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Cochlear Implantation in Far Advanced Otosclerosis: A Systematic Review and Meta-Analysis

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Objective: To evaluate speech outcomes and facial nerve stimulation (FNS) rates in patients with far advanced otosclerosis (FAO) after cochlear implantation.

Methods: A systematic review was performed using standardized methodology of Medline, EMBASE, PubMed, Cochrane, and Web of Science databases. Studies were included if adults with FAO underwent cochlear implantation. Exclusion criteria included concurrent otologic history (e.g., Meniere's disease, superior canal dehiscence), non-English-speaking implant users, case reports, abstracts, and letters/commentaries. Bias was assessed using the Newcastle-Ottawa Scale for cohort studies and the National Institute of Health Scale for case series. The primary outcome measure was speech discrimination and the secondary outcomes were rates of partial insertion and FNS.

Results: Twenty-seven studies evaluated cochlear implantation in FAO. Due to the heterogeneity of testing methods, statistical pooling of speech discrimination was not feasible, but qualitative synthesis indicated a positive effect of implantation. Pooled rates of FNS were 18% (95% confidence interval, CI 12%–27%) and the rate of partial insertion was 10% (95% CI 7%–15%).

Conclusion: Cochlear implantation in FAO demonstrates significant gains in speech discrimination scores with higher rates of FNS and partial insertion.

Key Words: cochlear implant, facial nerve stimulation, far advanced otosclerosis, partial insertion, speech perception.

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INTRODUCTION

Otosclerosis is a disease characterized by progressive dysplasia of the otic capsule with bony resorption, vascular proliferation, and sclerotic new bone formation.[1] Although initially presenting as a conductive hearing loss from stapes footplate fixation, the disease can advance to involve retrofenestral structures, causing progressive sensorineural hearing loss (SNHL).[2] In addition, a less common but distinct disease phenotype of cochlear otosclerosis may be initially present with SNHL.[3] In 1960, House and Sheehy[4] were the first to utilize the terminology of far advanced otosclerosis (FAO) for patients whose air conduction threshold was no better than 85 dB and whose bone conduction was non-measurable with standard testing. Although the common initial approach for these patients is stapes surgery with a hearing aid, there is a suggestion from the literature that these measures are sometimes unsatisfactory for advanced disease.[5,6]

Intuitively, the severe-to-profound SNHL of FAO patients would lend itself to the benefits of cochlear implantation (CI). Historically, the application of CI to otosclerosis was initially met with skepticism, with the diagnosis of FAO being recommended as a contraindication to implant candidacy.[7,8] FAO poses a unique challenge to the implant surgeon and patient. Intraoperatively, distorted bony anatomy from ossification may necessitate extra drilling and trauma to the cochlea, cochlear demineralization and obliteration may result in the incomplete insertion of the electrode array.[9] Postoperative complications can develop as a result of the altered current distribution of remodeled bone, leading to higher rates of non-auditory stimulation (NAS), such as facial nerve stimulation (FNS).[10–14] Despite these factors, a growing body of evidence supports the notion that CI is safe and beneficial in FAO.[5,15–17]

Although multiple studies have examined the merits of CI for FAO, systematic reviews and meta-analyses have been limited to studies comparing CI to stapes surgery.[18,19] This study aims to qualitatively and quantitatively analyze speech perception outcomes and pool complication rates for CI in FAO. The authors' hypothesis is that CI provides comparable hearing outcomes with marginally higher postoperative complication rates.

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METHODS

A systematic review and meta-analysis were conducted in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines (Appendix A).

Search Strategy

A database search was performed on Medline, EMBASE, PubMed, Cochrane, and Web of Science for relevant peer-reviewed studies from their respective date of inception. A comprehensive search strategy was compiled to represent the intersection of "otosclerosis," "cochlear implantation," and "speech perception" along with their key synonyms (Appendix B). Search terms were selected to create broad criteria with search strings combined using Boolean terms. The search strategy was conducted on the April 17, 2022. Search results were exported to the EndNote citation manager and duplicates were removed.

Study Selection

Studies were assessed for inclusion based on the inclusion criteria (Appendix C). Only adult patients who underwent CI for FAO were included. The original House and Sheehy definition of FAO was applied. Exclusion criteria included concurrent otologic history (e.g., Meniere's disease, superior canal dehiscence), alternate causes of deafness, non-English language speech testing, and pediatric subgroups (<18 years of age). Case reports and studies with mixed adult and pediatric datasets, whereby the data for adult patients could not be isolated, were further excluded. In studies with mixed etiology hearing loss, articles were included if otosclerosis data was able to be isolated from other etiologies. All article screening was performed by two authors (M.K. and K.V.) on Microsoft Excel. Disagreements between the two authors were resolved by input from the senior authors (N.J. and N.P.). Full-text manuscripts were then retrieved and reviewed by all authors to determine final eligibility for inclusion. The reference lists of included studies were screened for any additional articles that might meet the inclusion criteria.

