Early definition of type, degree and audiogram shape in childhood hearing impairment

La diagnosi audiologica precoce dell’ipoacusia dell’età pediatrica

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SUMMARY

In the context of permanent childhood hearing loss, early audiological diagnosis is a prerequisite for activation of an adequate rehabilitation program to prevent or limit the known effects that auditory deprivation determines on language development and cognitive skills in neonates. Audiological diagnosis consists schematically of three phases: identification of subjects at risk, definition of hearing loss and/or children features, verification of appropriateness of diagnosis itself and a rehabilitation programme. Strategies and methods of audiological diagnosis are well defined and include an integration of data coming from objective methods with clinical and behavioural data. Although the substantial effectiveness of procedures and a general consensus on their use and interpretation have been defined, there are several critical issues concerning the achievement of this objective, which will be discussed in this paper.

KEY WORDS: Infant hearing impairment • Early audiological diagnosis • Auditory pathways maturation • Universal newborn hearing screening (UNHS) • ABR • SWOT analysis

Introduction

Audiological diagnosis is the prerequisite to activate appropriate measures in order to prevent or limit the effects of permanent bilateral childhood hearing loss (PBHL), either on language development or cognitive skills 1. Three steps are schematically recognised in the process of audiological diagnosis 2 3. The first step is the identification of subjects at risk for hearing loss, and corresponds to the newborn hearing screening and to the surveillance programme; the second step is the audiological diagnosis itself, which must confirm the severity of hearing loss and define its features, before choosing the most suitable hearing aid parameters; a third phase is the follow-up during rehabilitation and hearing aid fitting process, to verify the appropriateness of care and with the option, if needed, of coming back to the second step.

After referral from newborn hearing screening, the adherence to the timeline of the diagnostic work-up allows achieving audiological diagnosis of PBHL at 3-6 months of age 4 5. Late onset hearing loss or lost-to-follow up cases, as well as children carrying audiological risk factors and children suspected for hearing loss by parents or caregivers should also be addressed to an adequate audiological evaluation and/or to follow up.

In confirmed cases of PBHL, the objective of diagnosis is to ascertain hearing loss and define its characteristics in view of the best prosthetic strategy through either amplification or cochlear implantation. Audiological diagnosis must therefore define the entity, nature (conductive, seno-
rineural, mixed), audiometric shape and pathophysiology/dynamic range of hearing, and which are requirements to define the type and operating parameters of hearing aids. Strategy and methods of audiological diagnosis are well defined and are essentially based on integration of data coming from objective methods (electrophysiology/auditory evoked potentials, impedance audiometry, otoacoustic emissions), with clinical data (medical history, otoscopy, clinical examination) and behavioural data. Although the substantial effectiveness of diagnostic and the general consensus on its use and interpretation is established, there are still several critical issues concerning the attainment of diagnosis. The present paper mainly discusses these issues using the methods of SWOT analysis. The present topic has been assigned to the Department of Head and Neck Surgery - Otorhinolaryngology Catholic University of the Sacred Heart “A. Gemelli” Hospital, in the framework of the Italian Ministry of Health project CCM 2013 “Preventing Communication Disorders: a Regional Program for early Identification, Intervention and Care of Hearing Impaired Children”.

Materials and methods

A group of professionals working in tertiary care referral centres was asked to complete a survey on the issues of paediatric hearing impairment evaluation. The survey asked the participants to report at least 2 strengths, weaknesses, opportunities and threats for their strategic planning. This phase was conducted by means of the principles of SWOT analysis. The acronym SWOT stands for Strength (S), Weaknesses (W), Opportunities (O) and Threats (T), and corresponds to what the comments of the participants have pointed out. The responses obtained were reviewed by the specialists responsible for the working group of the appropriate field of interest. A TOWS matrix was the used to match the external threats and opportunities with the internal weaknesses and strengths of the SWOT analysis to generate recommendations. The detailed description of the SWOT and TOWS matrix analysis procedure can be found elsewhere in this issue. The study and the survey were focused on this specific aim: how to define type, severity and morphology of PBHL within 3-6 months of age if the child has been referred from the universal newborn hearing screening, or within one month if the child has been referred from the hearing surveillance programme.

