AUDITORY BRAINSTEM EVOKED POTENTIALS IN CONGENITAL HYPOTHYROIDISM SCREENING PROGRAM GRADUATES

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ABSTRACT

Auditory brainstem response (ABR) was used to detect the prevalence of hearing loss among 2-4 months old thirty infants with early treated congenital hypothyroidism (CH), diagnosed during the screening program. It was also used to correlate such prevalence to the severity of the disease at time of diagnosis as well as the time of starting treatment. Bilateral mild to moderate and unilateral mild hearing losses were found in 5 and 2 CH infants respectively. When the thirty infants with CH were compared to 12 controls as regards different ABR variants, wave I absolute latency was found to be significantly prolonged in CH group, while I-III, III-V and I-V inter-peak latencies were significantly shorter than norms. Moreover, wave V threshold was sig-

nificantly higher in CH group than that of norms. CH infants were divided into severe and less severe subgroups (based on the pre-treatment level of FT4) and only wave V absolute latency was found to be significantly shorter in the severe subgroup. No significant correlation was found between pre-treatment level of FT4 and different ABR variants except for wave V absolute latency (positive correlation). CH infants were also subdivided into late and early onset treatment subgroups (based on the age at which treatment with Na-L-thyroxine started) and only V/I amplitude ratio was significantly higher in early onset treatment subgroup. There was a significant negative correlation between age at which treatment started and V/I amplitude ratio. It can be concluded that children with CH have some im-

pairment of hearing which can be a significant handicap for spoken speech despite early treatment and despite the current lower prevalence of impaired hearing after the introduction of screening for CH; it seems that biochemical severity of CH at diagnosis has no significant effects on the severity of hearing impairment; and finally, start of treatment of CH at any time within the first month of life can be considered as early treatment.

INTRODUCTION

The function of the thyroid gland is to concentrate iodide from the blood and to return it to the peripheral tissues in the form of thyroid hormone (TH). TH has well known effects on the body metabolism and growth and development manifested during the first two decades of life ⁽¹⁾. It plays a determining role in growth and differentiation of the brain including neuronal proliferation, nerve cell migration and myelination which takes place up to the end of the third postnatal year ⁽²⁾. Consequently, a deficit in TH and/or in iodine during early life will result not only in general hypometabolism but also in brain damage, expressed clinically by irreversible retardation in the brain functions. An excess of thyroid hormones during the

critical period of brain development can also result in alterations in brain growth and mental development. Congenital hypothyroidism is defined as hypothyroidism present at birth or even during fetal life ⁽¹⁾.

The clinical picture of CH may include slightly increased head size, prolonged physiologic icterus, feeding difficulties, enlarged abdomen, hypothermia (less than 35°C), cold, dry, scaly and mottled skin with little perspiration, heart murmurs; anemia, stunted growth, short extremities, widely open anterior and posterior fontanelles, broad and depressed nasal bridge, narrow palpebral fissures, swollen eyelids, thick and broad tongue, delayed dentition, and thick short neck. Hypothyroid infants appear lethargic and are delayed in sitting, standing and talking with hoarse voice. These manifestations may progress and is fully developed by 3-6 months of age ^{(3) (4) (5)}.

The prevalence of hearing loss is about tenfold higher in infants with CH. Therefore detection of hearing defects in order to avoid difficulties related to speech development in late-diagnosed cases makes screening crucial ⁽⁶⁾. The purpose of newborn

screening is primarily to screen all newborn infants for a given disorder in which symptoms would not clinically present until irreversible damage occurred and of which an effective treatment is available (7). By mid-1982, after a recommendation by the department of health, routine screening had been introduced throughout the UK and since that time, neonatal screening programs for CH have been set up in most countries with advanced health care systems (8). Two screening methods are used to detect CH: the TSH and the T4 methods. TSH is the most frequently used, because it is cheaper and easier to perform than the T4 method (6).

