ARTICLE

A Clinical Study of the Auditory Brainstem Evoked Potential in Down Syndrome Individuals

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Abstract

Sixty-one Down Syndrome and 31 control subjects were examined with a battery of audiological tests, including the Auditory Brainstem Evoked Potential (ABEP). Responses from a questionnaire, filled out by caregivers prior to testing of the Down Syndrome subjects, revealed that they were often unaware of the hearing status of the individuals in their care. The audiometric results showed a high incidence of hearing impairment in the Down Syndrome individuals (59%). A comparison of the ABEP results obtained for the normal hearing Down Syndrome group and the normal hearing control group pointed to a significantly shorter wave V absolute latency and wave I-V interpeak latency for the Down Syndrome group. When the ABEP is used in audiologic assessment of Down Syndrome clients, normative data from normal clients should be used with caution in the interpretation of results.

The audiology clinic at Glendale Lodge services residents of Vancouver Island with mental retardation or developmental delays. Many of our clients have Down Syndrome as it is one of the largest single causes of mental retardation, with an occurrence of approximately 1.5 per 1000 live births (Trimble & Baird, 1978). Previous research has pointed to a high incidence of hearing loss in the Down Syndrome population (Fulton & Lloyd, 1968; Balkany, Downs, Jafek, & Krajicek, 1979; Dahle & McCollister, 1986). The hearing loss can be of conductive, sensori-neural, or mixed origin. Early detection and remediation of the hearing loss are crucial for optimal speech and language development (Downs, 1980). In addition, Down Syndrome individuals may show evidence of premature aging, such as Alzheimer's disease (Zigman, Schupf, Lubin, & Silverman, 1987). Consequently, it is important to monitor hearing to ensure that they are not appearing confused because of a hearing loss.

Greenberg, Wilson, Moore, and Thompson (1978) found that most Down Syndrome children above 10 months mental age could be conditioned with Visual Reinforcement Audiometry. This was consistent with our impression that reliable test results were obtainable unless clients were very young, profoundly handicapped, or exhibiting behavioural problems. For these cases, the Auditory Brainstem Evoked Potential (ABEP) provides some objective information.

The decision to use ABEP in our clinic had been influenced by the perceived usefulness of the results. Our clinical experience had suggested that caregivers often attribute the lack of response to auditory stimuli as indicative of the client's functional level. Yarter (1980) found that only a small percentage of parents of children with Down Syndrome were aware of a hearing impairment. It was our concern that caregivers who do not recognize hearing loss may influence clinicians to abandon the special test procedure. Part of this study involved collecting information from caregivers to determine if they were able to predict the hearing sensitivity of their clients reliably.

In clinical work, the ABEP results from an individual are compared to data collected from normal hearing individuals. The normative data usually consists of peak and interpeak latencies (commonly waves I, III, V) at various intensities. The latencies and interpeak latencies are affected by such variables as hearing loss, gender, age, and rate of stimulus presentation (Picton, Stapells, & Campbell, 1981). Stockard, Stockard, Westmoreland, and Corfits (1979) indicated that the validity of the normative data is enhanced when the same sex and age matched controls are used.

ABEP testing gives a measure of hearing that is most highly correlated with thresholds between 2000 and 4000 Hz (Gorga, Worthington, Reiland, Beauchaine, & Goldgar, 1985). With a conductive hearing loss the latency of wave I and all subsequent peaks are prolonged. With a sensori-neural loss wave V has a normal latency at high intensity, but is not present or is prolonged at low intensity, resulting in a steeper latency intensity function (Galambos & Hecox, 1978; Coats & Martin, 1977). The ABEP's recorded from individuals with a steeply sloping high frequency hearing loss are difficult to separate from ABEP's obtained from individuals with conductive hearing losses as they have similar characteristics (Picton, Woods, Baribeau-Braun, & Healey, 1977).

With respect to gender, the wave V latency and the wave I-V interpeak latency are longer in adult males than females (Jacobson, Novotny, & Elliott, 1980; Jerger & Hall, 1980; Edwards, Squires, Buchwald, & Tanguay, 1981; Stockard, Stockard, Westmoreland, & Corfits, 1979). Aging has a clear effect on the latencies during the first 18 months of life. All the latencies and interpeak latencies decrease during that time

(Galambos & Hecox, 1978). The changes due to aging in older clients are much smaller or non-existent (Jerger & Hall, 1980; Beagley & Sheldrake, 1978).