Results

The respondents identified 34 items in the strength category, 43 items in the weaknesses category, 31 items in the opportunities category and 32 items in the threats category, accounting for 140 responses. Based on these responses, several specific themes were generated for each category (Table I).

Table I. Main key points extrapolated from the questionnaires.

| Table Ia. Strengths. |
|----------------------|
| Strength key points | N (%) |
| Performing audiological evaluation structured with all of the diagnostic procedures | 13 (38.2%) |
| Having a qualified, competent and motivated team | 13 (38.2%) |
| Having a standardised diagnostic workup | 4 (11.8%) |
| Having rapid evaluation times /good outpatient care services | 3 (8.8%) |
| Presence of anaesthesiologic care | 1 (3%) |
| Total | 34 (100%) |

| Table Ib. Weaknesses. |
|------------------------|
| Weakness key points | N (%) |
| Technical difficulty in identifying severe-medium frequencies | 10 (23.3%) |
| Lack of resources | 7 (16.3%) |
| Lack of paediatric anaesthesiologic care | 7 (16.3%) |
| Limited technical and specific skills | 6 (14%) |
| Problems linked to late maturation /electrophysiological responses unreliable | 6 (14%) |
| Difficult management of late onset or lost to follow-up | 4 (9.3%) |
| Difficult communication with parents | 2 (4.6%) |
| Difficult management of large volumes of diagnostic procedures | 1 (2.3%) |
| Total | 43 (100%) |

| Table Ic. Opportunities. |
|--------------------------|
| Opportunity key points | N (%) |
| Improvement of technologies | 13 (42%) |
| Presence of dedicated and competent team | 11 (35.5%) |
| Presence of a standardised diagnostic protocol | 3 (9.5%) |
| Better collaboration with the territory | 2 (6.5%) |
| Implementation of data collected by informal assessments | 2 (6.5%) |
| Total | 31 (100%) |

| Table Id. Threats. |
|--------------------|
| Threats key points | N (%) |
| Limited economic resources | 7 (21.9%) |
| Technical limits (false positives-long execution) | 6 (18.8%) |
| Difficult relationship with families for limited cooperation | 5 (15.6%) |
| Difficult management of extraterritorial patients/cultural background | 5 (15.6%) |
| Legal aspects | 4 (12.5%) |
| Difficult management of patient “late onset” or “lost to follow-up” | 2 (6.2%) |
| Inadequate hearing aid | 1 (3.1%) |
| Difficult execution of behavioural tests in children with cognitive deficit | 1 (3.1%) |
| Misinformation | 1 (3.1%) |
| Total | 32 (100%) |
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Strength key points analysis
The 4 most frequently cited strengths points were: being able to perform complete audiological evaluation with all the worthy diagnostic tools (38.2%), having a qualified, competent and motivated team (38.2%), having a standardised diagnostic workup (11.8%), having short waiting lists for evaluation plans thanks to good organisation of services and with easy access to a suitable environment (8.8%) (Table Ia). We describe and discuss the main themes below.

Performing an audiological evaluation enclosing all the worthy diagnostic procedures
In this group, we included all the answers that pertained to the correct evaluation of hearing function (n = 4) and, if PBHL is present, of its pathophysiological features. Evaluation should be quick, based on reliable instrumental equipment (n = 4), matched with an adequate and competent clinical examination (n = 3), and using objective and subjective measures (n = 2).

Having a qualified, competent and motivated team
In this group, we counted all the answers that refer to the presence of a medical/technical staff that is qualified and motivated (n = 7), which operates with devotion (n = 3) and specific competence in paediatric audiology (n = 3).

Having a standardised diagnostic workup
In this group, we enclosed all the answers that refer to the use of an efficient and accepted diagnostic protocol (n = 2), with the preparation, when necessary, of specific and adequate laboratory regulations (n = 1), that allows to obtain audiological diagnosis within a period compatible with an effective rehabilitation programme. We included an individual answer which emphasised that diagnostic procedures should be non-invasive and inexpensive (n = 1).

Having rapid evaluation times/good outpatient care services
In this group, we gathered all the answers that refer to the presence of good organisation of services, with suitable environments for medical procedures (diagnostic, assistance) and counselling, with short waiting lists (n = 2) and easy access to the structure (n = 1). In addition, when available, the strength of the availability of specific anaesthesiological care is reported (n = 1).