Auditory brainstem response (ABR) has become a standard and valuable component of the audiologic test battery a long time ago (9). It is generally agreed that the ABR is generated by the auditory nerve and subsequent fiber tracts and nuclei within the auditory brainstem pathways. ABR is used clinically both in estimation of auditory sensitivity and in neuro-otological diagnosis. The popularity of the ABR stems from the fact that its characteristics are quite similar and stable between people, making the response fairly easy to identify under

most circumstances. Moreover, response do not vary between wakefulness and sleep and is not affected by most medications. This means that children may be tested reliably during natural or sedation-induced sleep (10). However, in neonates, ABR waveform consists primarily of three peaks, corresponding to waves I, III, and V and assumes an adult structure during the first 18 months of life (11). Wave I amplitude is larger while, wave V amplitude is smaller in infants than in adults. Therefore the wave V/I amplitude ratio will be reduced for infants, often having a value less than 1.0 (12). The latencies of the ABR components are longer in neonates and decrease progressively throughout the neonatal period because of maturation of the cochlea and brainstem. A decrease in body temperature below normal causes an increase in ABR inter-peak latencies (IPL) that may be explained by slowed neural conduction velocity and synaptic transmission speed in hypothermia (13). A correction factors has been published for I-V IPL to compensate for the effects of hypothermia (14). Furthermore, the circadian variation and decrease in body temperature by 1°C have found to prolong latency of wave V by 0.2 msec (10).

SUBJECTS AND METHODS

Subjects were divided into: 1) Study group: consisted of 30 infants with CH. Infants of high-risk register for hearing were excluded and infants were further subdivided according to their FT4 level into 10 infants with severe CH ($FT4 < 6$ pmol/L) and 20 infants with less severe CH ($FT4 \leq 6$ & ≤ 10 pmol/L). Again, infants were also subdivided according to the age for onset of treatment with Na-L-thyroxine into late onset treatment group (≥ 14 days, up to one month old [16 patients]) and early onset treatment group (< 14 days old [14 patients]). 2) Control group: consisted of 12 infants with their age and sex matched as much as possible to the study group and were involved in the CH screening program with negative results.

Methods : All infants included in this study were subjected to thorough history taking, laboratory investigations (for study group only including serum FT4 measured by direct equilibrium dialysis and serum TSH measured by an immunoradio-metric assay), and X-ray knee (for study group only to detect bone age through assessment of the ossific centers of lower femur and upper tibia). All

subjects in the study were also subjected to audiological assessment including otoscopic examination, tympanometry, and ABR for threshold determination in 20 dB steps or down to 30 dBnHL, which ever came first, using Bio-logic System (version 5.64, model 317) with headphones TDH 39. Absolute latencies of waves I, III and V, inter-peak latencies I-III, III-V and I-V and of waves V/I amplitude ratio, all were measured for 90 dBnHL, in addition to wave V threshold.

RESULTS

This study included 30 infants with CH (17 males and 13 females) and 12 control infants (6 males and 6 females). They were subjected to pediatric assessment when firstly seen during their first month and audiological assessment at the age of 2-4 months. ABR was done for both study and control groups. Of the 30 patients, only 59 ears were tested as one patient with less severe CH and late onset treatment aroused before test completion and could not be re-tested. Tests of differences between right and left ears of the control group regarding ABR variants had been done. There was no inter-aural significant difference, and hence the 12

control infants have been expressed as 24 ears.

Table (1) shows that 59 ears of the study group were tested and shows that wave V could be detected down to 30 dBnHL in 47 ears (79.7%) [17 (85%) of the severe group and 30 (76.9%) of the less severe one]. The table also shows that 11 ears (18.6%) had mild hearing loss and that only one ear (1.7%) had moderate hearing loss. Five patients had bilateral hearing loss: 4 of them had bilateral mild hearing loss and one had moderate hearing loss in the right ear and mild hearing loss in the left ear. Two patients had unilateral mild hearing loss.

Absolute latencies of waves I, III and V and inter-peak latencies I-III, III-V and I-V at 90 dBnHL recorded for CH infants were compared to those of the controls. Only wave I absolute latency was found to be statistically prolonged than that of the controls (Table 2) and I-V and I-III inter-peak latencies in the CH were found to be statistically shorter than those of the control groups (Table 3).

To elaborate more, we compared between the control group and the severe subgroup regarding absolute la-

tencies of waves III and V as well as III-V inter-peak latency at intensity of 90 dBnHL. No significant differences between the two groups were found (Table 4). Again, when comparing the control group to less severe one regarding absolute latencies of waves III and V as well as III-V inter-peak latency at 90 dBnHL, there was significant difference between the two groups regarding III-V inter-peak latency (III-V inter-peak latency of the less severe group was shorter than that of the control group) (Table 5).

