30 Audiological Outcome with Cochlear Implantation

Hilal Burcu Ozkan, Betul Cicek Cinar, Gonca Sennaroglu, Esra Yücel, and Levent Sennaroglu

30.1 Introduction

Surgical and technological developments in CI technology have made a significant difference in the rehabilitation of hearing loss in individuals with inner ear malformations (IEMs). It is not surprising that there has been an increase in studies on the results of cochlear implantation in IEMs over the last decade. IEMs represent approximately 20–35% of the etiology of congenital sensorineural hearing loss (SNHL) cases based on radiology [1–4]. Incidence of IEMs was reported as 20% in our clinic [5].

There are certain challenges in the management of IEMs such as cerebrospinal fluid gusher, which is a risk for meningitis, facial nerve anomalies, decision making for the surgical approach and the type of electrode, choosing the correct implantation method; CI versus auditory brainstem implantation (ABI) and timing of surgery. It is very important to know about these possible risks for better rehabilitative counseling after surgery. Sennaroglu's classification correlates the surgical issues related to specific IEMs [5].

Classification of IEMs is based on differences in cochlear anatomy in various malformations. In addition to duration of deafness and preoperative auditory perception, IEMs have to be considered as an important limiting factor for successful CI outcomes. Children who qualify for and undergo CI surgery participate in follow-up testing at regular intervals for a period of 3 years.

H. B. Ozkan (✉) · B. C. Cinar · G. Sennaroglu
E. Yücel
Audiology Department, Faculty of Health Sciences, Hacettepe University, Ankara, Turkey

L. Sennaroglu
Department of Otolaryngology, Hacettepe University, Faculty of Medicine, Ankara, Turkey

30.2 Literature Review

Tucci et al. [6] reported CI outcomes in five children and one adult with IEMs. IEMs included common cavity (CC) deformity ($n = 1$), cochlear hypoplasia (CH) ($n = 2$), and incomplete partition (IP) ($n = 3$) anomalies. According to their results all patients showed improved performance after implantation. Four patients obtained open-set speech perception. Two remaining patients, whose poor language skills precluded administration of standard tests, showed increased awareness of environmental sounds and increased vocalization after implantation.

Luntz et al. [7] evaluated 10 CI users with IEMs: 3 CC deformity, 4 IP anomalies, 2 membranous deformity, and 1 enlarged vestibular aqueduct (EVA). Their study indicated that all 22 electrodes were inserted in 9 of 10 children. Each of the patients demonstrated speech awareness at 25 dB HL or better. Data was available after 30 months of experience in 4 of the 10 patients, and 3 (75%) of the 4 showed some degree of open-set word recognition. Six patients demon-

L. Sennaroglu (ed.), *Inner Ear Malformations*, https://doi.org/10.1007/978-3-030-83674-0_30

strated only speech detection and/or closed-set recognition scores.

Eisenman et al. [8] showed GASP-W scores of the 17 children with IEMs 12 months after implantation. Their results were worse, with slower rate of improvement than those of children with normal cochlea; however, by 24 months there was no significant difference between the two groups.

Buchman et al. [9] analyzed 28 pediatric CI users with the constellation of an IP, EVA, and dilatation of vestibule. Those with an isolated EVA or partial semicircular canal aplasia have relatively good levels of speech perception. Users with total semicircular canal aplasia, isolated IP, cochlear hypoplasia, or common cavity demonstrated lower levels of performance.

Papsin et al. [10] reported that children with IP obtained higher average speech perception outcomes because they were more likely to have a progressive hearing loss and, as a group, had superior linguistic skills before implantation. Children with CC deformity and CH demonstrated a tendency toward poorer performance despite inclusion in the data set of speech perception scores from children who were older and had significant language before implantation. Even the poorest performers with CH and CC showed speech perception gains with increased implant use.

Sainz et al. [11] reported word perception scores of CI users with common cavity and cochlear hypoplasia. They reported that these subjects demonstrated poor word perception and were unable to discriminate more than 50% of words and mostly relied on visual cues.

Isaiah et al. [12] illustrated that speech perception scores following cochlear implantation in children with IEMs were overall below than CI users with normal anatomy.

30.3 Results of Hacettepe University

Between November 1997 and September 2018, 2639 patients underwent CI and ABI in our department. Out of 2639 cases, 278 children have different IEMs, such as common cavity (CC), enlarged vestibular aqueduct (EVA), incomplete partition of the cochlea (IP-I, IP-II, IP-III), cochlear hypoplasia (CH), and dilatation of vestibule were implanted with CI. A retrospective study on auditory performance and language development of CI children with different IEMs was conducted in our clinic and these results were recently submitted for publication. In this chapter, auditory performance and language development of CI children with different IEMs were reported based on the results of this study. One hundred thirty-seven of 278 CI users were younger than 18 years old and had at least 1 year of cochlear implant experience.

All IEMs, aged between 12 months to 18 years, were matched with their peers in control group according to their chronological and implantation age (±8 months). All children are using their CI in daily basis. The distribution according to the number of IEMs and participants is shown in Table 30.1. These numbers were similar to those reported by a study in 2021 [13].

Depending on the wound healing, pediatric audiologists perform the initial activation of the electrodes between 3 days and 4 weeks after surgery. All children underwent hearing thresholds verification by audiometric testing after each CI programming session. Free field thresholds with CI were obtained at 0.25, 0.5, 1, 2, and 4 kHz using warble-tone or narrow-band stimuli and speech detection test was done through live voice using /ba/, /ss/, and /sh/ phonemes. Although some children get thresholds at 25–35 dB at 0.25, 0.5, 1, 2, and 4 kHz, others with limited benefit from CI get thresholds at 35–55 dB. Characteristic hearing thresholds of various IEMs and their free field tests are presented in Chapters 21 and 23–27.

