

AUDIOLOGY

Universal newborn hearing screening in the Italian Region of Sicily in 2018

Screening uditivo neonatale universale nella regione Sicilia nel 2018

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SUMMARY

Objectives. We have clarified the role of Universal Neonatal Hearing Screening (UNHS) for both early diagnosis and rapid treatment in order to improve the prognosis of the deaf child and reduce patient management costs. Although in Sicily UNHS has been progressively implemented, there is scarce data in the literature on this matter. Therefore, the main objective was to collect in the year 2018 the following data: number of newborns screened for hearing loss, number of infants “referred” to transiently evoked otoacoustic emissions (TEOAE), number of infants with pathologic auditory brainstem response (ABR) and number of infants affected by permanent hearing loss.

Methods. UNHS monitoring was conducted through the collection of data through a questionnaire, which was analysed evaluating the effectiveness and adherence to the screening program prepared by the Department for Health Activities and the Epidemiological Observatory (DASOE).

Results. In 2018, there were 40,243 newborns in Sicily. A total of 37,562 newborns were screened (93.3%). There were 1,328 “referred” infants with TEOAE (3.5%). On the 2nd level, “referred” newborns examined were 1,080 of 1,328 expected (missing 248 “refer” newborns, equal to 18.6%). The number of “referred” infants confirmed with TEOAE was 113 of 1,080, while “referred” infants confirmed with ABR were 71. On the 3rd level, 67 of 71 were infants examined: 28 infants were suffering from monolateral hearing loss (13 slight/mild, 13 moderate, 1 severe and 1 profound) and 39 from bilateral hearing loss (1 slight/mild, 19 moderate, 13 severe and 7 profound). Excluding 7 infants from the NICU, 60 of 37,562 infants had hearing loss (1.5%).

Conclusions. The monitoring of the UNHS in Sicily has allowed obtaining the data of individual centres, absent in the literature to date, to verify the effectiveness of the screening, according to JCIH criteria, to highlight some criticalities and, finally, to propose possible solutions.

KEY WORDS: neonatal hearing loss, universal newborn hearing screening, congenital deafness

RIASSUNTO

Obiettivi. Abbiamo chiarito il ruolo dello screening uditivo neonatale universale (UNHS) sia come diagnosi precoce che come trattamento rapido al fine di migliorare la prognosi del bambino sordo e ridurre i costi di gestione del paziente. Anche se in Sicilia l'UNHS è stato progressivamente attuato, vi sono scarsi dati nella letteratura in materia. Pertanto, l'obiettivo principale è stato quello di raccogliere nell'anno 2018 i seguenti dati: numero di neonati sottoposti a screening per la perdita dell'udito, numero di neonati risultati “refer” alle TEOAE, numero di lattanti con ABR patologico e numero di lattanti affetti da perdita permanente dell'udito.

Metodi. Il monitoraggio del UNHS è stato condotto attraverso la raccolta dei dati forniti dal questionario, che valuta l'efficacia dello screening e l'aderenza alle fasi successive.

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Conflict of interest

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Risultati. Nel 2018, i nati in Sicilia sono stati 40.243. Sono stati sottoposti a screening 37.562 neonati, pari al 93,3%. Sono risultati “refer” con le TEOAE 1.328 neonati (pari al 3,5%). Al 2° livello, i neonati “refer” esaminati sono stati 1.080 su 1.328 attesi (dispersi 248 neonati refer, pari al 18,6%). I neonati “refer” confermati con le TEOAE sono stati 113 su 1.080, mentre i neonati “refer” confermati con ABR sono stati 71. Al 3° livello, i bambini esaminati sono stati 67 su 71 previsti: ventotto bambini soffrivano di perdita monolaterale dell’udito (13 lieve, 13 moderata, 1 grave e 1 profonda) e trentanove soffrivano di perdita bilaterale dell’udito (1 lieve, 19 moderata, 13 grave e 7 profonda). Escludendo 7 bambini provenienti dalla NICU, i bambini con perdita dell’udito sono stati 60 su 37.562 (pari all’1,5%).

