairment within the first six months of life as accurately, rapidly and economically as possible.

Four papers were presented on electrophysiological methods at the Saskatoon conference, and the auditory brainstem response (ABR) was seen as a viable method to accurately identify hearing impairment in graduates of a neonatal intensive care unit (NICU).[5,6] Although it was agreed that more research on ABR was necessary, the clarion call of the Saskatoon conference was that it is possible to identify hearing impairment in the newborn. This was a major breakthrough. At the time, ABR equipment was expensive and its use was seen as being restricted to the screening of high-risk infants. One of the major recommendations of this conference was that infants should be diagnosed by 6 months of age and management initiated immediately.

In the 1980s, several centres in Canada began to carry out research using ABR, most often with babies from NICUs. The results of this research were presented at a symposium — the Canadian Experience in Neonatal Hearing Assessment by ABR,[7] which took place in 1983 during the biennial meeting of the Electric Response Audiometry Study Group in Ottawa. The research presented at the symposium by Canadian researchers clearly showed that ABR was a powerful tool for the identification of hearing impairment in newborns and infants.

Also in the 1980s, the Health Services Directorate of Health and Welfare Canada (now Health Canada) established a multidisciplinary Task Force on Childhood Hearing Impairment. One of the objectives of the Task Force was to document the activities taking place in each province and territory in the areas of prevention, early detection, diagnosis and management of children with a hearing impairment. Another objective was to develop consensus guidelines that would facilitate the development of strategies leading to the EHCD of children with a permanent hearing impairment. In addition, an awareness campaign on hearing impairment in children, funded by the Health Promotion Directorate, was launched to alert primary care physicians to the importance of early hearing detection and intervention in children with hearing impairment.[8]

To meet one of the objectives of the Task Force, questionnaires were sent to all provincial and territorial health and education ministers to gain information on the identification and management of children with hearing impairment in Canada. The results indicated that no province-wide policies existed at the time, that regions were developing their own individual programs, and that some programs existed in local hospitals as a result of local initiatives. The Task Force developed recommendations to address these issues which were published in a report in 1985.

In the 10 to 15 years that followed, significant progress took place in the development of rapid, valid, reliable and cost-effective technology based on the use of objective physiological measures. In addition, universal screening became the recommendation as targeted screening of high-risk newborns was seen as missing a large number of children with PCHI who had no identifiable risk factors.[9,10]

At a time when, in the United States, large universal newborn hearing screening (UNHS) programs were successfully developed,[11-14] a survey was carried out to determine the state of hearing screening programs in Canada. The survey results showed that only 10% (35 out of 384 respondents) of birthing hospitals in Canada had some kind of newborn hearing screening program, and that a wide variety of hearing screening approaches were used. The results of the survey, which was carried out 15 years after the report of the Task Force on Childhood Hearing Impairment was released, also showed that very few of the task force recommendations were in fact carried out.

In 2000, the Canadian Association of Speech-Language Pathologists and Audiologists and the Canadian Academy of Audiology published a Position Statement on Universal Newborn and Infant Hearing Screening in Canada.[15] It shows that large systematic programs are at different stages of development in many parts of Canada. In 2000, for example, the Ontario government announced that an Infant Hearing Program would be developed for the province — newborns have been screened since 2002. And in Alberta, a grant from the Alberta Health Innovation Fund has led to the development of a demonstration project on newborn hearing screening.

In addition, at the time of writing this report, the provinces of New Brunswick and Prince Edward Island had announced the development of provincial EHCD programs.

Conclusions

- The importance of the early identification and management of hearing impairment in children has been recognized in Canada for almost four decades.
- There was no systematic approach to the development of region-wide early hearing and communication development (EHCD) programs before 2000.
- Since 2000, programs have been developed in parts of Alberta, New Brunswick, Ontario and Prince Edward Island, and they are at different stages of development in many other provinces and territories.

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Brief International Overview

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Many countries are addressing questions and issues surrounding early detection of deaf and hard of hearing newborns and promotion of communication development.

In the United States, the National Campaign for Hearing Health (NCHH) reported that, as of May 2003, 86.5% of all babies born in the U.S. were being screened, and that 38 states as well as the District of Columbia had early hearing and communication development (EHCD) legislation or mandates. Interestingly, of the 13 states reported to have no legislation or policies in place, some reported screening rates as high as 97.6%. Screening rates by state ranged from 22% to 99.5%, but more than two thirds of all states reported that more than 90% of babies were being screened.[1]

The quality of the U.S. screening programs was also evaluated. The quality criteria were: the percentage of babies screened, the presence of a state-wide system of coordination, training, quality assurance and establishment of follow-up. Of all the state programs, 40 were rated "excellent" (90%+ coverage and the presence of the other quality indicators); 5 were "good"; and 6 were rated "unsatisfactory" (less than 79% of babies had been screened and there was an absence of the other quality indicators). These evaluations highlight the importance of considering not only screening coverage, but also the other key factors that contribute to program quality.[1]

In the United Kingdom, the Newborn Hearing Screening Programme (NHSP) aims to implement hearing screening for all newborns. It will be implemented gradually across the country in three phases, with all areas participating by 2006. As of early 2003, the 23 areas of the first phase had introduced the program and the second phase areas also started participating in early 2003. The NHSP website provides a wealth of information about this program (http://www.nhsp.info).

In Australia, New South Wales is the only state offering universal newborn hearing screening (UNHS) that is funded by the state government. In Western Australia, a state-funded newborn hearing screening program exists in several hospitals that cover about half of the state's births. In other states there is a combination of state-funded and individual hospital coordinated at-risk screening programs. All Australian states and territories have groups that are actively lobbying for UNHS.[2]

The great international interest in early identification and related issues has been evident in four international conferences that have been held in Como, Italy, since 1998: the European Consensus Conference on Neonatal Hearing Screening (1998), and the International Conference on Newborn Hearing Screening, Diagnosis and Intervention (2000, 2002 and 2004). These conferences have provided an international forum for delegates from over 50 countries to share ideas on a spectrum of topics relevant to the evolution of the world-wide phenomenon of ECHD. Information about these conferences can be found on the 2004 International Conference on Newborn Hearing Screening, Diagnosis and Intervention website (http://www.NHS2004.polimi.it).

As of early 2003, there is no central international inventory of programs but a number of useful electronic sources can be accessed for the most current information. The National Center for Hearing Assessment and Management (NCHAM), located at Utah State University in the United States, has a website (http://www.infanthearing.org) that provides a plethora of information including, for example, basic information for starting programs, available resources and related U.S. statistics. Also located on the website is a newsletter, *Sound Ideas*, published quarterly, which frequently provides international updates. For example, in November 2002, the newsletter published a report about Croatia, which stated that the country is "well on its way to having a nationwide newborn hearing screening, diagnosis and intervention program."

The European Commission in Brussels (Quality of Life Programme) also has an informative publication — the Newsletter of Project AHEAD II, which stands for Advancement of Hearing Assessment Methods and Devices — Immediate Intervention (http://www.biomed.polimi.it/aheadii/ahead_ii_frames.htm). This newsletter provides international updates and conference/meeting information several times a year for those interested in keeping abreast of the international situation.

Conclusions

- Many countries such as the United States and the United Kingdom have developed universal newborn hearing screening (UNHS) programs and programs related to communication development.
- Other countries are at different stages of program development.

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Chapter III: Burden of the Target Disorder

Target Disorder

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The "burden" of the target disorder is a term that refers to the overall set of negative consequences that may be a result of its occurrence. The target disorder is most commonly (albeit, not necessarily) defined in terms of parameters of hearing impairment, such as severity, laterality, type and time at expression. The burden of the disorder reflects its negative impact on the individual child and family, and upon society as a whole. The latter aspect of burden is governed primarily by the number of affected children and families (prevalence of the disorder) and by its cultural and socioeconomic effects.

The impact of a given hearing impairment on a particular child and family is widely believed to depend on many variables, and especially upon when the impairment is identified and what services are engaged to address it. These services may include, but are not limited to: family information and counseling; psychological, social and economic supports; assistive technologies such as personal amplification; other assistive devices; and cochlear implants. Instruction aimed at development of communication skills may include manual, oral or combined approaches. The effectiveness of all these services will depend on their nature, timeliness, accessibility, quality and acceptability. There is always *some* particular pattern of events related to service performance (or the lack of it), and so the impact of the target disorder must be considered as intimately connected to the current service pattern and its effects. For example, if it is assumed that high quality services are effective, then if services were inaccessible, inappropriate or of poor quality, the negative impact of a given spectrum of the target disorder would be increased.

For the purposes of this document, the primary focus in target disorder definition is upon hearing impairment that is congenital and is stable or progressive. This is referred to as permanent childhood hearing impairment (PCHI). Etiologically, most such impairment involves cochlear dysfunction and is medically irremediable. Structural conductive impairments which may arise from maldevelopment of the external or middle ears are usually included because they impose long-standing dysfunction, unless treated surgically.

Prevalence of Permanent Childhood Hearing Impairment

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The prevalence of permanent childhood hearing impairment (PCHI) in the age range 0–5 years is reviewed in this section. Most PCHI is sensorineural but structural conductive impairment (e.g., ear canal atresia) is included. **Congenital impairment** is defined as impairment recognized at birth or believed to have been present since birth. **Late-onset** impairment is impairment not present at birth and that cannot be attributed to an exogenous cause. **Acquired impairment** is impairment not present at birth and for which an exogenous cause can be identified. **Progressive impairment** is impairment that increases over time, regardless of its point of initial manifestation.[1]

These terms must be interpreted with great caution, because most datasets were obtained in the absence of comprehensive, valid and accurate hearing assessment in the neonate and infant. Thus, with the possible exception of recent, high quality, universal newborn hearing screening (UNHS) studies, prevalence of strictly congenital PCHI probably has been overestimated, and that of late-onset, acquired and progressive impairment underestimated.

Variation and Bias in Prevalence Estimates

There is large variation in reported prevalence. Major sources of variation include: random sampling error; the definition of the target disorder; the completeness and accuracy of the determination of hearing status; the definition of the target population; the methods by which that population is sampled; the age of the study population; demographic, cultural and behavioural factors; and the availability and quality of perinatal care.

Ascertainment Studies

Ascertainment studies involve case identification through notification systems or database surveys. Negative bias due to under-ascertainment is a concern, as are geographic consistency of case capture and the stability over time of the case-finding system, of the underlying population epidemiology, and of the hearing impairment measurement practices.

By far the most comprehensive ascertainment-based prevalence report to date is due to Fortnum et al.[1] This was a well-executed ascertainment study of a 15-year birth cohort in the U.K. (1980–1995), with 17,160 cases ascertained. The target disorder was PCHI of greater than 40 dBHL average over 0.5, 1, 2 and 4 kHz in the better ear. Adjusted prevalence was reported as 1.07 per 1,000 live births at 3 years and 2.05 per 1,000 live births at more than 9 years of age. Possible causes of the increase with age include progressive or delayed-onset PCHI, delayed confirmation of congenital PCHI, and acquired PCHI. There is insufficient information to resolve the relative contributions of these factors, all of which may contribute significantly.

The prevalence findings are reasonably consistent with those from other authors. Studies with substantial sample sizes were reviewed. They revealed significant effects of the target disorder criterion, as well as differences across demographic subgroups. The estimated congenital prevalence of moderate or greater hearing impairment for the accepted studies ranges from 0.09 to 1.16 per 1,000 live births. It should be noted that all these studies reported on hearing levels in the better ear, that is, on bilateral impairment. Also, they targeted average hearing levels over a frequency range. This represents a conservative approach to the definition of target PCHI.

Prevalence Estimates from Universal Newborn Hearing Screening Programs

Prospective prevalence estimates may be obtained from reports of large newborn hearing screening programs. Because only newborn screening referrals are followed up, UNHS programs are not cohort studies and they provide information exclusively about truly congenital PCHI. Other important limitations of UNHS studies are that prevalence estimates have large confidence intervals (due to the limited number of cases in typical study samples), and they may be biased due to incomplete screening coverage and follow-up or due to study-specific characteristics. Also, there is variation in target disorder definition, in the sensitivity of screening methods used, and in the accuracy and timing of hearing assessments.

The highest quality UNHS data was provided by the New York State UNHS demonstration project.[2] For this program, the adjusted prevalence of hearing impairment greater than 20 dB in either ear was 2.8 per 1,000 live births. For five acceptable program reports addressing at least mild, congenital PCHI in any ear (unilateral or bilateral impairment), the median unadjusted prevalence was approximately 2.2 per 1,000 live births. This value is biased negatively by incomplete follow-up. The adjusted median estimate accounting for children lost to follow-up is 3.2 per 1,000 live births.

The contrast between the UNHS estimates of congenital prevalence for PCHI criteria of *better-ear* >40 dBHL (Wessex and East London, average 1.06/1,000) versus the adjusted *any-ear* >20 dBHL value of 3.2 per 1,000 live births suggests that the prevalence of hearing impairment may triple if the severity threshold is changed from 40 dB to 20 dB and if unilateral impairment is included. However, there may be interactions among these variables, and the fact that common screening protocols will under-detect mild hearing impairments means that the true increase in prevalence may be greater than three-fold. At present, there are insufficient data to quantify prevalence in detail as a function of these three major variables.

Prevalence in At-Risk Groups

The U.S. Joint Committee on Infant Hearing (JCIH) has published a series of guidelines for risk indicators that predispose newborns and infants to congenital, progressive, late-onset or acquired PCHI.[3]

Prevalence estimates for PCHI in at-risk groups vary greatly. As well as the sources of variation noted earlier, risk determination itself adds further variability. Reported proportions of infants at risk vary with the risk indicator set and range from 3% to over 15%.

Accurate risk assignment requires that indicators be defined consistently and quantitatively, and that risk information be properly recorded, accessible and diligently sought. In practice, these conditions are never satisfied for all indicators. There is evidence that accurate determination of risk is time-consuming and far from straightforward. Also, geographic variations and advances over time in perinatal care quality may change both the prevalence of risk and its predictive value for impairment.

With respect to ascertainment studies, the overall prevalence of congenital impairment in the Trent study[4] was 1.12 per 1,000 live births for a 40 dB better-ear criterion; it increased from 0.54 per 1,000 in low-risk children to 3.2 per 1,000 for neonatal intensive care unit (NICU) graduates, and 7.6 per 1,000 live births for children with a family history.

In the New York State study,[2] the adjusted prevalences for mild or greater impairments in any ear were 1.2 per 1,000 and 11.2 per 1,000 live births in the well-baby nursery (WBN) and NICU groups, respectively; for bilateral impairment only, these values were more than halved, to 0.49 and 4.8/1,000. Vohr et al.[5] obtained similar values of 1.27 and 9.8/1,000, for the WBN and NICU, respectively. However, not all NICU graduates are at increased risk. The proportion may be as low as 60%, so estimates based on NICU attendance underestimate true at-risk prevalence. Also, not all WBN graduates are free from risk, so the prevalence of low-risk status may be overestimated. It is likely that these concepts of risk have been confounded in the literature, which may have contributed to bias and substantial variation among reports. It is also probable that diligent pursuit of risk information would frequently reveal substantial underestimation of the true proportion at risk.

In a high quality, prospective cohort study,[6] the prevalence of PCHI in the NICU was 1.5%. Of the 56 infants with PCHI, 30 (over half) had bilateral impairment. Other studies have yielded prevalence estimates as high as 4%, though many studies have significant methodologic limitations, especially relating to the timing and accuracy of audiologic assessment.

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Temporal Pattern of Permanent Childhood Hearing Impairment Detection in the Absence of Universal Newborn Hearing Screening

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Studies relating to the pattern of detection of permanent childhood hearing impairment (PCHI) in the absence of universal newborn hearing screening (UNHS) have been critically reviewed and are summarized here. The target disorder for the critical review is defined as a PCHI of 40 dBHL (0.5, 1 or 2 kHz) in the better ear. The review is restricted to the diagnosis of presumed congenital hearing impairment. Only literature published in 1990 or later is included, because it is felt that, before that time, technology may not have been available to accurately diagnose PCHI in infancy.

