Hearing Loss in Newborns with Cleft Lip and/or Palate
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Abstract

Introduction: This study aims to review the results of hearing screens in newborns with cleft deformities. Materials and Methods: A retrospective audit of 123 newborns with cleft deformities, born between 1 April 2002 and 1 December 2008, was conducted. Data on the results of universal newborn hearing screens (UNHS) and high-risk hearing screens, age at diagnosis, severity/type of hearing loss and mode of intervention were obtained from a prospectively maintained hearing database. Results: Thirty-one of 123 newborns (25.2%) failed the first automated auditory brainstem response (AABR). Seventy percent of infants (56 out of 80) who passed the UNHS failed the high-risk hearing screens which was conducted at 3 to 6 months of age. Otolaryngology referral rate was 67.5% (83/123); 90.3% of 31 newborns who failed the first AABR eventually required otolaryngology referrals. Incidence of hearing loss was 24.4% (30/123; 25 conductive, 2 mixed and 3 sensorineural), significantly higher than the hospital incidence of 0.3% (OR: 124.9, 95% CI, 81.1 to 192.4, P <0.01). In terms of severity, 8 were mild, 15 moderate, 5 severe, 2 profound. Eighteen out of 30 infants (60%) were detected from the high-risk hearing screens after passing the first AABR. Conclusion: These newborns had a higher risk of failing the UNHS and high-risk hearing screen. There was a higher incidence of hearing loss which was mainly conductive. Failure of the first AABR was an accurate predictor of an eventual otolaryngology referral, suggesting that a second AABR may be unnecessary. High-risk hearing screens helped to identify hearing loss which might have been missed out early on in life or which might have evolved later in infancy.

Key words: Malformation, Screening

Introduction

Cleft lip and/or palate (CL/P) is one of the commonest congenital malformations.1-3 A recent update collating data from international organisations for the years 2002 to 2006 reported an overall international rate of 7.94 in 10,000 live births with the highest prevalence of 10.2 occurring in the Asian continent.2 In Singapore, the incidence of cleft cases was reported to be 1.87 to 2.07 per 1000 live births.5,6

Hearing impairment has been reported in 20% to 60% of affected infants.7-12 In a child with cleft palate, there is poor tensor veli palatini function with velopharyngeal insufficiency and increased Eustachian tube compliance. This is postulated to result in persistent fluid collection in the middle ear, predisposing the young child to otitis media with effusion (OME) and thus, conductive hearing loss.13-17 The association of OME with CL/P has been clearly demonstrated in many studies across different racial groups.17-20 A previous study in Singapore21 reported an incidence of middle ear disease to be 23% in Chinese patients with cleft palate, whereas a recent study in China22 reported a higher incidence of 64% amongst 42 Chinese infants with non-syndromic CL/P and a consequently higher incidence of hearing loss of 86%. This should be given due concern

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as studies have shown that hearing loss due to persistent middle ear effusions can have long-term effects on speech, language, and cognitive development. In view of this, it is vital to detect hearing loss early and to institute timely intervention if it is present. Although the Joint Committee for Infant Hearing (JCIH) from the American Academy of Pediatrics has recommended that these high-risk infants should be referred for audiological assessment at least once by 24 to 30 months of age, an earlier high-risk hearing screen may be advantageous for them.

Universal newborn hearing screening (UNHS) is currently practised in many countries worldwide. This has led to early detection of hearing loss, leading to early intervention which in turn, facilitates improvement of speech and language outcomes. The UNHS programme in KK Women’s and Children’s Hospital (KKH), which was implemented in 2002, uses a 2-step automated auditory brainstem response (AABR) protocol (Fig. 1). The AABR measurements are obtained non-invasively via surface electrodes that record neural activity generated in the cochlea, auditory nerve, and brainstem in response to acoustic stimuli delivered. Results of the AABR reflect the status of the peripheral auditory system, the eighth cranial nerve, and the brainstem auditory pathway. Hence, it has the advantage of detecting peripheral (conductive and sensory) hearing loss as well as neural auditory disorders.

The first screen is typically performed before hospital discharge for all newborns on day 1 of life or after 34 weeks of gestation in premature infants. A “refer” result is conferred on patients who do not pass the AABR. They then undergo the second screen at 3 to 6 weeks of life at the outpatient setting. The high-risk hearing screen is performed at 3 to 6 months of life for those who have passed the UNHS but have been identified with high-risk for hearing impairment. The high-risk hearing screen is performed by the audiologist at the otolaryngology centre with the otoacoustic emission (OAE) test. Selected risk factors for hearing impairment include: craniofacial anomalies, positive family history of hearing impairment, intrauterine infections, neonatal intensive care unit stay with use of ototoxic drugs or mechanical ventilation etc. Infants who do not pass the high-risk hearing screen or UNHS are referred to the otolaryngologist for further evaluation. This practice recognises the fact that hearing impairment can occur later in life and will be missed by the UNHS. The UNHS office staff track the progress of the infant and record the outcome of audiological assessment.