As mentioned in Chap. 8, auditory perception skills were evaluated with a comprehensive test battery called as “Children’s Auditory Perception Skills Test in Turkish (CIAT)” [14]. All participants were evaluated before and after CI with Ling’s sound detection test, the Meaningful Auditory Integration Scale (MAIS) or Infant-

Table 30.1 Chronological age and duration of CI use of children with IEMs

	Common cavity		IP-I		IP-II		IP-III		EVA		Cochlear hypoplasia		Dilatation of vestibule	
	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)	Chr. age (months)	Drt. of CI use (months)
Mean	96.3	32.8	111.8	42	115.6	45.3	95.6	32.8	140.5	60	103.6	48.3	101.1	30
Min.	40	16	42	18	31	12	39	12	60	12	87	42	24	12
Max.	177	60	216	88	216	143	215	93	182	144	123	55	216	106

Toddler Meaningful Auditory Integration Scale (IT-MAIS), and CIAT's subtests such as closed-set Pattern Perception Test and open-set Daily Turkish Sentence Recognition Test. All testing was done at 1–6 months after CI activation and follow-ups were done during 1–3 years with different intervals.

Language development skills were assessed using the Test of Early Language Development-Third Edition (TELD 3). This test provides us with receptive and expressive language performances of children [15]. However, this test was applied only to the group with IEMs and the results were not compared with the group of children with NC.

The speech perception and language development outcomes are presented according to the classification of IEMs as follows:

30.3.1 Common Cavity (CC)

We evaluated eight children with CC in terms of auditory perception performances and language skills (Figs. 30.1 and 30.8). In the Ling's Sound Test and MAIS evaluation, preoperative performance of children with CC had lower scores compared to children with NC and statistically significant differences were found ($p < 0.05$). After cochlear implantation, for auditory perception Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed. There was a statistically significant difference between children with CC and NC in open-set and closed-set tests except Ling's Sound Test ($p < 0.05$). One to three years after CI, children with CC had a score of 25.38 points, while children with NC had a score of 40 full points in MAIS test. Similarly, closed-set Pattern Perception Test results were obtained 35.38% in children with CC and 82.63% in children with NC. The lowest score belongs to the Daily Turkish Sentence Recognition Test, which is an open-set 0% for CC and 46.13% for NC. Language skills assessments conducted with TELD-3 showed that the receptive language age of the children with CC is average 56.25 months, while the expressive language age is average 42.5 months. Children with CC obtained the lowest scores in terms of auditory perception performance among IEMs.

30.3.2 Cochlear Hypoplasia (CH)

We evaluated 26 children with CH in terms of auditory perception performances and language skills (Figs. 30.2 and 30.8). Before CI in the Ling's Sound Test, children with CH had lower scores than children with NC and statistically significant differences were found ($p < 0.05$). There was no statistically significant difference in MAIS test. One to three years after cochlear implantation, Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed for auditory perception. There was a statistically significant difference between children with CH and NC in open-set and closed-set tests except Ling's Sound Test ($p < 0.05$). In the MAIS test the children with CH had a score of 31.08 points, while the children with NC had a score of 38.73 points. Pattern Perception as closed-set test results showed that children with CH had lower scores (47.65%) than children with NC (85.12%). Similar to the results of CC, the lowest score belongs to the Daily Turkish Sentence Recognition Test (CH 5.96% and NC 45.15%). According to language skills assessments conducted with TELD-3, the receptive language age of the children with CH is average 58.34 months, while the expressive language age is average 41.53 months. Children with CH constituted the group of children with the second lowest scores in terms of auditory perception performance among IEMs.

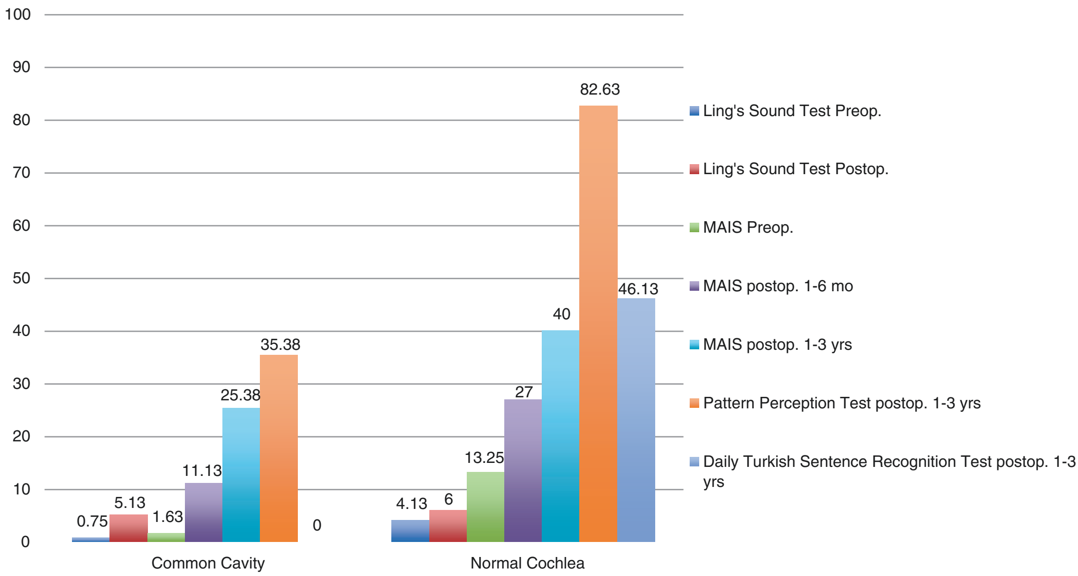

Fig. 30.1 Comparison of children with CC and NC in terms of auditory perception performance (CC = common cavity, NC = Normal cochlea)

