Pediatric Hearing Loss: Referral, Visiting and Treating – REVISIT RANU (Universal Neonatal Auditory Screening)

Review Article

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Abstract

Introduction: New risk and suspicion factors for childhood deafness were recently introduced in the literature, justifying the suggestion to review the algorithm associated with Portuguese Universal Neonatal Hearing Screening (UNHS).
Goals: Develop an algorithm that allows screening, early diagnosis, referral and rapid intervention in child deafness.
Material and methods: Algorithm creation based on literature review and existing clinical guidelines.
Results: Newborns after intrauterine Zika infection, admitted to a neonatal intensive care unit for more than 5 days or submitted to ExtraCorporal Membrane Oxygenation and children submitted to ventriculoperitoneal shunt were included in the UNHS risk group. In some cases, it is suggested to extend the audiological assessment until school age. Alarm signals were proposed at stages of development that should motivate referral for audiological examination.
Conclusions: The developed algorithm extends the scope of referral of auditory screening to other pediatric ages and suggests updates to the UNHS.
Keywords: Pediatric; Audiometry; Hearing; Evoked Response; Intensive Care, Neonatal; Extracorporeal Membrane Oxygenation; Zika virus

Introduction

Congenital or acquired hearing loss during childhood is relatively common, with approximately 1:1000 newborns (NB) presenting with severe-profound hearing loss and 1:500 developing some degree of hearing loss during childhood.[1–5]

In addition to its high prevalence, hearing loss in childhood interferes with the child's normal development, especially the development of spoken language, and may result in a delay in speech acquisition, limitation of cognitive abilities, and impaired communication and social interaction.[2–8]

The diagnosis should be made as early as possible to reduce the potential developmental consequences. Tools such as universal neonatal hearing screening (UNHS)[2], regular follow-up in a child health clinic by a general practitioner or pediatrician, awareness of the main risk factors for hearing loss during childhood, and rapid hospital referral protocols are essential for improving the prognosis of childhood deafness.

In Portugal, the Group for Screening and Intervention in Childhood Deafness (Grupo de Rastreio e Intervenção da Surdez Infantil - GRISI) has set the following goals: identify all children with hearing loss by three months of age and ideally start the intervention by six months of age.[2] Currently, the available audiological equipment for universal screening and diagnosis is easily accessible and manageable and permits objective, non-invasive, and even automatic assessment of hearing. The two major strategies used in this context are acoustic otoemissions (AOs) and auditory evoked potentials (AEP). AOs are sounds originating in the cochlea, namely in response to an auditory stimulus, which can be recorded by a microphone in the external auditory canal. There are two major groups of AOs, spontaneous and evoked. In the latter, we identify transient AOs and distortion products, both of which can be tested in neonatal hearing screening with equal efficiency.[9–11] In response to an auditory stimulus, AEPs record waves with different latencies corresponding to different points in the auditory pathway from the cochlea to the cerebral cortex. They are an objective measure of auditory function, allow estimation of the hearing thresholds of the patient, and may contribute to the topodiagnosis. Both AOs and AEPs, alone or combined, enable the identification of NBs with a hearing loss greater than 35[2] or 40 dB HL.[5,6,9] Nevertheless, even children who pass the screening test at birth and therefore do not appear to have neonatal hearing loss should be followed-up in a child health clinic for monitoring signs of late hearing loss and referred to a specialty physician if required.[2,8,12]

The risk factors newly reported in the medical literature justify the proposal of a revised algorithm for UNHS. The objective of this study was to develop and propose an algorithm for the screening, early diagnosis, and rapid referral of children at risk of hearing loss, to be applied not only in primary care child and adolescent health monitoring but also in pediatric and otorhinolaryngology clinics.

Materials and Methods

This literature review was conducted by searching the PubMed, NCBI, American Academy of Pediatrics (AAP), and Surgeon General (*Direcção Geral de Saúde*) websites using the terms “childhood hearing loss”, “infant hearing loss”, “neonatal screening”, “auditory screening”, and “hearing risk factors”. The following were consulted: the recommendations of the UNHS, GRISI (2007)[2], AAP[3,4], EUSCREEN Vision & Hearing 2021[5], Joint Committee on Infant Hearing (JCIH)[6], and Societé Française de Pédiatrie[13]. For topics introduced recently about premature NBs, we used the chronological age.