Weakness key points analysis
The six most frequently cited weaknesses were: technical difficulty in identifying audiological threshold for middle and low frequency range (23.3%), lack of resources (16.3%), lack of paediatric anaesthesiological care (16.3%), limited technical and specific skills (14%), problems linked to late maturation/unreliability of electrophysiological data (14%), difficult management of late onset cases or lost to follow-up (9.3%) (Table Ib).

We describe and discuss the main themes below.

Technical difficulty in identifying hearing loss at middle and low frequency range
In this group, we gathered all the answers that refer to the lack of a methodology that allows accurate definition of audiometric configuration. We reported the difficulty of an objective diagnosis threshold, for middle and low frequencies (below 1-2 kHz), and the consequent risk of overestimation (in down-sloping thresholds) or underestimation (in up-sloping thresholds) of overall hearing loss (n = 10).

Lack of resources
In this group, we included all the answers that refer to the lack of funds available for the recruitment of a competent and dedicated team (n = 4), and for adjusting and maintaining a complete and technologically updated instrumentation. Lack of funds (n = 2) also cause a shortage in logistical and organisational aspects (e.g. secretarial and archive management) and prolonged waiting lists, as well as poor accessibility to structures (n = 2).

Limited technical and specific skills
In this group, we counted all the answers that refer to the limited technical competence of the staff assigned to the management of paediatric hearing impairment (n = 4), such as in performing behavioural hearing tests, to ensure appropriate sedation and reliable and early diagnosis (n = 1).

Problems linked to late maturation/unreliable electrophysiological data
In this group, we embraced all the answers that refer to the topic of the interference of auditory pathway maturation (n = 5) and transient middle ear conditions (e.g. otitis media) (n = 1) with accurate and timely diagnosis.

Difficult management of late onset hearing loss or lost to follow-up
In this group, we gathered all the answers that refer to the complex management of “late onset“ hearing loss and “lost to follow-up“ patients (n = 2), especially in hospitals where the user comes from distant areas (n = 2), living very far from the referral audiological centre. Other weaknesses reported are issues in the communication with parents (n = 2), especially in the absence of reference professionals (paediatrician, psychologist, child psychiatrist) and the management of large volumes of diagnostic procedures (n = 1).

Opportunity key points analysis
The five most frequently cited opportunities were: possible improvement of technology (42%), presence of
dedicated and competent team (35.5%), presence of a standardised diagnostic protocol (9.5%), improved collaboration among healthcare professionals (paediatricians, speech therapists, hearing care professionals, etc.) of the area (6.5%) and implementation of data collected by informal assessments (6.5%) (Table Ic).

We describe and discuss the main themes below.

Improvement of technologies
In this group, we included all the answers that refer to the elaboration and implementation of new diagnostic technologies (n = 9), through collaboration among companies, basic and clinical research centres (n = 2), in order to improve completeness and reliability of objective audiological diagnosis (n = 2), with the development of new subjective procedures to be implemented as early as possible.

Presence of dedicated and competent team
In this group, we gathered all the answers that consider the availability of a dedicated and specifically trained team (n = 7), with the improvement of an internal program of organisation (n = 2), with better secretarial and logistic services, and easier access to the structure (n = 2).

Presence of a standardised diagnostic protocol
In this group, we enclosed all the answers that refer to the realisation of homogeneous standardised and accepted diagnostic pathways within a regional/national audiological “network” (n = 3).

Better collaboration in the territory
In this group, we took all the answers that refer to the opportunities to strengthen the connections between audiological centre and territorial network (n = 1), highlighting the role of families and paediatricians (n = 1). We also report, in addition to audiological measures, the data collected by informal assessments [information from parents (n = 1), observations of caregivers about auditory behaviour (n = 1)].

Threats key points analysis
The five most frequently cited threats are: limited economic resources (21.9%), technical limits (false positives, prolonged time of execution) (18.8%), difficult relationships with families and limited cooperation (15.6%), difficult management of extraterritorial patients and of families with different cultural backgrounds (15.6%) and legal aspects (12.5%) (Table Id).

We describe and discuss the main themes below.

Limited economic resources
In this group, we enclosed all the answers that refer to the lack of funds available to ensure establishment and strengthening of training and updating courses (n = 3) for all operators involved, to promote and support (n = 3) awareness-raising and information (families, paediatricians), to ensure an adequate technological support (n = 1) and to sustain the efficient organisation of referral centres.