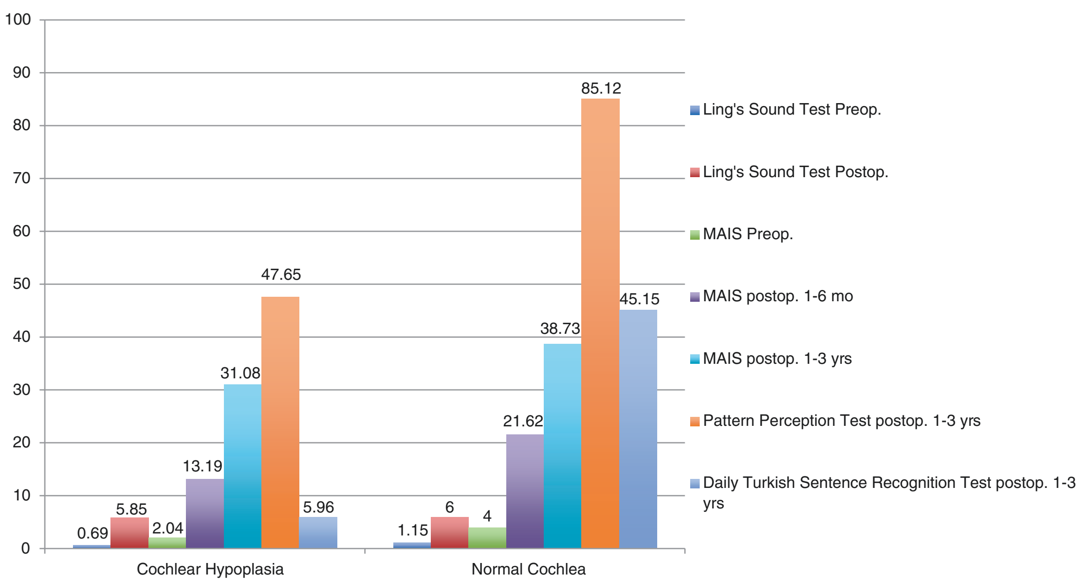

Fig. 30.2 Comparison of children with CH and NC in terms of auditory perception performance (CH = Cochlear hypoplasia, NC = Normal cochlea)

30.3.3 Incomplete Partition Anomalies of the Cochlea

30.3.3.1 Incomplete Partition Type I (IP-I)

We evaluated 36 children with IP-I in terms of auditory perception performances and language skills (Figs. 30.3 and 30.8). In the Ling's Sound Test and MAIS evaluation performed before the CI, children with IP-I had lower scores than children with NC and statistically significant differences were found ($p < 0.05$). After cochlear implantation, auditory perception tests such as Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were per-

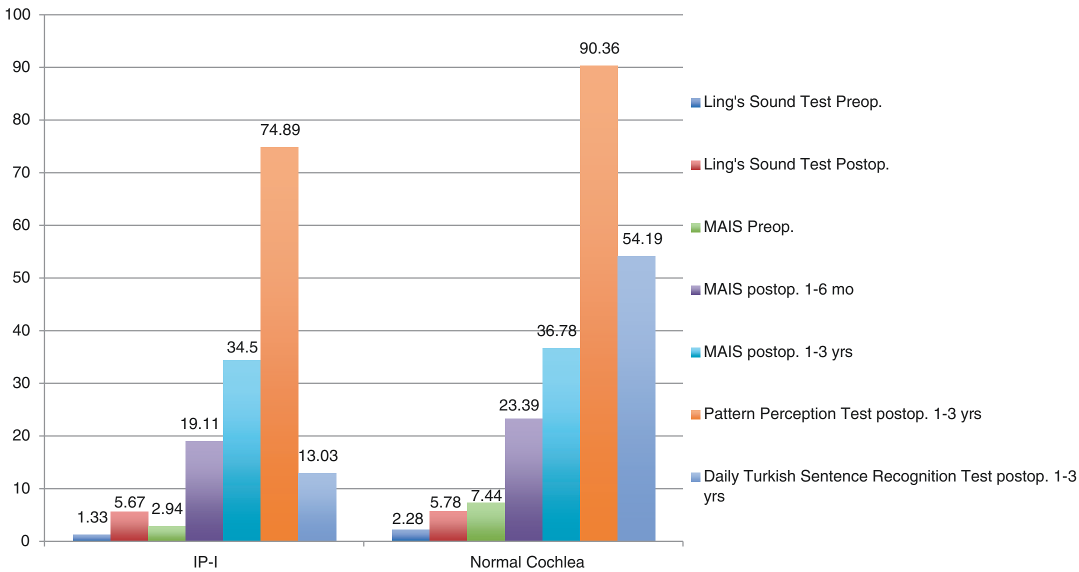

Fig. 30.3 Comparison of children with IP-I and NC in terms of auditory perception performance

formed. There was a statistically significant difference between children with IP-I and NC in Pattern Perception Test and Daily Turkish Sentence Recognition Test ($p < 0.05$). In the MAIS test between 1 and 3 years after CI, children with IP-I had a score of 34.5 points while the children with NC had a score of 36.78 points. According to Pattern Perception Test as closed-set results between 1 and 3 years after CI, children with NC had 90.36% and IP-I children had 74.89%. The lowest score belongs to the Daily Turkish Sentence Recognition Test (IP-I 13.03% and NC 54.19%). In addition to auditory perception evaluation, language skills assessments conducted with TELD-3, the receptive language age of the children with IP-I is average 67.61 months, and while the expressive language age is average 50 months. Children with IP-I constituted the group of children with the third lowest scores in terms of auditory perception performance among IEMs.

30.3.3.2 Incomplete Partition Type II (IP-II)

We evaluated 40 children with IP-II in terms of auditory perception performances and language skills (Figs. 30.4 and 30.8). In the Ling's Sound Test and MAIS evaluation performed before the CI, children with IP-II obtained similar scores to children with NC. After cochlear implantation, auditory perception tests such as Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed. There was no statistically significant difference between children with IP-II and NC in open-set and closed-set tests ($p > 0.05$). In the MAIS test between 1 and 3 years after CI, children with IP-II had a score of 37.98 points while the children with NC had a score of 38.28 points. According to Pattern Perception Test as closed-set results between 1 and 3 years after CI, children with NC had 88.5% and children with IP-II had 87.75%. The Daily Turkish Sentence Recognition Test showed close performance between the two groups (IP-II 48.48% and NC 50.05%). In addition to language skills assessments conducted with TELD-3, the receptive language age of the children with IP-II is 83 months, while the expressive language age is average 78 months. Children with IP-II completed all test items as the receptive language age. Children with IP-II constituted the group of children with the second highest scores in terms of auditory perception performance among IEMs.