Results

The main risk factors for childhood hearing loss requiring referral to an otorhinolaryngology consultation were determined based on the previously existing clinical standards in Portugal[2] and recommendations of international societies (Table 1).[3,5,6] Previous studies have also described reassessment timings for different clinical situations, such as the extension of serial audiological assessment until six years of age for children who underwent extracorporeal membrane oxygenation (ECMO)[14] or had intrauterine cytomegalovirus (CMV) infection (Table 2)[15]. In the context of child and adolescent health surveillance, warning signs have been proposed for different developmental stages that should lead to a referral to a specialty physician for audiological examination. [8,12]

Proposal for review of UNHS in Portugal

1. Risk factors to be included (Table 1)

a) Parental consanguinity

Population studies conducted in the Middle East have shown the existence of up to 3.5 times more cases of childhood hearing loss in the offspring of blood-related couples. [16,17] Therefore, it is pertinent to include NBs of blood-related couples in the at-risk group for UNHS in Portugal.

b) Intrauterine Zika virus infection

NBs with intrauterine Zika virus infection (mother with laboratory findings of Zika virus infection during labor and NBs with laboratory confirmation of infection) should be included in the at-risk group for UNHS according to the recommendation of the JCIH [6], even when asymptomatic. This risk group should preferably be evaluated using AEP.[6]

c) Intrauterine exposure to toxic substances and/or tobacco

Following the guidelines of the Société Française de Pédiatrie, all NBs with intrauterine exposure to tobacco and/or toxic substances were included. Some studies have demonstrated that NBs exposed to tobacco during pregnancy had lower amplitudes of OAE [18,19]. Intrauterine exposure to tobacco was recently associated with an increased risk of hearing loss in childhood [20] and adolescence. [21]

Intrauterine exposure to toxic substances such as cocaine and/or opioids has been shown to be associated with worse auditory outcomes in several studies. [22–26]

d) Hospitalization in the Neonatal Intensive Care Unit (NICU) longer than five days

All NBs admitted to NICUs for more than five days should be included, regardless of whether they underwent invasive mechanical ventilation, as suggested by the JCIH. An association between this population and the increased risk of auditory neuropathy has been established.[6] The use of aminoglycosides and hyperbilirubinemia are known risk factors for auditory neuropathy, and noise exposure in the NICU may also contribute to hearing loss.[6] EUSCREEN suggests that this group should be assessed by AEP.[5]

It should be noted that the *Société Française de Pédiatrie* also suggests the inclusion of preterm infants under 32 weeks of gestation in the at-risk group, as well as those with brain complications related to prematurity [13]. Because this is a controversial issue due to the overlap between this group of NBs and NBs requiring NICU admission or those exposed to other risk factors that were already included in the at-risk group [27-29], we chose not to include this risk factor as an isolated factor.

e) ECMO

Children undergoing ECMO were included by the JCIH due to the increased risk of late-onset hearing loss.[3,6,14]

f) Fetal Alcohol Syndrome

The *Société Française de Pédiatrie* recommends the inclusion of all NBs with documented intrauterine exposure to alcohol[13]. Despite the association shown between fetal alcohol syndrome and childhood hearing loss[30,31], studies conducted in recent years on children diagnosed with diseases of the fetal alcohol spectrum did not demonstrate an increased prevalence of hearing loss in this population compared to healthy children[32]. However, we deemed that NBs diagnosed with fetal alcohol syndrome should be included in the at-risk group of hearing loss.

g) Ventriculoperitoneal shunt

Recent evidence suggests that 40% of children (18 years old and less) develop hearing loss of at least 15 dB in three consecutive frequencies after ventriculoperitoneal shunt placement.[33,34] For this reason, EUSCREEN suggests that all children should be considered at risk of hearing loss after ventriculoperitoneal shunt placement and have their hearing evaluated in accordance with their ability to collaborate.[5]

Table 1
Indicators of risk of congenital permanent or late/progressive onset hearing loss in childhood