Having implemented UNHS and high-risk hearing screen in our hospital since 2002, this paper is the result of a systematic audit performed on hearing screening in high-risk infants to identify service gaps and to streamline the screening process. It reviews the results of the newborn and high-risk hearing screens in infants with CL/P born in KKH. There is little current literature on the outcome of the UNHS and early postnatal hearing screening in these infants.

Materials and Methods
The results of UNHS and high-risk hearing screen performed for newborns delivered in KKH, including newborns with CL/P, had been prospectively downloaded to a computerised database (HITRACK). Risk factors for hearing loss, the results of high-risk hearing screens, referrals to the otolaryngology department, audiological investigations and subsequent hearing outcomes had been entered manually by the UNHS staff. Hearing loss was confirmed by evoked response audiometry (ERA) with steady state evoked potential (SSEP) done in the outpatient clinic under conscious sedation or under the same general anaesthesia setting as the cleft repair. The following data on newborns with CL/P born between 1 April 2002 and 31 December 2008 were obtained from the HITRACK database and audited: results of UNHS and high-risk hearing screen, age at diagnosis, severity and type of hearing loss and the mode of intervention. Comparisons were made with the KKH cohort data involving 87,882 newborn infants. Interventions include: 1) medical treatment (oral or aural antibiotics and treatment for cerumen), 2) surgical treatment (myringotomy and tympanostomy), 3) amplification and auditory verbal therapy, and 4) speech and language intervention. Missing information was obtained from reviews of case records. The study was approved by the institutional review board.
Results

There were 123 newborns with CL/P during the audit period from 1 April 2002 to 31 December 2008, from a total of 87,882 births (0.14%). This incidence of 1.4 per 1000 births is comparable to the past reported incidence of cleft cases in Singapore.5,6

Results of Universal Newborn Hearing Screening (UNHS)

A total of 96.7% underwent hearing screen prior to hospital discharge. Figure 2 shows the outcomes of the UNHS in these patients. Out of the 4 newborns who missed the inpatient AABR, one had been discharged shortly after birth and did not return for the outpatient AABR. At 1.5 months of life, she was re-admitted for sepsis and right otorrhea, and was referred to otolaryngology, where a unilateral moderate conductive hearing loss was detected. Two of these 4 newborns were transferred to the Paediatric Intensive Care Unit (PICU) after cardiac surgery for coarctation repair and abdominal surgery for necrotising enterocolitis. They were not screened before discharge from the paediatric department but both returned for outpatient screening.

Results of High-Risk Hearing Screening

A total of 94 cleft infants were eligible for high-risk hearing screen (Fig. 3). These included the 88 infants who passed the inpatient AABR, 3 who passed after a first “refer” result, 1 who missed the repeat screen after a first “refer” result, and 2 who passed as outpatients after missing the inpatient screen. Despite sending reminder letters following failure to attend the high-risk hearing screen visit, 14 (14.9%) did not attend. Out of the 80 who underwent high-risk hearing screen, 56 (70%) had poor emissions and were referred to otolaryngology.

Referrals to Otolaryngology

In total, 83 were referred to the otolaryngology department for further evaluation, giving rise to an otolaryngology referral rate of 67.5% (Fig. 4). Out of these 83 infants, 30 were found to have hearing loss: one was directly referred from birth, 8 were referred from the UNHS, 3 were directly referred after the first AABR, and 18 were referred from the high-risk hearing screen (Fig. 5).

Hearing Loss

Thirty (24.4%) of the 123 infants with cleft deformities were diagnosed to have hearing loss at a median age of 39.9 weeks of life (range, 7.9 to 126.9 weeks). The age of diagnosis in each individual referral category is shown in Figure 5. The number needed to screen to pick up one case of hearing loss is 5. In the 30 cases of hearing loss, 23 (76.6%) were conductive, 2 (6.7%) were permanent conductive hearing loss, 2 (6.7%) were mixed and 3 (10%) were sensorineural; 12 had unilateral involvement (6 of mild severity, 3 of moderate severity, 2 of severe severity, 1 of profound severity); 18 had bilateral involvement (2 of mild severity, 12 of moderate severity, 3 of severe severity, 1 of
profound severity). Twenty-seven (90%) of the 30 affected infants underwent intervention; 22 underwent both surgical and medical treatment, 3 underwent medical therapy alone, 1 underwent surgical intervention and audiovisual therapy (AVT), and 1 underwent speech and language intervention.