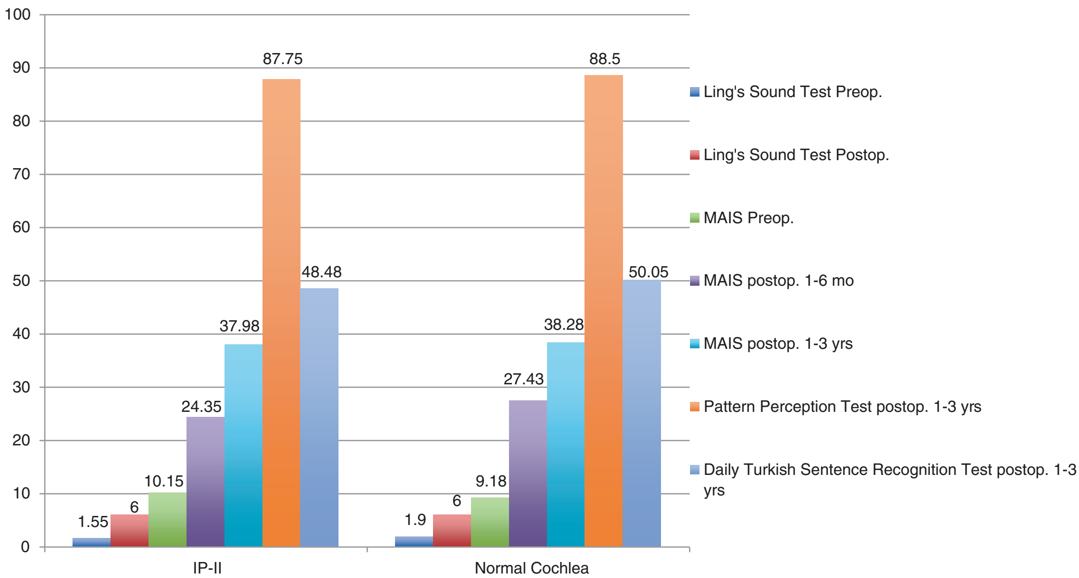

Fig. 30.4 Comparison of children with IP-II and NC in terms of auditory perception performance (IP-II = incomplete partition Type II, NC = Normal cochlea)

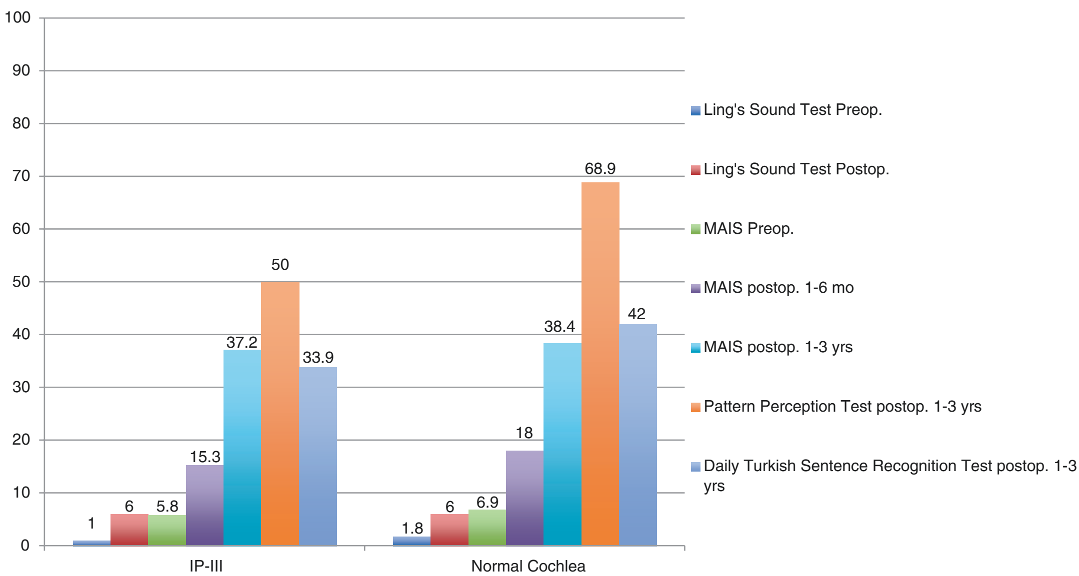

Fig. 30.5 Comparison of children with IP-III and NC in terms of auditory perception performance (IP-III = Incomplete Partition type III, NC = Normal cochlea)

30.3.3.3 Incomplete Partition Type III (IP-III)

We evaluated ten children with IP-III in terms of auditory perception performance and language skills (Figs. 30.5 and 30.8). In the Ling's Sound Test and MAIS evaluation performed before the CI, children with IP-III had scores close to children with NC and there was no statistically significant difference ($p > 0.05$). After cochlear implantation, auditory perception tests such as Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed. There was no statistically significant difference between children with IP-III and NC in open-set and closed-set tests ($p > 0.05$). In the MAIS test between 1 and 3 years after CI, the children with IP-III had a score of 37.2 points, while the children with NC had a score of 38.4 points. According to Pattern Perception Test as closed-set results between 1 and 3 years after CI, children with NC had 68.9% and children with IP-III had 50%. The Daily Turkish Sentence Recognition Test showed close performances between the two groups (IP-III 33.9% and NC 46.13%). Additionally, language skills assessments conducted with TELD-3, the receptive language age of the children with IP-III is average 68.9 months, while the expressive language age is average 54 months. Children with IP-III constituted the group of children with the fourth lowest scores in terms of auditory perception performance among IEMs.

30.3.4 Dilatation of Vestibule

We evaluated three children with dilatation of vestibule in terms of auditory perception performance and language skills (Figs. 30.6 and 30.8). In the Ling's Sound Test and MAIS evaluation performed before the CI, children with dilatation of vestibule had the scores close to children with NC and statistically significant differences were not found ($p > 0.05$). After cochlear implantation, auditory perception tests such as Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed. There was no statistically significant difference between children with dilatation of vestibule and NC in open-set and closed-set tests ($p > 0.05$). In the MAIS test between 1 and 3 years after CI, the children with dilatation of vestibule had a score of 39.33 points, while the children with NC had a score of 40 full points. According to Pattern Perception Test as closed-set results between 1 and 3 years after CI, children with NC had 93.33% and children with dilatation of vestibule had 56.67%. The Daily Turkish Sentence Recognition Test showed no statistically significant difference between the two groups (dilatation of vestibule 13.33% and NC 57%) but we found that children with dilatation of vestibule had difficulty in this open-set test. According to language skills assessments conducted with TELD-3, the receptive language age of the children with dilatation of vestibule is average 71.66 months, while the expressive language age is average 58 months. Children with dilatation of vestibule constituted the group of children with the third highest scores in terms of auditory perception performance among IEMs.