1	**Family**	Family history of hearing loss in childhood and/or use of hearing aid before the age of 50 years
2		Parental consanguinity
3	**Pregnancy**	Intrauterine infections (CMV, herpes, rubella, syphilis, toxoplasmosis, chickenpox, and laboratory findings of Zika infection in the mother and child)
		Intrauterine exposure to toxic substances (cocaine and/or opioids) and tobacco
4	**Perinatal**	Neonatal suffering (Apgar 0-4 at 1 min or 0-6 at 5 mins, Ø spontaneous breathing at 10 mins, hypotonia > 2 h after birth)
5		NICU: hospitalization for > 5 days or ECMO or invasive ventilation or hyperbilirubinemia requiring exchange transfusion
6		Weight at birth < 1,500 kg
7	**Congenital**	Craniofacial anomalies. Especially anomalies involving the external ear (e.g., external ear atresia), EAC, or temporal bone
8		Physical changes associated with syndromes that usually include conductive/sensorineural deafness, such as the congenital white forelock (Waardenburg syndrome)
9		Neurodegenerative diseases (Hunter syndrome, leukodystrophies) or sensory-motor neuropathies (Friedreich ataxia, Charcot-Marie-Tooth syndrome)
10		Syndromes associated with hearing loss (trisomy 21, 22q11 deletion, fetal alcohol syndrome, neurofibromatosis, osteopetrosis, Usher syndrome, Waardenburg syndrome, Alport syndrome, Pendred syndrome, Jervell and Lange-Nielsen syndrome)
11	**Other**	Culture-confirmed infections associated with sensorineural hearing loss, including bacterial and viral meningitis (especially herpes and chickenpox) and severe neonatal sepsis
12		Ototoxic substance administration > 5 days (gentamicin, trobramycin, or loop diuretics)
13		Recurrent or persistent (more than 3 months) otitis media
14		Chemotherapy (especially platinum derivatives)
15		Caregiver's concern about hearing loss or delayed speech/discourse development or learning difficulties
16		Cranial trauma requiring hospitalization, especially involving the skull base or temporal bone fracture
17		Ventriculoperitoneal shunt

Abbreviations: CMV, Cytomegalovirus; NICU, Neonatal Intensive Care Unit; APGAR, Appearance, Pulse, Grimace, Activity, Respiration; ECMO, Extracorporeal membrane oxygenation; EAC, External Auditory Canal.
* Suggested alteration to the current recommendations.

h) Cranial trauma requiring hospitalization

The AAP suggests that all children who experienced cranial trauma requiring hospitalization should have their hearing evaluated, in particular, but not exclusively, in cases of temporal bone fractures, unlike what is currently recommended in Portugal.[2–4]

2. Timings of the audiological re-evaluation with otorhinolaryngology (ORL) consultation (Table 2)

Based on the JCIH, we introduced the recommendation of evaluating children (up to school age) who had undergone ECMO or were exposed to intrauterine CMV infection. Children diagnosed with bacterial/viral meningitis and severe neonatal sepsis who exhibited some degree of hearing loss in the audiological evaluation at the time of discharge should also remain under follow-up until school age, because of the risk of worsening.[6] The JCIH recommends audiological re-evaluation up to nine months of age in cases of other intrauterine infections (other than CMV and Zika), hospitalization in the NICU for longer than five days,

Table 2
Periods of reevaluation according to the clinical entity or ORL consultation with audiological evaluation suggested by the Joint Committee on Infant Hearing

1	Family history of hearing loss in childhood	**Until 9 months** (extend if late onset)
2	ECMO	**Until school age** (3-6 months intervals)
3	Intrauterine CMV infection	
4	Culture-confirmed infections associated with sensorineural deafness, in particular bacterial and viral meningitis (especially herpes and chickenpox) and severe neonatal sepsis	
5	Intrauterine Zica infection (mother and child with laboratory findings)	**AEP at 1 M and 4-6 M**
6	Other intrauterine infections (herpes, rubella, syphilis, toxoplasmosis, chickenpox)	**Until 9 months**
7	NICU: hospitalization > 5 days or hyperbilirubinemia requiring exchange transfusion	
8	Aminoglycosides > 5 days (gentamicin, tobramycin, streptomycin)	
9	Neonatal distress	
10	Craniofacial anomalies, namely those involving the external ear, EAC, or temporal bone	
11	Physical changes associated with syndromes that usually include conductive/ sensorineural deafness, such as the congenital white forelock (Waardenburg syndrome)	
12	Neurodegenerative diseases (Hunter syndrome, leukodystrophies) or sensory-motor neuropathies (Friedreich ataxia, Charcot-Marie-Tooth syndrome)	
13	Syndromes associated with hearing loss (neurofibromatosis, branchiootorenal syndrome, osteopetrosis, Usher syndrome, Waardenburg syndrome, Alport syndrome, Stickler syndrome, Pendred syndrome, Jervell and Lange-Nielsen syndrome)	
14	Chemotherapy (especially platinum derivatives)	**< 3 months after occurrence**
15	Cranial trauma requiring hospitalization, especially involving the skull base or temporal bone fracture	
16	Otitis media, recurrent or persisting for more than 3 months	**Immediate**
17	Caregiver's concern about hearing loss or delayed speech/discourse development or learning difficulties	