As regard wave V/I amplitude ratio at intensity of 90 dBnHL, only 54 ears of the patients group were recorded, three of them were abnormally very high causing bias in the results, and hence had been excluded. There was no statistically significant difference between patients and controls. On the other hand, highly significant difference was found between the two groups as regard wave V threshold that was higher in CH infants (Table 6).

Tables (7), (8) and (9) show the statistical comparisons between severe and less severe groups of CH as regard absolute latencies of waves I, III and V, I-III, III-V and I-V inter-peak

latencies, amplitude ratio of waves V/I (at 90 dB nHL), as well as wave V threshold. No significant difference was found between the two groups except for absolute wave V latency which was shorter in the severe group more than the less severe one.

Tables (10), (11) and (12) show results obtained when comparing the late onset treatment group and the early onset treatment one as regard the ABR variants. Only amplitude ratio of waves V/I was significantly higher in the early treatment onset group.

Table (1) : Number (%) of normal and abnormal hearing tests in 59 ears tested in 30 infants with CH.

	Infants with congenital hypothyroidism		
	Severe (No.=20)	Less severe (No.=39)	Total (No.=59)
Normal hearing (Wave V threshold: 30 dB nHL)	17 (85%)	30 (76.9%)	47 (79.7%)
Mild hearing loss (30-50 dB nHL)	3 (15%)	8 (20.5%)	11 (18.6%)
Moderate hearing loss (30-50 dB nHL)	0 (0%)	1 (2.6%)	1 (1.7%)

Table (2): ABR absolute latencies (msec) for waves I, III and V among total patients group, as compared to control group using t-test

	Total patients group			Control group			t	P
	No	Mean	S.D.	No	Mean	S.D.		
Wave I	59	2.21	±0.58	24	1.62	±0.04	7.78	0.00
Wave III	59	4.56	±0.91	24	4.66	±0.12	-0.51	0.61
Wave V	59	6.42	±0.28	24	6.43	±0.16	-0.18	0.85

Table (3): ABR I-III, III-V and I-V inter-peak latencies (msec) among total patients group, as compared to control group using t-test

	Total patients group No.=59		Control group No.=24		t	P
	Mean	S.D.	Mean	S.D.		
I-III	2.35	±0.65	3.03	±0.09	-7.92	0.00*
III-V	1.65	±0.38	1.78	±0.16	-2.16	0.03
I-V	3.99	±0.87	4.81	±0.16	-6.9	0.00*

* Highly significant as $p < 0.01$

Table (4): ABR absolute latencies (msec) for waves III and V and III-V inter-peak latency among severe group, as compared to control group using t-test

	Severe group No.=20		Control group No.=24		<i>t</i>	<i>P</i>
	Mean	S.D.	Mean	S.D.		
Wave III	4.6	±0.5	4.66	±0.12	-0.54	0.59
Wave V	6.33	±0.21	6.43	±0.16	-1.92	0.06
III-V	1.76	±0.24	1.78	±0.16	-0.27	0.78

Table (5): ABR absolute latencies (msec) for waves III and V and III-V inter-peak latency among less severe group, as compared to control group using t-test

	Less severe group No.=39		Control group No.=24		<i>t</i>	<i>P</i>
	Mean	S.D.	Mean	S.D.		
Wave III	4.56	±1.12	4.66	±0.12	-0.43	0.67
Wave V	6.47	±0.3	6.43	±0.16	0.56	0.57
III-V	1.59	±0.43	1.78	±0.16	-2.45	0.02*

* Significant as $p < 0.05$

Table (6): ABR amplitude ratio for waves V/I and wave V threshold (dBnHL) among total patients group, as compared to control group using t-test

	Total patients group			Control group			<i>t</i>	<i>P</i>
	No.	Mean	S.D.	No.	Mean	S.D.		
V/I	51	1.8	±1.72	24	1.02	±1.46	1.87	0.06
Wave V threshold	59	34.41	±9.15	24	30	±0	3.7	0.00*