30.3.5 Enlarged Vestibular Aqueduct (EVA)

We evaluated 14 children with EVA in terms of auditory perception performance and language skills (Figs. 30.7 and 30.8). In the Ling's Sound Test and MAIS evaluation performed before the CI, children with EVA obtained scores very close to children with NC and statistically significant differences were not found ($p > 0.05$). After cochlear implantation, auditory perception tests such as Ling's Sound Test, MAIS, Pattern Perception Test, Daily Turkish Sentence Recognition Test were performed. There was no statistically significant difference between children with EVA and NC in open-set and closed-set tests ($p > 0.05$). In the MAIS test between 1 and 3 years after CI, the children with EVA had a score of 35 points while the children with NC had a score of 38.57 points. According to Pattern Perception Test as closed-set results between 1 and 3 years after CI, children with NC had 82.64% and children with EVA had 89.71%. The Daily Turkish Sentence Recognition Test showed close performances between the two groups (EVA 79.64% and NC 87.29%). According to language skills assessments conducted with TELD-3, the receptive language age of the children with EVA is 83 months, while the expressive language age is average 83 months. Children with EVA completed all test items as the receptive and expressive language age. Children with EVA obtained the highest scores in terms of auditory perception performance among IEMs.

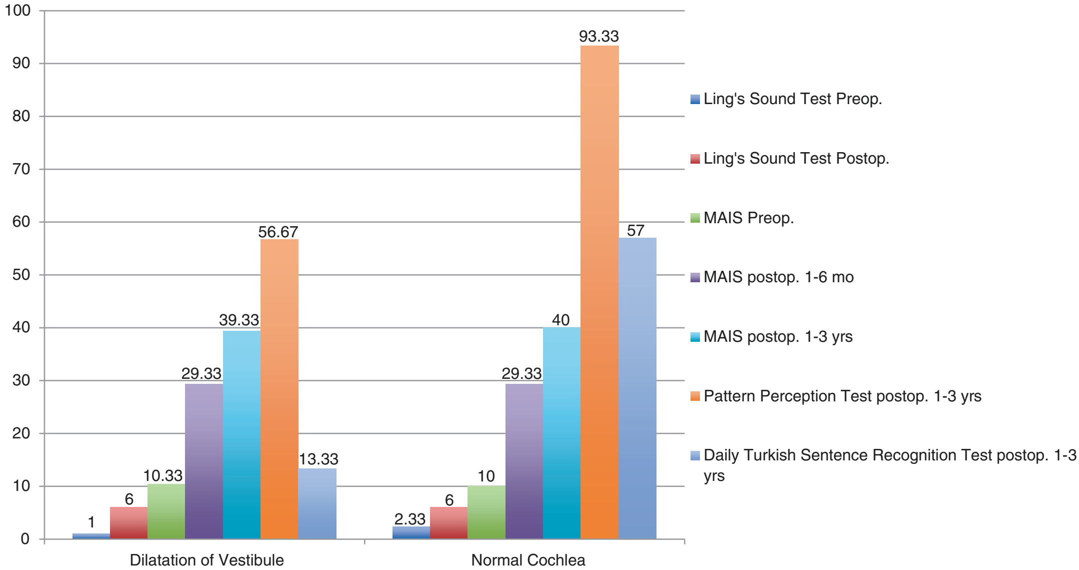

Fig. 30.6 Comparison of children with dilatation of vestibule and NC in terms of auditory perception performance (NC = Normal cochlea)

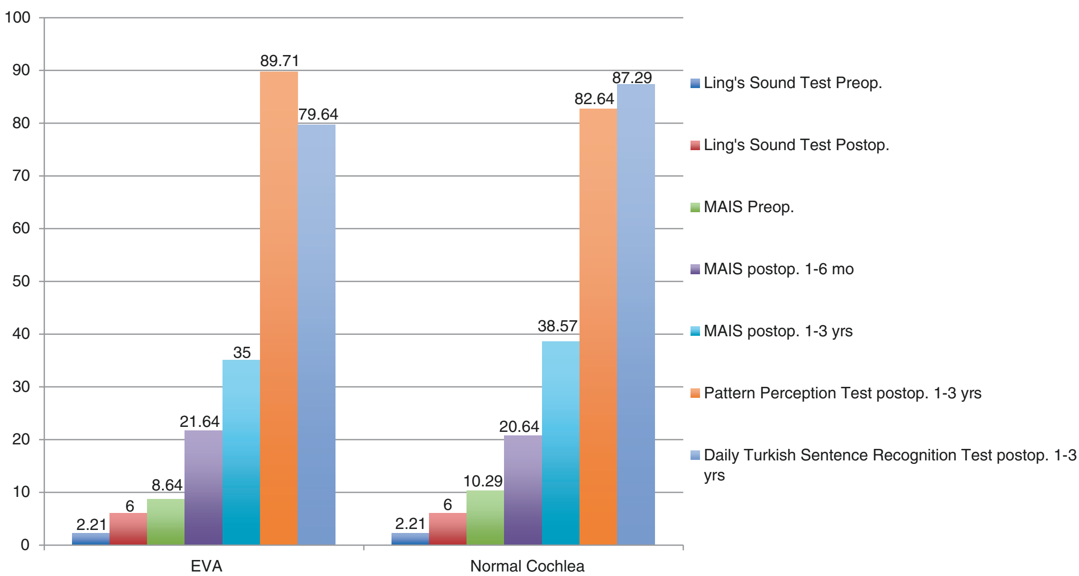

Fig. 30.7 Comparison of children with EVA and NC in terms of auditory perception performance (EVA = Enlarged vestibular aqueduct, NC = Normal cochlea)

Our data showed that children with IEMs might receive considerable benefit from CI. Taking into consideration variation in children with IEMs using CI, it is very difficult to establish outcomes. Nevertheless, with other studies, our results demonstrated that CI is a successful treatment modality in deaf children with and without IEMs. Depending on the subgroup of IEMs, their outcomes also showed variability. Additionally, this variation can also be caused by several other factors, such as age of implantation, preoperative residual and functional hearing, cognitive skills, parental and environmental support.

