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Factors contributing to hearing impairment in patients with cleft lip/palate in Malaysia: A prospective study of 346 ears

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ABSTRACT

Objective: To determine the factors contributing towards hearing impairment in patients with cleft lip/palate.
Method: A prospective analysis was conducted on 173 patients (346 ears) with cleft lip and palate (CL/P) who presented to the combined cleft clinic at University Malaya Medical Centre (UMMC) over 12 months. The patients' hearing status was determined using otoacoustic emission (OAE), pure tone audiometry (PTA) and auditory brainstem response (ABR). These results were analysed against several parameters, which included age, gender, race, types of cleft pathology, impact and timing of repair surgery.
Results: The patients' age ranged from 1–26 years old. They comprised 30% with unilateral cleft lip and palate (UCLP), 28% with bilateral cleft lip and palate (BCLP), 28% with isolated cleft palate (ICP) and 14% with isolated cleft lip (ICL). Majority of the patients (68.2%) had normal otoscopic findings. Out of the 346 ears, 241 ears (70%) ears had passed the hearing tests. There was no significant relationship between patients' gender and ethnicity with their hearing status. The types of cleft pathology significantly influenced the outcome of PTA and ABR screening results ($p < 0.001$). There was no significant difference between the repaired and unrepaired cleft groups and the outcome of hearing tests. However, hearing improvement occurred when palatal repair was performed at the age of <1year old (OR = 2.37, CI 1.2 = 4.6, $p = 0.01$).
Conclusion: Majority of the cleft patients had normal hearing (70%). Hearing threshold varied significantly between the different types of cleft pathology. Surgery conferred no significant impact on the hearing outcome unless surgery was performed at the age of <1 year old.

1. Introduction

Cleft lip and palate (CL/P) are common defects that result in abnormal facial configuration, hearing and speech. These defects are due to partial or complete lack of fusion of the maxillary prominence with the medial nasal prominence, on one or both sides [1]. Incidence of cleft lip and palate has been reported to range from 1.19 to 170 per 1000 live births, with ethnic and geographic variation [2,3].

Patients with cleft lip and palate (CLP) frequently present with conductive hearing loss due to otitis media with effusion (OME) [4]. In a Malaysian study conducted by Loh and colleagues on the incidence of middle ear effusion in cleft palate patients, the incidence of middle ear effusion was high (57.6%) [5]. In a healthy child, Eustachian tube function improves with age. The critical period when the Eustachian tube function fully normalizes is at the age of 5–6 years [6].

Hearing problems in CL/P could be influenced by several factors such as the patients' age, gender, ethnicity, repair surgery and the types of cleft pathology. Hanzic et al. reported that the hearing improves as the child grows older for all cleft types. They also found that bilateral and unilateral cleft lip and palate patients were more likely to suffer from mild to moderate conductive hearing loss as compared to isolated cleft palate due to the high volume clefts [7]. However, Chu et al. reported that patients' age, gender, race and types of cleft did not significantly affect the hearing [8]. In another study, infants with cleft palate were reported to have normal middle ear function up to the age of four months [9]. Surgical closure of palatal clefts at or before the age of four months significantly lowers the incidence of hearing impairment [9].

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2. Methods

The objective of this study is to investigate the factors contributing towards hearing impairment in patients with cleft lip/palate.

We conducted this prospective observational study over a period of 12 months from February 2013 to February 2014. The total number of patients with cleft lip and palate were 173 (346 ears). The patients comprised of new cases of any age group, referrals and follow-up cases. We excluded patients lost to follow-up and those who were reviewed at multiple medical centres.

We collected data from all the patients with cleft lip and palate who attended the Combined Cleft Clinic (CCC) at the University of Malaya (UM) during the said period. We obtained standard demographic information like age, gender, ethnicity as well as medical history such as co-morbidities, history of palatal surgery and developmental milestones for speech and language. We examined the oral cavity and oropharynx to establish the type of cleft pathology and categorized them into unilateral cleft lip and palate (UCLP), bilateral cleft lip and palate (BCLP), isolated cleft lip (ICL), isolated cleft palate (ICP) and submucous cleft accordingly.

We proceeded with otoscopic examination on all patients and documented findings like perforated tympanic membrane, retracted tympanic membrane, ear discharge, tympanosclerosis and the presence of grommets. Retracted tympanic membrane appeared dull, lustreless, with a foreshortened handle of the malleus and/or a distorted cone of light.

Standard diagnostic instruments like otoacoustic emission (OAE), pure tone audiometry (PTA), auditory brainstem response (ABR) and tympanometry were utilized to evaluate all patients. Choice of diagnostic and screening tools would depend on patient's age and maturity.

