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Original Article

The Audiological Profile and Rehabilitation of Patients with Incomplete Partition Type II and Large Vestibular Aqueducts

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BACKGROUND: Incomplete partition type II (IP-II) malformation is often accompanied by a large vestibular aqueduct (LVA). In IP anomalies, the patient's auditory rehabilitation requirements are decided according to the presence of inner ear structures and the degree of hearing loss (HL). There has been limited research on auditory rehabilitation (AR) requirement selection in patients diagnosed with IP-II and LVA. This study investigated the typical characteristics of HL and AR choices in patients diagnosed with IP-II and LVA.

METHODS: Patients with IP-II and LVA (n = 55; 25 women and 30 men) were identified, and audiological evaluations were performed. The patient's demographic characteristics, the type and degree of HL, the AR method, age at diagnosis, and educational status were retrospectively compared.

RESULTS: The distribution of our 55 patients according to cochlear implants, hearing aids (HA), and bimodal applications was 29.1% (n = 16), 43.6% (n = 24), and 27.3% (n = 15), respectively. Statistical analyses using chi-square tests found no significant differences in the incidence of dizziness/imbalance, tinnitus, HL progression, or the degree and onset of HL among the patients.

CONCLUSION: The data revealed different audiological characteristics among patients with IP-II and LVA, as well as different AR solutions. The most widely used AR modality was found to be HA. Prediction of sudden versus progressive HL development among patients is challenging, and the characteristics of IP-II vary. Therefore, they should be interpreted with caution.

KEYWORDS: Audiological rehabilitation, auditory training, hearing loss, iIncomplete partition type II, inner ear malformations, large vestibular aqueduct

INTRODUCTION

Inner ear malformations (IEMs) represent 20%-35% of congenital and sensorineural hearing loss (SNHL) etiology. Inner ear malformations are divided into types depending on embryological development and radiological findings. They are significant causative agents in congenital SNHL.[1-5] Incomplete partition (IP) malformations with normal external dimensions and abnormal internal cochlear structures comprise the most common IEM subgroup. IP malformations can be divided into IP-I, IP-II, and IP-III. In cases of Mondini deformity, the cochlea has one and a half instead of two and a half turns, and the middle and apical turns combine to form a cystic crest.[6] Incomplete partition-II is often accompanied by a large vestibular aqueduct (LVA), which is an anatomical anomaly characterized by enlargement of the vestibular aqueduct in the temporal bone.[7] Incomplete partition-II with LVA is one of the most common anomalies. It generally occurs bilaterally.[7-8] Considering the etiology of LVA, the anomaly may be caused by either a secondary genetic mutation during embryonic development or environmental factors during early childhood.[5] The etiology of hearing loss (HL) in patients with LVA has not been fully elucidated. However, many mechanisms have been proposed, including cochlear dysplasia, damage to hair cells due to increased endolymphatic fluid pressure, and genetic factors.[9] Although bilateral SNHL is the most common auditory disorder in LVA, conductive HL is sometimes seen.[10] The auditory rehabilitation (AR) options for patients with IP malformations are usually either hearing aids (HAs) or cochlear implants (CIs), depending on the presence of inner ear structures and the degree of HL.[1] Rarely, audiological follow-up without intervention is the appropriate course in cases with minimal HL.[1] To the best of our knowledge, there have been few studies on the characteristics of HL and suggested AR

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methods for patients with IP-II and LVA. Batuk et al[1] have investigated the type and severity of HL and the recommended AR options for IP-II malformations but did not analyze the results or take auditory training and other audiovestibular findings into account. Berettini et al[16] examined some of the audiological features of LVA patients with various anomalies, but there were no patients with LVA-related IP-II malformations in their sample, and the number of participants was limited. Therefore, this study aimed to characterize the auditory profile of patients diagnosed with IP-II and LVA and to classify the AR options for patients with various types of HL.

MATERIAL AND METHODS

The medical database of the Department of Audiology at Hacettepe University Hospital was retrospectively searched for patients previously diagnosed with IP-II and LVA by an ear, nose, and throat physician. This study was approved by the Hacettepe University Non-Invasive Clinical Research Ethics Committee (Approval No:GO 20/1156, Date: March 16, 2021). All participants gave written informed consent for study participation.

