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Hearing Loss in Children with Associated Disabilities (AD+): CODEPEH Recommendations 2021

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FIAPAS
CONFEDERACIÓN ESPAÑOLA DE FAMILIAS DE PERSONAS SORDAS
Declarada de Utilidad Pública
FOMENTANDO INCLUSIÓN. APOYANDO PERSONAS. AVANZANDO SOLIDARIAMENTE.

SPANISH CONFEDERATION OF FAMILIES OF DEAF PEOPLE

MINISTERIO DE SANIDAD, CONSUMO Y BIENESTAR SOCIAL
REAL PATRONATO SOBRE DISCAPACIDAD

SPANISH MINISTRY OF HEALTH, CONSUMER AFFAIRS AND SOCIAL WELFARE
ROYAL PATRONAGE FOR DISABILITY

RESUMEN

Aproximadamente el 40% de los niños con sordera tienen añadido un trastorno del desarrollo o un problema médico importante, que puede retrasar la edad de diagnóstico de la hipoacusia y/o precisar de la intervención de otros profesionales. Esta situación se designa como “hipoacusia o sordera con discapacidad añadida” (DA+). El motivo por el que la población de niños con problemas auditivos es más propensa a asociar discapacidades añadidas (40% versus 14% en la población oyente) radica en que los factores de riesgo para la hipoacusia se superponen con los de muchas otras discapacidades. Estos factores pueden influir en diversos aspectos del desarrollo, incluida la adquisición del lenguaje. Es importante comprobar que se recibe la adecuada atención, la efectividad de audífonos o implantes, así como de las estrategias de intervención logopédica, y la adherencia de la familia a sesiones y citas. Los desafíos que plantea la DA+ son su detección precoz, para permitir una temprana y adecuada intervención, y la necesidad de una colaboración transdisciplinar fluida entre todos los profesionales que han de intervenir, junto con la implicación de la familia.

PALABRAS CLAVE

Sordera, discapacidad añadida, desarrollo del lenguaje, trastorno del desarrollo, discapacidades, intervención temprana, transdisciplinariedad.

SUMMARY

Approximately 40% of children with deafness have an additional developmental disorder or major medical problem, which may delay the age of diagnosis of hearing loss and/or require intervention by other professionals. This situation is referred to as “hearing loss or deafness and additional disability” (AD+). The reason why the population of hearing-impaired children is more likely to have associated additional disabilities (40% versus 14% in the hearing population) is that the risk factors for hearing impairment overlap with those for many other disabilities. These factors can influence various aspects of development, including language acquisition. It is important to check that appropriate care is received, the effectiveness of hearing aids or implants, as well speech therapy intervention strategies, and family adherence to sessions and appointments. The challenge posed by AD+ is early detection, to allow early and appropriate intervention, and the need for fluid cross-disciplinary collaboration between all professionals involved, together with the involvement of the family.

KEY WORDS

Deafness, deafness with additional disabilities, language development, developmental disorder, disabilities, early intervention, cross-disciplinary.

CONTENTS

1. INTRODUCTION

Neonatal hearing screening programmes for hearing loss have led to major advances in the early identification and treatment of children with hearing loss, resulting in improved speech and language development (Kennedy *et al.,* 2006).

Although the audiometric threshold is an important factor influencing the language development of a deaf child, it is not the only one. There are other variables that must be taken into account such as sex, cognitive skills, the presence of an additional disability, the educational or socio-economic level of their parents, the age at which they were diagnosed and received audiological treatment, the type of device chosen to alleviate hearing loss, the quality of their fitting and their appropriate use (Cupples *et al.,* 2018b).

Approximately 40% of children with deafness have an associated developmental disorder or major medical problem which, in addition to delaying the age at which hearing loss is diagnosed in many cases, also requires the intervention of other professional specialists (Gallaudet Research Institute, 2008).

The situation of these children is known in the literature as being "hard of hearing or deaf plus" (Wiley *et al.,* 2021), although it seems more appropriate in our language to use the term "hearing loss or deafness and additional or associated disability", applicable both to diagnosed deafness accompanied by other disorders, and to diagnosed disabilities associated with deafness.

The percentage of children with hearing loss with an additional disability is increasing considerably in number and diversity, posing a major challenge for early detection and diagnosis programmes, and early intervention for childhood hearing loss (Jackson *et al.,* 2015).

The reason why children with hearing problems are more likely to have other additional disabilities (40% versus 14% in the hearing population) is that risk factors for hearing loss (Núñez-Batalla *et al.,* 2012) overlap with risk factors for many other disabilities, such as the presence of certain genetic syndromes, prematurity, congenital infections, and meningitis. These factors can disrupt various aspects of development as well as language acquisition.

One of the challenges posed by the association of a disability with hearing loss is early detection, making the assessment of speech and language development in any child with deafness every 6 months extremely important, since identifying an additional disability to deafness allows for prompt and appropriate intervention. Another challenge to be addressed is the need for fluid, coordinated cross-disciplinary collaboration by all professionals involved.

This CODEPEH Recommendations document analyses hearing loss children with additional disability, as well as disability associated with deafness, as well as the effect that both entities (AD+) have on language development and strategies for early identification and treatment, with the aim of trying to reduce the gap between these situations and deaf children without additional disability.

2. INCIDENCE OF AD+ ACROSS CLINICAL CONTEXTS

2.1. Diagnosed deafness and other associated disabilities

Data published in the 2017 global morbidity study on the prevalence of childhood epilepsy, intellectual disability, and vision and/or hearing loss show that 11.2% of children and adolescents worldwide had one of these four disabilities. The prevalence increased with age, from 6.1% among 1-year-olds to 13.9% among 15- to 19-year-olds. Although 94.5% lived in low- and middle-income areas and countries, predominantly in South Asia and sub-Saharan Africa, this problem is not unknown in developed countries (Olusanya *et al.,* 2020).

Approximately 40% of children with deafness have an associated developmental disorder or major medical problem.

The most common disability associated with hearing loss is intellectual (ID), which occurs in 8.3% of these children (Gallaudet Research Institute, 2013).

