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SPECIAL ARTICLE

Early hearing detection and intervention: 2010 CODEPEH recommendation

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Abstract

Newborn hearing screening is currently performed routinely in many regional health-care systems in Spain. Despite the remarkable expansion in newborn hearing screening since 2000, its feasibility and the benefits of early identification and intervention, many major challenges still remain. In this article, the Committee for the Early Detection of Hearing Loss (Comisión para la Detección Precoz de la Hipoacusia, CODEPEH) updates the recommendations that are considered important for the future development of early hearing detection and intervention (EDHI) systems in the following points: 1) Screening protocols: Separate protocols are recommended for neonatal intensive care units (NICU) and well-infant nurseries. 2) Diagnostic audiology evaluation. Professionals with skills and expertise in evaluating newborn and young infants should provide diagnosis, selection and fitting of amplification devices. 3) Medical evaluation. Risk factors for congenital and acquired hearing loss have been combined in a single list rather than grouped by time of onset. A stepwise diagnostic paradigm is diagnostically more efficient and cost-effective than a simultaneous testing approach. 4) Early intervention and surveillance. All individuals providing services to infants with hearing loss should have specialised training and expertise in the development of audition, speech and language. Regular surveillance should be performed on developmental milestones, auditory skills, parental concerns and middle ear status. 5) Quality control. Data management as part of an integrated system is important to monitor and improve the quality of EDHI services. © 2009 Elsevier España, S.L. All rights reserved.

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PALABRAS CLAVE Sordera; Hipoacusia; Trastornos de la audición

Recomendaciones de la Comisión para la Detección Precoz de la Hipoacusia (CODEPEH) para 2010

Resumen

Actualmente, el cribado auditivo neonatal se lleva a cabo de forma rutinaria en muchos de los sistemas de salud autonómicos en España. A pesar de la importante expansión del cribado de la hipoacusia desde 2000, su viabilidad y los beneficios de la identificación e intervención tempranas, aún existen importantes retos. En este artículo, la CODEPEH actualiza las recomendaciones que se consideran importantes para el futuro desarrollo de los sistemas de detección e intervención precoz en los siguientes puntos: 1. Protocolos de cribado: se recomienda seguir distintos protocolos para los niños ingresados en cuidados intensivos neonatales y los procedentes de maternidad. 2. Evaluación audiológica: se precisa contar con profesionales con experiencia en evaluación de recién nacidos y niños pequeños para completar tanto el diagnóstico como para la selección y adaptación de audioprótesis. 3. Evaluación médica: los factores de riesgo para la hipoacusia neonatal y adquirida se recogen en una única lista en lugar de estar agrupados por el momento de su aparición. Un protocolo de diagnóstico paso a paso es más eficiente y de coste efectivo que efectuar todas las pruebas simultáneamente. 4. Intervención temprana y seguimiento: todos los profesionales que atienden a niños con hipoacusia deberían contar con un entrenamiento especializado y experiencia en la audición, el habla y el lenguaje. Debe realizarse un control periódico del desarrollo de las habilidades auditivas, si existen sospechas paternas y del estado del oído medio. 5. Control de calidad: la gestión de la información como parte integral del sistema es importante para monitorizar y mejorar la calidad del servicio. © 2009 Elsevier España, S.L. Todos los derechos reservados.

Introduction

In recent years, the development of programmes for the early detection of hearing loss has been important in Spain. This allows us to look towards 2010 with optimism, when it is predictable that all regional communities will have launched their own programmes and a decade will have passed since the Commission for the Early Detection of Hearing Loss (CODEPEH) submitted in 2000, before the National Health Council (Ministry of Health and Consumption and regional communities), its first draft protocol for the screening and early diagnosis of childhood deafness.

CODEPEH was established in 1995¹ (CODEPEH 1999) with the objective of promoting neonatal screening for hearing loss in Spain. To this end, and after various initiatives undertaken since its formation, a consensus was reached in April 2003 on the basic and minimum content for the establishment of programmes for early detection of deafness at a national level by the Ministry of Health and Consumption, together with the Regional Communities. In November of the same year, the Minimum Data Registry was approved in the context of the Inter-Territorial Health Board. This collects indicators of coverage, process and outcome of programmes for early detection of deafness.

