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Endoscopic findings and long-term hearing results for pediatric unilateral conductive hearing loss

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ABSTRACT

Objectives: Analyze reasons for unilateral conductive hearing loss (CHL) with unknown etiology in children.
Introduction: Unilateral conductive hearing loss (HL) without known etiology can be undiagnosed despite of hearing screening programs. It can be difficult to find the reason for HL and to make a treatment plan. Middle ear endoscopy gives hard-evidence diagnosis and basis for an individual treatment plan.
Methods and material: Prospective clinical follow-up study for a cohort of generally healthy elementary school age children with unilateral conductive HL with unknown etiology. The study population was 192 children, of which 46 had a HL of at least 25 dB with more than 10 dB conductive component. Mean age was 8.7 years. Preoperative tests included otomicroscopy, bone- and air-conduction audiogram, tympanometry, stapes reflex tests, Rinne and Weber test and Otoacoustic emissions. The children underwent endoscopy of the middle ear with an individual treatment plan and long-term follow-up. The aim was to explore etiology and to give a treatment plan for hearing loss. Follow-up included air- and bone conduction hearing tests annually or every other year. Mean follow-up was 5.2 years.
Results: A clear etiological finding was found in 36 (78%) ears, stapes anomaly (23) as the most common (64%) finding. Other findings were two cholesteatomas, 2 status after trauma, 5 middle ear anomalies, 5 incus fixations and one incus erosion. Air conduction hearing improved spontaneously during follow-up in 81% (17/21, 2 dropouts) of the stapes anomaly ears (mean 11,3 dB, range 4–32 dB), and none of these ears showed hearing deterioration. In the incus fixation group, one ear showed hearing deterioration. There were no major complications for exploration, and 5 minor postoperative infections.
Conclusions: The most common reason for pediatric unilateral conductive hearing loss was stapes anomaly/fixation. The HL does not deteriorate. Hearing loss in stapes anomalies shows a tendency for spontaneous recovery. Stapes surgery can be postponed or avoided.

1. Introduction

Pediatric unilateral conductive hearing loss (CHL) is mainly bass dominated hearing loss and it is usually due to anomalies in the ossicles [1–3]. The anomaly of the stapes can give diagnosis of pediatric otosclerosis. Other malformations of the ossicular chain include fixation of the incudomalleal or incudostapedial joint or one of the ossicles to the Scutum or attic ridge [4,5].

If the hearing loss (HL) is unilateral, it can be difficult to detect before the child is old enough to be tested with clinical pure tone audiogram with bone conduction. A child can be habituated to unilateral bass dominated HL and this kind of HL can be undiagnosed until school age. Unformal tests were used as hearing screening before universal newborn hearing screening become gold standard. These tests are often performed by other that trained audiologist. In Finland the first screening audiogram with a pass level of 20 dB is usually done at 4 years age and then at school start. Unilateral bass defect did not necessarily lead to referral to clinical level audiometry tests. Therefore, diagnosis of unilateral bass dominated HL can be delayed to school start. At this age, it is also possible to perform reliable bone conduction tests.

In Finland universal newborn hearing screening become standard during the first decade of this millennium. The present screening devices are set to 35 dB pass level. This applies both Otoacoustic emission and automatic auditory brain stem (aABR) pass levels. For children the zero level of Hearing Sensation Level (HSL) in bone conduction can be −10 to −15 dB [6]. This means that clinically relevant conductive HL can hide behind passed screening test. The old screening methods and

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relatively little obvious handicap from unilateral bass dominated HL led to accumulation of these type referrals to ENT clinic when the children reached school age.

