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*Original Research*

# Etiological Factors and Audiological Evaluation of Children with Congenital Unilateral Hearing Loss

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**Abstract**

**Objectives:** The aim of the present study is to evaluate the audiological, radiological, and etiological in terms of clinical findings relating to babies and children with congenital unilateral hearing loss. **Materials and Methods:** Audiometric tests, tympanometric and acoustic reflex measurements, otoacoustic emission tests, and auditory brainstem response assessments were conducted. Twenty-nine babies and children (13 F ;16 M) diagnosed with congenital unilateral hearing loss, between the ages of 3–87 months, were included in this study. **Results:** Of these patients, 65.5% (n:19) were diagnosed with sensorineural hearing loss and 34.5% (n:10) with conductive hearing loss. Of the subjects with sensorineural hearing loss, 57.9% were diagnosed with profound hearing loss. Of the subjects with conductive hearing loss, 7 of the 10 (70%) had been diagnosed with microtia. Of the patients with conductive hearing loss, all those who had received CT scans were diagnosed with ossicular chain malformations. The most frequent risk factor for sensorineural hearing loss is intermarriage (26.3%), whereas the most frequent risk factor for conductive hearing loss is craniofacial anomalies (30%). **Conclusion:** The interdisciplinary work in otology, audiology, and radiology is essential for early diagnosis and effective treatment of congenital unilateral hearing loss cases.

***Keywords:*** *Unilateral Hearing Loss, Congenital, Audiologic Findings*

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## Introduction

Newborn hearing screening is the most effective tool to diagnose congenital hearing loss (Laury, Casey, McKay, & Germiller, 2009). Children with unilateral hearing loss (UHL) are being diagnosed at younger ages because of newborn hearing screening. Even when identified in the first months of life, children with UHL show a tendency to lag behind their normal hearing peers in functional auditory listening and in receptive and expressive language development (Fitzpatrick et al., 2019).

The diagnosis of hearing loss is usually followed by a search for an underlying etiology. The most commonly reported known etiologies of UHL include viral complications (approximately 25%), meningitis (approximately 15%), head trauma (approximately 8% to 12%), prenatal or perinatal disorders (12%) and genetic disorders. In addition, prematurity, enlarged vestibular aqueduct syndrome, sudden idiopathic hearing loss, auditory neuropathy/ dysynchrony, noise induced hearing loss, bacterial complications, and unilateral atresia or microtia are among the causes of unilateral hearing loss (Laury et al., 2009; van Wieringen, Boudewyns, Sangen, Wouters, & Desloovere, 2019). The aim of the study is to review unilateral neural hearing loss in babies and infants, to better understand its etiology, clinical and audiologic features. Additionally, it was hypothesized that there were no differences in hearing loss according to gender.

## Materials and Methods

The study protocol was approved by Hacettepe University Non-Interventional Clinical Research Ethics Committee (No: LUT 12/164 – 12, 13.02.2013). Babies and children with unilateral hearing loss who cannot pass the national newborn hearing screening in one ear or who had physical findings associated with unilateral hearing loss were included in this study. Data were collected on age, gender, affected ear, severity of hearing loss and risk factors for hearing loss (e.g. hyperbilirubinemia, prematurity, drug use, CMV). Audiological test battery was included the tympanometry, acoustic reflex measurements, otoacoustic emission measurements and Auditory Brainstem Response (ABR) measurements. The degree of hearing loss was classified according to the American Speech, Language, Hearing Association (ASHA) (Clark, 1981). Magnetic Resonance Imaging (MRI) and Computerized Tomography (CT) scan were performed and interpreted by radiologist.

The following criterion was applied to determine the participants included: (1)

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diagnosed with unilateral hearing loss, (2) granting permission from legal guardian to participate to the study.

**Tympanometric Evaluation**

Tympanometry was performed by GSI Tympstar Version 2 Middle Ear Analyzer. The assessment was made automatically, occluding the external auditory canal tympanic membrane. Changing pressure was applied from +200 daPa (decapascals) to -400 daPa. Applications were performed with the speed of 200/600 daPa/sec. In our study, 226 and 1000 Hz probetones were used, depending on the patient's age. The tympanometry measurements analyzed were the peak pressure, equivalent ear canal volume and the static compliance.