Researchers have noted problems in using the ABEP with the mentally handicapped population. In some cases, the peaks were absent while quantifiable hearing was recorded (Cummings & Wells, 1986; Worthington & Peters, 1980). This has generally been reported in cases where there was severe neurological damage. Another problem in using ABEP has been reported in studies of the Down Syndrome population. These studies have shown that this group generally has shorter wave V latencies and wave I-V interpeak latencies as well as a steep latency-intensity function (Folsom, Widen, & Wilson, 1983; Squires, Aine, Buchwald, Norman & Galbraith, 1980; Squires, Ollo, & Jordon, 1986; Widen, Folsom, Thompson, & Wilson, 1987). Possible explanations for this difference have included structural brain differences (Squires et al., 1986) and cochlear abnormalities (Widen et al., 1987).

This study was undertaken to establish norms for our own clinical use and to assist other clinics in which individuals with Down Syndrome are seen less frequently. The study assessed differences in ABEP latencies between a group of Down Syndrome individuals with and without hearing impairment, and a control group with normal hearing. In addition, an attempt was made to determine the validity of the caregiver's perception of the hearing status to substantiate our impression that they underestimate the hearing loss in individuals with Down Syndrome.

Method

Subjects

All of the subjects were tested at Glendale Lodge. The Down Syndrome subjects were individuals with Trisomy 21 either living in the community or in this institution. Although some subjects had been seen previously, in most instances, the initial referral was not for a hearing test. Control subjects were of normal intelligence, either working at Glendale or attending university.

The mean age of all subjects and the Intelligence Quotient (IQ) of the Down Syndrome subjects is shown in Table 1. IQ data were obtained from medical records and were the scores on the most recent psychological tests administered.

Audiometric Assessment

All subjects were administered the audiometric assessment. Pure tone air conduction thresholds were obtained binaurally under headphones at 0.25, 0.5, 1, 2, 3, 4, 6, and 8 kHz using standard, play, or visually reinforced audiometry on a Grason-Stadler 1701 audiometer. Five decibel ascending steps and ten decibel descending steps were used. Where clinically indicated, bone conduction thresholds were obtained. Speech reception thresholds were obtained using spondees, picture board, and occasionally body parts.

Immittance audiometry was performed on all subjects using a Madsen ZO72 Electro-Acoustic Impedance Bridge. Tympanograms and contralateral (0.5, 1, 2 and 4 kHz) and ipsilateral (1 and 2 kHz) acoustic reflexes were measured using a pure tone stimulus. An otoscopic examination also was performed.

ABEP Assessment

All subjects were tested in a dimly lit, radio frequency shielded room while lying on a hospital bed. The caregiver for the Down Syndrome subjects was present throughout the test, and test equipment was in the same room so that the examiner was close to the subject. Subjects were instructed to lie still with their eyes closed and relax. No sedation was used.

Gold plated GRASS electrodes were placed on the forehead and mastoids with GRASS EC-2 electrode cream. The active electrode was on the forehead and the ground electrode was on the mastoid contralateral to the stimulated ear. Electrode impedance was 5 kilohms or less.

The ABEP was measured using a Cadwell 5200A Auditory Evoked Potential System. The stimuli were 100 microsecond rarefaction clicks presented at 22.2 per second through TDH-49 headphones with MX-41/AR cushions. Intensity was increased in 15 dB steps from 30 to 90 dBnHL. Stimuli were presented monaurally with 60 dBnHL of white noise masking in the contralateral ear. A 10 millisecond window and a band-pass filter of 100-3000 Hz were used.

Table 1. Age and I.Q. of subjects.

SUBJECTS		AGE			I.Q.	
		N	MEAN	RANGE	MEAN	RANGE
CONTROL	MALE	15	36	21-53		
	FEMALE	16	28	18-41		
DOWN SYNDROME	MALE	33	28	13-62	38	19-60
	FEMALE	28	26	5-53	42	18-71

Table 2. Audiometric findings.

SUBJECTS	NORMAL HEARING	HEARING LOSS
	NUMBER OF SUBJECTS	NUMBER OF SUBJECTS
CONTROL MALE	11	4
CONTROL FEMALE	16	0
TOTAL	27 (87%)	4 (13%)
DOWN SYNDROME MALE	12	16
DOWN SYNDROME FEMALE	11	17
TOTAL	23 (41%)	33 (59%)

One thousand clicks were averaged per waveform, and two or three replications were completed at each intensity. If two consistent waveforms could not be obtained at a particular intensity, no latency was recorded. Consistency was judged on a clinical basis by one of the authors (JL). If repeatable waveforms were obtained, they were averaged, and latencies were measured from the composite waveform. The latency of wave V was measured for each intensity; waves I and III and the wave I-V interpeak latencies were measured at 75 and 90 dBnHL.