Quality Assessment

All included articles were graded as per the Oxford Centre for Evidence-Based Medicine 2011 Levels of Evidence[20] Risk of bias for each article was assessed using the Newcastle-Ottawa Scale (NOS) for cohort and case-controlled studies and the NIH quality assessment tool for case series.[21,22] Both scales entailed a checklist evaluating case selection, confounders, and comparability and outcome reporting.

Data Extraction

Information obtained regarding each article included: the study title, authorship, publication year, study design, and total numbers. In subjects with FAO, the following patient characteristics were collected: gender, age at implantation, implant type, years of deafness, and previous stapedectomy. Outcome data collected included:

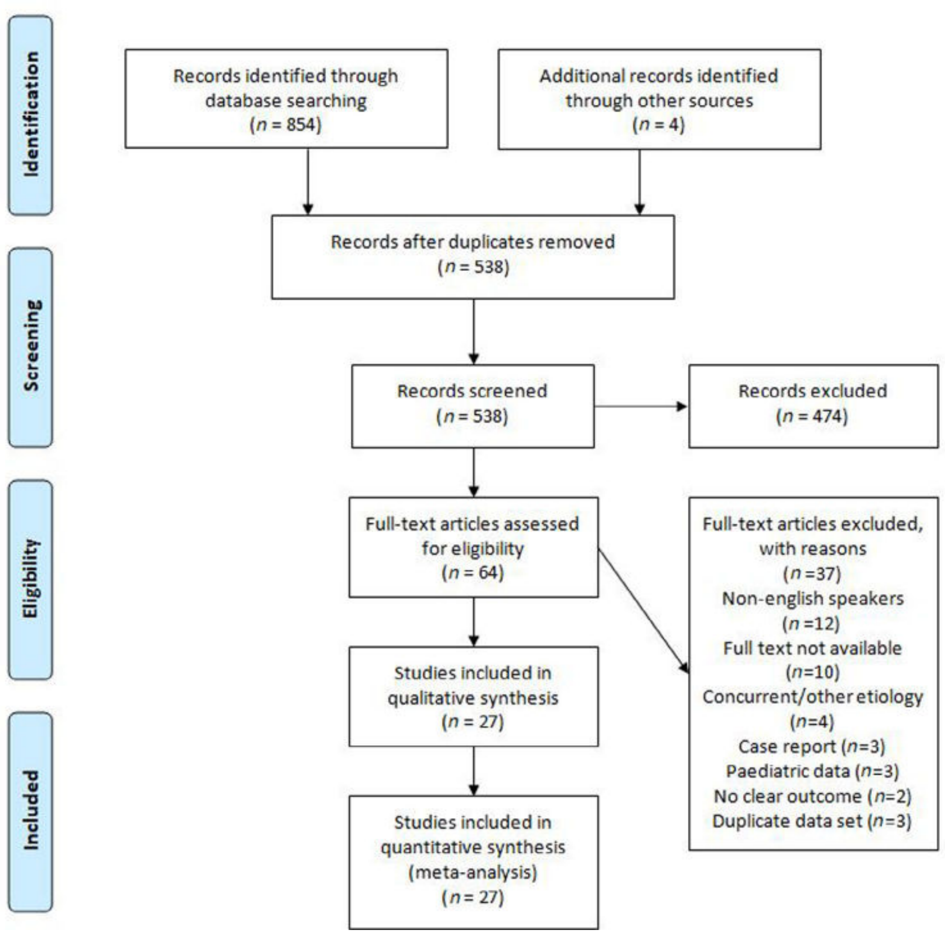

Fig. 1. Preferred Reporting Items for Systematic Reviews and Meta-Analyses flow diagram. [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]

TABLE I.
Newcastle-Ottawa Scale Bias Assessment for Case Controlled Studies.