**Acoustic Reflex Measurements**

Acoustic reflexes were measured by GSI Tympstar. The appropriate probetone for the child's age (226 Hz or 1000 Hz) was chosen to elicit the acoustic reflexes. The reflex threshold was searched manually by sending sound to the ipsilateral ear at frequencies of 500, 1000, and 2000 Hz and to the contralateral ear at frequencies of 500, 1000, 2000, and 4000 Hz. Three repetitive responses at the same intensity level were accepted as the acoustic reflex threshold.

**Otoacoustic Emission (OAE) Measurement**

Transient otoacoustic emissions (TEOAE) and distortion product otoacoustic emissions (DPOAE) were conducted separately to each ear with an ILO 292 Echo Port USB II device with ILO V6 Clinical OAE Software Otodynamics, (London, UK). A DPOAE was accepted present at a particular frequency region when the signal-to-noise ratio was ≥ 6 dB. The TEOAEs were accepted ''pass'' when the signal to noise ratio was ≥ 6 dB and the confidence ratio was ≥ 80%.

**Diagnostic ABR Measurement**

Diagnostic ABR tests were applied with an AUDERA and/or VIVOSONIC tester during the infant's/child's natural sleep. Cup (AUDERA) or patient-mounted (VIVOSONIC) electrodes were used. ABR stimuli in air conduction measurements were presented separately in both ears with ER 3 headphones. For cases in which the stimulus intensity was more than 70 dB, the contralateral ear was masked with 35–50 dB of noise. Click stimuli was used to evaluate hearing sensitivity. The stimulus repetition rate was determined as 11.1/sec. The tests were performed by rarefaction stimulus polarity. Both measurements were performed with a repetition rate of 2000. A 3 kHz low-pass filter was used to create the stimulus and was set to be a 30 Hz

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high-pass filter. A 15-millisecond (ms) analysis window was used to record the sweeps. Placing B71 headphones on the mastoid bone of the ear being tested made bone conduction ABR (AUDERA) stimulus measurements. Masking noise was applied in the contralateral ear. Stimulus rate was set to 9.1. Alternating stimulus polarity was used. Double traces were collected at intensities of 40 dBnHL, 30 dBnHL, and 20 dBnHL. If there was a risk of waking the child during the session, the test was started with 30 dBnHL stimuli, and if a wave was detected, 40 dBnHL stimuli were not applied. The high- frequency filter was at 30 Hz, and the low-frequency filter was at 3 kHz to form the waves. The type of hearing loss was predicted by analyzing bone conduction and air-conduction ABR results together and/or analyzing the latencies, amplitudes and morphology of air-conduction ABR waves.

**Magnetic Resonance Imaging**

MRIs were performed with a 1.5 Tesla scanner with a standard head-coil (Symphony, Siemens, Erlangen, Germany). In the steady state, standard temporal bone protocol, including transverse T1-weighted imaging, transverse T2-weighted imaging and axial and sagittal oblique three-dimensional (3D) constructor was applied.

**Statistical Analyses**

The study design was single group descriptive study. Data were analyzed using SPSS 21.0 (Statistical Package for Social Sciences) program. Quantitative data were described as mean ± standard deviation (X ± SD), and qualitative data were described in percentage values. The normality of data was evaluated with visual (histogram and stem-leaf plots) and analytic (Kolmogorov–Smirnov/Shapiro–Wilk tests) methods. Comparison of the side and type of hearing loss in girls and boys with unilateral hearing loss was analyzed by chi-square test.

## Results

Twenty-nine children (13 girl and 16 boy) age-ranged from 3 to 87 months with unilateral hearing loss were included in this study. The population sample characteristics are given in Table 1. None of the patients in this study were diagnosed with congenital mixed hearing loss.