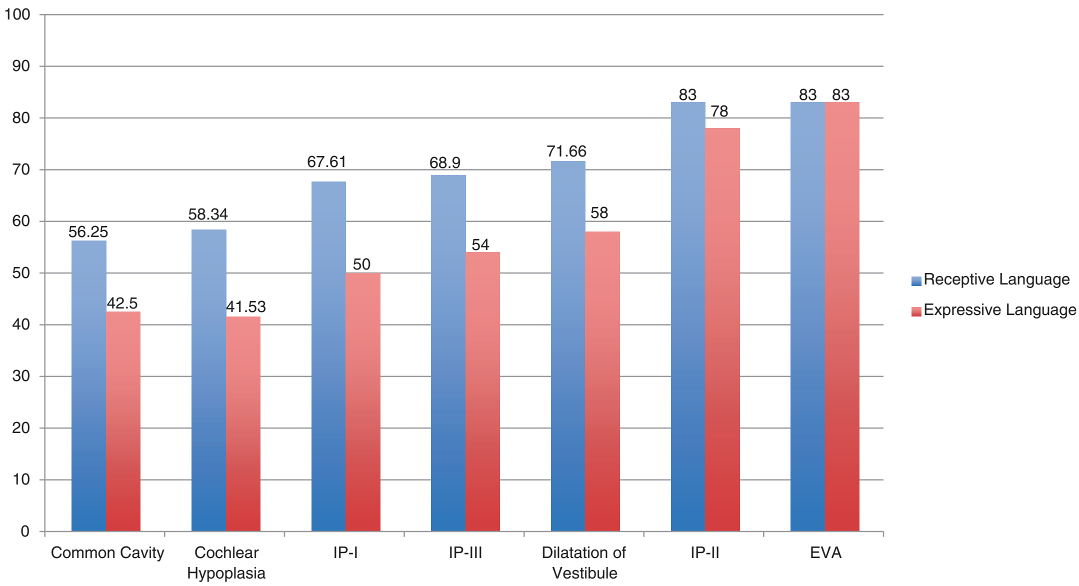

Fig. 30.8 Results of TELD-3 in children with all IEMs

30.4 Hacettepe Experience of Cochlear Implantation in Children with IEMs

Auditory perception performance in speech sounds and closed-set pattern perception assessed with CIAT test battery. All children with or without IEMs detected Ling 6 sounds in various frequencies. However, speech sounds, which are, used in this task only represent frequencies in wide ranges in speech frequency bands. Considering the results of speech perception tasks, children with all kinds of IEMs showed varying degrees of auditory benefit by the end of 3 years of CI experience.

Speech perception and language development performance variations between children with IEMs are summarized below:

30.4.1 Common Cavity (CC)

Children with CC had the lowest scores in terms of auditory perception performance among IEMs. Although they used their CIs regularly in daily basis, it was found that they develop identification and comprehension of environmental sounds 1 year after but not developed this proficiency for speech sounds.

One child with CC who could not perform any speech perception skills had only four active electrodes and had facial stimulation as side effect. He underwent ABI for his contralateral ear after 1 year of cochlear implant experience. He performed significantly better with his contralateral ABI and rejected to use CI.

Functionality of auditory perception performances was mostly evaluated with MAIS in clinical studies. The results indicated an obvious delay in children with CC; however, their performance changes with time. Unfortunately, children with CC are unable to reach the full MAIS score even 1–3 years. It is necessary to follow children with CC more closely and to study in this direction in terms of the functionality of auditory perception performances.

Children with CC showed better performance with regard to a closed-set situation, but their vocabulary was weak; they struggled in attention and memory skills. Auditory training programs should have activities that support attention and memory. Further developmental stages involve thinking and predicting words in sentences using clues in the context to maintain conversation; use of language-based visual clues would be a difficult skill to develop. Therefore, rehabilitation programs should be encouraged to improve these skills [16]. Their pattern perception scores changed due to their chronological age, duration of CI use, and cognitive development.

Unfortunately, open-set sentence recognition is not improved in children with CC between 1 and 3 years.

When the language skills of children with CC were analyzed, these children have a language development of about 4.5 years old. As higher-level language skills have increased, it has been determined that these children have difficulty with comprehension skills. Results indicate that duration of CI is significant than chronological age for both receptive and expressive language development. When a child becomes older, the gap between chronological age and language development scores becomes wider. In the later years, language development tasks will become harder, and catching up with these tasks would be more difficult [17].

30.4.2 Cochlear Hypoplasia (CH)

Children with CH constituted the group of children with the second lowest scores in terms of auditory perception performance among IEMs.

Children with CH used their CI regularly on a daily basis, and they were found to develop identification and comprehension toward environmental sounds and speech sounds after 1 year.

Functionality of auditory perception performances was mostly evaluated with MAIS in researches. The results indicated an obvious delay with CH children; however, their performance changes with time. Unfortunately, children with CH are unable to reach the full MAIS score even 1–3 years, but it has been found that children with CH can use auditory perception more functionally than children with CC. Also, similar results show it is necessary to follow children with CH more closely and to study in this direction in terms of the functionality of auditory perception performances.

Children with CH showed better performance with regard to a closed-set situation, but their vocabulary was weak; they struggled in attention and memory skills. Auditory training programs should have activities that support attention and memory. Further developmental stages involve thinking and predicting words in sentences using clues in the context to maintain conversation; use of language-based visual clues would be a difficult skill to develop. Therefore, rehabilitation programs should be encouraged to improve these skills [12]. Their pattern perception scores changed due to their chronological age, duration of CI use, and cognitive development. It was also found that children with CH could use closed-set pattern perception skills better than children with CC.

Open-set sentence recognition is improved in children with CH between 1 and 3 years. This is a better result compared to children with CC, even though only some children with CH have already open-set sentences recognition.

Analysis of the language skills of children with CH showed that they have a language development of about 4.8 years old. As higher-level language skills have increased, it has been determined that these children have difficulty with comprehension skills. Results indicate that duration of CI use is more significant than chronological age for both receptive and expressive language development. When a child becomes older, the gap between chronological age and language development scores becomes wider. In the later years, language development tasks will become harder, and catching up with these tasks would be difficult [15].

30.4.3 Incomplete Partition Anomalies of The Cochlea

30.4.3.1 Incomplete Partition Type I (IP-I)

Children with IP-I constituted the third lowest scores in terms of auditory perception performance among IEMs. They used their CI regularly on a daily basis, and they were found to develop identification and comprehension toward environmental sounds and speech sounds after 1 year.

