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COMISIÓN PARA LA DETECCIÓN PRECOZ DE LA HIPOACUSIA
COMMISSION FOR THE EARLY DETECTION OF HEARING LOSS

Update of early detection programmes for pediatric hearing loss: 2019 CODEPEH recommendations

(Levels 2, 3 and 4 Diagnosis, Treatment and Follow-up)

Actualización de los programas de detección precoz de la sordera infantil: recomendaciones CODEPEH 2019

(Niveles 2, 3 y 4 Diagnóstico, Tratamiento y Seguimiento)

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The present CODEPEH Recommendations 2019 document has been prepared in the context of the project *"Update on the Programs for the Early Detection of Infant Hearing Loss (Diagnosis, Treatment and Monitoring)"*, developed by the Spanish Federation of Families of Deaf People (Confederación Española de Familias de Personas Sordas-FIAPAS), in collaboration with the Commission for the Early Detection of Infant Hearing Loss (Comisión para la Detección Precoz de la Sordera Infantil-CODEPEH), and with co-organization by the Royal Patronage for Disability (Real Patronato sobre Discapacidad).

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ABSTRACT

With the review of recent scientific evidences on the processes of diagnosis, treatment and follow-up, CODEPEH completes its recommendations regarding neonatal hearing screening programs. It is necessary to update some aspects of the etiological diagnosis, ordered sequentially in a previous publication. It reminds that it should be carried out without delaying the audiological diagnosis, which incorporates the routine use of steady state auditory evoked potentials that allow knowing the auditory thresholds in a more extensive frequency range than those of the brain stem. It is recognized the importance of binaural hearing by warning that unilateral deafness is not without negative consequences for the child's development, also requiring treatment and follow-up. Bilateral cochlear implantation is recommended in cases of bilateral profound hearing loss. Organizing an interdisciplinary team, with an adequate information system, is essential to achieve the objectives of early treatment of congenital deafness, highlighting the role of the families in this process and the support point offered by the Family Associative Movement.

KEY WORDS: neonatal hearing screening, infant hearing loss, cochlear implant, hearing diagnosis techniques, binaural hearing, interdisciplinarity, information systems, role of the families.

RESUMEN

Con la revisión de recientes evidencias científicas sobre los procesos de diagnóstico, tratamiento y seguimiento, la CODEPEH completa sus recomendaciones en relación con los programas de cribado neonatal de la hipoacusia. Es necesario actualizar algunos aspectos del diagnóstico etiológico, ordenado secuencialmente en una publicación anterior, recordando que se lleve a cabo sin retrasar el diagnóstico audiológico, que incorpora el uso rutinario de los potenciales evocados auditivos de estado estable que permiten conocer los umbrales auditivos en un rango frecuencial más extenso que los de tronco cerebral. Se reconoce la importancia de la audición binaural alertando de que la sordera unilateral no está exenta de consecuencias negativas para el desarrollo del niño, requiriendo tratamiento y seguimiento. Se recomienda la implantación coclear bilateral en los casos de hipoacusia profunda bilateral. Organizar un equipo interdisciplinar, con un adecuado sistema de información, es imprescindible para alcanzar los objetivos del tratamiento precoz de la sordera congénita, destacando el rol que desempeñan las familias en este proceso y el punto de apoyo que ofrece el Movimiento Asociativo de Familias.

PALABRAS CLAVE: cribado neonatal, hipoacusia, hipoacusia infantil, implante coclear, técnicas de diagnóstico auditivo, audición binaural, interdisciplinariedad, sistemas de información, rol familias.

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The reading of this Document is complemented by the CODEPEH's Recommendations Document, on the Detection phase, issued in 2018.

All CODEPEH documents and complementary information brochures can be consulted in FIAPAS's Virtual Library (http://bit.ly/DocCODEPEH). Free download:

– CODEPEH (Núñez, F. et al.) (2014): "Late-onset and acquired deafness in children: 2014 CODEPEH recommendations." *FIAPAS Journal,* 151. Supplement

– CODEPEH (Núñez, F. et al.) (2015): "Aetiological diagnosis of pediatric hearing loss: 2015 CODEPEH recommendations". *FIAPAS Journal,* 155. Supplement

– CODEPEH (Núñez, F. et al.) (2016): "Diagnosis and treatment of secretory otitis media in children: 2016 CODEPEH recommendations". *FIAPAS Journal,* 159. Supplement

– CODEPEH (Núñez, F. et al.) (2017): "Diagnosis and treatment of unilateral or asymmetric hearing loss in children: 2017 CODEPEH recommendations". *FIAPAS Journal,* 163. Special supplement

– CODEPEH (Núñez, F. et al.) (2018): "Update of early detection programmes for pediatric hearing loss: 2018 CODEPEH recommendations (Level 1 Detection)", *FIAPAS Journal,* 167. Special supplement

– CODEPEH (Núñez, F. et al.) (2019): "Update of early detection programmes for pediatric hearing loss: 2019 CODEPEH recommendations (Diagnosis, Treatment and Follow-up)". *FIAPAS Journal,* 171. Special supplement

1. INTRODUCTION

Over the last few decades we have seen sustained growth in knowledge about the causes and optimal treatment of pediatric sensorineural hearing loss. Important advances fundamentally in the field of molecular genetics, as well as in imaging diagnosis, together with the lack of a consensus-based protocol on the diagnostic process once neonatal screening has confirmed hearing loss, led the Commission for the Early Detection of Infant Hearing Loss (CODEPEH) to publish its recommendations on the etiological diagnosis (Núñez *et al*., 2016a: 193-218). This document established a protocolised sequence for investigation of the causes of congenital hearing loss, in which genetic studies are recommended as a second step after exploration and study of the clinical history, since among the available complementary tests they offer the best diagnostic performance by identifying a genetic cause in 44% of all patients with bilateral sensorineural hearing loss (Sloan-Heggen *et al*., 2016: 441-50).

We are living a growth in knowledge about the causes and optimal treatment of hearing loss in children.

This step also recommended the study of cytomegalovirus (CMV) - the most prevalent infectious cause of congenital hearing loss, and which has been the focus of special attention in the latest recommendations of the CODEPEH (Núñez *et al*., 2019: 201-220). The impact of congenital CMV (cCMV) infection in neonatal hearing loss screening programs is so important that it has led to modification of their algorithms (Núñez *et al*., 2015: 163-186).

Imaging tests, as the third step in diagnostic evaluation, are intended to identify anatomical causes of the disorder and assess candidates for surgical treatment. The two techniques used, computed tomography (CT) and magnetic resonance imaging (MRI), offer diagnostic performance proportional to the severity of hearing loss - with performance being higher for MRI than for CT (Beeck Calkoen *et al*., 2018: 180-185).

Whenever possible, hearing tests should be performed in parallel to the etiological diagnosis. Audiometric testing constitutes a priority to ensure compliance with the objectives of the neonatal screening program. Regardless of whether hearing aids are fitted or hearing implants are chosen, the ultimate aim is the early and optimal development of listening, speech and language, within the appropriate developmental stages, in order to place the child with hearing loss on an equal footing with their hearing-able peers at the start of the education and schooling process (Leigh *et al*., 2013: 443-450; Núñez *et al*., 2018: 259-280; Núñez *et al*., 2019: 201-220).

Cochlear implants represent one of the most important advances in the treatment of profound hearing loss, and were approved as treatment for deafness in adults in 1985 and in children in 1990. Since then, the indications have evolved and expanded with clinical experience, and now include implantation with lower hearing thresholds, electroacoustic stimulation in the presence of residual hearing, and implantation in unilateral hearing loss (UHL) (Carlson *et al*., 2015: 43-50; Gantz *et al*., 2016: e118-125; Peters *et al*., 2016: 713-721).

All the above addressed by the CODEPEH in its recommendations on the screening, diagnosis and treatment process takes on meaning in both this new work and that corresponding to 2018. In both cases the aim has been to review all the steps of the process, incorporating the knowledge arising from the abundant scientific evidence. In this regard, and as has been scientifically recognized, the families play a role in this entire process and, in general terms, in the educational and social development of children with hearing loss. Family associative movements therefore must serve as a point of support in all the stages, as part of the interdisciplinary team.