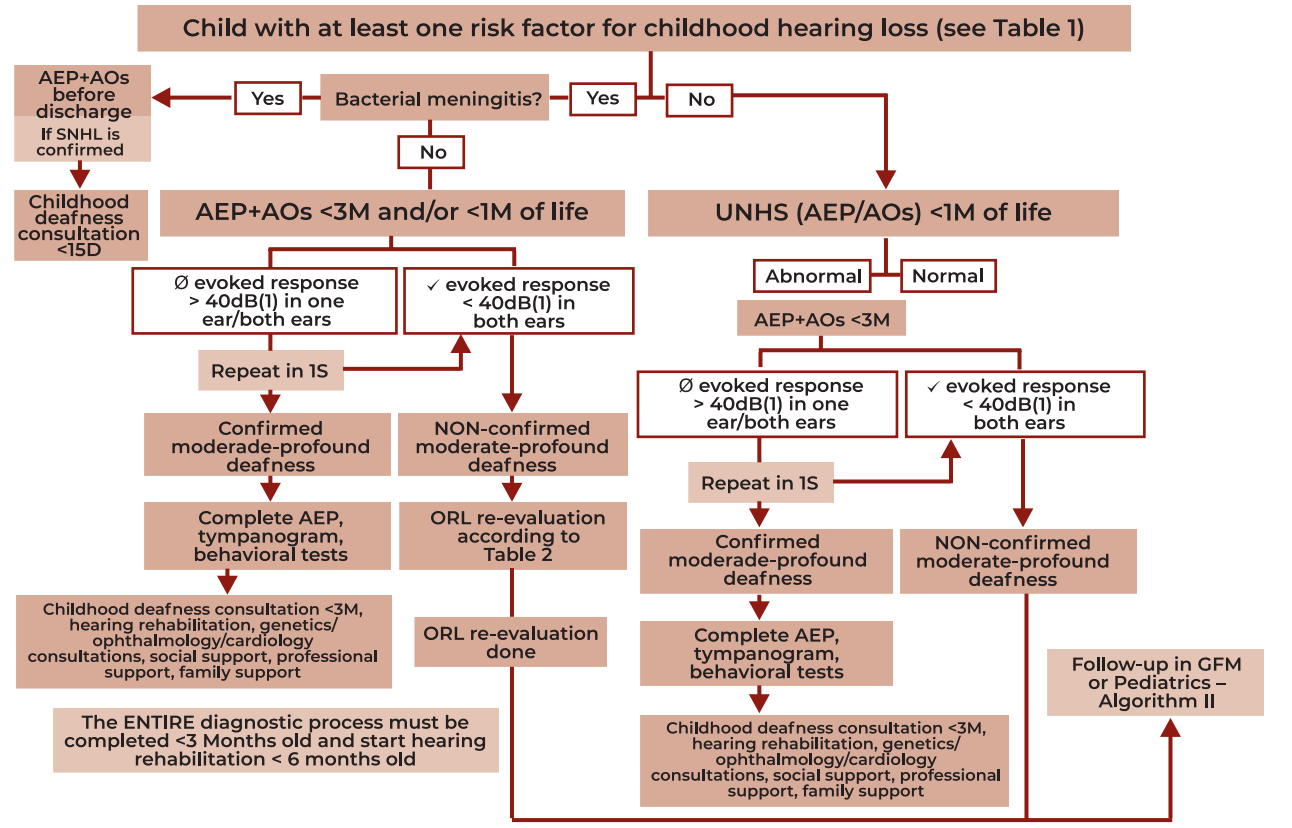
Abbreviations: ECMO, Extracorporeal membrane oxygenation; CMV, Cytomegalovirus; APGAR, Appearance, Pulse, Grimace, Activity, Respiration; EAC, External Auditory Canal
* Suggested alteration to the current recommendations.