	Selection				Confounder	Outcome			
Study	Representative of the Exposed Cohort	Selection of External Control	Ascertainment of Exposure	Outcome of Interest Not Present at Start of the Study	Comparability of Cohorts	Assessment of Outcome	Follow Up Long Enough for Outcomes to Occur	Adequacy of Follow-up of Cohorts	Total
Baijin 2020	*	0	*	0	*	*	*	*	6/8
Berrettini 2004	*	*	*	*	*	*	*	*	8/8
Bredberg 2003	*	0	*	0	0	*	*	0	4/8
Calmels 2007	*	*	*	*	0	*	0	*	6/8
Castillo 2014	*	*	*	*	*	*	*	*	8/8
Marshall 2005	*	*	*	0	*	*	*	*	8/8
Muckle 1994	*	*	*	0	0	*	*	*	6/8
Rotteveel 2004	*	0	*	0	0	0	0	*	3/8
Rotteveel 2010	*	0	*	0	0	*	*	*	5/8
Ruckenstein 2001	*	0	*	*	0	*	0	*	5/8
Sainz 2009	*	*	*	*	*	*	*	*	8/8
Semaan 2012	*	*	*	*	*	*	*	*	8/8
Silveira 2017	*	*	*	0	*	*	*	0	6/8
Tokat 2022	*	*	*	0	*	*	*	*	7/8
Zaghis 2003	*	*	*	*	*	*	*	*	8/8

Note: * Signifies the study possesses the quality in the column heading above. "0" signifies that the study does not possess this quality.

preoperative and postoperative PTA and speech perception scores, incidence of FNS, electrodes switched off due to NAS, incidence of operative complications, and incidence of other non-NAS complications. Image J software (Image J software, National Institutes of Health, United States) was utilized to extract data present in published figures when it was not obtainable from the manuscript or authors. This method has been described previously.[23]

Meta-Analysis

Weighted pooled prevalence for complication rates was conducted using R version 4.1.1 (R Core Team, Vienna, Austria) assuming a random-effects model with inverse variance. Heterogeneity for all outcome analyses was measured using the I^2 statistic. Forest plots were created for comparison of rates amongst studies.

RESULTS

Identification of Eligible Studies

The search generated 854 unique results, of which 790 were excluded after initial screening based on the inclusion and exclusion criteria. Thirty-seven full-text articles were excluded after full-text review, leaving

TABLE II.
National Institute of Health Quality Assessment Tool for Case Series.

Study	Objective Clearly Stated	Study Population Well-Described	Consecutive Cases	Comparable Subjects	Intervention Clearly Described	Outcome Measure Defined and Consistent	Adequate Follow-up	Statistical Method Well-Described	Results Well-Described	Total
Balkany 1988	0	*	*	*	*	*	0	0	*	6/9
Bird 1999	*	*	0	*	*	*	*	0	*	7/9
Burmeister 2017	*	*	*	*	*	0	0	0	*	6/9
Fayad 1990	*	*	*	*	*	0	0	0	*	6/9
Flook 2011	*	*	0	*	0	*	*	0	*	6/9
Hodges 1999	*	*	*	*	*	*	*	*	*	9/9
Kabbara 2015	*	*	*	*	*	*	*	0	*	8/9
Matterson 2007	*	*	*	*	0	*	*	*	*	8/9
Mosnier 2007	*	*	*	*	0	0	*	*	*	7/9
Roland 2008	*	*	*	*	*	0	*	*	*	8/9
Vashishth 2017	*	*	*	*	*	*	*	0	*	8/9
West 2017	*	*	*	*	*	0	0	0	*	6/9

Note: * Signifies the study possesses the quality in the column heading above. "0" signifies that the study does not possess this quality.

TABLE III.
Study Characteristics.