Päijät-Häme Central hospital serves as a single referral hospital with an ENT department for a population of about 210 000 people. The population in the hospital district is relatively stable in the last decades with little moving in or out of the hospital district. This leads to reliable data about hearing problems for children in this hospital district. When the author started at the ENT department in 1999, there was a substantial group of children with referral to diagnostic tests and treatment plan for unilateral conductive hearing loss. A special subgroup for these children is those without any obvious reason or anamnestic explanation to HL. At that time, the CT scan with relatively high exposure to radiation was not good enough method as diagnostic tool for unilateral CHL. The load of referrals gave the incentive to investigate unilateral CHL for these children. Uncertainty and worry of prognosis for HL was the main concern for parents. The aim for the study was to find out the reason for unilateral CHL and to make a treatment plan and start a long-time follow-up. The method to reach this goal was to make endoscopy of the middle ear. The parents were as a rule relieved when they received the proposition of this study and the informed consent of pros and cons for the treatment.

2. Material and methods

The study setting was a prospective clinical follow-up study for a cohort of generally healthy elementary school age children with unilateral conductive hearing loss with an unknown etiology. The ethical committee for the hospital accepted the study plan. All parents and children received an informed consent of the study as a treatment protocol in the department. The normal out-patient examinations with otomicroscopy, tuning fork tests, pure tone and speech audiograms, otoacustic emissions, stapedial reflexes and tympanometry did not reveal the etiology for HL. Patients with major ear infections with rupture of tympanic membrane, tympanostomy tube treatment or other middle ear intervention were excluded. The tympanic membrane status should be intact and tympanometry should be of type A. Children with known syndromes or hereditary hearing loss were excluded. The average air conduction hearing level should be at least 25 dB PTA. This is the limit level for hearing aid rehabilitation in Finland for children. The frequencies selected were 0.5, 1, 2 kHz because the hearing loss was as a rule bass dominated. The average conductive hearing loss (AB gap) should be at least 10 dB. The audiometers used were first Madsen and later Aurical clinical audiometers with standardized and calibrated air (THD 39) and bone conduction headsets.

The parents and the children were informed that the study included endoscopy of the middle ear, which could be extended to explorative tympanotomy if necessary and minor defects would be repaired in the same anesthesia. The main goal was to find etiology for hearing loss and to make a plan for further treatment. The children were then included in a long-term hearing control protocol. The child and the parents were informed that the risks included for the treatment were middle ear infection or perforation of the tympanic membrane. The risk could be estimated to be at the same level as for tympanostomy tube treatment, which is about 2% [7].

The study started in 2000 and ended in 2012. Original amount of children referred was 192. Forty-six children fulfilled the inclusion criteria and the child and the parents were informed of the possibility for endoscopy of the middle ear. All parents and children accepted the study plan. Mean age was 8.7 years (range 5–16 years of age). The original study group included 20 boys and 26 girls. The rest of the children (146) did either not have a conductive hearing loss or the reason was obvious, like perforation of the tympanic membrane or secretory otitis media as main findings.

The children were examined in general anesthesia as a day-care procedure. The endoscopy was made thorough the posterior superior quadrant of the tympanic membrane. The tympanic membrane was treated with 90% Phenol to reduce bleeding and pain and to keep the anesthesia level moderate. Phenol as 70 or 90% solution has been used for many decades as local anesthesia for paracentesis in Finland. It can be considered as a safe local anesthesia if used with precaution and only with a minimal amount that cannot reach the middle ear. A small paracentesis was carefully performed without touching the incus. The initial small paracentesis was widened with micro forceps. An Olympus or Stortz 1,9 mm 0 and 30° wide angle endoscopy was used to examine the middle ear systematically. The status of the middle ear was recorded in the patient chart and operation procedure text. The procedure was made in the first phase of the study in direct eye control thorough the endoscope, as the first video cameras were too heavy. The middle ear cavity was systematically examined with status of the stapes, incus, malleus, facial nerve, oval and round window, and promontory. The mobility of the ossicular system (the incus and the incudostapedial joint and the stapes) was examined thorough the same paracentesis with a pick under otomicroscopy. If the finding was unclear or there was need of better visibility, the procedure was converted to explorative tympanotomy with endomeatal incision. The paracentesis was closed with a silk paper patch. The endoscopy gave the diagnosis and tentative prognosis for future treatment plan.

