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.
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 follow.\(^1\)\(^-\)\(^3\)\.

Significant bilateral hearing impairment, if undetected, can cause profound effects on speech, language and cognitive development, and can thus hamper emotional and social well-being.\(^4\)\(^-\)\(^6\).

In Italy and Western countries, it is estimated between 0.5 and 2 cases per 1,000 live births have profound hearing loss.\(^5\)\(^-\)\(^7\) Nevertheless, in some pediatric 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 (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 loss.\(^17\)

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 1-2-3.\(^18\)

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 DASOE.\(^19\) 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 bi-laterally “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 system. 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 dB), 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%. It should be noted that, in the year 2018, the screening cover-
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             |
| Acreale      | 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 Grettin  | 568   | 568   | 100.0%     | 6     | 1.1%    | ND             |
| CDC Falcidin| 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          |
| Castelvetran | 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-Annas 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 | Bilateral Hearing loss |
|----------|---------|------------|-----------------|-----------------|-----------------|-----------------|
|          | 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  | 15 - A.O.U. Policlinico | 6 | 11 | 3 | 2 | 1 |
|          | 18 - ARNAS Civico | 7 | 17 | 2 | 2 |
|          | 21 - Bucchieri/ReFerla | 6 | 21 | 6 | 2 | 2 |
|          | 9 - Ingrassia | 4 | 9 | 3 | ND |
|          | 31 - Cervello | 8 | 16 | 6 | 2 | 4 |
|          | 6 - CdC Zancia | 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 |
|          | 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 - Castelvetrano | 1 | 1 | 1 | 1 |
|          | 1 - Osp di Trapani | 0 | 1 | 0 | 1 |
|          | 1 - Ospedale di Erna | 1 | 1 | 1 | 1 |
| Termini Im. | 5 - Cimino Term. Im. | 5 | 5 | 0 | 1 |
|          | 9 - Ingrassia Palermo | 9 | 12 | 3 | 2 |
|          | 1 - Civico di Corleone | 1 | 1 |
|          | 2 - Civico di Partinico | 2 | 2 |
| Catania  | 267 - Policlinico | 1 | 1 | 1 | 1 |
| A.O. Policlinico | 210 - NICU Policlinico | 5 | 3 | 1 | 1 |
|          | 170 - S. Bambino | 3 | 3 | 3 | 1 | 1 |
|          | 11 - NICU S. Bambino | 0 | 11 | 0 | 1 |
|          | 2 - Garibaldi Catania | 2 | 2 | 1 | 1 |
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).

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

| ENT Unit                  | 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           |                          |            |
|                           | 0 - ASP Caltagirone              | 0           | 0         | 0           |                          |            |
|                           | 1 - CdiC Grettar                 | 1           | 1         | 0           |                          |            |
|                           | 0 - CdiC 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           |                                  |             |           |             |                          |            |
| Messina                   |                                  |             |           |             |                          |            |
| A.O.U. Policlinico        | 125 - Policlinico                | 7           | 7         | 3           | 2                        | 1          |
|                           | 91 - NCIU Policlinico            | 10          | 10        | 6           | 3                        | 2          | 1       |
| Patti                     | 1 - Barone di Patti              | 1           | 1         | 1           | 1                        |            |
|                           | 1 - Sant’Agata Milletto          | 1           | 1         | 1           | ND                       |            |
| Taormina                  | 2 - Osp. di Taormina             | 2           | 2         | 2           | 1                        |            |
| Ragusa                    | 1 - Civile di Ragusa             | 1           | 1         | 1           | 1                        |            |
| A.O. Civile               | 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                   | 3 - Osp. di Trapani              | 3           | 3         | 1           |                          |            |
| A.O. Sant’Antonio Abate   | 1 - Osp. di Marsala              | 1           | 1         | 0           |                          |            |
|                           | 10 - Castelvetrano               | 10          | 6         | 3           | 2                        |            |
|                           | 7 - Mazara del Vallo             | 7           | 5         | 0           |                          |            |
|                           | 2 - CdiC Sant’Anna Erice         | 2           | 2         | 1           |                          |            |
| Siracusa                  | 0 - Osp. di Siracusa             | 0           | 1         | 1           | 1                        | 19         | 10      | 9       |    |
| TOTAL                     |                                  |              |           |             | 13                       | 13         | 1       | 1       | 19 | 10 | 9 |

**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.
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 NICU. 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 positives.

The percentage of “referred” newborns to screening and concluding the diagnostic procedure was 81.0%, below the benchmark of 90%. 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 under-line 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 loss. 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 JCIH 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 population. 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 diagnosis.

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.

