Special supplements

FIAPAS

Number 180 / 2023

CODEPEH
COMMISSION FOR EARLY DETECTION OF HEARING LOSS

Universal newborn hearing screening. Clinical problems and frequently asked questions: CODEPEH recommendations 2022

Cribado auditivo neonatal universal. Problemas clínicos y preguntas frecuentes: recomendaciones CODEPEH 2022

Author: CODEPEH

(Faustino Núñez, Carmen Jáudenes, José Miguel Sequí, Ana Vivanco, José Zubicaray)

To cite this article:

Núñez, *F. et al.* (2022). Cribado auditivo universal. Problemas clínicos y preguntas frecuentes: recomendaciones CODEPEH 2022. *Revista FIAPAS,* (180).

GOBIERNO DE ESPAÑA
MINISTERIO DE DERECHOS SOCIALES Y AGENDA 2030
SECRETARÍA DE ESTADO DE DERECHOS SOCIALES

GOVERNMENT OF SPAIN
MINISTRY OF SOCIAL SERVICES AND 2030 AGENDA
SECRETARY OF STATE FOR SOCIAL SERVICES

Fundación ONCE
ONCE Foundation

This CODEPEH Recommendations Document 2022 has been prepared within the framework of the project on *Universal Newborn Hearing Screening. Clinical Problems and Frequently Asked Questions,* developed by the Spanish Confederation of Families of Deaf People-FIAPAS, in collaboration with the Commission for the Early Detection of Infant Hearing Loss-CODEPEH, with the co-organisation of the Royal Board on Disability.

The following are members of the CODEPEH:

Dr Faustino Núñez-Batalla, Chairperson
ENT Service, Hospital Universitario Central de Asturias-Oviedo
Representing the Spanish Society of Otorhinolaryngology

Ms Carmen Jáudenes Casaubón, Member
Director of FIAPAS
Representing the Spanish Confederation of Families of Deaf People

Dr José Miguel Sequí Canet, Member
Head of the Paediatrics Service, Hospital Universitario de Gandía-Valencia
Representing the Spanish Association of Paediatrics

Dr Ana Vivanco Allende, Member
Paediatric Clinical Management Area, Hospital Universitario Central de Asturias-Oviedo
Representing the Spanish Association of Paediatrics

Dr José Zubicaray Ugarteche, Member
Paediatric ENT Service, Complejo Hospitalario de Navarra-Pamplona
Representing the Spanish Society of Otorhinolaryngology

GOBIERNO DE ESPAÑA
MINISTERIO DE DERECHOS SOCIALES Y AGENDA 2030
GOVERNMENT OF SPAIN
MINISTRY OF SOCIAL SERVICES AND 2030 AGENDA

REAL PATRONATO SOBRE DISCAPACIDAD
ROYAL BOARD ON DISABILITY

FIAPAS
THE SPANISH CONFEDERATION OF FAMILIES OF DEAF PEOPLE (FIAPAS)
Fomentando Inclusión. Apoyando Personas. Avanzando Solidariamente.
(Fostering Inclusion Supporting People Progressing Together)

ABSTRACT

The quality and effectiveness of newborn screening programmes for hearing loss have improved considerably since their implementation almost two decades ago, thanks to the incorporation of technological advances and progress in knowledge about the screening process, diagnosis and early intervention. However, there are still unknowns on different aspects of and lack of training in the causes of congenital hearing loss and the pathways and referral destinations for cases that do not pass screening.

This document of the CODEPEH analyses the current situation and aims to provide answers, from the most recent scientific evidence, on how to recruit newborns for inclusion in the screening process, what information to provide the family prior to the test, how to improve the competences and performance of the team assigned to the programme, in addition to reviewing the advantages, disadvantages and limitations of the technologies available to perform the testing. It details the process of the screening, including how to document and report the results to families. It also addresses the loss of cases during the process and how to mitigate this, and highlights the importance of a well-structured information system to assess the quality and effectiveness of the programme.

KEYWORDS

Newborn hearing screening; health professionals; otoacoustic emissions; auditory evoked potentials; losses during the process; information for families.

RESUMEN

La calidad y efectividad de los programas de cribado neonatal de la hipoacusia ha mejorado considerablemente desde su implantación hace casi dos décadas, incorporando avances tecnológicos y progresando en el conocimiento sobre el proceso de cribado, el diagnóstico y la intervención precoz. Pero existen lagunas sobre diferentes aspectos y falta formación sobre las causas de hipoacusia congénita y los itinerarios y destinos de derivación de los casos que no superan el cribado.

Este documento de la CODEPEH analiza y pretende dar respuesta, desde la evidencia científica más reciente, a cómo efectuar la captación de los recién nacidos para su inclusión en el proceso de cribado, qué información proporcionar a la familia previa a la prueba, cómo mejorar las competencias y funcionamiento del equipo adscrito al programa, además de revisar ventajas, inconvenientes y limitaciones de las tecnologías disponibles para realizar la prueba. Se detalla el desarrollo del cribado, incluyendo cómo documentar y comunicar los resultados a las familias. Asimismo, se aborda la pérdida de casos en el proceso y cómo paliarla, y se resalta la importancia de un sistema de información bien estructurado para evaluar la calidad y efectividad del programa.

PALABRAS CLAVE

Cribado auditivo neonatal; profesionales de la salud; otoemisiones acústicas; potenciales evocados auditivos; pérdidas en el proceso; información familias.

CONTENTS

1. INTRODUCTION

2. BEFORE SCREENING

2.1. NEWBORN RECRUITMENT
2.2. CONSENT AND INFORMATION FOR FAMILIES

3. DURING SCREENING

3.1. PERSONNEL

3.1.1. COORDINATION OF THE SCREENING PROGRAMME
3.1.2. PROCESS EXECUTION

3.2. TECHNOLOGIES

3.3. PROCEDURE

3.3.1. SCREENING
3.3.2. RE-SCREENING
3.3.3. DOCUMENTATION AND REPORTING OF RESULTS

4. AFTER SCREENING

4.1. LOSSES IN THE PROCESS
4.2. INFORMATION SYSTEMS

4.2.1. QUALITY CONTROL

5. INVOLVEMENT OF FAMILIES

6. CODEPEH RECOMMENDATIONS 2022

7. FIGURE (Action algorithm)

8. REFERENCES

1. INTRODUCTION

The success of newborn screening programmes for congenital hearing loss depends on early identification and early diagnosis and treatment of children with hearing loss by a multidisciplinary team of professionals, whose technical knowledge, execution of the protocol, and ability to provide appropriate clinical procedures are crucial.

The quality and effectiveness of hearing loss screening programmes have improved considerably since their implementation almost two decades ago, thanks to the incorporation of technological advances and progress in the knowledge about the screening process, diagnosis and therapeutic intervention in childhood hearing loss (Ravi *et al.,* 2017).