Information was transferred from the ABEP unit to a Tandy 1200 HD computer and stored for future analysis. Total test time, including behavioural and ABEP, was approximately 75 minutes.

Results

Audiometric Test Results

The results of the audiometric testing are presented in Table 2. Subjects were divided into groups based on their audiometric findings. The normal hearing groups had pure tone thresholds up to 4000 Hz of 25 dBHL or better, binaurally.

Down Syndrome subjects with hearing loss were divided into three groups: conductive, sensorineural, and mixed. Subjects were placed in the conductive loss group, if there was abnormal immittance or the presence of a 15 dB or larger air bone gap; in the sensori-neural loss group, if there was normal immittance or no air bone gap; and in the mixed loss group, if there was a combination of conductive and sensori-neural loss. Table 3 shows this data. Control subjects with a hearing loss

Table 3. Hearing loss in Down Syndrome subjects.

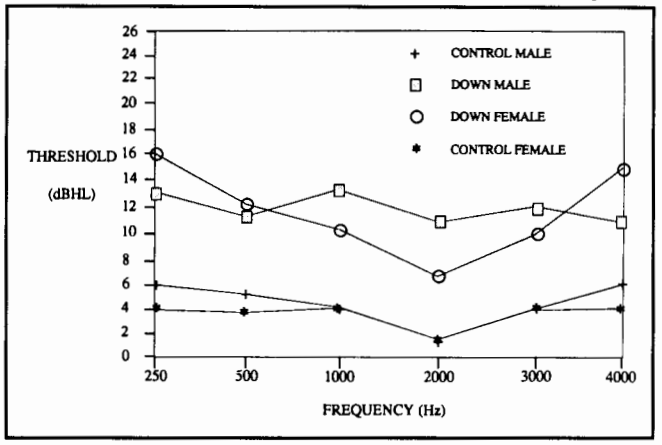
GROUP	N	MEAN AGE	AVERAGE HEARING THRESHOLD IN dB HL					
			250 Hz	500 Hz	1000Hz	2000 Hz	3000 Hz	4000 Hz
CONDUCTIVE	11	25	26	29	29	25	25	30
SENSORINEURAL	16	36	31	30	30	27	32	44
MIXED	11	29	32	34	40	40	51	54

Questionnaire

Before testing began, the caregivers of each of the subjects with Down Syndrome completed a questionnaire based on the *Speech and Hearing Checklist* from the *Information Kit on Childhood Hearing Impairment* (Durieux-Smith, Gibson, Shea, Schloss, Bernard, & Going, 1985). The questionnaire was designed to elicit observations of behaviours indicative of a hearing loss, even though the caregiver may have been unaware of the loss. The questionnaire is included in the Appendix.

were excluded from further analysis. There were seven Down Syndrome subjects from whom we were unable to obtain reliable pure tone results. Two of these seven were the only profoundly mentally handicapped individuals in the study; four were severely mentally handicapped; and one was moderately mentally handicapped. Five of these seven Down Syndrome subjects could not be assigned to either the normal hearing group or the hearing loss group on the basis of pure tone thresholds and impedance measurements.

Figure 1. Right ear thresholds for normal hearing subjects.



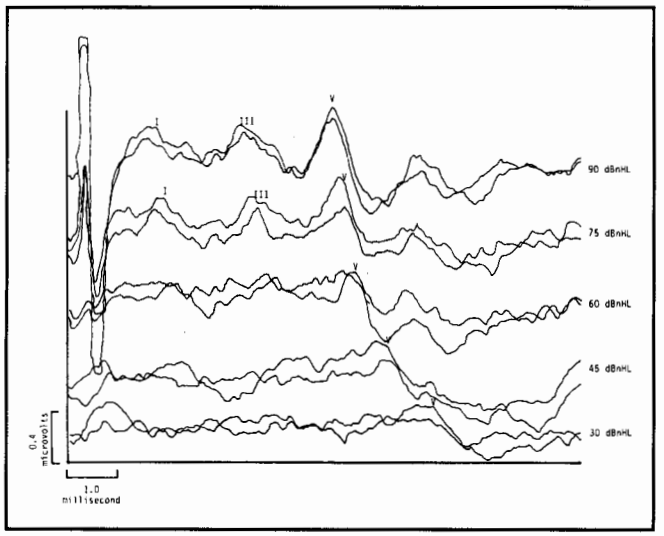
The thresholds of individuals with normal hearing are presented in Figure 1. A two-way ANOVA (group by gender) was done at each frequency, and there was a significant difference ($p<0.01$) between the Down Syndrome group and the control group. There was no significant main effect between males and females. The interaction was not significant.