Table 1 shows that much larger percentages of infants with CL/P do not pass the newborn AABR either at birth or at 3 to 6 weeks, and are referred to otolaryngology for audiological assessment and have hearing loss compared to the general population of non-cleft newborns born in KKH during the same period of time (unpublished data from HITRACK database of UNHS in KKH, 1 April 2002 to 31 December 2008).

Table 1. Comparison of Screening Results in Infants with Cleft Lip and/or Palate with the General KKH Non-Cleft Cohort

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<table>
<thead>
<tr>
<th></th>
<th>KKH Cohort Excluding Cleft Infants (n = 87,759)</th>
<th>Infants With Cleft Lip and/or Palate (n = 123)</th>
<th>Odds Ratio (95% CI)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Did not pass inpatient AABR</td>
<td>3.3% (2857/87,759)</td>
<td>25.2% (31/123)</td>
<td>10.0 (6.6 – 15.1)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Did not pass outpatient AABR*</td>
<td>13.8% (407/2940†)</td>
<td>72.4% (21/29‡)</td>
<td>16.3 (7.2 – 37.1)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Referral to otolaryngology</td>
<td>0.5% (395/87,759)</td>
<td>67.5% (83/123)</td>
<td>458.9 (310.7 – 677.8)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Diagnosed with hearing loss</td>
<td>0.3% (226/87,759)</td>
<td>24.4% (30/123)</td>
<td>124.9 (81.1 – 192.4)</td>
<td>&lt;0.01</td>
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**Confounders**

Univariate analyses were carried out with the other known risk factors such as positive family history of hearing loss, low birth weight less than 1.5 kg, meningitis, use of ototoxic drugs, associated syndromes, hyperbilirubinaemia, intrauterine infections, craniofacial anomalies, use of mechanical ventilation, asphyxia, gestation less than 32 weeks and neonatal intensive care unit stay for more than 48 hours. They showed no significant confounding effect on the outcome of hearing screening ($P >0.05$).

**Discussion**

**Diagnosis of Hearing Loss**

More than 95% of affected infants were screened before hospital discharge, as recommended by JCIH. The percentage of infants with CL/P who did not pass the UNHS is comparable with other studies. There are several possible reasons that could explain this. First, the confirmatory audiometric test done in the otolaryngology clinic or in the operating theatre with the cleft surgery may have been performed after some medical treatment had been instituted by the otolaryngologist as deemed appropriate during the initial reviews. Second, it could also be due to the fact that we had an unfortunately high default rate (14.9% at the high-risk hearing screen and 25.3% at the otolaryngology evaluation).
**High-Risk Hearing Screen**

The majority of infants with hearing loss in this audit were identified by the high-risk hearing screen. This further reinforces the importance of this high-risk hearing screen at 3 to 6 months of age for these newborns as hearing impairment may evolve later in their infancy and thus may not be apparent during the first AABR. JCIH 2007 recommended a referral for audiological assessment at least once by 24 to 30 months of age. Our current protocol reassesses the child at risk at an earlier age with the high-risk hearing screen. It is effective for the detection of late-onset hearing loss even when done at an age earlier than the recommended age stipulated by JCIH. It allowed intervention before the age of spoken language.

**Age of Diagnosis**

The age of diagnosis was at a median age of 39.9 weeks of life (range, 7.9 to 126.9 weeks). It is preferable to diagnose hearing impairment under 1 year of age, before the onset of speech. This is crucial for early intervention and the possibility of normal speech.

The cases which were diagnosed (i.e. underwent the confirmatory audiometric test) beyond the age of 1 year were usually already identified earlier through a failed hearing screen and seen by the otolaryngologist before 6 months of age for initial assessment and perhaps a trial of treatment. Only 1 infant defaulted multiple otolaryngology visits due to medical illnesses and was first seen by the otolaryngologist at 8 months of age. At times, when it is difficult or inappropriate to sedate the young infant for the outpatient audiometric test, the otolaryngologist may defer the confirmatory audiometric test to the date of the cleft palate surgery so that it can be done under the same general anesthetic setting.

The age of diagnosis is only slightly earlier for the group which was referred from the UNHS compared to that referred from the later high-risk hearing screen (Fig. 5). This should not be misinterpreted to mean that there is no significant advantage in early screening for hearing loss. An earlier otolaryngology referral prompted by a failure to pass the UNHS widens the window period for intervention whilst awaiting the confirmatory audiometric test. As mentioned above, the confirmatory audiometric test may only be done at the otolaryngology clinic after some initial assessment and trial of medical treatment, and may at times be deferred to coincide with the date of the cleft surgery.