Therefore, the programmes have evolved and adapted to the emergence of new technologies and to advances in the knowledge and understanding of childhood hearing loss, which has required changes in screening protocols. Also, as a result of the implementation of the programmes, deficiencies have been identified that may compromise the aims of said programmes. Studying and rectifying them is a continued obligation of the staff involved.

Since many professionals are aware of the need to acquire and update their knowledge (Ravi *et al.,* 2018; Danhauer *et al.,* 2006), it is important to promote and provide information to ensure ongoing training and refreshers on the foundations for early detection of congenital hearing loss, particularly as regards the details of the screening process and how it works (Moeller *et al.,* 2006a). There is also, on occasion, a lack of information on the causes of congenital hearing loss and its risk factors (Moeller *et al.,* 2006b), as well as on pathways and referral destinations for cases that fail screening or require diagnostic confirmation and treatment (Arnold *et al.,* 2006a).

It is important to know the roles of the personnel, the specific training they must have and the skills they must possess; the technologies available to perform the test: otoacoustic emissions (OAS) and automated brainstem auditory evoked potentials (BAEPs), both currently in use, with their respective advantages, disadvantages and limitations; as well as the procedures for documenting and reporting their results.

Other issues, such as the way in which newborn recruitment should be performed, together with the information that must be provided to the family to obtain the consent for the test (a compulsory procedure), are vital to ensuring participation and compliance with the applicable regulations.

The significant and concerning problem of cases lost during the programmes' process has recently been highlighted, as it can seriously compromise their effectiveness. To provide measures aimed at mitigating the percentage of children who do not attend the appointment for the follow-up test indicated by the protocol, to find out the reasons why, emphasising the importance of a well-structured information system, which also serves to assess the quality and effectiveness of the programme.

The need to keep families informed throughout all steps of the screening protocol must be addressed as a priority because they are, together with the newborn, at the core of all programme actions.

This paper addresses the update on clinical problems and the most frequently asked questions related to universal newborn hearing screening, with special emphasis on the aspects that need to be brought up to date. In this regard, CODEPEH has established the pertinent recommendations.

The information needs of families throughout all steps of the screening protocol must be addressed as a priority

2. BEFORE SCREENING

Different studies indicate that newborn disease screening is very stressful for families, given the potential for a disease being detected in their newborn children. The information provided to families is essential and must be adapted to reduce their stress when subjecting their baby to the various examinations and tests, to be able to work with them (DeLuca *et al.,* 2011).

The aim of the newborn screening programmes for hearing loss is to confirm that the infant is hearing correctly and, if not, to provide the diagnostic means and treatments necessary to prevent or minimise the sequelae of hearing loss.

2.1. Newborn recruitment

Most births in Spain occur in the hospital setting. However, there is a segment of the population that may be interested in delivery in the family home, most often with medical care, but there are circumstances, particularly in marginalised populations, in which pregnant women do not seek healthcare or follow-up during pregnancy or delivery. According to the Spanish National Statistics Institute, INE, only 0.32% of all deliveries are performed outside the hospital setting in Spain, a figure that has decreased since 1980 when this percentage was 16.9%. There is also a growing minority of people who reject all types of preventive action in children, and adequate decision-making and information is critical in these cases (Riaño *et al.,* 2013).

Therefore, an important time to start the information process regarding care, monitoring and tests to be performed for the newborn are the visits that the mother-to-be has with her midwife, where she will receive plentiful information about pregnancy, childbirth and the perinatal period on a regular basis. This initial contact and information makes it easier to recruit babies for the hearing health examination, tests and follow-up. The Public Health Services of each autonomous community are responsible for providing these professionals with adequate training to fulfil this task.

The paediatrician at the health centre must ensure that the required neonatal tests have been performed and, if not, inform the family in order to resume the screening process

Another important time for recruitment and inclusion in the programme and review of adequate hearing screening is during visits to the paediatrician at the health centre, because the latter must ensure the mandatory newborn tests have been performed (including hearing screening) and, if not, inform the family in order to resume the screening process (Núñez *et al.,* 2019).

This entire recruitment process should be managed by the persons responsible for the newborn hearing screening programme in the autonomous community, who will ensure adequate follow-up of each case at all times.

2.2. Consent and information for families

Spanish legislation establishes, in *Ley 41/2002, de 14 de noviembre, básica reguladora de la autonomía del paciente y de derechos y obligaciones en materia de información y documentación clínica,* [the Patient Autonomy and Rights and Obligations regarding Clinical Information and Documentation Act] the principles and reasons for exercising the right and obligation of informed consent to the patient or his/her family. Specifically, Article 4.1 (Right to healthcare information) states: *Patients have the right to know, as a result of any action in the field of their health, all information available about them, except for the cases excepted by law. In addition, everyone has the right to have their wishes to not be informed respected. Information, which as a general rule will be provided verbally and recorded in the clinical history, includes, at a minimum, the purpose and nature of each intervention, its risks and its consequences.*

Although the newborn hearing screening procedure is non-invasive, informed consent is required, given the importance of the diagnosis if the child does not pass the test: not only because of the emotional impact on the family at the time of reporting, but because it will lead to a battery of tests, studies and treatments over time.

In newborn screening programmes, both of the parents, or the legal guardians, must consent or reject the screening procedure. Consent should always be given verbally and in writing to the extent possible.

The mothers' condition at the time of delivery may vary greatly. Sometimes the mother cannot be spoken with due to health problems or impending delivery. Or, even when access to the mother is possible, given the circumstances, it is not always an appropriate time to carry out this information and consent procedure. The recommended time for information and consent is therefore the prenatal period, either provided by the midwife or during birthing preparation courses, though it would be advisable for someone from the hearing screening programme itself to perform it (American Academy of Pediatrics, 2000; Queiro *et al.,* 2007).

In any case, it must be ensured that, before performing the test on the newborn, the person who is going to perform it provides the information to the family and checks that he/she has the consent to perform it. Consent must be documented in the clinical history.

The information provided must be truthful, specific and complete, provided in a language that is understandable and not alarmist.

If the parents, or the legal representatives, of the newborn refuse to have the screening performed, it is important that this informed dissent be reflected in writing. If they do not wish to sign it, this must be recorded, for legal reasons, in the clinical history. Dissenting in this way can help to give room for further reflection on the decision (Ministry of Health. Working Group on Newborn Hearing Screening of the Population Screening Presentation, 2021).

Although there are various ways to grant consent, the minimum data it should contain are (Arnold *et al.,* 2006b):

- Newborn's information. Place and site of performance of screening.
- Brief explanation of the test and the other steps to follow if the baby fails the first phase.
- Declaration of the person providing the consent, stating that he/she has understood the information and is aware that the consent can be withdrawn at any time and without stating the reason.
- Name of the person leading the consent discussion. Name of the person consenting (parent or legal representative). Signature and date.
- Name of the person dissenting or withdrawing consent (parent or legal representative). Signature and date.