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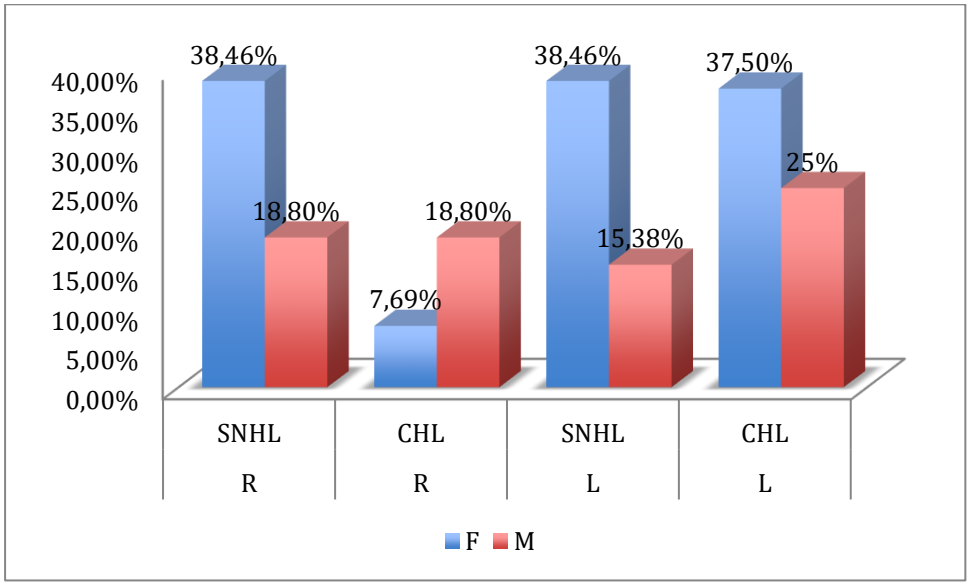
**Table 1:** The population sample characteristics.

| | Number | % |
|---|---|---|
| **Girls** | 13 | 44.8 |
| **Boys** | 16 | 55.2 |
| **Right UHL** | 16 | 55.2 |
| **Left UHL** | 13 | 44.8 |
| **SNHL** | 19 | 65.5 |
| **CHL** | 10 | 34.5 |

UHL:Unilateral hearing loss, SNHL:Sensorineural hearing loss, CHL:Conductive hearing loss

Gender distribution of laterality and type of the hearing loss, are analyzed in Figure 1. The number of boys with unilateral hearing loss was observed to be slightly higher than girls (M:F ; 53.3%:46.7%). This difference is not statistically significant.




SNHL: Sensorineural hearing loss, CHL: Conductive hearing loss

**Figure 1:** Gender distribution of laterality and type of hearing loss.

Severity of sensorineural hearing loss is given in Table 2. Six of ten with conductive hearing loss were diagnosed microtia. Therefore, it was unable to diagnose the severity of hearing loss in these patients. Severity of conductive hearing loss is given in Table 3.

Risk factors in sensorineural hearing loss and conductive hearing loss vary. Therefore, risk factors in our study for sensorineural and conductive hearing loss and imaging results are presented in Table 4. CT results are available for 17 of 29 patients (58.6%). Six (20.7%) patients were not at a suitable age for CT. The parents of 6 (20.7%) patients did not consent to CT imaging. CT imaging of 10 (34.5%) patients

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with SNHL and 7 (24.1%) patients with CHL were available. Risk factors and imaging results are shown in Table 4. Imaging finding of the patient with CHL was given in Figure 2.

**Table 2:** Severity of sensorineural hearing loss.

| SNHL | Number | % |
|---|---|---|
| **Profound** | 12 | 63.2 |
| **Severe** | 4 | 21 |
| **Moderate** | 1 | 5.3 |
| **Mild** | 2 | 10.5 |
| **Total** | 19 | 100 |

**Table 3:** Severity of conductive hearing loss.

| CHL | Number | % |
|---|---|---|
| **Profound** | 1 | 10 |
| **Moderate** | 2 | 20 |
| **Mild** | 1 | 10 |
| **Total** | 4 | 40 |