CODEPEH believes that, at present, the target of screening before the first month, diagnosis at 3 months and treatment at 6 months is being met with varying degrees of success depending on the regions, but generally at higher rates than those reached only 2 or 3 years ago. It should be noted that even communities that have established their programme later are progressing at a rapid pace to obtain similar results to those that have been working for several years^{2,3} (data collected in 2006 in a survey of the health councils of the regional communities and the autonomous cities of Ceuta and Melilla on the percentage of newborns who are subjected to screening, presented at the VI National Meeting of CODEPEH in A Coruña, April 2009).

The experience of the consolidated programmes shows that early detection of hearing loss is possible and that early attention with prosthetic intervention and speech therapy offers children with hearing problems access to oral language at early ages and, consequently, development of learning that depends on it later on (reading, reasoning and understanding), allowing greater possibilities for family, educational and labour integration⁴ in an eminently oral society.^{5,6} Thanks to early identification of hearing problems, the education of these children gives them a level of integration impossible just a few years back.7,8 Spanish programmes with samples greater than 30,000 children and several years of experience are confirming with their data the significant differences that exist between children who receive early attention and those cases in which that care is delayed because of a late identification in a context where there is no universal neonatal screening.9-11

CODEPEH congratulates health administrations for the significant effort being made to expand programmes to all hospitals and for the enthusiasm and effort with which the various professionals are developing them. In addition, CODEPEH believes that the time has come to carry out new recommendations designed to improve the quality of the established programmes and to unify criteria and set standards that provide the maximum uniformity in evaluation of objectives and results.

It is desirable that these new recommendations will encourage the evolution towards standard programmes that, respecting certain diversity, will allow metaanalysis and comparison between them as a method for understanding their effectiveness and efficiency and for correcting deviations from the ultimate goals. Therefore, CODEPEH raises the following recommendations to the Administration, to both health and non health professionals and to families:

Recommendations for screening

There are two internationally accepted tests for the performance of hearing screening: the Transient Evoked Otoacoustic Emissions (TEOAE) and Automated Auditory Brainstem Response (AABR). Both have shown high sensitivity in the early detection of hearing loss and are not mutually exclusive, but rather complementary. Given that the goal of the screening is to detect any type of hearing loss, the use of both tests prevents the occurrence of false negatives. Although the sequential application of the two tests takes very little time, their joint use should be limited to those specific cases that present risk factors for retrocochlear hearing loss or where there is suspicion of auditory neuropathy. Other infants can be analysed with either of the two techniques.^{12,13}

Therefore, infants with no history or risk of retrocochlear hearing loss may be tested in the screening phase by either TEOAE or by AABR. If the AABR screening is not passed, then a second test would not be needed and they could be referred for diagnostic confirmation. However, if the TEOAE are used, especially if performed before the child is older than 72 hours, then the test should be repeated at least once before referral to the diagnostic phase.

In screening programmes based on TEOAE, children who present risk factors for retrocochlear hearing loss must be subjected to a complementary test by AABR or by Auditory Brainstem Evoked Potentials (ABEP) for diagnosis even if they have passed the Otoacoustic Emissions, to avoid false negatives associated with the existence of auditory neuropathy. For the same reason, in infants screened by AABR, the TEOAE should be applied jointly in cases where the first test is not passed, to document the existence of a possible auditory neuropathy. Ongoing monitoring of children is important, even if they have passed the screening in the neonatal period. This monitoring is required within the Healthy Children Programme at health centres, ensuring that the communicative and language development of children is appropriate every 6 months and at least up to age 3. For those who have risk factors associated with hearing loss, the timing and number of auditory re-evaluations should be adapted and individualised depending on the factor identified.14-16 Schooling is associated with a new opportunity to evaluate the communication skills of children within the school health programme, thereby ensuring that any late-onset congenital or acquired audiological disorders will not go undetected and untreated. 17, 18

These auditory controls, which can be carried out with objective or subjective tests adapted to age, should be extended to the entire paediatric stage. At the slightest suspicion of hearing loss, the child should be referred to diagnostic units with expertise in diagnosing hearing loss in children.