Participants

The study participants were patients diagnosed with IP-II and LVA at our institution. IP-II malformations were diagnosed by an experienced neuroradiologist and a neuro-otologist using axial sections on high-resolution computerized tomography (SOMATOM Plus 4 Volume Zoom, Siemens, Erlangen, Germany).[11] Among patients in the clinical database diagnosed with different IEMs, information was retrieved from 120 patients with bilateral LVA and IP-II anomalies. Their data were retrospectively reviewed, and those aged 8–42 years diagnosed with IP-II and LVA were included in the study. Of the 120 patients, 55 (25 females and 30 males) met these inclusion criteria. The rest were excluded. The mean age of the participants was 26 $\pm$ 1.17 years. Individuals who were diagnosed and treated between 2000 and 2015 were included in this study. Hearing aid (HA) and CI users in our study used their devices bilaterally. Bimodal users had a hearing aid in 1 ear and a cochlear implant in the other. All the study's hearing aid and cochlear implant users use their devices regularly (at least 8 hours a day). All individuals included in the study were followed audiologically at regular intervals (3-6 months). Hearing aid applications were performed on individuals who used hearing aids and experienced progressive hearing loss during their audiological follow-up. Patients who could not benefit from hearing aids due to hearing loss and speech comprehension scores in free field measurements were evaluated for cochlear implant candidacy by a multidisciplinary team consisting of an otolaryngologist, radiologist, psychologist, and expert audiologist. As a result of anatomical structure and audiological evaluations, CIs were applied to suitable candidates. The fittings of individuals using CIs and hearing aids were checked during their routine follow-up.

Audiological Evaluation

The audiological findings of all patients were retrospectively analyzed. Information on family history, tinnitus, dizziness, imbalance complaints, type of HL (conductive, mixed, or sensorineural), AR selections, and auditory training status was extracted from the database records. A two-channel audiometer (GSI 61, Grason-Stadler, Inc., USA) was used for pure tone audiometry tests. Experienced audiologists performed these tests with patients in a soundproof cabinet. TDH-39 headphones and B-71 bone vibrators were used to measure air and bone conduction thresholds, respectively. Relevant thresholds at related frequencies were recorded in decibels (dB). The requisite masking procedures were applied to the respective tests' air and bone thresholds. Degrees of HL were organized according to Goodman's (1965) classification.

Statistical Analysis

All statistical analyses were performed using Statistical Package for the Social Sciences Statistics software, version 23.0 (IBM SPSS Corp.; Armonk, NY, USA). Frequency statistics (number of cases [n] and percentage [%]) were used for categorical variables, including the demographic characteristics of patients. Patients were divided into subgroups based on the type and degree of their HL and AR. Chi-square tests were used to compare the relationship of HL type, chronological age, and age at diagnosis with the presence of progression; the effects of HL degree on audiovestibular symptoms such as dizziness/imbalance and tinnitus, as well as on HL onset (congenital versus sudden), AR (CI, HA, or bimodal), and auditory training; and the associations between AR type and HL type and auditory training. Statistical significance was set at $P < .05$.

MAIN POINTS

- This study is the first to examine the audiological profile and intervention options exhaustive in patients with LVA anomaly accompanying IP-II.
- In order to determine the path to be followed in the management of inner ear anomalies, it is important to determine the comparison of the audiological profile and intervention methods of this group in detail.
- Although it is known in the literature that these two anomalies often accompany each other, no study examining the characteristics of such a high number of patient groups has been found.
- The study provides a comprehensive analysis, highlighting important factors to consider in the rehabilitation of these patients.
- The results show that IP-II and LVA patients have different auditory characteristics and cochlear implants are the most commonly used rehabilitation method. These data highlight the importance of individualized treatment approaches to assess patients' specific needs.

RESULTS

The audiological profiles of patients diagnosed with IP-II and LVA were investigated in detail, and comparisons were made between the suggested AR options and the factors affecting HL. The demographic characteristics of patients are shown in Table 1.

