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Audiological Findings in Children with Congenital Hypothyroidis (CH)

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Abstract: The incidence of congenital hypothyroidism (CH) in population of children amount to 1:3500 – 1:4000. The experiments on animals showed that the thyroid hormones play a considerable role in the proces of maturaton of the structures of the internal ear, and their deficiency increases the risk of hearing damage in children.

The aim of this study is not only the complex evaluation of the hearing organ in children with CH but also assessment of the correlation between results of audiometry and the time of initation of the therapy.

Material: The investigations included 111 children 3–16 years: the children were classified ito three groups: Group I – 75 children with congenital Hypothyroidism (CH), Group II – 3 children with transient hypothyroidism (TH), Group III – 6 children with Pendred Syndrome (PS)

Methods: Audiometric evaluation of hearing was carried out using tests of subjective audiometry, (pure tone audiometry and speachh discrimination if possible to perform) as well as tests of objective audiometry, that is, ABR, EOAE and tympanometric tests.

Results: Normal Threshold hearin values in PTA and ABR were observed in 79,4% of children from Group I and in 93,4% of children from Group II. In all 6 children from Group III the hearing impairment or complete deafness was noted.

Conclusion: Hearing impairment in children with CH dependes not only on the severity of the damage and its duration time but also on the child's age when the hormonal therapy is initiated.

Zusammenfassung: *Audiologische Befunde bei Kindern mit congenitaler Hypothyroidose (CH).* Die Häufigkeit einer angeborenen Schilddrüsenunterfunktion bei Kindern beträgt 1:3500 bis 1:4000. Untersuchungen bei Tieren haben gezeigt, daß die Schilddrüsenhormone eine beträchtliche Rolle bei der Ausreifung der Strukturen des inneren Ohres spielen und ein Mangel das Risiko einer Hörschädigung bei Kindern erhöht.

Das Ziel dieser Studie ist nicht nur die komplexe Bewertung der Hörfähigkeit bei Kindern mit Schilddrüsenunterfunktion, sondern auch die Bewertung der Korrelation zwischen den Ergebnissen der Audiometrie und des Zeitpunktes des Beginns der Therapie.

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Untersuchungsbedingungen: Die Studie bezog sich auf 111 Kinder von 3–16 Jahren; die Kinder wurden in drei Gruppen eingeteilt: Gruppe I – 75 Kinder mit angeborener Schilddrüsenunterfunktion, Gruppe II – 30 Kinder mit gelegentlicher Schilddrüsenunterfunktion, Gruppe III – 6 Kinder mit einem Pendred-Syndrom (erbliche Kombination von angeborener Innenohrschwerhörigkeit mit einem Enzymdefekt im Jodstoffwechsel).

Methoden: Bei der audiometrischen Messung wurden Meßmethoden der subjektiven Audiometrie (Audiometrie des Tonhörens und, wenn möglich, der Sprachdiskrimination) wie auch Methoden der objektiven Audiometrie verwand, also ABR, EOAE und tympanometrische Tests.

Ergebnisse: Normale Schwellenwerte beim Hören bei PTA und ABR wurden in Gruppe I bei 49,4% der Kinder beobachtet und bei 93,4% der Kinder in Gruppe II. Bei allen 6 Kindern der Gruppe III wurde eine starke Schwächung des Hörvermögens oder völlige Taubheit festgestellt.

Schlußfolgerung: Die Hörminderung bei Kindern mit angeborener Schilddrüsenunterfunktion hängt nicht nur von der Schwere und der Dauer der Schädigung ab, sondern auch vom Alter des Kindes, in dem mit der Hormontherapie begonnen wurde.

*

Introduction

Dysfunctions of the nervous system and hearing impairment are parts of the clinical pisture of hypothyroidism. They are observed in genetically determined hypothyroidism, Pendred syndrome [2, 3, 4], endemic cretinism [5] as well as in adults and children with congenital or acquired hypothyroidism [1].

Hoprman and Held [10] state the Pendred syndrome which is manifested by three symptoms, that is, congenital perceptive hearing loss, goiter and an abnormal perchlorate test is the most frequent of 124 manifestations of genetically determined deafness.

According to Kopp et al. [13] and Everrett et al., this syndrome refers to over 10% of inherited deafness. Elamin et al. [7] maintain that refers to 4–10% of inherited deafness and the cause of the hearing loss are bilateral malformations of the cochlea of Mondini type which was first confirmed in MRI by Nakagawa et al. [17] in 1994.