Studies on the age of diagnosis or confirmation of PCHI in the absence of UNHS fall into different categories. These include studies on the age of diagnosis in the absence of any screening activities, in the presence of some screening activities, and studies that have included some populations screened during the neonatal period as well as unscreened populations.

Studies on the age of diagnosis in the absence of any screening activities report on data collected by retrospective chart reviews and analyses of existing databases,[1-3] by parental questionnaires[4] and by a combination of methods.[5] The results indicate that, on average, the age at diagnosis of PCHI in children exceeds 12–18 months; that there is an inverse relationship between degree of hearing impairment and age of identification; and that children with risk factors, additional medical or handicapping conditions are diagnosed earlier than children without such conditions.

In some studies, the age at PCHI confirmation is reported in the presence of some behavioural screening at age 7–9 months. Such studies have been done in the United Kingdom,[6-8] Australia,[9,10] Finland[11] and Denmark.[12] In some cases, the screening at 7–9 months was replaced by a vigilance program that includes questions for parents and professionals.[6,7] Some of these studies also report some screening of high-risk neonates,[6-10] although no data are provided separately for the screened infants. Also included in this category of studies are those that have used a high-risk birth certificate registry.[13] The results of all these studies are not very different than those reported in the absence of screening. The mean or median ages at diagnosis exceed 12 months; children with risk factors are identified earlier than those without and there is an inverse relationship between degree of hearing impairment and age at diagnosis. Finally, studies in which some targeted newborn hearing screening was taking place report a lowering of the age at diagnosis for the entire birth cohort during that time. Studies that have compared different birth cohorts report a lowering of age at diagnosis over time.[12] This indicates an increased awareness of hearing impairment in children, most likely by physicians and health care professionals.

Studies comparing neonatally screened and unscreened populations include results for UNHS versus unscreened populations[14,15] and for targeted screened (high-risk and/or neonatal intensive care unit (NICU) babies) and non-screened populations.[16,17] Results of these studies clearly show that the age of diagnosis of babies with a PCHI is significantly lower for those identified through screening than for those identified through the traditional medical referral route.

The results of three major studies on the outcome of UNHS programs[18-20] are presented for comparison. These results indicate that, in the presence of UNHS, the median age of diagnosis for children with PCHI ranges from 2.1 months[18] to 3 months.[19] In addition, studies that have been collecting data over several years indicate an improvement in the ages of diagnosis over time because of experience in the development and refinement of programs.[20]

Relationship Between Severity of Hearing Impairment and Pattern of Detection

Information on the temporal pattern of detection in relation to the degree of hearing impairment severity was extracted from most of the articles already reviewed in which age at diagnosis for children in different types of health systems were presented. The results of these studies indicate that in the absence of UNHS, children with profound hearing impairments are identified sooner than children with lesser degrees of impairment, although rarely before 12 months of age. Children with moderate hearing impairments are identified between 20 and 42 months of age.[1-4,16] For children identified through UNHS, there is no significant difference in the ages at diagnosis for children with different degrees of hearing impairment.

Conclusions

- The prevalence of permanent childhood hearing impairment (PCHI) is about 1/1,000 live births in infancy if one uses 40 dBHL in the better ear as the cut-off. This rate increases to about 2/1,000 live births over the first decade of life.
- In the absence of systematic screening, the detection, confirmation, diagnosis and management of hearing impairment are significantly delayed.
- With universal newborn hearing screening (UNHS), the median age of diagnosis is less than 3 months.

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Chapter IV: Screening

Definition of the Target Disorder

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In the screening context, the "target disorder" is defined most conveniently as the set of all hearing impairments that the screening is intended to detect. Precise definition of the target disorder is fundamental to appropriate program design and evaluation.

The target disorder for a hearing screening program is most commonly quantified in audiometric terms, that is, in terms of parameters of impaired sensitivity to sound. The precise definition adopted affects many aspects of screening programs, including the prevalence and individual impact of the disorder so defined, as well as the nature, timing, intrinsic operating characteristics and actual field performance of screening tests. Generally, the more conservative the definition of the target disorder, the better the screening test performance will be. Some key dimensions of the target disorder are impairment severity, frequency range, laterality (one or both ears), and permanence, as well as the site of the disorder in the auditory system and the associated categories of impairment type (e.g., conductive, sensory, neural). Hearing disorders that are mild, frequency-specific, time-variant or located in the inner cochlear hair cells or afferent auditory pathway (e.g., auditory neuropathy) tend to give rise to higher rates of screening errors. The disorder definition must also address the child's age when the impairment is first expressed. For example, screening in the neonatal period alone will not detect progressive, late-onset or acquired disorders that express later in infancy.

The most common target disorder is congenital permanent childhood hearing impairment (PCHI). Most PCHI is of the "sensorineural" type, originating either in the inner ear (sensory) or in the nerve pathways to higher brain centres (neural), or a combination of the two. The PCHI definition may also include structural conductive impairment arising in the external ear or middle ear. There is a lack of consensus on the merit of detecting transient hearing impairment, which is usually conductive in nature, such as may arise from middle ear infection. Screening targets typically range from a lower sensitivity threshold limit of 25 dBHL (hearing level) at any of several frequencies in any ear (a liberal definition) through to a lower threshold limit of 40 dBHL in the better ear and affecting a wide range of frequencies (a conservative definition).[1]

In general, the evidence base relating specific impairment characteristics to specific outcomes in infant development is limited, and the specification of target disorders for screening programs is often based on ad hoc rationales and emerging conventions of practice. On psychoacoustical grounds, it is certain that a child with a 40dB better-ear, permanent impairment would experience major difficulty hearing conversational speech.[2] The direct evidence base in support of targeting lesser degrees of impairment is less well established. However, it must be stressed that absence of such evidence does not logically equate to absence of impact for smaller

degrees of impairment. Even a 25 dB loss of hearing sensitivity at important frequencies would be expected to confer significant limitations in perception of real world signals, on acoustical principles alone.[3]

In practice, the intrinsic operating characteristics of feasible screening tests strongly influence the target disorder criteria. The current, low limit of impairment that appears to be detectable with reasonable accuracy is typically reported to be about 30 dBHL.[2] It should be noted, however, that there are several unresolved technical issues relating to the meaning of hearing level in the context of newborn screening. The hearing level scale reference zero level is defined in relation to adult ears, and the effect of delivering a given stimulus to the ear of a newborn may differ from that in an adult, because of differences in anatomy and function of the immature ear.

The question of whether unilateral impairment should be targeted is important because, currently, there is no consensus, and the decision has a large effect on screening test performance and program resource requirements. Important considerations are the likelihood that a unilateral impairment will progress to a bilateral one, or if a child with a unilateral impairment will be seriously disadvantaged, even temporarily, by a middle ear disorder in the better ear. Even if there were no generally acknowledged, effective steps to ameliorate a unilateral impairment, such children may merit close observation and it can be argued that they should be detected by a screening program. This is an apparent departure from the World Health Organization (WHO) tenet that an effective intervention must be available,[4] but an interpretation is that the intervention in this example is to monitor for adventitious or progressive exacerbation of the target disorder. It is the *risk* of significant functional limitation that could be considered a justifiable, immediate target.

Definitive (Gold Standard) Measures of Hearing Sensitivity in Infants

Hearing screening tests must be evaluated in relation to definitive tests of hearing. There are two general approaches to "definitive" assessment: behavioural and electrophysiologic; their strengths and limitations are discussed in more detail in Chapter V.

Behavioural tests are commonly quoted to be the gold standard measure. Behavioural observation audiometry (BOA) has been discredited because of poor reliability.[1] Behavioural testing using operant conditioning (visual reinforcement audiometry (VRA)) usually becomes feasible at a developmental age of 6–9 months.

For audiometry to be definitive it must be ear specific and frequency specific, that is, it must test hearing sensitivity for specific frequencies of sound, and in each ear separately. Also, it must be able to be done by both air-conduction (AC) and bone-conduction (BC) stimuli, where clinically indicated. Testing by AC (with an earphone) measures the sensitivity of the entire auditory system including the external and middle ears, and reflects both conductive and sensorineural impairment. Testing by BC (with a transducer usually placed behind the ear) stimulates the inner ear

directly by skull vibration, bypassing the external and middle ears, and reflects only sensorineural impairment.

Above about 30 months of age in a developmentally normal child, conditioned play audiometry becomes practicable. Children between 18 and 30 months of age can be very difficult to test accurately by behavioural methods. In children with significant developmental delay, visual or motor impairment, or other co-morbidities, it may be difficult or impossible to obtain accurate hearing assessment by behavioural means at any age.

Auditory evoked potentials (AEPs) may be used to estimate perceptual hearing thresholds in infants and young children. Generally, these potentials are recorded in response to rapidly repeated sounds that are delivered by earphone or BC transducer. Electroencephalogram (EEG) recording electrodes register minute electrical responses from the neurones of the auditory pathway from the cochlea to the cerebral cortex. The AEPs are extracted from ongoing, spontaneous electrical activity of the brain and scalp musculature, using computer averaging of responses that is synchronized to many rapidly repeated stimuli.

The auditory brainstem response (ABR) is an AEP that is a widely accepted proxy gold standard measure of hearing sensitivity in newborns and infants.[5] ABR measurements can yield reasonably accurate and ear-specific, frequency-specific estimates of perceptual threshold, as well as other information about the functional status of auditory neural pathways.

Measures of Screening Test Performance

A successful screening test yields a binary (pass or refer*) outcome in each ear, for some criterion set of stimulus parameters that are linked as closely as possible to the target disorder definition. If unilateral impairment is within the target disorder, then a refer result in either ear is sufficient to refer the child; if only bilateral impairment is targeted, each ear must refer for an overall refer result. Common measures of screening test performance include test sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV) and refer rate. Sensitivity and specificity are intrinsic operating characteristics of the screening test (or multi-stage test protocol), whereas the other measures depend strongly on the prevalence of the target disorder. For a given target disorder, sensitivity and specificity vary inversely as the screening refer criterion is altered, that is, made more liberal or more conservative. Therefore, it is necessary to specify both sensitivity and specificity in order to quantify intrinsic test performance adequately. More sophisticated measures include the relative operating characteristic (ROC), which is a curve relating test sensitivity to false-positive rate, as the screening refer criterion is changed.

*The term "refer" is used to indicate that the appropriate response is diagnostic investigation and to minimize the use of the word "fail" which has a more negative connotation.

A limitation of current evidence on screening test performance is that to estimate sensitivity accurately it is necessary to determine the true hearing status of *all* infants screened, regardless of their screening result. In real world screening programs, only infants who have a refer result from screening are followed up with confirmatory testing. There are very few studies in which the complete screened cohort approach was used.

Follow-up studies restricted to screening refers cannot assess true sensitivity, because the total number of true positive cases is not determined. The number of true cases found is a lower bound on the actual number present. Published reports vary in the appropriateness with which this issue is handled. Also, reliance on extant case-finding systems to identify false-negative screens is clearly questionable in all but the most controlled environments. Such errors or omissions in the literature will tend to lead to inflated estimates of screening test sensitivity.

Loss of cases to follow-up is a source of negative bias in sensitivity estimation from the referred group. In some reports, the true number of cases is estimated by prorating observed yield at follow-up on the basis of the proportion of cases successfully followed. This is a first approximation but is simplistic in that it assumes an equal probability of follow-up attendance for true cases and non-cases, which seems unlikely.

In contrast to the situation for sensitivity, screening test specificity can be estimated accurately from large screening programs. When the prevalence of the target impairment is low, as it is in the population at large, very few babies who pass the screen will have hearing impairment, so the error in assuming the specificity to approximately equal the proportion of infants who pass the screen will be small.

When evaluating screening tests, it is crucial that the tests be known to be conducted with appropriate conditions and techniques. In this regard, it is important to note that screening test performance depends on many variables, and that performance observed in the context of a research study may differ from that seen under field operational conditions. Other factors that may bias estimates of screening test performance include change in hearing status in the interval between screening and gold standard assessment, and less than perfect accuracy of the gold standard assessment itself.

In many of the studies reviewed, screening protocols were not optimized, and this accounts for some of the variation observed. Another important source of variation is sample size. Only large, well-designed studies using appropriate screening and confirmatory test protocols should be considered to yield representative data.

Automated Auditory Brainstem Response Screening

Conventional ABR testing for definitive hearing testing typically involves manual selection of stimulus and recording parameters, and subjective interpretation of averaged EEG/ABR waveforms.[6] Automated auditory brainstem response (AABR) screening is an adaptation that usually involves a single stimulus condition, with

computer-controlled stimulation and recording and computer-based response detection. The usual stimulus is a click at a sound pressure level that corresponds to about 35 dBnHL (normal hearing level), namely, 35 dB above the average subjective perceptual threshold for the click in a group of young adults with normal hearing. A successful test will yield a binary outcome, specifically a pass or refer result, for each ear.

AABR screening is fully automated, objective and non-invasive. It can be performed on any neonate or very young infant who is asleep or at least at quiet rest, in a moderately quiet test environment. Sensors are attached to the scalp and the stimuli delivered by insert earphone or supra-aural earphone. The test typically takes about 10 minutes per baby. The automation of the measurement and interpretation radically reduces the knowledge and skills required in the screening personnel, and renders the procedure much less costly and generally more accurate. Some skill is required of the tester, especially in choosing an appropriate behavioural state of the child at test, the placement of the recording electrodes, and the application of the earphone.

The studies that address the accuracy of screening ABR tests describe results for click ABR screening relative to subsequent definitive audiometry and are of fair to good quality.[7] There are significant sensitivity limitations of the click ABR, because the click is a broadband stimulus that stimulates a wide range of frequencies. It appears that frequency-specific screening AABR tests are not currently available.

The accuracy of click AABR depends on the definition of the target disorder. It cannot detect hearing impairment at low frequencies or at isolated, specific frequencies, but it has reasonable performance for hearing impairments that are defined in terms of average impairment across the speech frequencies (between 500 and 4,000 Hz). Typical reported sensitivities are from 80–90%, with false-positive rates (FPRs) from 5–10%. It is likely that this is an underestimate of true performance, because factors such as transient impairment at the time of screening or emergent impairment between screening and confirmatory testing degrade apparent test accuracy. Failure to detect a hearing impairment that is not present at the time of screening is not a screening test error, nor is a refer result in the presence of a transient disorder that resolves prior to definitive assessment. The false negative screening error rates of the statistical algorithms for AABR response detection are typically set in the range 0.01–0.001. Discrepancies between these parameters and the higher apparent rates of missed cases in follow-up studies may reflect emergent impairment, frequency-specific impairment and confirmatory test error.

Automated Otoacoustic Emission Screening

Otoacoustic emissions (OAEs) are very faint sounds that arise from the inner ear and may be recorded by a miniature microphone in the external auditory canal.[8] Evoked OAEs arise in response to controlled acoustical stimulation. There are two types of evoked OAE: transient- and distortion product-evoked OAE; these are denoted as TEOAE and DPOAE, respectively. TEOAE measurement involves delivery of a rapid series of click stimuli, with computer averaging and frequency analysis in order

to extract the minute OAE signal from concurrent environmental sound or body-generated sounds. DPOAE measurement involves delivery of two simultaneous, continuous tones that have a specific relationship in intensity and frequency. In response, the inner ear generates a third tone that is related in frequency to the two stimulus tones. It arises because of non-linear distortion in the patterns of excitation within the cochlea, hence the name DPOAE.

Like the AABR, an automated otoacoustic emission (AOAE) screening test is fully automated, objective and non-invasive. It can be done on any neonate or infant who is asleep or at least at quiet rest, and it requires a quiet acoustical environment. A transducer is placed in the ear, and both delivers the stimuli and records the OAE for immediate computer processing. A binary (pass/refer) result is obtained for each ear. The test typically takes less than five minutes per baby.