Update points:

- 1. Separate protocols are recommended for children from the Neonatal Intensive Care Unit (NICU, level 2-3) and those from maternity. Infants with stays in NICU over 5 days should mandatorily be explored with ABR to prevent a diagnostic error of neural hearing losses. NICU infants who do not pass the AABR test should be referred directly to ENT for reassessment, including ABR and TEOAE, if these were not performed in the screening phase.
- Infants in whom a second test is needed (re-screening) should be evaluated bilaterally even if only one side did not pass the initial test.
- 3. Children who are re-admitted during the first month of life, when combining auditory risk factors (e.g. hyperbilirubinemia with exchange transfusion and sepsis) should repeat auditory screening before being discharged.
- 4. Children with risk factors should be tracked individually according to the probability of late onset of hearing loss. Even if they pass the neonatal test, they should be re-evaluated at least once before age 24-30 months. In children with high risk, this should be done earlier and more frequently. Within this group, those children who present a special suspicion of retrocochlear lesion should be evaluated with ABR, as should children from NICU, regardless of the outcome of the TEOAE.
- 5. All children should be re-evaluated systematically in health controls established by the Healthy Child Programme. There should be an emphasis on the development of spoken language (comprehension and expression), middle ear status and overall development, as well as taking into consideration the suspicions of family, teachers and/ or caregivers.

Recommendations for diagnosis

Diagnostic confirmation of children who have not passed the newborn screening must be completed in the third month of age to enable early audiological diagnosis. This diagnosis should be based on a set of tests, which should always include ABR, tympanometry, stapedial reflex (using 1,000 Hz as sound carrier) and TEOAE, repeated at least 2 times with a difference of one to 4 weeks.

The first contact can be used to give parents advice for conducting conditioning to sound stimuli tests at home with the child, as well as exercises to facilitate audiometry through visual reinforcement, which should be feasible around the sixth month of life. At that age, the child is often also able to respond to the test of Ling and the "name" test.

Audiological diagnosis should not be delayed beyond the fourth to fifth month, to allow early initiation of care, which should always consider speech therapy intervention and the prosthetic adaptation necessary in each case before 6 months.

Aetiological diagnosis should be carried out simultaneously with audiometric assessment without delaying early child stimulation due to not having completed it. It is important to have a multidisciplinary team (otolaryngologist, paediatrician, paediatric neurologist and geneticist) and to carry out laboratory tests including imaging, genetic and any other tests deemed appropriate in light of each case. Every boy and girl with the confirmed diagnosis of hearing loss must pass at least one eye examination. We must evaluate if the hearing loss is isolated or integrated in a syndrome or if it is a disorder associated with certain diseases or neurological or neuropsychological deficits.¹⁹

To date, no consensus has been reached allowing a complete diagnostic protocol to be followed, once the presence of hearing loss in a newborn is confirmed, as identified by the screening programme.²⁰

The aetiology of congenital sensorineural hearing loss can often be discovered by anamnesis, as acquired environmental causes (such as intrauterine infections. ototoxic medication, metabolic disorders, substance abuse, prematurity, perinatal hypoxia or anoxia or exposure to teratogens) are identified in 35% of cases. 21,22 Physical and neurological examinations can add information about associated malformations and syndromes.²³ However, hereditary non-syndromic sensorineural hearing loss is difficult to diagnose through only the history, and clinical examination must be completed by diagnostic tests that have not yet been protocolised. The tests to be performed according to the clinical history (personal and family) and physical and neurological examinations and audiological tests are genetic tests, 24-26 radiological tests, 27 analytical determinations and "other complementary tests" (ECG, electroretinography and electrophoresis).