3. DURING SCREENING

3.1. Personnel

3.1.1. Coordination of the Screening Programme

The coordination of the Screening Programme is essential. For this, there must be a professional figure with experience in the management of newborn hearing screening and knowledge about the functioning of the technology required for the implementation of the programme, as well as with competence and capacity to assume the responsibilities required in performing this responsibility.

The functions to be assumed include:

- Being responsible for the equipment, personnel and protocol to be followed.
- Ensuring that each new professional who joins the team has received appropriate training according to established procedures and standards of patient care.
- Plan the agendas of the personnel performing the screening to ensure coverage 365 days of the year.
- Schedule ongoing staff training when necessary, and take steps to correct, improve and maintain the performance of the programme.
- Monitor the documentation of the results in accordance with the provisions of the hospital and/or health administration.
- Check the availability of consumables.
- Perform monthly monitoring of the screening programme database to check that information has been entered for each child screened.
- Coordinate follow-up and consultation of children requiring follow-up.
- Inform the relevant regulatory authorities in accordance with the applicable regulations.

There must be a professional figure for coordinating the programme with experience in managing screening and knowledge about the technology, as well as the competence and capacity to assume the required responsibilities

3.1.2. Process execution

To perform this function, nursing staff is used in Spain. There is case-law in this regard to grant this role to these professionals.

In any case, the personnel responsible for the screening should:

- Comply with all requirements determined by the hospital regarding this competence and responsibility, with specific training.
- Be able to work autonomously and demonstrate the skills required to perform tasks such as: following the sequence of instructions in the screening protocol, correctly managing the screening equipment, or the skill required to manage a newborn and to place the probe in the newborn's ear.
- Be an active part of the multidisciplinary team, being able to interact and exchange information with the rest of its components.

In order to ensure that the personnel in charge of screening are competent at all times to perform their functions correctly, protocols should be developed and implemented for their training and

for the verification of the skills of newly recruited personnel in the programme (Joint Committee on Infant Hearing, 2019). To do this:

- A continued training plan must be in place for all programme staff.
- According to the manufacturer of the equipment used as the screening technology, the physician responsible for the programme must provide training to the personnel for its proper use.
- The programme physician should provide training to the screening personnel in order to optimise the status of the newborn at the time of testing.

Screening should examine both ears, because unilateral congenital deafness must also be detected and diagnosed early to be treated

The audiological diagnosis of children is the exclusive competence of professionals with specific skills, knowledge and access to all the equipment necessary to carry out the audiological diagnosis of babies only months old. When the necessary experience and equipment is not available, it is imperative to refer the child to those centres that can undertake this important task, which is the basis for accurate diagnosis and ensuring access to early care services without delays.

The establishment of a network of professionals that includes personnel from maternity wards, paediatric units, ear, nose and throat (ENT) services, under the direction of public health services, is mandatory for the development and practical application of a successful hearing screening programme. In addition to meetings of a multi-professional network led by health authorities to discuss quality issues as part of the screening programme, screening workshops to exchange experiences and for training purposes are another key component (Joint Committee on Infant Hearing, 2019; Holzinger *et al.,* 2021).

3.2. Technologies

When defining the primary characteristics of universal newborn screening for hearing loss, it should be taken into account that the child will not collaborate and that a large population will be included. Therefore, procedures that are sensitive, specific and objective should be chosen, but that, given the number and characteristics of the population to be screened, mostly healthy newborns, these procedures must also be non-traumatic, simple, repeatable, rapid and inexpensive. It should be noted that the purpose of screening is not a firm diagnosis, but rather the identification of newborns suspected of having hearing loss to focus subsequent efforts for diagnostic confirmation (Bussé *et al.* 2021a; Bussé *et al.,* 2021b; Mackey *et al.,* 2021).

Universal newborn hearing screening with Automated Brainstem Auditory Evoked Potentials (BAEPs) has some major advantages, but also some disadvantages when compared with the use of Evoked Otoacoustic Emissions (OAEs).

When selecting one technology or another, the different conditioning factors adequately reflected in the literature must be taken into account (Johnson *et al.,* 2005).

Changes in tympanic membrane movement greatly affect screening with OAEs, which requires a normal middle ear, but have very little effect on screening performed with BAEPs. A Doyle study reported that in children with decreased mobility of the tympanic membrane, the percentage with normal tests ranged from 33% if we use OAEs to 95% using BAEPs (Doyle *et al.,* 2000).

False positives in the first three days of the child's life are more common when using OAEs than when screening with BAEPs. This is probably also influenced by the accumulation of vernix in the external auditory canal or amniotic fluid in the middle ear.

Since the failure rate using OAEs is higher than if BAEPs is used, the need for re-screening is higher if the former technology is used, which increases the likelihood of loss to follow-up. If we screen in a single phase with BAEPs, we would reduce this loss of cases during the process (Benito-Orejas *et al.,* 2008).

As regards the time taken to perform the test, screening with BAEPs was increased (between 4 and 15 minutes, as compared to 2-5 minutes with OAEs). Thus, the time for performing BAEPs implies a significant increase in the expense related to personnel costs associated with this type of screening. The higher cost of consumables using BAEPs should also be added to this.

However, the total cost may be similar in the long term if BAEPs are considered to account for fewer children sent to the diagnostic stage. Referral of children with

risk factors, screened with OAEs, to testing with BAEPs is also avoided. This reduces the number of screening phases.

BAEPs are the ideal test for diagnosing retrocochlear pathology such as auditory neuropathy. Children at risk of developing this include those with risk factors for hearing loss due to their stay in the neonatal ICU for more than 5 days. BAEPs would therefore be the ideal method for screening in neonatal units, where most children with risk factors for retrocochlear diseases are admitted. If OAE screening is performed, the number of false negatives (children who pass screening and, however, have hearing loss) could be increased because auditory neuropathy is underdiagnosed (Berg *et al.,* 2005). The incidence of auditory neuropathy in healthy babies is very low, 6-30/10,000 births. Less than 1% of infants in the maternity ward had a fail with BAEPs then sent to OAEs, and none showed this pattern in outpatients, so it is specifically acceptable to re-examine with OAE after failing with BAEPs, with the warning that a baby with auditory neuropathy would not be diagnosed with this protocol (Joint Committee on Infant Hearing, 2019).

3.3. Procedure

As discussed above, universal newborn screening presents some challenges in the organisation and distribution of tasks related to the responsible hospital service and the healthcare staff in charge of performing the test, where and when the examination of the newborn should be performed and, finally, whether the study should be performed in the presence of the family.

Newborn hearing screening should test both ears and be universal (>95% of newborns). For some infants admitted to the Neonatal Intensive Care Unit (NICU), for example, infants on respirators, it may not be feasible or practical to perform a hearing test before one month of age due to the high probability of secretory otitis, noise interference and electrical interference from equipment, or infants not being in adequate and medically stable conditions to perform the test. In the latter case, measures should be taken to complete hearing screening when this situation reverses.