Conclusioni. Il monitoraggio dell’UNHS in Sicilia ha permesso di ottenere i dati dei singoli centri, dati assenti nella letteratura, di verificare l’efficacia dello screening, secondo criteri JCIH, di evidenziare alcune criticità e, infine, di proporre possibili soluzioni.

PAROLE CHIAVE: ipoacusia neonatale, screening universale uditivo neonatale, sordità congenita

Introduction

Significant permanent hearing impairment (more than 30-40 dB HL in both ears) is the most common sensory disability, and represents a relevant problem not only for the health of the child, but also for the economic and social costs that follow¹⁻³.

Significant bilateral hearing impairment, if undetected, can cause profound effects on speech, language and cognitive development, and can thus hamper emotional and social well-being⁴⁻⁶.

In Italy and Western countries, it is estimated between 0.5 and 2 cases per 1,000 live births have profound hearing loss⁵⁻⁷. Nevertheless, in some paediatric subpopulations, the rates are significantly higher (about 4%), especially in the presence of risk factors or in neonates hospitalised in the Neonatal Intensive Care Unit (NICU)⁶⁻¹².

The latest universal screening diagnostic protocols using TEOAE and automated auditory brainstem response (A-ABR) have shown a sensitivity (percentage of children with abnormal hearing who fail the test) close to 100% and specificity above 90%¹³⁻¹⁶.

To be successful, a neonatal hearing screening programme should be universal (i.e., include all neonates), because selective screening, based on high-risk criteria, detects at most half of all infants with congenital hearing loss¹⁷.

A further control process is the one modified in 2019 by JCHI which, noting promising results from the respect of benchmarks 1-3-6 (complete screening within 1-month, audiological diagnosis within 3 months, early intervention within 6 months), invited the audiological reference centres to perform a reduced temporal sequence 1-2-3¹⁸.

Although in Sicily the UNHS has been progressively implemented since 2011 and currently the coordination is entrusted to the Department for Health Activities and the Epidemiological Observatory (DASOE), there is scarce data in the literature on this matter. Interestingly, from 2011 to 2017, the percentage of children screened increased every year as follows: 6%, 18%, 32%, 57%, 87%, 92%, 98%. However, in 2018 a lower coverage of 37,303 (91.6%) out of 40,645 screened newborns was reported¹⁹.

In 2016, 2,160 (5.7%) were registered as “referred” out of a population of 37,250 newborns, of whom 95 were diagnosed with deafness (2.5 per 1,000)¹⁹.

From these reported rates it follows that, in order to evaluate the efficiency of the UNHS correctly, it is necessary not only to evaluate the coverage rate, but equally essential to know information relating to the rate of “referred” and newborns diagnosed with hearing loss.

Through these assumptions, we have developed and proposed a questionnaire sent to birth centers and Otolaryngology Units in Sicily in the year 2018 to collect the following data: number of newborns screened for hearing loss, number of infants resulted refer to TEOAE, number of infants with pathologic ABR and number of infants affected by permanent hearing loss.

Materials and methods

The search covered the 9 Sicilian provinces: Palermo, Catania, Messina, Agrigento, Trapani, Ragusa, Caltanissetta, Siracusa and Enna. The screening was performed following the protocol designed by DASOE¹⁹. The UNHS program is structured in three levels, each performed in an adequately equipped centre.

The first level is represented by public and private birth points. The second level is represented by the Audiology Services, autonomous or aggregate to ENT-Unit. The Regional Reference Centres represent the third level.

According to the screening protocol designed by DASOE, all birth points must screen all newborns by searching for (TEOAE), during spontaneous sleep and before hospital discharge (generally, within 48-72 hours).