For an OAE screen to be accurate, the earphone or earphone/microphone assembly must be placed appropriately in the external ear canal, and the canal must be free of vernix or debris. Also, OAEs are especially vulnerable to the presence of middle ear fluid and conductive hearing impairment, partly because the middle ear must first conduct the stimulus to the cochlea and then conduct the emission back to the external ear. It is probably for this reason that AOAE screening in the first few hours after birth is generally reported to yield elevated rates of test failure or false-positive refer results. The AABR, in contrast, appears to be less affected by most minor conditions of the middle ear or external ear.

The most definitive, experimental study to date involved screening of about 5,000 neonatal intensive care unit (NICU) graduates and well babies, using ABR, TEOAE and DPOAE screens with optimized test parameters and objective, statistical screening failure criteria.[8] Screening results were compared with high quality, confirmatory VRA at 8–12 months corrected age in the complete cohort.[9] For all optimized screens, with a 10% FPR the sensitivity was typically 80–90%. Because of the possibility of intercurrent or progressive impairment in longitudinal validation studies, these values should be considered as lower bounds on true sensitivity. ABR was found to be the most accurate predictor of hearing impairment at lower frequencies (1 kHz), probably because OAE is especially vulnerable to ambient noise at lower frequencies. At the higher frequencies more commonly associated with PCHI (2–4 kHz), all screening tests performed similarly, with less than a 6.4% refer rate. Multi-stage testing with DPOAE followed by ABR resulted in lower refer rates than TEOAE followed by ABR, the best rate being 2%.

The most definitive screening program report to date addressed 69,761 newborns screened using a two-stage screening prior to hospital discharge (usually OAE and AABR), and an outpatient AABR re-screen after four to six weeks.[10] The program performance improved over three years, the overall refer rate improving from 5.9–2.6% and the pre-discharge FPR achieving less than 3% (specificity 97%). There is increasing evidence that multi-stage screening that includes AABR in at least one stage can achieve overall referral rates as low as 1–2%, which sets an upper bound for the true FPR.[11]

A final issue to be considered is auditory neuropathy (AN), a condition that may affect up to 10% of all infants with PCHI.[12] Neuropathy is usually characterized by sensorineural hearing impairment (SNHI) of any degree, usually normal OAEs, normal cochlear microphonic potentials (CM), absent acoustic reflexes and an absent or grossly abnormal ABR. OAE screening will miss such babies, but AABR screening will detect them. Because the majority of babies with AN are likely to have attended an NICU, the use of AABR for screening in all NICU graduates is necessary in order to identify most babies with AN.

Potential Harms Associated with Screening Outcomes

False-positive screens increase the burden on follow-up diagnostic services, and may increase family anxiety and stress. These concerns figure prominently in reviews of universal newborn hearing screening (UNHS) and have led to a maximum FPR of 3% being proposed as a benchmark for UNHS programs.[13] It is clear that high quality screening protocols can achieve overall FPRs of less than 2%. For hospital-based programs, low FPRs at hospital discharge usually require series screening protocols that involve at least two pre-discharge screening tests, the second screen being conditional on a refer outcome for the first screen. The AABR is especially effective at least as the second screen. The lowest overall rates of referral for diagnostic, audiologic assessment may be achieved by including a third screen with AABR, shortly after hospital discharge. These compound protocols reduce the immediate family burden of false positives, as well as the resource impact on both the family and the program, with respect to attendance for audiologic follow-up.

When examining parental responses to universal programs, it was found that negative attitudes were rare and positive attitudes common.[14] The limited body of evidence available does not support the belief that parents generally suffer anxiety or stress due to early screening or to false-positive tests. A substantial majority of parents endorse early identification of hearing impairment. Any concerns are more closely related to timeliness and quality of professional interactions. The rate of significant family anxiety in screening referrals does not appear to differ from the population base pattern of anxiety scores on standard, psychometrically validated measures.

Screening Coverage and Follow-Up Compliance

The overall effectiveness of a screening program depends not only on the performance of the screening tests themselves but on the extent to which subjects are successfully accessed for screening and are successfully followed up after a screening refer result. The overall sensitivity of a program, for example, is the product of the screening coverage, the screening protocol sensitivity, and the follow-up rate. It is commonly reported that there is increased follow-up of screening failures and increased coverage over the first two to three years of screening program implementation.[15] Unpredictability of discharge or transfer has been cited as a reason for higher miss rates in NICU groups. Reports generally indicate that incomplete follow-up coverage is a major area of concern, with reported follow-up rates rarely exceeding 80%.

Surveillance and Referral Components

The proportion of PCHI expressed in infancy but not present congenitally is poorly understood. Estimates lie in the 5–15% range but may be underestimates, due to over-attribution of congenital expression in the absence of comprehensive early detection programs. There is evidence that certain risk indicators, including cytomegalovirus (CMV) infection, persistent pulmonary hypertension of the newborn (PPHN) and several syndromes, are strongly associated with progressive or late-onset impairment. These impairments cannot be detected by neonatal screening and require a program component that includes tracking and screening at a later date, such as at 1 year of age and beyond. It should be noted that re-screening infants at risk (including NICU graduates) with OAE will miss hearing impairment associated with AN. AABR re-screening will not miss neuropathies but has the disadvantage that it will often require use of sedation, in infants older than about 6 months. Also, because not all such cases will be determined to be at risk perinatally, it is probable that education of both families and professionals (e.g., primary care physicians) about early warning signs of hearing impairment will be necessary to maximize overall detection performance. However, to date there appear to have been no systematic studies of the effectiveness of such efforts.

Finally, a comprehensive system for early identification would include a program component that ensures prompt referral for screening and/or audiologic assessment of infants with postnatal risk factors that are associated with acquired hearing impairment, such as meningitis.

Key Differences Between Universal Screening and High-Risk (Targeted) Screening

The target population for screening is usually defined as either all children (universal screening, UNHS) or children at risk for hearing impairment (targeted screening). Typically, the proportion of the general newborn population considered to be at risk for hearing impairment is reported to be in the 8–15% range.[1,16] It follows that it will usually require substantially fewer resources to screen only the high-risk group, relative to those required for universal screening.

Populations at risk for hearing impairment differ from the general population most obviously in the prevalence of the target disorder. The prevalence of hearing impairment in typical high-risk groups, as defined currently, is typically about eight to ten times greater than in babies currently considered to be not at low risk. Relative to universal screening, this increase in base prevalence substantially increases the positive predictive value (PPV) of a screening refer result (non-pass), and substantially reduces the number needed to screen (NNS) in order to identify an additional case.

The most obvious limitation of targeted screening in high-risk newborns and infants is that a substantial proportion of infants who actually have the target impairment will not be screened. It is reported that between 50–75% of young children with

significant PCHI manifest a risk indicator, with the most common estimates being closer to 50%.[15,17] This means that even with a perfect screening test, the overall sensitivity of high-risk screening could be no better than 75%, and would probably be closer to 50%. Therefore, relative to targeted screening, the incremental yield of UNHS is the 25–50% of all infants who have hearing impairment and who would not be screened in a targeted program.

Another significant limitation of targeted screening is the difficulty of accurately identifying the sub-population genuinely at risk. Comprehensive and accurate risk identification is a difficult and time-consuming task. Many important risk indicators are not routinely determined or accurately documented. These include perinatal infections such as asymptomatic CMV, as well as manifestations of a variety of syndromes known to be associated with PCHI, including craniofacial anatomical anomalies. Global, proxy risk indicators, such as NICU attendance for over 48 hours, are simple to implement but are very inaccurate indicators of genuine risk. Familial childhood hearing impairment is an important indicator but is very difficult to determine accurately.

In a targeted program, the risk assessment can be viewed conceptually as a documentation-based screening test with very poor sensitivity and specificity, that is, in series with one or more of the physiologic screening tests. The sensitivity and specificity of a compound, series screening protocol are no better than the poorest performance parameter for each component screen. Furthermore, it should be noted that to actually perform a physiologic screening test in a given child may be more reliable and substantially less resource consumptive than to carry out a comprehensive risk determination, so it would seem potentially more accurate and efficient to screen all newborns. However, if risk indicator information were considered absolutely necessary for other activities, such as to define a subpopulation for targeted surveillance, then the risk assessment is required for all infants, and the efficiency argument is weakened. Alternatively, if a program were to incorporate routine surveillance of all infants, then the relevance of specific risk indicators is reduced and the relative accuracy and efficiency of universal, physiologic screening again become a dominant consideration.

Because only about 10% of newborns will manifest a risk indicator detectable with current methods in widespread use,[18,19] UNHS typically involves screening about 10 times as many babies as in targeted high-risk programs, but will increase the yield of true cases by a factor of 33–100%. The actual increase in yield will depend on many variables that reflect individual programs, among them the relative proportions of progressive and early-onset hearing impairment in the at-risk and no-risk groups. These proportions are currently unknown.

It is important to note that a large number of variables affect the optimal design of a screening program. While there are certain, generalizable principles that appear to affect the likelihood of good program performance, it is unlikely that any one

particular program design will be optimal under all circumstances. Good screening program design must take proper account of local contextual and infrastructural factors, resources and constraints.

Conclusions

- Hearing screening is currently based on automated otoacoustic emission (AOAE) testing and automated auditory brainstem response (AABR) testing.
- The sensitivity of AABR is 85% and the specificity is 90–95%; for AOAE testing, the sensitivities are only slightly less.
- Current screening protocols frequently involve series or parallel combinations of tests that achieve overall referral rates of <2%.
- Loss to follow-up is the largest single factor limiting effective sensitivity and preventing delivery of hearing and communication development options. Future studies should include determination of predictive models of parental participation, such as optimal communication of screening results to families and psychosocial causes of non-participation.

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Chapter V: Assessment

Audiologic Assessment

Context

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Audiologic assessment here refers to detailed determination of the hearing status of an infant. Three routes to audiologic assessment include universal newborn hearing screening (UNHS), surveillance of children at risk for permanent childhood hearing impairment (PCHI), and risk-based referral. Respectively, these cover neonates who express congenital PCHI, infants at risk for late-onset or progressive impairment, and infants at risk through postnatal risk discovery or new risk events. Most audiologic assessment candidates arise through UNHS, and are accessible under 3 months corrected age (see Prieve et al.[1]). Surveillance and postnatal risk routes may yield candidates at any time throughout infancy (age 0–24 months). For a comprehensive overview, see Thompson et al.[2]

Audiologic assessment must be as accurate as possible.[3] Errors have occurred in the past, with significant consequences for affected children and families. The importance of high quality test protocols applied consistently and program wide, adequate tester training and skills maintenance, as well as rigorous quality management, cannot be overstated. Definitive quantification of hearing may require several test sessions, either to improve audiometric accuracy or completeness, or to monitor possible changes in hearing. The later audiologic assessments may occur after provision of amplification or other communication enhancement strategies.

Objectives

The purpose of the audiologic assessment is to provide the audiometric information that is necessary and sufficient to fully inform the subsequent course of events, which may include medical diagnosis and treatment, prognosis, provision of assistive technology such as hearing aids, and/or communication development options. There are two kinds of measurement in common use: (i) **estimates** of perceptual threshold for pure tones at specific frequencies; and (ii) **measures** of function of specific parts of the auditory system. The latter measurements can inform about the **site** of dysfunction (e.g., middle ear, cochlear, retrocochlear) and the related **type** of dysfunction (e.g., conductive, sensory, neural, mixed). Also, the relationships among the measurements can help to validate the overall inferences about hearing status.

Age at Assessment

The chronological and developmental age of the infant affects the choice of tests. For infants under about 9 months developmental age, behavioural observation audiometry (BOA) is often recommended as the assessment tool of choice. However, there is clear evidence that BOA is not a reliable procedure.[4] It is widely recommended that

audiologic assessment in such infants must be based on physiologic measures.[5] These include: threshold estimation by evoked potentials (EPs); acoustic immittance and middle ear muscle reflexes; transient-evoked or distortion product-evoked otoacoustic emissions (TEOAE or DPOAE); and testing the functional integrity of the afferent cochleoneural pathways by cochlear microphonic potentials (CM) and also by the auditory brainstem response (ABR). A limitation of electrophysiologic audiometry is that EPs are epiphenomena of hearing and so their relationship to perception is not causal, but correlational. Thus, EP thresholds are proxy statistical estimators of actual perceptual thresholds.

At 6–9 months' developmental age, most infants can give reliable hearing thresholds using operant-conditioned head-turn responses — visual reinforcement audiometry (VRA). Infants with multiple disorders or severe cognitive disorders may not be testable behaviourally, and it may be necessary to resort to physiologic methods of estimating perceptual thresholds.

Electrophysiologic Threshold Estimation

Hearing threshold estimates are required at a set of frequencies, typically in the range 0.5 through 4 kHz. This range includes frequencies that are important for speech perception as well as for detailed specification of hearing aids, and which are conventionally required for medical evaluation. Of the many EPs that can be elicited from the cochlea to the cerebral cortex, only the frequency-specific (FS) ABR and auditory steady state responses (ASSR) are appropriate candidates for threshold estimation.[6]

Auditory Brainstem Response

ABR thresholds can be measured for click and tonepip stimuli. The click excites a broad cochlear region, so click ABRs cannot provide accurate, frequency-specific audiometry.[6-8] In contrast, there is evidence that tonepip ABRs can yield clinically acceptable estimates of puretone thresholds by air conduction (AC), but only given appropriate test protocols and tester skills.[9] The key elements of the test protocol are that the infant must be in natural or sedated sleep, recording electrodes must be properly positioned and have low and approximately equal contact impedances. In addition, recording bandwidth must be from about 30–1,500 Hz, the data window must be at least 25 milliseconds (ms) in length, the gain and artifact rejection limits must be set appropriately, and averages must be of at least 2,000 accepted sweeps with replication.[6] Because test time is limited, the stimulation rate must be at least 30 per second and the selection strategies for stimulus frequency, intensity and route must be very efficient. It is necessary to restrict testing to only those frequencies and routes of stimulation that are required for clinical decision making. Furthermore, in order to resolve the conductive and sensorineural components of hearing impairment, testing by bone conduction (BC) may be necessary.[10-13] Data on the accuracy of BC FS-ABR threshold estimates are limited, as is the dynamic range of BC stimulation.

Errors in FS-ABR threshold estimation are reported to arise most commonly from misjudgment of response presence or absence.[6] Currently, reliable and well-validated objective statistical response detection algorithms are not widely available clinically. Thus, it is essential that testers be properly trained in test conduct and response recognition, that caseloads are sufficient to maintain skills, and that ABR results be assessed critically in the light of all other sources of evidence.

ABR thresholds are *not* equal to perceptual thresholds, and it is necessary to adjust for bias when estimating true hearing levels. FS-ABR thresholds are biased positively by about 0–20 dB, the exact amount depending on the stimulus frequency and possibly on stimulus level.[6,9] Due to this bias and the intensity limitations of the transducer, it is not possible to resolve hearing impairments greater than about 80 dBHL by tonepip ABR techniques.

Careful attention to stimulus calibration is essential. Levels may be expressed in dB peak equivalent sound pressure level (SPL) in the individual infant ear, in a standard coupler that simulates an average adult ear, or in dBnHL (normal hearing level). Neither dBnHL nor dBHL (hearing level, as defined by the American National Standards Institute (ANSI) in American National Standard S3.6-1996) is directly applicable to thresholds in young infants. An unresolved question is whether it is necessary to apply SPL adjustments reflecting the acoustical properties of the average infant ear, or to take real ear SPL measurements, in order to express infant hearing thresholds appropriately (see, for example, Sininger et al.[14]).

In the vast majority of infants under about 6 months of age, successful FS-ABR can be accomplished during natural sleep. However, it can be difficult to test infants over 4 months in natural sleep, and those over 6 months will most likely require sedation or light, general anesthesia. While electroencephalogram (EEG) conditions in sedated sleep are usually very good, test efficiency remains an important consideration, because the duration of sedated sleep can be unpredictable and limited.

