European Archives of Oto-Rhino-Laryngology (2019) 276:2763–2768
https://doi.org/10.1007/s00405-019-05534-1

OTOLOGY

Cochlear implantation in incomplete partition type I

Ali Eftekharian[1] · Kourosh Eftekharian[2] · Nooshin Mokari[1] · Maryam Fazel[1]

Received: 29 March 2019 / Accepted: 25 June 2019 / Published online: 29 June 2019

Abstract

Objectives To share our experience in cochlear implanted patients with incomplete partition type I, to compare it with the literature results and to disclose difficulties facing cochlear implant teams dealing with these patients.

Materials and methods Clinical records of 1089 cochlear implant procedures in a cochlear implant center were reviewed and data of patients who had incomplete partition type I were enrolled in this study. Their auditory and speech performances were evaluated 3 years after the implantation.

Results Eighteen cases (1.65%) had incomplete partition type I. Cerebrospinal fluid gusher was encountered during opening the cochlea in 15 patients (83.3%). There were no cases of persistent CSF leak or postoperative meningitis. In 61.1% of patients, some additional anomalies were found during the operation. Although in 55.6% of cases no electrically evoked compound action potential was detected even in long-term follow-up, all patients had satisfactory auditory and speech outcome.

Conclusion Cochlear implantation is a relatively safe and effective treatment for patients who have incomplete partition type I, even if the procedure may be somehow challenging.

Keywords Cochlear implantation · Incomplete partition · Inner ear malformations · Gusher

Introduction

Inner ear malformations are present in about 20% of patients with congenital sensorineural hearing loss [1].

Incomplete partition (IP) type I is defined by Sennaroglu [2, 3] as a cystic cochleovestibular malformation in which the cochlea lacks the entire modiolus and interscalar septa and is accompanied by a large dilated vestibule. Due to the defective development of the cochlear aperture and absence of the modiolus, there is a defect between the internal auditory canal and the cochlea.

Although malformation of the inner ear is no longer considered as absolute contraindication for cochlear implantation, abnormal anatomy of the cochlea and facial nerve still raise concerns regarding safe and effective electrode insertion [4].

✉ Kourosh Eftekharian
kourosheftkharian@gmail.com

[1] Cochlear Implant Center, Loghman Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran

[2] School of Medicine, Tehran University of Medical Sciences, Enghlab Street, Tehran 14155-6559, Iran

Hearing outcome and speech perception improvement following cochlear implantation in patient with inner ear malformations seems to be as not good as the normal population, but there is a lack of adequate data for patients with IP type I malformations in the literature [5].

Cochlear implantation teams who work on these patients must be aware of the difficulties that may rise not also during the surgery but also afterwards.

The aim of this study is to share our experience, to compare it with the literature results and to disclose difficulties facing cochlear implant teams dealing with these patients.

Materials and methods

Between July 2008 and December 2018, 1089 cochlear implant procedures have been done in our department. Their clinical records were reviewed and the data of patients who had incomplete partition type I were enrolled in this study.

Their demographic features as well as surgical findings and their follow-up issues were evaluated.

Auditory and speech perception, speech intelligibility and speech production skills of the implanted children

before implantation and at the third year of implantation were evaluated and compared with three tests: categories of auditory performance (CAP) test [6], speech intelligibility rating (SIR) test [7], and meaningful use of speech scale (MUSS) test [8].

Statistical analysis of the data was conducted with SPSS 24.0.0 (SPSS Inc.S, Chicago, IL, USA). Comparison was done using Wilcoxon signed ranks test and "*P*" value < 0.05 was considered significant.

Results

Out of 1089 cochlear implantations, 18 cases (1.65%) had incomplete partition type I: seven males (38.9%) and eleven females (61.1%). Their data are shown in Table 1. All patients had unknown congenital hearing loss except case number 5 who had a history of maternal Cytomegalovirus infection. Eight patients (44.4%) were born from parents with consanguineous marriage. The preferred approach of opening cochlea and electrode insertion in our department is via round window that was possible in 12 patients (66.7%). Cochleostomy is chosen just when anatomically adequate view of the round window does not exist. In four patients (cases number 8, 10, 15 and 17), no round window was found at all, and cochleostomy was done on the promontory approximately 2 mm below the oval window. Cerebrospinal fluid (CSF) gusher was encountered during opening the cochlea in 15 patients (83.3%). Fortunately, that was controlled during the surgery in all cases and no case of persistent CSF leak or postoperative meningitis was encountered. In seven patients (38.9%), no additional anomaly was found during the surgery while in eleven patients (61.1%) some were found during the operation (Table 1).

Full insertion of the electrodes was achieved for all the patients. Thirteen patients (72.2%) had no electrically evoked compound action potential (e-ECAP) during surgery which in three cases it was recovered during follow-up. Five patients (27.8%) experienced some facial nerve stimulation at the device switch on. All were revealed by their device program modifications.