* Highly significant as $p < 0.01$

Table (7) : ABR absolute latencies (msec) for waves I, III and V among severe group, as compared to less severe group using t-test

	Severe group No.=20		Less severe group No.=39		<i>t</i>	<i>P</i>
	Mean	S.D.	Mean	S.D.		
Wave I	2.14	±0.39	2.25	±0.66	-0.71	0.48
Wave III	4.57	±0.22	4.56	±1.12	0.04	0.97
Wave V	6.33	±0.21	6.47	±0.3	-2.13	0.04*

*Significant as $p < 0.05$

Table (8) : ABR I-III, III-V and I-V inter-peak latencies (msec) among severe group, as compared to less severe group using t-test

	Severe group No.=20		Less severe group No.=39		<i>t</i>	<i>P</i>
	Mean	S.D.	Mean	S.D.		
I-III	2.43	±0.56	2.3	±0.69	0.69	0.49
III-V	1.76	±0.24	1.59	±0.43	1.64	0.11
I-V	4.19	±0.51	3.89	±1	1.22	0.23

Table (9) : ABR amplitude ratio for waves V/I and wave V threshold (dB nHL) among severe group, as compared to less severe group using t-test

	Severe group			Less severe group			<i>t</i>	<i>P</i>
	No.	Mean	S.D.	No.	Mean	S.D.		
V/I	13	2.09	±2.1	38	1.7	±1.62	0.58	0.57
Wave V threshold	20	33	±7.33	39	35.13	±9.97	-0.84	0.4

Table (10) : ABR absolute latencies (msec) for waves I, III and V among late onset treatment group, as compared to early onset treatment group using t-test

	Late onset treatment group No.=31		Early onset treatment group No.=28		<i>t</i>	<i>P</i>
	Mean	S.D.	Mean	S.D.		
Wave I	2.13	±0.72	2.3	±0.37	-1.12	0.27
Wave III	4.45	±1.24	4.67	±0.25	-0.92	0.36
Wave V	6.46	±0.3	6.38	±0.27	-1.04	0.3

Table (11): ABR I-III, III-V and I-V inter-peak latencies (msec) among late onset treatment group, as compared to early onset treatment group using t-test

	Late onset treatment group No.=31		Early onset treatment group No.=28		<i>t</i>	<i>P</i>
	Mean	S.D.	Mean	S.D.		
I-III	2.39	±0.65	2.37	±0.48	0.17	0.87
III-V	1.6	±0.49	1.7	±0.2	-1.11	0.27
I-V	3.92	±1.13	4.08	±0.54	-0.71	0.48

Table (12) : ABR amplitude ratio for waves V/I and wave V threshold (dB nHL) among late onset treatment group, as compared to early onset treatment group using t-test

	Late treatment onset group			Early treatment onset group			<i>t</i>	<i>P</i>
	No.	Mean	S.D.	No.	Mean	S.D.		
V/I	26	1.21	±1.45	25	2.42	±1.85	-2.14	0.04*
Wave V threshold	31	35.16	±10.3	28	33.57	±7.8	-0.66	0.51

DISCUSSION

It has long been recognized that CH can be associated with hearing impairment. The prevalence of hearing loss was found to be about tenfold high in infants with CH, and so audiological assessment is highly recommended during the first 2 months of life or not later than the age of 3 months (6) . A few studies have been carried out on children with sporadic CH before widespread introduction of neonatal screening and these have shown severe hearing impairment in 20-30% of cases (15) .

The current study included 30 CH infants who started treatment within the first month of their lives (17 males and 13 females) and the study also included 12 controls (6 males and 6 females). The results of this study indicate that children with CH have some impairment of hearing despite early treatment and a significant number may have elevated thresholds (more than 30 dBnHL), which can be a significant handicap for spoken speech (16) . Of the 59 ears tested in this study, there was hearing impairment in 12 ears (20.3%); 11 with mild hearing loss (18.6%) and one with moderate hearing loss (1.7%); 7 infants (23.3%) had hearing

impairment; 4 with bilateral mild hearing loss, 1 with mild hearing loss in one ear and moderate hearing loss in the other one, and 2 with unilateral hearing loss. These results are in agreement with Abo-El-Saad (17) who had detected hearing impairment in 9 infants (30%) from 30 infants with CH, despite early treatment. Of those 9 infants; 3 had severe to profound, 3 had moderate to severe, and 1 had mild to moderate high frequency sensorineural hearing loss (SNHL) and 2 had mild conductive hearing loss.