Technical limitations
In this group, we enclosed all the answers that refer to the limitations of current available diagnostic technologies, meaning either the uncertainty of the audiometric threshold for the middle and low frequencies (n = 3), or the need to use sedation during objective tests (n = 3).

Difficult relationship with families for limited cooperation
In this group, we incorporated all the answers that refer to the management of relationship with families that do not always allow an ideal setting for performing subjective tests (n = 2) and or to perform sedation when needed (n = 3).

Difficult management of extraterritorial patients/cultural background
In this group, we gathered all the answers that refer to the difficulties encountered in achieving diagnostic information on the family, which is not always willing to accept the audiological diagnosis or the diagnostic workup. This could be an issue in centres where users are often multicultural (n = 2), with the difficulties encountered in the management of diagnostic pathway for the extraterritorial patient (n = 3), considering the lack of a good internal and territorial organisational network.

Legal aspects
In this category, we included all the answers concerning the improvement of the polices (n = 4) that are prerequisites to obtain the necessary legal and economic support. In addition, other reported threats are: difficult management of patient “late onset” cases or “lost to follow-up” patients (n = 2), inadequate fitting of hearing aid (n = 1), low reliability of behavioural tests in children with cognitive deficits (n = 1) and misinformation (n = 1).

Discussion
The objective of this analysis was to obtain general recommendations regarding the activity of “defining the type, degree and audiogram shape of hearing impairment within 3-6 months of life if the hearing loss was identified through the UNHS, or within one month from hearing impairment identification if the child was referred after a neonatal age”. Starting from the SWOT analysis data, a TOWS matrix was created, which compares Strengths-Opportunities, Weaknesses-Threats, Weaknesses-Opportunities, eventually offering recommendations and directions that can constitute an effective starting point for prospective planning and innovation, in particular for tertiary care audiology centres.
Analysing and discussing the data obtained from this research, 16 recommendations were obtained (Table II), grouped in four main areas, which identify strategies directed to minimise external obstacles and enhance opportunities:  
1. To have a competent and dedicated working group, constantly updated, able to make a comprehensive audiological evaluation, supported by all reference professional.  
2. To use a validated diagnostic protocol, with reference to scientific progress and national and international standards, which includes integration of clinical and instrumental data; to promote the development of clinically available procedures through collaboration between companies and research centres, in order to obtain improvement and spreading of diagnostic technologies.  
3. To improve collaboration and communication between tertiary care audiological centres and territorial references (families, family paediatrician, territorial rehabilitation), promoting awareness and information at the local level, thus facilitating the access to tertiary care audiology centres for families.  
4. To obtain or to improve legislation and regulations regarding early hearing impairment detection and interventional programmes, in order to create conditions that facilitate economic and organisational support by institutions.  

The first strategy includes establishing and maintaining of a competent and dedicated working group that is able to comprehensively and reliably manage the audiological evaluation of a newborn or a young child, which respects the time needed to activate the rehabilitative programme. The audiological team needs to be supported in the diagnostic phase by other referral professionals (i.e. clinical geneticist, paediatric neuropsychiatrist, anaesthesiologist) who are skilled in childhood audiological and developmental issues. The anesthesiologist is often essential in audiological diagnosis to ensure adequate conditions of examination, especially in complex cases. The importance of presence/availability of a paediatric anaesthesiologist was strongly emphasised by the participants of this SWOT analysis.  

Having a competent and cohesive working group that operates in logistically and structurally appropriate conditions is the foundation for the second most complex strategy: implementation of a validated diagnostic protocol, which refers to scientific evidence and to national and international standards, and that is based on integration of clinical and instrumental data. Clinical data essentially include general clinical and otoscopic examination and medical history, paying special attention to audiological risk factors revision and to parental observations about the child’s auditory behaviour.  