“Pass” test infants leave the screening process, while “referred” infants (mono or bilaterally) must be re-examined, within the first month of life, with TEOAE. If the test result is “referred”, it is necessary to perform the ABR at the Audiology Services (Level 2). In case of “referred” confirmation, the newborn is sent to the Regional Reference Centre (Level 3) within the third life month for definitive diagnosis, and starting prosthetic rehabilitation no later than the sixth month of life.

The infants hospitalised at the NICU, or the ones who bilaterally “pass” both screening tests (TEOAE and ABR), but who have risk factors for late-onset hearing loss, are sent to level II centres for an audiological surveillance programme that includes assessment every 6-12 months in the first three years of life (Tab. I).

UNHS monitoring was conducted through the collection of data from a questionnaire sent to and filled in by the UNHS program’s coordinator of each birth point of the National Health System (NHS) in Sicily in the year 2018. All coordinators invited to participate in the surveys returned the questionnaires filled in. For each birth point participating in the survey, the following figures were collected:

1. total births in the year;
2. number of newborns screened for hearing loss;
3. number of infants “referred” to TEOAE;
4. number of infants transferred to NICU, premature, deceased.

For each ENT-Unit (level 2), participating in the survey, the following figures were collected:

1. number of expected infants resulting “refer” and their birth point;
2. number of confirmed infants “referred” to TEOAE;
3. number of infants subjected to ABR;
4. number of infants with pathologic ABR.

For each Regional Reference Centre (level 3) participating in the survey, the following figures were collected:

1. number of infants affected by monolateral hypoacusis (mild, moderate, severe and profound);
2. number of infants affected by bilateral hypoacusis (mild, moderate, severe and profound).

The data collected by this survey, expressed in numbers, concern procedural aspects and are used for the generation of reports useful for the strategic planning of the health sys-

tem. The collected data cannot be traced back to a specific patient according to the privacy rights of the children and their families.

In the absence of regional indicators of screening efficiency, the data collected in the questionnaire were analysed considering the indicators recommended by JCIH, which allow evaluating the effectiveness of the screening and adherence to the subsequent phases.

More specifically, the quality indicators for screening and confirmation of deafness are the following:

- the percentage of infants who completed the screening should be more than 95% of the neonatal population during the first month of life;
- the percentage of infants, without risk indicators, who passed neither the initial testing nor the subsequent ones - “referred” confirmed should be below 4% of evaluated infants;
- the audiological evaluation must be completed in at least 90% of children who do not pass the screening.

Hearing loss can be classified or defined in many ways and categories. This study has used the classification of the WHO: slight/mild (26-40 B), moderate (41-60 dB, severe (61-80 dB) and profound (over 81 dB). In the case of moderate hearing loss, the range for children is 31-60 dB.

Results

This report shows the data relating to UNHS of all birth points of Sicily in the year 2018.

The data relating to the activity of the UNHS included 46 of 47 birth points active in 2018, of which 36 public hospitals, 3 University-Hospitals (Palermo, Catania and Messina) and 7 private structures affiliated with the NHS.

The birth points of Bronte and Biancavilla (province of Catania) were unified in the chart (Tab. II), while the data of the birth point of Pantelleria were not received (number of newborns < 50).

Table II shows the data for each birth point: in total there were 40,243 newborns, of which 37,562 were screened (equal to 93.3%) and, of these, 1,328 infants resulted “referred” to TEOAE (equal to 3.5%) and were sent to the 2nd level.

Only a few coordinators have reported the number of newborns transferred to NICU, the premature ones and the deceased ones. The territorial screening coverage, a key indicator to assess the adherence to the program, was 93.3%, above 92% reported by DASOE, but still below the international standard (95%).

In the years 2015, 2016 and 2017, the territorial coverage had been, respectively, 87%, 93% and 98%¹⁹.

It should be noted that, in the year 2018, the screening cover-

Table I. Risk indicators associated with permanent congenital hearing loss in childhood (DASOE, 2016)¹⁹.