The CODEPEH therefore considers it necessary to establish new recommendations to complete the proposal for the updating of hearing screening programs based on the latest evidence on progress related to implementation of the programs at levels 2, 3 and 4: diagnosis, treatment and monitoring.

2. DIAGNOSIS

2.1. Hearing diagnosis

Although a previous CODEPEH document (Recommendations 2015) on the etiological diagnosis of hearing loss conducted a comprehensive review on the state of the art (Núñez *et al*., 2016a: 193-218), new advances are continuously being made for the correct use of the available diagnostic resources.

There are currently no variations in the recommended diagnostic sequence (Figure 1). A series of objective and subjective hearing tests are available, adapted to the age and cooperative capacity of the child. The different hearing tests are complementary, and so a single type of test cannot be used to establish proper diagnosis and treatment.

2.1.1. Objective tests

- **Tympanometry and impedanciometry**

Both of these tests assess the anatomical situation of the middle ear and its function, using a probe to introduce a pure tone of 1000 hertz (Hz) in the external auditory canal (EAC) in infants under one year of age versus 226 Hz in the rest of cases (Kei *et al*., 2003: 20-28), with the induction of pressure variation in the canal by means of a pump. As a result, we obtain three basic Jerger curves (Jerger, 1970: 311-324) and the Baldwin curves in infants under 9 months of age (Baldwin, 2006: 417-427).

By means of the stapedial (acoustic) reflex we can study the reflex arc of the VII and VIII cranial nerves to obtain information on cochlear and retrocochlear lesions.

- **Otoacustic emission (OAE)**

Since Kemp first described OAEs in 1978 (Kemp, 1978: 1386-1391), these have become a standard element in the battery of pediatric hearing tests. The two clinically most widely used techniques are transient evoked otoacoustic emissions (TEOAEs), which are mainly applied in neonatal screening programs, and distortion product otoacoustic emissions (DPOEAs) - which are more useful for the study of cisplatin ototoxicity.

At diagnostic level, OAEs are essential for differentiating cochlear or retrocochlear injury (6.5% of all cases of hearing loss), since their presence associated to pathological auditory evoked potentials (AEPs) helps define the topography of the injury (Boudewyns *et al*., 2016: 993-1000; Zubicaray *et al*., 2014: 1-15).

Steady state auditory evoked potentials (SSAEPs) constitute a safe and essential test

- **Brainstem auditory evoked potentials (BAEPs)**

These potentials were described in 1967 by Sohmer and Feinmesser (Sohmer and Feinmesser, 1967: 427-435). They are based on the electrophysiological response to stimulation of the auditory pathway to the inferior colliculus within the brainstem.

In addition to determining the threshold by means of the V-wave, analysis of the latencies and inter-latencies of the rest of the waves contributes to determine the topographic location of the lesion (Sohmer and Feinmesser, 1967: 427-435). The response obtained informs us about the condition of the high frequencies between 2000-4000 Hz, though sensitivity is poor at low frequencies. The latency times do not normalize until the first year of life - a fact that must be taken into account particularly in very preterm infants (Bakhos *et al*., 2017: 325-331).

- **Steady state auditory evoked potentials (SSAEPs)**

These potentials were first described in relation to visual stimuli and were defined by Regan (Regan, 1989). Stable state potentials are periodic quasi-sinusoidal responses with amplitude and phase characteristics that remain stable over time (Pérez-Ábalo *et al*., 2003: 42-50).

They allow thresholds to be determined at different frequencies (500, 1000, 2000 and 4000 Hz), without requiring participation of the subject. Correlation studies with pure-tone audiometry have confirmed their validity in neonatal, pediatric and young adult populations with normal hearing (Torres-Fortuny *et al*., 2016: 375-379). They also serve to detect residual thresholds in young children in the absence of BAEPS response (Grasel *et al*., 2015: 1-7).

In addition to their total objectivity, since they do not depend on the patient or on the explorer, SSAEPs offer the advantage that they can be performed at various frequencies to obtain a graphic display superimposable to an audiometric recording. In addition, they are more effective at detecting traces of hearing function in individuals with profound hearing loss. They can be performed in both ears simultaneously, thereby affording speed to the study. In nursing infants, the thresholds experience practically no changes with age, though they are affected by maturation (Martinez-Beneyto *et al*., 2014: 171-181). In preterm children, an increase in the thresholds is observed (Yang *et al*., 2017: 661-667). In hearing neuropathy and in cases secondary to brain injury, the thresholds are altered.

In contrast to BAEPS, a topographic lesion diagnosis cannot be established with SSAEPs.

There is still not enough scientific evidence to recommend SSAEPs in the screening phase, though it is expected that they may be used in the future, offering broader frequency information on the thresholds.

This exploration method should be included in the study of hearing in neonates and in children to obtain an early estimate of the hearing thresholds in cases of suspected hearing loss, with a view to facilitating early hearing aid adaptation in very young children (Núñez, F. *et al*., 2016b: 193-200; Vlastarakos *et al*., 2017: 464-468).

2.1.2. Subjective tests

It should be remembered that subjective tests are essential for the confirmation and evolutive assessment of hearing loss.

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- **Liminal tone audiometry**

Measurement is made in decibels (dB), and the frequencies of 250, 500, 1000, 2000, 3000, 4000 and 8000 Hz are evaluated. The thresholds are confirmed when the emitted tone response rate is 50%. Airway and bone transmission should be measured.

- **Verbal audiometry or speech audiometry**

This technique involves the use of words which the child hears and must repeat or point to in drawings. The threshold is established when the child correctly repeats 50% of the words, and it moreover allows us to evaluate how the patient discriminates language at different intensities.

- **Behavioural observation audiometry**

This type of audiometry was first described in 1944 (Ewing and Ewing, 1944: 309-338). It is used in infants under 9 months of age and can be performed with voice, warble tones (FM) or narrow band tones. The responses may consist of: head or limb reflex action, startled body response, suction, blinking, raising of eyebrows, or the cessation of certain types of behaviour such as movement or suction. Behavioural observation is a subjective measure of hearing ability and does not afford specific frequency information.

- **Visual reinforcement audiometry**

This technique can be performed from 6 months of age until about one year. The child is visually rewarded with light and animated toys on turning the head towards the sound source. Prior conditioning training is used. This technique can provide information at frequencies from 250 to 8000 Hz.

- **Play audiometry**

This technique is usually performed in infants from 30 months to 5 years of age, though in some cases it can be applied from 24 months and in patients with disabilities who do not collaborate in conventional tone audiometry. The child is instructed to perform a simple task such as placing a block on a bucket or similar object each time he/she hears a sound (Pitarch *et al*., 2014: 217-228).

2.2. Imaging studies

Imaging tests are an essential element in studies for cochlear implantation, though there is no consensus on the most appropriate study technique. Although a previous CODEPEH document on the etiological diagnosis of hearing loss (Núñez *et al*., 2016a: 193-218) reviewed the preferential indications of both computed tomography (CT) and magnetic resonance imaging (MRI), it is interesting to complement it with the current situation referred to the indications of imaging studies when assessing candidates for cochlear implants.

Historically, high-resolution CT has been the standard imaging test for presurgical cochlear implant evaluation. Although maximum information is obtained with combined high-resolution CT and MRI, this strategy might not be the most efficient way to use the available resources, and moreover exposes children to needless radiation and anesthesia.

Magnetic resonance imaging (MRI) should be the test of choice

Many retrospective studies have examined the combined use of these imaging tests, with diverse conclusions. According to some authors, both tests are important for analysing inner ear status in children with sensorineural hearing loss of unknown origin (Jallu *et al*., 2015: 341-346). Other studies suggest that MRI alone is sufficient, offering direct visualization of the VIII cranial nerve and identifying elements relevant for implantation at inner ear and central nervous system level (Liming *et al*., 2016: 251-258).