Regarding instrumental data, it is widely accepted that audiological evaluation in the first year of life must be based on all objective data gathered from the following measurements: immittance measures (including tympanometry, preferably with probe tone of 1000 Hz in the first 6 months of life, ...
and ipsilateral and contralateral cochleo-stapedial reflexes thresholds); diagnostic oto-acoustic emissions (transients or distortion products, i.e. TEOAEs or DPOAEs); electrophysiology (i.e. electrocochleography, auditory brainstem responses (ABR), and cortical auditory potentials). An ABR is obtained by air conducted transient stimuli (click), eventually integrated by bone conduction testing, when there is a doubt about the conductive component/nature of hearing loss. The examination should include the use of frequency-specific stimuli (low frequencies) when there are doubts or it is necessary to better define the audiometric configuration. This aspect can become critical both in hearing impairment with differential high- or low-frequency involvement, in which click ABR measurements can cause false positive (overestimation of hearing threshold) or false negative results (underestimation of hearing threshold), respectively. Along with tone-burst stimuli, the results obtained with the recently introduced “CHIRP”, spectral default composition stimuli, appear promising.

The specific informative content of each diagnostic procedure contributes to a comprehensive audiological evaluation that allows in most cases the definition of the main characteristics of hearing impairment that are propedeutic to the therapeutic choices. This SWOT analysis emphasised the possibility of potential ambiguity and complexity in diagnostic evaluation, which can be attributed to various conditions. The first and most common problem is the coexistence of a conductive component of the hearing loss, which is a frequent condition in early childhood, this condition can be evidenced by otoscopy and tympanometry. The condition does not represent a significant problem if the estimated ABR hearing threshold is measured within 40 dB HL. Beyond this limit, electrophysiological data cannot always differentiate the main type of hearing impairment, i.e. the distinction between a conductive or sensorineural hearing loss or the amount of the conductive component of a mixed hearing impairment. The ambiguity is not always easy to overcome in these cases, and can lead to wrong therapeutic choices or delayed diagnosis.

An additional aspect of variability and diagnostic error (mainly overestimation of hearing impairment) involves the “development” of the hearing system, especially in its neural component and electrophysiological data. Maturational phenomena involve the whole auditory system, from the periphery to the cortex, but they are especially supported by processes of myelination and synaptogenesis of neural structures. These phenomena are well known and demonstrated in many studies concerning the effects on evoked potentials in time domain in normal subjects.

In the neonatal period, in particular in pre-term infants or in infants hospitalised in intensive care units (ICU) various conditions are frequently associated with alterations of electrophysiological epiphenomena of maturational processes (increased the latency and threshold of evoked potentials).

Other conditions of possible “interference” during audiological diagnosis are represented by cases of “downstream sensory function” compartment (cyto-neural/neural junction, VIII nerve, central auditory pathways) dysfunction, where the impairment of temporal and dynamic characteristics of afferent neural activation may cause important clinical effects on verbal perception and a deterioration or disappearance of electrophysiological responses. These audiological profiles were initially named “auditory neuropathy” and later included in the group of diseases described as “synaptopathy” or “dyssynchrony”. They are also found in the neonatal period and, again, most commonly in pre-term infants usually hospitalised in the ICU. These conditions are able to sustain a mismatch between good peripheral capabilities (middle ear-cochlea) and an abnormal function of afferent pathway “downstream” to the cochlea. It may cause a wrong diagnosis of hearing impairment or its even extreme overestimation that is not always predictable by medical history.

The presence and the possible coexistence of these interfering conditions can complicate audiological diagnosis. An extreme situation is represented by cases with no ABR responses that lead to a diagnosis of a severe/profound sensorineural hearing impairment, with an estimated threshold equal to or higher than 90 dB HL. This result may, in fact, match several conditions: a profound hearing impairment that represents a primary indication for a cochlear implantation; a cochlear hearing impairment rapidly decreasing for frequencies beyond 500/1000 Hz; a moderate or moderate-severe hearing impairment associated with a coexistent conductive component or neural dysfunction. These uncertainties may not be always resolved, even with careful integration of results by different diagnostic methods. It is therefore mandatory to apply careful clinical/audiologic tools and frequent subjective and objective audiometric monitoring. It is author’s opinion that severe/profound hearing impairment cases should be considered for a cochlear implant selection process after 6-8 months of age in infants without associated audiologic risk factors, and by 70-85 weeks of corrected age in pre-term infants. Complex cases should be considered for electrocochleography (ECoG) with trans-tympanic derivation, as a second choice procedure. The trans-tympanic optimises the assessment of auditory periphery level and provides unique information for differential diagnosis of cytoneural/neural compartment dysfunctions. At this stage of audiological childhood diagnosis, we must also consider the steady-state auditory evoked potentials, which are able to provide a high frequency specificity in estimating the audiometric threshold (80-100 Hz SSR), even if there are no data concerning reliability and applicability of this time consuming method. Audiologic centres of second or, even more, third level (where comprehensive diagnosis is performed) must be able to apply and interpret all diagnostic procedures. While in most cases test is obtained in spontaneous sleep, it is nec-
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Audiologic centres should adhere to strict standards and it is hoped that initiatives are taken at national level to define shared practices. The centre should, however, take all measures (development of own rules, calibration and maintenance of instruments) aimed at controlling the biological (age/maturity, etc.) and non-biological factors (type and method of acoustic stimulation, etc.) which can be a source of diagnostic errors.