ABEP Results

Typical ABEP waveforms for a Down Syndrome individual with normal hearing are shown in Figure 2. These show the usual decrease in amplitude and lengthening of latencies as the intensity of the stimulus is decreased.

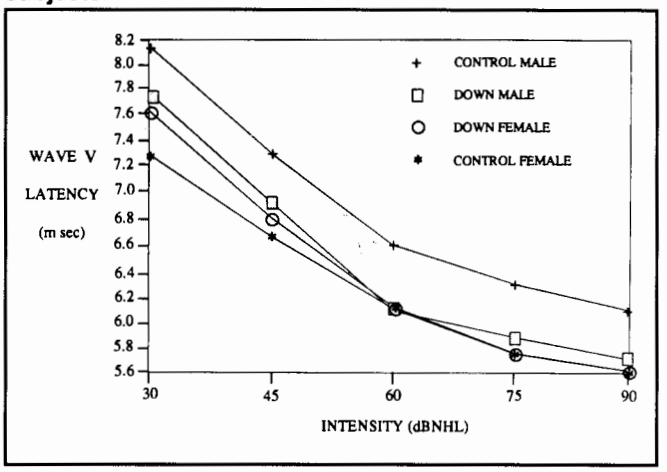
The average latency of wave V at all intensities for the normal hearing groups are shown in Figure 3. Two-way ANOVAs were done for data at 75 and 90 dBnHL (group by gender) and results were significant ($p<0.01$). Significant main effects for group and gender were found for both intensities. Controls had significantly longer latencies than the Down Syndrome group, and males had longer latencies than females. Post hoc testing was completed using *t* tests and showed that the latency of wave V was significantly longer in the normal hearing male control group than in the other three groups. No other post hoc tests were significant. The statistical findings for ABEP are detailed in Table 4.

Figure 2. Auditory brainstem evoked potential waveforms from a Down Syndrome subject with normal hearing.



The wave I-V interpeak latency (IPL) was measured, and two-way ANOVAs were done at 75 and 90 dBnHL (group by gender). Again, there were significant main effects ($p<0.01$).

Figure 3. Latency-intensity curves for normal hearing subjects.



At each intensity, control subjects had longer IPLs than the Down Syndrome subjects, and males had longer IPLs than females. Mean IPLs at 75 dBnHL for individuals with normal hearing are shown in Table 5 and are similar to those reported by Squires et al. (1986). Mean wave V latency for control subjects was calculated at each intensity. One standard deviation from the mean was used as normative data to compare with the mean wave V latency in Down Syndrome subjects with hearing loss. No statistical analysis was completed on subjects with hearing loss. The effect of hearing loss on the wave V latency was similar to that recorded in previous studies. The means of both the conductive and sensori-neural loss groups fall within the range of normal hearing for control subjects. As an example, Figure 4 shows the average latency of wave V at each intensity for the conductive hearing loss group superimposed on the normative data. It should be noted that there were no subjects with a conductive hearing loss that showed a wave V peak at 30 dBnHL.

As stated previously, there was no significant difference between the wave V latencies of the female control subjects and the Down Syndrome subjects, and therefore, the mean wave V

Table 4. Statistics for normal hearing control and Down Syndrome subjects: two-way ANOVA (group by gender).

	75 dBnHL							
	WAVE V LATENCY				CCT			
	p		*F*		*p*		*F*	
	RIGHT	LEFT	RIGHT	LEFT	RIGHT	LEFT	RIGHT	LEFT
GROUP	0.001	.000	14.16	16.810	0.006	.000	8.20	19.440
GENDER	0.000	.001	18.00	12.103	0.000	.005	18.19	8.770
INTERACTION	0.001	.028	6.99	5.033	0.020	.060	5.60	3.633
POST HOC-*t* TESTS ONLY RECORDED IF *P*<0.5	*p**		*t*		*p**		*t*	
	RIGHT	LEFT	RIGHT	LEFT	RIGHT	LEFT	RIGHT	LEFT
CONTROL MALE>DOWN MALE	0.005	.007	3.13	3.0052	0.016	.0062	2.56	3.05
CONTROL MALE>CONTROL FEMALE	0.002	.008	3.47	2.87	0.003	.02	3.36	2.47
CONTROL MALE>DOWN FEMALE	0.000	.0002	5.62	5.0781	0.000	NS	5.09	NS

*Values of *p* are for a two tailed test.