It has also been shown that the overall burden of developmental disabilities has not significantly improved since 1990, suggesting inadequate attention to children's developmental potential (Global Research on Developmental Disabilities Collaborators, 2018).

As part of the process of assessing congenital hearing loss, we must take into account that approximately 30% to 40% of children have an additional disability (mainly cognitive impairment) and about 20% of this population have more than two. For appropriate language development in the event of hearing loss, not only is early diagnosis and treatment of hearing impairment required, but other non-hearing factors influencing this progress must

also be excluded. In a Spanish study of children with deafness diagnosed at less than 3 years of age, 22% had psychomotor delay from this early age (1 in 4 to 5 children) (Benito-Orejas *et al.,* 2017).

Communication and language development may be more delayed in children with hearing loss associated with one or more disabilities, making it important to identify this population (Bruce and Borders, 2015) as the additional disability is often identified later in children with hearing loss than in those with normal hearing (Wiley and Meinzen-Derr, 2013).

The term "deafness and additional or associated disability" (AD+) applies to both diagnosed deafness accompanied by other disorders, and diagnosed disabilities with associated deafness.

Though not intended to be an exhaustive list, the most prominent syndromes that may be associated with deafness are described below, distinguishing between those that do not affect cognitive skills and those with intellectual disability:

a) With intellectual disability (to varying degrees)

Down syndrome (DS)

The prevalence of hearing loss in Down syndrome ranges from 2% to 78%, since they usually have chronic otitis media (Jackson *et al.,* 2015). Despite this, and with adequate audiological management, less than 2% have permanent hearing loss (Shott and Heithaus, 2001).

Usher syndrome (US)

This is the most common condition affecting both hearing and vision, with three clinical subtypes. It occurs in 3-6% of children with congenital deafness and up to 50% of the population with deafness and blindness (Liu *et al.,* 2008).

Any child with severe-deep bilateral neurosensory hearing loss should have an ophthalmology assessment to rule out Usher syndrome (American Academy of Pediatrics, 2007). Children with type I US have absent or limited vestibular function, and so the mean onset of ambulation is at 23 months (Mets *et al.,* 2000).

The prevalence of intellectual disability in children with US is unknown. One study showed that 15% of the sample had some degree of intellectual deficit (Dammeyer, 2012), while another study found similar levels of cognitive development when comparing deaf children with US and cochlear implants with other deaf children without this syndrome with hearing aids (Henricson *et al.,* 2012).

CHARGE syndrome

It is the second-largest cause affecting both hearing and vision (Jackson *et al.,* 2015). Complex heart defects, swallowing and breathing disorders often make it difficult to assess hearing and visual loss, as well as cognitive delay (Raqbi *et al.,* 2003). Most patients with CHARGE syndrome have mixed or deep neurosensory hearing loss and reduced visual field (Arndt *et al.,* 2010). The incidence of severe to deep hearing loss ranges from 34% to 38% (Lanson *et al.,* 2007).

Approximately 50% of children with CHARGE have preserved intellectual capacity, 25% have a moderate reduction and another 25% are severely affected.

b) No cognitive alterations

Treacher Collins syndrome (TCS)

Most sufferers have facial malformations, cleft palate and bilateral auricular atresia, which usually causes transmission hearing loss. TCS usually does not cause cognitive deficits, although the alterations may affect speech and language development (Jackson *et al.,* 2015).

Waardenburg syndrome (WS)

This is a group of autosomal dominant genetic defects, which may also cause hearing loss and changes in pigmentation of the hair, skin and eyes (Ahmed jan *et al.,* 2021). There is no evidence of a relationship between WS and cognitive deficits.

c) Deaf-blindness

As seen above among associated disabilities, deaf-blindness resulting from the combination of two sensory impairments (visual and hearing) is noteworthy, as it generates unique communication problems and special needs, essentially due to the difficulty for globally perceiving, knowing, express-

ing interest and developing in the environment around them. Deaf-blindness should be considered as a distinct disability, requiring specialised servic-es (Ruiz, 2017).

The most common causes of deaf-blindness are often hereditary syndromes and complications with prematurity (Bruce and Borders, 2015). Early detection is critical in providing visual and auditory aids and optimising development (Parker and Nelson, 2016).

d) Other

There are many other syndromes or entities with associated deafness. Among others, a significant number of cases with deep hearing loss have been documented as also having autism spectrum disorder, a proportion that is not reflected equally the other way around (ASD with suspected hearing loss).

2.2. Diagnosed disability with suspicion of hearing loss

As mentioned above, there is much less information about the presence of hearing loss in individuals diagnosed with an autism spectrum disability or disorder (ASD).

Children and young people with intellectual disabilities are at high risk of other disabilities as well. Intellectual disabilities are found to be more strongly associated with sensory disabilities and/or physical disabilities, but individuals diagnosed with ASD have also been found to be independently associated with intellectual disability.

The combination of intellectual disability and/or ASD with hearing loss alters communication more than expected, leading to increasing complexity in the assessments and treatment of such cases (Kinnear *et al.,* 2020).

In the case of ASD, the risk of associated hearing loss is five times higher, visual impairment is eight times higher and intellectual disability is almost 50 times higher, so it is important to raise awareness among professionals about the high degree of comorbidity of the disorder and to take into account the prevalence data required to plan prevention and intervention measures (Rydzewska *et al.,* 2019).

In addition, there are other disorders commonly associated with cognitive retardation, such as global developmental delay (GDD) and cerebral palsy (CP), which are more likely to have hearing loss, with a prevalence of between 4% and 39% in the case of CP, aggravating the delay in the development of speech, language and cognitive function.

Hearing loss not identified in individuals with AD+ due to intellectual disability (ID) can significantly worsen their quality of life (Herer, 2012), so it is crucial to identify early if hearing loss is present in order to establish immediate care and treatment. Assessment of the hearing capacity of individuals with ID may require more time and better preparation to be able to use techniques such as auditory evoked potentials. The impact of hearing loss in combination with ID may vary, depending on the type and degree of hearing loss and the age at which it presents (Carvill, 2001).