hyperbilirubinemia requiring exchange transfusion, neonatal distress and suspicion or confirmation of syndromes or neurological diseases associated with hearing loss, due to the risk of worsening of hearing in this period that may be missed in the first evaluation.[6] In cases in which the parents, speech therapists, educators, or other professionals that follow the child's development suspect a problem, the audiological evaluation may be further extended. We emphasize that children at risk due to intrauterine Zika infection, hospitalization in the NICU > five days, or hyperbilirubinemia requiring exchange transfusion should be preferentially assessed with AEP.[6]

3. Algorithm for hearing screening of newborns or children at risk (Algorithm 1) (figure 1)

According to the clinical standards currently in place in Portugal [2], all NBs should be assessed for the existence of risk factors for childhood deafness (Table 1). Thus, UNHS can be divided into screening of children at risk and those not at risk of hearing loss. NBs not at risk of hearing loss should be evaluated with AEP or OAE in the first month of life, as is currently recommended in Portugal.[2] In cases In which

Figure 1
Algorithm 1. Scheme of organization for conducting hearing screening in newborns and children with risk factors for hearing loss



Abbreviations: AEP, Auditory Evoked Potentials; OAE, Otoacoustic emissions; C, Consultation; D, Days; W, Weeks; M, Months; ORL, Otorhinolaryngology; UNHS, Universal Neonatal Hearing Screening; GFM, General and Family Medicine

a response is detected through these methods at 40 dB HL or less, the NB is referred to the family physician or pediatrician for follow-up, as described in algorithm II. NBs without a response at 40 dB HL or more in AEP or OAE should undergo AEP and OAE before reaching the age of three months to confirm moderate to profound deafness, because the combined use of AEP and OAE may reduce the rate of false negatives in this subgroup.[3] In cases in which moderate to profound deafness is confirmed, complete AEP, tympanogram, and behavioral tests should be ordered and the child should be referred for a childhood deafness consultation for subsequent etiological investigation and clinical guidance regarding hearing and social and family rehabilitation. NBs at risk of RN bacterial meningitis should be evaluated with AEP and OAE before being discharged from the hospital and referred for a childhood deafness consultation within less than 15 days if hearing loss is confirmed, because of the risk of progression to labyrinthitis ossificans.[4,6] All other at-risk NBs should undergo AEP and OAE in the first month of life and their audiological evaluation should be finished at three months[4,6], because the combined use of AEP and OAE may reduce the rate of false negatives in these subgroups.[3] In at-risk cases in which moderate-profound deafness is not confirmed, the children should be reevaluated according to the intervals indicated in Table 2. In cases in which there is no evoked response at 40 dB HL or more in one or both ears, the clinical guideline is the same as for NBs without risk of hearing loss who similarly did not demonstrate a response in UNHS at birth. Children of any age, who throughout their development, have one of the risk factors for hearing loss listed in Table 1 should be screened in the same way as NBs at risk, as shown in Table 2.

4. Algorithm for the identification of children at risk of hearing loss, namely during consultation of child and adolescent health monitoring in the primary health care setting (Algorithm 2) (figure 2)

Children followed in the child and adolescent health monitoring consultation in the primary health care setting should undergo an audiological evaluation with an otorhinolaryngologist if a delay in development (namely in speech development) or behavioral problems such as autism spectrum disorders are detected.[3,4] Because of the negative impact on the child's development, a high degree of suspicion of hearing loss should be maintained even in children who apparently do not have a developmental delay or behavioral issues. If a risk factor for hearing loss is identified at any stage of the follow-up, the child should be referred for an audiological evaluation with an otorhinolaryngologist within less than 15 days in case of bacterial meningitis.[4]