Click Auditory Brainstem Response and Auditory Neuropathy

The click ABR has limited clinical value for diagnostic audiometric assessment. For example, the latency of wave I or wave V can imply a significant conductive impairment component, and this is informative if BC threshold measurements prove impractical in a given infant. Also, if EEG conditions were marginal, the click ABR might be detected when tonepip ABRs were not, because of the superior neuronal synchrony induced by a click. However, these applications are relevant only if sedated testing for FS-ABR is deemed to be unfeasible.

The click ABR is useful to detect cochleoneural dysfunction beyond the level of the cochlear outer hair cells. The most common cause is auditory neuropathy (AN), which is a set of cochleoneural transduction disorders that have in common reduced temporal synchrony of afferent neuronal activity.[15] This leads to absence or gross abnormality of the ABR, and its predictive accuracy for hearing threshold is lost.

Most infants who have AN are graduates of neonatal intensive care units (NICUs).[16] AN usually yields absent ABRs, present otoacoustic emissions (OAEs), absent acoustic middle ear muscle reflexes, and any degree of hearing impairment from mild to profound.[17,18] OAEs may be absent or may decline over time, but cochlear microphonic potentials (CM) are present. Accordingly, an appropriate sub-protocol for CM measurement is essential. When AN is detected, ABR thresholds are usually not measurable and in any event are not reliable, whereas behavioural thresholds, such as by VRA, are more accurate. AN typically imposes an auditory perceptual deficit in the temporal resolution of complex signals such as speech. It is reported that about 50% of infants with AN receive significant benefit from hearing aids[19] but, currently, the prediction of such benefit is difficult. A substantial proportion of infants with AN appear to do well with cochlear implants.

Emerging Technology: Auditory Steady State Response

The auditory steady state response (ASSR) is an evoked potential that is recorded from the scalp and, like the ABR, can be used to estimate auditory thresholds.[20] It is frequency specific and can be elicited at a single frequency or at multiple frequencies simultaneously.[21] The ASSR has been measured in newborns, children and adults[22,23] when asleep or awake.[24,25] The stimulus is a tone that is amplitude- and/or frequency-modulated, evoking periodic scalp potentials at the modulation frequency. The stimulus activates a place on the basilar membrane determined by the carrier frequencies, typically 0.5, 1, 2, and 4 kHz.[26]

The ASSR is usually displayed as amplitude and phase spectra of the response and background noise. Presence or absence of response is determined statistically, which makes the ASSR an objective test.[22,27] The multi-frequency stimulus/multi-ear stimulation technique has the potential to shorten the time needed to determine auditory thresholds,[28] relative to FS-ABR, which would be a major advantage given the time pressures in infant testing. However, this advantage would be gained only if no substantial interaction occurs between the responses for multiple frequencies and ears. Further analysis of the techniques and large-scale clinical trials are required to quantify the usefulness of this technique.

Otoacoustic Emissions

Otoacoustic emissions (OAEs) are sounds produced by the cochlea and recordable in the ear canal.[29] Since the cochlea is developed by 34 weeks' gestation, OAEs can be measured in newborns.[30] The two types of OAEs in common clinical use are transient-evoked OAE (TEOAE) and distortion product-evoked OAE (DPOAE).[29] TEOAEs are normally elicited by clicks and comprise a complex waveform lasting up to about 15 ms and reflecting progressive activation of the cochlear partition from base (high frequency) to apex (low frequency). Response components associated with various frequency regions of the cochlea are extracted by spectral analysis. DPOAEs are elicited by simultaneously stimulating the cochlea with two tones

(at frequencies f1 and f2) making the response frequency specific. The result is an emission at 2f1–f2 caused by intermodulation distortion occurring on the basilar membrane of the cochlea.

OAEs are useful for screening, in part because they are an objective and frequency-specific measure.[29] However, they are limited as a tool for detailed audiologic assessment, because they relate more to cochlear function than to audiometric threshold. OAE measures cannot predict an individual's puretone threshold, so they are considered as a test of cochlear function from which inferences can be drawn about hearing status.[31] OAEs are typically not present in ears with significant negative middle ear pressure, middle ear effusion or permanent, conductive disorders.[32,33] In ears with sensorineural impairment, OAEs may or may not be present depending upon the frequency and severity of the impairment. An absent OAE suggests a severity of at least 30–35 dBHL in the frequency region of the OAE, but only if the impairment is of cochlear origin.[34,35] Regardless of the true size of the cochlear impairment component, even a slight conductive impairment could abolish the OAE, and there is no direct clue from the OAE itself about the impairment type. However, OAEs are a useful contributor to the audiometric assessment process by providing a limited cross-check of the EP or VRA results.

Middle Ear Analysis

Middle ear analysis (MEA) typically includes tympanometry and acoustic middle ear muscle reflex (MEMR) measurement (see ASHA[36] for review). Basic tympanometry yields acoustic immittance versus static pressure, and the parameters of the relation reflect the middle ear function. The most common inferences relate to middle ear pressure and to fluid in the middle ear, associated with otitis media (OM). These findings increase the likelihood of conductive hearing impairment (CHI), and thereby provide a weak inferential cross-check on a conductive component measured by ABR or VRA. It should be noted that reflexes are absent even with a slight conductive hearing loss.

The differential diagnostic value of MEA lies in four inferences. First, finding a significant conductive impairment component by ABR implies middle ear abnormality, so normal MEA would suggest that the ABR results be examined critically (especially BC ABR results). Second, BC stimulus levels above about 50 dBnHL are not achievable, so air-bone gaps cannot be measured accurately when AC thresholds are above 50 dB. Abnormal MEA implies a possible conductive component. Third, abnormal MEA is often associated with reduced or absent OAE, so when MEA is abnormal, absence of OAE has little diagnostic value in assessment of possible AN. Fourth, presence of an acoustic reflex (AR) is normally associated with AC thresholds not greater than 10 dB below the AR threshold, so an inference of severe hearing impairment or greater by ABR must be examined very critically if an AR is observed. Conversely, absence of the AR is not informative about thresholds if the tympanometry is markedly abnormal.

To optimize these cross-checks, MEA probe frequency should be much higher than the conventional 226 Hz used in adult testing.[37] There is fair evidence that in infants under about 7 months of age, a normal 226 Hz immittance curve can be obtained despite the presence of middle ear fluid.[38] A probe tone frequency of 678 Hz or greater reduces these false-negative findings.[39,40] Similarly, ARs are frequently absent in young infants with 226 Hz probes, but not with higher frequency probes.[41] Thus, use of high-frequency probes is likely to improve the diagnostic value of MEA in young infants.

Visual Reinforcement Audiometry

Visual reinforcement audiometry (VRA) procedures in common use are less than optimal because they involve sound field stimuli and/or speech stimuli which has limited value for precise threshold estimation. These VRA methods, therefore, do not measure ear-specific and frequency-specific thresholds accurately. However, Widen et al.[42] describe VRA procedures that make obtaining these measurements possible at 8–12 months in over 90% of candidates, given skilled testers and a high quality protocol. Key elements of a high quality protocol are: stimulation by insert earphones at key frequencies; use of frequent, valid control trials; and careful documentation of the sequence of stimulus, control and response events. Infrastructural aspects, such as the use of two properly trained testers, appropriate test environment, and optimal conditioning, reinforcement and distraction methods, are also important.

Minimum response levels (MRLs) obtained by VRA are likely to be biased positively, relative to true perceptual thresholds.[43] The bias will be small if and only if the infant has adequate cognitive, visual and motor function and is well conditioned, the assessment of which is partly subjective but can be made more reliable and verifiable by careful response documentation. In infants for whom adequate operant conditioning cannot be established, one may always resort to ABR methods, bearing in mind an increasingly likely need for sedation in infants over about 6 months of age.

Audiologic assessment based on VRA should include OAE and MEA testing wherever feasible. If cognitive development and responsiveness are deemed sufficient for accurate VRA, then normal OAEs and VRA threshold elevation beyond about 45 dBHL are clearly discrepant, in which case AN is a possibility and ABR testing is indicated, usually under sedation. As was the case for ABR threshold inferences, abnormal tympanometry precludes inferences from OAE and from AR absence. Given normal tympanometry, AR presence should prompt careful review of the significance of a finding of severe or profound hearing impairment by VRA.

Auditory Brainstem Response / Visual Reinforcement Audiometry Relationships

In the present, programmatic context, VRA will be used most frequently in infants with prior ABR audiometry, and often after fitting of hearing aids. If the VRA and ABR thresholds differ significantly, questions arise about hearing aid adjustment as well as diagnostic and prognostic significance of the possible "changes." Available

data are limited, but discrepancy clearly demands careful review of the quality of all audiometric findings. Consistent and reliable VRA thresholds well below ABR estimates should lead to revised management based on the VRA, but the validity of the VRA reliability judgment is crucial. When VRA thresholds are higher than ABR thresholds, the latter must be carefully reviewed for false-positive response identification, and re-testing under sedation may be indicated. The potential for progressive impairment requires careful and sustained audiometric monitoring.

Finally, it is known that the age period between about 18 and 30 months can be problematic for reliable audiometry. The child's attention may not be sustainable for operant conditioning, yet adequate cooperation for play audiometry may not yet be readily achieved. Frequency-specific ABR testing under sedation remains a viable option for the determination of hearing thresholds across all age groups, if neuropathy is not suspected.

Conclusions

- Complete audiologic assessment, which can be achieved in healthy children under 6 months of age, is essential to appropriate hearing aid fitting and to family decisions about communication development options.
- Further studies are needed to determine the true prevalence of congenital, late-onset, progressive and acquired impairment, and to clarify its relationship to risk indicators.
- Existing literature is unclear with respect to the management of auditory neuropathy (AN) and the value of auditory steady state response (ASSR) testing.

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Medical Evaluation of a Child with Bilateral Sensorineural Hearing Impairment

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Universal newborn hearing screening (UNHS) programs target earliest identification of sensorineural hearing impairment (SNHI) and facilitate entry into an early hearing and communication development (EHCD) program. As such, clinicians will encounter children with SNHI at a younger age. Medical evaluation of a newborn with hearing impairment should be initiated prior to 3 months of age and intervention initiated prior to 6 months of age. Establishing the etiology for SNHI is important to allow patients at risk for SNHI to be identified early, to allow physicians to intervene against the causative factor (i.e., stop potentially ototoxic drug therapy), and to provide the patient and family with prognostic information.

A detailed history, physical examination and audiological evaluation are the most important steps in diagnosing the etiology of SNHI.[1] Many syndromes and infectious etiologies can be diagnosed by these methods. This critical review provides a discussion of the etiology of childhood bilateral SNHI (>40 dBHL) and a review of processes for diagnostic evaluation.

Etiology of Bilateral Sensorineural Hearing Impairment

Advances in genetic testing and aggressive management of perinatal infections have altered the frequency of diagnoses of etiologies of SNHI.[2-8] Recent studies suggest that autosomal recessive genes are responsible for most cases of unknown etiology (i.e., connexin 26 (cx26), see below). The etiologic categories and their prevalence are:

1. Unknown (37.7%)
2. Genetic — nonsyndromic (29.2%) and syndromic (3.2% — e.g., Waardenburg syndrome)
3. Nongenetic — prenatal (12% — e.g., rubella, cytomegalovirus), perinatal (9.6% — e.g., prematurity, asphyxia, kernicterus), postnatal (8.2% — e.g., meningitis). Auditory neuropathy (AN) is presumed to be a nongenetic postnatal cause. Its frequency and etiology requires further study.

Presentation of Common Etiologies

The presentation of the more common etiologies of bilateral SNHI and the results of investigations are discussed in this section.

Unknown

With careful investigation, a presumptive cause can be determined in some children with bilateral SNHI previously labeled as "unknown."[8,9] Genetic nonsyndromic is often the presumed cause following the obtaining of further details regarding family history.

A recent study[10] retrospectively tested stored neonatal blood of children diagnosed with SNHI, and identified five children (12% of children with SNHI) believed to have SNHI secondary to intrauterine cytomegalovirus (CMV) infection (four unilateral and one bilateral hearing impairment). The role of prenatal infections is an area that requires further investigation.

Genetic Nonsyndromic

Most of the nonsyndromic recessive gene mutations produce congenital profound deafness, although there is variation. Genetic nonsyndromic hearing impairment is highly heterogeneous. At the time of this review, 30 recessive genes have been localized with 7 of the genes identified; 39 dominant genes have been localized with 11 identified; and 7 X-linked genes have been localized with one identified.[11]

The most common recessive nonsyndromic mutation is in the beta-2 gene on chromosome 13 that produces the protein connexin 26 (cx26). Mutations in the cx26 gene produce hearing impairments with considerable range in severity, from the mild-moderate range to profound. Cx26 forms gap junctions between cells and is thought to help recirculation of ions in the cochlear endolymph.[12] Mutations in this gene account for half of all nonsyndromic recessive deafness, meaning that they cause 30–50% of genetic nonsyndromic deafness. The carrier frequency in European populations is about one in forty.[13]

Genetic Syndromic

An example within this classification is Waardenburg syndrome, an autosomal dominant syndrome with variable penetrance.[14] Clinical features include lateral displacement of the medial canthi of the eyelids, high nasal root, hypertrichosis, confluent eyebrows, pigmentary disorders and SNHI. Hearing impairment, unilateral or bilateral, occurs in 30–50% of patients with Waardenburg syndrome.[15] The possibility that such features may appear fortuitously in some family members can make it difficult to ascertain whether the syndrome is present. There are several classifications for Waardenburg syndrome, defined by physical characteristics. Individuals with dystopia canthorum have Type 1. SNHI occurs in about 20% of individuals with Type 1. Patients without dystopia canthorum (Type 2) have a 50% prevalence of SNHI and the hearing impairment is more likely to be progressive.[16] The SNHI may be unilateral or bilateral.[15,17] A radiological abnormality of the inner ear was detected in 17% of patients.[18] Genetic testing is possible for Waardenburg syndrome (i.e., PAX3, EDNRB, EDN3; Smith et al.[19]) but not widely available. The sensitivity and specificity of genetic testing for this condition is not well described in the literature.

Nongenetic

Prematurity

Prematurity is defined as birth before completion of the 37th week of gestation. Although several risk factors for SNHI, such as perinatal hypoxia, sepsis and kernicterus, may be present in premature neonates, prematurity constitutes a unique reliable risk indicator. Premature infants are 20 times more likely to be severely hearing impaired than infants of normal weight and gestational age.[20] The mechanism for this is not well understood, but is believed to involve recurrent apnea attacks. The natural history of the SNHI associated with prematurity has not been well described. If the potential for prematurity cannot be established prior to birth, based on risk factors or ultrasound dating, there are characteristic neonatal signs and symptoms which include: weak cry, hypotonia, abnormal posturing and poor feeding.[21-23] The reliability of these clinical measures is not well described.

Asphyxia

There is evidence that birth asphyxia can result in damage to the central auditory pathways and to the cochlea. Autopsy studies have shown ischemic lesions of the cortical gray matter, basal ganglia and brainstem in infants with perinatal asphyxia.[24] The severity of pathologic findings correlates with the duration of hypoxia.[25] The pathophysiology would indicate that the hearing impairment is present at birth. However, the natural history of the SNHI is not well described in the literature.