Pure tone audiometry was performed for patients with a developmental age of more than four years. By using the GNO Madsen Itera II Audiometer with the patient in a sound proof booth, we recorded the threshold intensity at 500 Hz, 1000 Hz and 2000 Hz. The sound proof booth is calibrated yearly and is certified by the Department of Occupational Safety and Health (DOSH). The patient presses and releases a signal switch for the duration a presented pure tone is heard. Once a positive response is obtained, the presented tone is reduced by 10 dB and then increased by 5 dB to accurately determine the hearing threshold. Normal hearing was defined as a pure tone average of 0–25 dB at speech frequencies of 500, 1000 and 2000 Hz. These frequencies were tested as most phonemes fall within this frequency range. The pure tone average of 0–25 dB on PTA was taken as normal hearing to have it closely standardised with the OAE value of 30 dB or better, being labelled as a pass. Also, considering the wide age distribution in our study population, the adult upper limit of normal hearing threshold was used.

Tympanometry was performed on all patients using an impedance audiometer. Jergers Classification was used to define the results. Type A indicates normal tympanic membrane compliance, Type B suggests middle ear effusion (in a patient with an intact tympanic membrane) and type C is seen with negative middle ear pressure.

We performed transient evoked otoacoustic emission (TEOAE) tests for all children under the age of 4 years old. The test result was labelled as a “pass” when we detected TEOAE signal, suggesting a hearing sensitivity at 30 dBHL or better.

With the screening/automated auditory brainstem response (ABR), the Bio-logic ABaer Cub was used. Brainstem response was elicited by multiple auditory click stimuli at 35 dB. Appearance of wave V in the first 10ms was defined as a normal screening BERA in this study, which was labelled as ‘pass’.

Data entry and analysis was performed using SPSS version 20. Skewness and Kurtosis tests were utilized to determine if the age distribution among all types of cleft patients fall within the normal Gaussian distribution. Comparative statistical tests such as the Kruskal Wallis, Pearson Chi-square, Fisher's exact, Mann-Whitney and logistic regression tests were used to determine the correlation between the different parameters and the mean hearing loss. Binary logistic regression was performed to determine whether a particular type of cleft pathology would predict higher odds of failed hearing tests. We carried out Spearman correlation to examine the level of association between hearing loss and age of patients.

3. Result

The study comprised of 173 non syndromic cleft patients (346 ear samples) with the median age of 4 years old. The patients' age ranged from 1 to 26 years old. Data was analysed in terms of number of ears. The age distribution among all the cleft types differed from the normal Gaussian graph (Skewness = 1.645; Kurtosis = 2.285). Female to male ratio was 1:1.34. There was no statistical significant difference between male and female cleft patients in the outcome of hearing test (OR = 1.25, 95% CI 0.78–1.99, $p = 0.355$). None of the patients had sensorineural hearing loss.

Four major types of cleft pathology were studied in our patients. Fifty two were diagnosed with unilateral cleft lip and palate (30.1%); 49 patients with bilateral cleft lip and palate (28.3%); 48 patients with isolated cleft palate (27.7%); isolated cleft lip in 24 patients (13.9%). There was no gender difference between the unilateral and bilateral cleft lip and palate patients; however, we noted male preponderance in the group with isolated cleft palate (OR = 2.20, 95% CI 1.08–4.482, $p = 0.03$).

Otoscopic examination was performed on all the 346 ears. We reported abnormal findings in 31.8% of the ears. Common sequelae of serous otitis media noted in our patients were perforated tympanic membrane, retracted tympanic membrane, tympanosclerosis, grommet in-situ and dull tympanic membrane. Patients with cleft lip and palate were 3.75 times more likely than patients with isolated cleft lip to develop these sequelae (OR = 3.75, 95% CI 1.54–9.20, $p = 0.004$).

The study comprised three major ethnic groups, namely Malay (51.4%), Chinese (24.3%), Indian (19.7%) and others (4.6%). No significant difference was noted in the racial distribution of all four cleft types (Pearson Chi-Square, asymp. sig. 2-sided, $p = 0.48$). There was also no statistically significant difference between the three ethnic groups and the outcome of hearing tests. (Pearson Chi-Square, sig. 2-sided, $p = 0.28$).

Patients with bilateral cleft lip and palate were more likely to have failed otoacoustic emission (OAE) tests, when compared to the non-palatal cleft group, i.e. isolated cleft lip group (OR = 12.7, 95% CI 3.4–46.7, $p < 0.001$). We found a significant difference in the result of OAE between the four types of cleft pathology ($p < 0.001$).

Pure tone audiometry (PTA) was performed in 81 patients (162 ears) aged four years old and above. A total of 108 out of the 162 ears (67%) had passed the PTA test (hearing threshold less than 26 dB). We found a statistically significant difference in pure tone average at speech frequencies between the four major groups of cleft patients.

We conducted correlation test to study the degree of relationship between the patients' age and their pure tone average. When all the patients were analysed, we reported no significant linear correlation between patients' age (346 ears) and their pure tone average ($rs = 0.132$, $p = 0.095$). The patients were further divided into three different age categories of less than five years, 5–15 years and more than fifteen years of age. In the group of <5 years old,