Results of auditory and speech performance tests; CAP, SIR and MUSS tests are shown in Table 2.

These patients had statistically significant progress in face of speech perception, speech intelligibility and speech production in all three tests.

Patients which their e-ECAP was not measurable even in their last follow-up have also statistically significant progress in CAP, SIR and MUSS tests. (Table 3).

To date, no complication was seen in our cases.

Discussion

Sennaroglu and Saatci were the first who classified incomplete partition anomalies of the cochlea [2]. Cochlear implantation in incomplete partition type I patients is not widely reported in the literature [5, 9].

To our knowledge, the largest reported series of cochlear implantation in this malformation is by Suk et al. who reported on 25 surgeries in 2015 [10]. Surgery may be challenging in patients with IP type I. One important issue concerning cochlear implantation in inner ear malformation, and particularly in IP type I, is the higher incidence of CSF gusher [9]. The incidence of CSF gusher in this malformation is varied in different reported series. Berrettini et al. [9] had no CSF gusher in their four cases. Sennaroglu [3] reported nine cases out of his 23 patients (39%). The reported incidence by Suk et al. [10] and Kontorinis et al. [11] is 56% and 63.6%, respectively. Catli et al. [12] had CSF gusher in all their five cases (100%). Farhood et al. in a review of literature found its incidence to be 45.9% of the cases [5]. The incidence of CSF gusher in our cases was 83.3%. As there is a defect between the internal auditory canal and the cochlea in this anomaly, it is not surprising to have this high incidence but the reason behind this widely various reports is not clear for us. In all our patients, a watertight seal around the cochlear implant array was done with a customized technique using four to five pieces of fresh muscle and no other attempt was necessary. There was no case of persistent CSF leak or meningitis after the operations. Our management technique for controlling CSF gusher was previously described [13].

One of the most important factors determining the final position of the facial canal is the development of the inner ear. Certain abnormalities of the inner ear may cause anomalies in the location of the facial canal, making its course even more complicated. Severe facial nerve anomalies preventing facial recess access are possible in IP type I [3]. In our patients, no severe facial nerve anomaly was found and just in three cases we had anterior displacement of the mastoid segment of the nerve (Table1). Nevertheless, we agree with Sennaroglu [3] and Berrettini [9] that facial nerve monitoring should be used in all cases with inner ear malformations undergoing cochlear implantation.

In five patients (27.8%), some facial nerve stimulation was detected at first switch on of the device. As it is apparent in Table 1, in the majority of them no facial nerve anomaly was found during their operation. The reported incidence by Farhood [5] is 23.7% while in the study of Suk et al. [10] it is 60% and they stated that it could be due to proximity of the facial nerve to the electrode array, aberrant course or dehiscence of the facial nerve.

Table 1 Demographic data

No.	Patient	Sex	Age (years)	Cons	Op side	Type of C.O	CSF Gusher	Other surgical findings	Intra-op e-ECAP	Facial nerve stimulation at switch-on	Device model
1	M. B	F	8	+	R	C	+	Stapes footplate defect with CSF leak	−	−	CI24R(ST)
2	S. C	F	9	−	R	R.W	+	No LSCC	−	−	HiRes90k
3	F. M	F	14	−	L	R.W	+	−	−	+	CI24RE
4	H. K	M	5	+	R	C	+	−	−	−	HiRes90k
5	M. M	M	6	−	L	R.W	+	No LSCC	−	−	HiRes90k
6	F. I	F	6	+	R	R.W	+	No LSCC stapes was fixed	−	+	HiRes90k
7	A. M	F	2	−	R	R.W	+	−	−	−	HiRes90k
8	A. Y	M	4	+	R	C	+	No round window	+	−	HiRes90k
9	M. S	M	9	+	L	R.W	+	−	+	+	CI24RE
10	S. J	F	6	+	L	C	+	No LSCC, no round window	−	−	HiRes90k
11	Z. F	F	13	+	L	R.W	+	−	+	−	HiRes90k
12	F. P	F	15	−	L	R.W	+	Stapes was fixed	−	−	HiRes90k
13	F. S	F	2	−	L	R.W	−	Facial nerve anterior displacement in mastoid segment	−	−	HiRes90k
14	F. H	F	13	−	R	R.W	+	−	−	+	HiRes90k
15	P. A	F	6	−	R	C	+	No round window	−		HiRes90k
16	M. D	M	1.5	−	L	R.W	−	Facial nerve anterior displacement in mastoid segment	−	+	HiRes90k
17	A. B	M	3	+	L	C	+	No round window, Facial nerve anterior displacement in mastoid segment	+	−	HiRes90k
18	M.M	M	4.5	−	R	R.W	−	−	−	−	HiRes90k