Family history of permanent childhood hearing loss
Birth weight less than 1500 g
Neonatal intensive care
Hyperbilirubinemia that requires exchange transfusion
Exposure to ototoxic medications
In utero infections, such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis
Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits
Syndromes or physical findings, such as white forelock, that are associated with hearing loss
Neurodegenerative disorders or sensory motor neuropathies
Meningitis

Table II. Overall data for hospitals.

Birth points	Borns	TEOAE	% screened	Refer	% refer	Trasf/Prem/Dec
Palermo-Poi.	611	608	99.5%	18	3.0%	ND
PA-Arnas C.	1374	1193	86.8%	13	1.1%	ND
PA- Buccheri	2050	2046	99.8%	84	4.1%	ND
PA-CDC Zancla	1123	1106	98.5%	12	1.1%	16
PA-CDC Serena	1014	1014	100.0%	28	2.8%	ND
PA-Ingrassia	578	567	98.1%	9	1.6%	ND
PA-Cervello	1617	1550	95.9%	11	0.7%	ND
PA-CDC Candela	1176	1150	97.8%	13	1.1%	20-0-0
Corleone	225	225	100.0%	1	0.4%	ND
Partinico	452	296	65.5%	2	0.7%	ND
Cefalu'	400	385	96.3%	1	0.3%	ND
Termini Imerese	614	602	98.0%	5	0.8%	12
Catania-Po I	2009	1692	84.2%	267	15.8%	ND
CT-Cannizzaro	1276	1101	86.3%	23	2.1%	27-0-0
CT-S.Bambino	1818	1658	91.2%	246	14.8%	5-0-6
CT-Garibaldi Nes	2082	2082	100.0%	64	3.1%	ND
Acireale	524	198	37.8%	0	0.0%	ND
Biancavilla-Brante	689	603	87.5%	2	0.3%	4-0-0
Caltagirone	587	438	74.6%	7	1.6%	ND
CDC Gretter	568	568	100.0%	6	1.1%	ND
CDC Falcidia	808	795	98.4%	16	2.0%	13-0-0
Messina Poi	1404	1281	91.2%	155	12.1%	91-ND-ND
Me-Papardo	689	640	92.9%	3	0.5%	0-74-2
Patti	843	839	99.5%	1	0.1%	3-0-1
Milazzo	714	492	68.9%	1	0.2%	5-0-0
Sant'Agata M.	307	307	100.0%	1	0.3%	ND
Taormina	549	274	49.9%	7	2.6%	16-0-0
Ragusa	1464	1443	98.6%	14	1.0%	9-2-6
Vittoria	880	838	95.2%	2	0.2%	35-0-5
Modica	856	832	97.2%	66	7.9%	33-0-0
Agrigento	1523	1484	97.4%	128	8.6%	ND
Canicatti	611	586	95.9%	9	1.5%	ND
Licata	270	265	98.1%	ND	ND	ND
Sciacca	607	595	98.0%	8	1.3%	8-0-2
Trapani	961	950	98.9%	12	1.3%	0-12-0
Marsala	530	522	98.5%	3	0.6%	8-0-0
Castelvetrano	376	332	88.3%	21	6.3%	ND
Mazzara del Vallo	463	431	93.1%	26	6.0%	ND
CDC-Sant'Anna	510	506	99.2%	16	3.2%	4-0-0
Siracusa	1473	1473	100.0%	ND	ND	ND
Lentini	1113	1113	100.0%	ND	ND	ND
Caltanissetta	582	567	97.4%	3	0.5%	15-0-0
Gela	757	751	99.2%	7	0.9%	6-0-0
Enna	975	975	100.0%	17	1.7%	ND
Nicosia	191	189	99.0%	0	0.0%	2-0-0
TOTAL	40,243	37,562	93.3%	1,328	3.5%	