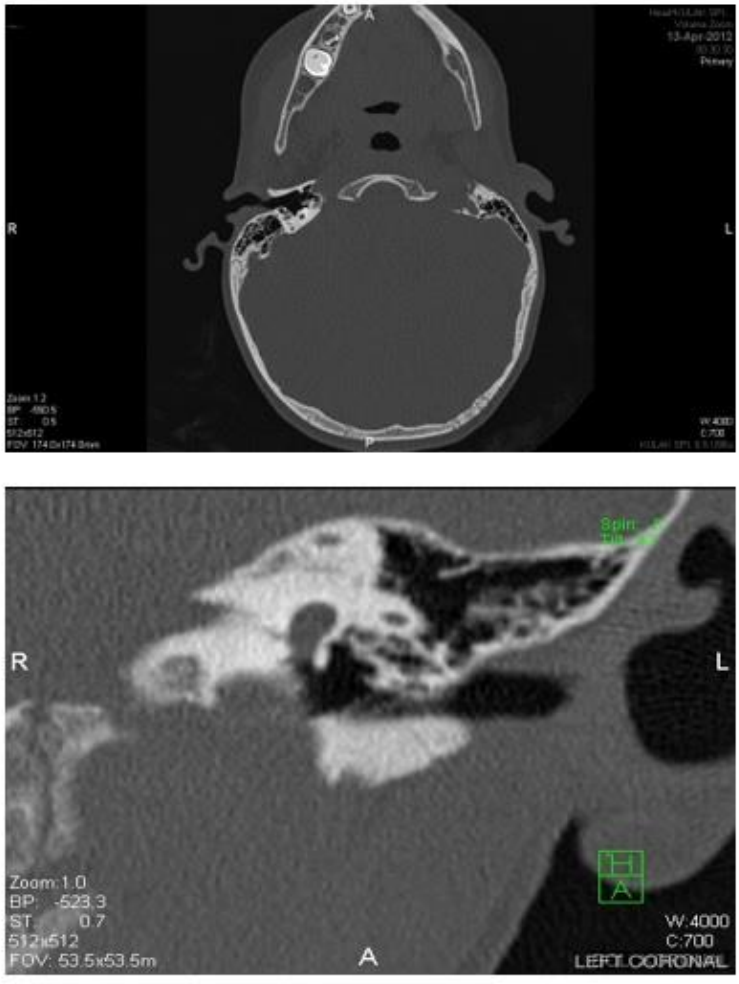


**Figure 2:** Imaging findings of patient (No: 10) with congenital unilateral CHL.

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**Table 4:** Risk factors and imaging results.

| Subject | Gender | Affected Ear | HL Type | HL Degree | Syndroms/ Risk Factors | Imaging |
|---|---|---|---|---|---|---|
| **1** | M | R | S/N | Moderate | Penicilin use in pregnancy | CT Scan: Delay of otic capsule ossification<br>MRI: Cochlear nerve aplasia in left ear |
| **2** | M | L | S/N | Profound | - | CT Scan: -<br>MRI: - |
| **3** | F | L | S/N | Severe | Anbiotics use in pregnancy | CT Scan: Normal<br>MRI: - |
| **4** | F | L | S/N | Profound | Family history of HL | CT Scan : Normal<br>MRI: Narrowness of internal acoustic canal, cochlear nerve aplasia in left ear |
| **5** | M | R | S/N | Profound | - | CT Scan: Normal<br>MRI: - |
| **6** | M | R | S/N | Profound | Family history of HL and antibiotics use in newborn | CT Scan: Normal<br>MRI: - |
| **7** | F | L | S/N | Severe | Family history of HL + Urinary tract infections at pregnancy | CT Scan: Normal<br>MRI: - |
| **8** | F | R | S/N | Profound | Family history of HL + Blood incompatibility + CMV at pregnancy | CT Scan: Bilateral normal inner ear structures, adjacent to the temporal bone in the antero superus at the beginning of bilateral malleus |
| **9** | M | R | S/N | Severe | - | CT Scan: Common cavity<br>MRI: - |
| **10** | M | R | S/N | Profound | Prematurity | CT Scan: Normal<br>MRI:- |

