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Main Article

Long-term outcome of children undergoing surgery for suspected perilymph fistula

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Abstract

A number of authors have suggested that surgery for suspected perilymph fistula is effective in preventing deterioration of hearing and in improving hearing in some cases in the short term. We present long-term hearing outcome data from 35 children who underwent exploration for presumed perilymph fistula at The Children's Hospital, Sydney, Australia, between 1985 and 1992.

Methods: The pre-operative audiological data (mean of 500, 1000, 2000 and 4000 Hz results) were compared with the most recently available data (range two to 15 years) and the six-month post-operative data.

Results: The short-term results showed no significant change in hearing at six months, with a subsequent, statistically significant progression of hearing loss in both operated and non-operated ears (Wilcoxon signed rank test: operated ear, $p < 0.017$; non-operated ear, $p < 0.009$).

Conclusion: In this case series, exploratory surgery for correction of suspected perilymph fistula did not prevent progression of long-term hearing loss.

Key words: Perilymph; Fistula; Sensorineural Hearing Loss; Otologic Surgical Procedures

Introduction

It has been suggested by some authorities that children with sudden or fluctuating hearing loss should undergo exploratory surgery to exclude perilymph fistula as the underlying cause for their symptoms and, where necessary, to block the fistula in an effort to prevent progression of symptoms. The potential sites of these fistulae and the methods used to close them have been variously described. However, the diagnosis of a perilymph fistula remains controversial, and most series rely on subjective opinion at the time of operation. Although short-term hearing outcomes have been reported, the long-term outcome for the operated ear in such children (and comparison with the non-operated ear) has not been well documented; this study addresses this issue.

Method

The senior author (EJB) prospectively collected data on all children undergoing exploration for perilymph fistula at The Children's Hospital, Sydney, Australia, between 1985 and 1992. Pre- and post-operative hearing levels were documented, together with the site of, and method used to close, the fistula where appropriate. Any relevant radiographic findings, blood tests, and medical, genetic or family history were also recorded.

Ethical approval was obtained to contact these patients, and consent was subsequently obtained to access current medical records, scans and hearing tests, which were reviewed by AHJ. The pre-operative audiological data (mean of 500, 1000, 2000 and 4000 Hz results) were compared with the most recently available data (patient age range two to 15 years) and with the six-month post-operative data. The hearing outcomes for both the operated and non-operated ears were analysed using the Wilcoxon signed rank test.

Results

A total of 49 operations were undertaken in 39 children (20 male and 19 female) aged one to 16 years (Figures 1 and 2); long term results were obtained for 35 of these children. Presenting symptoms included fluctuating deafness, sudden hearing loss, progressive hearing loss and vertigo, consistent with other studies.

A review of patients' case notes for other potential causes of hearing loss identified the following risk

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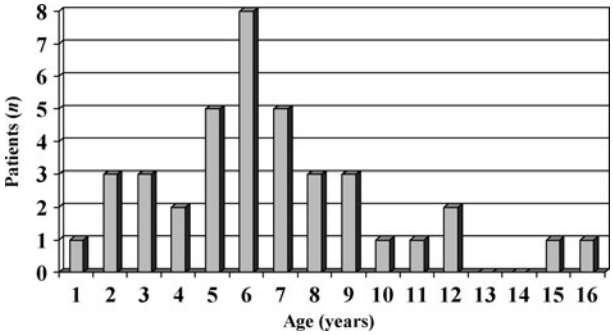

FIG. 1
Patients' age distribution at first operation.

factors. Five children had evidence of cytomegalovirus, mumps or toxoplasmosis infection. Five children had a family history of hearing loss. Three children had a history of meningitis. One child each had a diagnosis of hydrops, Pendred's syndrome or chondrodysplasia.

A review of patients' computed tomography scan reports showed the following diagnoses: reported as normal ($n = 21$); osteoneogenesis post-meningitis (one); Mondini defect (nine); large vestibular aqueduct (two).

A review of patients' operative findings to establish the presence and site of fistula showed the following results: oval window ($n = 36$); round window (two); oval and round window (two); no fistula found (nine); abnormal stapes (12).

A variety of packing materials was used to pack the oval and round windows, including connective tissue, fat and perichondrium.

Full details of the presenting symptoms, presence or absence of fistula, and other findings for the 35 children with long-term follow up are given in Table I.