Screening should examine both ears, because unilateral congenital deafness must also be detected and diagnosed early to be treated, given its consequences for the development of the child (Trinidad *et al.,* 2010; Núñez *et al.,* 2018; Joint Committee on Infant Hearing, 2019).

Examination in the maternity ward before discharge is advised in order to avoid loss of cases. If for any reason this is not possible, an appointment should be made in an outpatient clinic linked to the programme

3.3.1. Screening

In terms of the site for performing the first audiological examination, examination in the maternity ward prior to discharge is recommended to prevent losses of cases. If for any reason this is not possible, an appointment should be made in an outpatient clinic linked to the programme (Sequí-Canet *et al.,* 2005).

Regarding inpatient hearing screening, there are a number of general and specific aspects to consider:

- For all newborns, the test should be performed in a quiet room. It should be performed by trained healthcare personnel who will also be responsible for recording the results.
- Babies should have the hearing screening test as close as possible to the time of hospital discharge, allowing sufficient time for it to be repeated if the baby does not pass it on the first attempt.
- The second attempt, if needed, should not be made immediately after the first one, but should be made at least several hours later, preferably if there has been a feed in between.
- In limited staffing settings, it is advisable for all nurses in maternity wards and neonatal units to be trained in the hearing examination, as this allows the test to be performed in all shifts, throughout the day and have coverage even during bank holidays and holiday periods. Nevertheless, in large hospitals, a special team of healthcare professionals can be dedicated to performing the screening (this improves reliability, but worsens continuity due to holidays and leave).
- It is advisable to have several testing devices so as not to have to stop testing altogether if one of them needs to be calibrated or repaired. For the same reason, several probes, headphones and cables must be prepared.
- As a general rule, the test should not take more than 5 minutes. If this cannot be done because

of the child's restlessness or other causes, it should be stopped and repeated later. Attempts should be made to do this after feeding, in a calm place, or while breastfeeding (Sequí-Canet et al., 2014; Sequí-Canet *et al.,* 2020).

– Do not insist on testing if, after two attempts, the result is "fail". Refer the patient to the next level after ensuring that the ear and probe have been properly adjusted and there were no technical problems (Joint Committee on Infant Hearing, 2019).

During screening, hearing risk factors should be assessed and documented. Necessary follow-up appointments should be scheduled with the ENT before discharge of neonate

When performing BAEPs, it is important to check the electrical noise present and disconnect as many electronic devices as possible (including mobile phones, pulse oximeters, lights, etc.) and to prepare the skin well before to reduce impedance (use a special gel).

3.3.2. Re-screening

The second test (re-screening) should be performed by expert personnel before 2-3 weeks of age (to be able to successfully perform a cytomegalovirus test in a timely manner if the newborn fails the test). The test should be bilateral, not just studying the ear that did not pass the test previously, to ensure that fluctuations or losses of hearing levels are not present in both ears, and controlling the time spent on repetitions. If one ear does not pass the test, refer the newborn to the ear, nose and throat (ENT) specialist immediately.

The Joint Committee on Infant Hearing (JCIH) recommends only one high-quality attempt to perform this second test, since, by simple statistical probability, if performed repeatedly to make "pass" / "fail" decisions, the probability of obtaining a "pass" result increases by chance alone (type I error) (Joint Committee on Infant Hearing, 2019).

A high-quality test implies that the child is sleeping or resting quietly without moving during the test, and that the permeability of the ear canal is ensured as much as possible before starting screening.

The JCIH states that the result of screening both ears, in the same session, using any technology, before hospital discharge and in re-screening, is acceptable as a hearing test passed in newborns not admitted to neonatal intensive care, which may help reduce the percentage of losses to follow-up.

This is not the case for infants who have received care in the NICU for more than 5 days, among which a higher prevalence of high hearing thresholds has been shown compared to infants in maternity wards. Not only is there an increased prevalence of hearing loss in this population, but there is also an increased risk of auditory neuropathy. For this reason, the exclusive use of BAEPs is recommended for the study of hearing in infants who have been admitted to the NICU. It is advised that infants who do not pass automated BAEP screening in the NICU be referred directly to the ENT for further screening or diagnostic hearing evaluation, rather than scheduling a new outpatient screening.

During screening, hearing risk factors should be assessed and documented. In these cases, BAEPs are recommended, and the necessary revisions by the ENT must be scheduled before discharging the newborn.

3.3.3. Documentation and reporting of results

All results of newborn hearing screening should be documented in the clinical history. The minimum documentation should include the date of screening, test method, result of each ear ("pass" / "fail" or not performed), and any hearing risk factors.

The results of the screening must be reported in writing to the child's family doctor in his/her health record and the final hearing screening result must be included in the discharge report.

The results of the newborn hearing screening programme should also be regularly reported to Public Health (Núñez *et al.,* 2020).

4. AFTER SCREENING

4.1. Losses in the process

Programmes for early detection of congenital hearing loss through universal newborn screening are faced with the significant problem posed by losses to follow-up and documentation of each case, which significantly delays timely diagnosis and early treatment. Loss to follow-up refers to the lack of compliance with the next phase of the screening, diagnosis or treatment process; while loss to documentation refers to incompletely recorded data that would make it possible to know the details of screening, as well as its adequate follow-up within the Congenital Hearing Loss Detection Programme (Cunningham *et al.,* 2018).

Loss rates during the process have gone from greater than 60% to currently less than 30% (Nicholson *et al.,* 2022). According to the data of the U.S. Centers for Disease Control and Prevention (CDC)(2014), 97.9% of newborns were screened and, of them, 34% of those who "failed" the first test were lost to follow-up or incompletely documented. To reduce these losses, the Joint Committee on Infant Hearing (JCIH) recommends that, in addition to documenting the results in the clinical history, programmes should ensure that there is a professional at the hospital where screening is performed who communicates directly with the primary care physician in case any child has not been screened or has not reported for the next step of the process (Subbiah *et al.,* 2018).

The Public Health Services should have a protocol to check, within the register of births in the autonomous community, those who, due to different circumstances, have not started screening and contact social services to try to recruit them.

A number of maternal and newborn factors have been identified that are associated with losses to follow-up in the process or losses to documentation. In the USA, studies have shown that children living in rural areas, who belong to ethnic and racial minorities, certain health insurance systems, children of adolescent mothers, children of mothers who are smokers, and children of mothers with a low level of education, are at an increased risk of being lost to follow-up or poorly documented (Chia-Ling *et al.,* 2008; Holte *et al.,* 2012; Bush *et al.,* 2014).