The results of MAIS indicated obvious delay children with IP-I; however, their performance changes with time. Unfortunately, children with IP-I are unable to reach the full MAIS score even 1–3 years, but it has been found that children with IP-I can use auditory perception more functionally than children with CC and CH. Also, similar results show it is necessary to follow children with IP-I more intensely and to study in this direction in terms of the functionality of auditory perception performances.

Children with IP-I obtained better performance with regard to a closed-set situation, also their vocabulary was not weak but they struggled in attention and memory skills. Auditory training programs should have activities that support attention and memory. Further developmental stages involve thinking and predicting words in sentences using clues in the context to maintain conversation; use of language-based visual clues would be a difficult skill to develop. Therefore, rehabilitation programs should be encouraged to improve these skills [12]. Their pattern perception scores changed due to their chronological age, duration of CI use, and cognitive development. Also it has been found that IP-I children can use closed-set pattern perception skills better than children with CC and CH.

Open-set sentence recognition is improved in children with IP-I between 1 and 3 years. This is a better result compared to children with CH, even though only some children with IP-I have already open-set sentences recognition.

When analyzed at the language skills of children with IP-I that these children have a language development of about 5.5 years old. As higher-level language skills have increased, it has been determined that these children have difficulty with comprehension skills. Results indicate that duration of CI is significant than chronological age for both receptive and expressive language development. When a child becomes older, the gap between chronological age and language development scores becomes wider. In the later years, language development tasks will become harder, and catching up with these tasks would be difficult [15].

30.4.3.2 Incomplete Partition Type II (IP-II)

Children with IP-II obtained the second highest scores in terms of auditory perception performance among IEMs.

Children with IP-II used their CI regularly on a daily basis, and they were found to develop identification and comprehension toward environmental sounds and speech sounds after 1–3 months.

MAIS results indicated better performance children with IP-II; however, their performance changes with time. Children with IP-II are close to reach the full MAIS score 1–3 years, and it has been found that children with IP-II can use auditory perception more functionally than children with IP-I and IP-III.

Children with IP-II demonstrated better performance with regard to a closed-set situation, also their vocabulary was strong.

Open-set sentence recognition showed improvement in children with IP-II between 1 and 3 years. Most of the children with IP-II can repeat sentences correctly.

When the language skills of children with IP-II were analyzed, their receptive language was found to be same as their chronological age.

30.4.3.3 Incomplete Partition Type III (IP-III)

Children with IP-III constituted the fourth lowest scores in terms of auditory perception performance among IEMs.

Children with IP-III used their CI regularly on a daily basis, and they were found to develop identification and comprehension toward environmental sounds and speech sounds after 1 year.

MAIS results indicated obvious delay children with IP-III; however, their performance changed with time. Children with IP-III are close to reach the full MAIS score 1–3 years, and it has been found that children with IP-III can use auditory perception more functionally than children with IP-I.

Children with IP-III did not show a good performance with regard to a closed-set situation. In addition, their vocabulary was weak but they struggled in attention and memory skills. Auditory training programs should have activities that support attention and memory. Further developmental stages involve thinking and predicting words in sentences using clues in the context to maintain conversation; use of language-based visual clues would be a difficult skill to develop. Therefore, rehabilitation programs should be encouraged to improve these skills [12]. Their pattern perception scores changed due to their chronological age, duration of CI use, and cognitive development.

Open-set sentence recognition has improved in children with IP-III between 1 and 3 years. This is a better result compared to children with IP-I; more children with IP-III have already open-set sentence recognition.

Children with IP-III have a language development of about 5.7 years old. As higher-level language skills have increased, it has been determined that these children have difficulty with comprehension skills. Results indicate that duration of CI is more significant than chronological age for both receptive and expressive language development. When a child becomes older, the gap between chronological age and language development scores becomes wider. In the later years, language development tasks will become harder, and catching up with these tasks would be difficult [15].

30.4.4 Dilatation of Vestibule

Children with dilatation of vestibule constituted the third highest scores in terms of auditory perception performance among IEMs.

Children with dilatation of vestibule used their CI regularly on a daily basis, and they were found to develop identification and comprehension toward environmental sounds and speech sounds after 3–6 months.

MAIS results indicated better performance children with dilatation of vestibule; however, their performance changes with time. Children with dilatation of vestibule are close to reach the full MAIS score 1–3 years.

Their performance with regard to a closed-set situation was not good. In addition their vocabulary was weak but they struggled in attention and memory skills. Auditory training programs should have activities that support attention and memory. Further developmental stages involve thinking and predicting words in sentences using clues in the context to maintain conversation; use of language-based visual clues would be a difficult skill to develop. Therefore, rehabilitation programs should be encouraged to improve these skills [12]. Their pattern perception scores changed due to their chronological age, duration of CI use, and cognitive development.

Open-set sentence recognition is improved in children with dilatation of vestibule between 1 and 3 years.

Children with dilatation of vestibule have a language development of about 6 years old. As higher-level language skills have increased, it has been determined that these children have difficulty with comprehension skills. Results indicate that duration of CI is significant than chronological age for both receptive and expressive language development. When a child becomes older, the gap between chronological age and language development scores becomes wider. In the later years, language development tasks will become harder, and catching up with these tasks would be difficult [15].

30.4.5 Enlarged Vestibular Aqueduct (EVA)

EVA is the only group that Pattern Perception Test and Turkish Daily Sentence test results were better than NC group. SNHL since birth EVA is a congenital anomaly with progressive SNHL. At the beginning their hearing may be normal. The process is progressive. Final outcomes are better than children with NC.

EVA–IP-II difference is modiolar defect in IP-II. In general outcome of IP-II is almost similar to NC. EVA is better than IP-II because there is no modiolar defect in EVA, which is the reason to make the outcome slight worse.

Children with EVA had a higher score than children with NC in the closed-set test. In these children, we can say that the children with EVA perform better than children with NC. It is thought that the reason for this is that some of the children with EVA may have been suffering from hearing loss in the peri- or post-lingual period rather than the prelingual period.

Children with IP-II and EVA scores reached implanted children with NC but children with CC, CH, and IP-I improved slower. Preoperative counseling for the parents is advised in order to explain the possible impact of the diagnosed disabilities on performance and habilitation. Nevertheless, factors influencing the success of implantation are multiple, including a thorough preoperative radiological examination, a well-performed surgery, and an individually tailored postoperative rehabilitation program.