First Author	Year	Journal	Country	Design	LOE	Study Purpose
Baijin	2020	*Turk Arch of Otorhinolaryngol*	Turkey	RC	3	Compare hearing outcomes between stapes surgery and cochlear implant in FAO
Balkany	1988	*Laryngoscope*	USA	CS	4	Examine the use of long multichannel electrodes in partially ossified cochleae in comparison to non-ossified cochleae
Berrettini	2004	*J Otolaryngol*.	Italy	RC	3	Compare hearing outcomes between stapes surgery and CI in FAO
Bird	1999	*Ann Otol Rhinol Laryngol*	USA	CS	4	Examining the CLARION CI in cochlear ossification with intraoperative findings and hearing outcomes
Bredberg	2003	*Cochlear Implants Int*	Sweden	RC	3	Comparing open-set speech recognition in CI in subjects with ossified cochleae compared to open cochleae
Burmeister	2017	*Am J Otolaryngol*	USA	CS	4	Evaluating outcomes of CI in patients with otic capsule otosclerosis
Calmels	2007	*Acta Otolaryngol*	France	RC	3	Comparing stapedotomy with CI as first intention treatment
Castillo	2014	*Am J Otolaryngol*	Spain	RC	3	Comparing the results and complication rates of CI in patients with FAO versus unknown origin hearing loss (UOHL)
Fayad	1990	*Am J Otol*	USA	CS	4	Comparing surgical data and hearing outcomes between groups with and without ossified scala tympani
Flook	2011	*J Int Adv Otol*	UK	CS	4	Review for preoperative predictors of difference in audiological outcome, complications, and patient satisfaction for CI in FAO
Hodges	1999	*Am J Otol*	USA	CS	4	Compare speech perception results between CI in obstructed versus open cochleae
Kabbara	2015	*Otol Neurotol*	France	CS	4	Compare stapedotomy versus CI as best initial approach in terms of word recognition, PTA, and radiology
Marshall	2005	*Laryngoscope*	USA	CC	4	Correlate implant performance in cochlear otosclerosis to various factors.
Matterson	2007	*Otol Neurotol*	Australia	CS	4	In CI for otosclerosis: examine the correlation between speech perception and total duration of deafness, age.
Mosnier	2007	*Adv Otorhinolaryngol*	France	CS	4	Evaluate hearing outcomes, surgical difficulties, and complications of CI in patients with FAO
Muckle	1994	*Am J Otol*	USA	CC	3	Evaluate facial nerve stimulation and its incidence in the setting of otosclerosis
Roland	2008	*Otol Neurotol*	USA	CS	4	Examine electrode insertion and open-set speech performance with partial and double-array implantation
Rotteveel	2004	*Otol Neurotol*	N'lands	RC	3	Examine the clinical characteristics, CT and surgical findings, and complications of CI in FAO
Rotteveel	2010	*Audiol Neurotol*	N'lands	RC	3	Analyze the speech perception performance of CI in FAO and evaluate factors influencing performance
Ruckenstein	2001	*Otol Neurotol*	USA	PC	3	Evaluate speech perception and complication rates from CI in FAO
Sainz	2009	*Otol Neurotol*	Spain	PC	3	Describe midterm complications and hearing outcomes in CI for otosclerosis
Semaan	2012	*Am J Otolaryngol*	USA	RC	3	Compare hearing outcomes in CI for FAO versus age-matched controls and review complications
Silveira	2017	*J Otolaryngol ENT Res*	Brazil	RC	3	Compare hearing outcomes of CI for FAO versus other etiologies of deafness
Tokat	2022	*J Int Adv Otol*	Turkey	RC	3	Evaluate the surgical and auditory outcomes of cochlear implantation in patients with cochlear ossification
Vashishth	2017	Otol Neurotol.	Italy	CS	4	Compare auditory outcomes and FNS rates in CI between ossified cochleae and non-ossified cochleae
West	2017	*J Int. Adv Otol*	Denmark	CS	4	Report surgical results and complications of CI in patients with FAO
Zaghis	2003	*J Otolaryngol*	Italy	PC	3	Compare objective and subjective outcomes of CI in totally ossified versus patent cochlea

CC = case-controlled study; CI = cochlear implantation; CS = case series; FAO = far advanced otosclerosis; FNS = facial nerve stimulation; LOE = Oxford Level of Evidence; PC = prospective cohort study; RC = retrospective cohort study.

27 for the systematic review meta-analysis (Fig. 1). Twelve full texts were excluded on the basis of non-English speaking subjects and 10 full texts were not attainable despite contacting the respective authors. Three case reports were excluded from screening. Three articles were excluded due to duplicate data sets from identical authors, but 2 articles by Rotteveel et al. were both included,[14,24] as one included complication data,

whereas the other included speech perception data. There were several studies in which outcomes of interest were reported and analyzed but data was not retrievable either because the data was presented ambiguously or in graph form not amenable to previously described image analytics. These studies were included in the qualitative synthesis but excluded from quantitative analysis for the respective outcome.

Assessment of Bias

Out of 27 studies, 15 cohort studies were evaluated with the NOS, with 73% ($n = 11$) of studies scoring at least 6 out of 8 (Table I). Fifty-three percent ($n = 8$) of studies lacked preoperative outcome reporting, and 40% ($n = 6$) were limited in comparability between cohorts. Twelve case series were evaluated with the NIH scale, with 58% ($n = 7$) receiving at least 7 out of 9 (Table II). Thirty-three percent ($n = 4$) of case series lacked an adequate follow-up interval of >6 months and 67% ($n = 8$) lacked a description of the statistical method.

Bibliometrics

A summary of publications is shown in Table III. All studies were at Oxford level 3 or 4, with 14 studies (52%) at level 3 and 13 studies (48%) at level 4. There were no randomized control trials and 3 prospective cohort studies. There was no trend toward a higher level of evidence over time.