Correct screening does not guarantee a lower number of losses to follow-up, which shows that the barriers faced by families with low economic levels, minorities and those living in rural areas cannot be mitigated by measures limited to the scope of hospital practice. This emphasises the importance of the roles of families, primary care and public health services to ensure that children are properly monitored after a screening failure. There are routines in the hospital setting that help prevent missing screening or related documentation, such as, to name a few:

- Obtaining a daily list of all newborn admissions to the hospital ward and Neonatal Intensive Care Unit.
- Identifying children transferred to other parts of the hospital and document this in the screening programme.
- Properly identifying and documenting newborns who have died.
- Identifying newborns with special situations, such as those who must avail themselves of child protection services or those who are in the process of adoption.
- Documenting in the clinical history of each newborn the date and time of performing the screening test, its result, and if follow-up is required.
- Identifying newborns whose legal representatives have refused screening tests and collect written and signed evidence to be included in the clinical history.

In the USA, losses to follow-up have significantly decreased (Subbiah *et al.,* 2018). As a result of several programmes seeking solutions to this problem, simple actions have also been identified to take into account:

- Ensure adequate identification and communication with the primary care paediatrician assigned to the newborn before hospital discharge.
- Record additional family contact phone numbers before discharge.
- Document, in writing, the information that has been transferred to the family when the newborn does not pass the initial screening.
- Set a date for the next test appointment (re-screening) before the family leaves the hospital, explaining the importance of attending.
- Call the family to check the follow-up appointment and offer them helpful information (transportation, location of the visit…).

Extreme caution should be exercised in the following situations in order to avoid commonly associated losses (Joint Committee on Infant Hearing, 2019):

- Home births. A protocol should be developed to routinely offer screening in such cases.
- Births outside the autonomous community. The autonomous communities must establish agreements between them to share information on the outcome of screening and follow-up of children.
- Outpatient screening. This should also be arranged for children whose parents, mother and father, or legal guardians have refused or have been unable to complete hearing screening and later decide to have the hearing test performed for their baby.
- Hospital discharges before screening. There must be a procedure in place to schedule an outpatient appointment.
- Transfers to other hospitals. The report must state whether screening was performed and its results. The destination hospital should perform screening if not done previously or if a new risk factor for development of hearing loss has emerged.

The lack of an information system that makes it possible to share reliable data across newborn screening programmes for hearing loss has been a historical burden to ascertaining results

In order to recuperate lost cases, the participation of primary care professionals is essential, and patient follow-up is required in a longitudinal database accessible to all specialists involved.

No cases should ever be considered definitively lost, and for all children, regardless of age, it must be confirmed that at some time in their life, they underwent a hearing study, and if there is any doubt, perform it then.

4.2. Information Systems

The lack of an information system that makes it possible to share reliable data from newborn screening programmes for hearing loss has been a historical burden to ascertaining results in most countries, including Spain. For this reason, it is difficult to make comparisons between programmes and draw conclusions that help improve programme performance. Moreover, this lack of real perspective of the situation may call into question the favourable cost/benefit ratio that must exist to maintain them (Joint Committee on Infant Hearing, 2019; Sequí-Canet *et al.,* 2021).

It is essential to have an information system capable of meeting the objectives required to ascertain the functioning of the screening programme and patient follow-up, in addition to allowing adequate quality control of the entire process. An information system of this nature and purpose must be capable of achieving at least the following objectives (Ministry of Health. Working Group on Newborn Hearing Screening of the Population Screening Presentation, 2021):

- To allow, at the regional and state levels, monitoring and assessment of the newborn screening programme for hearing loss included in the common services portfolio of the National Health System (NHS).
- Be the official source of information that provides the data for the annual technical assessment report.
- Collect data and indicators that will allow measurement of quality objectives.
- Enable the sharing of information on results.

To ensure that these objectives are achieved, the challenges that a complex information system involves, such as the variability in the collection and recording of follow-up data between the different screening programmes, which has an impact on data integrity and quality, must be overcome. It should also be borne in mind that there may be differences in the infrastructure and functionality of information systems, which limit the ability of some programmes to accurately identify data from all births, in order to avoid duplicates that are individually identifiable.

Another problem is the heterogeneity of the definitions of data to be entered and of the performance measures between the programmes, which causes differences in the results reports that make their evaluation difficult; not to mention the practical obstacles to sharing data among the professionals involved and the workload that managing an adequate information system entails. It should be noted that there is still considerable dependence on the technology and processes of the twentieth century, which have not been brought up to date with measurement and data management technology such as Big Data and Machine Learning.

For all the above, whenever possible, nationally approved indicators and standard data elements or uniform coded value sets should be used.

4.2.1. Quality control

Hospital newborn hearing screening programmes must work with all institutions to achieve and maintain high-quality services in their autonomous communities. Indicators from a more basic high-quality newborn hearing screening programme include (Joint Committee on Infant Hearing, 2019):

- Studying both ears in all newborns.
- Assess risk indicators for hearing loss in all newborns.
- Perform the screening test before discharge of the mother in the maternity ward and always in the child's first month of life.
- Have a false-positive rate of 3% or less and a false-negative rate that tends to 0.
- Achieve high quality screening that allows for screening referral to ENT services to not exceed 4% of newborns for diagnostic confirmation.
- Provide information to both parents or legal representatives regarding hearing screening tests, outcomes, referral centres and family support.
- Referral to the ENT service, for follow-up, of newborns with risk indicators for developing late onset hearing loss.
- Record data on newborns in the programme's computer record.
- Record the results in the corresponding section of the Child Health Booklet.
- Conduct the evaluation and monitoring of the programme results by analysing the data from the computerised registry.
- Ensure that more than 10% of infants who fail initial screening are not lost to follow-up.

The effectiveness of screening programmes implies not only measures of internal validity (sensitivity, specificity, coverage, etc.), but also measures of external validity, whose key would be the age of identification or confirmation of hearing loss. However, these are still indirect measures of the final benefit, as identification and initiation of prosthetic adjustment are only the starting point of a long journey throughout the child's life, which should manifest in other and more general effects (quality of life, quality of life of the family, academic achievement and work). Measurement of age of identification is not a measurement of a result *per se,* but it is usually used to describe short- or medium-term results. It is therefore necessary to incorporate other long-term indicators in order to provide a more complete picture of the whole process (Ramos, 2003).

Hospital newborn hearing screening programmes must work with all institutions to achieve and maintain high-quality services in their autonomous communities

5. INVOLVEMENT OF FAMILIES AND FOLLOW-UP

Before screening, the family should be offered information about the screening programme, providing them with the necessary information so that they can make any decision they consider appropriate about their participation in it (Núñez *et al.,* 2016; Núñez *et al.,* 2019; Núñez *et al.,* 2021).

Regardless of the communication route and the medium used, the following information must be provided:

- Programme objective. Voluntary nature of participation.
- Importance of early detection of hearing loss. Expected benefits. Risks and adverse effects.
- What the screening test entails, when and how it is done.
- What the test result means. Steps to take in the event of a "fail" result.
- Informed consent/dissent.
- How to get more information. Family support structures.