Audiological data

Figures 3 and 4 show graphical representations of hearing change data for both the operated and non-operated ears. Analysis of audiological data (mean of 500, 1000, 2000 and 4000 Hz results) using the Wilcoxon signed rank test showed no statistically significant progression of hearing loss (compared with pre-operative levels) in either the operated or non-operated ears in the short term. In the longer term however, the Wilcoxon signed rank test showed significant progression of hearing loss in both the operated and non-operated ears (operated ear, $p < 0.017$; non-operated ear, $p < 0.009$). However, it should also be noted that if a 10 dB hearing change is taken as the cut-off level for test–retest variation, then seven of 44 ears in the operated group showed an improvement of 10 dB or more in the longer term, compared with zero of 17 in the non-operated group.

Ten per cent of the children in this study subsequently progressed to cochlear implantation.

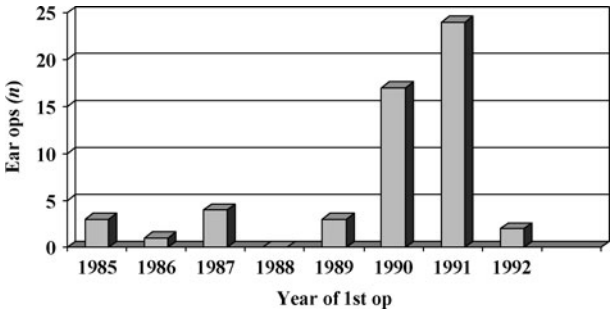

FIG. 2
Number of initial ear operations (ops) carried out per year, to 1992.

Discussion

Perilymph fistula is easily defined as an abnormal connection between the perilymph space of the inner ear and the middle ear. However, the diagnosis and treatment of such cases remains much more controversial. No consistent collection of symptoms and signs are described, with any combination of hearing loss, tinnitus and vestibular symptoms being possible.[1] Although beta-2 transferrin has been used to support the diagnosis,[2] the specificity of the test is unclear and it does not enable pre-operative diagnosis.[3] Careful history-taking and exclusion of other causes have been used to support surgical exploration, with packing of the round and/or oval windows if a fistula is seen[4] or, in some cases, if no evidence of a fistula is found at surgery.[3] The outcomes from these studies are variable, with stable or improved hearing in 23 to 87 per cent of patients.[3,5,6]

Our study represents a retrospective analysis of a case series with meticulous data collection. Most of the data are comparable to other studies in this area, in terms of patient age, variety of presenting symptoms and positive findings at surgery.

- **Several authors have suggested that surgery for suspected perilymph fistula is effective in preventing deterioration of hearing and in improving hearing in some cases in the short term**
- **This paper reports long-term hearing outcome data from 35 children undergoing exploration for presumed perilymph fistula at The Children's Hospital, Sydney, Australia, between 1985 and 1992**
- **Short-term results showed no significant change in hearing at six months; however, a subsequent significant progression of hearing loss was observed in both the operated and non-operated ears**

External advice was sought from a medical statistician regarding formal analysis and presentation of data. Our results suggest that exploration and repair of suspected perilymph fistula does not adversely