F female, *M* male, *Cons* born from parents with consanguineous marriage, *Op side*, operation's side, *C.O.* Cochlea's opening, *C* Cochleostomy, *R.W.* round window, *CSF* cerebrospinal fluid, *LSCC* lateral semicircular canal, *Intra-op e-ECAP* intraoperative electrically evoked compound action potential

Table 2 Auditory and speech performances

No	Patient	Sex	Age (years)	Type of C.O	e-ECAP		PTA	CAP			SIR			MUSS			Device model	Follow-up (months)
					IO	L		B	T	*P*	B	T	*P*	B	T	*P*		
1	M. B	F	8	C	−	−	20	2	4	< 0.001*	2	3	< 0.001*	10	30	< 0.001*	CI24R(ST)	108
2	S. C	F	9	R.W	−	+	35	2			2			4			HiRes90k	Lost follow up
3	F. M	F	14	R.W	−	+	20	2	6		3	4		25	31		CI24RE	84
4	H. K	M	5	C	−	+	20	3	6		2	4		4	25		HiRes90k	75
5	M. M	M	6	R.W	−	−	40	1	5		1	2		4	17		HiRes90k	74
6	F. I	F	6	R.W	−	−	25	3	7		1	4		3	36		HiRes90k	72
7	A. M	F	2	R.W	−	−	20	4	6		1	4		4	35		HiRes90k	68
8	A.Y	M	4	C	+	+	25	5	7		2	4		26	38		HiRes90k	66
9	M. S	M	9	R.W	+	+	30	5	7		1	4		3	25		CI24RE	66
10	S. J	F	6	C	−	−	30	2	6		1	4		2	32		HiRes90k	63
11	Z. F	F	13	R.W	+	+	30	6	7		4	5		33	40		HiRes90k	62
12	F. P	F	15	R.W	−	−	55	2	5		1	3		3	21		HiRes90k	60
13	F. S	F	2	R.W	−	−	25	1	7		1	4		1	29		HiRes90k	59
14	F. H	F	13	R.W	−	−	40	2	5		1	3		5	23		HiRes90k	56
15	P. A	F	6	C	−	−	30	0	6		1	3		0	16		HiRes90k	52
16	M. D	M	1.5	R.W	−	−	40	0	6		1	2		0	11		HiRes90k	52
17	A. B	M	3	C	+	+	20	1	6		1	2		2	17		HiRes90k	40
18	M.M	M	4.5	R.W	−	−	–	1			1			3			HiRes90k	Recently implanted

F female, *M* male, *C.O.* Cochlea's opening, *C* Cochleostomy, *R.W.* round window, *e-ECAP* electrically evoked compound action potential, *IO* intraoperative, *L* last measurement, *PTA* pure-tone average at the last measurement: average of the thresholds at 0.5, 1, 2, and 3 kHz, *CAP* categories of auditory performance test score, *B* before the operation, *T* 3 years after the implantation, *SIR* speech intelligibility rating test score, *MUSS* meaningful use of speech scale test score, *PP* value

*Statistically significant

Table 3 Auditory and speech performances of patients with no detectable electrically evoked compound action potential

No	Patient	Age (years)	e-ECAP		PTA	CAP			SIR			Muss			Device model	Follow-up (months)
			IO	L		B	T	*P*	B	T	*P*	B	T	*P*		
1	M.B	8	No	No	20	2	4	0.005*	2	3	0.005*	10	30	0.005*	CI24R(ST)	108
5	M.M	6	No	No	40	1	5		1	2		4	17		HiRes90k	74
6	F.I	6	No	No	25	3	7		1	4		3	36		HiRes90k	72
7	A.M	2	No	No	20	4	6		1	4		4	35		HiRes90k	68
10	S.J	6	No	No	30	2	6		1	4		2	32		HiRes90k	63
12	F.P	15	No	No	55	2	5		1	3		3	21		HiRes90k	60
13	F. S	2	No	No	25	1	7		1	4		1	29		HiRes90k	59
14	F.H	13	No	No	40	2	5		1	3		5	23		HiRes90k	56
15	P. A	6	No	No	30	0	6		1	3		0	16		HiRes90k	52
16	M. D	1.5	No	No	40	0	6		1	2		0	11		HiRes90k	52

e-ECAP electrically evoked compound action potential, *IO* intraoperative, *L* last measurement, *PTA* pure-tone average at the last measurement: average of the thresholds at 0.5, 1, 2, and 3 kHz, *CAP* categories of auditory performance test score, *B* before the operation, *T* 3 years after the implantation, *SIR* speech intelligibility rating test score, *MUSS* meaningful use of speech scale test score, *PP* value

*Statistically significant

It was shown in the literature that program modifications of the device may resolve these facial nerve stimulations [5, 10, 14] which was also true in all our cases.