Of the 55 patients included, 45.5% were female, and the remaining 54.5% were male. The onset of HL in patients was 85.5% prelingual, 14.5% post-lingual, 76.4% congenital, and 23.6% sudden. The type of HR was 65.4% sensorineural and 34.6% mixed. The degree of HL was slight-to-mild in 29%, mild-to-moderate in 23.63%, moderate-to-severe in 20%, and severe-to-profound in 27.27% of the patients. There was a progression of HL in 56.4% of the patients and no progression in the remaining 34.6%. Tinnitus was observed in 63.6% of the patients. The AR options of the patients in the study were 43.6% HA, 29.1% CI, and 27.3% bimodal. Auditory training was received by 72.72% of the patients.

Table 1. Audiological and Demographic Information of All Patients

	Group	n	Percent (%)
Gender	Female	25	45.5
	Male	30	54.5
HL onset	Prelingual	47	85.5
	Postlingual	8	14.5
	Congenital	42	76.4
	Sudden HL	13	23.6
Type of HL	Sensorineural	43	78.2
	Mixed	12	21.8
Degree of HL	Slight to Mild	16	29
	Mild to Moderate	13	23.63
	Moderate to Severe	11	20
	Severe to Profound	15	27.27
Progress of HL	Yes	31	56.4
	No	24	43.6
Family history	Yes	35	63.6
	No	20	36.4
Dizziness/imbalance	Yes	7	21.8
	No	48	78.2
Tinnitus	Yes	12	21.8
	No	43	79.2
Audiological rehabilitation	HA	24	43.6
	CI	16	29.1
	Bimodal	15	27.3
Auditory training	Yes	40	72.72
	No	15	27.28

CI, Cochlear implant; HA, hearing aid; HL, hearing loss.

When the type of HL was evaluated for progression, mixed HL was found in 18.18%. The progression of HL in those with SNHL was 41.81%. There were no significant relationships between the presence of progression and HL type, chronological age, or age at diagnosis ($P > .05$) (see Table 2).

When the effects of HL grade on audiovestibular symptoms such as dizziness/imbalance and tinnitus, the onset of HL (congenital or sudden), AR (CI, HA, or bimodal), and receipt of auditory training were compared, no statistically significant differences were found ($P > .05$). Details are given in Table 3.

Table 2. Comparison of Progression with Type of Hearing Loss, Age of Hearing Loss, and Chronological Age

		Progression		*P*
		No	Yes	
Hearing loss type	Mix	9 (16.36)	10 (18.18)	>.05
	Snik	13 (23.63)	23 (41.81)	
Chronologic age		29 (52.72)	26 (47.27)	>.05
Hearing loss diagnosis age		24 (43.63)	31 (56.36)	>.05

The effects of the degree of HL degree on audiovestibular symptoms such as dizziness/imbalance and tinnitus; HL onset (congenital versus sudden); AR (CI, HA, and bimodal); and auditory training were not statistically significant ($P > .05$).

Table 4 compares the HL and auditory training types according to the AR options. No statistically significant differences were found. There were also no significant differences in AR according to HL type or receipt of auditory training ($P > .05$) (See Table 4).

DISCUSSION

Although many studies have analyzed the characteristics of IEM to our knowledge, this is the first to examine the relationship between audiological characteristics and AR options in patients with IP-II and LVA.[1-12,13] Several studies have shown different pathophysiological features in different IP anomalies.[11-14] Despite the frequency with which LVA malformations accompany IP-II, the exact mechanism of HL in this population remains unknown, leading to controversy regarding the appropriate interventions. Previous studies have reported fluctuating or progressive moderate-to-severe HL among this patient population. For instance, Rose et al[15] have reported moderate-to-severe HL in isolated LVA malformations. They also found that, although different types and degrees of HL were observed, the percentage of patients with severe-to-profound HL was 76% in IP-II.[15] Regarding the AR options, interventions are selected according to the type and degree of HL in IP-II and LVA.[3] Hearing aids and CIs are appropriate AR options for these patients.[1-5] In this study, we compared the audiological characteristics of patients with IP-II and LVA and their rehabilitation options. We compared the degree of HL with the presence of dizziness/imbalance, the presence of tinnitus, the progression of HL, HL onset, and the receipt of auditory training. We also compared the effects of HL progression and type and investigated the relationship between auditory training and rehabilitation.