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|---|---|---|---|---|---|---|
| **11** | F | L | S/N | Severe | Family history of HL | CT Scan: Aplasia of left cochlear aperture, duplication of left facial canal labyrinth segment<br>MRI: - |
| **12** | F | R | S/N | Mild | Drug use in pregnancy + antibiotic use in newborn | CT Scan: -<br>MRI: - |
| **13** | M | R | S/N | Profound | - | CT Scan: -<br>MRI: - |
| **14** | F | R | S/N | Profound | - | CT Scan: -<br>MRI: - |
| **15** | F | L | S/N | Profound | Phototherapy | CT Scan: -<br>MRI: - |
| **16** | M | R | S/N | Profound | - | CT Scan: -<br>MRI: - |
| **17** | F | R | S/N | Mild | Family history of HL + Drug use in pregnancy + Phototherapy and antibiotics use in newborn | CT Scan: -<br>MRI: - |
| **18** | F | L | S/N | Profound | - | CT Scan: -<br>MRI: - |
| **19** | M | L | S/N | Profound | Diabetes | CT Scan: -<br>MRI: Compression to 8$^{th}$ cranial nerve |
| **20** | F | R | CHL | Moderate | Pregnancy of multiples + Prematurity + NICU | CT Scan: Possible congenital stapes fixation<br>MRI: - |
| **21** | M | R | CHL | Microtia | - | CT Scan: -<br>MRI: - |
| **22** | M | R | CHL | Microtia | Urinary tract infections + Drug use in pregnancy | CT Scan: -<br>MRI: - |

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| | | | | | | |
|---|---|---|---|---|---|---|
| **23** | M | L | CHL | Profound | Blood incompatibility + Antibiotics use in newborn | CT Scan: Ossicle chain malformation<br>MRI: - |
| **24** | M | L | CHL | Mild | Family history of HL | CT Scan: Outer ear canal stenosis, possible malleus fixation<br>MRI: - |
| **25** | M | R | CHL | Moderate | Intermarriage + Antibiotics use in pregnancy + Urinary tract infections + Hyperbilirubinemia | CT Scan: Ossicle chain malformation<br>MRI: - |
| **26** | M | L | CHL | Microtia | Family History of HL + Diabetes | CT Scan: Outer ear canal atresia, fusion of incus to temporal posterior bone<br>MRI: - |
| **27** | F | R | CHL | Microtia | Intermarriage + Antibiotics use in pregnancy + Urinary tract infections + | CT Scan: Outer ear canal atresia, no ossicular chain, severe hypoplasia of middle ear<br>MRI: - |
| **28** | M | R | CHL | Microtia | Antibiotics use in pregnancy + Urinary tract infections | CT Scan:-<br>MRI: - |
| **29** | F | L | CHL | Microtia | Drug use in pregnancy + Urinary tract infections | CT Scan: Outer ear canal atresia, abnormal fusion of ossicular chain, deformation<br>MRI: - |

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## Discussion

The aim of this research was to contribute to the understanding of the characteristics of unilateral hearing loss. Several studies indicate that gender and other demographic factors might impact the prevalence of unilateral hearing loss, which is more common in males than in females (Genç et al., 2013; Jakubíková, Kabátová, Pavlovčinová, & Profant, 2009; Newton, 2008; Vartiainen & Karjalainen, 1998). In this study, the number of males with unilateral hearing loss was slightly higher than females (M:F; 53.3%:46.7%). This difference is not statistically significant and its clinical significance is controversial.

There are varying data in the literature about the affected side of UHL. Brookhouser et al. (Brookhouser, Worthington, & Kelly, 1991) documented that the left ear was affected in 52% of patients whereas the right ear in 48% of patients in their study. According to data from the Centers for Disease Control and Prevention and from the Early Hearing Detection & Intervention Center in 2015, 50,32 % of 2460 unilateral hearing loss on the right side and 49,43 % of were on the left side (Centers for Disease Control and Prevention, 2015). In this study, the left ear was affected in 44,8 % of patients and the right ear in 55,2%. That is, the present study showed a similar tendency to the aforementioned literature. The side of hearing loss has a significant influence on the development of intellectual functions. Children withright-sided hearing loss had limited aspects of abstract thinking and classifying. Children with left-sided hearing loss had limited intellectual abilities within non-verbal intelligence (Niedzielski, 2006).

There are many risk factors for UHL and it's important to identify these factors for accurate diagnosis and effective rehabilitation of hearing loss. A study published in 2013 by Yelverton et al. (Yelverton et al., 2013), noted that craniofacial anomalies, family history of hearing loss, and the syndromic appearance associated with hearing loss are the highest risk factors in UHL. In their study, Brookhouser et al. (Brookhouser et al., 1991) reported hereditary factors in 12.6% of sensorineural UHL cases and head trauma in 10.8% cases. In a study of 34 patients with UHL by Dodson et al. (Dodson et al., 2012), a family history of hearing loss was reported in 59% of the cases. Forrester and Merz (Forrester & Merz, 2005) stated that microtia is more prevelant in pregnancy with multiples with low birth weight (<2500 grams) and birth at a gestation of <38 weeks. In the present study, the most common risk factor for conductive hearing loss is craniofacial anomalies (30%) whereas the most common risk factors for SNHL are intermarriage (26.3%), hyperbilirubinemia (21.1%), and ototoxic drug use (21.1%). It is thought that our findings support previous findings in the literature about the risk factors of UHL.