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%

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 costs. 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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References

1. Mehl AL, Thomson V. Newborn hearing screening: the great omission. Pediatrics 1998;101(1):E4. https://doi.org/10.1542/peds.101.1.e4
2. Erenberg A, Levenson J, Sia C, et al. Newborn and infant hearing loss: detection and intervention. American Academy of Pediatrics. Task Force on Newborn and Infant Hearing. 1998-1999. Pediatrics 1999;103:527-530. https://doi.org/10.1542/peds.103.2.527
3. Bess FH, Tharpe AM. Case history data on unilaterally hearing-impaired children. Ear Hear 1986;7:14-19. https://doi.org/10.1097/00003446-198602000-00004
4. Neumann K, Chadha S, Tavartkiladze G, et al. Newborn and infant hearing screening facing globally growing numbers of people suffering from disabling hearing loss. Int J Neonatal Screen 2019;5:7. https://doi.org/10.3390/ijns5010007
5. Mehl AL, Thomson V. The Colorado newborn hearing screening project, 1992-1999: on the threshold of effective population-based universal newborn hearing screening. Pediatrics 2002;109:E7. https://doi.org/10.1542/peds.109.1.e7
6. Morton CC, Nance WE. Newborn hearing screening - a silent revolution. N Engl J Med 2006;354:2151-2164. https://doi.org/10.1056/NEJMra050700
7. Bubbico L, Rosano A, Spagnolo A. Prevalence of prelingual deafness in Italy. Acta Otorhinolaryngol Ital 2007;27:17-21.
8. Ricci G, Molini E, Giommetti G, et al. Screening Audiologico e intervento precoce in Umbria: un progetto “A tutto tondo”. In: Orzan E, editor. Dalla diagnosi all’autonomia comunicativa - XXV Relazione Ufficiale SIOP 2019. Bagheria-Palermo: Plenuma Editore; 2019. pp 16-20.
9. Martini A, Marchisio P, Bubbico L, et al. Permanent childhood hearing impairment: universal newborn hearing screening, PCHI management. Minerva Pediatr 2013;65:231-250.
10. Yoshinaga-Itano C, Sedey AL, Coultzer DK, et al. Language of early- and later-identified children with hearing loss. Pediatrics 1998;102:1161-1171. https://doi.org/10.1542/peds.102.5.1161
11. Moeller MP. Early intervention and language development in children who are deaf and hard of hearing. Pediatrics 2000;106:E43. https://doi.org/10.1542/peds.106.3.e43
12. Ferlito S, Cocuzza S, Grillo C, et al. Complications and sequelae following tympanostomy tube placement in children with eustachian otitis media: single center experience and review of literature. Acta Medica Mediterranea 2020;36:1905-1912. https://doi.org/10.19193/0393-6384_2020_3_298
13. Berrettini S, Ghirri P, Lazzerini F, et al. Newborn hearing screening protocol in tuscany region. Ital J Pediatr 2017;43:82. https://doi.org/10.1186/s13052-017-0397-1
14. National Institute of Health Consensus Development Conference. Early identification of hearing impairment in infants and young children. NIH Consensus Statement 1993 Mar 1-3;11:1-24.
15. European Consensus Statement on Neonatal Hearing Screening. Acta Paediatri 2007;88:107-108. https://doi.org/10.1111/j.1651-2227.1999.tb01282.x
16. Joint Committee on Infant Hearing. JCIH year 2000 position statement: Principles and guidelines for early hearing detection and intervention programs. Ann J Audiol 2000;9:9-29.
17. Davis A, Bambard J, Wilson I, et al. A critical review of the role of neonatal hearing screening in the detection of congenital hearing impairment. Health Technol Assess 1997;1:i-iv, 1-176.
18. Joint Committee on Infant Hearing, Year 2019 position statement: Principles and guidelines for early hearing detection and intervention programs. J Early Hear Detect Interv 2019;4:1-44.
19. Assessorato della Salute. DASOE. Implementazione dello screening udittivo neonatale anno 2016. Regione Siciliana; Palermo, 2016.
20. Butcher E, Dezauteau C, Cortina-Borja M, et al. Prevalence of permanent childhood hearing loss detected at the universal newborn hearing screen: systematic review and meta-analysis. PLoS One 2019;14:e0219600. https://doi.org/10.1371/journal.pone.0219600
21. Frezza S, Catenazzi P, Gallus R, et al. Hearing loss in very preterm infants: should we wait or treat? Acta Otorhinolaryngol Ital 2019;39:257-262. https://doi.org/10.14639/0393-100X-2116
22. Haller T, Shoup A, Park AH. Should targeted screening for congenital cytomegalovirus infection be implemented? Int J Pediatr Otorhinolaryngol 2020;134:110055. https://doi.org/10.1016/j.ijporl.2020.110055
23. Palma S, Roversi MF, Bettini M, et al. Hearing loss in children with congenital cytomegalovirus infection: an 11-year retrospective study based on laboratory database of a tertiary paediatric hospital. Acta Otorhinolaryngol Ital 2019;39:40-45. https://doi.org/10.14639/0393-100X-2020
24. Bubbico L, Ferlito S, Antonelli G, et al. Hearing and vision screening program for newborns in Italy. Ann Ig 2021;33:433-442. https://doi.org/10.7416/ai.2020.2401