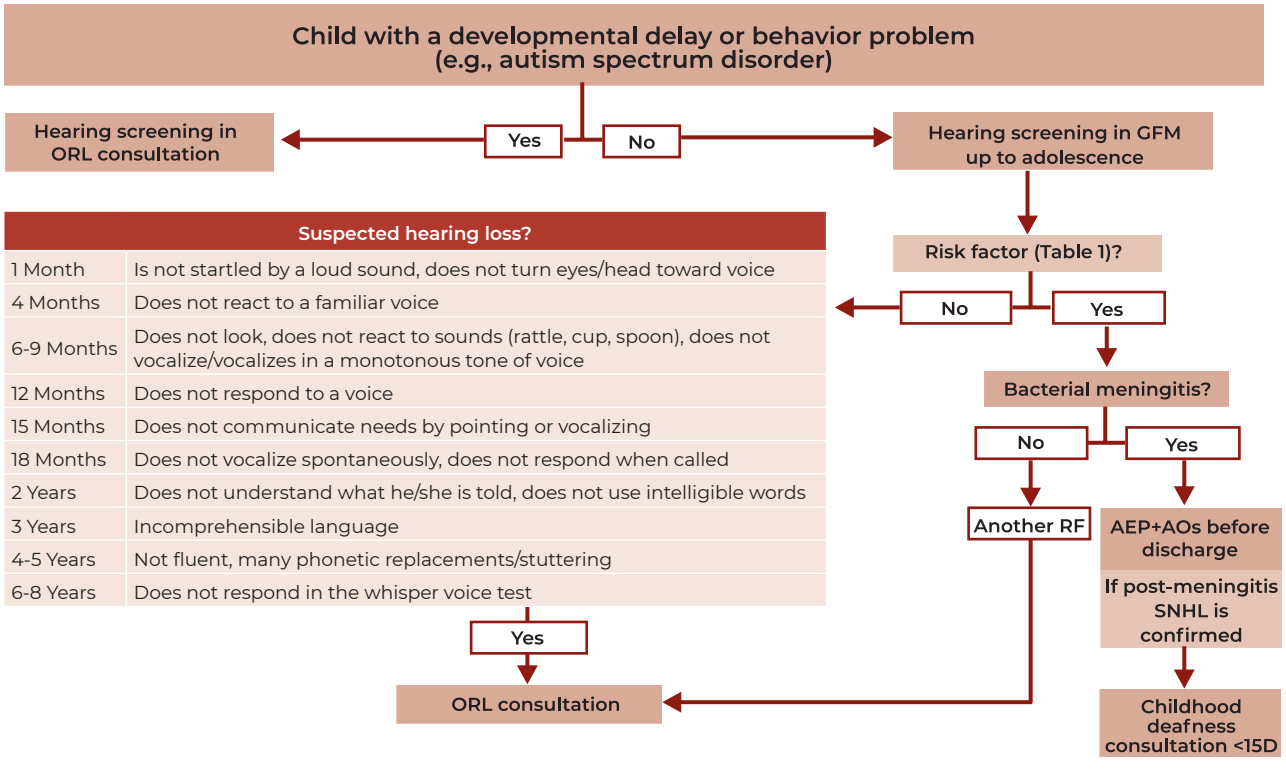
Based on the suggestions of Yoshinaga-Itano *et al* [8] and of the National Program for Child and Adolescent Health (Programa Nacional de Saúde Infanto-Juvenil)[12], which has been in force in Portugal since 2013, we created a list of potential changes in the child's normal development that may indicate a higher risk of hearing loss and the need for an ORL evaluation with an audiological assessment.

Some European countries have recently established a new universal hearing screening at the age of four years. However, because this measure still lacks cost-benefit evidence, we chose not to include this suggestion in the designed algorithm.[35,36]

Discussion

The present algorithm proposes to extend the current scope of referral for pediatric hearing screening in Portugal to other pediatric ages and suggests updates to the present clinical guidelines with the introduction of new risk

Figure 2
Algorithm 2 – Scheme of organization to identify children at risk of hearing loss during child and adolescent health monitoring consultation in the primary health care setting



Suspected hearing loss?	
1 Month	Is not startled by a loud sound, does not turn eyes/head toward voice
4 Months	Does not react to a familiar voice
6-9 Months	Does not look, does not react to sounds (rattle, cup, spoon), does not vocalize/vocalizes in a monotonous tone of voice
12 Months	Does not respond to a voice
15 Months	Does not communicate needs by pointing or vocalizing
18 Months	Does not vocalize spontaneously, does not respond when called
2 Years	Does not understand what he/she is told, does not use intelligible words
3 Years	Incomprehensible language
4-5 Years	Not fluent, many phonetic replacements/stuttering
6-8 Years	Does not respond in the whisper voice test

Abbreviations: AEP, Auditory Evoked Potentials; OAE, otoacoustic emissions; C, Consultation; ORL, Otorhinolaryngology; FM, Family Medicine; SNHL,sensorineural hearing loss

factors and re-evaluation intervals according to the clinical entity seeing the child.