A recent review (Siu *et al*., 2019: 2627-2633), in which an algorithm was proposed (Figure 2) for the indication of imaging tests in candidates for cochlear implantation, showed that in many cases MRI alone can be used for preoperative evaluation of children with hearing loss requiring a cochlear implant, since most patients present normal anatomical features, and implantation can be performed without problems. The additional need for a CT scan arises in a small percentage of implant candidates, and can be predicted based on the study of certain characteristics found in the clinical history or physical examination, or directly on abnormalities detected in the initial MRI scan. In these cases, CT is useful for visualizing the trajectory of the facial nerve and for detailing abnormalities in bone anatomy that could make it difficult to find the surgical references for access to the cochlea during implantation.

Reducing unnecessary radiation exposure in children is another important reason for limiting the use of CT. There is a well established relationship between radia-

tion exposure from imaging studies in childhood and the subsequent development of malignancies (Pearce *et al*., 2012: 499-505) and cataracts. This constitutes sufficient evidence to justify limiting exposure as far as possible (Niu *et al*., 2012: 1020-1026).

2.3. Genetic diagnosis

Up to 60% of all cases of congenital or early-onset sensorineural hearing loss are due to genetic factors, commonly manifesting in the absence of a family history of deafness. As reflected in the CODEPEH document of 2018 on universal screening (Núñez *et al*., 2019: 201-220), such programs have limitations in detecting mild and moderate hearing loss, cases of late or sudden onset and/or those progressing after birth. Since many cases of hearing loss of genetic origin cannot be detected by the current universal screening practices, some studies have combined them with genetic screening for the most frequent mutations associated with hearing loss (Wang *et al*., 2011: 535-542). These studies show that the percentage detection of cases of hearing loss using combined hearing and genetic testing exceeds the percentage of cases detected isolatedly by one test or the other independently (Sun *et al*., 2015: 766-770).

The genetic diagnosis offers many benefits. In the future, it will help improve treatment

In genetic diagnostic studies, the most commonly detected alterations are located in GJB2 (connexin 26), which is the most common variant worldwide (Morton and Nance, 2006: 2151-2164).

Mutations of the mitochondrial MTRNR1 gene, which can manifest with hearing loss induced by aminoglycoside treatment, are also relatively frequent (del Castillo *et al*., 2002: 243-249). Although alterations of the GJB2 and GJB6 genes (locus DNFB1) account for 10-40% of the cases (Burke *et al*., 2016: 77-86; Chan and Chang, 2014: E34-53), depending on the population involved, many patients remain undiagnosed after the study of these genes. This is not surprising, since hearing loss is present in over 400 syndromic alterations, and more than 100 genes have been implicated in non-syndromic sensorineural hearing loss. In this regard, next-generation sequencing (NGS) has been a breakthrough in the study of these patients (Sabatini *et al*., 2016: 319-328; Cabanillas *et al*., 2018: 58; Mutai *et al*., 2013: 172).

Considering the recommendations of CODEPEH, once a new case of non-syndromic sensorineural hearing loss has been confirmed, a study of cytomegalovirus (CMV) infection should be made, and genetic screening is advised (Núñez *et al*., 2016a: 193-218). The fact that a genetic diagnosis can be established has many benefits for both the patient and the parents, since it provides information about inheritance, helps distinguish syndromic causes, contributes data on the course of deafness, and could prevent certain triggering factors that cause or worsen hearing loss. In the future, it will help improve treatment, which may be specifically targeted to each detected mutation (Sommen *et al*., 2016: 812-819; Géléoc *et al*., 2014: 1241062; Alford *et al*., 2014: 347-355).

At present, tests based on next-generation sequencing (NGS) in the context of hearing loss are replacing methods based on individual gene sequencing, but are limited by our current knowledge of the genes involved in deafness. Other tests use disease-targeted exon capture, and whole exome sequencing (WES) seeks to find variations in all the exomes of the genome. It therefore could identify genes related to hearing loss that are not yet known. In turn, whole genome sequencing (WGS) is not limited to exons alone but can identify changes outside exons that may be related to hearing loss.

The recommendations of the American College of Medical Genetics and Genomics (ACMG) include genetic study based on the following guidelines: if syndromic hearing loss is suspected, genetic studies targeting suspected genes should be conducted, and if non-syndromic hearing loss is suspected, studies of isolated genes such as GJB2/GJB6, gene panels, or NGS based on clinical suspicion are indicated (Alford *et al*., 2014: 347-355).

The latest recommendations suggest NGS sequencing as a first option, limited to a panel of genes determined by the ethnicity of the study population (D'Aguillo *et al*., 2019: 1-17).

One of the problems with next-generation sequencing (NGS) is the clinical interpretation of the findings. Some studies have observed that up to 30% of all the genetic variants described as a cause of disease in the literature may have been misinterpreted (Boycott *et al*., 2013: 681-691).

Within NGS, genetic panels may be established, targeting a particular disease or alteration, or whole genome sequencing (WGS) may be used (Shearer *et al*., 2013: 627-634). The latter technique increases the risk of secondary findings, understood as identified variants of genes unrelated to the disease or to the reason for performing the genetic test (Green *et al*., 2013: 565-574). Comparisons have been made between WGS and disease-targeted panels in other disorders, e.g., eye diseases, and the targeted panels were found to improve precision. This is the main reason why these panels are used in the etiological study of hearing loss (Consugar *et al*., 2017: 253-261; Rehm *et al*., 2013: 733-747).

Progressive cost reductions, as well as improvement in the quality of WES/WGS (whole genome sequencing),

will probably lead to future replacement of the panels with non-targeted studies (Cabanillas *et al*., 2018: 58). In fact, in some published studies, WES has been used as a method for establishing the cause of hearing loss, combined with other genetic techniques (microarray) (Downie *et al*., 2017: e000119).

2.4. Vestibular study

Children with hearing loss are at an increased risk of developing vestibular disorders (Cushing *et al*., 2008: 1814-1823) due to the close embryological relationship between the auditory and vestibular structures of the inner ear (Tribukait *et al*., 2004: 41-48). Depending on the vestibular function study protocol employed, the etiology and the degree of hearing loss, vestibular disorders affect between 38-91% of all children with sensorineural hearing loss, even when mild or unilateral (Martens *et al*., 2019: 196-201). The vestibular system plays an essential afferent sensory functional role for balance control. Alterations of this system therefore result in imbalance and delays in the motor development of the child. Together with hearing loss, this will negatively affect the development of spatial orientation and attention and, indirectly, will influence self-esteem, psychosocial development, reading, writing and learning ability.

Vestibular screening would allow the early diagnosis of any alterations in motor development and balance

Although the literature alerts to the risk of developing vestibular disorders in children with hearing loss, this function is currently not routinely assessed. The evaluation of vestibular function is usually limited to candidates for cochlear implantation (Jacot *et al*., 2009: 209-217) and children with overt vestibular dysfunction.

The study of vestibular function in general is complex, though it has been found to be feasible in children, with certain adaptations (Dhont *et al*., 2019: 490-493). In order to compensate for the lack of vestibular assessment in children with congenital hearing loss, vestibular screening has been proposed to allow an early diagnosis of disorders at this level, with a view to then conducting due motor assessment and rehabilitation, if necessary. Vestibular screening is performed using a technique called cervical vestibular evoked myogenic potential (cVEMP) testing: it is auditorily stimulated through bone (tone burst from 500 Hz to 59 dBHL) while recording the electromyographic activity of the sternocleidomastoid muscle, consisting of the observation of a reproducible response with two peaks. If the test result proves normal, the child is not subjected to further evaluations, though if it proves anomalous, patient referral is made for the assessment of adequate development (Dhont *et al*., 2019: 490-493).

The cVEMP test can be successfully performed in young children. Although the sensitivity of the test may vary depending on the conditions under which it is performed, it is able to provide diagnostic information in most cases.

The cVEMP technique can be performed in any audiology clinic in which an evoked potentials device is available. It is therefore advised that professionals involved in pediatric audiology should incorporate this technique for the assessment of vestibular disorders, which are so often associated to congenital hearing loss (Zhou *et al*., 2014: 1-6).

3. TREATMENT

3.1. Hearing aid adaptation in infants

The primary aim of hearing amplification is to give infants with hearing problems the opportunity to have access to a sound-enriched environment and especially to speech. The objectives of such amplification include minimal sound distortion, the development of an appropriate signal processing strategy, the selection of features that maximize desired signal audibility and noise reduction, as well as flexibility, ease of connection to external devices, and physical comfort allowing constant daily use.