Meningitis

SNHI secondary to meningitis is due to cochlear damage with a reduction in neurons in the spiral ganglia and destruction of outer and inner hair cells.[26] Most patients with meningitis sustain permanent, bilateral, severe-profound SNHI, but in a series of 64 cases of meningococcal meningitis, 38% had bilateral asymmetric SNHI and 11% exhibited a unilateral SNHI.[27] The reported incidence of SNHI after meningitis has varied from 3–40%, with most reports clustered in the 15–20% range.[28] Eighty-nine percent of those who suffered post-meningitic hearing impairment contracted meningitis before the age of 3 years.[29] Post-meningitic hearing impairment has been described as occurring as late as six months after an episode of meningitis, although patients who exhibit a normal auditory brainstem response (ABR) after the first few days of hospitalization and antibiotic therapy are unlikely to develop SNHI.[30]

Kernicterus

Hyperbilirubinemia is defined as a serum bilirubin greater than 1.5mg/100ml.[31] Hyperbilirubinemia during the first week of life is most often due to overproduction of bilirubin through hemolysis and defective conjugation. Kernicterus defines a syndrome of neurologic sequelae secondary to bilirubin crossing the blood-brain barrier. It is often seen at values greater than 1.8–2.0mg/100ml. Early symptoms and signs include extreme jaundice, absent Moro (startle) reflex, poor suck and lethargy. Late features include high-pitched cry, arched back with neck hyperextension (opisthotonos),

bulging fontanel and seizures.[32] The association of kernicterus with SNHI is well documented and believed to be secondary to deposition of bilirubin in the cochlear nuclei and basal ganglia.[33] It is also associated with auditory neuropathy (see below).

Rubella

Defects attributed to congenital rubella infection include SNHI, cataracts, microphthalmia, buphthalmos, ventricular septal defect, pulmonary stenosis, microcephaly, cerebral palsy, mental retardation, thrombocytopenic purpura, rash, hepatomegaly, splenomegaly and osteopathy.[34] Not all associated defects present concurrently. In fact, 40% of children included in the National Congenital Rubella Surveillance Program (NCRSP) had a single organ defect,[35] and hearing impairment is present as an isolated defect in 22% of children.[36] The greatest risk occurs with maternal infection during the first trimester.[37] This risk has been estimated in various prospective studies to be between 60% and 90%.[38,39]

SNHI is the most common defect secondary to intrauterine exposure to rubella. It is usually bilateral but can be unilateral. Reported damage to the auditory system has included degenerative and inflammatory changes affecting the organ of Corti, stria vascularis, Reisener's membrane and the tectorial membrane.[40]

In some children with SNHI secondary to rubella, it may be possible to elicit a maternal history of contact with rubella or of a rash during pregnancy. However, 24% of mothers of children registered with the NCRSP were unable to give any history of contact, rash or illness during pregnancy.[35] In a proportion of children with no evidence of damage other than a hearing impairment, there may be signs of rubella retinopathy to indicate that a congenital infection has taken place. The retinopathy is the result of alternate areas of hyperpigmentation and hypopigmentation, and the appearance is described as a "salt and pepper" effect. It does not usually affect visual acuity. Fifty percent of patients with congenital rubella display the typical retinopathy.[41] Sera obtained in pregnancy which show seroconversion or a significant rise in antibody titre, and detection of specific IgM antibody provide evidence of definite infection.[42] Demonstration of rubella-specific IgM in cord serum is diagnostic of a congenital infection, as immunoglobulin of the IgM class does not cross the placental barrier.

With routine immunization programs, rubella as a cause for SNHI has practically been eradicated in developed countries.

Auditory Neuropathy

The syndrome of auditory neuropathy (AN) has only recently been described. AN is defined by the presence of otoacoustic emissions (OAEs), an abnormal ABR, absence of middle ear muscle responses and elevated or absent behavioral responses to sound. Speech intelligibility is affected out of proportion to pure tone thresholds.[43,44]

The audiometric pattern is variable but typically demonstrates a rising or flat configuration. The hearing may fluctuate over time. Children with AN can achieve favourable results with cochlear implantation.[45]

AN has been associated with hyperbilirubinemia, neurodegenerative diseases, neurometabolic diseases, demyelinating diseases, hereditary motor sensory neuropathology (e.g., Charcot-Marie-Tooth syndrome), inflammatory neuropathy, hydrocephalus, severe and/or pervasive developmental delay, ischemic-hypoxic neuropathy, encephalopathy, meningitis and cerebral palsy.[46] A genetic factor may exist as AN has been described in families.[47] The postulated site of lesion is the inner hair cell/cochlear afferent system.[48]

Diagnostic Yield of Tests Used to Determine the Etiology of Sensorineural Hearing Impairment

When ordering investigations for SNHI, the clinician needs to know what the diagnostic certainty is associated with a positive or negative test result.

Genetic Testing

Studies have shown that the carrier rate for the 35delG mutation of connexin 26 (cx26) is between 2% and 3%, similar to the carrier rate for the gene for cystic fibrosis. The polymerase chain reaction assay for cx26 has a sensitivity and specificity of 97.4% and 96.9%, respectively.[13] In addition to its high yield, proponents of early genetic testing advocate its minimal morbidity. Although blood sampling allows for a greater yield of DNA, buccal smears are an alternative means of obtaining DNA.[49] Interest in the role of genetic testing continues to expand.

CT Scan

Radiological abnormalities may be present on CT scans in up to 37% of children with SNHI.[50] Large vestibular aqueducts (LVA) are the most common isolated findings, followed by lateral semicircular canal dysplasia, otic capsular lucency, small internal auditory canals and hypoplastic cochlea. At least 40% of patients with LVA will develop profound SNHI.[51] The presence of LVA may also indicate additional malformations and has been associated with stapes gusher syndrome, lateral semicircular canal dysplasia and Mondini deformity.[52]

Laboratory Studies

The low diagnostic yield for blood tests (CBC, platelet count, autoimmune evaluation and blood glucose), in the absence of other specific disease manifestations, does not justify their routine use.

The diagnosis of Pendred syndrome depends on the demonstration of the triad of congenital SNHI, goitre and abnormal perchlorate discharge test.[53] Given the rare abnormalities on thyroid function studies, they should only be performed in the presence of clinical signs and symptoms of hypothyroidism, presence of goitre, or when there is radiological evidence of LVA or Mondini deformity.

Other Studies

An electrocardiogram (ECG) may be valuable to detect conduction abnormalities associated with Jervell Lange Neilson (JLN) syndrome. It is a rare disorder with an estimated incidence of one to six cases per million.[54] However, in a review of the

reported cases of JLN ("hereditary Q-T prolongation syndrome"), the frequency in deaf children was found to be 0.3%.[55] Identification of patients with JLN can be lifesaving. An ECG is particularly valuable when a history of syncope, arrhythmias, or a family history of sudden death in a young child is elicited.

Urinalysis alone may not be adequate for the diagnosis of Alport syndrome. Examination of urine for glomerular basement membrane proteins, however, did provide a high diagnostic yield.[56,57] However, the cost of this test may be prohibitive to be used for routine screening. An unanswered question is the value of detecting hematuria and/or proteinuria on routine urinalysis. Routine urinalysis is inexpensive and simple to perform. Its role relative to the child with SNHI is unresolved.

Conclusions

- Medical evaluation of an infant with hearing impairment should be initiated at less than 3 months of age and intervention should be started by 6 months of age.
- Common etiologies of bilateral sensorineural hearing impairment (SNHI) include nonsyndromic gene mutations (such as connexin 26 (cx26) mutations), genetic syndromes (such as Waardenburg syndrome) and nongenetic causes involving preterm birth, asphyxia, meningitis, kernicterus, intrauterine infection and auditory neuropathy (AN).
- There is a need for evidence-based rational decision strategies, embracing history taking, physical examination, risk assessment and genetic testing and their interpretation, in children with bilateral SNHI.

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Management of Middle Ear Disease in Children Less than 2 Years of Age with Sensorineural Hearing Impairment

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Seventy to ninety percent of children will experience fluctuating conductive hearing impairment (CHI) secondary to otitis media with effusion (OME) with or without acute otitis media (AOM) in their first two years of life. The child less than 2 years of age with fluctuating CHI and underlying sensorineural hearing impairment (SNHI) presents a challenging diagnostic and therapeutic dilemma. Given the attention to and implementation of early hearing and communication development (EHCD) programs, physicians are increasingly requested to review young children and infants to assess and manage medical conditions associated with hearing impairment.

Measures of speech and language development have been shown to be negatively correlated with duration of time spent with OME in a child's first years of life.[1,2] Fluctuating CHI may have a greater effect on speech discrimination than a comparable SNHI.[3] The conditions that cause CHI affect the performance of various diagnostic tests,[4] further compromise hearing in a child with SNHI and lead to delays in diagnosis and hearing and communication development options.[5] Otoacoustic emissions (OAEs), for example, can be influenced by debris in the ear canal and the presence of middle ear fluid.[6]

This critical review focuses on the child with fluctuating CHI and SNHI. It does not address the management of children with permanent CHI (e.g., secondary to congenital ossicular fixation or external auditory canal atresia). These conditions pose less of a management dilemma for the physician and audiologist, and are beyond the scope of this review. In addition, the modification of behavioural and environmental risk factors for OME and AOM is not comprehensively addressed. Current evidence suggests the risk of AOM or OME in an otherwise healthy child 1 to 3 years of age is increased with exposure to passive smoking, group daycare attendance and bottle feeding.[7] The rationale for modification of these risk factors applies equally well to children with and without SNHI. Finally, audiological tests for OME were also not specifically reviewed, i.e., tympanometry. It is recognized that these tests are an important adjunct to clinical examination.

Diagnosis of Otitis Media with Effusion

Accurate assessment of OME may prevent delays in diagnosis and unnecessary treatment of children with a permanent childhood hearing impairment (PCHI). Pneumatic otoscopy is strongly recommended by the Otitis Media Guideline Panel[7] for the diagnostic evaluation of OME in otherwise healthy children ages 1 to 3 years, as this allows the observer to directly see the effect of positive and negative pressure on the mobility of the tympanic membrane.

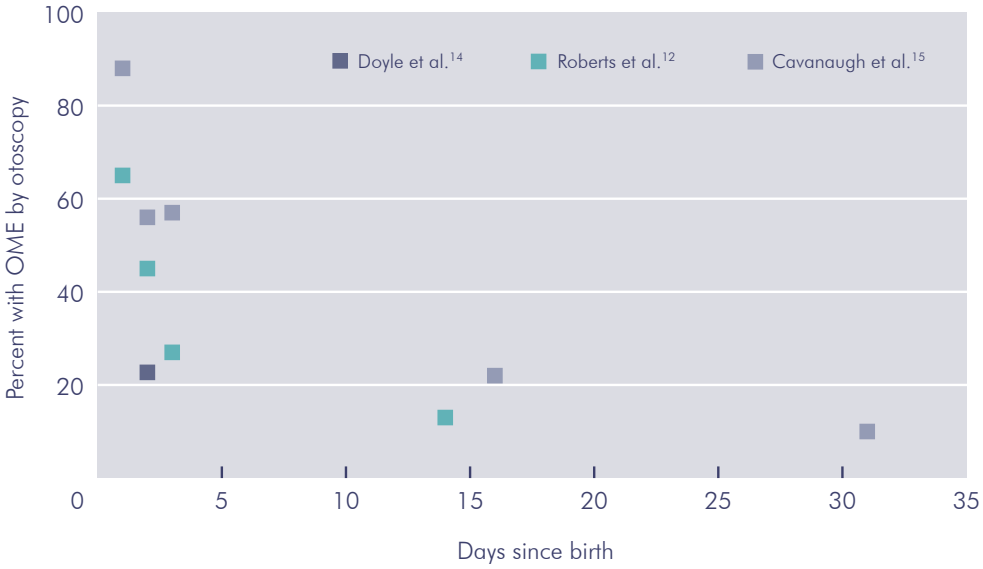
There is no published literature regarding the accuracy of pneumatic otoscopy in diagnosing OME in a child less than 2 years of age with SNHI. Studies in healthy children suggest a sensitivity of 85–98% and a specificity of 71–90%.[8-10] As with other procedures, tester training significantly improves diagnostic accuracy.[11] Clinical

examination results should be interpreted in conjunction with audiological assessments performed when diagnosing OME. Tympanometry and acoustic reflexometry are possible in children less than 2 years of age. Of note, the diagnostic agreement among the three tests is poor in the newborn period,[12] but improves significantly after 2 to 3 months of age.[13]

Middle Ear Fluid: Prevalence and Clearance

Most newborns referred for audiologic diagnostic evaluation from EHCD programs are subsequently found not to have PCHI. Retained amniotic fluid is a hypothesized cause for false-positive results on newborn hearing screening. Amniotic fluid takes many days to clear from the newborn's middle ear (see Figure 1).

Figure 1: Natural History of Clearance of Amniotic Fluid from the Middle Ear following Birth



The peak prevalence of OME occurs between 6 and 12 months of age and is significantly more common in high-risk born infants.[16] Unilateral OME will clear after an average of five weeks with or without a history of AOM; bilateral OME will clear on average nine weeks following AOM, and after eight weeks without a history of AOM.[17]

Managing Otitis Media with Effusion

Many clinicians recommend an aggressive approach to the management of OME in a child with an underlying SNHI. Developing guidelines for the treatment of children with OME and co-existing SNHI requires extrapolation from studies on otherwise healthy children. However, many experts recommend more aggressive management.[18]

Ventilation Tubes

Bilateral myringotomy and ventilation tube (BM&T) placement reduce the mean duration of OME over the year following their placement from 277 days to 142 days and result in improved hearing thresholds by a mean of 5–6 dB.[19] However, some of the complications of ventilation tubes are associated with CHI, i.e., otorrhea and perforations (2.2% for short-term tubes; 16.6% for long-term tubes).[20]

Behavioural problems may be more common in children with OME and improvement of the CHI by BM&T may improve the behavioural problems.[21] Verbal and expressive language scores in children with OME may be delayed compared to healthy children.[22] There is some evidence to suggest that BM&T will improve these measures as well.

Many physicians advocate an aggressive approach to children with OME and underlying SNHI. Based on this review, if OME persists for eight to twelve weeks, BM&T with short-term tubes (lower complication rate) should be discussed with the parents.

Antibiotics

Antibiotics offer a small short-term increase in the likelihood of resolution of OME in otherwise healthy children.[18] Seven children require treatment with an antibiotic for one to benefit — number needed to treat (NNT) = 7. This small benefit must be weighed against the potential adverse effects of antibiotic use which includes an increase in bacterial resistance to the antibiotic in the child's community.

In a child with co-existing SNHI and OME for four to six weeks, a course of 10 days of a first-line antibiotic (amoxicillin 40–80 mg/kg/day) may be warranted.

Other Treatments

Adenoidectomy is not indicated in this age group for the treatment of OME due to a lack of studies to support its efficacy.[7] Steroids, antihistamine-decongestants and tonsillectomy are not recommended.[7]

Management of Acute Otitis Media

Fluctuating CHI secondary to AOM in the child less than 2 years of age is presumed to cause similar problems to that caused by OME. Concerns of a lower rate of clinical resolution, and possibly an increased risk of complications from AOM in children less than 2 years compared to children more than 2 years of age, currently justify a 10-day course of a first-line antibiotic (amoxicillin 40–80 mg/kg/day).[23] Treatment should be individualized, as some children may be at a greater risk of long-term complications of middle ear disease warranting more aggressive treatment of AOM. For example, Canadian Inuit children may have a greater risk of developing CHI as a result of middle ear disease.[24]

A child with three or more episodes of AOM in six months, or four or more episodes in one year, can be managed with either prophylactic antibiotics or BM&T.[18] If BM&T is considered to be necessary in a child with underlying SNHI, intubation with short-term rather than long-term ventilation tubes is indicated given the lower risk of chronic perforation.[20] Alternatively, a child with recurrent AOM can be treated with a trial

of one to six months of prophylactic antibiotics (amoxicillin 20 mg/kg/day). This would avoid the risk of otorrhea and/or perforation secondary to BM&T but may increase the development of resistant bacteria in the community. There is good justification for restricting the use of prophylactic antibiotics due to an increase in bacterial resistance in the community following widespread use.

Conjugate pneumococcal vaccine will reduce the overall risk of AOM by 6–7%, the risk of pneumococcal AOM by 25% and the need for BM&T by 20% in healthy children who are less than 3 years of age.[25,26] Prevention of AOM in children with SNHI may be an additional reason to implement publicly-funded programs with this vaccine, if not universally, then for those at higher risk of the detrimental effects of AOM. Similarly, influenza vaccine, in particular intranasal formulations, may be beneficial in reducing the risk of AOM in children.[27] Vaccinated children had 6–30% fewer episodes of AOM compared to controls, although the study was not specifically designed to assess reduction in episodes of AOM. Further studies in this area are required.