Many children with cognitive retardation have a history of prematurity, ranging from 26% to 83%. This prematurity is a known risk factor for ASD and CP, and is also a risk factor for hearing loss with a frequency between 1% and 19% (Trudeau *et al.,* 2021; Wit de *et al.,* 2018).

The association of deafness and intellectual disability may also result from congenital cytomegalovirus (CMV) infection, which is one of the most common causes (up to one case per 200 newborns) (Blázquez-Gamero *et al.,* 2020).

Congenital CMV often combines disabilities associated with neurosensory hearing loss. In a recent review, the prevalence of hearing loss at birth was more than 33% among infected symptomatic newborns and less than 15% among asymptomatic infections. This difference in prevalence was maintained during childhood with more than 40% prevalence detected for symptomatic CMV and less than 30% for asymptomatic CMV. Late onset and progressive hearing loss appears to be characteristic of this congenital infection. In terms of associated disabilities, a high incidence of developmental delays (81%), microcephaly (93%), seizures (33%), neonatal encephalopathy (10%) and eye abnormalities (14%) has been reported, which may result in a poorer prognosis of overall development (Vos *et al.,* 2021; Gowda *et al.,* 2021).

3. RISK FACTORS AND WARNING SIGNS OF CHILDREN WITH AD+

Early detection and early treatment of hearing loss through currently available technological advances have improved access to sound for children with hearing loss and prevented delayed development of spoken language.

There is therefore a new pathway for these children, who have benefited from these advances that clinicians need to know in order to suspect and detect when progress in language acquisition and development is not adequate, so as to intervene appropriately (Wiley *et al.,* 2021).

It is important to note that not all children will respond equally to traditional therapeutic strategies for language development *(Table 1).* When it is wrongly assumed that developmental delay is solely due to hearing loss, we are missing the opportunity to intervene early and adequately to address associated disabilities, thereby improving communication skills in the long term, among other developmental milestones.

Thus, in the field of early care and educational intervention with deaf children, the next step should be to focus efforts on minimising the disparity in the level of language and communication development of this large group of children.

There is a high percentage of additional disabilities in children with deafness (40% versus 14% in the hearing population) because risk factors for hearing loss overlap with risk factors for many other disabilities.

3.1. Risk factors for associated disabilities

Developmental disabilities may include a number of factors that indicate a higher risk of adding another disability to hearing loss. These risk factors can be classified as prenatal, perinatal and postnatal. Recognising them helps alert families and to monitor that children have all their needs met early so as to support the maximum development of their abilities, as shown in *(Table 2)* (Wiley and Moeller, 2007).

3.2. Warning signs of developmental delay in children with hearing loss

Although the risk factors listed in *(Table 2)* may help in identifying children with different learning needs, not all children necessarily have one of these identifiable risk factors. This is important to remember, particularly in cases where there is a known cause of deafness that does not a priori lead to developmental problems.

Indeed, having an identified cause, e.g. genetics, does not protect the child from their development being influenced by other genes or other causes. Clinicians, speech therapists, educators and teachers must therefore all be vigilant, to detect atypical learning that causes a child with hearing loss not to progress appropriately:

Motor development

Children with hearing loss generally have typical motor development (Liberman *et al.,* 2005). If this is not the case, vestibular problems, vision problems or the presence of cerebral palsy or myopathies of various kinds are the main causes that need to be ruled out.

Warning signs suggesting a delay or disorder in motor development include: observation of poor control of the posture of the trunk and head; if the child does not walk around 15 months of age; if the child suffers frequent falls, has not developed a preference for a dominant hand by around 2.5 years of age, or shows a pattern of immature grasping and releasing.

Visual disorder

Children diagnosed with deafness have an increased risk of developing visual disorders, as discussed, since certain syndromes or diseases that cause hearing loss also have an impact on vision. In children with deafness, a visual disorder must always be ruled out, especially in cases diagnosed with a syndrome (Usher, CHARGE, Waardenburg, etc.), congenital infections causing retinopathy (CVM, retinopathy of prematurity) or brain damage, such as that caused by childhood cerebral palsy.

The warning signs are: lack of eye fixation, poor eye tracking, oscillating eye movement, wandering eyes, cephalic tilt, stumbling over objects, poor night vision, and difficult acclimatisation from darkness to light or vice versa.

Learning difficulties

Although deafness has been excluded from consideration as a specific learning disorder, it is common for children with deafness to have some learning difficulties or a specific disorder.

It is difficult for professionals to separate out the impact of deafness or the specific learning disorder itself on children's academic performance. Professionals with a good understanding of the differences between the two entities can plan an appropriate intervention.

Communication disorders

It is also difficult to determine whether there is a specific language disorder, which extends beyond the expected impact of deafness on communicative development.

It is clear that specific communication disorders and autism spectrum disorder may co-exist with deafness in a child. In addition, some children seem to have significant language processing difficulties that would not be explained solely by their hearing loss. For these children, receptive language skills are far behind what might be expected based on their cognitive potential and ability to learn. This pattern is easier to recognise in cases with mild or moderate hearing loss, and so early identification allows for the most appropriate educational support and intervention.

Intellectual disability

Cognitive skills can have an impact on a child's progress in all aspects of their development (Meinzen-Derr *et al.,* 2010). Although intellectual capacity is not the only factor influencing language development, it is highly correlated with language outcomes, so having a reliable assessment of cognitive abilities (typically determined by non-verbal cognitive measurements) can help provide a better structure to identify whether a child is making the progress that could be expected over time.

The algorithm described in *(Figure 1)* for the diagnostic and therapeutic approach to hearing loss in children with AD+ may serve as a guide for professionals who, knowing the individual characteristics and learning characteristics of the child, can then take step-by-step action to determine the intervention needs suited to each case.

4. LANGUAGE DEVELOPMENT IN AD+

The development of oral, receptive and expressive language in children with deafness treated with hearing aids (hearing aids and/or implants) is determined, among other factors, by the severity of hearing loss, its onset, the age of hearing aid fitting, cognitive development and the communicative model used in early intervention.

There are two other variables that behave differently in children fitted with hearing aids and implanted children. On the one hand, the level of maternal education, which in any event promotes the development of speech and language, especially in children fitted with hearing aids. On the other, the presence of additional disability to deafness negatively affects this development, especially in the case of implanted children with AD+ (Cupples *et al.*, 2018b).