Hearing evaluation and hearing aid treatments have a number of peculiarities in infants. The age range from 0-6 years is the period posing the greatest difficulties in terms of hearing evaluation and habilitation.

The adaptation of hearing aids and hearing implants is oriented towards the early and optimal development of hearing and listening, speech and language - with speech therapy being an essential element

The external ear of the child is continuously growing, and this means that individualized assessment of the external auditory canal is required through real ear - coupler difference (RECD) measurements. Such measures must be contemplated in the prescription and adjustment of hearing aids. The natural communication environments of children are noisier and more reverberant than those of adults; continuous monitoring of correct use and fitting of the hearing aid by an experienced professional is therefore essential. It is very important and necessary

for the whole process to be focused on the child and the family, with due interdisciplinary coordination (Olleta *et al*., 2018: 91-198) in collaboration with the family.

Thanks to programs for the early detection of hearing loss, the diagnosis is made in the first months of life (Núñez *et al*., 2015: 163-186). This implies an important challenge for the audioprosthetist, who has to deal not only with anatomical difficulties, but also with children who, because of their early age, are unable to indicate whether they perceive sound or not. It therefore will be necessary to guide the evaluation through observation (behavioural audiometry) and the results of objective electrophysiological tests (Kerkhofs and de Smit, 2013: 17-25).

If there is a hearing neuropathy spectrum disorder, and provided it cannot be confirmed that the patient responds to conversational speech without hearing aids, we should establish a test period with hearing amplification aids to observe the response. Hearing aid adaptation is advised before deciding a cochlear implant (American Academy of Audiology Clinical, 2013: 5-60; British Society of Audiology, 2019: 21).

That unilateral deafness is not without negative consequences for the child's development, also requiring treatment and follow-up

An appropriate digital signal processing hearing aid should be chosen, taking into account the size and shape of the external ear. In this regard, because of the very rapid anatomical changes occurring in the external auditory canal, the optimal choice is the use of behind the ear (BTE) hearing aids. These hearing aids can be adapted to the external auditory canal by means of a custom-made closed mould that should be changed as the ear grows. It is advisable to regularly check the integrity of the connection between the ear mould, tube, elbow and hearing aid. In the case of older children there are other more aesthetic options than BTE hearing aids. However, it is important to be aware that they cannot be connected to magnetic induction, FM or equivalent systems. This implies significant restrictions in acquiring information from the environment, particularly in the educational setting, and in benefiting from accessibility measures.

Another factor to be considered is the microphone. The use of omnidirectional microphones is recommended, since directional microphones can reduce the audibility of people speaking outside their reach. This limits incidental learning at a time particularly important for language development and other learnings.

In terms of safety, it is advisable to use tamper-proof battery carriers in order to reduce the risk of battery swallowing. Likewise it is advisable to deactivate or block the volume controls, or to use wide dynamic range compression - eliminating the need for volume adjustment for audibility and comfort. The bandwidth used must allow for maximum versatility in the amplification of all frequencies, creating programs that will gradually adapt to the daily life context of the child. If the magnetic induction coil is used, it is important for it to be activated automatically. The connectivity options now available for hearing aids are also of interest, though always in conjunction with the induction coil, since this is the system that provides the child with the hearing accessibility needed in public spaces such as educational centres and cultural and leisure areas, etc.

The RECD (real ear - coupler difference) values must be taken into account in fitting the hearing aid. The RECD values may vary substantially from infant to adult age (children usually have greater RECDs than adults) (Zenker, 2001: 10-15). These values allow checking of compliance with the technical characteristics of the hearing aids, verifying their correct repair, measuring variations in the different parameters after modifying their controls, and allowing the objective evaluation of hearing aid adaptation by performing the measurements in the real ear.

The discomfort threshold defines the limit of amplification, and is one of the most important aspects in adaptation of the hearing aid (Calvo and Maggio de Maggi, 2003: 57-59). In the case of pediatric adaptations, the most recommended gain prescription methods are DSLv5a and NAL-NL1. In addition, compression in the dynamic range is recommended, minimally altering the speech signal. High frequency bandwidth amplification will improve the audibility of acute spectrum sounds.

Electroacoustic verification of adaptation is essential. The lack of patient cooperation and the impossibility of checking what is expressed by the child make the use of objective tools very important. Free field use of hearing aid functional gain is not recommended in young children and should not be used for checking the adaptation of hearing aids. It also must be taken into account that microphone probe measurements using real ear insertion gain (REIG) protocols are likewise not recommended. The electroacoustic characteristics of the hearing aid then must be verified, comparing gain prescription and maximum output pressure in the real ear of the patient or in a 2-cc coupler (Calvo and Maggio de Maggi, 2003: 57-59). Hearing aids must be validated on a continuous basis, since at these ages it is common for a loss of transmission due to middle ear problems to be added to the sensorineural hearing loss. The possibility of progressive hearing loss also must be taken into account. Due consideration is required of the hearing age of the child, the chronological age and the medical history, together with the evaluation of patient speech, cognitive and psychomotor development before and after adaptation. The correct localization of sound with hearing aids, functional frequency discrimination (Ling sounds), and appropriate discrimination in noisy environments also need to be explored.

Advice to parents and their instruction in the management of hearing aids and in the checking of their proper function must be continuous. Caregivers, speech therapists and teachers should also be involved in these aspects (Olleta *et al*., 2018: 91-198).

3.2. Indications of cochlear implantation

Although audiometric criteria remain the most important factors in the decision to place a cochlear implant, technological advances and the development of surgical techniques have profoundly changed the indications of cochlear implantation, with the opening up new perspectives in this field (Manrique *et al*., 2015: 289-296).

Binaural hearing is fundamental for hearing processing

The indications of cochlear implantation in childhood are well established and accepted in the case of bilateral severe to profound sensorineural hearing loss, with little benefit from the use of hearing aids after a trial period. However, there are further indications that must be taken into account in pediatric patients: some are already established, such as bilateral implantation, while others are gaining support from scientific evidence, such as implantation in cases of unilateral deafness. All this is fundamented upon the well known benefits of binaural hearing: improved comprehension in noisy environments, and greater ability to localize sounds in space. The existing studies show that if possible, simultaneous implantation is preferable, and if this does not prove possible, then the time interval between implantation in both ears should be a brief as possible.

Another consolidated indication is cochlear implantation in asymmetric hearing loss. These patients have moderate to severe sensorineural hearing loss in one ear and profound hearing loss in the other, and thus could simultaneously use the implant in the ear with the poorest hearing, and a hearing aid in the other ear. This form of stimulation is known as a dual mode strategy. With this strategy it has been shown that patients of this kind achieve stereophonic hearing and improved levels of language discrimination, both in a quiet and in a noisy environment, compared with those who use hearing aids or only a cochlear implant.

However, cochlear implantation in unilateral deafness is not yet a consolidated indication. To date, the management approach to patients with unilateral deafness was no treatment; the use of a bone conduction implant; or the use of a hearing aid with a contralateral routing of signal (CROS) system. Cochlear implantation constitutes a new alternative for certain patients. Cochlear implants have been shown to offer superior performance versus the rest of the alternatives, with no interferences with contralateral normally hearing ear. This indicates that central integration of electrical and acoustic stimulation is possible, even with contralateral normal hearing. One of the reasons for advocating cochlear implantation in children with unilateral deafness is to facilitate complete development of the central auditory system within the critical or more sensitive period, corresponding to the first years of life. This indication is considered particularly interesting in children with important vision problems or with fragile conditions in the normally hearing ear, such as certain labyrinthine malformations (dilation of the vestibular aqueduct, incomplete cochlear partitions, etc.).

Despite the above, a meta-analysis (Peters *et al*., 2016: 713-721) has indicated that there is little evidence for cochlear implantation in unilateral deafness, and that the supporting data are limited to case series; no firm conclusions therefore can be drawn regarding the effectiveness of implantation in unilateral infant deafness. Nevertheless, based on the findings of this systematic literature review, the promising results obtained in adults, and the importance of binaural hearing, suggest that cochlear implantation in children with unilateral hearing loss may be effective.