TABLE IV.
Patient Characteristics.

Studies	*n*	Age (Years)	Sex (% Female)	Duration of Deafness (Years)	Previous Stapedectomy (%)	Implant Type(s)
Baijin 2020	13	56	23	—	53.8	—
Balkany 1988	1	35	—	—	—	Nucleus
Berrettini 2004	5	48	40	—	40	Nucleus 24M
Bird 1999	1	70	100	—	100	Clarion
Bredberg 2003	2	—	—	9	—	Med El Combi 40/40+ GB split electrode
Burmeister 2017	6	68	0	—	40	Nucleus Contour Advance peri-modiolar electrode array
Calmels 2007	7	64	71	10	57	Nucleus system 22, Nucleus system 24
Castillo 2014	17	56	77	—	65	Pulsar (Med-El)
Fayad 1990	20	—	—	—	—	Single-channel 3 M/House cochlear implant
Flook 2011	35	64	—	13	28.6	Cochlear CI24K-CON, Cochlear CI22M, Cochlear Freedom, Med-El C40+, Med-El Sonata Flex, Ineraid
Hodges 1999	3	74	—	16	—	Clarion, Nucleus 22
Kabbara 2015	34	—	—	—	73.5	Straight electrode, perimodiolar of unspecified type
Marshall 2005	30	—	—	5	60	Clarion C1.2 enhanced bipolar, Clarion HiRes90 K, Clarion HiFocus II, Nucleus 24
Matterson 2007	59	66	44	23	—	Nucleus Straight, Nucleus Contour
Mosnier 2007	19	61	56	3	68.7	Nucleus 22, Nucleus 24, Bilateral Med-El Combi-40
Muckle 1994	3	58	33	—	33	Nucleus 22
Roland 2008	1	55	—	3	—	Single array but unspecified which implant type
Rotteveel 2004	56	62	—	—	62.2	Nucleus, Clarion, Combi 40+ devices
Rotteveel 2010	28	64	—	11	41%	Nucleus, Clarion, Combi 40+ devices
Ruckenstein 2001	8	62	25	9	100	Nucleus 22, Nucleus 24, Clarion
Sainz 2009	15	59	60	27	100	Med-El Combi 4-+ device standard electrode array
Semaan 2012	34	72	53	—	42.8	Nucleus Freedom, Nucleus 24, Nucleus 5, HiRes
Silveira 2017	17	50	53	8	—	Cochlear Nucleus 24k, 24M, and Contour
Tokat 2022	10	51	—	8	—	Cochlear Slim Straight, Medel Standard, and Split, Advanced Bionics HiRes Standard
Vashishth 2017	38	60	58	29	63.9	Cochlear straight electrodes, Medel and MXM
West 2017	10	71	38	—	100	Nucleus Contour, Freedom, Freedom Advance, 512, Advanced Bionics
Zaghis 2003	2	60	50	15	—	Nucleus M24, Digisonic

Patient Characteristics

Across these studies, there were 474 patients undergoing cochlear implantation identified with a mean age of 64 years and an average duration of deafness of 16 years prior to implantation. There were 48% females and 52% males. 63% of patients had previous stapes surgery. Findings, including implant types, are summarized in Table IV.

Speech Perception Outcomes

Twenty-six studies reported speech perception outcomes. The preoperative and postoperative outcomes and testing information are detailed in Table V. Thirty-eight percent ($n = 10$) of studies had both preoperative and postoperative speech outcomes, whereas the majority (62%) had only postoperative data. There was a wide array of testing used, most commonly the Central Institute of Deafness sentence score ($n = 6$). In studies with both preoperative and postoperative speech discrimination data, significant improvements were noted in all except Roland et al., in which case the single subject scored 0% before and after. There were no cases of deterioration in speech discrimination.

Operative Complications

Studies examined a wide variety of complications. Twenty studies reported FNS, showing an overall rate of 18% (95% confidence interval, CI 12%–27%). The partial insertion rate across 21 studies was 10% (95% CI 7%–15%). FNS and partial insertion rate analysis had heterogeneity with I^2 of 46% and 7%, respectively (Figs. 2 and 3). Postoperative electrode deactivation was mentioned in 16 studies, with electrodes switched off in 11 of

TABLE V.
Speech Discrimination Outcomes.