One of the challenges raised by disability associated with hearing loss is early detection.

Previous studies have described, as one of the most reliable predictors in children's speech and language development, the presence of additional disability, as well as hearing loss. In this regard, in a study by Cupples, most children were diagnosed with autism spectrum disorder, cerebral palsy and global developmental delay, disabilities that have a greater impact than others on speech and language development (Cupples *et al.*, 2018a).

Thus, additional disabilities to hearing loss most often have been classified into two groups based on their impact on speech and language development *(Table 3)*. The first group identifies the disabilities with the greatest impact (ASD, cerebral palsy, ASD associated with cerebral palsy, global developmental delay associated with another syndrome and isolated global developmental delay). The second group includes vision disorders, speech and articulatory disorders, other non-developmental syndromes, and various medical and health problems. Research in 3-year-olds found that children with autism, cerebral palsy, and/or developmental delay score significantly worse on language development compared with children with other disabilities (such as visual impairments, speech disorders, syndromes not associated with developmental delay and disease).

Children with an additional disability to hearing loss achieve worse levels of language development than children without hearing loss, namely 1 to 2 standard deviations (SD) below normal hearing children of the same age. In contrast, non-verbal cognitive ability is approximately 0.3 SD off the standard mean (Ching *et al.,* 2013).

Past literature points to the variability of language development in implanted children with associated disability. One factor that may explain this variability is the different cognitive ability that each child or group of children may have.

Many studies indicate that better language development is related to better levels of cognitive development (Cupples *et al.*, 2018a), although other explanatory factors may also be found: non-verbal skills, less profound degree of hearing loss, use of spoken language in early intervention, higher educational level of the mother and early fitting of hearing aids or cochlear implants.

5. ADAPTATION OF CHILD HEARING DEFICIT PROGRAMMES TO CHILDREN WITH AD+

Widespread neonatal hearing screening programmes and early intervention have significantly increased the detection of children with deafness with additional disability. While demographics have changed, educational complexity and service needs for this population have also increased. Technological advances have improved access to sound and language development, while shortcomings may be seen in updating the knowledge of staff providing services to such a diverse group of children (Jackson *et al.,* 2015).

5.1. Diagnosis of deafness with additional disability

The first challenge to be faced by an early screening and treatment programme for childhood hearing loss is the detection or recognition of deaf children with additional disability. The identification of additional disabilities as soon as possible is essential so as to ensure access for these children to appropriate intervention and thus to achieve the maximum possible development in each case. Because screening and early care make it possible for language development delays in children with hearing problems to become less frequent, it may seem easier than in the past to diagnose other disabilities, such as ASD, in a child with deafness.

The recommendations of the Joint Committee on Infant Hearing (JCIH) and the CODEPEH establish that all children with hearing loss under 3 years of age should undergo a biannual evaluation of their speech, language and cognitive skills development (American Academy of Pediatrics, 2007; Núñez *et al.,* 2015), allowing detection of cases in which delays or deviations from the milestones to be reached and the expected results are identified.

5.2. Diagnosis of hearing loss in severe developmental disorders

Audiometric assessment to rule out a hearing deficit in a child with a disability can be a major challenge. In fact, many children undergo multiple unsuccessful attempts at behavioural audiometry. The inability to determine auditory thresholds in this context makes it necessary on many occasions to resort to sedation to carry out objective audiological tests. While it is always desirable to minimise sedation or anaesthesia, repeated and unsuccessful attempts to perform behavioural audiometry may cause diagnostic delays, leading to greater financial cost and concern for families.

Communication and language development may be more delayed in children with hearing loss associated with one or more disabilities.

Patients with cognitive retardation, autism spectrum disorder, global developmental delay or cerebral palsy are those at increased risk of developing additional hearing loss, even if they have passed neonatal screening tests for hearing loss (Trudeau *et al.,* 2021). They must therefore be followed more closely in order to rule out the presence of additional hearing loss (Núñez *et al.,* 2015).

Given the great benefit obtained from early intervention with hearing loss added to a cognitive or developmental disorder, it is imperative to avoid diagnostic delays, which some studies suggest may amount to up to two years (between 2 or 3 failed attempts of behavioural audiometry on average before auditory evoked potentials) (Trudeau *et al.,* 2021). This evidence supports the prompt indication of objective audiological tests to avoid delays in diagnosis, using sedation or general anaesthesia if necessary.

It is important to remember that some of the commonly used audiological tests do not confirm normal central auditory function (Davis and Stiegler, 2005). There is a tendency to report that hearing is appropriate to ensure speech and language development based on the normality of these tests, although it should be borne in mind that there are problems with central auditory processing that prevent proper development. Children and adults with ASD report difficulties in listening in noisy environments, in maintaining attention to auditory stimulation, and in using auditory information in situations of sensory stress (Cloppert and Williams, 2005), which may point to a central auditory processing disorder.

5.3. Audiological treatment recommended in cases of AD+

Once hearing loss is diagnosed in a child with AD+ and fitting hearing aids is indicated, the hearing aids should be selected and fitted using a prescriptive method that takes into account the acoustics of the auditory ducts and thresholds. If the behavioural thresholds are not reliable, the programming of the hearing aids should be adjusted based on the electrophysiological results of steady-state

evoked potentials, so that the fitting can be subsequently corrected as information on the behavioural thresholds is obtained in subsequent assessments (American Academy of Pediatrics, 2007).

The goal of any hearing aid fitting is to provide the child with access to acoustic speech information without the hearing aid exceeding the recommended amplification limits. Children with ASD are no different in terms of this goal, but it should be noted that some children with ASD experience a higher perception of sound and may exhibit behaviour consistent with an exaggerated response to an auditory stimulus (Tharpe *et al.,* 2006). It may therefore be necessary to limit the levels of prosthetic stimulation in order to facilitate acceptance of the new device. Another characteristic of these children is poor tolerance to the tactile sensation of contact with other people, which may hinder RECD (real-ear to coupler differences) measurements or taking impressions for ear moulds in hearing aid fitting, so it would not be unusual to resort to a sedation of the child for this task (Egelhoff *et al.,* 2005). As with any child fitting, access to hearing aid batteries must be blocked, and the volume and programming control buttons disabled.