To increase diagnostic accuracy and minimize parental stress, molecular study of the *GJB2* gene has been proposed as the first step of the process to be followed in cases of sensorineural hearing loss in which no aetiology is identified in the medical history or upon physical examination.²⁸ The diagnostic performance of this test was of 22% in a population of children with severe or profound sensorineural hearing loss. If the mutation is found in only one gene, then the search should be performed in serial combination with the search for the GJB6 gene deletion. In the Spanish population, we observe that a significant number of cases in which there is a mutation in *GJB2* and no other is found (appearing as "healthy" heterozygotes and thus that the cause for hearing loss is other than genetic), are compound mutants of *GJB2/GJB6*.²⁹

Imaging tests, particularly computed tomography (CT) of the petrosal region, show alterations in approximately 30% of cases.³⁰ It has been shown statistically that the presence of mutations in the *GJB2* gene makes it unlikely to find these changes, so omitting this test is recommended, considering the expense and disruption that performing a CT scan causes for families and children. For the same reason, the *GJB2* genetic testing should also be omitted for those children shown to be affected by the CT, if this had been requested as the first study.

Test results have a very low diagnostic outcome, not contributing to the identification of the aetiology in any of the 150 children in the study, a fact already documented in other studies^{31,32} that have established that the routine request of many laboratory tests has become irrelevant. Although the electrocardiogram has very reduced diagnostic performance, its request is recommended in all cases with severe or profound hearing loss to exclude the prolonged Q-T interval associated with Jervell-Lange-Nielsen syndrome; it could thus save lives.³³

Although it could be thought that the determination of thyroid hormones might be of interest to exclude Pendred syndrome (sensorineural hearing loss with goitre), it is known that 56% of children with this syndrome are euthyroid³⁴ and that the perchlorate discharge test is the test of choice when the syndrome is suspected. Therefore, there is no need to use it as screening for all children with hearing loss. Moreover, it necessary to rule out hearing loss in all hypothyroid children.

Carrying out the diagnosis of childhood hearing loss in an orderly way and step by step is more efficient and cost effective than requesting all the available evidence indiscriminately.

Children with severe or profound hearing loss should be evaluated genetically as a first step. In contrast, children with mild or moderate deafness have to undergo a CT scan first. Those who have genetic disorders should not undergo imaging, which saves the trouble and expense involved. Figure shows the recommended diagnostic algorithm^{35,20} for the systematic and orderly study of congenital hearing loss.

Update points:

- 1. Besides the audiological evaluation of children with hearing loss, there should be the ability to prescribe the fitting of hearing aids, if indicated.
- 2. To confirm a permanent hearing loss in children under 3 years old, it is necessary to have carried out at least one ABR test.
- 3. The re-evaluation of hearing in children with risk factors has to be programmed individually, so as to adapt to each case according to the likelihood of late-developing hearing loss. Children with risk factors who have passed the screening must be audiologically re-evaluated before 24-30 months of age. Children with cytomegalovirus infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma or infection associated with hearing loss or children who have undergone chemotherapy or extracorporeal oxygenation should be assessed earlier and more often and also when there is suspicion of hearing loss by parents or a family history of deafness.
- 4. When adaptation of prosthetics is indicated by the otolaryngologist, this should take place no later than one month and, likewise, speech therapy should be initiated early. Without this, early diagnosis is sterile and prosthetic adaptation is insufficient.⁴
- 5. Families of children with hearing loss should be offered the option of requesting a genetic consultation.
- 6. Every child diagnosed with hearing loss should be assessed at least once by an ophthalmologist.
- 7. Updated risk factors for congenital and acquired hearing loss should be listed in a single list and not by age of onset as has been done so far (Table).

Recommendations for treatment and follow-up^{36,37}

With regards to the confirmation of the existence of hearing loss, any necessary prosthetic fitting (hearing

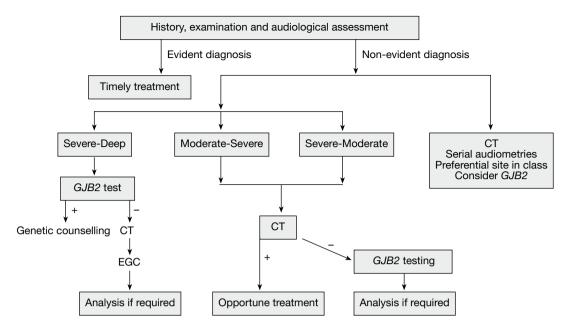


Figure Algorithm for the evaluation and treatment of childhood sensorineural hearing loss. CT: computed tomography; ECG: electrocardiogram.

aids and implants) or speech therapy should be carried out.