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TABLE I

PATIENT DETAILS

Pt ID	Presenting symptoms	Fistula?	PTA* (dB)			CT	Other information†
			Pre-op	Post-op	Recent		
2‡	Progressive, fluctuating HL	Yes	116	117	120	Normal	Pneumococcal meningitis
2	Progressive, fluctuating HL		112	109	110	Normal	Pneumococcal meningitis
3‡	Progressive, fluctuating HL	Yes	109	81	96	Normal	Congenital
3	Progressive, fluctuating HL		96	97	>120	Normal	Congenital
5‡	Progressive, asymmetrical HL	Yes	82	105	90	Normal	Family history
5	Progressive, asymmetrical HL		55	86	79	Normal	Family history
7‡	High frequency HL	Yes	20	22	26	No scan	Previous surgery
7	High frequency HL		5	9	10	No scan	
9‡	Vertigo	No	62	92	120	Mondini	Congenital
9	Vertigo		62	79	75	Mondini	
10‡	Vertigo & fluctuating HL	Yes	102	102	108	Possible LVA	Family history
10‡	Vertigo & fluctuating HL	Yes	100	97	110	Possible LVA	
11‡	Sudden HL	Yes	102	70	72	Bulbous cochlea	Congenital (toxoplasmosis)
11‡	Sudden HL	Yes	62	77	79	Bulbous cochlea	
12‡		Yes	90	72	96	Normal	Congenital
12‡		Yes	72	75	87	No scan	
13‡		Yes	112	103	82	Mondini	
13			64	60	61	Mondini	
14‡	Asymetric HL	Yes	30	16	21	No scan	
14	Asymetric HL		15	10	14	Normal	
15‡	Sudden HL	No	80	75	65	No scan	
15	Sudden HL		55	59	46	No scan	
16‡	Fluctuating HL	Yes	86	74	84	Normal	CMV (unaffected twin)
16‡	Fluctuating HL	Yes	102	100	>120	Normal	
17‡	Fluctuating HL	Yes	77	76	102	Normal	Congenital
17	Fluctuating HL		75	77	107	Normal	Congenital
19‡	Fluctuating HL	Yes	25	39	85	Normal	Mumps & CMV +ve
19	Fluctuating HL		27	32	45	Normal	Mumps & CMV +ve
20‡	Ataxia & sudden HL	No	64	95	102	Mondini	
20	Ataxia & sudden HL		64	95	>120	Mondini	
21‡	Progressive HL	Yes	70	69	95	Normal	Congenital & family history
21	Progressive HL		67	74	93	Normal	Congenital & family history
22‡	Fluctuating HL	Yes	112	91	90	Possible Mondini	Congenital + previous surgery
22‡	Fluctuating HL	Yes	99	82	83		
28‡	Sudden HL	Yes	94	87	102	LVA	
28‡	Sudden HL	No	92	95	105		
29‡	Sudden HL	Yes	82.5	116	116	Mondini / LVA	CHARGE
29	Sudden HL		79	90	96	Mondini / LVA	CHARGE
31‡	Progressive & fluctuating HL	Yes	56	92	112	Normal	
31‡	Progressive & fluctuating HL	Yes	105	80	112	Normal	
32‡	Progressive HL	Yes	70	81	89	Normal	Congenital
32	Progressive HL		72	84	91	Normal	Congenital
33‡	Sudden HL	Yes	45	31	37	Normal	Congenital
33	Sudden HL		45	32	37	Normal	Congenital & family history
38‡	Sudden HL	No	85	90	108	Normal	Congenital
38	Sudden HL		92	87	106	Normal	Congenital
40‡	Sudden HL	Yes	62	71	90	Normal	Pneumococcal meningitis
40‡	Sudden HL	Yes	56	50	64	Normal	
41‡	Vertigo & fluctuating HL	Yes	65	61	60	Possible Mondini	
41‡	Vertigo & fluctuating HL	Yes	57	35	62	Possible Mondini	
42‡	Progressive HL	Yes	50	55	62	Possible Mondini	
42‡	Progressive HL	Yes	46	48	62	Possible Mondini	
43‡	Fluctuating HL	Yes	77	80	62	No scan	
43	Fluctuating HL		75	82	71	No scan	

*Average of 500, 1000, 2000 and 4000 Hz results. †Other potential risk factors for hearing loss. ‡Operated ear. Pt ID = patient identification number; PTA = pure tone audiometry; CT = computed tomography; pre-op = pre-operative; post-op = 6 months post-operative; HL = hearing loss; LVA = large vestibular aquaduct; CMV = cytomegalovirus; +ve = positive

affect the short-term hearing results, and may appear to offer protection against further loss. However, our longer-term results show that this hearing stabilisation was not maintained. Both operated and non-operated ears showed a statistically significant progression of hearing loss in the longer term, as judged by the Wilcoxon signed rank test. However, seven of the 44 ears in the operated group showed a hearing improvement of 10 dB or more in the longer term, compared with zero of 17 in the non-operated group. We were unable to find any correlations with symptoms, investigations, or radiological or surgical findings in the sub-group showing improvement, which might identify those patients who would benefit from exploratory surgery.