Written information should be provided, including at least the aforementioned sections. In order to support written information, information leaflets with simple, synthetic and direct content are very useful, making them easier to understand. All information must be adapted to the needs and characteristics of users.

Families should receive information throughout the screening process that includes risk factors for hearing loss, normal language development, and resources for more information, if desired.

Families of newborns who "fail" the hearing screening will receive information about why their baby may not have passed the screening, the importance of follow-up and the next steps, as well as subsequent visits they are advised to attend.

The Family Association Movement is a necessary reference point in the family referral pathway and as part of the interdisciplinary organisation

The family should be informed of the results as soon as possible. It is important to clearly explain what the results mean. It is fundamental that the ideal time is chosen to communicate the results and clearly explain what screening involves, emphasising the differences between screening and diagnosis. On the other hand, it is important to note the possibility of late-onset hearing loss despite the fact that the result was a normal screening test (Núñez *et al.,* 2015; Núñez *et al.,* 2020; Sequí-Canet *et al.,* 2021).

If the result shows normal hearing, the information can be provided in writing only. However, if the result is otherwise and other diagnostic tests are needed, in addition to a written report, it is important that a health-care professional explain to the family, verbally and in depth, the meaning of this result and the steps to follow thereafter. The results of the test must be recorded in the documentation of the child (child health booklet and clinical history).

This information, as well as guidance and counselling for the family, play a prominent role in the hearing loss intervention (Pendleton and Hasler, 1983; Núñez *et al.,* 2016; *Núñez et al.,* 2020).

When reporting the diagnosis, we must prepare, before providing the information, the severity and prognosis of deafness and the special characteristics of the child under study. The physician is usually responsible for reporting the diagnosis and prognosis in these cases, though this news should be understood as part of a process and assumed by the rest of the team.

Referral circuits and care pathways must be established for both the child's care and family support, coordinated with each other and ensuring the continuity of the process. In this regard, having a stable specialised structure for meeting other families, with information, guidance and accompaniment services, is a fundamental source of support for the family after learning about the diagnosis and having to start the next steps for early care, prosthetic adaptation and speech therapy intervention. The Family Association Movement is a necessary reference point in the family referral pathway and as part of the interdisciplinary organisation involved with the child with deafness and his or her family (Núñez *et al.,* 2016; Núñez *et al.,* 2020; Núñez *et al.,* 2021).

6. CODEPEH RECOMMENDATIONS 2022

Starting with the update on the clinical problems and the most frequently asked questions regarding universal newborn hearing screening, addressing the aspects requiring an update, the CODEPEH then presents the appropriate recommendations (*see Figure 7*).

Who should perform the screening tests?
In Spain there is case-law specifying that, according to Act 44/2003 of 21 November 2003, on the Regulation of Healthcare Professions, personnel with a degree in Medicine and/or Nursing must be responsible for performing screening tests on newborns.
The screening personnel must be trained and have the necessary skills to apply the instructions specified in the protocol and properly handle the equipment.

When is the best time to perform the screening test?
Healthy newborns. Can be screened from 6 hours after birth. However, for optimal results, it is recommended to wait until at least the first 24 hours of life and to perform the test as close as possible to the time of hospital discharge.

Newborns admitted to the Intensive Care Unit. Perform screening when the child's condition is stable or before hospital discharge.

Newborns born at home. Screening is recommended before the first two weeks of life within the outpatient schedule.

Where should screening take place?
Choose any room with a quiet environment, either in or out of hospital, and with minimal electromagnetic pollution.

Which technique should be used to perform screening?
In the case of a healthy child without risk factors, both otoacoustic emissions (OAEs) and brainstem auditory evoked potential (BAEP) tests are valid.
In the case of children with risk factors and/or admitted to the neonatal ICU, the use of the BAEP is recommended.

How do you ensure that screening equipment is in optimal condition?
Check that the equipment has been regularly calibrated according to the manufacturer's specifications.
Cleaning and maintenance of the probe and equipment should be performed daily.

How many times are the tests repeated?
If the result of the first screening test in healthy children, performed correctly, is "fail" in two attempts, refer to the next level.

How to check that every newborn has been screened?
It is helpful to obtain the hospital's daily newborn admissions census.
The date and time of screening, the results in each ear and whether follow-up of the case is required should be documented in the newborn's clinical history.
Cases where the families have refused the test need to be documented.
The possibility of including out-of-hospital births in screening should be enabled.

How to manage the follow-up of failed screening cases or those who did not report for testing?
In addition to documenting the screening result in the database, the need to repeat the test due to a failure in the first attempt according to the protocol should be recorded in the newborn's clinical history. Families and the primary care paediatrician shall be informed.
The newborn should be scheduled for subsequent protocol tests before leaving the hospital.

How to proceed when identifying risk factors for hearing loss requiring follow-up?
The screening personnel is responsible for the identification of children with risk factors who, having passed the screening, may have late-onset or progressive hearing loss, ensuring appropriate follow-up within the programme protocol.

What actions can decrease the rate of cases lost in the process (screening, re-screening or diagnostic confirmation)?
The primary care paediatrician should be properly informed and family contact details checked.
It is useful to appoint a person from the multidisciplinary team to contact families who need assistance and guidance to attend appointments.

Which technique should be used to re-screen healthy children?
A normal result in both ears in the same session using any technology is accepted as a successfully passed hearing test.
Only one attempt in both ears made under optimal conditions is recommended in re-screenings.

Why is an information system needed?
An information system is needed to ensure quality control of the whole process and compliance with the programme objectives, as well as for adjustment of its functioning according to the quality standards set and appropriate follow-up of patients.