The indications for cochlear implantation have evolved, and new perspectives have been opened in this regard

Auditory neuropathy or auditory dissynchrony is a hearing disorder that is difficult to diagnose and is characterized by a varied clinical behaviour. Hearing aids in this case offer only limited benefit. In turn, cochlear implants afford variable results, since when the lesion is located in the nerve (demyelination), electrical stimulation may have the same limitations as acoustic stimulation. However, 75% of all neuropathies are known to be due to presynaptic alterations of inner hair cell function - a circumstance that would allow optimal performance with an implant.

Children with hearing neuropathy require specific evaluation. Because each child with hearing neuropathy is different, a number of clinical situations arise that need to be taken into account. A first clinical situation refers to a child with normal otoacoustic emissions or robust cochlear microphonic potentials, but with poor hearing thresholds. In these cases, the hearing thresholds may possibly recover over time, indicating the recovery of neural function. A second clinical situation is that arising from the identification of behavioural hearing thresholds not compatible with severe or profound hearing

loss (which do not meet audiometric criteria for implantation), but in patients which nevertheless suffer very important delays in speech and language development with respect to what would be expected from their hearing situation. This context illustrates the great dilemma which sometimes occurs when indicating an implant in these patients with seemingly good thresholds, since it is not clear whether the delayed speech and language development is caused by the hearing disorder or by neurocognitive comorbidities.

These clinical situations serve to explain the delay in implantation observed in children with auditory neuropathy, who are implanted later (at a mean age of 3.3 years) compared to children without this disorder (who are implanted at 1.9 years of age). This monitored waiting period is necessary, because the audiometric thresholds suggest that hearing aids will suffice for speech and language development. Since the critical period of 5 years of age is not exceeded, this delay does not seem to be detrimental in terms of the expected outcomes of implantation (Harrison *et al*., 2015: 1980-1987).

3.3. Future lines

New genetic techniques have been developed, such as clustered, regularly interspaced, short palindromic repeats (CRISPR) associated to nuclease 9 (CRISPR/cas 9), which are very potent tools both for the study of genes and for the treatment of genetic disorders. One of the main examples would be hearing loss of genetic cause, since most such cases are due to single-gene mutations (Zou *et al*., 2015: 102-108).

New genetic techniques allow the study of gene functions causing hearing loss

This technique allows gene editing (adding, stopping or changing gene sequences), as well as the elimination of genes or the introduction of mutations to observe the effect. In fact, experiments are being conducted in mice, attempting to edit and correct genes causing a form of inherited dominant genetic hearing loss (Gao *et al*., 2018: 217-221). These techniques open up the possibility of developing gene- or cell-based treatments in the future that could preserve or restore hearing with more natural sound perception. Such studies have been guided by similar research in eye therapy, since both organs have many similarities (Zhang *et al*., 2018: 221) and allow us to envisage a very promising future.

4. MONITORING

In order to establish a monitoring protocol, a first essential requirement is an adequate information system.

In the case of children detected in the screening phase, their degree of hearing loss must be known with certainty before 6 months of age, reliably determining hearing thresholds and establishing an etiological diagnosis. After that moment, a periodic review program should be established with the pediatric otolaryngologist.

It must be taken into account that hearing loss does not remain constant throughout childhood, and that hearing deterioration can occur at any time. This situation is more difficult to perceive in the first years of life, since infants do not have the capacity to explain whether they hear worse, and clinicians therefore must be guided by the subjective impressions of the caregivers. This phase is particularly sensitive, because it is the time when the child is acquiring language skills (Núñez *et al*., 2015: 163-186).

Reviews are advised with the following frequency:

- Within the first 18 months of life, on a continuous basis as required by each situation
- From 18 months to 3 years, every 3 months
- From 3 to 6 years, every 6 months
- In the case of children over 6 years of age with stable deafness, annual reviews are indicated

Special care should be taken with children presenting hearing loss secondary to cCMV infection and with cochlear malformations, such as a dilated vestibular aqueduct (Lin *et al*., 2005: 99-105), since they are at an increased risk of worsened hearing. Strict monitoring is required in children with hearing neuropathy spectrum disorders, due to their difficulties in terms of language intelligibility, fluctuation and worsening of hearing (Hood, 2015: 1027-1040).

Children with deafness secondary to cCMV infection require special attention

Regardless of the established protocols, parent perceptions of possible changes or difficulties in their children should always be taken into account (Fitzpatrick *et al*., 2016: 34-43).

In the case of children at risk of developing deafness due to risk factors, the timing and number of hearing reviews or checks should be individualized according to the relative likelihood of late onset hearing loss. Children who pass neonatal screening but have a risk factor should undergo at least a diagnostic hearing assessment between 24 and 30 months of age. Early and more frequent assessment may be particularly indicated in the presence

of certain risk factors, as recommended by several publications (Núñez *et al*., 2015: 163-186; Joint Committee on Infant Hearing Pediatrics, 2007: 898-921).

In the course of the life of the child, multiple causes can lead to deafness, in the absence of prior risk factors. The CODEPEH has produced recommendations (Núñez *et al*., 2015: 163-186) to detect hearing problems in infants between 6 months and 4 years of age based on a simple questionnaire to be completed by the pediatrician. The American Academy of Pediatrics recommends audiometry for all children at 4, 5, 6, 8 and 10 years of age.

Children with deafness secondary to cCMV infection require special attention

In addition, in children over 10 years of age, the assessment of frequencies of 6000 and 8000 Hz is advised at least once between 11 and 14 years of age, between 15 and 17 years of age, and between 18 and 21 years of age. This strategy improves the outcomes referred to early detection and intervention in the absence of risk factors, compensating the costs and inconveniences.

4.1. Information systems

Hearing impairment is a clear example of how convergence is needed among the specific aspects of each administrative sector involved in the pediatric age (Health, Social Services and Education), together with the different professionals (paediatrician, ENT doctor, audiologist, speech therapist, teacher...), with intervention from an interdisciplinary and comprehensive perspective targeted to the child and family, and adopting planned joint, coordinated and convergent actions referred to resources and services (Jáudenes, 2014).

In order to ensure effective and efficient organization, such crucial convergence requires a database accessible to all those involved, to facilitate the exchange of information, and an analytical system allowing the study of this information to identify the strong and weak points of the screening programs with a view to correcting defects and improving the outcomes.

These information systems are intended to guide activities, help with planning, ensure implementation and support program evaluation, and are moreover useful in formulating research hypotheses. They can range from simple single-source data collection systems to electronic systems receiving data from many sources and in different formats, ensuring compliance with the applicable data protection regulations.

The information collected will serve not only to improve the services provided for both children and families, but also to monitor the quality of screening, diagnosis and early treatment, as well as to facilitate the compilation of demographic data on congenital hearing loss, as outlined by the CODEPEH in previous recommendations (Trinidad *et al*., 2010: 69-77; Núñez *et al*., 2015: 163-186). An absolute need in this respect is the existence of a single national database in Spain. However, to date no central registry collects the information of the different early hearing loss detection programs which all the different regions (Autonomous Communities) in the country are applying - though these regions do have their own individual information systems. Since these databases probably have matching fields, they would easily allow for the creation of a common database.

In fact, a very convenient option would be to integrate all of the current neonatal screening programs into a single database, making data registration and analysis easier for the staff involved (Feresin *et al*., 2019: 193-199).

These information systems also contribute to improve screening programs - the weak point of the latter being the follow-up dropout rate of children with altered results or the absence of records of cases that may have received a diagnosis and/or treatment, and in which the result has not been reported. In order to correct this situation, it is absolutely necessary to have reliable and accessible patient contact details in order to allow the recovery of these individuals.

Pediatric monitoring allows the early detection and diagnosis of hearing loss occurring between 6 months and 4 years of age

Another cause of loss related to the information systems is when children entered in the program move from one geographic area to another between initial screening and medical monitoring, since the responsibility for monitoring also moves from one centre to another. As a result, these cases are lost to follow-up for the centre that had initial responsibility for monitoring the patient. This reinforces the recommendation that the database should be a national registry (Matulat *et al*., 2017:1008-1013; Chung *et al*., 2017: e227-231).