Table 4. (continued)

	90 dBnHL							
	WAVE V LATENCY				CCT			
	p		*F*		*p*		*F*	
	RIGHT	LEFT	RIGHT	LEFT	RIGHT	LEFT	RIGHT	LEFT
GROUP	0.002	.001	10.57	13.87	0.0001	.001	14.67	15.073
GENDER	0.000	.005	17.20	8.91	0.000	.029	19.40	4.99
INTERACTION	0.004	.016	9.27	6.11	0.027	.835	5.06	.041
POST HOC - *t* TESTS ONLY RECORDED IF *P*<0.5	*p**		*t*		*p**		*t*	
	RIGHT	LEFT	RIGHT	LEFT	RIGHT	LEFT	RIGHT	LEFT
CONTROL MALE>DOWN MALE	.005	.0088	3.07	2.91	0.008	NS	2.91	NS
CONTROL MALE>CONTROL FEMALE	.001	.0083	3.69	2.86	0.002	NS	3.43	NS
CONTROL MALE>DOWN FEMALE	.000	.0003	5.20	4.67	0.000	NS	5.90	NS

*Values of *p* are for a two tailed test.

latency and one standard deviation were calculated at each intensity for the female control subjects. All of the hearing loss groups fell outside of this normative range except for the sensori-neural loss group at high intensities. As an example, Figure 5 shows the average latency of wave V at each intensity for the conductive hearing loss group superimposed on female control normative data.

Questionnaire Results

Questionnaire results are presented in Table 6. The first question revealed that caregivers were not always aware of previous hearing tests. The caregivers of 15 out of the 45 subjects that had a previous test at Glendale were not aware of the test on record. The remaining questions produced relatively few "no" responses. As 33 of the Down Syndrome subjects had a hearing loss, it can be seen that only a small percentage of caregivers were noting "no" responses to these questions. In most cases, caregivers were attributing the "no" response to reasons other than a hearing loss.

Discussion

Questionnaire results supported our impression that caregivers were often unaware of the hearing status of Down Syndrome individuals. Down Syndrome individuals appear to relocate

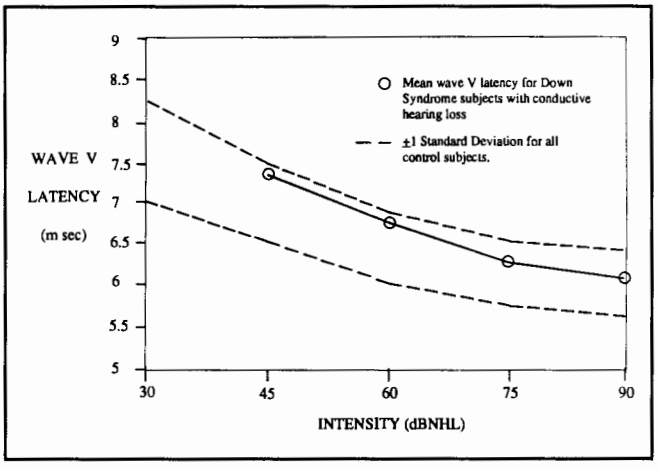
Table 5. Interpeak latency (IPL) at 75 dBnHL for subjects.

SUBJECTS	MALE	FEMALE
CONTROL	4.35	3.83
DOWN SYNDROME	3.94	3.79

often, and there is a substantial turnover of staff in homes for the mentally handicapped. Furthermore, often there are several caregivers for each individual. These were the most likely reasons that some of the caregivers were unaware of previous hearing testing. In order to provide the best service to this population, audiologists need to educate those working with the mentally handicapped about the importance of knowing the hearing status.

The questionnaire was designed to ask questions about tasks that even low functioning Down Syndrome subjects should be able to do, if they had normal hearing. The results indicate that either the subjects were able to do the tasks even with impaired hearing or that the reports of caregivers were inaccurate. It is our impression that it was a combination of the above reasons. There were individual subjects with a moderate to severe bilateral hearing loss of which the caregivers were totally unaware.