Batuk et al[1] researched the degree of HL in 74 patients diagnosed with IP anomalies and found normal hearing in 2 patients, mild HL in 1 patient, mild-to-moderate HL in 15 patients, and severe-to-profound HL in 56 patients. Of these, 2 patients had conductive HL, 52 had mixed HL, and 20 had SNHL.[1] Berrettini et al[16] examined 17 patients with LVA anomalies and observed moderate HL in 2 patients and profound HL in 15, while 10 patients had conductive HL. In the present study, 16 of the 55 patients had slight-to-mild HL, 13 had mild-to-moderate HL, 9 had moderate-to-severe HL, and 18 had severe-to-profound HL. Furthermore, 19 had mixed HL, and 36 had SNHL.

Gopen et al[17] found conductive HL to be the least common type in patients with LVA. Other studies have reported that, while most LVA patients are diagnosed with SNHL, some have an air-bone gap at low frequencies.[17-19] In these patients, the masking dilemma may occur with profound HL, and the inability to reliably determine the bone thresholds in children may explain the common misdiagnosis of patients with LVA who show a conductive component on audiograms.[17-19] In the early 2000s, some researchers suspected fixation, relaxation, or discontinuity of the ossicles among these patients. However, the recent increase in exploratory tympanotomies has shown that these characteristics do not relate to the middle ear.[20-22] Moreover, the improvement in the bone conduction mechanism

Table 3. Comparison of Hearing Loss Degree with Audiological Characteristics

Hearing Loss Degree							
		Slight to Mild	Mild to Moderate	Moderate to Severe	Severe to Profound	Total	*P*
		n (%)	n (%)	n (%)	n (%)	n (%)	
Congenital hearing loss		13 (23.63)	10 (20)	7 (12.72)	13 (23.63)	43 (78.18)	>.05
Sudden hearing loss		3 (5.45)	3 (5.45)	2 (3.63)	5 (9.09)	12 (21.81)	>.05
Progression	No	8 (14.54)	8 (14.54)	9 (16.36)	6 (10.90)	31 (56.36)	>.05
	Yes	8 (14.54)	5 (9.09)	2 (3.63)	9 (16.36)	24 (43.63)	>.05
Dizziness/imbalance	Yes	1 (1.81)	3 (5.45)	4 (7.27)	5(9.09)	7 (12.72)	>.05
	No	15 (27.27)	10(18.18)	8 (14.54)	9(16.36)	48 (87.27)	>.05
Tinnitus	No	13 (23.63)	10 (18.8)	7 (12.72)	13 (23.63)	43 (78.18)	>.05
	Yes	3 (5.45)	3 (5.45)	4 (7.27)	2 (3.63)	12 (21.81)	
Auditory training	No	2 (3.63)	6 (10.9)	1 (1.81)	6 (10.90)	15	>.05
	Yes	14 (35)	7 (12.72)	10 (18.18)	9 (16.36)	40	
Audiological rehabilitation	CI	-	1 (1.81)	5 (9.09)	10 (18.18)	16	>.05
	HA	2 (3.63)	6 (10.90)	10 (18.18)	6 (10.90)	24	
	Bimodal	1 (1.81)	1 (1.81)	5 (9.09)	8 (14.54)	15	