Iodine deficiency disorder (IDD) is a transient form of congenital hypothyroidism resulting from insufficient iodine intake by mother during pregnancy which leads to deficiency of iodine being a substrate in the thyroid hormone production.

Numerous experiments on animals [14, 15, 16, 17] showed that the thyroid hormones play a significant role in maturation of hypothyroidism increases the risk of hearing loss in children.

The aim of the retrospective complex audiological assessment of the hearing organ in children was:

1. To find the relationship between various forms of hypothyroidism and congenital deafnes.
2. To localize the site of defect in the cochlea or auditory pathways in case of hearing impairment.

Materials and Methods

The study included 111 children aget between 3 to 16 years treated in Endocrynological Department of Polisch-American Children Hospital, Collegium Medicum, Jagiellonian University in Kraków. Audiological tests were performed in the Audiological Laboratory of the Otolaryngological University Hospital CM Jagiellonian University.

The children were divided into three groups:

- Group I included 75 children with congenital hypothyroidism (CH); mean age 7.4 years.
- Group II included 30 children with iodine deficiency disorders (IDD); mean age 5.9 years.
- Group III included 6 children with Pendred syndrome; mean age 13.5 years.

The additional Group IV comprised 30 otologically healthhy children (60) ears in the same range of age with no past history of severe or chronic ear disease or the use of ototoxic drugs and with normal results of otological examinatons (normal otoscopy and middle ear pressure). Pure tone audiometry in all children of this group revelaed hearing threshold of 5 to 10 dB nHL in frequency range of 250 to 8000 Hz.

The complex evaluation of the hearing organ in each child was carrie out with the use of subjective pure tone audiometry and objective tests which registered:

1. Auditory brainstem responses (ABR), if necessary.
2. Click evoked otoacoustic emissions (CLick EOAEs).
3. Tympanometry

In these children hearing acuity was asswssed using pure tone average (PTA).

Taking into consideration the categories of hearing loss used by Hayes [9] and Hyde et al. [11], hearing impairment was indicated by hearing threshold greater than 10 dB in one or both ears. Hearing loss was considered mild (21–40 dB0), moderate (41–65 dB) and severe if hearing threshold was highter than 65 dB.

Results

The analisys of the mean hearing threshold values obtained in pure tone audiometry (PTA) and results of electrophysiological tests (ABR) showed that in the study group of 111 children, the correct hearing threshold was obserwed most frequently in Group II (IDD), that is, in 56 ears (93.34%) Table 1 whereas in Group I (CH) correct hearing threshold was found in 119 (79.33%) of 150 ears examined. Mild hearing loss in Group I was obserwed in 31 ears (20:7%) and in Group II only in 4 ears (6.7%).

Tympanometric examinations of children with CH in Group I revealed aut of 131 ears with mild hearing loss, 16 ears (10.7%) with sensorineural hearing loss (mean hearing threshold of 28.12 dB) localized in the cochlea and confirmed by the presence of stapedius reflex. Of 15 ears (10.0%) with conducyive hearing impairment in 12 ears (8.0%) it was caused by serous otitis media (SOM) confirmed by tympanomtry type B and mean middle ear pressure of 312 daP. In the rest of 3 ears (2.0%) the hearing impairment was found to be related to the Eustachian

Table 1. Hearing threshold in particular groups of children.

GROUP OF CHILDREN	NORMAL	HEARING LOSS		
	HEARING LEVEL	MILD	MODERATE	SEVERE
I-CH N=150 EARS	119 79,4%	31 20,06%	-	-
II-IDD N=60 EARS	56 93,4%	4 6,6%		
III-PENDRED,S SYNDROME N=12 EARS			3 25,0%	9 75,0%

Table 2. Threshold values PTA and EOAE in particular types of tympanograms.

Group I	Type of Tympanograms			EOAE Type of Tympanograms					
	A	B	C	A		B		C	
				+	-	+	-	+	-
Normal Hearing Level	101 67,4%		18 12,0%	99 66,0%	2 1,4%			16 10,7%	2 1,4%
Mild Hearing Loss	16 10,7%	12 8,0%	3 2,0%	3 2,0%	13 8,7%	2 1,4%	10 6,7%	2 1,4%	1 0,7%

tube dysfunctions; the mean thresholds value was 25 dB and middle ear pressure was 202 daP in tympanogram type C.