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Several studies indicated that the sensorineural hearing loss is most common type in unilateral hearing loss (Genç et al., 2013; Kuppler et al., 2013; van Wieringen et al., 2019). In the present study, 67% of the subjects diagnosed SNHL. A possible reason is that the development of the inner ear is substantially independent from that of the outer and middle ear (12), and development of the inner ear occurs at the earliest stage. The degree of hearing loss is important to treatment strategies. In sensorineural UHL, profound HL is the most common HL level (Centers for Disease Control and Prevention, 2015). Genç et al. (2013) revealed that 19 single-sided sensorineural hearing loss cases were diagnosed, and 63.2% were profound, 21% of severe, 5.3% moderate, and 10.5% mild hearing loss. The most common congenital hearing loss type is profound hearing loss because it is thought to be a problem arising during the embryonic period, affecting a large part of the emerging system.

Although outer-, middle-, and inner-ear malformations are generally observed separately they may be co-existent. Studies indicate a high prevalence of both inner-ear malformations and malformations of the internal auditory canal together in unilateral sensorineural hearing loss detected by high-resolution temporal CT (Dodson et al., 2012; Masuda, Usui, & Matsunaga, 2013; Song et al., 2009). Parrish and Amedee (Parrish and Amedee, 1990), noted that atresia of external ear canal is usually accompanied by auricular deformities. Our study also supports these findings. CT imaging is available in three of four patients with atresia of the external ear canal, and external ear canal stenosis was diagnosed in one patient. It was reported middle ear ossicles malformations in all patients who underwent CT Scan in our study. In some UHL cases, individuals with unilateral hearing loss should be referred for MRI. Laury et al. (Laury et al., 2009) reported the neural hearing loss in 11 of 480 patients with unilateral sensorineural hearing loss. Clemmens et al. (Clemmens et al., 2013) stated that stenosis of the internal auditory canal (<17mm) was generally an indication of the lack of a cochlear nerve. In this study, among 10 patients who had MRIs, 2 (20%) were identified with hearing loss due to cochlear nerve aplasia. In one of these cases, internal auditory canal stenosis was reported. When our findings were interpreted with the literature, it might be considered that the CT Scan and MRI are important to diagnose UHL.

CMV infection, which has a high rate in the literature, has not been encountered in the etiological factors of the patients in this study. This may be due to the low number of subjects. Another issue in the analysis of the etiologic factors was the fact that in patients with UCHL and USNHL, intermarriage rates were as high as 20% and 25%. Here, it is thought that, in our country, the degree of intermarriage is high, and that the etiologic factors in the hearing loss population are the reason for the high degree of intermarriage. This factor, which is more

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frequently seen in bilateral hearing loss in the literature, may be important for UHL (for some populations) and it is thought that it is important to increase future research in this direction.

The heterogonous distribution of the participants according to hearing loss type is one of the major limitations of the study. One other limitation that caused the heterogonous distribution of the participants was the sample size of the study. When these two limitations considered together, it was assumed that the generalization of the findings would be criticized. It is suggested for future studies to enroll equal numbers of diagnosis with larger sample size.

The characteristics of unilateral hearing loss in this study were defined by performing etiologic, otologic, audiologic, and radiologic evaluations of congenital unilateral hearing loss. It has been found that sensorineural hearing loss is most frequently observed in congenital unilateral hearing loss. Mixed hearing loss was not detected in any of these cases. It is thought that the multidisciplinary work of otology, radiology and audiology plays a critical role in the early diagnosis and rehabilitation of even in the diagnosis of unilateral hearing loss.

## Conflict of Interest

The authors whose names are listed immediately certify that they have NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

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## References

Brookhouser, P. E., Worthington, D. W., & Kelly, W. J. (1991). Unilateral hearing loss in children. *The Laryngoscope, 101*(12), 1264-1272.