Studies	*n*	SD Test	Testing Conditions	Timing After Implantation	Preop SD	Postop SD	% Change
Baijin 2020	13	Speech recognition	—	12 mo	—	78%	—
Balkany 1988	1	Iowa sentences	Single-walled sound booths, 72-73 dB SPL as single presentation	—	—	35%	—
Berrettini 2004	5	Open Set Word Recognition Score	With amplification, without lip-reading, in a quiet room	12 mo	4%	98%	+94%
Bird 1999	1	CID sentences	Test materials were presented at 70 dB SPL in quiet	46 mo	—	32%	—
Bredberg 2003	2	Sentence Test Scores	70 dB in quiet without lip-reading	12 mo	—	81%	—
Burmeister 2017	6	HINT	Contralateral ear occluded, no other details	6 mo	1%	75%	+74%
Calmels 2007	7	Dissyllabic words	Words voiced at 70 dB	—	0%	80%	+80%
Castillo 2014	17	CID sentences	—	12 mo	11%	100%	+89%
Fayad 1990	20	MTS Stress	—	—	—	87%	—
Flook 2011	35	CUNY sentences	In quiet	9 mo	—	83%	—
Hodges 1999	3	CID sentences	70 dB SPL in sound field without lip-reading	10 mo	—	87%	—
Kabbara 2015	34	Word Recognition Scores	60 dB without contralateral hearing aid	12 mo	—	73%	—
Marshall 2005	30	CID sentences or HINT (mixed)	—	12 mo	—	75%	—
Matterson 2007	59	Phonemes	—	12 mo	—	61%	—
Mosnier 2007	19	Sentence score	—	12 mo	—	89%	—
Muckle 1994	3	CID sentences	Without lip-reading	12 mo	—	15%	—
Roland 2008	1	CNC words	—	—	0%	0%	0%
Rotteveel 2010	28	BKB test	Sound-treated booth, 65 dB SPL	9 mo	—	66%	—
Ruckenstein 2001	8	CID sentences	—	6 mo	0%	93%	+93%
Sainz 2009	15	Discrimination of common phrases	—	12 mo	0%	83%	+83%
Semaan 2012	34	HINT sentences	Best aided conditions	12 mo	18%	92%	+74%
Silveira 2017	17	Open set sentences	Open set sentences, live voice	12 mo	—	85%	—
Tokat 2022	10	CAP-II score	70 dB in silent room with live voice	12 mo	—	7.6	—
Vashishth 2017	38	Open set sentences scores	Live voice through sound field at 70 dB SPL	12 mo	20%	68%	+48%
West 2017	10	Discrimination scores	—	—	12%	—	—
Zaghis 2003	2	Open set sentences	Without lip-reading	24 mo	0%	25%	+25%

AB = Arthur Boothroyd; BKB = Bamford-Howal-Bench; CAP-II = Central auditory processing II; CID = Central Institute of Deafness; CNC = Consonant-Nucleus-Consonant; CUNY = City University of New York; HINT = hearing in noise test; MTS = Monosyllable Trochee-Spondee; NU6 = Northwestern University 6; SD = speech discrimination; SPL = sound pressure level.

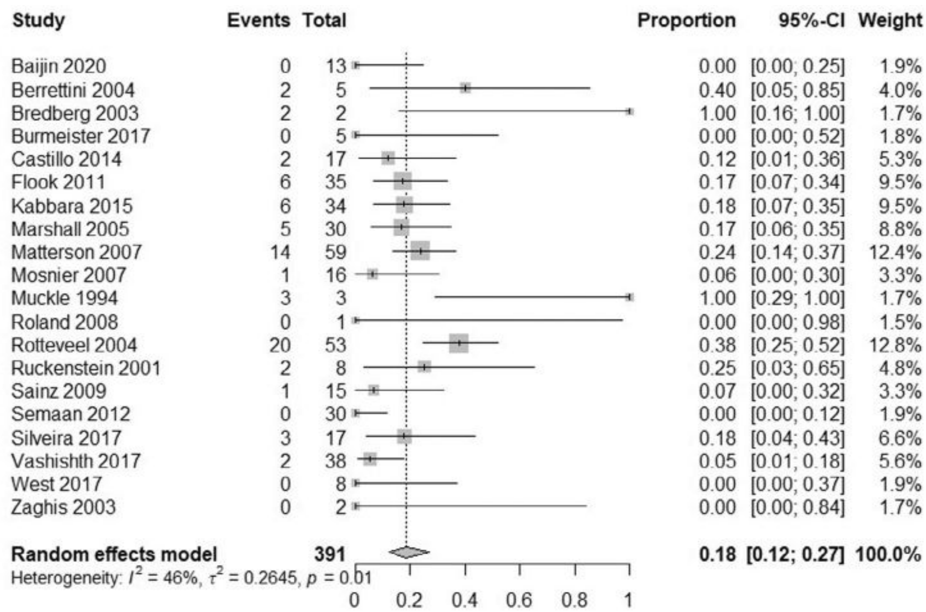

Fig. 2. Forest plot of weighted pooled prevalence of facial stimulation rates.

these studies, mostly for FNS. In most cases, only 1–2 electrodes were switched off, with the maximum being 8. Furthermore, out of 310 patients across studies reporting all other relevant complications, there were 4 cases of CSF gusher, 5 cases of otalgia, 2 cases of severe vertigo, and 2 cases of severe progressive tinnitus, dizziness, and headache.