According to Berrettini [10], hearing results in these patients are variable, but in many cases satisfactory. Our patients had statistically significant progress in speech perception, speech intelligibility and speech production (Table 2). Majority of them reached CAP score of 6 or 7. It means that most of them have the ability to communicate without lip reading and 31.3% of them can also use telephone.

Intra-operative e-ECAP was not measurable in 77.8% of our patients. As is it is apparent in Table 3 even for those which their e-ECAP remained constantly unmeasurable, CAP, SIR and MISS tests shows statistically significant progress. Cochlear implant teams should not be disappointed, not even during intraoperative e-ECAP evaluation when facing no results, but also thereafter.

Minami et al. [15] emphasized that e-ECAP recordings depend largely on spinal ganglion cells, which are very often defective in modiolus deficiency-type malformed cochlea, such as in IP type I, and also excessive stimulus artifacts preclude the successful acquisition of e-ECAP in these cases. In these patients, they suggested the use of electrically evoked auditory brainstem responses instead, which could be recorded even in the malformation cases that lacked e-ECAP responses.

Fortunately, all of our patients have satisfactory hearing after implantation and as Kontorinis [11] stated, most of the cochlear implant recipients with IP type I achieved open speech perception, showing that even in this severe cochlear malformation, cochlear implantation must be considered.

We did not compare these patients with cochlear implanted ones who have normal inner ear; it was not the goal of our study. It is written in the literature that hearing and speech performances of these patients may not be as good as normal inner ear ones and their progress may be more slowly. We would like to show with our large series, the difficulties facing cochlear implantation teams in handling these patients—which are often resolvable—and also long-term hearing and speech benefit of cochlear implantation in this inner ear malformation.

Conclusions

Cochlear implantation is a relatively safe and effective treatment for patients who have IP type I, even if the procedure may be challenging for CI teams. There is a high risk of CSF gusher occurrence and surgeons should be ready to manage it.

Funding The authors do not have any financial or material support for this work.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Jackler RK, Luxford WM, House WF (1987) Congenital malformations of the inner ear: a classification based on embryogenesis. Laryngoscope 97:2–14
2. Sennaroglu L, Saatci I (2002) A new classification for cochleovestibular malformations. Laryngoscope 112:2230–2241

3. Sennaroglu L (2010) Cochlear implantation in inner ear malformations—a review article. Cochlear Implants Int 11:4–41
4. Luntz M, Balkany T, Telischi FF, Hodges AV (1997) Surgical techniques for cochlear implantation of the malformed inner ear. Am J Otol 18(6 Suppl):s66
5. Farhood Z, Nguyen SA, Miller SC, Holcomb MA, Meyer TA, Rizk HG (2017) Cochlear implantation in inner ear malformations: systematic review of speech perception outcomes and intraoperative findings. Otolaryngol Head Neck Surg 156:783–793
6. Archbold S, Lutman ME, Marshall DH (1995) Categories of auditory performance. Ann Otol Rhinol Laryngol Suppl 166:312–314
7. Allen C, Nikolopoulos TP, Dyar D, O'Donoghue GM (2001) Reliability of a rating scale for measuring speech intelligibility after pediatric cochlear implantation. Otol Neurotol 22:631–633
8. Robbins AM, Osberger MJ (1992) Meaningful use of speech scale. Indiana University School of Medicine, Indianapolis
9. Berrettini S, Forli F, De Vito A, Bruschini L, Quaranta N (2013) Cochlear implant in incomplete partition type I. Acta Otorhinolaryngol Ital 33:56–62
10. Suk Y, Lee JH, Lee KS (2015) Surgical outcomes after cochlear implantation in children with incomplete partition type I: comparison with deaf children with a normal inner ear structure. Otol Neurotol 36:e11–17
11. Kontorinis G, Goetz F, Giourgas A, Lenarz T, Lanfermann H, Giesemann AM (2012) Radiological diagnosis of incomplete partition type I versus type II: significance for cochlear implantation. Eur Radiol 22:525–532
12. Catli T, Uckan B, Olgun L (2015) Speech and language development after cochlear implantation in children with bony labyrinth malformations: long-term results. Eur Arch Otorhinolaryngol 272:3131–3136
13. Eftekharian A, Amizadeh M (2014) Cerebrospinal fluid gusher in cochlear implantation. Cochlear Implants Int 15:179–184
14. Kamogashira T, Akamatsu Y, Kashio A, Ogata E, Karino S, Kakigi A, Iwasaki S, Yamasoba T (2016) Development of auditory skills after cochlear implantation in children with inner ear malformations. Acta Otolaryngol 136:78–82
15. Minami SB, Takegoshi H, Shinjo Y, Enomoto C, Kaga K (2015) Usefulness of measuring electrically evoked auditory brainstem responses in children with inner ear malformations during cochlear implantation. Acta Otolaryngol 135:1007–10150