As per JCIH 2007 guidelines for congenital permanent bilateral or unilateral hearing loss, permanent conductive hearing loss and neural loss, it would have been preferable for these infants to have undergone comprehensive audiological evaluation at no later than 3 months of age and receive appropriate intervention at no later than 6 months of age if they were confirmed to have hearing loss. In our audit, there were 2 infants diagnosed with permanent conductive hearing loss and 3 infants with sensorineural hearing loss, with the age of diagnosis ranging from 3 to 22 months (median age of 8 months). Two of these 5 infants were diagnosed before the age of 3 months. The remaining 3 infants who were diagnosed after 3 months of age were all reviewed by the otolaryngologist before 3 months of age — one was an ex-premature 26-weeker neonatal intensive care unit graduate who had a stormy neonatal period and required surgical treatment for bilateral OME; another had Goldenhar syndrome with left microtia and normal hearing on the right ear; the last infant had recurrent OME requiring multiple surgical interventions.

**Nature of the Hearing Loss**

Similar to other studies, hearing loss was mainly conductive in this study. This is not surprising as OME has been found to be almost universally present in infants with cleft palate. Conductive hearing loss due to OME is potentially treatable by intervention with antibiotics or grommet insertion. Hence, it is beneficial and imperative to initiate otolaryngology referral upon failure of the hearing screens done in the first 6 months of life.

**Default Rate**

Fourteen out of 94 infants (14.9%) defaulted the high-risk hearing screen. Out of the 80 infants who attended the high-risk hearing screen, 56 (70%) did not pass. Using this same percentage, it is postulated that at least 9 infants out of the 14 defaulters would have not passed the high-risk hearing screen. Twenty-one infants out of 83 (25.3%) did not attend the otolaryngology referral. Out of the 62 infants who attend the otolaryngology referral, 30 of them (48.4%) were documented to have hearing loss. Using this same percentage, it is postulated that at least 10 of the otolaryngology defaulters and 4 of the high-risk hearing screen defaulters would have hearing loss. Assuming the worst case scenario, if all 14 high-risk hearing screen defaulters and all 21 otolaryngology defaulters had hearing loss, the proportion of infants with cleft abnormalities who have hearing loss will be increased to 52.8%.

While it is heartening to know that all of the infants did undergo at least one hearing screen, the increasing rate of defaulting at the later stages of the screening process, when the risk of hearing loss is rising, is alarming. This high default rate is similarly seen in other centres, and in infants with other risk factors for hearing impairment. It would be useful to review the characteristics of the infants who default and to explore the reasons for defaulting.
possible reason could be that parents of these infants with cleft deformities may be more focused on the surgical management and feeding issues related to the cleft deformities. Not realising the associated high incidence of hearing loss, they may overlook the need for hearing screens in early infancy. This highlights the importance of parental education. It has been shown that speech and language development is more dependent on hearing ability than the severity of cleft or surgical repair.33

**Recommendations for Improvement**

**Direct Referral for Otolaryngology Assessment After a First Failed Automated Auditory Brainstem Response (AABR)**

Out of 31 infants who did not pass the first AABR, 5 were referred to otolaryngology, 2 passed away and 1 did not attend. The majority (87.0%) of the remaining 23 had the same result on the outpatient screen and were referred to otolaryngology. All 3 infants who passed the repeat outpatient AABR subsequently failed the high-risk hearing screen and were also referred to otolaryngology. In total, 28 out of the 31 infants (90.3%) who did not pass the first AABR required an eventual otolaryngology referral. Thus, we recommend a direct referral to otolaryngology if the infant does not pass the first AABR. This will be discussed with the Department of Otolaryngology and the birth defect team of the Department of Neonatology.

**Reduction of the Default Rate**

Infants with CL/P are followed up in the neonatal birth defect clinic and in the craniofacial unit, both by a plastic surgeon and a speech and language therapist. These visits can serve as an excellent platform to reiterate advice to parents regarding the high incidence of hearing loss and recurrent otitis media. Both teams of staff can be prompted to reinforce compliance with hearing screens.

**Future Follow-Up**

Other studies have reported that the median resolution of conductive hearing loss in infants with cleft palate was 5 years9,40 and persisted in 24.8%.9 Follow-up of this study cohort would be definitive to determine if early diagnosis and intervention of hearing loss facilitated by systematic screening will lead to its earlier resolution and reduction in the percentage that persist. The results will be reviewed in 2 to 3 years after the appropriate amendments to the screening programme have been instituted.

**Conclusion**

Newborns with CL/P are at a higher risk of failing to pass both their newborn and high-risk hearing screens, thus requiring otolaryngology referrals. The incidence of hearing loss in these newborns with cleft deformities is significantly higher than the general population. Failure of the first AABR was an accurate predictor of an eventual otolaryngology referral, suggesting that a second AABR may be unnecessary. The high-risk hearing screen is beneficial for those who may have missed earlier screens, or whose hearing problems evolve later in their infancy. The high default rate should be addressed by reinforcing compliance to hearing screens and otolaryngology assessments.

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**REFERENCES**