Similar results were also obtained when 101 children were studied retrospectively and followed longitudinally during screening for CH (18) . Of the 75 children who had their hearing tested, 15 children (20%) had hearing loss; 9 (12%) SNHL [1 unilateral, 8 bilateral], 5 (6.7%) conductive [2 unilateral, 3 bilateral] and one child (1.3%) had bilateral mixed loss. The hearing loss was mild in 10 cases (13.3%), moderate in 2 cases (2.7%) and moderately severe in 3 cases (4%). Also SNHL has been detected in 18 children (47%) from 38 children with early treated CH. Of the 76 ears tested, 25 ears (32.9%) showed mild hearing

loss and 2 ears (2.6%) showed moderate hearing loss (15). Our results are correlated to their results in that no severe hearing impairment was detected. Forty two early treated CH children were examined: 14 children (33.3%) had SNHL (12 had hearing loss of 31-50 dB at 8 kHz and 2 had bilateral mild hearing loss at conversational frequencies) (19). In addition, nine cases (14.3%) were reported to have SNHL from 63 children with CH (20).

In comparison with earlier studies carried out before the introduction of screening for CH, the current prevalence of impaired hearing appears lower. For example it has been found that over 50% of their subjects had some degree of SNHL (21), compared with 23.3% in this study; and bilateral SNHL in 20% of cases, compared with 16.7% in this study (22). Also, unlike these earlier studies, none of our cases showed evidence of severely impaired hearing. These results suggest that the effects of CH on hearing can be partially prevented by early treatment, even though relatively minor degrees of hearing impairment which are of less clinical consequence can still be documented.

This study showed significant prolongation of wave I latency (representing peripheral neural conduction) in infants with CH as compared to age and sex matched controls. Meanwhile, no significant difference was found as regards absolute latencies of waves III and V (representing central conduction) and similar results were also reported (17). This also comes in agreement with the findings that reported prolongation of only wave I latency in 11 CH newborns (19). Increased wave I latency was recorded in a group of 6 children with CH when they were examined before treatment with some improvement when they were reassessed 2 weeks after initiation of therapy (23). In 1987, the same teamwork examined 48 treated CH children and found that increased wave I latencies were the more frequent abnormalities (24). Also when 34 CH children under thyroid hormone therapy were evaluated, prolonged wave I latency was found in 7 children (20.6%) (25). On the other hand, abnormal ABR tracings were found in 8 patients (25%) from 32 patients with hyperthyrotropinemia diagnosed during neonatal screening (7 with CH and one with transient hypothyroidism) (26). However, of these 8 patients; 4 were with prolonged

wave I latencies and 4 were with prolonged wave III or V latencies. Again, (15) recorded 2 children (5.5%) from 36 children with early treated CH having prolonged waves I and V latencies above the upper normal departmental limits. Also, a significant delay in ABR absolute latencies in 20 patients with primary hypothyroidism had been also reported (27) .

Thyroid hormones stimulate calorigenesis and increase oxygen consumption through increasing transport of sodium and potassium across the cell membrane by the enzyme Na⁺, K⁺ ATPase (3) . A prolonged wave I absolute latency encountered in these studies can be attributed to affected cochlear metabolism due to CH, the effect which may not be corrected even with early treatment. On the other hand, prolongation of waves III and V absolute latencies found in some studies may be referred to a conductive element such as secretory otitis media which is common during infancy.

In this study, there was significant difference between infants with CH and controls as regard inter-peak latencies I-III, III-V, I-V which were shorter in patients than controls. The

shortness of inter-peak latencies I-III and I-V may be referred -in part- to the obvious prolongation of wave I absolute latencies as well as the possibility of the presence of recruitment, abnormal rapid growth of loudness, in the subjects with SNHL within the study group. The shortness of III-V inter-peak latency in patients was significant only in the less severe subgroup when they were studied apart from the severe one. These results concord with those reported shortened I-V inter-peak latencies in 10 children (29.4%) from 34 children with CH under therapy (25) . However, in another study, no significant difference between 48 congenital hypothyroid treated children and controls as regard ABR inter-peak latencies was found (24) . The same findings were reported by the same investigators in the year 1986 when they examined 6 patients with CH before initiation of therapy (23) . Contrary, others found significant delay in ABR inter-peak latencies when 20 patients with primary hypothyroidism were examined (27) . Moreover, 7 cases (20.6%) from 34 early treated CH children showed prolonged intervals between waves I and V (28) . Seven infants with CH were examined using ABR (29) . Results were abnormal in 3 of them who

showed bilateral conduction delays in caudal brainstem regions.