Borns: Total number of births in the year; TEOAE: Transient Evoked Otoacoustic Emissions screened infants; % Screened: Relation between number of screened infants and borns; Refer: Infants number resulted Refer to TEOAE and sent to the ENT Unit 2nd level; % Refer: Relation between number of infants resulted Refer and screened; Trasf/Prem/Dec: Number of infants transferred to NICU, premature, deceased; Palermo-Pol: Policlinic of Palermo; PA: Palermo; CdC: Nursing home; Catania-Pol: Policlinic of Catania; CT: Catania; ME: Messina; PA-Arnas C.: ARNAS Civic Hospital of Palermo; CT/ S. Bambino: S. Bambino Hospital of Catania; Garibaldi Nes.: Garibaldi Hospital of Catania; Messina-Pol.: Policlinic of Messina; S. Agata M.: Sant'Agata di Militello (ME) Hospital; ND: Data not available.

age was more than 95% in 7 of 9 provinces, while it was lower in the provinces of Catania and Messina; the percentage of screening in the provinces of Catania and Messina was 88.2% and 85.1% respectively. The reasons for these low percentages reported by the persons in charge of the birth points were device breakage and temporary absence of trained personnel.

In 2018, the percentage of infants “referred” to the test and retest was 3.5%, equal to 1,328 children. This data, which is not present in literature, satisfies the quality indicator (4%); however, in 7 birth points, it exceeds the threshold value.

The Neonatology Unit of the Catania Policlinic screened 1,692 infants out of 2,009 (84.2%): 1,482 (73.7%) at the Nursery and 210 (10.4%) at the NICU. Newborns resulted “referred” were 267 (15.8%) of 1,692: of which 262 (15.5%) at the Nursery and 5 (0.3%) at NICU.

The Neonatology Unit of Santo Bambino Hospital of Catania screened 1,658 infants out of 1,818 (91.1%): 1,405 (84.7%) at the Nursery and 253 (15.3%) at NICU. Newborns resulted “referred” were 246 (14.8%) out of 1,658: of which 211 (85.8%) at the Nursery and 35 (14.2%) at NICU. Fifty-seven infants did not show up for the test, which had been postponed due to the temporary absence of staff.

The neonatology data of the Policlinic of Messina were recovered thanks to the ENT Unit: 1,281 infants out of 1,404 have been screened (91.2%). The infants resulted “referred” were 155 (12.1%). Newborns transferred to the NICU were 91. Moreover, it was not possible to trace the causes that led to the dispersion of the “referred” babies due to the lack of a paper or computer tracking system.

On the 2nd level (Tab. III), “refer” newborns examined

Table III. Overall data on the 2nd and 3rd level.

ENT Unit	Level 2					Level 3								
	Newborn examined and Birth points	Refer TEOAE	ABR perf.	ABR Pathol.	Monolateral Hearing loss			Bilateral Hearing loss						
					L	M	S	P	L	M	S	P		
Palermo A.O.U. Policlinico P. Giaccone	15 - A.O.U. Policlinico	6	11	3							2			1
	18 - ARNAS Civico	7	17	2									2	
	21 - Buccheri/LaFerla	6	21	6		3					2	1		
	9 - Ingrassia	4	9	3	ND									
	31 - Cervello	8	16	6		2					4			
	6 - CdC Zancla	4	6	2							1	1		
	8 - CdC Serena	2	8	2	1						1			
	5 - CdC Candela	1	5	1									1	
	1 - CdC Sant'Anna Erice	0	1	0										
	1 - Mazara del Vallo	1	1	1										1
	1 - Licata	1	1	1					1					
	8 - Civile Termini Im.	1	3	1								1		
	4 - Osp di Agrigento	2	4	1										1
	3 - Giglio di Cefalù	1	3	1								1		
	1 - Castelvetro	1	1	1										1
	1 - Osp di Trapani	0	1	0										
	1 - Ospedale di Enna	1	1	1								1		
Termini Im. A.O. Cimino	5 - Cimino Term. Im.	5	5	0										
	9 - Ingrassia Palermo	9	12	3							1	2		
	1 - Civico di Corleone	1												
Catania A.O.U. Policlinico	2 - Civico di Partinico	2												
	267 - Policlinico	1	1	1										1
	210 - NICU Policlinico	5	3	1		1								
	170 - S. Bambino	3	3	3	1	1				1				
	11 - NICU S. Bambino	0	11	0										
2 - Garibaldi Catania	2	2	1											1

continues ►

Table III. Overall data on the 2nd and 3rd level (follows).