All children were included in a follow-up set-up after the endoscopy. It included an annual or every other year control with pure tone audiogram, air and bone conductions, and examination of the ear with otomicroscopy. The average follow-up was 5.2 years (minimum 1, maximum 11 years). Totally six patients were drop-outs in the study, three early and three late.

Hearing results are based on pure tone air conduction pre- and postoperatively as this is the most reliable hearing test for children. Thus, air-bone gap was not used as a study parameter.

The pure tone bone conduction was also measured, but this test showed more variation and was considered as unreliable. However, the bone conduction test was used to confirm conductive hearing loss as well as stapes reflex, Rinne and Weber test.

3. Results

3.1. Endoscopy results

An etiological finding was found in 36 (36/46, 78%) ears.

Stapes fixation and anomaly (23/36, 64%) was the most common finding. Pure footplate fixation was found in 12 ears. The superstructure of stapes was abnormal in addition to footplate fixation in another nine ears. The crura were abnormal and the arch of the crura was small. The overall variation for stapes anomaly was large from a thickened footplate to a true columella type single column. In two ears also the incudomalleal complex showed reduced mobility. Two patients in this group were drop-outs in the follow-up. The findings and hearing results for this group is presented in Table 1.

Two ears had congenital cholesteatomas. Two ears showed post-trauma findings without any anamnestic background explanation (one ear with crura fracture and one ear with both stapes and incus fracture). Five ears showed middle ear anomalies without stapes anomaly. Ponticulus was missing in three ears and in two ears all ossicles were anomalous. Incus was fixed in 5 ears. In two of these ears, also stapes was fixed, as mentioned and included in the stapes group. The long process of incus had an atypical look without any uniform anomaly type. If the incus was fixed, most probably also malleus was fixed, but it could not be verified by this method. Two ears showed probable sequela of otitis media chronica. One of these ears had adhesions but otherwise normal middle ear and hearing proved normal in the long-term follow-up. The other ear showed incus erosion. One ear showed secretory otitis media without preoperative status findings. Two ears had no findings in the middle ear that could explain conductive hearing loss. It remained unclear if the original conductive hearing loss was true

Table 1
Stapes fixation group.

Patient	Age (years)	dB A Pre	Endoscopy finding	Subjective handicap	Hearing aid	Follow-up (years)	dB A Post	Change in dB A
1	7	35	Footplate fixed and thick	1	1	9	31	4
2	7	32	Footplate fixed	0	0	4	15	17
3	5	30	Crura anomaly	0	0	2	30	0
4	9	30	Footplate fixed	0	0	3	30	0
5	7	30	Footplate fixed	0	0	9	8	22
6	8	35	Footplate fixed and Crura anomaly	1	0	4	15	17
7	13	55	Footplate fixed			drop-out		
8	8	65	Footplate fixed	1	0	2	33	32
9	10	30	Footplate fixed and Crura anomaly	0	0	4	30	0
10	8	30	Footplate fixed	1	1	8	25	5
11	5	33	Footplate fixed	0	0	6	26	7
12	8	35	Footplate fixed	0	0	10	30	5
13	6	65	Footplate fixed			drop-out		
14	13	51	Footplate fixed and Crura anomaly	1	1	6	43	8
15	6	45	Footplate fixed and Crura anomaly	1	1	6	30	15
16	6	35	Footplate and Incus fixed	0	0	7	30	5
17	8	27	Footplate fixed and Crura anomaly	0	0	7	5	22
18	8	25	Footplate fixed	0	0	4	0	25
19	14	30	Footplate and Incus fixed, Crura anomaly	0	0	9	30	0
20	5	47	Footplate fixed and thick, Crura anomaly	0	0	1	42	5
21	13	46	Footplate fixed and thick, Crura anomaly	1	0	6	25	21
22	11	25	Footplate fixed	0	0	7	15	10
23	11	50	Stapes and Incus anomaly	0	0	2	33	17
Mean	8.5	38.5				5.5	25	11.3

0 = no, 1 = yes.
dB A = PTA 500, 1000 and 2000 Hz, Air conduction.
The dB A Preop. is the preoperative hearing measurement and the dB A Postop. is the last postoperative measurement.