With regard to cochlear implantation of children with ASD, improvements have been found in social communication, behaviour, attention to their surroundings, as well as an increase in vocalisations, eye contact and reaction to music (Donaldson *et al.,* 2004). Comparing children with ASD and others with different additional disabilities, it has been observed that the former have limited development of auditory perception. However, cochlear implantation is not contraindicated in children with ASD, provided that parents are informed that successful implantation cannot be predicted and it is important that expectations are adjusted. In such cases, it is desirable to have an experienced team open to consulting with ASD experts (Beers *et al.,* 2014).

It is essential to detect atypical learning that causes a child with hearing loss not to progress appropriately, as this may be influenced by a cause other than deafness that has not been identified.

An understanding of hearing loss is important as it is commonly seen in children with ASD. It consists of a pattern of exaggerated behavioural reactions to auditory sensory stimuli. Although its presence is not universal, it is so common in these cases that a differential diagnosis of ASD is required (Baranek *et al.,* 2005).

Psychoacoustic tests have been used to measure the perception of acoustic intensity in children with ASD, revealing a narrow dynamic range and a reduced tolerance, which is consistent with the hearing loss observed in this group (Khalfa *et al.,* 2004). The prevalence of this symptom ranges from 18% to 53% in children with ASD, whereas it is not found in children with typical development. The presence of hearing loss may correspond to certain characteristics of the subject, such as mental age, since it is attenuated as mental age increases. It is thus considered to be a general deficit associated with developmental disabilities, and is not specific to children with ASD (Baranek *et al.,* 2007).

The results of cochlear implantation vary widely. Specifically, the review of several studies regarding cochlear implants in children with WS indicates that they develop capacities similar to those implanted for typical neurosensory deafness. There are several studies on improvement in both hearing and speech intelligibility after cochlear implantation in children with WS (Cejas *et al.,* 2015). Results in patients with CHARGE have also shown that the most continued to have mild hearing loss, although on the other hand there was an improvement in paternal/maternal perception of hearing response. Most children used gesture communication and were enrolled in special education classes (Lanson *et al.,* 2007).

5.4. Early intervention in children with AD+

Another challenge to be addressed by a child hearing deficit programme is the planning of services to be made available to each child with AD+. The approach to dealing with such cases should be based on the individual needs of children, avoiding categorisation or labelling of their disability. The specific aspects of each case will thus serve as the basis to reinforce their strengths and provide tailored responses to their individual needs. This person-based approach allows families to play a major role in designing the care that will be given to their children.

AD+ children, if not detected early and adequately treated, risk missing the critical learning period and thereby consolidating a gap in their development with respect to deaf children without additional disability (Jones and Jones, 2003).

The variability of language development in implanted children who have AD+ may be explained by the different cognitive skill that each child or group of children may have.

Early care requires interdisciplinary teams to care for children with AD+, involving multiple service providers that include teachers, speech therapists, psychologists, educationalists and audiologists, and can be extended to other professions such as physiotherapists, likewise requiring the use of supportive products and technologies according to the needs of the child.

Although early care professionals see collaboration with families and other professionals as an important aspect of their work, interdisciplinary collaboration is often a challenge. One reason may be the poor preparation for such cooperative work, as collaboration is a subtle and very complex field requiring explicit instructions for professionals.

A continuum of collaborative models has been used in order to provide services to children with special educational needs: multidisciplinary, interdisciplinary and cross-disciplinary. The multidisciplinary model involves professionals working with children separately. This model results in fragmented services and even conflicting activities and recommendations being offered (Ewing and Jones, 2003). Using the interdisciplinary model, professionals communicate with each other and make decisions by consensus, but assessments and implementation tend to be carried out separately. This model does not allow professionals to fully coordinate their activities, and may result in a child having to work with multiple professionals, which may be difficult for younger children. The cross-disciplinary model not only includes communication between professionals and families, but also transfers the skills of one team member to another (Cloninger, 2004).

Cross-disciplinary collaboration is also known as an indirect therapy model and is characterised by a planned freeing up of roles, which are shared and exchanged, as are responsibilities among team members. This model allows for fewer team members directly providing services, while other members act as consultants. Such a strategy may be less stressful for young children.

However, the most effective programmes for children with AD+ should be highly flexible and individualised, with early intervention services focused on the child and family. The cross-disciplinary approach allows for the provision of coordinated services based on the contributions of a team of professionals with diverse experience and expertise.

5.5. Care and support for families of children with AD+

Among the actions that determine the effectiveness of treatment and intervention for a child with disabilities, one key need is to provide comprehensive care and, as has been stated, to focus on the child and his/her family, avoiding compartmentalised, decontextualised and uncoordinated responses, providing confidence and reassurance to families, reducing their disorientation and a procession of different specialists and services, while also initiating intervention as early as possible (Núñez *et al.,* 2019).

Coming to terms with the news and a lack of information are in turn two of the main difficulties the family faces in the initial moments after learning of the diagnosis. In the case of children with AD+, this situation is further heightened as they are often undergo overlapping or successive diagnoses, causing families greater emotional stress, which interferes with both acceptance of the situation and decision making. Families must therefore be able to rely on stable teams and structures with specialised and qualified professionals capable of dealing with the case on its individual terms, and on the basis of an analysis of the specific socio-familial situation.

The role of the professionals involved at each level is fundamental, especially in dealing with the initial news, and in general with conveying all the information to families, hence the importance of training programmes aimed at developing effective teamwork skills (Núñez *et al.,* 2015). It has also been shown that family support needs go beyond receiving information on health issues or hearing technology, and extends to the emotional plane. Different studies have collected information on the needs of families, some of which were related to the way information is passed on to them, the lack of coordination among professionals during the different stages of the screening programme, partial information on treatment and the difficulty of referral to appropriate services and the lack of knowledge of available resources (Núñez *et al.,* 2015).