In the current study, no significant difference was found between CH infants and controls as regard waves V/I amplitude ratio. It also showed no significant difference between severe and less severe subgroups of infants with CH as regard all ABR recorded variants except for wave V absolute latency, which was shorter in the severe subgroup. While there was a trend for the children with more severe hypothyroidism to show increased hearing loss, the differences were not of statistical significance (15). This partially differs from what was found in this study, as the mean for hearing threshold in our patients was 33 dBnHL in severe subgroup compared to 35.13 dBnHL in less severe one, but without statistical significance. Moreover, these results are in agreement with those found no correlation between the ABR inter-peak latencies and the L-thyroxine serum values at the time of the test or just prior to treatment initiation (25).

On the other hand, reports showed significant partial correlation between ABR and thyroxine (T4) serum levels at diagnosis and TSH serum levels at

the time of recording (24). When 75 children with CH were studied, the children with hearing loss did not differ from the children without hearing loss in disease severity or duration (18). In adults, a relationship between ABR abnormalities and the degree of hypothyroidism was estimated (30).

In this study, when comparing late onset treatment group (14 days up to one month old) to early onset treatment group (before age of 14 days) regarding ABR variants, there was no significant difference between the two groups except for waves V/I amplitude ratio which was higher in the early onset treatment group. Of the 75 CH infants studied by Rovet et al (18); mean age of treatment onset in infants with hearing loss was 22 days, compared with 12 days in this study, and mean age of treatment onset in infants without hearing loss was 14 days, compared with 13 days in this study. In the current study, all infants with CH have actually started treatment within their first month, and hence there was no statistical difference between the two groups except for waves V/I amplitude ratio. This very narrow difference in the onset of treatment between the two subgroups is not appropriate to justify the statisti-

cal difference of waves V/I amplitude ratio by a maturational element. Also, waves V/I amplitude ratio is affected by many variables as electrode montage and age of the tested subject.

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الملخص العربى

لقد تم استخدام تسجيل استجابة جزع المخ للأصوات لمعرفة نسبة حدوث ضعف السمع فى ثلاثين طفلاً تتراوح أعمارهم بين ٢-٤ أشهر وتتم معالجتهم من نقص هرمون الغدة الدرقية الوراثى أثناء برنامج المسح الخاص لهذا المرض فى المواليد. ولقد تم استخدام استجابة جزع المخ للأصوات أيضاً فى إيجاد العلاقة بين نسبة ضعف السمع وشدة المرض عند التشخيص من ناحية وبداية العلاج من ناحية أخرى.

وتم إكتشاف ضعف سمع فى ٧ حالات (٥ حالات ضعف سمع بسيط إلى متوسط فى الجهتين و ٢ حالة ضعف سمع بسيط فى جهة واحدة). وأيضاً تم ملاحظة ببطء فى ظهور بعض موجات استجابة جزع المخ للأصوات فى مرضى نقص هرمون الغدة الدرقية الوراثى عنها فى الأطفال الأسوياء. وعند دراسة مدى تأثير شدة النقص الهرمونى على إستجابة جزع المخ للأصوات وجد قصر فى الموجة الخامسة فقط فى الأطفال شديدي النقص الهرمونى. وكما وجد أن النسبة بين قوة الموجة الخامسة للأولى تختلف فى الأطفال الذين تمت معالجتهم مبكراً عنها فى الأطفال الذين تأخر علاجهم.

وبذلك يمكن توقع حدوث لبعض حالات الخلل السمعى فى الأطفال الذين يعانون من نقص هرمون الغدة الدرقية الوراثى والذي يخشى من تأثيره على نمو الكلام عندهم .