Table 2
Stapes mobile group.

	dBA pre	Age	Findings	Folllow-up	dBA post	
1	27	8	IM fixation, Corda position abnormal	10	32	
2	30	8	IM fixation	3	40	
3	30	6	IM fixation, Crura anomaly, footplate mobile, Ponticulucs missing	1	30	
4	75	10	Crura fracture, Fracture in the ear canal, Facial nerve position abnormal	2	75	
5	65	16	Stapes and Incus fractura	11	65	
6	30	6	Anomaly in all ossicles, footplate normal	9	30	
7	60	9	Incus and Stapes superstructure missing, footplate normal, Facial nerve position abnormal	4	60	
8	35	7	Ponticulus missing, Anterior crura for Stapes missing, footplate normal	0		drop-out
9	30	14	Posterior crus and Stapedial tendon missing, footplate normal	4	20	
10	43	5	Ponticulus missing	6	38	
11	26	6	Congenital cholesteatoma	9	43	Operated
12	27	16	Congenital cholesteatoma	4	0	Operated
13	67	8	Status post otitis media chronica, Incus erosion	7	60	
14	30	5	Status post otitis media chronica, adhesions	3	17	
15	30	6	Secretory otitis media	6	0	
16	32	9	Tinnitus, high Bulbus jugularis and large Hypotympanum, masking problem?	3	10	
17	90	13	Possible barotrauma, no finding, masking problem?	1	100	
Average	43	8.9		4.9	39	

0 = no, 1 = yes. IM= Incudo Malleal
dB A = PTA 500, 1000 and 2000 Hz, Air conduction.
The dB A Pre is the preoperative hearing measurement and the dB A Post is the last postoperative measurement.

or if there were problems with masking. It is also possible that these ears had periodical secretory otitis media as the reason for conductive hearing loss. Thus, thirteen ears in this group had a clear etiological finding in endoscopy. There was one drop-out in this group. The findings for these ears are presented in Table 2.

The operation was converted to explorative tympanotomy with endaural or endomeatal incision in the two ears with fracture findings.

There were no major complications for exploration, and no permanent TM perforations. Five ears showed minor postoperative infection, which was treated with local antibiotics.

3.2. Hearing results

3.2.1. Stapes group

Seven children had subjective handicap of the hearing loss and four of these used a hearing aid.

One child received stapedotomy at the age of 18. The finding was the same as with endoscopy. ME status was normal otherwise, but stapes was fixed. The whole footplate was thick as a sign of footplate anomaly.

The mean preoperative air conduction PTA in the stapes group was 36 dB (38 dB, included dropouts) and 25 dB at the end of the follow-up.

The difference in means is statistically significant with paired *t*-test, $p < 0.05$. The mean bone conduction PTA was 5.1 dB preoperatively and 6.6 dB postoperatively. The difference is not statistically significant. The mean age for the whole group was 8,5 years and average follow-up was 5,5 years.

Nine children showed over 10 dB of hearing improvement. The average follow-up for these children was 4,9 years. Their mean age at the beginning of the follow-up was 8,4 (range 6–13) years and at the end of follow-up 13,3 years (range 10–18).

Air conduction hearing improved during follow-up in 81% (17/21, 2 drop-outs) of the ears (mean 11,3 dB, range 4–32 dB), and no ear showed hearing deterioration. The improvement was gradual and the preoperative and last postoperative measurement was used to calculate statistics.

All kind of stapes anomaly showed improvement in hearing, so the stapes status did not predict the long-term hearing. In the stapes fixation group, three children had some kind of hearing loss in family in extended family history survey, but none of these had otosclerosis diagnose. In these ears, there were no typical otosclerotic fixation focuses on the footplate, but the whole footplate was thick. The diagnosis remains unclear as only histological samples or CT scan could reveal true otosclerotic focus in the ante-fenestra region. Clinical audiometry revealed that six children had a clinically not significant (air conduction better that 25 dB) bass dominated conductive hearing loss also in the other ear. The Rinne test was negative for all but one ear. This ear showed combined hearing loss in clinical audiometry.