A cross-disciplinary, co-responsible team working in partnership with the family is therefore essential at all levels of case management, especially when AD+ is involved. This team must provide quality, complete, understandable, objective and truthful information so as to adjust expectations as to the prognosis. This must cover

medical diagnosis, treatment, use of hearing devices, options for early intervention and speech therapy, as well as referral to family support structures and resources. Fami-ly support programmes must be integrated as an element of support and cooperation within the team (Núñez *et al.,* 2015; Joint Committee on Infant Hearing, 2007).

The widespread deployment of neonatal screening programmes for hearing loss has significantly increased early detection of children with deafness plus additional disability (AD+).

The message must be coherent and consistent between the different professionals involved. It is also impor-tant to sequence and group appointments within a short timeframe, coordinating the assistance and provision of information between them, in compliance with current regulations regarding personal data protection. The fam-ily should not be responsible for conveying information between professionals or mediating their coordination, as it is advisable to control the amount of information provided, the timing and setting for communication. Inter-administrative and inter-sectoral coordination is also needed with regard to the services and care for children and their families, with simplified and coordinated proce-dures for access to all of them. Referral circuits and care pathways must be established for both child and family, coordinated with each other and ensuring the continuity of the process (Núñez *et al.,* 2019).

The family must receive information, help and advice, bearing in mind that their collaboration and intervention is indispensable and irreplaceable, since the effective-ness and/or optimal outcome of the treatments depends largely on them.

6. CODEPEH RECOMMENDATIONS 2021

- Approximately 40% of children with deafness have an additional developmental disorder or major medical problem which, in addition to delaying the age at which hearing loss is diagnosed in many cases, also requires the intervention of other professional specialists.

  Early detection of this additional disability is vital, so assessing overall development every 6 months in any deaf child is extremely important.

- Similarly, deafness as an associated disability should be ruled out in children who have already been diagnosed with a disability and who do not reach the expected developmental milestones.

- The situation of these children is known in literature as “hard of hearing or deaf plus”, although the term “deafness and associated disability (AD+)” is preferred.

- Identifying AD+ allows for prompt, appropriate intervention that will be seen, among other areas, in improved language and communication skills.

- Relevant audiological testing should be performed as soon as possible, with sedation if necessary.

- Early care should include cross-disciplinary teams to care for children with AD+, using an approach based on the individual needs of the child and in collaboration with the family.

7. TABLES AND FIGURES

TABLE 1. Incidence of associated disabilities
(Gallaudet Research Institute, 2008)

TYPE OF DISABILITY	POPULATION WITH HEARING LOSS	GENERAL POPULATION
No associated disability	60%	86%
Intellectual disability	8.3%	0.71%
Cerebral palsy	4-39%	0.39%
Visual disorders	5.5%	0.13%
Attention deficit hyperactivity	5.4%	5-10%
Specific learning disorder	8%	5-10%
Autism spectrum disorder	7%	1%

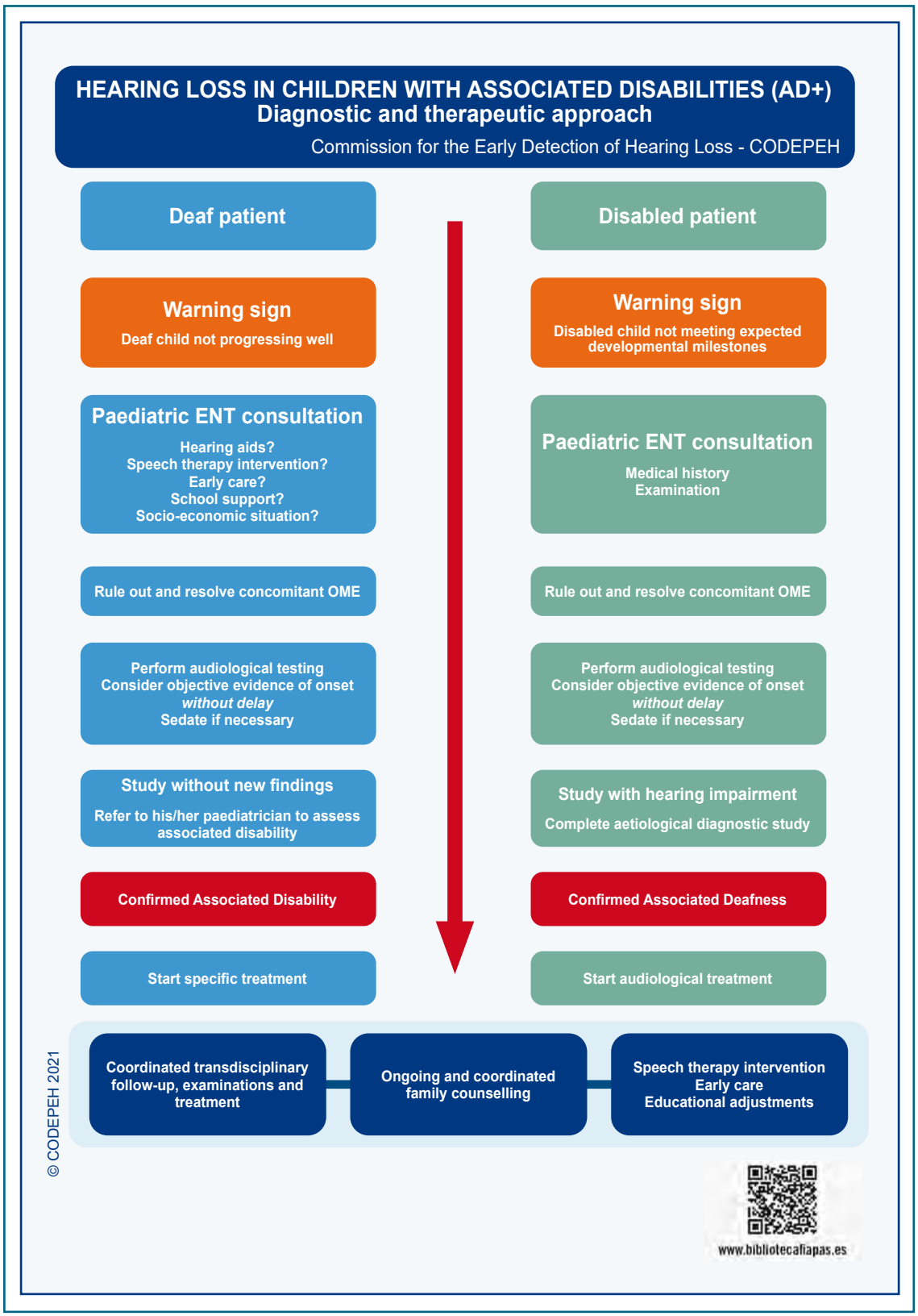
TABLE 2. Risk factors for associated disabilities
(Wiley & Moeller, 2007)