DISCUSSION

Although CI in FAO is practiced across many institutions, this study has set out to formally aggregate speech outcomes and complication rates in this group to formally validate its utilization. Overall, the results from this study have shown that improvements in speech recognition from CI in FAO are equivalent to general adult CI outcomes. However, there are higher than usual rates of FNS and partial insertion.

Improvement in speech recognition at 12 months in this study was on par with outcomes in a recent scoping review of all adult CIs from 2000–2018.[25] This scoping review reported a mean preoperative score of 20% and a mean postoperative score of 75% for open set sentences in quiet at 12 months' follow-up, an improvement of 55%.

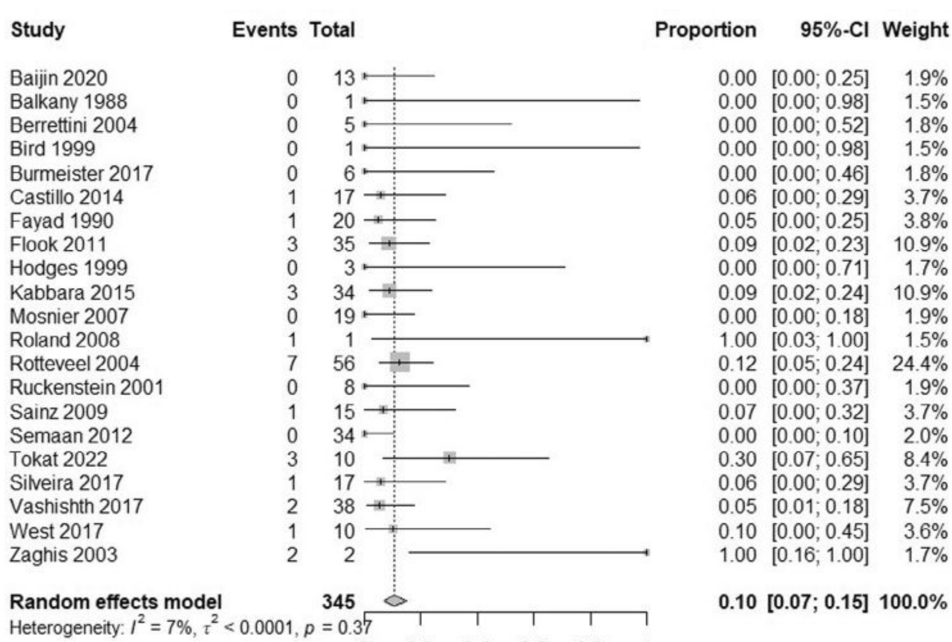

Fig. 3. Forest plot of weighted pooled prevalence of partial insertion rates.

Five studies representing 96 patients included in this review fulfilled these similar testing criteria and had a weighted aggregate improvement from 16% to 82% (66% improvement). This finding is consistent with the conclusion reached in individual comparative studies included in this review.[12,13,26,27]

Histologically, otosclerosis affects the endosteal lining and lateral wall of the cochlea with relatively little effect on spiral ganglion neurons.[28] However, concerns raised in the literature pertaining to reduced impedance of the otic capsule causing shunting of electrical currents through bone, lead to an increased current requirement to stimulate auditory nerve fibers and a potential excess current spread to the facial nerve.[11,29] This phenomenon is in addition to increased NAS, requiring more frequent adjustment of the electrical map. Indeed, these challenges may be addressed with proper postoperative follow-up and optimization. Therefore, studies comparing speech perception outcomes between otosclerosis and non-otosclerosis patients exhibit trends but do not reach significance.[30,31] However, it is still imperative to acknowledge that long-term progressive structural disturbances that are characteristic of the disease can cause increasing difficulties in the propagation of the electrical stimuli, affecting thresholds, electric charges, pulse widths, stimulation rates, and eventually, the number of active electrodes.[2,10,11]