In Group II of children with IDD, in 56 ears the hearing was within normal values and only in 4 ears mild range conductive hearing impairment resulting from the Eustachian tube dysfunction was observed (Table 3) After pharmacological treatment, the hearing returned to the norm, In groups of children with CH and IDD, sewere sensorineural hearing impairment was no observed.

In Group III of 6 children with Pendred syndrome moderate hearing loss was observed in 3 children (5 ears); meann hearing threshold was 63.4 dB. In the rest of 3 children (6 ears) severe hearing loss woth mean threshold values of 85.4 db was found. Tympanometric examinations in this group of children confirmed in all ears normal functioning of the middle ear, that is, the presece of tympanogram type A and normal middle ear pressure, that is, not higher than 85

Table 3. Mean values of hearing threshold (PTA) and mean values of EOAE parameters.

Group II	*Tympanograms A*			*Tympanograms B*			*Tympanograms C*		
	PTA	EOAE		PTA	EOAE		PTA	EOAE	
N=60 ears	indB	% of reproduction	amplitude	indB	% of reproduction	amplitude	indB	% of reproduction	amplitude
Normal Hearing Level N=56 ears	6,6	92,01	19,55					-	-
Mild Hearing Loss N=4ears							26,25	-	-
Control Group N=60ars	5,0	94,80	22,80	-	-	-	-	-	-

daPa. In two children with moderate hearing loss, stapedius reflex was observed which confirmed cochlear localiztion of the hearing impairment. All the children with Pendred syndrome were equipped with hearing aids and reffered for auditory rehabilitation.

Evoked otoacoustic emission could be registered in 122 ears (81.33%) of children in Group I (CH); it was absent in 28 ears (18.66%), that is, in 4 earswith normal hearing threshold and in 24 ears with mild hearing loss and tympanometry type B and C. In Children from Group II (IDD), EOAE was registered in 51 ears (85%) in children with normal hearing level; their absence was found in 9 ears (15%) mainly in children with tympanometry type B and C. In none of the children from Group III EOAE was registered.

The analysis of the EOAE parameters in the study groups in comparison with the control group revealed that even slight increase in the hearing theshold decreases both the amplitude and percentage of otoemission reproduction.

Discussion

The results of the examinations show that 20% of children with CH have hearing impairment. In all children, disregarding the severity of hearing loss, only mild range of hearing impairment was observed. The hearing impairment in all children but one was bilateral and symmetric. The rate of the inner ear conductive hearing impairment was nearly equal to the rate of the inner ear sensorineural hearing impairment. The observed rate of hearing impairment in children with CH is consistent with this obtained by Rovet et al. [18] Deubryne et al. [6] and Vandershueren-Lodeweyck at al. [20] who reported mild hearing loss in 18 to 20% of cases but no cases of severe hearing loss. In the group of children with IDD abnormal hearing threshold was obserwed only in two children who had conduc-

tive mild hearing loss diagnosed. The main cause of hearing loss diagnosed. The main cause of hearing loss were Euystachian tube dysfunctions effectively treated pharmacologically with complete hearing restoration.

Coakley et al. [2], Jamal et al. [12], Cramers et al. [3] underline the progressive nature of hearing loss in patients with Pendred Syndrome which was confirmed by our investigations. The slightest hearing impairment of 55 dB was observed in the youngest 7-year-old boy, whereas the most severe hearing loss of 95 dB was noted in the oldest 16-year-old boy. Taking into account progressive nature of hearing loss, it seems that tympanometric check-ups are necessary in order to ensure the highest quality of life using hearing aids that all our patients with pendred Syndrome were supplied with.

In conclusion, it shold be stressed that

1. Complet audiological assesment with the use of subjective and objective audiometry allowed hearing evaluation in all 111 children, disregarding their age and co-operation abilities during examination.
2. Addition of the tympanometry to the battery of the subjective tests together with EOAE enables topodiagnosis of the hearing impairment in children.
3. In the diagnosed congenital hypothyroidism, the impairment of the hearing organ can occur on its various levels, that is, in the middle year, the cohlea or the extracochlear portion of the auditory pathway.
4. In our retrospective investigation of children treated for hypothyroidism we found 47 (21.17%) ears with various degree of hearing loss:
   - 31 (13.96%) ears of children with CH sensorineural usually bilateral symetric and mild range.
   - 4 (1.8%) ears of children with IDD have conductive hearing loss, which improved by appropriate pharmacological trreatment.
   - 12 (5.40%) ears of children with Pendred's Syndrom had moderate and severe sensorineural hearing loss atenuated by the hearing aid and auditory rehabilitation.

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