The presence of a perilymph fistula is difficult to confirm and is undoubtedly a subjective diagnosis in the operative setting. Our group of patients was

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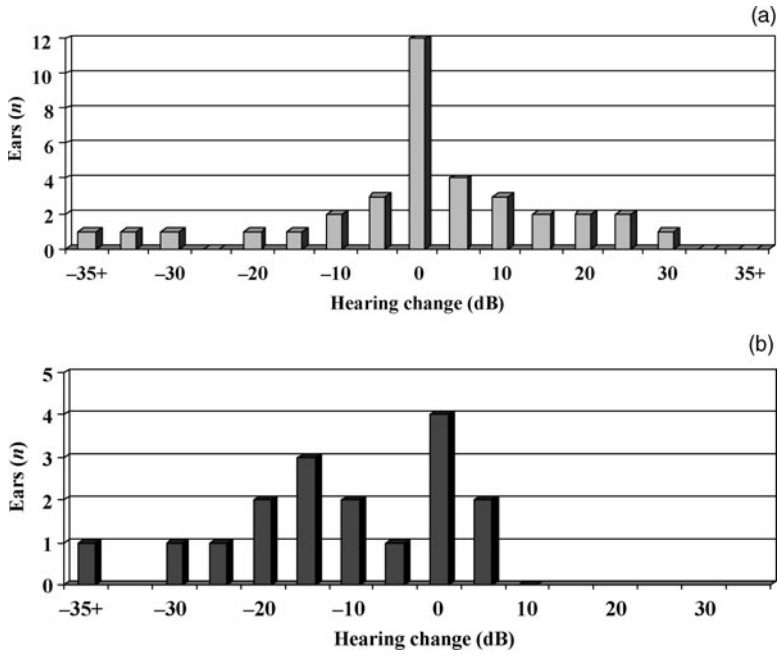

FIG. 3

Patients' short-term hearing outcomes. Change in hearing (as average of 500, 1000, 2000 and 4000 Hz results) in the (a) operated and (b) non-operated ears, compared with pre-operative average hearing levels. Worsening hearing is indicated by negative values. For operated ears: mean hearing change = 0.39 dB; standard deviation (SD) = 16.4 dB; 95% confidence intervals (CI) = −5.0 to 5.8. For non-operated ears: mean hearing change = −6.0 dB; SD = 12.0 dB; 95% CI = −0.2 to −11.8.

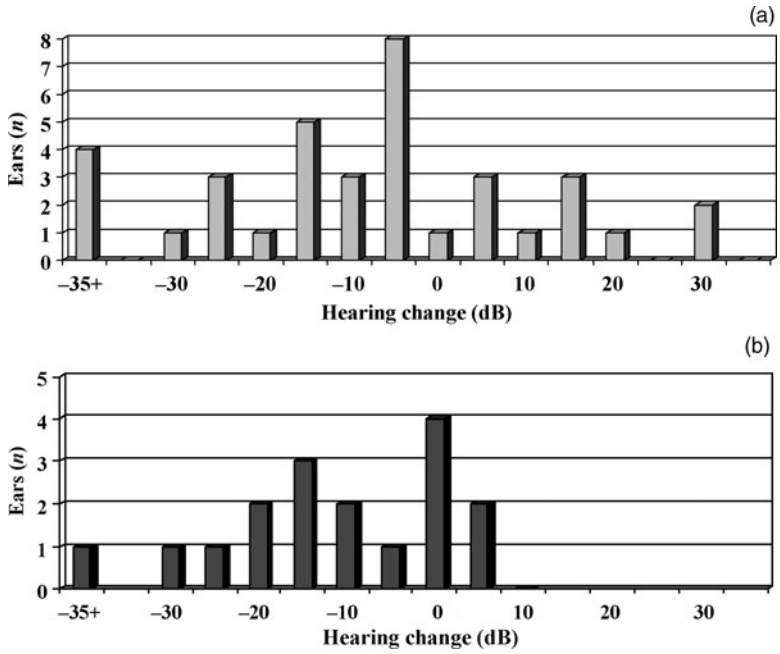

FIG. 4

Patients' long-term hearing outcomes. Change in hearing (as average of 500, 1000, 2000 and 4000 Hz results) in the (a) operated and (b) non-operated ears, compared with pre-operative average hearing levels. Worsening hearing is indicated by negative values. For operated ears: mean hearing change = −9.7 dB; standard deviation (SD) = 22.0 dB; 95% confidence intervals (CI) = −2.6 to −17.0. For non-operated ears: mean hearing change = −13 dB; SD = 17.1 dB; 95%CI = −4.7 to −21.4.

investigated at a time when exploratory surgery and treatment for suspected perilymph fistula was accepted practice for a variety of symptoms. It is unlikely that all of these patients would undergo such surgery today, and we accept that a number of different aetiologies may have been responsible for these children's symptoms. However, the short-term results are in keeping with other published series

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which have suggested that exploration and surgical repair may prevent further deterioration in hearing. Our results suggest that this preservation may not be maintained in the longer term.

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