In order to reduce these losses in monitoring, it is essential for the database to automatically generate alerts and facilitate the recovery of these patients (Ravi *et al*., 2016: 29-36).

In this regard, recent data from countries with years of experience in screening have identified the weaknesses that most often need to be corrected in information

systems in order to improve the effectiveness of the program. These amendments involve ensuring adequate data referral, the definition of a program coordinator, the creation of a common webpage, data standardization and verification, and software adequacy (Greczka *et al*., 2018: 13-20).

On the other hand, in relation to screening programs, there are shortcomings in their planning, as well as organizational difficulties and problems referred to the availability of resource that impede the guarantee of diagnostic confirmation and access to quality early intervention. Without a database containing information on the screening results, it is impossible to know these difficulties and correct those planning or execution errors that prevent us from reaching the desired objectives.

4.2. Information and family support. Early management

As previously commented, one of the most positive and outstanding aspects of the program for the early detection of infant hearing loss, approved in Spain by the Ministry of Health and the Autonomous Communities in 2003, is the incorporation of a monitoring phase as an essential part of the process, and which includes the early management of children with hearing loss and their families (Public Health Commission, Ministry of Health and Consumer Affairs, 2003).

For nearly two decades now, in our country we have evidenced a significant and differentiating qualitative leap between previous generations of deaf people and the deaf children and young people of today. Application of the program for the early detection of hearing loss, and the medical, audiological and prosthetic advances, are clearly and directly responsible for this change (Jáudenes. *et al*., 2007: 51-53; Silvestre and FIAPAS, 2010: 75-79; FIAPAS [vvaa], 2017). While it is true that the early detection and diagnosis of hearing loss have substantially changed the educational and social perspective of these children, they must be followed by two decisive actions in order to be truly effective:

- Prompt initiation of optimum hearing aid measures and speech therapy in each case, facilitating early and natural access to spoken language; and
- Provision of global care focused on the child and family, avoiding divided, decontextualized and uncoordinated responses, giving parents trust and confidence, lessening their disorientation and the need to go through various specialists and departments.

Thanks to early hearing stimulation, which takes advantage of the critical neuronal plasticity period, and the acquisition of verbal language from the environment at the pertinent evolutive moment, sharing communication codes with the family - since over 95% of these children are born into hearing families (Mitchell and Karmchmer, 2002) - infants with hearing loss reach global and communicational development comparable to that of their hearing peers, and can access schooling having acquired the necessary cognitive and language skills to do so. This in turn explains the highly significant differences between early-stimulated deaf people and those who have received specific management later in time and/or inadequately - thereby seriously compromising their learning (Jáudenes *et al*., 2007: 51-53).

A national information system is essential for effective monitoring, and allows evaluation of the programs and the definition of lines of research, the planning of services and resources, etc.

The aims of an early hearing loss detection program therefore extend beyond mere detection and diagnosis. The ultimate and desired objective must be early, planned and coordinated management directed on one hand to stimulate and enhance global development of the child - in this case through two types of intervention: prosthetic adaptation and specialized speech therapy (hearing stimulation and language development) - and on the other to support the family and strengthen its skills as first educators, through emotional support and counselling.

From the analysis of the data collected on a representative sample of 600 families in Spain, participating in a study carried out by the Spanish Confederation of Families of Deaf People-FIAPAS (Jáudenes, 2006: offprint), it is highlighted that one of the main difficulties faced by parents in the early stages, after receiving the diagnosis, is the need to assimilate the news of their child's deafness, and the lack of information. Thus, in relation to communication with the family, not only the message itself but also the way and environment in which it is transmitted are important. We must remember that, particularly in the moments when hearing loss is confirmed, the attitude of the professionals dealing with the family and providing the information should be characterized by key elements such as listening, impartiality, objectivity, clarity and accessibility (Federación Estatal de Asociaciones de Profesionales de Atención Temprana, 2011: 19-38). Families therefore should be able to count on stable teams and structures characterized by specialized and qualified professionals able to analyse the family situation, considering the individuality of each case and family, as well as the life and social circumstances, and establishing the most appropriate intervention program (Jáudenes, 2012a: 11-24).

This evidences the need for a global approach, immediate to the time of diagnosis, with referral routes easily identifiable by the family. At the same time, we again must underscore the importance of cooperative and co-responsible interdisciplinary intervention, with the involvement of the Family Association Movement (Jáudenes, 2012b: 177-189; Jáudenes and Patiño, 2013: 288-302).

An interdisciplinary team is essential, working in collaboration with the family, and with the support of the Family Association Movement

Optimum results are achieved when programs for the early detection of infant hearing loss achieve high quality levels at all stages, from suspicion to diagnostic confirmation and provision of the most appropriate treatment in each case - including prosthetic adaptation and early care (Yoshinaga-Itano, 2014: 143-175). Ensuring that the process is carried out, under quality standards that guarantee its effectiveness, must be an obligation for all parties involved. In sum, the detection, diagnosis, monitoring and early care process must be set within a continuum that requires qualified and experienced professionals, as well as resource and service planning with coordinated and undivided and non-decontextualized responses (Marco *et al*., 2004: 103-106; Joint Committee on Infant Hearing, 2007: 898-921; Trinidad, 2010: 69-77; Núñez *et al*., 2015: 163-186).

Lastly, it should be noted that the implementation of a program for the early detection of infant hearing loss mobilizes and integrates different resources, actions and procedures, as well as the necessary management and evaluation of outcomes. Beyond a first level of clinical and methodological evaluation, which must meet certain quality standards, the future development of knowledge and measurement of the efficacy of the program must include the functioning and effectiveness of the referral circuits, the communicational and educational level reached by the children with an early diagnosis, and the level of satisfaction expressed by their families.

We thus need to integrate family management and support as part of the development of the programs for the early detection of infant hearing loss, including mutual help among families, family involvement at each intervention level, and increased knowledge of the role of the family among all the parties involved. Recognizing and responding to the needs of the families is the responsibility of all the levels and professionals implicated in these programs. Likewise, some studies (Benito, 2017: 85-103) indicate that from a clinical and healthcare perspective, knowing the development of language skills acquired by the children with an early diagnosis of prelingual hearing loss, and following a standardized intervention protocol, inherently constitutes an indicator for validation of the diagnostic and treatment process in infant hearing loss.

4.3. Interdisciplinary team

A number of professionals from different settings must intervene in the evaluation, treatment and monitoring of children with hearing loss: ENT specialists, pediatricians, prosthetic audiologists, speech therapists, pedagogues and teachers, psychologists and sometimes also other specialists (Figure 3), integrated within a multidisciplinary team and working in coordination with the family (Joint Committee on Infant Hearing, 2007: 898-921; Jáudenes, 2012a: 11-24; Jáudenes, 2012b: 177-189; Núñez *et al*., 2015: 163-186).

The interdisciplinary team must provide quality information regarding medical diagnosis and treatment, as well as instruction on the use of hearing devices, the rehabilitation process to be followed, and the support structures and resources for families. The message must be coherent and consistent among the different professionals involved. Family-centred care requires the family to be involved in the child's hearing health throughout the process, though the family should not be responsible for transmitting information between professionals or for mediating in their coordination (Findlen *et al*., 2019: 141-146).

Inter-administrative and inter-sectorial coordination is also needed with regard to the services for the children and their families, with simplified and coordinated procedures for access to all of them. For this purpose, the coordinator and the participation of Public Health, together with the Educational and Social Services Administration, are crucial elements. In turn, the Family Association Movement acts as a social network and agent, developing family support programs and forming an integral part as an element of support and cooperation within the interdisciplinary activities.

5. CODEPEH RECOMMENDATIONS 2019

The CODEPEH considers it necessary to establish recommendations to complete and update the implementation of programs for the early detection of infant hearing loss, improving their performance in accordance with the following sequence referred to the detection, diagnosis, treatment and monitoring process (Figure 4).