Conclusions

- 70–90% of children will experience fluctuating conductive hearing impairment (CHI) secondary to otitis media with effusion (OME), with or without acute otitis media (AOM) in the first two years of life.
- Pneumatic otoscopy is recommended for the clinical evaluation of OME in otherwise healthy children under 2 years of age who have sensorineural hearing impairment (SNHI).
- Unilateral OME clears after an average of five weeks with or without a history of AOM, and bilateral OME clears on average after eight to nine weeks.
- Bilateral myringotomy and ventilation tube (BM&T) placement reduces the mean duration of OME and improves hearing thresholds, as well as some behavioural problems and expressive language scores in some children.
- Antibiotic therapy and conjugate pneumococcal vaccine should be considered in relation to middle ear disease in children.
- Further research is needed to explain the natural history of OME in children with SNHI, optimal antibiotic regimes and complications of myringotomy tubes.

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Chapter VI: Hearing and Communication Development

Amplification

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There is evidence that properly fitted hearing aids can improve auditory performance in children with hearing impairment.[1-3] Therefore, the early use of binaural amplification in infants with measurable residual hearing bilaterally is likely to be beneficial. To take full advantage of the early diagnosis of hearing impairment, the accurate fitting of hearing aid amplification must be undertaken in a timely manner, optimally within one month of the confirmation of hearing impairment and, whenever possible, no later than 6 months of age.[4] The fitting of amplification should proceed when: (1) hearing impairment has been confirmed; (2) hearing aid fitting has been chosen by the family following a review of expectations and options that has been presented in a complete and objective manner; and (3) when there is an absence of specific contraindications following a medical evaluation preferably performed by an otolaryngologist with extensive pediatric experience. Lastly, to ensure the accurate fitting of amplification with young infants, highly qualified professionals and pediatric-specific hearing instrument fitting protocols are required.[5,6]

Key Elements in Fitting Amplification in Young Infants

Assessment for Hearing Instrument Fitting

The accurate fitting of amplification in infants is dependent on the results of a valid and comprehensive assessment. Assessment data that are required before proceeding with the fitting of amplification include:

- frequency-specific and ear-specific estimates of hearing sensitivity
- determination of the type of the impairment — i.e., conductive, sensory, mixed (conductive and sensory), auditory neuropathy (AN)
- medical clearance for hearing instrument fitting
- measurement of the infant's occluded external ear acoustics using the real-ear to coupler difference (RECD) procedure[7]

Frequency-specific air conduction (AC) and, when indicated, bone-conduction (BC) threshold estimates must be obtained before proceeding with the prescription and fitting of amplification for young infants. For newborns and infants under the developmental age of 6 months, frequency- and ear-specific estimates of hearing sensitivity can be obtained by auditory brainstem response (ABR) measures.[8] Frequency-specific ABR data are necessary for accurate estimation of the degree and configuration of the hearing impairment in each ear. A click ABR measure alone is not sufficient for the accurate fitting of amplification. For older infants, ear-specific behavioural threshold measures should be obtained using visual

reinforcement audiometry (VRA). Acoustic immittance measures, including high-frequency tympanometry and middle ear reflex testing, and otoacoustic emission (OAE) are required to determine the type of hearing impairment present.[6]

The acoustic properties of the external ears of infants show high between-subject variability.[9-11] For accurate hearing instrument selection and fitting, this variable needs to be accounted for and applied at several stages in the amplification selection and fitting process. The RECD measure provides a reliable and valid means by which to capture the individual external ear acoustics for the purposes of fitting amplification.[7,12,13] Thus, in addition to conventional diagnostic audiometric test procedures, the prescription and fitting of amplification should not proceed until the infant's RECD has been quantified. This information is applied to the threshold assessment data to individualize the estimated hearing level to sound pressure level (SPL) transform used by modern pediatric hearing instrument prescription procedures.[14] In addition, this information is used at a later stage in the amplification fitting process to define 2cc coupler electroacoustic performance characteristics for the individual infant, as well as to predict real-ear hearing instrument performance from coupler-based electroacoustic measures.[13]

Hearing Aid Selection

A systematic, evidence-based prescriptive method specifically developed for pediatric applications should be applied in the fitting of amplification in young infants.[5,6] The prescriptive method should: (1) provide target values for the required frequency/gain function for a range of input levels; (2) provide target performance values by frequency for the maximum hearing instrument output; (3) systematically account for the developmental variations and changes over time in external ear acoustics; and (4) ensure audibility for a wide range of speech input levels and frequencies.[5,6]

Given the rapid advancement and ongoing introduction of new hearing instrument technology, further research is needed to determine the benefits and/or limitations of new digital signal processing options for application with infants.[6]

Electroacoustic Verification

The purpose of the electroacoustic verification stage of the hearing instrument fitting process is to ensure that the measured performance of the instrument to be fitted meets the prescribed criteria for each infant. Comprehensive electroacoustic verification for hearing instruments to be fitted to young infants requires that:

- measurements be made with speech or simulated speech test signals to accurately predict the hearing instrument gain in use environments
- measurements be made at multiple input levels to predict audibility for a range of everyday speech input conditions
- measurements of hearing instrument maximum output be obtained using pure tone test signals[6,15]

At present, the evidence suggests that a "simulated real-ear" approach to electroacoustic verification be applied in fitting amplification with the infant population. This includes the verification of all electroacoustic parameters in a 2cc coupler within a hearing instrument test chamber. Subsequently, an individualized acoustic transform, that includes the infant's RECD, is applied to predict real-ear hearing instrument performance.[7] A study by Seewald and colleagues[13] confirmed that this approach can be used to derive accurate predictions of real-ear aided gain (REAG) and the real-ear saturation response (RESR) in children. One advantage of this "simulated real-ear" approach to verification is that it does not introduce measurement error that can occur with conventional sound field probe-microphone measurements.

Monitoring Performance with Amplification

Once an infant has been properly fitted with hearing aids, the infant's performance with amplification must be closely monitored and evaluated. Over time, consideration is given to candidacy for additional assistive hearing technologies (e.g., FM systems) and/or alternative devices (e.g., cochlear implants).*

Frequent reassessment is important to ensure appropriate amplification parameters and hearing aid benefit over time. While there is a lack of research involving the validation of amplification in infants, there is a possibility that this may change as infant hearing screening programs progress.[10,16]

Studies have revealed contradictory findings that hearing aid use can cause marked threshold shifts on a child's aided ear. Poorly controlled aspects of retrospective studies and the use of group data contribute to these discrepancies. Therefore, frequent monitoring of thresholds and hearing aid functionality as part of a specific follow-up program for infants fitted with hearing aids is recommended.[5]

Conclusions

- Hearing aids can improve auditory performance in children with auditory impairment who have some hearing bilaterally.
- Coupler-based verification in conjunction with individual real-ear coupler difference (RECD) measurement is a valid procedure for the electroacoustic verification of hearing instruments.
- Further research is needed to explore new signal processing options.

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*Note: Cochlear implants have not been addressed within this section because this report deals predominantly with the infant in the first six months of life.

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Communication Development

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The majority of children with a permanent childhood hearing impairment (PCHI) are born to parents who have normal hearing and limited knowledge of hearing impairment. After a diagnosis of PCHI has been confirmed, parents need to choose an appropriate hearing and communication development (HCD) strategy. Some may choose amplification and a communication development option emphasizing the development of spoken language while others may choose not to amplify their child and pursue a manual mode of communication.

Historically there have been two major philosophies aimed at developing communication skills: oralism and manualism. The goal of oralism is the development of spoken language skills. Several variants have been developed. These include the aural-oral (A-O) approach which advocates the use of hearing through the use of amplification devices, which in turn is facilitated by speech reading. The highly structured auditory verbal therapy (AVT), which is another form of the oral approach, relies almost exclusively on residual hearing and the development of listening skills.

The manualists, on the other hand, do not feel that it is necessary for individuals with a permanent hearing impairment to develop spoken language and see signing or visual language systems as a natural language of the deaf. This communication approach is seen as facilitating the full participation in the deaf culture. Many sign language systems (such as American Sign Language (ASL) and Langue des signes quebecois (LSQ)) are visual languages with their own grammar and syntax. For this approach to be effective, the child must be in contact with adults who are fluent in the chosen sign language system.

The total communication (TC) approach, which until recently was a popular option, combines elements of the oral and manual approaches. Various methods of communication, including sign language, finger spelling, natural gestures, speech reading and spoken language, were all integrated. The simultaneous use of spoken and signed language was seen as a way to facilitate the development of communication skills.[1] TC was a multisensory approach where amplification and signing were used together. Proponents of this approach believed that signing would facilitate the development of spoken language.

Regardless of the approach, the belief is that early intervention is key to the development of communication and social skills and of academic functioning. Early intervention is being facilitated by the development of universal newborn hearing screening (UNHS) programs. In addition, technological developments such as cochlear implants are providing increased access to auditory information by children with a PCHI.

In order to select an appropriate HCD approach for their child, parents need to be provided with adequate information. The choice of an HCD option for parents must be an informed choice based on scientific evidence. In order to determine the effectiveness of the four main types of approaches used until recently (A-O, AVT,

ASL, TC) for children with a congenital PCHI, a systematic review of the empirical evidence was carried out by the Chalmers Research Group in Ottawa.[2] This section provides a brief summary of the results of the systematic review.

The search strategy consisted of a variety of existing electronic databases. Studies were eligible for inclusion if they were characterized by any level of evidence above opinion, if they involved children with a congenital PCHI, and if at least one subject in the study had received one of the four HCD options of interest to the review. Several independent reviewers were involved in the selection of the studies.

Two types of studies were found to be relevant. These included direct and indirect evaluations of the impact of the approaches of interest to the review. In direct studies, a variety of designs were identified (uncontrolled case study to randomized controlled trials (RCTs)) and these included any or no comparators, and were conducted specifically to determine the effectiveness of one of the approaches of interest. Indirect studies established the potential utility of at least one of these types of programs.

Of the two types of studies, direct and indirect, the direct types of evaluation were more likely to have instituted scientific methods of control to assure the study's internal validity. Indirect studies had a different focus and were somewhat less rigorous. Of 625 citations, 194 unique studies were entered into data abstraction. This included 91 direct and 103 indirect evaluations of review-relevant programs. Evidence tables were derived with variables highlighted by clinical content experts. An examination of these tables indicates a preponderance of missing data — examples of missing data included sample size, degree of hearing impairment, route to identification and types of amplification.

The observations obtained in the systematic review do not permit the confirmation or disconfirmation of the absolute or comparative effectiveness of any of the four HCD options reviewed. One of the major reasons, as previously stated, was the preponderance of missing data in the majority of studies. This was attributed to the failure of investigators to recognize the importance of investigating the collected data according to key population- and intervention-based variables, the inability to reliably measure the variables or to a failure to report the data.

The systematic review indicates that it was impossible to ascertain the exact number and other characteristics of children meeting the review's eligibility criteria. This gave rise to an incomplete qualitative synthesis. The authors of the systematic review conclude that the state of the research literature on HCD approaches appears to be one of disarray with considerable amounts of missing and divergent information. The question concerning the effectiveness of the four review-relevant approaches for children with a congenital PCHI cannot at the moment be answered in the affirmative or the negative. The authors acknowledge that the questions concerning the effectiveness of habilitation programs are complex as a multitude of variables can have an impact on outcome.

A key goal of future research could be the ascertainment of the approach that would be most appropriate for *each* child *and his or her family*. HCD options need to be scientifically based while taking into account social and cultural factors. Well-designed and controlled studies are needed, as parents require the scientific evidence that will allow them to make informed choices on the most appropriate HCD option for their child.

Conclusions

- A systematic review of the research on outcomes of the most common communication development options (aural-oral (A-O), auditory verbal therapy (AVT), American Sign Language (ASL) and total communication (TC)) revealed that inter-study variability and limitations of study design and analysis preclude the establishment of the effectiveness of any of the four communication development options.
- The lack of definitive proof, however, does not mean these communication development options are ineffective, but that further studies are required.
- Progress in this area would be facilitated by development of quantitative measures of oral and manual language development applicable to infants.

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Chapter VII: Outcomes

Universal Newborn Hearing Screening: The Evidence

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Permanent childhood hearing impairment (PCHI) has been associated with delays in speech, language development and learning. It has been reported that deaf students graduate from high school with language and academic levels corresponding to those of fourth grade students with normal hearing.[1,2] One of the factors which has been linked with the delays in language and academic development has been the age at diagnosis. Universal newborn hearing screening (UNHS) is seen as a strategy that leads to the early identification of children with PCHI and to the provision of early hearing and communication development (EHCD) programs. EHCD programs are seen as ways to reduce the gap in language and academic skills between hearing and deaf students. The determination of the evidence in support of UNHS programs has, therefore, focused predominantly on two main aspects: (1) Do UNHS programs lead to a greater number of children with PCHI being identified and treated earlier? (2) Does EHCD improve the development of language and communication?

The U.S. Preventive Services Task Force (USPSTF)[3] represents one of several initiatives which review evidence to ensure that the development of clinical practice guidelines is based on scientific evidence rather than on expert opinion. Systematic searches of multiple bibliographic research databases help to identify relevant literature in an unbiased and thorough manner. Quality criteria developed by methodologists are used to guide judgments of the strengths and weaknesses of individual studies. Two independent members of the topic team usually review abstracts of all articles. Once a decision has been taken to include an article, information is abstracted on patient population, study design, interventions (where applicable), quality indicators and findings.

The strongest empirical support comes from experimental designs involving randomized controlled trials with large numbers of subjects. This is viewed as the only design that permits clear linkages between the intervention and associated outcomes. The second type of evidence is usually obtained from quasi-experimental designs, often seen in cohort studies. These are prospective studies in which a large group of individuals with a common characteristic are followed over time and a particular outcome investigated. Control groups are usually involved allowing for intergroup comparisons. These types of studies are seen as providing less compelling evidence than randomized controlled studies. A third type of study usually involves the use of non-experimental designs that can often be retrospective in nature. These types of studies are often seen as lacking experimental controls and are often criticized. Once a review of the literature has been carried out, the supporting evidence is reviewed and rated and statements and recommendations are issued.

The USPSTF limits the areas to be reviewed to those conditions that cause a large burden of suffering to society and which can potentially be alleviated by some form of prevention. Good or fair quality evidence for an entire preventive service must

include studies of sufficient design and quality to provide linkages that connect the preventive service with the health outcome. Newborn hearing screening is seen as a preventive measure that should be associated with enhanced speech and language development in children with a PCHI. The topic of UNHS has been reviewed by the USPSTF, through the Oregon Health Sciences University evidence-based practice centre and the results of this review appear in an article published by Thompson and colleagues in 2001,[4] which is summarized in this report.

Universal Newborn Hearing Screening and Early Identification of Hearing Impairment

The ages of diagnosis of children identified in the absence of UNHS and in the presence of UNHS have been addressed in Chapter III in this document. Some of the articles presented in that chapter will be reviewed briefly in this section. The Wessex Universal Neonatal Hearing Screening Trial[5] reported an increase in the number of cases with a significant hearing impairment who were identified and treated early. Seventy-one more babies with a moderate to severe PCHI per target population of 100,000 were referred before 6 months of age during periods with neonatal screening than during periods without. In addition, UNHS led to an increase in confirmation and management of hearing impairment by 10 months of age. Fifty-seven percent of children with moderate or severe hearing impairment were diagnosed with UNHS in comparison with 14% without UNHS. In the best quality U.S. study,[6] in the presence of UNHS, the age of diagnosis for mild to moderate and severe hearing impairment was approximately 6 months and below. Both the Wessex trial[5] and the Dalzell et al. study[6] have been rated as providing good evidence by the Oregon Health Sciences University evidence-based practice centre. The studies were rated as being well designed — the U.S. study being a cohort study and the U.K. study being a controlled non-randomized trial. Based on the review published by this centre, the USPSTF has stated that there is good evidence that newborn hearing screening leads to earlier identification and treatment of infants with hearing impairments. The next question which needs to be answered is whether early identification and treatment resulting from UNHS improve language and communication development.