3.2.2. Incus group

Three ears showed only incudomalleal fixation. In this group one ear showed hearing deterioration and two had stable CHL.

Otoacoustic emissions showed various, non-consistent results. Stapes reflex tests showed as a rule negative reflexes, but many children disliked or refused the test and it was not possible to take the whole test set for most of the children. Tympanometry results showed an A type finding for all children.

4. Discussion

The cumulative evidence of hearing loss and problems in cognitive and academic thrive in life makes pediatric hearing problems important to detect and treat [8,9]. Unilateral conductive hearing loss without obvious reason is usually not severe. However, the hearing loss can be compared with HL found with secretory otitis media. Conductive hearing loss can be detected and treated. The treatment can be conservative with different kind of hearing aids and technical devices, like microphone system in a schoolroom and special education. The problem is that this kind of hearing loss is easily overseen in the newborn hearing screening, which is based on 35 dB general pass level. This problem is even more obvious if the screening is based on ABR, which measures discant weighted frequencies. Children are masters to adapt to hearing losses. The parents are righteously worried about the educational challenges for their children. It is frustrating for the clinician not to know the exact diagnosis or prognosis of conductive unilateral hearing loss. This study confirms earlier findings of stapes and incus anomaly and fixation. This study also shows that the major reason is stapes anomaly and this hearing loss showed tendency to improve spontaneously. The reason for this could be either the problems to measure exactly air and bone conduction for children or that the hearing improvement is real. The latter explanation is more plausible because of several reasons. All children were in school age and there were no problems to get the child to understand the study or to respond in a correct way to the air conduction test. Rinne test was negative. All audiometricians (audionoms) were used to test children and the hearing improvement was statistically significant. The air conduction thresholds improved, but the bone-conduction thresholds remained unchanged.

There are two theories for congenital fixation of annular ligament: it may fail to differentiate from the otic capsule at the lamina stapedius or it may develop normally and become ossified secondarily [5]. The results from this study supports the former theory. The incudostapedial joint did not show anomaly, but the footplate was as a rule thick. Earlier studies have shown variable results for pediatric otosclerosis [10,11]. This study could not confirm pediatric otosclerosis, as the diagnosis needs either CT or histological verification. However, the finding that HL was either stabile or improved makes the otosclerosis diagnose unlikely and supports the minor anomaly of stapes as the reason for CHL.

HL did not improve in the incus fixation group as the malleus was probably also fixed. The incus anomaly group showed deterioration in HL in only one ear.

This study shows that endoscopy as a minor procedure gives explanation to unilateral CHL and explorative tympanotomy is not necessary. Stapedotomy is not necessary in early age as the hearing loss has a tendency to improve spontaneously. Modern high-resolution CT scans still expose the child to radiation but do not always give diagnosis [12]. Endoscopy is therefore a good alternative, but CT remains a gold standard before more invasive ear surgery is planned. The study exposed a large variation of hidden reasons for CHL. Most of these can and should be treated at early stage. The disadvantage of endoscopy is the need of anesthesia and a minor risk for perforation or infection. In this study material these problems were minor. Compared with the large-scale use of tympanostomy tubes and similar risks for the procedures, this study justifies the unilateral CHL as indication to endoscopy. This study is in accord with earlier [2,5] and recent [13,14] reviews of the literature, but shows that endoscopy is a useful method for unilateral CHL and that stapes footplate related CHL has a tendency to improve spontaneously during long-term follow-up.

5. Conclusion

Endoscopy is useful and safe method for diagnosis and treatment plan for unilateral CHL without obvious reason.

There is no need for early CT scans. Wait and see is a good policy for stapes fixation.

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Declaration of competing interest

The author declares no conflict of interest.

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