LEVEL 1. DETECTION

- Either of the two available screening techniques (otoemissions and/or automated potentials) are appropriate in this phase.
- Proper identification of the hearing risk factors is needed to ensure optimal subsequent monitoring.
- The study of congenital cytomegalovirus (cCMV) infection is recommended in children who do not pass hearing screening.
- The detection of cCMV must be performed before 15 days of life.
- Passing neonatal screening does not rule out the possibility of developing late or sudden hearing loss.
- Combined hearing, genetic and cCMV screening would overcome the limitations of current screening practice.

LEVEL 2. DIAGNOSIS

- A correct examination and medical history remain the cornerstone of diagnosis.
- A single hearing test is not enough for proper diagnosis and treatment. Steady state auditory evoked potentials (SSAEPs) constitute a safe and essential test.
- A diagnosis of cCMV beyond 21 days of life requires confirmation, based on a biological sample in the neonatal period.
- In the case of known syndromes, genetic study targeting the causal genes is advised. For non-syndromic deafness, next-generation sequencing panels are recommended.
- Magnetic resonance imaging (MRI) should be the test of choice, with the individualized consideration of a supplementary computed tomography study..
- It is advisable to assess vestibular function in all children with hearing loss, based on cervical vestibular evoked myogenic potential (cVEMP) testing.

LEVEL 3. TREATMENT

- After diagnosis, it is essential to start the appropriate treatments in each case as soon as possible, from a global approach, focused on the child and family.
- Early speech therapy is essential for auditory stimulation and the development of spoken language, as well as for the cognitive and learning skills that derive from them.
- Hearing aid adaptation must be carried out at no more than 6 months of age, prescribing BTE aids with a magnetic induction coil.
- A trial period with hearing aids is recommended before making a decision about cochlear implantation.
- There are new indications regarding cochlear implantation referred to unilateral hearing loss and the type and severity of hearing loss.
- In the future, thanks to cochlear gene therapy, treatments will be developed that can preserve or restore hearing..

NIVEL 4. MONITORING

- The ultimate aim of an early deafness detection program should extend beyond the diagnosis, focusing on early, planned and coordinated care.
- Interdisciplinary teams, working in collaboration with the family and organized by a coordinator, are required at all levels of the program.
- The Family Association Movement must be a point of support in all the stages, as part of the interdisciplinary team.
- A national information system, accessible to the professionals involved, is needed to facilitate the exchange of information and know the state of the art, unify protocols, reduce losses during the process, and improve the development and outcomes of the program through ongoing evaluation.

6. FIGURES

FIGURE 1. Diagnostic sequence (Núñez *et at.,* 2016a: 193-218)

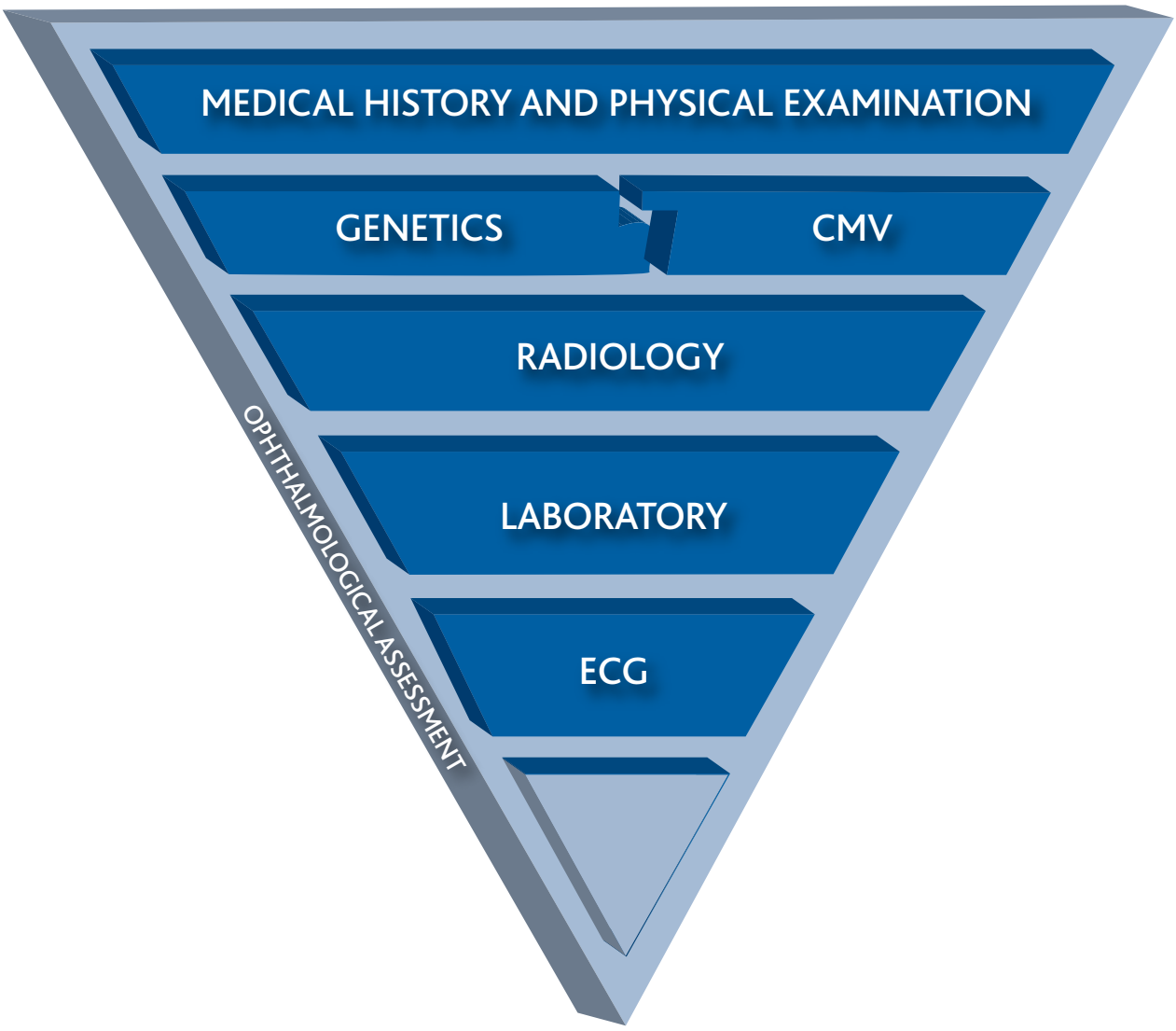


FIGURE 2. Algorithm of an MRI-based imaging protocol (Siu *et al.*, 2019: 1-7)