30.5 Summary

Our major goal is to provide meaningful sound information through CI in children with IEMs who have severe to profound hearing loss. Because of the complexity of different subgroups, it is not possible to explain the performance of children with IEMs under a single group. The critical point is that the degree of malformation should be taken into consideration while evaluating functional hearing with implantation. The readiness of the children in auditory perception, language skills cognitive, psychosocial, and similar areas should be evaluated comprehensively.

According to our results, in terms of auditory perception and language skills, the children with IEMs can be arranged from poor to good performance as **CC, CH, IP-I, IP-III, Vestibular Dilatation, IP-II, and EVA**.

Children with CC obtained the lowest scores among children with IEMs. All children in this group make benefit from CI almost after 3 years CI usage for auditory perception. By evaluating these low scores, we have determined that children with CC cannot bring their auditory perception and language skills to the level of children with NC at the end of the third year. Therefore, if we do not see an acceptable progress in these children, our team proposes an ABI on their contralateral ears.

Children with CH and IP-I are the second and third groups that obtained the least benefit from CI, respectively. However, these children showed heterogeneous scores in terms of auditory perception and language development. Although several children with CH and IP-I achieved closer scores to CI children with NC, some others were able to show improved performance after 3 years of CI use. Children with CH and IP-I definitely need bilateral implantation. As we know, children with CH and IP-I can be a candidate for CI or ABI. When CI was applied to one ear, according to their benefit from CI in that ear and cochlear nerve status on the contralateral ear (demonstrated with MRI), these children could also be an ABI candidate on the contralateral side. If there is well-developed cochlear nerve, bilateral CI should be done. In case of CN deficiency, contralateral ABI is advisable. In case of bilateral CH-I, which is the least developed type of CH, contralateral ABI can be proposed.

Children with IP-III and dilatation of vestibule showed poor performance in open-set scores. Consequently, in the earliest period, bilateral CI should be recommended to this group. The risk of gusher in IP-III is 100%; therefore, bilateral CI should be staged in IP-III. In vestibular dilatation bilateral CI can be done simultaneously or staged. IP-III cases do not need an ABI as they have well-developed cochlear nerves.

It was found that the open-set, closed-set, and language scores of children with IP-II and EVA were similar to children with NC. Therefore, children with IP-II and EVA were able to obtain good results with unilateral CI. Bilateral CI is also proposed to these groups to obtain the advantages of bilateral hearing.

Children with IEMs should be involved in intensive rehabilitation and follow-up at regular intervals. The open-set, closed-set, language, and speech production of children in this group should be examined one by one. The alternative surgical method should be considered when there is a failure in these skills.

References

1. Chadha NK, James AL, Gordon KA, Blaser S, Papsin BC. Bilateral cochlear implantation in children with anomalous cochleovestibular anatomy. Arch Otolaryngol Head Neck Surg. 2009;135:903–9.
2. Ha JF, Wood B, Krishnaswamy J, Rajan GP. Incomplete cochlear partition type II variants as an indicator of congenital partial deafness: a first report. Otol Neurotol. 2012;33:957–62.
3. McClay JE, Tandy R, Grundfast K, Choi S, Vezina G, Zalzal G, et al. Major and minor temporal bone abnormalities in children with and without congenital sensorineural hearing loss. Arch Otolaryngol Head Neck Surg. 2002;128:664–71.
4. Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int. 2010;11:4–41.
5. Sennaroğlu L, Bajin MD. Classification and current management of inner ear malformations. Balkan Med J. 2017;34(5):397.
6. Tucci DT, Telian SA, Zimmerman-Phillips S, Zwolan TA, Kileny PR. Cochlear implantation in patients with cochlear malformations. Arch Otolaryngol Head Neck Surg. 1995;121:833–8.
7. Luntz M, Balkany T, Hodges AV, Telischi FF. Cochlear implants in children with congenital inner ear malformation. Arch Otolaryngol Head Neck Surg. 1997;123:974–7.
8. Eisenman DJ, Ashbaugh C, Zwolan TA, Arts HA, Telian SA. Implantation of the malformed cochlea. Otol Neurotol. 2001;22(6):834–41.
9. Buchman CA, Copeland BJ, Yu KK, Brown CJ, Carrasco VN, Pillsbury HC III. Cochlear implantation in children with congenital inner ear malformations. Laryngoscope. 2004;114(2):309–16.
10. Papsin BC. Cochlear implantation in children with anomalous cochleovestibular anatomy. Laryngoscope. 2005;115(S106):1–26.
11. Sainz M, Garcia-Valdecasas J, Fernandez E, Pascual MT, Roda O. Auditory maturity and hearing performance in inner ear malformations: a histological and electrical stimulation approach. Eur Arch Otorhinolaryngol. 2012;269(6):1583–7.
12. Isaiah A, Lee D, Lenes-Voit F, Sweeney M, Kutz W, Isaacson B, Lee KH. Clinical outcomes following cochlear implantation in children with inner ear anomalies. Int J Pediatr Otorhinolaryngol. 2017;93:1–6.
13. Ozkan HB, Cicek Cinar B, Yucel E, Sennaroglu G, Sennaroglu L. Audiological performance in children with inner ear malformations before and after cochlear implantation: a cohort study of 274 patients. Clin Otolaryngol. 2021;46(1):154–60.
14. Yücel E, Sennaroglu G. Çocuklar İçin İşitsel Algı Testi. Advanced Bionics. 2011.
15. Topbaş S, Güven OS. Reliability and validity results of the adaptation of TELD-3 for Turkish speaking children: implications for language impairments. In: Oral presentation, 12th Congress of the International Clinical Phonetics and Linguistics Association, Istanbul, Turkey;2008.
16. Yücel E, Aslan F, Özkan HB, Sennaroglu L. Recent rehabilitation experience with pediatric ABI users. J Int Adv Otol. 2015;11(2):110.
17. Merkus P, Di Lella F, Di Trapani G, Pasanisi E, Beltrame MA, Zanetti D, et al. Indications and contraindications of auditory brainstem implants: systematic review and illustrative cases. Eur Arch Otorhinolaryngol. 2014;271:3–13.