At the same time, parents must provided with specialised care to enable them, first, to have the information necessary to make decisions regarding the deafness of their child and, second, to play the critical role they have in the process of (re)habilitation. It is within the family that communication and oral language acquisition of children begin and are developed and where they will forge their full and autonomous future life.³⁸ The intervention, therefore, should be focused on the family, which must be supported by a well coordinated multidisciplinary team that carries out the surgical, hearing-aid, speech and education, treatment appropriate to the condition and age of the child.

The period between confirmation of hearing loss before 3 months of age and schooling at age 3 is critical for ensuring access for children with hearing loss to auditory stimulation and oral language needed to provide the cognitive tools and instruments that will help them to participate in schooling on an equal footing with the rest of the students. This objective is achieved with adequate and early fitting of prosthetics without waiting for more than one month from diagnosis, combined with effective early care provided by skilled professionals, duly certified and gualified. It is imperative that the government should provide sufficient financial coverage, which allows families to have both professional and appropriate hearing-aid benefits to ensure care that is adequate, sufficient and specialised enough for the child and family, regardless of the parental sociocultural and economic capacity. This stage is key to the future development of children if it is intended that they become autonomous, independent adults integrated into society. 19,38

Proper prosthetic adaptation, performed by audioprosthetists with accredited degrees, and continued use of hearing aids allows the use of the child's residual hearing.³⁹ In cases where it is found that the correct prosthetic adaptation and early attention do not achieve the expected results between 3 and 6 months, then the inclusion of children in the cochlear implant programme must be considered.

If tests carried out are consistent with auditory neuropathy, it is advisable to conduct a genetic study to investigate the existence of a disease related with the otoferlin gene.⁴⁰ If the child is a carrier of this genetic alteration, he/she is a candidate for cochlear implant, which can be carried out once this alteration is confirmed. However, in other neuropathies, especially those caused by hyperbilirubinemia, the attitude must be expectant and with early speech therapy stimulation. In any case, cochlear implants should be delayed until the lack of response from the child is clear.

Update points:

- 1. The government should provide all children with any degree of unilateral or bilateral permanent hearing loss with the adequate prosthetic adaptation and the necessary early speech therapy. Furthermore, the administrations must provide the necessary resources for the guidance and support of their families.
- 2. Early intervention services (speech therapy and hearing aids) must be provided by staff with appropriate qualifications and sufficient experience in infant hearing loss.
- 3. It is recommended that all children (even if they do pass neonatal screening) be checked at the Health Centre by

Table Update of the risk factors for infant hearing loss adapted from JCIH 200742

- 1) Suspicion by the caregiver about delays in speech development and abnormal hearing
- 2) Family history of permanent childhood hearing loss
- 3) Stay in neonatal intensive care for more than 5 days, including readmissions to the init within the first month of life
- 4) Having undergone extracorporeal membrane oxygenation, assisted ventilation, ototoxic antibiotics, loop diuretics (furosemide). Hyperbilirubinemia requiring exchange transfusion.
- 5) Intrauterine infections of the TORCH group (cytomegalovirus, herpes, rubella, syphilis and toxoplasmosis)
- 6) Oraniofacial anomalies including the ear, ear canal, appendixes or preauricular pits, cleft lip or sunken palate and abnormalities of the temporal bone and asymmetry or hypoplasia of the facial structures
- 7) Findings related to physical syndromes associated with sensorineural or transmission hearing losses, such as a patch of white hair, heterochromia iridis, hypertelorism, telecanthus or abnormal skin pigmentation
- 8) Syndromes associated with hearing loss or progressive or late-onset hearing loss such as neurofibromatosis, osteopetrosis and the syndromes of Usher, Waardenburg, Alport, Pendred, Jervell and Lange-Nielsen, among others
- 9) Neurodegenerative diseases such as Hunter syndrome and sensory-motor neuropathies such as Friedrich's ataxia and Charcot-Marie-Tooth disease
- 10) Postnatal infections with positive cultures associated with hearing loss, among which are included viral meningitis (especially chickenpox and herpes) and bacterial meningitis (particularly Hib and pneumococcal)
- 11) Craniofacial trauma, particularly fractures of the temporal bone and skull base requiring hospitalisation
- 12) Chemotherapy
- 13) Endocrine diseases. Hypothyroidism