ENT Unit	Level 2				Level 3								
	Newborn examined and Birth points	Refer TEOAE	ABR perf.	ABR Pathol.	Monolateral Hearing loss			Bilateral Hearing loss					
					L	M	S	P	L	M	S	P	
Catania A.O.U. Policlinico	3 - Cannizzaro CT	3	3	0									
	5 - ASP di Acireale	5	5	0									
	2 - Biancavilla/Bronte	2	2	1									1
	0 - ASP Caltagirone	0	0	0									
	1 - CdC Gretter	1	1	0									
	0 - CdC Falcidia	0	0	0									
	1 - Umberto I - Enna	1	1	0									
	1 - Civile di Ragusa	1	1	1	1								
	2 - Osp. Lentini	2	2	0									
1 - Umberto-Siracusa	1	1	1	1									
ARNAS Garibaldi	ND												
Messina A.O.U. Policlinico	125 - Policlinico	7	7	3		2					1		
	91 - NICU Policlinico	10	10	6		3					2		1
Patti	1 - Barone di Patti	1	1	1	1								
	1 - Sant'Agata Militello	1	1	1	ND								
Taormina	2 - Osp. di Taormina	2	2	2		1					1		
Ragusa A.O. Civile	1 - Civile di Ragusa	1	1	1							1		
	0 - Osp. di Vittoria	0											
	0 - Osp. di Modica	0											
Agrigento	6 - Osp. Agrigento	6	6	6	6								
Sciacca	3 - Osp. di Sciacca	3	3	2			1						1
Trapani A.O.	3 - Osp. di Trapani	3	3	1									1
	1 - Osp. di Marsala	1	1	0									
Sant'Antonio Abate	10 - Castelvetro	10	6	3	2								1
	7 - Mazara del Vallo	7	5	0									
	2 - CdC Sant'Anna Erice	2	2	1									1
Siracusa Umberto I	0 - Osp. di Siracusa		26	0									
TOTAL	1,080	113	235	71	13	13	1	1	1	19	10	9	
									67				

ENT: Otorhinolaryngology; TEOAE: Transient Evoked Otoacoustic; ABR perf.: ABR performed; ABR pathol.: Pathological ABR; L: slight/mild; M: moderate; S: severe; P: profound; NICU: neonatal intensive care unit; ND: data not available.

were 1,080 out of 1,328 expected (missing 248 newborns). The number of “referred” infants confirmed with TEOAE was 113 out of 1,080 and the number of “refer” infants confirmed with ABR was of 71. In ENT Unit of the Policlinic of Messina, the “referred” infants examined with the combination of TEOAE and A-ABR were 125 of 145 (20 children contacted didn't show up), of which 3 children were confirmed “referred” and inserted in the 3rd level of screening. All infants from the NICU, 91 in total, were examined at 2nd level by TEOAE and ABR and, of

these, 6 were confirmed “referred” and inserted in the 3rd level of screening.

On the 3rd level, 67 of 71 infants were examined: 28 infants were suffering from monolateral hearing loss (13 slight/mild, 13 moderate, 1 severe and 1 profound) and 39 from bilateral hearing loss (1 slight/mild, 19 moderate, 13 severe and 7 profound).

The percentage of “referred” newborns at 1st level of screening that ended the diagnostic process was 81.0% (1,080 – 4 = 1,076 of 1,328).

Excluding 7 infants from the NICU, 60 out of 37,562 infants had hearing loss (1.5%).