that results from LVA is thought to be a potential cause of conduction component production.[18,19,23,24] There has been no thorough general analysis of the types of HL associated with this malformation because the origin of this abnormality remains unknown, and there is no comprehensive data on its progression. In the present study, as in a study by Gopen, the number of patients with conductive HL was lower than that with SNHL. Despite the absence of patients with conductive HL in our study, 35% (n = 19) had mixed HL. The type of HL may change depending on the retrospectively obtained data, including age at diagnosis and the progression of HL. Concerning HL progression, there may be several AR options for IP-II and LVA. Batuk et al evaluated AR options and found that patients with IP-II generally use HAs and CIs.[1] In the present study, 24 (43.6%) patients were HA users, 16 (29.1%) were CI users, and the remaining 15 (27.3%) were bimodal users. Thus, according to these findings, the most commonly used AR is CI. There was no statistically significant difference between IP-II and LVA in terms of AR and the degree and type of HL ($P > .05$). It is very important to provide early hearing rehabilitation to children with hearing loss. As the degree of hearing loss increases, the performance of CIs becomes better than that of traditional acoustic HAs. Determining the transition point from HA to CI is very important. Clinical decisions regarding cochlear implant candidacy are made individually by multidisciplinary teams, considering factors such as audiometric thresholds, speech perception, language development, health, history of hearing, previous device use, anatomy, and additional needs. Assessing speech and language in young children is challenging; this underlines the importance of audiometric thresholds in candidacy decisions.[25,26] The decision to offer a patient a cochlear implant can significantly impact many aspects of his or her life. Although many studies have reported outcomes in HAs and cochlear implant users, studies of individuals with profound hearing loss have focused on a limited number of outcomes with users of acoustic HAs only. With recent technological advances, improvements have been made in microphone directing and noise reduction in CIs. In children with hearing loss, recommending the use of HAs or CIs for those with better residual hearing thresholds than candidates with typical hearing loss may affect a variety of outcomes not routinely assessed.[27-32] These outcomes include spatial hearing, listening effort, psychosocial well-being, balance function, tinnitus, and music perception. Although optimizing these outcomes is not the primary goal of hearing rehabilitation, it is critical to the child's overall quality of life, mental health, social participation, and academic success.[33-39] In our study, CIs were applied to patients who were diagnosed and treated between 2000 and 2015, did not benefit from HAs, had progressive hearing loss, developed receptive and expressive language skills, and had severe-profound hearing loss. In addition, CIs were applied to patients whose speech perception was not within the HA thresholds, who could not obtain sufficient gain from the HA in the high-frequency region of their audiogram, and whose threshold for distinguishing speech from the HA was low. Additionally, in this study, individuals with moderate to severe hearing loss who had hearing thresholds remaining at low frequencies were followed audiologically with HAs, assuming that they would be able to use low-frequency speech cues more effectively with the use of HAs.

Table 4. Comparison of Audiological Rehabilitation with Type of Hearing Loss and Receiving Auditory Training

		Audiological Rehabilitation			Total	*P*
		HA n (%)	CI n (%)	Bimodal n (%)		
Hearing loss type	Mix	14 (30.9)	2 (3.63)	3 (5.45)	19	>.05
	Snik	10 (12.72)	14 (25.45)	12 (21.81)	36	>.05
Auditory training	No	7 (12.72)	4 (7.27)	4 (7.27)	15	>.05
	Yes	17 (30.9)	12 (21.81)	11 (20)	40	

Batuk et al[1] stated that the cochlear nerve is often normal in patients with IP-II since there is no internal acoustic canal damage. Among

incomplete partition anomalies, IP-II anomalies are the anomalies in which the internal organization of the cochlea is most developed. Although the apical part of the modiolus is malformed, the basal part is intact. IP-II anomalies, which have a relatively more intact anatomy compared to other anomalies, constitute the group that can benefit most from CIs among inner ear anomalies. Additionally, Kocabay et al[40] stated that children with IP-II malformations benefit from cochlear implantation in terms of sound localization and speech understanding skills as much as children with normal cochlear morphology. Furthermore, Budak et al[41,42] stated that the quality of life of CI users with IP-II anomalies is as good as that of cochlear implant users with normal anatomy. In line with the retrospective information obtained from the tests performed in the past years of the patients in our study, it was observed that the thresholds measured in the free field were within the speech field and that the speech discrimination thresholds with CIs were compatible with the measurement results. These results suggest that this patient group benefits from CIs. Hearing aid and auditory implant manufacturers are introducing new sound processing algorithms and microphone directionality options today.[33,43] Considering the evolving practice and technology environment, evaluating cochlear implantation candidacy or HA use in individuals with moderate to severe hearing loss is important, considering the patient's quality of life, speech understanding in noise, and optimizing spatial skills.