the paediatrician for developmental milestones, auditory skills, questions arising from parents and the condition of the middle ear. At this level of assistance, it should be possible to perform a standard hearing screening test with a validated and objective test at 9, 18, 24, and 30 months of age or at any other time if there are suspicions of hearing loss.

- 4. IChildren who do not pass the language tests of the global screening conducted within the Health Centre or those for whom there is suspicion about their audition should be referred for full audiological evaluation and assessment of speech and language, following the indications of the Guide for comprehensive assessment of children with hearing impairment developed by the Spanish Committee of Audiophonology (CEAF).¹⁹
- 5. IPeferral pathways should be established to prevent the peregrination of families among the various professionals involved in caring for their child with hearing loss and to ensure the necessary coordination for the proper interprofessional and comprehensive care required by people with hearing loss and their families.⁴¹

Recommendations for quality control of the programme

It is important that the programme for early detection of hearing loss is a public health programme regularly monitored to determine whether it meets the appropriate quality criteria. These quality criteria should ensure that this initiative will enable the detection and treatment of hearing impaired children, who will need constant vigilance to provide the best medical, educational and social care. This makes it necessary to establish quality parameters that clearly define its objectives and that have been defined by the CODEPEH.⁴² The general objectives of the programmes have to be conducting the screening before the first month of life to obtain diagnostic confirmation before 3 months and starting treatment before 6 months. To achieve this, it would be necessary to meet the parameters below, collected from the JCIH⁴³ and amended by the CODEPEH.

- The programme aims to discover all unilateral or bilateral hearing loss present at birth, regardless of severity and aetiology. As quality criteria, screening procedures must ensure that false negatives tend towards 0%^(a).
- Quality indicators of universal screening(^b): both ears of all children born in the Regional Community will be explored.
 - a) To be universal, screened children must be more than 95% of newborns.
 - b) To be neonatal, the first test should be performed before the first month of life on more than 95% of children.
 - c) Pe-screening(°): all children who did not pass the first test will be explored again. The objective should be 100% of the children referred to second or third test, but it will be considered satisfied by 95%.
- *3.* Quality indicators for diagnostic confirmation: all children referred from the screening phase will be explored.
 - a) It should not exceed 4% of the rate of referral to confirmatory testing.
 - b) The aim should be 100% of children with confirmed diagnosis carried out during the third month of life, but it will be considered satisfied if it exceeds 90% in the third month.

- 4. Quality indicators for treatment:
 - a) The establishment of adequate early care before 6 months of age should aim toward 100% of the children with confirmed diagnosis of hearing loss, but will be considered satisfied if it exceeds 90%.
 - b) If prosthetic adaptation is decided, no more than one month should pass between the indication and adaptation in 95% of candidates.
 - c) For boys/ girls with late-onset congenital or acquired hearing loss, 95%should have started treatment within 45 days after diagnosis.
 - d) The percent age of children with permanent hearing loss who have received a development control (cognitive and linguistic) before 12 months should be 90%
- 5. Quality criteria for the monitoring of the programme:
 - a) Epidemiology: computer registry of all children and the results of the different phases. It is advisable to have data on:
 - 1. Number of newborns screened before leaving the hospital.
 - 2. Number of children with confirmed hearing loss before 3 months of age.
 - 3. Number of infants enrolled in a programme of early intervention before 6 months of age.
 - Number of children with suspected or confirmed hearing loss who are referred to an Infant Hearing Loss Unit.
 - Number of children with non-syndromic hearing loss who have adequate development of language and communication skills at the beginning of school age.
 - 6. Number of children referred for cochlear implant programme.
 - b) Clinical: Control of all children identified in ENT consultation.