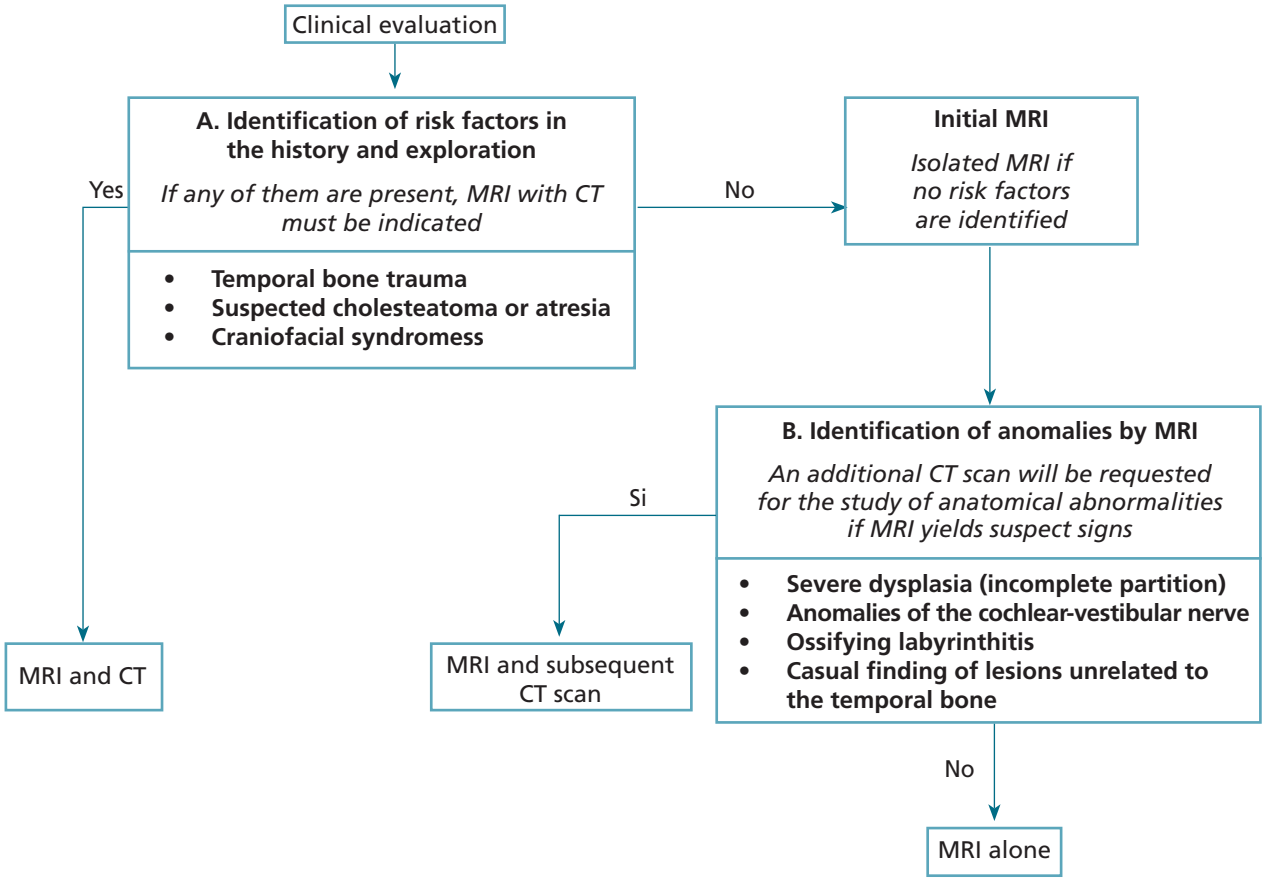


FIGURE 3. Interdisciplinary coordination. Infant hearing screening programs

CINTER-DISCIPLINARY COORDINATION PROGRAMS FOR THE EARLY DETECTION OF INFANT HEARING LOSS[1]

OEAR, NOSE AND THROAT SPECIALIST

- Hearing assessment
- Diagnosis and medical orientation
- Medical and/or surgical treatment
- Prescription of hearing aids / implants
- Monitoring

PEDIATRICIAN

- Verification of the screening process
- Control of children with risk factors
- Assessment of hearing and communicational development
- Referral to ENT specialist
- Monitoring of health and development

COCHLEAR IMPLANT PROGRAM

- Evaluation of candidates
- Surgery
- Programming
- Monitoring

PROSTHETIC AUDIOLOGIST

- Hearing aid study and adaptation
- Supporting products. Inductive coil activation for hearing accessibility in the environment
- Monitoring

SPEECH THERAPIST

- Evaluation, diagnosis and intervention in communicational and spoken language development
- Functional hearing evaluation. Hearing stimulation and training
- Support of the reading-writing acquisition and development process
- Monitoring

OTHER PROFESSIONALS IN THE FIELDS OF HEALTH, EDUCATION AND SOCIAL SERVICES

(Ophthalmologists, Geneticists, Neuropediatricians, Teachers, Pedagogues, Psychologists, Social Workers, etc.)

FAMILY ASSOCIATION MOVEMENT

- Information and counselling
- Training
- Family care and support service. Interfamily mutual aid program (guide fathers and mothers)
- Early management and specialized speech therapy intervention
- Monitoring

HEALTHCARE

- Public health. Program for the early detection of infant hearing loss
- Healthcare and hearing aid provision
- Medical-functional rehabilitation (speech therapy)
- Information systems
- Other services related to early management

EDUCATION

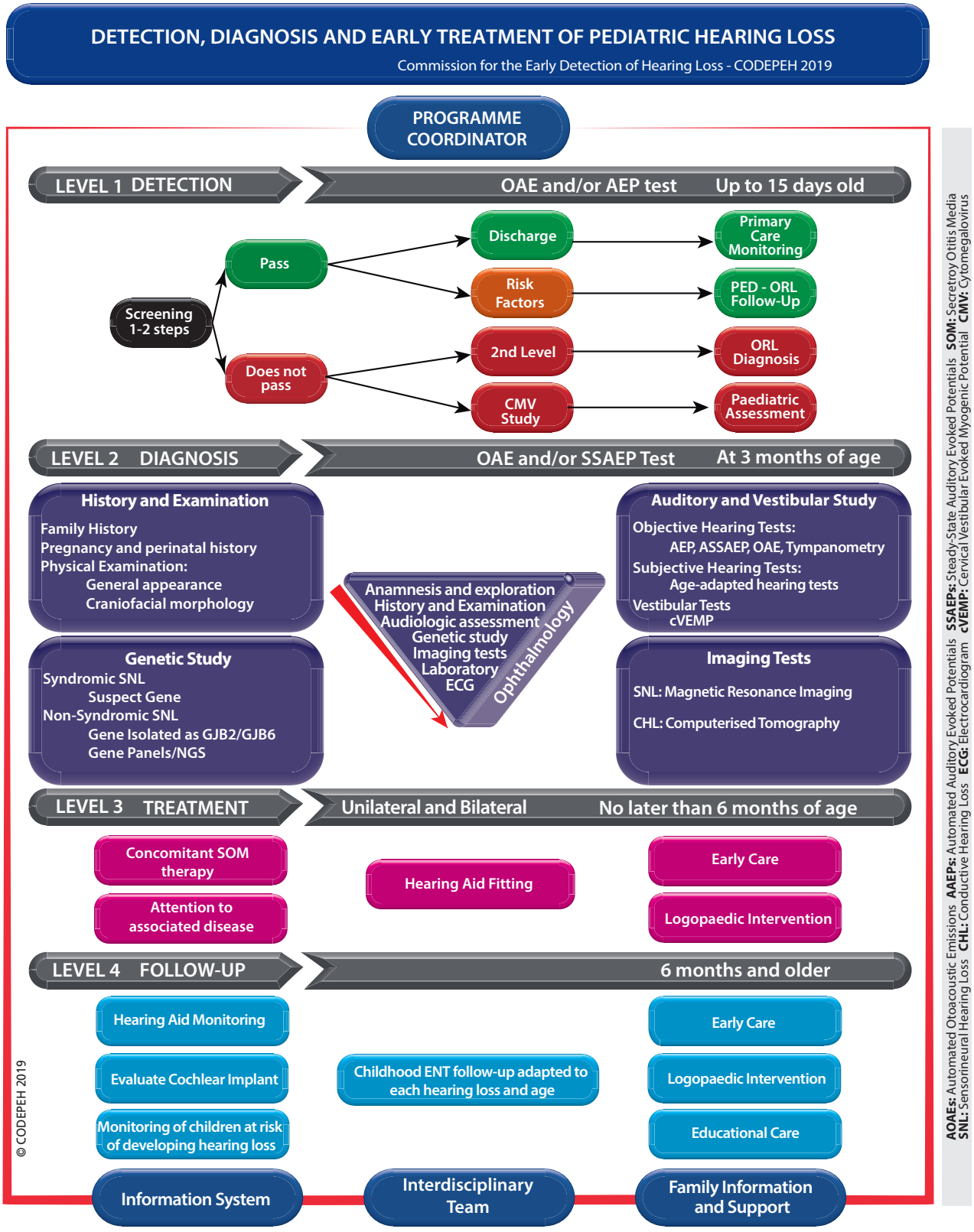
- Psychoeducational assessment
- Schooling
- Specialist intervention Hearing Language and Therapeutic Pedagogy
- Accessibility of information and communication in schools, as well as in complementary and extracurricular activities

SOCIAL SERVICES

- Assessment of the degree of disability
- Economic and social support
- Other services related to early management

[1] Adapted and updated from FIAPAS and CODEPEH (1998): "Do not wait. From diagnosis to treatment of hearing impairment" (Brochure). Madrid: FIAPAS 2017 (20th edition)

FIGURE 4. Update of the infant hearing screening programs



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