RISK FACTORS (AD+)
Prenatal risk factors
Exposure to toxic substances, such as alcohol and lead
Obstetric factors such as
Placental insufficiency
Twin pregnancy
Gestational hypertension
Gestational diabetes
Infections such as cytomegalovirus
Genetic factors
Syndromes or a family history of learning difficulties
Atypical embryonic development, such as spina bifida or brain abnormalities
Perinatal risk factors
Acute perinatal hypoxia
Prematurity
Perinatal infections
Hyperbilirubinaemia
Postnatal risk factors
Environmental exposure to tobacco smoke or lead
Malnutrition
Infections, such as meningitis or encephalitis
Complex medical issues such as
Heart disease
Visual problems
Neurological problems
Traumatic brain injury
Physical or emotional abuse
Inappropriate environmental situations

TABLE 3. Disabilities frequently associated to deafness
(Cupples et al., 2018a)

TYPE OF DISABILITY	
GROUP A	**GROUP B**
Autism spectrum disorder	Vision disorders
Childhood cerebral palsy	Speech disorders
Delayed development	Syndromes without developmental delay
Other syndromes with delayed development	Other medical conditions

FIGURE 1. Diagnostic and therapeutic approach to hearing loss in children with AD+ (CODEPEH, 2021)



REFERENCES

Ahmed jan, N., Mui, R.K. y Masood, S. (2021). Waardenburg Syndrome. En *StatPearls* [Internet]. Treasure Island (FL): StatPearls Publishing. https://www.ncbi.nlm.nih.gov/books/NBK560879/

American Academy of Pediatrics, Joint Committee on Infant Hearing. (2007). Year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics,* 120(4), 898–921.

Arndt, S. *et al.* (2010). Spectrum of hearing disorders and their management in children with charge syndrome. *Otology & Neurotology,* 31(1), 67–73.

Baranek, G. T., Parham, L. D., y Bodfish, J. W. (2005). Sensory and motor features in autism: assessment and intervention: Assessment and Intervention. En F.R. Volkmar, R. Paul, A. Klin y D. Cohen (Eds.), *Handbook of autism and pervasive developmental disorders: Assessment, interventions, and policy* (831–857). John Wiley & Sons, Inc.

Baranek, G. T. *et al.* (2007). Hyperresponsive sensory patterns in young children with autism, developmental delay, and typical development. *American Journal of Mental Retardation: AJMR,* 112(4), 233–245.

Benito-Orejas, J.I. *et al.* (2017). Etiología de la hipoacusia infantil. *Revista Orl,* 8(2), 69–83.

Beers, A.N. *et al.* (2014). Autism and peripheral hearing loss: a systematic review. *International Journal of Pediatric Otorhinolaryngology,* 78(1), 96–101.

Blázquez-Gamero, D. *et al.* (2020). Prevalence and clinical manifestations of congenital cytomegalovirus infection in a screening program in madrid (piccsa study). *The Pediatric Infectious Disease Journal,* 39(11), 1050–1056.

Bruce, S.M. y Borders, C. (2015). Communication and language in learners who are deaf and hard of hearing with disabilities: theories, research, and practice. *American Annals of the Deaf,* 160(4), 368–384.

Carvill, S. (2001). Sensory impairments, intellectual disability and psychiatry. *Journal of Intellectual Disability Research,* 45(6), 467–483

Cejas, I., Hoffman, M. F., y Quittner, A. L. (2015). Outcomes and benefits of pediatric cochlear implantation in children with additional disabilities: a review and report of family influences on outcomes. *Pediatric Health, Medicine and Therapeutics,* 6, 45–63.

Ching, T.Y.C. *et al.* (2013). Outcomes of early- and late-identified children at 3 years of age: findings from a prospective population-based study. *Ear and Hearing,* 34(5), 535–552.

Cloninger, C. J. (2004). Designing collaborative educational services. En Orelove, F.P. *et al.* (Eds.), *Educating children with multiple disabilities: A collaborative approach.* Baltimore, MD: Brookes, 1–29.

Cloppert, P., y Williams, S. (2005). Evaluating an enigma: what people with autism spectrum disorders and their parents would like audiologists to know. *Seminars in Hearing,* 26(04), 253–258.

Cupples, L. *et al.* (2018a). Language development in deaf or hard-of-hearing children with additional disabilities: type matters! *Journal of Intellectual Disability Research: JIDR,* 62(6), 532–543.

Cupples, L. *et al.* (2018b). Spoken language and everyday functioning in 5-year-old children using hearing aids or cochlear implants. *International Journal of Audiology,* 57(Sup2), S55–S69.

Dammeyer, J. (2012). Children with Usher syndrome: mental and behavioral disorders. *Behavioral and Brain Functions: BBF,* 8(16).

Davis, R. y Stiegler, L. (2005). Toward More Effective Audiological Assessment of Children with Autism Spectrum Disorders. *Seminars in Hearing,* 26(4), 241–252.

de Wit, E. et al. (2018). Same or different: the overlap between children with auditory processing disorders and children with other developmental disorders: a systematic review. Ear and Hearing, 39(1), 1–19.

Donaldson, A. I., Heavner, K. S. y Zwolan, T. A. (2004). Measuring progress in children with autism spectrum disorder who have cochlear implants. *Archives of Otolaryngology–Head & Neck Surgery,* 130(5), 666–671.

Egelhoff, K. *et al.* (2005). What audiologists need to know about autism spectrum disorders. *Seminars in Hearing,* 26(4): 202–209.