Large vestibular aqueduct is an inner ear anomaly that can cause deterioration of the auditory and vestibular systems.[8] In Yang et al[44], 6 (22%) of 27 children with LVA reported symptoms of vertigo.[45] In Berettini et al[16], vestibular evaluations showed that, while 13 of 17 patients had vestibular weakness, only 7 (41.1%) had vestibular symptoms. In the present study, 7 (12.7%) of the 55 patients reported dizziness/imbalance. The low percentage of vestibular symptoms may be due to the retrospective nature of this study. Although our patients reported symptoms of active vestibular disorders in the past, no active symptoms were reported during follow-up. Vestibular disorders are common among patients with LVA; however, tinnitus and ear fullness rarely accompany this syndrome.[7,8] Among our participants, 12 (21%) reported tinnitus. This rate suggests that tinnitus is relatively frequent in LVA and IP-II malformation patients. Therefore, anamnesis should be carefully performed.

Masuda et al[46] investigated the relationship between family history and the incidence of unilateral and bilateral IEM and found that 26 of 81 patients with bilateral IEM had a family history of these malformations. A family history of IP-II was observed in 35 of our 55 patients (63%). This may be indicative of the genetic etiology of IEM.

Batuk et al[1] have suggested that HL progression is a feature of IP-II malformation. Berettini et al[16] examined the audiological characteristics of patients with LVA and observed progressive HL in 11 of 17 patients. Similarly, we observed a high rate of 56% (n = 31) of progressive HL in our patients. We compared the progression, degree, and type of HL, age at diagnosis, and chronological age, but no statistically significant differences were observed ($P > .05$).

Despite the progressive characteristics of IP-II malformations, chronological age and age at diagnosis were not associated with the degree or type of HL. As a result, it was difficult to determine the effects of IP-II and LVA on the progression of HL.

Decreased hearing sensitivity in the first few years of life can negatively affect language, verbal communication, cognition, and educational progress. Early intervention can reduce the harmful effects of HL.[10] To the authors' knowledge, there have been no previous studies on the rate of auditory training in IP-II. In our study, 72% (40 of 55) of the patients had received auditory training. No significant difference was observed between patients who received auditory training and the degree of HL ($P > .05$). We also found no significant differences between patients who received auditory training and those who did not ($P > .05$). In our study, individuals receiving auditory training were between the ages of 8 and 17. When diagnosed with hearing loss, these individuals started auditory training and used any auditory rehabilitation option. The rate at which people received auditory training decreased with age. None of the adult users received auditory training.

Casselman et al[47] have reported sudden HL in 2.3% of patients with inner ear anomalies. However, Birman and Gibson[48] reported a much higher rate of sudden SNHL in patients with LVA than those without inner ear anomalies. They found that an enlarged internal acoustic canal (IAC) might be associated with partial detachment of the lateral fundus and other temporal bone anomalies. This leads to increased communication between the IAC and the inner ear and progressive or fluctuating SNHL.[47,48] Sudden HL was reported in 5 of 17 (29%) of Forli et al's[7] patients' with LVA malformations. Similarly, sudden HL was observed in 12 (23%) of our 55 patients. We found no relationship between sudden HL and the degree of HL. This finding is thought to be due to the variability in IP-II malformations.

Limitations and Strengths

The retrospective nature of this study was a limitation. Future clinical studies are needed with larger samples to evaluate all aspects of the audiological profiles of this patient population. However, the present study will contribute to the knowledge of such audiological characteristics and planned audiological interventions in patients with IP-II and LVA. To the authors' knowledge, the audiological characteristics and AR options have yet to be investigated in IP-II and LVA patients. Furthermore, selection from the AR options could be made based on the examination of IP-II patient characteristics during the follow-up period.

Incomplete partition type II and LVA are the most common comorbid malformations in audiology. The most widely used AR modality was found to be CIs. Predicting the development of sudden versus progressive HL among patients is challenging. The findings of this study have shown variable features among patients with the IP-II anomaly.

Ethics Committee Approval: This study was approved by Ethics committee of Hacettepe University (Approval No:GO 20/1156, Date: March 16,2021).

Informed Consent: Informed consent was obtained from the patients who agreed to take part in the study.

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