Discussion

The adverse effects of childhood hearing loss are mitigated through universal newborn hearing screening and early action. In a recent systematic review and meta-analysis, conducted in highly developed countries, the prevalence of childhood hearing loss was 1.1 per 1,000 screened children and was 6.9 times higher among those admitted to NICU²⁰. In our study, from the overall data of the 1st level, some aspects of inhomogeneity emerged. In 11 of 46 birth points, the activity had been interrupted for a few months, due to the TEOAE device breakdown, with consequent less program adherence. The absence of a secondary device and a recovery procedure has caused a large dispersion number of newborns. However, a significant number of children did not show up on the 1st level check-up. It is necessary to issue guidelines to reduce dispersion.

In 7 of 46 birth points a high percentage of “referred” infants was present, which overcomes the quality indicator mentioned above (< 4%); some coordinators assumed that it is due to the replacement of health professionals assigned to the screening.

In order to minimise the number of infants to be addressed to audiological follow-up, an outpatient rescreening protocol should be provided within the first month of discharge. In the literature, it has been shown that the use of A-ABR in addition to TEOAE in neonates without audiological risk factors can reduce the number of false positives¹⁶.

The percentage of “referred” newborns to screening and concluding the diagnostic procedure was 81.0%, below the benchmark of 90%¹⁶.

It was not possible to trace the causes that led to the dispersion of the “referred” babies due to the lack of a paper or computer tracking system. The paediatrician could be useful for this; however, the role of the family paediatrician in neonatal audiological screening and audiological surveillance is not defined in the regional plan of prevention 2014-2018.

The analysis of the data collected at points of birth underline that no distinction was made between newborns with or without risk factors for hearing loss. Therefore, in centres where births are more numerous or where there is intensive care, a close relationship between the birth point and the Audiology centre is desirable.

Another criticality of data communication was found in second and third level centres, in which no discrimination between the group of new-borns from neonatology and the one from NICU was made. Therefore, the protocols for

communication of data to the regional register should be well defined.

The JCIH states that infants with risk factors should perform both the TEOAE (often present) and the A-ABR before discharge from the NICU, because the A-ABR is more sensitive to recognising retrocochlear hearing loss¹⁶. Infants “referred” to one or both ears should be sent to a 3rd level, within the third/fourth month of life corrected for gestational age and should undergo periodic audits every 6 months during the first 3 years of life. The JCHI 2019 Declaration approved, only for infants with failed A-ABR screening, that the review and transition to TEOAE are acceptable, given the very low incidence of auditory neuropathy in this population¹⁸. Recently, Frezza et al. reported a trend towards improvement of hearing threshold in 47% of very preterm infants with normalisation of initial mild-moderate hearing losses at final diagnosis²¹.

Furthermore, no diagnostic strategy has been identified regarding the infant with a suspicion of hearing loss due to congenital cytomegalovirus (CMV) infection. As strongly recommended by a review in the literature, in all infants, referring to the 1st level screening, it is necessary to search for CMV-DNA in urine no later than 15 days after birth by PCR^{22,23}.

Finally, recording of the screening results is desirable to take place in an online database. The questionnaire highlighted that the 1st level screening operators are not informed about the results of the audiological checks following screening. The commitment of all the professionals involved could improve adherence to screening. In the organization of the screening, it is crucial to know the rate of “referred” infants who complete the diagnostic process; it must be over 90%¹⁶.

The Region should implement a data management and traceability system to monitor the quality of screening, to measure outcomes and to report on the adequacy of the service.

Conclusions

The study addresses a very important critical issue, especially in a context in which the mandatory application of UNHS is recent and the data published by the regional reference organisation are scarce and partial. Even in the absence of a shared protocol, the UNHS in Sicily is carried out, and the direct data collection has made it possible to verify the screening effectiveness at each birth point and, above all, the possibility of making known some data absent in the literature. However, it is evident that too many children are lost between not passing the initial screening and rescreening and between rescreening and diagnosis.

The use of both ABR and TEOA tests in the audiological screening decreases the number of newborns sent for audiological evaluation with a notable reduction of costs²⁴. Lastly, there is a need to establish a shared project, a regional network with a reference element and coordination of information, monitoring and quality controls of regional data.

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