Universal Newborn Hearing Screening and Language and Communication Development

Thompson et al.[4] reviewed the articles which have investigated speech and language development in children with a PCHI identified through UNHS. Their literature search indicates that at the moment there are no prospective, controlled studies that have directly examined whether newborn hearing screening and early intervention give rise to improved speech, language and educational development. Eight recent cohort studies from three intervention programs are summarized in the Thompson et al. paper.[4] The studies reported used standardized receptive and expressive tests to evaluate the speech and language skills of pre-schoolers.

All of the studies reported statistically significant associations between age of diagnosis and language development at ages 2 to 5 years. Six of the eight studies reported results on children in the Colorado Home Intervention Program. One of these studies[7] compared the language performance of hearing impaired children born in hospitals with UNHS programs to that of children born in hospitals without. The results showed that the mean scores for expressive, receptive and total language were within normal ranges for the screened group and significantly higher than for the unscreened group. The evidence provided by this study was rated as poor because the authors used a convenience sample, the assessment of outcome was unblinded and the exclusion criteria not specified. In another study by the same group,[8] which was also rated as poor, children identified before 6 months of age were seen to have language scores at or near their cognitive test scores, whereas children identified after 6 months performed on a significantly lower level than their cognitive test scores. This study was criticized based on the statistical method used in the data analysis, on the fact that no information on dropout rates was provided and that the assessments were not masked.

The articles reviewed by Thompson and colleagues[4] were all seen as having several limitations. The subject populations were comprised mostly of convenience samples. The inclusion criteria were unclear and the assessments were not blinded. In addition, none of the studies provided information on attrition and follow-up rates. The USPSTF rated the quality of the evidence linking early intervention with language outcomes as inconclusive and the quality of the evidence as fair to poor.

The USPSTF concluded that there is a need for population-based studies that begin with inception cohorts which carefully report outcome on all possible subjects, as well as rates of follow-up and attrition. There is a need for prospective, longitudinal studies which report on the speech, language and education development of children with PCHI over time.

Since EHCD programs are relatively new, there is a need for additional well-controlled research to determine the efficacy of UNHS. Such research is complex and difficult. Many factors have the potential to impact on the communication development of children with a PCHI, such as parental involvement,[9] degree of hearing impairment, additional handicapping conditions and quality of pediatric care. Many variables can contribute to developmental outcome and not all of them can be included in a research design. Nevertheless, new EHCD initiatives are providing opportunities to prospectively follow children with PCHI who have been identified early, taking into account, as much as possible, the many variables which can have an impact on outcome.

Conclusions

- Some studies have concluded that early identification and strong family involvement improve the development of speech and language in infants and young children with hearing impairment.
- There is a need for more research in this area, in particular whether universal newborn hearing screening (UNHS) programs and early hearing and communication development (EHCD) lead to improved speech, language and education development.

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Chapter VIII: Infrastructure

Context

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The scope of an effective early hearing and communication development (EHCD) program for hearing impaired infants is very broad. A population/public health approach, rather than the traditional doctor/audiologist/patient medical model, is likely to support most appropriately all of the processes necessary to ensure the earliest possible access to communication and literacy development for hearing impaired infants. A variety of professional groups and agencies must be able to work together collaboratively if an EHCD program is to be successful.[1]

The World Health Organization (WHO)[2] and the National Screening Committee of the British National Health Service[3] have identified specific criteria that justify implementation of a screening program. Both of these bodies identify acceptable diagnostic tests and access to effective treatment or hearing and communication development (HCD) options, among other criteria, as necessary ingredients of screening programs. In this context, EHCD programs are comprised of successive stages, or subprograms, that include screening, surveillance, audiologic assessment, medical assessment, family support and provision of options for communication development. Hearing screening, the starting point of an EHCD program, has traditionally been the component that has received the greatest attention. However, a preoccupation with screening may result in insufficient attention to the other, critical components of EHCD programs. Bess and Penn[4] have reiterated that: "it is inappropriate to screen for any disorder without certainty that facilities for suitable follow-up care of individuals who fail the screen are readily available."

The performance characteristics of each successive sub-program of the EHCD program are critical because deficiencies in each will compound serially and may seriously compromise the overall EHCD program performance.[1] The weakest link in the chain will dominate overall program effectiveness. For example, limitations in screening coverage, imperfect attendance for definitive audiologic assessments and limited compliance with follow-up recommendations can result in a serious, cumulative program shortfall. For an EHCD program to be "successful," each sub-program must have excellent performance characteristics and its goals/objectives must align with all the others, and all sub-elements must communicate and link effectively.[5]

Ideally, the development of an EHCD program, including each of its sub-programs, should conform to standard principles of program design.[6] Prior to implementation, each component should have clearly articulated, predetermined goals and objectives, preferably in quantifiable terms. Structures, processes and outcomes should be defined in such a way as to make program evaluation and quality management possible. To the greatest extent practicable, these considerations should have been dealt with before the first baby is screened.

A poorly-structured program that does not deliver what it seems to promise (i.e., to promote effective family communication) can have negative consequences that are difficult to anticipate. For example, an EHCD program, even though it may have poor performance characteristics, may result in other systems or services for dealing with this population to be altered or withdrawn, on the assumption of the existence of appropriate, replacement mechanisms. A comprehensive examination of infrastructure will anticipate such performance issues before they become problems and identify possible solutions. For example, initial funding of a universal screening program may be insufficient to provide definitive audiologic assessment for all infants who have a refer result on screening, or to provide for appropriate HCD options. A temporary solution to such a program deficiency might be to identify a smaller target population (e.g., a high-risk group) for whom the full spectrum of services is deliverable within the funding envelope. Such a model program approach could pave the way for a larger, more comprehensive and fully funded EHCD program.

Infrastructure

Infrastructure is the "glue" that keeps all program components together and in synchrony. It relates to those elements that support, sustain and link all program components to achieve the ultimate program goal. The main components are: human resources, information systems, administrative systems and communication systems.

1. Human Resources

Good human resources (HR) management is a critical feature of a successful EHCD program. In addition to the traditional attributes of good HR practice (e.g., appropriate qualifications, clear roles and responsibilities, job descriptions, performance management, appropriate orientation and training, and quality assurance programs), it is important to consider the "softer" side of staff recruitment and retention. Staff who are committed to the program and perceive their work to be valuable will do their best to make the program succeed. Staff who have been co-opted or have had their workload stretched to breaking point are unlikely to maximize program performance.

While it is common to discuss EHCD programs in terms of technology choices, pass/refer rates, and compliance with follow-up as though they were the same discreet entities from program to program, such features are often governed primarily by staff attitudes towards the program. Sometimes, for example, the obvious choice for screening personnel does not play out in the "real world" — overloaded nursing staff may resent additional work unless they are convinced of its value to their patients, whereas support staff may find the work challenging and interesting. Local circumstances often dictate the best choice of staffing for some tasks, unless specific expertise is required (e.g., an audiologist for detailed hearing assessments).

Any EHCD program will need staff with a variety of skill sets if it is to be sustainable. A coordinator who oversees the whole program is a key individual and local circumstances usually will determine the person best suited for this important task.

Typically, there is also need for some level of clerical support to take care of, for example, data entry, filing and supplies. The professional staff involved include the screeners, the audiologist, auditory verbal therapists, American Sign Language (ASL) and Langue des signes quebecois (LSQ) providers, aural/oral therapists, counselors and hearing aid dispensers. Information systems (IS) and information technology (IT) support staff who can maintain and service all of the computing and test equipment are also key individuals. The coordinator should be an individual with excellent "people" skills and diplomacy to meld what is typically a disparate group of individuals into a smoothly operating team.

In addition to staff dedicated to and funded by the EHCD program, other professional groups such as otolaryngologists, neonatologists, paediatricians, family practitioners, social workers, nurses, speech-language pathologists and deaf educators will be involved with identified infants. Full-time EHCD staff will have to devote effort to developing effective linkages and communication systems with these professional colleagues, many of whom may be less familiar with the deaf/hard of hearing neonatal/infant population. The relevant professional groups will vary regionally, but the importance of identification and inclusion of all the key players cannot be underestimated.

2. Information Systems

The importance of a high quality, automated IS for tracking, follow-up and seamless transition from one program stage to the next, as well as for program evaluation, cannot be overemphasized. The problems with manual methods have been described.[7] Several standard software packages are available, so it is important not to undertake development of custom systems without a great deal of thought, expertise and resources. Identification of critical data fields requires a deep understanding of the goals of the program and the ultimate intended use of the captured information. A shotgun approach to data collection is likely to consume resources and yield little useful information. If physical forms are utilized, they should be user-friendly and be formatted so that they facilitate accurate and rapid data entry.

Consent and confidentiality are especially important considerations in relation to information management. Parental consent is commonly required before any EHCD procedure is performed or any results are sent to third parties.[7] If an institution were to adopt hearing screening as part of its standard of care, then the consent for screening itself would be subsumed under a global consent. But care must be taken to ensure optimal and timely flow of communication regarding individual infants and families. However, even in that situation, consent to release results to third parties would probably be required. Knowledge of local legislation is important when designing an EHCD program: in one province of Canada, for example, it is legislated that consent must be obtained before any information can be transmitted electronically. Program design must also take into account that families/caregivers have the right to decline consent, and the program must be able to deal with such circumstances effectively, for example, by providing helpful information that may encourage consent subsequently.

Confidentiality is an important feature of our medical system and it is important that all EHCD program staff be aware of the need to treat all personal client information as confidential.[4] Confidentiality requirements extend to communication of patient information by any means — spoken, written or electronic. For example, when designing EHCD program facilities, fax machines and computers should be located in areas accessible only to staff, and telephones should be located so that the public is not privy to staff conversations. Program quality assurance systems will have to address the integrity of their confidentiality and consent systems. For example, the IS should be structured so that it is possible to audit who has accessed patient files and whether consents were recorded.

3. Administrative Structures

EHCD programs, like all other programs, need administrative structures to ensure that program mission, goals and objectives are defined and support its clinical functions. The program co-coordinator is the person most likely to be responsible for these functions. They include setting up systems for maintaining financial, staff and clinical records. Budgeting and fund raising are critical features of program infrastructure. The program should have a transparent accountability/reporting structure that includes all program personnel and is accessible to anyone who has a right to access program information. The administrative system would ensure that the program has developed and defined standard protocols and that they are disseminated and adhered to by all staff. Forms development is an important function — staff will follow standard protocols and enter all required data fields when they have efficient, user-friendly forms. The administrative structure would also ensure that staff has access to appropriate, ongoing training and educational opportunities. Program evaluation and continuous quality improvement initiatives are administrative responsibilities that will ensure program efficiency, effectiveness and systematic evolution.

4. Communications and Public Relations

A broad-based communications/public relations program is an invaluable means of securing support and demand for an EHCD program. Parents, professional groups (e.g., audiologists, otolaryngologists, neonatologists, paediatricians, family physicians, nurses, etc.) and consumer groups should receive information about the EHCD program by as many means as possible. Television and radio spots, Internet web pages, videos, newspaper articles, professional journal articles, consumer group newsletters, pamphlets and brochures are all possible means of "getting the information out there." Multiple means are more effective than one or two approaches because different groups seem to have different preferences for information content and presentation. For example, many physicians seem to prefer relatively brief, factual information, written in bullet point form, as opposed to videotapes. A good promotional effort will galvanize all stakeholders and thereby improve consent for screening, compliance with follow-up recommendations and, ultimately, long-term sustainability of the EHCD program when the inevitable competition for resources arises.

Every EHCD program exists within social, cultural and political values and contexts that influence how a particular region chooses to spend its resources. Catching the brass ring is often a question of being known by the right person (the key "influencer") at the right place and the right time (the luck component), but usually that alone is not sufficient. Being prepared to provide potential sponsors with evidence that is easily understood is usually vital for making the case to support an EHCD program.

Conclusions

- Public health system models and linkages seem more appropriate than traditional medical models for effective delivery of comprehensive early hearing and communication development (EHCD) programs.
- Human resources are critical components of EHCD programs.
- Careful attention is also required for information management, administrative structures and external communications if an EHCD program is to be effective and sustainable.

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Chapter IX: Program Evaluation

Program Evaluation and Quality Improvement

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Program evaluation and quality improvement (PEQI) are essential components of any high quality program for early hearing and communication development (EHCD). Both components must exist as a sub-program of the overall EHCD program. The PEQI sub-program should address structure, process and outcome elements of the entire EHCD program. The PEQI sub-program requires explicit and precise *a priori* definition of quantitative and realistic objectives for the EHCD program as a whole, as well as for each and every one of its major, programmatic components. Additionally, PEQI programs should include clear and demonstrably effective mechanisms for EHCD program adjustment in response to observed deficiencies.

Program evaluation is a formal method by which the EHCD program directorate can determine whether the overall goals and specific objectives of the program are actually being achieved. To be worthwhile, such an evaluation must lead ultimately to concrete changes in each and every component of the program that is thought to contribute to any observed shortfall in achievement of objectives. Quality improvement is a related conceptual approach that incorporates ongoing and continuous re-examination of the program components in order to determine proactively whether the program is functioning as effectively and efficiently as possible, and to determine when and where problems or concerns arise, so that the components can be maintained, repaired and, wherever feasible, enhanced (see, for example, Donabedian[1]).

In accordance with a widely-accepted health services evaluation conceptual framework, a good PEQI program can be formulated in terms of the structural, process and outcome components that define the overall EHCD program.[2] The structural elements of the EHCD program include but are not limited to:

- administration
- personnel training, performance management and continuing education
- information management, including tracking and reporting
- instrumentation procurement, calibration, maintenance and manufacturer liaison
- clinical protocol development, dissemination and updating
- family consenting, infant risk assessment; contact, screening and follow-up compliance procedures
- audiologic assessment procedures
- communication development options provision
- family support provision, surveillance procedures, referral procedures, measurement procedures for outcomes and proxy outcomes

- quality management
- public and professional education
- ongoing technology
- evidence assessment

Program Evaluation

Program evaluation requires that the program and its component sub-programs have associated outcome measures that are not only explicitly definable but also quantifiable. The next step is to develop realistic indicators and benchmarks for each of the outcome measures; the program can then use these to determine if it is meeting its intended performance targets. The benchmarks are levels to which programs should aspire, in order to know that what is being achieved satisfies defined standards of care. They are important in EHCD programs to prove to health professionals and decision makers that EHCD is both beneficial and cost-effective.[3] It should be recognized that there is clear evidence of program improvement over time and that benchmarks might be difficult to achieve, particularly at the outset. However, asymptotic performance benchmarks should be achievable in the second or third year of implementation. Program administrators should encourage and assist individual sites in the pursuit of performance benchmarks that are reasonably uniform and consistent with the program's overall benchmarks. Failure to achieve benchmark performance should be clearly evidentiated and improvements aggressively sought and documented.

Outcome Measures

The outcome measures that a high quality EHCD program should routinely quantify and document are listed below. Measurements may include both true outcomes and process events that may serve as proxies for true outcomes. The latter type of measurement is common in situations wherein true outcomes are difficult, expensive or impossible to obtain. An example is use of a habilitative service event as a proxy for the desired effect of that service on language development.

- The number and proportion of the overall target population successfully screened by 1 month of age, or within one month of discharge from the birth hospital admission.
- The numbers and proportions of infants with a refer result from screening overall and broken down by screening site, screening personnel and risk status.
- The numbers and proportions of screening referrals for whom audiologic assessment is initiated by 3 months of age and within two months of the initial screening result.
- The numbers and proportions of screening referrals with completed audiologic assessments by 4 months of age and within one month of assessment initiation.