Ewing, K. M., y Jones, T. W. (2003). An educational rationale for deaf students with multiple disabilities. *American Annals of the Deaf,* 148(3), 267–271.

Gallaudet Research Institute (2008). *Regional and national summary report of data from the 2007-08 annual survey of deaf and hard of hearing children and youth.* Washington, D.C.: Gallaudet University.

Gallaudet Research Institute (2013). Regional and national summary report from 2011-2012 annual survey of deaf and hard of hearing children and youth. Washington D.C.: Gallaudet University.

Global Research on Developmental Disabilities Collaborators (2018). Developmental disabilities among children younger than 5 years in 195 countries and territories, 1990-2016: a systematic analysis for the Global Burden of Disease Study 2016. *The Lancet. Global health,* 6(10), e1100–e1121.

Gowda, V. K., Kulhalli, P., & Vamyanmane, D. K. (2021). Neurological manifestations of congenital cytomegalovirus infection at a tertiary care centre from southern india. *Journal of Neurosciences in Rural Practice,* 12(1), 133–136.

Henricson, C. *et al.* (2012). Cognitive skills in children with usher syndrome type 1 and cochlear implants. *International Journal of Pediatric Otorhinolaryngology,* 76(10), 1449–1457.

Herer, G. R. (2012). Intellectual disabilities and hearing loss. *Communication Disorders Quarterly,* 33(4), 252–260.

Jackson, R. L. W., Ammerman, S. B. y Trautwein, B.A. (2015). Deafness and diversity. *American Annals of the Deaf,* 160(4), 356–367.

Jones, T. W. y Jones, J. K. (2003). Educating young deaf children with multiple disabilities. In Bodner-Johnson, B. y Sass- Lehrer, M. (Eds): *The young deaf or hard of hearing child: A family-centered approach to early education.* Baltimore MD: Brookes, 291–329.

Khalfa, S. *et al.* (2004). Increased perception of loudness in autism. *Hearing Research,* 198(1), 87–92.

Kennedy, C.R. *et al.* (2006). Language ability after early detection of permanent childhood hearing impairment. *The New England Journal of Medicine,* 354(20), 2131–2141.

Kinnear, D. *et al.* (2020). The relative influence of intellectual disabilities and autism on sensory impairments and physical disability: a whole-country cohort of 5.3 million children and adults. *Journal of Applied Research in Intellectual Disabilities,* 33(5), 1059–1068.

Lanson, B.G. *et al.* (2007). Cochlear implantation in children with charge syndrome: therapeutic decisions and outcomes. *The Laryngoscope,* 117(7), 1260–1266.

Lieberman, L. J., Volding, L., y Winnick, J. P. (2004). Comparing motor development of deaf children of deaf parents and deaf children of hearing parents. American Annals of the Deaf, 149(3), 281–289.

Liu, X.Z. *et al.* (2008). Cochlear implantation in individuals with usher type 1 syndrome. *International Journal of Pediatric Otorhinolaryngology,* 72(6), 841–847.

Meinzen-Derr, J. *et al.* (2010). Language performance in children with cochlear implants and additional disabilities. *The Laryngoscope,* 120(2), 405–413.

Mets, M. B., Young, N. M., Pass, A., y Lasky, J. B. (2000). Early diagnosis of usher syndrome in children. *Transactions of the American Ophthalmological Society,* 98, 237–242.

Núñez-Batalla, F. *et al.* (2012). Indicadores de riesgo de hipoacusia neurosensorial infantil. *Acta Otorrinolaringológica Española,* 63(5), 382-390.

Núñez, F. *et al.* (2015). Recomendaciones CODEPEH 2014: sorderas diferidas y sobrevenidas en la infancia. *Revista Española de Discapacidad,* 3(1): 163–186.

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(1): 201–220.

Olusanya, B.O. et al. (2020). Global burden of childhood epilepsy, intellectual disability, and sensory impairments. Pediatrics, 146(1), e20192623.

Parker, A. T., y Nelson, C. (2016). Toward a comprehensive system of personnel development in deafblind education. *American Annals of the Deaf,* 161(4), 486–501.

Raqbi, F., Morisseau-Durand, M. P., y Dureau, P. (2003). Early prognostic factors for intellectual outcome in CHARGE syndrome. *Developmental Medicine and Child Neurology,* 45(7), 483–488.

Ruiz, M. (2016). La sordoceguera, su tratamiento normativo y atención a las personas que la presentan. *Siglo Cero. Revista Española Sobre Discapacidad Intelectual,* 47(3), 29–54.

Rydzewska. E. *et al.* (2019). Prevalence of sensory impairments, physical and intellectual disabilities, and mental health in children and young people with self/proxy-reported autism: observational study of a whole country population. *Autism: The International Journal of Research and Practice,* 23(5), 1201–1209.

Shott, S. R., Joseph, A. y Heithaus, D. (2001). Hearing loss in children with down syndrome. *International Journal of Pediatric Otorhinolaryngology,* 61(3), 199–205.

Tharpe, A. et al. (2006). Auditory characteristics of children with autism. Ear and Hearing, 27(4), 430–441.

Trudeau, S. *et al.* (2021). Diagnosis and patterns of hearing loss in children with severe developmental delay. *American Journal of Otolaryngology,* 42(3), 102923.

Vos, B. *et al.* (2021). Cytomegalovirus-a risk factor for childhood hearing loss: a systematic review. *Ear and Hearing,* 42(6), 1447–1461.

Wiley , S. *et al.* (2021). *Chapter 6: Children who are deaf of hard of hearing plus.* En National Center for Hearing Assessment and Management e book. *A Resource Guide for Early Hearing Detection and Intervention (EHDI).* Utah State University Eds. Logan (Utah), 1-6. https://www.infanthearing.org/ ehdi-ebook/ index.

Wiley, S., y Meinzen-Derr, J. (2013). Use of the ages and stages questionnaire in young children who are deaf/hard of hearing as a screening for additional disabilities. *Early Human Development,* 89(5), 295–300.

Wiley, S., y Moeller, M. P. (2007). RED FLAGS for disabilities in children who are deaf/hard of hearing. *ASHA Leader,* 12(1), 8-9, 28-29.

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