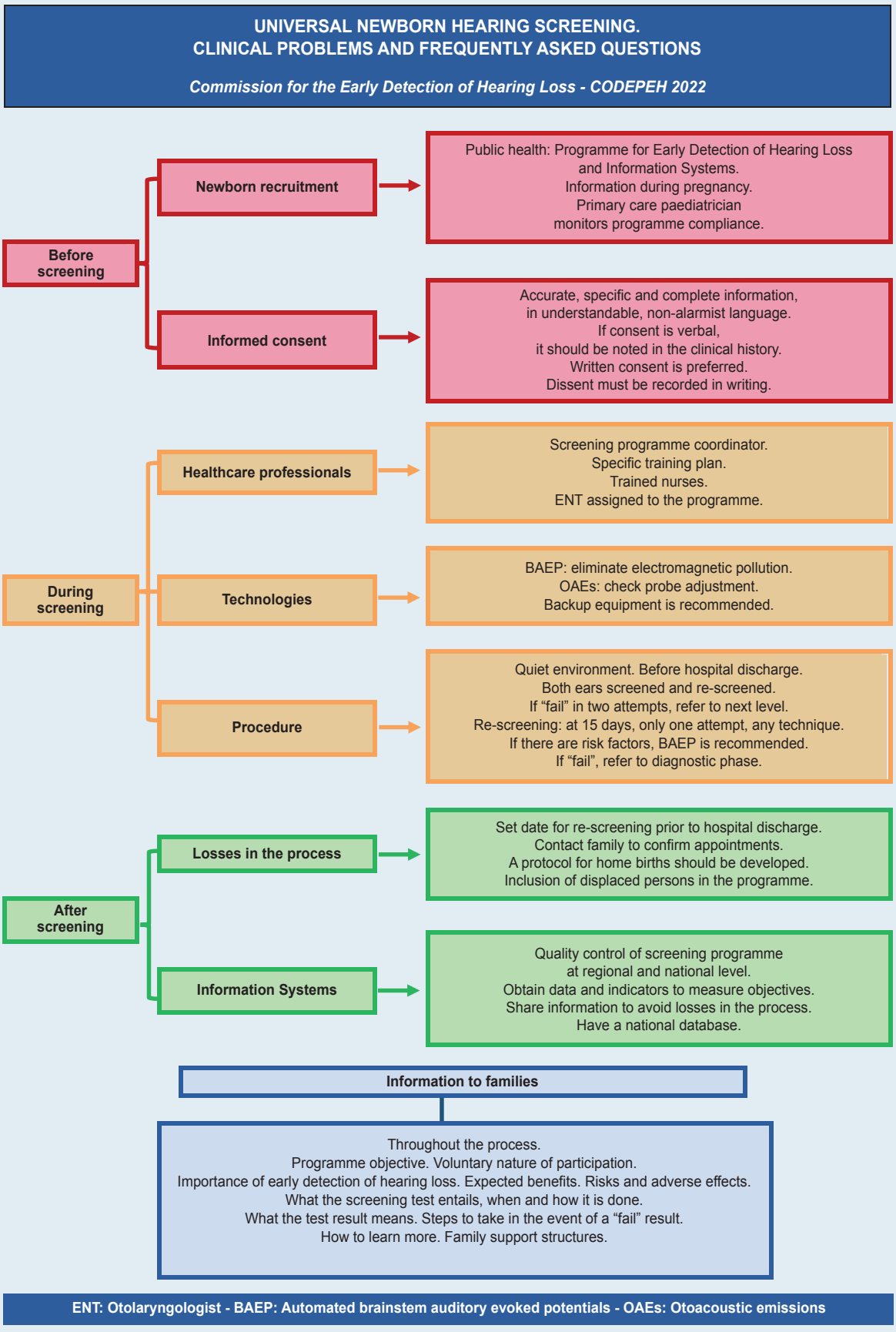
What information should be provided to families prior to screening?
Information should preferably be provided in writing on:
- Programme objective. Voluntary nature of participation.
- Importance of early detection of hearing loss. Expected benefits. Risks and adverse effects.
- What the screening test entails, when and how it is done.
- What the test result means. Steps to take in the event of a "fail" result.
- Informed consent. Informed dissent.
- How to get more information. Family support structures.

When and where is information given about hearing screening?
It is recommended that families be routinely informed as part of the pregnancy monitoring programme, as well as in the place of birth (hospital or home).

What information should be given if the newborn "fails" screening?
Families will receive information about why their baby may not have passed the screening, the importance of follow-up and the next steps, as well as subsequent visits they are advised to attend.

The **Newborn Hearing Loss Screening Programme must have a professional responsible for its coordination**, with experience in the management of newborn hearing screening, with in-depth knowledge of the equipment required to perform it and the responsibilities involved, among others, with regard to the personnel involved in the implementation of the programme, their knowledge and training for this task, planning and supervising their ongoing training.
The Programme Coordinator must also monitor and report on compliance with the quality indicators, as well as verify that all newborns have been registered in the database, coordinating referral and care by other services and follow-up of cases when required.

7. FIGURE Action algorithm (SOURCE: own preparation, CODEPEH 2022)



8. REFERENCES

American Academy of Pediatrics (2000). Serving the family from birth to medical home. A report from the newborn task force convened in Washington DC. *Pediatrics,* 106(2), 383-427.

Arnold, C. L. et al. (2006a). Infant hearing screening: stakeholder recommendations for parent-centered communication. *Pediatrics,* 117, S341-S354.

Arnold, C. L. et al. (2006b). Assessment of newborn screening parent education materials. *Pediatrics,* 117, S320-S325.

Benito-Orejas, J. I. et al. (2008). Comparison of two-step transient evoked otoacoustic emissions (TEOAE) and automated auditory brainstem response (AABR) for universal newborn hearing screening programs. *Int J Pediatr Otorhinolaryngol,* 72(8), 1193-1201.

Berg, A. L. et al. (2005). Newborn hearing screening in the NICU: profile of failed auditory brainstem response/passed otoacoustic emission. *Pediatrics,* 116(4), 933-938.

Bush, M. L. et al. (2014). Delays in diagnosis of congenital hearing loss in rural children. *J Pediatr.* 164(2), 393-397.

Bussé, A. M. L. et al. (2021a). Assessment of hearing screening programmes across 47 countries or regions I: provision of newborn hearing screening. *Int J Audiol,* 60(11), 821-830.

Bussé, A. M. L. et al. (2021b). Assessment of hearing screening programmes across 47 countries or regions III: provision of childhood hearing screening after the newborn period. *Int J Audiol,* 60(11), 841-848.

Chia-Ling, L. et al. (2008). Evaluating loss to follow-up in newborn hearing screening in Massachusetts. *Pediatrics,* 121(2), e335-e343.

Cunningham, M. et al. (2018). Infant, maternal, and hospital factors' role in loss to follow-up after failed newborn hearing screening. *Academic Pediatrics,* 18(2), 188-195.

Danhauer, J. L. et al. (2006). A national survey of pediatric otolaryngologists and early hearing detection and intervention programs. *J. Am. Acad. Audiol,* 17(10), 708-721.

DeLuca, J. M. et al. (2011). Parents' experiences of expanded newborn screening evaluations. *Pediatrics,* 128(1), 53-61.

Doyle, K. J. et al. (2000). External and middle ear effects on infant hearing screening test results. *Otolaryngol Head Neck Surg,* 122(4), 477-481.

España. Ley 41/ 2002, de 14 de noviembre, básica reguladora de la autonomía del paciente y de derechos y obligaciones en materia de información y documentación clínica. *Boletín Oficial del Estado,* de 15 de noviembre de 2002, núm. 274, pp. 2002-22188.

España. Ministerio de Sanidad. Grupo de trabajo de cribado de hipoacusia neonatal de la Ponencia de Cribado Poblacional (2021). *Indicadores de calidad y sistema de información del Programa de Cribado Neonatal de Hipoacusia del Sistema Nacional de Salud.* Madrid, 2021.