Current procedures of subjective behavioural audiometry are considered as having a limited role in early audiological diagnosis, although their value grows in the first year of life and acquires a decisive role in the diagnostic and rehabilitative process, especially in cases selected for CI. There is a strong need to develop new and reliable behavioural evaluation procedures that can be appropriate since the first months of life. Improvements and further developments of subjective and objective procedures is expected, although there are methodologies that guarantee adequate reliability in early audiological diagnosis. They can be achieved only by enhanced collaboration between clinical and basic research centres, and companies of diagnostic equipment production and distribution.

Another recommendation involves the diagnostic “formalisation” and professional-family alliance. A full audiological diagnosis should include the results and their significance in a complete and clear style, including explicit indications for rehabilitation. The report should be given to the family and to the operators involved in the prospective management of the young patient (hearing aid professional, speech therapist, paediatrician etc.)

Clear and complete information is the core of childhood audiological diagnosis and is the foundation of the management alliance among all those professionals (healthcare operators, pedagogical and social workers, family) who are part of the rehabilitation programme. SWOT participants consider this approach as an appreciated and an essential element for its success.

The third identified strategy concerns access to the diagnostic process and services through an extensive network in the area, in cooperation with families and family paediatricians, which recalls the principle of an approach based on a well-founded alliance among all the figures and institutions involved in the diagnostic and intervention programme for hearing impairment. Having good internal organisation would reduce the number of “lost to follow-up” cases and obtain maximum accuracy and timeliness in diagnosing “late onset” hearing impairment cases. This has been reported as particularly important for the large referral centres that frequently treat children and families that come from far away. Creating a single network between the third level centre and the territory resources would allow more effective management of medical treatment of common respiratory infections and associated conductive hearing impairment, or other conditions that affect the auditory system. Moreover, they can ease the organisation of training and update courses for healthcare professionals and public information and awareness campaigns. Diagnostic information has to be provided to the family in a clear and explicit manner, in terms of both therapeutic indications and prognosis and expectations. Diagnosis has to be accurate and formalised in an understandable report. It should be addressed to the family and the family paediatrician, whose cooperation is central for diagnosis, control of variability factors or diagnostic confusion, and of course future follow-up and treatment. The existence of an effective network can guarantee accurate information and ensure an easy access to audioligic centres, achieving the goal of making easier and faster the contact with the family.

The fourth strategy concerns legislative and regulatory issues that are the prerequisites for obtaining adequate legal and administrative support. It is certainly possible to achieve operational improvements through organisational and procedural adjustments inside the centres, although this analysis highlighted that substantial changes cannot be separated from normative, legislative and, accordingly, economic aspects. Enhancement of didactical aspects, improvement of information and connection methods, acquisition of new tools and validation of new procedures are all necessarily subordinated to economic aspects. Considering the current situation characterised by limited resources and a progressive contraction in healthcare expense, the group opinion emphasised the requirement of a commitment that can raise the awareness of national policymakers and administrative managers, of local authorities and in its operating area about the importance and the effectiveness of early diagnostic and treatment programs for childhood hearing impairment.

Conclusions

In the field of audiological diagnosis, first of all diagnosis should come from a competent and constantly updated dedicated working group. Secondly audiological evaluation should be based on a validated diagnostic protocol, with reference to national and international standards, which includes integration of clinical and instrumental data. Thirdly collaboration and communication between the audiological centre and territorial referent should be optimised, largely obtaining or improving legislation and regulations regarding the program of early hearing loss detection are necessary requirements to create the conditions to facilitate economic and organisational support by institutions.
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