Centers for Disease Control and Prevention, (2015). Summary of Laterality, Type and Severiy of Identified Hearing Losses: By Ear. 2010 statistics at glance. Erişim: https://www.cdc.gov/ncbddd/hearingloss/2015-data/14-2015-type-and-severity.html

Clark, J.G. (1981). Uses and abuses of hearing loss classification. *ASHA, 23*, 493-500..

Clemmens, C. S., Guidi, J., Caroff, A., Cohn, S. J., Brant, J. A., Laury, A. M., . . . Germiller, J. A. (2013). Unilateral cochlear nerve deficiency in children. *Otolaryngology--Head and Neck Surgery, 149*(2), 318-325.

Dodson, K. M., Georgolios, A., Barr, N., Nguyen, B., Sismanis, A., Arnos, K. S., . . . Pandya, A. (2012). Etiology of unilateral hearing loss in a national hereditary deafness repository. *American journal of otolaryngology, 33*(5), 590-594.

Fitzpatrick, E. M., Gaboury, I., Durieux-Smith, A., Coyle, D., Whittingham, J., & Nassrallah, F. (2019). Auditory and language outcomes in children with unilateral hearing loss. *Hearing research, 372*, 42-51.

Forrester, M. B., & Merz, R. D. (2005). Descriptive epidemiology of anotia and microtia, Hawaii, 1986–2002. *Congenital anomalies, 45*(4), 119-124.

Genç, G. A., Konukseven, Ö., Muluk, N. B., Kirkim, G., Başar, F. S., Tuncer, Ü., . . . Dizdar, H. T. (2013). Features of unilateral hearing loss detected by newborn hearing screening programme in different regions of Turkey. *Auris Nasus Larynx, 40*(3), 251-259.

Jakubíková, J., Kabátová, Z., Pavlovčinová, G., & Profant, M. (2009). Newborn hearing screening and strategy for early detection of hearing loss in infants. *International journal of pediatric otorhinolaryngology, 73*(4), 607-612.

Kuppler, K., Lewis, M., Evans, A. (2013). A review of unilateral hearing loss and academic performance: Is it time to reassess traditional dogmata? *International journal of pediatric otorhinolaryngology, 77, 617-622.*

Laury, A. M., Casey, S., McKay, S., & Germiller, J. A. (2009). Etiology of unilateral neural hearing loss in children. *International journal of pediatric otorhinolaryngology, 73*(3), 417-427.

Masuda, S., Usui, S., & Matsunaga, T. (2013). High prevalence of inner-ear and/or internal auditory canal malformations in children with unilateral sensorineural hearing loss. *International journal of pediatric otorhinolaryngology, 77*(2), 228-232.

Newton, V. E. (2008). *Paediatric audiological medicine*: John Wiley & Sons.

Niedzielski, A., Humeniuk, E., Blaziak, P., Gwizda, G. (2006). Intellectual efficiency of children with unilateral hearing loss. *International Journal of Pediatric Otorhinolaryngology, 70*, 1529-1532.

Parrish, K.L., Amedee, R.G. (1990). Atresia of the external auditory canal. *The Journal of the Louisiana State Medical Society: official organ of the Louisiana State Medical Society*, 142(9), 9-12.

Song, J.-J., Choi, H. G., Oh, S. H., Chang, S. O., Kim, C. S., & Lee, J. H. (2009). Unilateral sensorineural hearing loss in children: the importance of temporal bone computed tomography and audiometric follow-up. *Otology & Neurotology, 30*(5), 604-608.

van Wieringen, A., Boudewyns, A., Sangen, A., Wouters, J., & Desloovere, C. (2019). Unilateral congenital hearing loss in children: Challenges and potentials. *Hearing research, 372*, 29-41.

Vartiainen, E., & Karjalainen, S. (1998). Prevalence and etiology of unilateral sensorineural hearing impairment in a Finnish childhood population. *International journal of pediatric otorhinolaryngology, 43*(3), 253-259.

Yelverton, J. C., Dominguez, L. M., Chapman, D. A., Wang, S., Pandya, A., & Dodson, K. M. (2013). Risk factors associated with unilateral hearing loss. *JAMA Otolaryngology–Head & Neck Surgery, 139*(1), 59-63.