- The numbers and proportions of the target birth, screened and referred cohorts of infants who have confirmed permanent childhood hearing impairment (PCHI) at 6 months of age, also broken down by ear and severity categories, risk status and assessment site/personnel.
- The numbers and proportions of infants with PCHI who have been recommended for hearing aids by 6, 9 and 12 months of age.
- The numbers and proportions of infants with PCHI who have received medical intervention for otitis media (OM) by 6 months of age.
- Documentation of reasons for non-fitting of hearing aids by 6, 9 and 12 months of age, broken down by risk status.
- The number of infants with confirmed PCHI at 12 and 24 months of age who passed newborn screening and were detected by surveillance and referral-in routes.
- Family satisfaction with EHCD program processes.
- Communication development outcomes, broken down by type and degree of PCHI.

Performance Benchmarks

Using the outcome measures above, programs can determine if benchmarks have been achieved. The following are examples of feasible benchmarks. (A more comprehensive set of benchmarks can be found in JCIH.[4])

- Within six months of program initiation, hospitals or birthing centres screen a minimum of 95% of infants during their birth admission or before 1 month of age.
- The referral rate for audiologic and medical evaluation following the screening process should be 4% or less within one year of program initiation.
- The agency within the EHCD program with defined responsibility for follow-up documents efforts to obtain follow-up on a minimum of 95% of infants who do not pass the hearing screening.
- Infants referred from universal newborn hearing screening (UNHS) begin audiologic and medical evaluations before 3 months of age, or three months after discharge for neonatal intensive care unit (NICU) infants.
- Infants with hearing impairments are enrolled in a family-centred early intervention program before 6 months of age.

Long-Term Outcomes

Measures that may appear to be proxies for long-term outcomes may constitute true outcomes at the sub-program level. For example, the UNHS component of an EHCD program exists solely in order to deliver all correctly identified infants with PCHI to appropriate follow-up services, with minimal delivery of false positives.

In the larger sense, however, EHCD programs exist not to screen or to diagnose hearing impairment but to deliver effective interventions and communication development services. Ultimately, therefore, the true measures of a successful EHCD program lie in outcomes such as family satisfaction with services, improved hearing ability at an early age, and improved communication development over the long term. There is an acute need to develop and apply valid and accurate measures that reflect these diverse outcome domains. Because randomized clinical trials of EHCD programs are clearly unfeasible, the ultimate validation of the benefits of EHCD will lie in comparisons of a variety of outcomes with their historical controls. This approach has been strongly advocated in several critical evaluations of UNHS/EHCD initiatives, including formal, evidence-based systematic reviews.

Economic Evaluations

Aside from the outcome measures listed above, which provide information service processes and associated outcomes, funding agencies routinely require both budgetary and more comprehensive economic evaluations of EHCD programs. These are relevant for accountability and sustainability issues, especially in relation to cost containment and to opportunity costs of alternative health care programs. The functionality of the EHCD program is contingent upon its continued funding. Therefore, economic data need to include, but are not limited to, the capital and operating direct costs of *all* program structures and processes, including infrastructural elements such as information systems. A common problem in such analyses is to differentiate the true costs of the EHCD program from other costs that are associated with the health care environment within which the EHCD program operates. These analyses must also take into account the actual costs that would ultimately be sustained in the absence of the EHCD program, and this is a difficult area for which quantitative data are frequently lacking. An example of the difficulty is the weighing of direct EHCD costs against the actual, historical costs of the ad hoc and frequently inadequate assessment and intervention services that are the norm in the absence of EHCD programs.

Basic measures that are commonly used for EHCD direct cost evaluation are the overall cost per infant screened and the cost per infant identified with a PCHI.[5] Such analyses are fundamental and are a useful facet of PEQI, especially for comparative cost-effectiveness analysis of various process options. Cost-effectiveness analysis (CEA) will be discussed later in this chapter. For example, cost per infant identified may be very sensitive to the referral rates to audiologic assessment that are achieved by specific screening strategies, and this may be more meaningful than simple use of process proxies such as the raw referral rates. However, a much more comprehensive analysis is required to approach true cost-benefit issues, because cost-benefit analysis (CBA) requires attribution of quantitative, monetary costs to long-term outcomes such as educational achievement, earning capacity and quality of life. This is an area that is at a rudimentary stage and requires much further investigation.

False-Positive Referrals

Some of the "costs" associated with program performance are extremely difficult to quantify. An important example are the costs associated with false-positive screening referrals. Much concern has been expressed in the literature about the impact of false positives on both parental anxiety and child bonding, as well as on more easily determined costs such as those of unnecessary audiologic assessments, assessment errors and inappropriate interventions. At present, a major focus of program quality is based upon minimization of the screening false-positive rate, as a proxy cost measure. A related key focus is upon audiologic assessment error minimization.

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Cost-Effectiveness Analysis

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The decision to implement a systematic program for early hearing and communication development (EHCD) must take account of the net costs to the individual family and to society as a whole. This is especially relevant in the context of a limited health care cost envelope, within which expenditure on one type of service implies restriction of some other health services. Cost analysis increases in importance if the proposed program does not clearly reflect an ethical imperative or a core societal value, for example, that "all children and families have a basic right to early, effective communication." Debates about such value statements in relation to EHCD are as yet at an immature stage.

Cost-effectiveness analysis (CEA) seeks to comparatively evaluate different service models, usually in terms of cost per unit valued outcomes, such as cases successfully identified, diagnosed or achieving specific long-term criteria of success. Cost-benefit analysis (CBA) can be viewed as an extension of CEA, within which *all* activities, outcomes and side effects of a program are expressed in a common, monetary unit.

When the net costs of a program are assessed comprehensively and are negative (i.e., a net monetary gain), then the cost issue is moot, except for the politically common issue of short-term cost of long-term benefit. When there is significant, positive net cost, society must consider value judgments, typically relating the proposed program to other, comparable activities already occurring (such as other newborn mass screening activities). Society must also consider the costs associated with the status quo (in the absence of the proposed program) and how they may change, especially in relation to the quality of life of affected individuals and families. It is often difficult and contentious to assign monetary values to such domains, although several sophisticated methods are available (such as utility analysis).

There have been several published cost-related analyses in connection with EHCD programs. The approaches and models used vary in sophistication and completeness, and the field is characterized by variability of methods, assumptions and results, as well as limitations of data underlying rational choice of key parameters. By far the most sophisticated analysis to date is a report by Keren et al.,[1] which includes an extensive bibliography of earlier publications in the area. Keren et al.[1] attempted to identify the best data available for the many key parameters in a comprehensive model. In addition to the usual program capital and operating costs, they estimated long-term savings from reduced, special educational costs, as well as improved work force productivity. The key methods, findings and limitations of the study are presented here.

The target disorder for the CEA was defined as a bilateral hearing impairment of $\geq$40 dBHL. This is a typical selection, but it is the most conservative criterion within reason. Many EHCD programs, especially in the United States, have much more liberal criteria, typically including lesser degrees of hearing impairment and unilateral impairments. A change in criterion definition has a strong effect on the prevalence of the disorder, and increases the incremental yield of both targeted

and universal systematic screening. This limits the direct applicability of the analysis results to many current screening programs, although the methodology remains generally appropriate.

Many probabilities used in the model were available through data from current hearing screening programs. Reasonable estimates were available for the prevalence of hearing impairment in low- and high-risk infants, the proportions who complete testing at each stage of the protocol, and the proportions of infants with hearing impairments detected prior to 6 months of age and who had intervention prior to 12 months of age. Some probabilities were estimated — for example, sensitivity and specificity of screening tests. While there are excellent data available on specificity, current data on sensitivity are limited, so actual program sensitivity may diverge considerably from the assumed values of 0.95. The assumption of identical sensitivities for otoacoustic emissions (OAE) and auditory brainstem response (ABR) is also questionable, given that the ABR is sensitive to auditory neuropathy (AN), whereas the OAE are not, and that AN may be present in up to 10% of all infants with bilateral permanent childhood hearing impairment (PCHI).[2]

Screening costs included capital costs for equipment, as well as labour costs. Long-term societal costs accounted for lost productivity, special education, vocational rehabilitation, medical costs and assistive devices.

With the nominal values of all base parameters in the model, three situations of no screening, targeted screening for high-risk infants and universal screening detected 30, 66 and 99 of 128 infants with PCHI, respectively. Costs per infant diagnosed were $2,300, $10,100 and $21,400, respectively. The incremental (marginal) cost per additional case diagnosed was $16,400 for targeted screening, and $44,300 for universal screening (all values in $U.S.). These values were considered comparable to those that apply in detection of hypothyroidism and phenylketonuria.

When lifetime costs were considered, both high-risk and universal screening programs were found to result in overall cost reduction, assuming that intervention prior to 12 months of age improved speech and language outcomes. For universal screening, a majority of cases detected were required to achieve normal language outcomes, in order for there to be a net cost reduction.

An interesting feature of the model is that the numbers of hearing impaired children with normal language outcomes were 53, 59 and 65 for the three situations noted; clearly, the assumptions used were such as to yield these very modest differences in this important outcome.

Keren et al.[1] used sensitivity analysis to examine the extent to which their result varied for different values of key parameters in the model. This is an essential component of any such analysis, if it is to be relevant, valid and generalizable. The incremental cost per case diagnosed by universal screening was found to be very sensitive to the assumed proportions of successful long-term language outcomes and to the impact on lifetime productivity. It was moderately sensitive to the assumed success rate for targeted screening, and to the losses to follow-up in a universal program.

The Keren et al. model[1] highlights a need for better data on long-term impact of EHCD programs on language levels and on lifetime productivity. It also underscores the importance of high rates of follow-up diagnostic evaluation in infants who do not pass screening; this is a common area of deficiency in EHCD program reports to date. It is important to note that, for example, a 20% loss to follow-up of screening referrals translates to a 20% reduction in the effective sensitivity of the screening process, whatever the estimated, intrinsic sensitivity of the screen.

A final caveat is that overall performance of a screening program is a function of many parameters of program operation, as well as contextual variables relating to the health care system in which the program is embedded. Moreover, simple sensitivity analysis involving adjustment of one or even two parameters may not reveal the true effects of program improvement in several aspects simultaneously. Because of these and other limitations, the Keren et al. report[1] may be considered as illustrating an appropriate methodological approach to CEA in relation to early detection of hearing impairment. However, further work in this area is required to ensure its generalizability to specific program structures within specific health care systems.

Conclusions

- Universal newborn hearing screening (UNHS) must be accompanied by appropriate, accessible services for confirmation, audiologic and etiologic diagnoses, and effective hearing and communication development (HCD) options for all children referred through screening programs.
- Early hearing and communication development (EHCD) programs should reflect demographic and cultural factors as well as existing systems, infrastructure and well-developed collaborative linkages with other health care, social support and educational systems.
- A well-designed program will include ongoing evaluation and continuous quality improvement components as well as cost-effectiveness analyses (CEA) and cost-benefit analyses (CBA).

Key References

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Chapter X: Conclusion

Based on formal reviews of published scientific evidence, as well as on expert opinion and consultations with a broad range of stakeholders throughout Canada, the Canadian Working Group on Childhood Hearing (CWGCH) has determined that early hearing and communication development (EHCD) programs incorporating universal newborn hearing screening (UNHS) are feasible and are likely to yield significant overall benefit, relative to traditional methods of identifying permanent hearing impairment in very young children in Canada.

It is necessary, however, to place these inferences in a broad context and perspective. There are geographic variations in demographics, epidemiology and health systems infrastructure. These will affect system performance, many aspects of design and implementation of new programs, and the incremental costs and benefits from such programs.

The evidence reviewed indicates that, while it is possible to develop successful EHCD programs, meticulous attention to all aspects of program design and quality management is necessary in order to achieve substantial net benefits with high cost efficiency. Such programs constitute a chain of events, and the integrity and performance of the overall chain are dominated by its weakest links. The ultimate goal is not merely to screen all babies, but to actually deliver effective services to *all* children and families in need.

The lack of full consensus on the merits of EHCD based on UNHS can be attributed to several factors. First, there has been an emphasis on long-term language outcomes as the primary index of benefit. While such outcomes are important, they are complex and are mediated by a host of variables that are poorly understood. Yet, the target disorder is hearing impairment and, therefore, amelioration of such impairment and reduction of its duration are the most direct, primary health outcomes. Also, there has been relatively little exploration of concomitant, potential benefits from early identification, such as its impact on family communication, decision making and quality of life. Such possible benefits, as yet poorly understood, may underlie evidence that most families endorse early identification.

Second, the pace of developments in this field is rapid, with the result that lengthy clinical trials, and inferences based upon them, may not reflect current performance accurately. This is most likely in the areas of false-positive screening referrals, diagnostic errors and quality of intervention processes. New evidence has come to light, even throughout the period of the deliberations of the CWGCH.

Finally, the rationale for new programs must reflect societal values and ethics, but these have received little attention, despite their importance.

These issues are crucial and must be weighed, along with the available evidence, when considering whether to implement a new EHCD program.

In summary, then, newborn hearing screening leads to early identification of hearing impairment. Such early identification leads to improved hearing and facilitates communication development.

List of Acronyms

AABR — automated auditory brainstem response
ABR — auditory brainstem response
AC — air conduction
AEP — auditory evoked potential
AHEAD II — Advancement of Hearing Assessment Methods and Devices — Immediate Intervention
AN — auditory neuropathy
ANSI — American National Standards Institute
A-O — aural-oral
AOAE — automated otoacoustic emissions
AOM — acute otitis media
AR — acoustic reflex
ASL — American Sign Language
ASSR — Auditory Steady State Responses
AVT — auditory verbal therapy

BC — bone conduction
BM&T — bilateral myringotomy and ventilation tube
BOA — behavioural observation audiometry

CBA — cost-benefit analysis
CEA — cost-effectiveness analysis
CHI — conductive hearing impairment
CM — cochlear microphonic potentials
CMV — cytomegalovirus
CWGCH — Canadian Working Group on Childhood Hearing
cx26 — connexin 26

dBnHL — decibels normal hearing level
DPOAE — distortion product-evoked otoacoustic emissions

ECG — electrocardiogram
EEG — electroencephalogram

EHCD — early hearing and communication development
EHDI — early hearing detection and intervention
EP — evoked potential

FPR — false-positive rate
FS — frequency specific
FS-ABR — frequency-specific auditory brainstem response

HCD — hearing and communication development
HL — hearing level
HR — human resources

ICF — International Classification of Functioning, Disability and Health
ICIDH — International Classification of Impairments, Disabilities and Handicaps
IS — information systems
IT — information technology

JCIH — Joint Committee on Infant Hearing
JLN — Jervell Lange Neilson syndrome

LSQ — Langue des signes quebecois
LVA — large vestibular aqueducts

MEMR — middle ear muscle reflex
MRL — minimum response level
ms — milliseconds

NCHAM — National Center for Hearing Assessment and Management
NCHH — National Campaign for Hearing Health
NCRSP — National Congenital Rubella Surveillance Program
nHL — normal hearing level
NHSP — newborn hearing screening programme
NICU — neonatal intensive care unit
NNS — number needed to screen
NNT — number needed to treat
NPV — negative predictive value

OAE — otoacoustic emissions
OM — otitis media
OME — otitis media with effusion

PCHI — permanent childhood hearing impairment
PEQI — program evaluation and quality improvement
PHAC — Public Health Agency of Canada
PPHN — pulmonary hypertension of the newborn
PPV — positive predictive value

RCTs — randomized controlled trials
REAG — real-ear aided gain
RECD — real-ear to coupler difference
RESR — real-ear saturation response
ROC — relative operating characteristic

SNHI — sensorineural hearing impairment
SPL — sound pressure level

TC — total communication
TEOAE — transient-evoked otoacoustic emissions

UNHS — universal newborn hearing screening
USPSTF — U.S. Preventive Services Task Force

VRA — visual reinforcement audiometry

WBN — well-baby nursery
WHO — World Health Organization