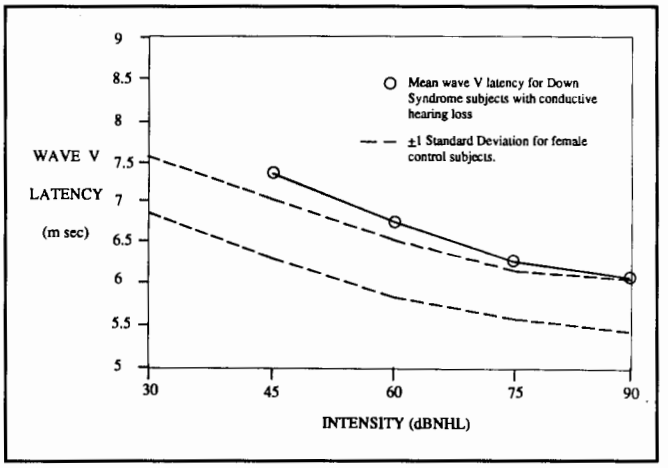
Figure 4. Mean latency wave V of Down Syndrome subjects with conductive hearing loss superimposed on normative data of all control subjects.



As indicated in previous research, there were relatively few Down Syndrome subjects that could not be tested with pure tone audiometry. In these cases, the results of our questionnaire data indicated that the clinician should not dismiss the possibility of completing an ABEP because the caregiver is making statements like "they hear well but they only hear what they want to." This is not to say that statements of the caregiver should be ignored or that the caregivers were not aware of the client's abilities, but that the audiologist should recognize the difficulty in determining hearing status without controlled test conditions.

In addition to the knowledge that caregivers often underestimate the degree of hearing loss, our results indicated that the Down Syndrome population is at high risk for hearing impairment. These two points should encourage the clinician to use ABEP when pure tone testing cannot be obtained.

Figure 5. Mean latency of wave V of Down Syndrome subjects with a conductive hearing loss superimposed on normative data of female control subjects.



The significant differences between the Down Syndrome group and the control group in wave V latency and in the IPL are of concern when using the ABEP clinically. Ideally, separate normative data should be collected for each clinic; however, this is not practical for most settings.

Our main concern is that a hearing loss will appear to be less serious than it really is. To avoid this problem, we recommend following the suggestions of Squires et al. (1986) and Jerger and Hayes (1976): Use the ABEP as part of a complete testing battery and never in isolation. In addition, we would agree with Widen et al. (1987) and recommend that audiologists observe the level at which wave V disappears in addition to the wave V latencies at higher levels. On the basis of our results, the use of female normative data would be more accurate than the use of male and female combined data for this population.

Table 6. Questionnaire results.

			RESPONSES	
QUESTION NUMBER	"YES"	"NO"	NO RESPONSE	"IF NO, IS IT DUE TO A HEARING LOSS?"
1	27	12	20	0
2	46	12	0	3
3	48	10	0	1
4	56	2	0	0
5	56	2	0	0
6	53	5	0	0
7	54	4	0	2
8	54	4	0	1
9	56	0	2	0
10	46	12	2	6

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Appendix

Questionnaire

Name_____

Form completed by_____

How long have you known him?_____

1. Has he had a previous hearing test? yes no
   If yes, where? _____
   and when? _____
   What were the results? _____
2. Does he respond to your voice when he can't see you? yes no
   If no, is this due to a hearing loss?
   or other cause _____
3. Does he turn to the source of sound? yes no
   If no, is this due to a hearing loss?
   or other cause _____
4. Does he acknowledge or react to environmental sounds (dog barking, telephone ringing, someones voice, his own name)? yes no
   If no, is this due to a hearing loss?
   or other cause _____
5. Can he point to familiar objects or people when asked? yes no
   If no, is this due to a hearing loss?
   or other cause _____
6. Can he point to body parts on request without gesture? yes no
   If no, is this due to a hearing loss?
   or other cause _____
7. Can he follow simple commands? (Bring me your ball.) yes no
   If no, is this due to a hearing loss?
   or other cause _____
8. Can he point to features of pictures in a book on request? yes no
   If no, is this due to a hearing loss?
   or other cause _____
9. Does he recognize meaningful sounds? (telephone) yes no
   If no, is this due to a hearing loss?
   or other cause _____
10. Does he respond appropriately when you call him from another room? yes no
    If no, is this due to a hearing loss?
    or other cause _____