The present study determined the incidence of FNS in CI recipients with otosclerosis to be 18%. FNS was the most common NAS in this patient group, and these figures are higher than a rate of 6% reported across reviews for all adult CI patients.[32] FNS, in all cases, prompted switching off of electrodes, in most cases 1–2, and in certain cases up to 8 electrodes.[33] Studies suggested the benefit of the use of perimodiolar electrodes to achieve lower rates of FNS.[13,17]

Partial insertion rates of 10% found in this study are higher than rates of 2% in a review of adult CI complications.[34] Although the difficulty of electrode insertion in otosclerosis is often attributed to ossification of the basal turn, a histopathological analysis by Lee et al.[35] showed that cases of partial insertion were associated with cochlear lumen obstruction in only a small number of cases. In most cases, cochlear obstruction is bypassed with the drilling of the basal turn, thus enabling full insertion of the array. Alternatively, scala vestibuli insertion was utilized to a varied extent, with the upper limit being 21% of Kabbara et al.'s reported cases.[5] Despite electrode switch-off due to NAS and higher partial insertion rates, CI in FAO had comparable speech discrimination rates to normal adult CI. The authors propose that speech perception has an inverse exponential decay relationship to electrode deactivation, such that there is a threshold of stimulation. For instance, significant deterioration in speech perception is seen with a higher number of electrodes deactivated[36] whereas not observed with smaller numbers deactivated.[37] Furthermore, a recent study by Atanasova-Koch et al. also notes that de-activated electrodes did not make a significant difference in speech differentiation.[38] The patients included in the papers reviewed in this study had minimal electrode deactivation (in the order of 1–4). Partial insertion rate across 96 patients in the five studies qualifying for weighted aggregate was 4%, likely not having a significant bearing on speech discrimination. However, higher rates of partial insertion in FAO overall—10% as shown in this study—would likely have an impact on postoperative speech perception outcome in a larger group quantitative analysis.

With regard to other complications, implantation in FAO had higher incidences of CSF gushers and lower rates of vertigo. This is compared to a review of 168 adult CI cases, which recorded no cases of CSF gusher, 13 cases of vertigo, 7 cases of tinnitus, and 1 case of facial palsy.[39]

Ultimately, there are no widely accepted decision-making guidelines on treatment modalities in FAO. Several studies recommend a phased approach where stapedotomy and hearing aids are offered as a primary treatment and CI as the preferred salvage technique.[18] This is supported by studies showing that salvage implantation carries the same benefits as primary implantation.[15]

In the design of this study, patients with concurrent otologic history and non-English testing were excluded in anticipation of confounding etiology and cultural and linguistic differences. Risk of bias was low to moderate across the studies, with most studies scoring favorably on the NOS or the NIH scale. Outliers included Bredberg et al., which was a multinational study with inconsistent methodology and multiple confounders affecting comparability.[40]

This study encountered numerous limitations that impacted the synthesis of results. Across the studies, there was a large variety of testing modalities with varied or limited reporting of testing conditions and follow-up timeframes. Many studies were noted to use custom speech discrimination tools that were poorly described and not validated. Therefore, meta-analysis and statistical pooling of speech discrimination were not feasible due to incomparability of outcomes. Furthermore, the limited reporting of preoperative speech discrimination precluded change score generation for effect size estimation. Additionally, limitations stemmed from the inherent clinical diversity of the populations, with differing patient characteristics, implant types, and surgical techniques. These factors limited the comparability of cohorts across studies and further added to the challenge of data aggregation. Several of the gathered patient characteristics and outcome data points were unable to be analyzed due to the sparse reporting across studies. For instance, radiological grading data was seldom reported in studies, and when reported, it often varied widely in classification systems. Lastly, the papers included in this review were largely cohort studies and case-reviews with no randomized control trials and few prospective studies, which ultimately limited the quality of the analysis.

This review is the first to provide numerical validation for CI in FAO. Future reviews should be conducted with the availability of sufficient studies with similar testing modalities, conditions, and timeframes with both preoperative and postoperative speech discrimination scores, enabling quantitative analysis in a larger sample size and incorporating higher-level evidence in the form of prospective studies and randomized control trials.

CONCLUSION

Cochlear implantation in FAO demonstrates significant gains in open-set speech discrimination scores with higher FNS rates (18%) and partial insertion rates (10%). With appropriate surgical planning, patient counseling, and postoperative care, implantation in this cohort is likely to provide patient benefit, particularly as salvage surgery. However, quantitative analysis was limited by the lack of homogeneity in testing modalities, patient characteristics, and sample sizes. Continued efforts to standardize data reporting will likely facilitate future reviews.

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