Final considerations and operational proposals

Clearly there are significant challenges and barriers in the system to be overcome in the next few years. Therefore, as final considerations that also attempt to offer a sequence of operational proposals to successfully achieve the ultimate goal of screening, early diagnosis and care and early speech therapy intervention, the committee recommends an increased focus on the following aspects:

• Strengthen the implementation of universal screening throughout the State, giving effect to the agreement

reached between the Ministry of Health and the regional communities and within the Inter-Territorial Board of Health in 2003, with the approval of the consensus for the implementation of the Programme for Early Detection of Hearing Loss.

- Establish the necessary procedures and resources for effective monitoring of programmes for early detection of hearing loss, according to the commitment made by the Ministry of Health and the health administrations in 2003.
- Confirm in the shortest possible time the "no pass" of newborns not passing the screening, in accordance with the established protocol and the various phases of the programme.
- Emphasise the need to establish "diagnosis units for infant hearing loss" to which children who are suspected of hearing loss can be derived and which will have trained personnel and adequate equipment for infant diagnosis.
- Appoint a medical chief for the hearing loss screening programme in each hospital in which the screening programme is being carried out.
- Train nurses specifically to perform tests included in the screening programme and also in the management of newborns with safety.
- Ensure that the re-screening phase is also conducted by experienced staff and in adequate premises, especially with regard to its soundproofing.
- Establish specific procedures for cooperation between centres and outpatient recovery mechanisms for infants who are lost without undergoing screening (births at home or in a different country, regional community or hospital; readmissions in NICU with auditory risk pathology, etc.) for referral centres that conduct testing and diagnosis.
- Enhance the key role of primary care paediatricians in monitoring correct auditory development, as well as communication skills and oral language development of all children, in addition to their role in identifying risk factors in paediatric cases.
- Ensure adequate multidisciplinary treatment that addresses both the medical needs of and the support and guidance for children with hearing loss and their families. All professionals involved in diagnosis and treatment will therefore be coordinated for the effective exchange of information and adequate follow-up of child development and progress, as well as for analysis and decision-making in relation to the different strategies of intervention, speech therapy and prostheses.
- Designate an individual responsible for the programme for early detection of hearing loss in each regional community. This person must be a physician with experience in audiological screening, diagnosis and treatment of childhood hearing loss and who has the appropriate administrative means to carry out the supervisory work of the various phases of the programme and who is also responsible for coordinating the multi-professional teams.
- Guarantee early care to ensure the joint responsibility of health, education and social services administrations and the necessary inter-administrative coordination. Once in school, the educational system has to provide the care and support resources that are most adequate and appropriate to each individual case.

^aFalse negative: child under 3 years who passes the screening programme with normal result and appears with hearing loss and lack of oral speech and for whom no underlying cause is found that justifies it (congenital postneonatal-onset hearing loss or acquired hearing loss).

^bScope of the programme: percentage of children studied in relation to the number of children who are offered the programme. Being a universal programme, it should be offered to all children.

^cRe-screening: a second or even third screening test can be considered before referral to diagnosis.

- Adopt legal measures so that auditory prosthetics (implants and hearing aids) are financed entirely by the National Health System and incorporated into the Portfolio of Ortoprosthetics Services with no age limit, regardless of whether they are implantable or not. Likewise, there should be adequate coverage provided to ensure maintenance and update.
- Plan long term cost-benefit studies promoted by the Ministry of Health and the health administrations, to assess the quality and effectiveness of the implementation of programmes for early detection of hearing loss, early prosthetic adaptation, speech therapy and intervention, as well as the adaptation to the quality standards established for each of these areas.

We must remember that the ultimate goal of all screening and early treatment of congenital hearing loss initiatives is: "optimising communication and social, academic and professional development for every child with permanent hearing loss"⁴² and "facilitating early and natural access to spoken language through hearing and thus making use of brain plasticity in the early years of life to stimulate communication and language development in children."⁴³

Conflict of interests

The authors declare no conflict of interests.

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