With regard to the introduction of new risk factors, intrauterine infection with the Zika virus was only linked to neonatal hearing loss in 2016; therefore, it has not yet been included in the currently used clinical standards in Portugal.[2,5,6] We think it is essential to introduce this risk factor, given the increasing prevalence of this infectious disease in Brazil[37] and the fact that Brazilians accounted for 25.67% of the Portuguese immigrant community in 2019.[38]

Another recent factor is the exponential growth in the use of ECMO in recent years. From 1989 to March 2022, 46,667 NBs have undergone ECMO, according to the records of the Extracorporeal Life Support Organization. NBs undergoing ECMO, regardless of the duration of the extracorporeal support, are a subgroup at major risk of hearing loss, which may present at a later stage: the literature describes correlations up to 13 years of age.[14] Therefore, we suggest that these infants are included in the at-risk group under the existing standards in Portugal and that their hearing is monitored in ORL clinics at least until school age.[6] Between 6 to 13 years of age, they should be followed by their assistant physician and referred for an ORL consultation when there is a suspicion of hypoacusis, either reported by the child or by his/her caregivers.

There are other risk factors in the context of NICU hospitalization, namely exposure to noise. This may exceed the mean 45 dB recommended by the AAP[39–41] and be related to the degree of hearing loss six months after discharge from the NICU.[42] Due to the exposure to noise and coexistence of other comorbidities associated with hearing loss (such as hyperbilirubinemia, exposure to ototoxic substances, and invasive mechanical ventilation), these NBs are a subgroup at risk of hearing loss. We believe that unlike the current standards in use in Portugal[2], all NBs who were in the NICU for more than five days should be included in the at-risk subgroup of UNHS, regardless of the NB's comorbidities. Similar to the JCIH and EUSCREEN recommendations and due to the risk of auditory neuropathy, we suggest that this subgroup should also be preferentially assessed by AEP.[5,6]

With regard to the introduction of new intervals between evaluations, we propose that some groups, already included in the clinical standards in effect in Portugal, should be monitored until school age (given the increased risk of late-onset hearing loss), such as NBs with intrauterine CMV infection, because deafness may develop up to school age.[15] Children diagnosed with bacterial/viral meningitis who have shown some degree of hearing loss in the audiological evaluation at the time of discharge should also be monitored until school age due to the risk of worsening.[43]

Retrospective studies conducted in maternity units of the Portuguese national health service have shown rates of neonatal screening effectiveness between 98% and 98.7%, with 2% of the NBs being referred for an ORL consultation, which is in line with the goals defined by the JCIH.[44,45] The introduction of new risk factors and warning signs for referral to child health and pediatric consultations aims at increasing the rate of effectiveness of UNHS and increasing the referrals to childhood deafness consultations so that the prognosis of children with deafness in Portugal is improved.

The algorithm developed in this study still requires an evaluation of its impact on the improvement of health care in Portugal and a formal cost-benefit analysis.

Some of the parameters introduced in this algorithm require further studies with larger samples to allow establishing a stronger correlation between the risk factor and childhood hearing loss. In this group, we highlight intrauterine exposure to toxic substances and/or tobacco, as suggested by the Societé Française de Pédiatrie[13].

In addition, we believe that it is essential to create a national database of the audiological results of all the tests of the population with confirmed or at-risk of hearing loss to avoid delayed diagnoses due to absenteeism or difficulties in referral to a specialist. Moreover,

it is a practical, independent, and pragmatic way of assessing the real impact of health care on the quality of life of these patients while evaluating the efficiency of resolution and monitoring of childhood hearing loss.

Conclusion

The proposed algorithm is a tool that can enable an earlier diagnosis of hearing loss during childhood through the identification of the main risk factors associated with deafness in this population in light of the current scientific knowledge. Moreover, we extended the criteria of referral for pediatric hearing screening to other pediatric ages by seeking to define the warning signs for a referral for audiological evaluation in a consultation in the primary healthcare setting. This algorithm may accelerate and increase the number of referrals to childhood deafness consultations, thereby improving the hearing outcomes of children in Portugal.

Conflicts of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

Data Confidentiality

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

Protection of humans and animals

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the 2013 Helsinki Declaration of the World Medical Association.

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Availability of scientific data

There are no datasets available, publicly related to this work.

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