Holte, L. et al. (2012). Factors influencing follow up to newborn hearing screening for infants who are hard of hearing. *Am J Audiol,* 21(2), 163-174.

Holzinger, D. et al. (2021). Development and implementation of a low-cost tracking system after newborn hearing screening in Upper Austria: lessons learned from the perspective of an early. *Intervention Provider Children* (Basel), 8(9), 743.

Johnson, J. L. et al. (2005). A multicenter evaluation of how many infants with permanent hearing loss pass a two-stage otoacoustic emissions/automated auditory brainstem response newborn hearing screening protocol. *Pediatrics,* 116(3), 663-672.

Joint Commitee on Infant Hearing (2019). Position statement: principles and guidelines for early hearing detection and intervention programs. *Journal Early Hearing and Intervention,* 4(2), 1-44.

Mackey, A. R. et al. (2021). Assessment of hearing screening programmes across 47 countries or regions II: coverage, referral, follow-up and detection rates from newborn hearing screening. *Int J Audiol,* 60(11), 831-840.

Moeller, M. P. et al. (2006a). Strategies for educating physicians about newborn hearing screening. *J. Acad. Rehabil. Audiol,* 39, 11-32.

Moeller, M. P. et al. (2006b). Primary care physicians' knowledge, attitudes, and practices related to newborn hearing screening. *Pediatrics,* 118(4), 1357-1370.

Nicholson, N. et al. (2022). Analysis of health disparities in the screening and diagnosis of hearing loss: early hearing detection and intervention hearing screening follow-up survey. *Am J Audiol,* 31(3), 764-788.

Núñez, F. et al. (2015). Recomendaciones CODEPEH 2014. *Revista Española de Discapacidad,* 3(I), 163-186.

Núñez, F. et al. (2016). Diagnóstico etiológico de la sordera infantil: recomendaciones CODEPEH 2015. *Revista Española de Discapacidad,* 4(I), 193-218.

Núñez, F. et al. (2018). Diagnóstico y tratamiento precoz de la hipoacusia unilateral o asimétrica en la infancia: recomendaciones CODEPEH 2017. *Revista Española de Discapacidad,* 6(I), 259-280.

Núñez, F. et al. (2019). Actualización de los programas de detección precoz de la sordera infantil: recomendaciones CODEPEH 2018 (Nivel 1: Detección). *Revista Española de Discapacidad,* 7(I), 201-220.

Núñez, F. et al. (2020). Actualización de los programas de detección precoz de la sordera infantil: recomendaciones CODEPEH 2019 (Niveles 2, 3 y 4: diagnóstico, tratamiento y seguimiento)". *Revista Española de Discapacidad,* 8(I), 219-246.

Núñez, F. et al. (2021). Sordera infantil con discapacidad asociada (DA+): recomendaciones CODEPEH 2021. *Revista Española de Discapacidad,* 10(2), 209-227.

Pendleton, D., y Hasler, J. (1983). Comunicación médico paciente. Nueva York: *Prensa Académica.*

Queiro, T. et al. (2007). Información a padres sobre cribado neonatal de metabolopatías: evaluación de la situación actual y establecimiento de estándares de información basada en la evidencia (Informes, Estudios e Investigación; avalia-t num. 2007/07). Ministerio de Ciencia e Innovación. *https://www.sergas.es/docs/Avalia-t/avaliat200704_crib-Metabolopatias.pdf.*

Ramos, A. (2003). Control de calidad de los sistemas de screening de la audición. Coste-beneficio. En CODEPEH y Ministerio de Sanidad y Consumo (Eds): *Libro blanco sobre hipoacusia. Detección precoz de la hipoacusia en recién nacidos* (161-76). Madrid: Ministerio de Sanidad y Consumo.

Ravi, R. et al. (2017). A national survey of knowledge, attitude and practices among pediatricians towards newborn hearing screening in India. *Int. J. Pediatr. Otorhinolaryngol,* 95, 9-14.

Ravi, R. et al. (2018). Systematic review of knowledge of, attitudes towards, and practices for newborn hearing screening among healthcare professionals. *Int Jour Pediatr Otorhinolaryngol,* 104, 138-144.

Riaño, I. et al. (2013). Recomendaciones para la toma de decisiones ante la negativa de los padres para la vacunación de sus hijos: análisis ético. *Anales de Pediatría,* 79(1), 1-50.

Sequí-Canet, J. M. et al. (2005). Hearing screening based on otoacoustic emissions in infants born in secondary-level hospitals: feasible, efficient and effective. *Acta Pediátrica Española,* 63(11), 465-470.

Sequí-Canet, J. M. et al. (2014). Perinatal factors affecting the detection of otoacoustic emissions in vaginally delivered, healthy newborns, during the first 48 hours of life. *Acta Otorrinolaringologica* (English Edition), 65(1), 1-7.

Sequí-Canet, J. M. et al. (2020). Breastfeeding results in better hearing in newborns compared to bottle-feeding. *Journal of Clinical and Translational Research,* 6(3), 81-86.

Sequí-Canet, J. M. y Brines-Solanes, J. (2021). Keypoints to successful newborn hearing screening. Thirty years of experience and innovations. *Healthcare* (Basel, Switzerland), 9(11), 1436.

Subbiah, K. et al. (2018). Progress in documented early identification and intervention for deaf and hard of hearing infants: CDC`s hearing screening and follow-up survey, United States, 2006-2016. *J Early Hear Detect Interv,* 3(2), 1-7.

Trinidad, G. et al. (2010). Recomendaciones CODEPEH 2010. *Revista FIAPAS,* 131.

FIAPAS

THE SPANISH CONFEDERATION OF FAMILIES OF DEAF PEOPLE (FIAPAS)

EXCELENCIA INNOVACIÓN SOSTENIBILIDAD 400 EFQM 2020 - 2023

FOMENTANDO INCLUSIÓN. APOYANDO PERSONAS. AVANZANDO SOLIDARIAMENTE.
(FOSTERING INCLUSION SUPPORTING PEOPLE PROGRESSING TOGETHER)

Pantoja, 5 (Local) 28002 Madrid
Tel.: 91 576 51 49 Fax: 91 576 57 46
Telesor Service
fiapas@fiapas.es www.fiapas.es www.bibliotecafiapas.es

Follow us on:

Legal Deposit: M-26488-1988

This publication is available for download in PDF at www.bibliotecafiapas.es

Edition 2023 funded by:

GOBIERNO DE ESPAÑA
MINISTERIO DE DERECHOS SOCIALES Y AGENDA 2030

FOR SOLIDARITY
OTHER PURPOSES OF SOCIAL INTEREST
GOVERNMENT OF SPAIN
MINISTRY OF SOCIAL SERVICES AND 2030 AGENDA

Fundación ONCE
ONCE Foundation