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A 10-Year Review of Brainstem Auditory Evoked Response Testing at the Philippine Children's Medical Center: Patient Demographics and Outcomes

ABSTRACT

Objective: The study aims to present the clinical and demographic profile of subjects who have undergone Auditory Brainstem Response (ABR) test at the Philippine Children's Medical Center over a 10-year period.

Methods:

Design: Retrospective chart review
Setting: Tertiary children's hospital
Subjects: All patients referred for ABR testing from January 1996 to December 2005.

Results: A total of 2783 cases were included in the study with 1.63:1 male-to-female ratio. Almost 50% belonged to the 2-to 5-year-old age group. There were 111 different indications for referral, with speech and language disorders ranking first at 38%. Patients with Congenital Rubella had the highest incidence of pathologic ABR results with 90.62%. There was no significant difference in the degree of hearing loss between the pre-school (2-5 years old) and school age (>5 to 10 years old) group. Our patients who presented with speech delay had a much older average age of hearing loss detection by ABR compared to foreign studies.

Conclusion: Speech and developmental delays were the leading causes for ABR referral across age groups with most belonging to the 2-to-5-year-old age group. There was no statistically significant difference in the degree of hearing loss between the preschool and school-age groups with speech delay. ABR in hearing screening of neonates and children constitutes only a small fraction of the total indications for ABR Testing at the Philippine Children's Medical Center. Detection of hearing loss at an earlier age may reveal the true burden of illness and facilitate earlier intervention. Universal hearing screening should be performed for all newborns and not just for high risk infants.

Key words: *hearing loss, speech delay, Auditory Brainstem Response, ABR*

The American Speech-Language Hearing Association estimates the prevalence of newborns with congenital hearing loss in the United States at between 1 to 6 per 1,000.[1,2] The average age of detection in the pediatric population is between 12 and 25 months[3, 4, 5,6] with patients at risk and more severely impaired children being identified earliest. Children with no risk factors for hearing loss and children with mild to moderate hearing loss typically were not identified until about 28 months of age; with many undetected until identified at preschool and kindergarten

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hearing screening programs, or when hearing was tested because of concerns regarding speech, language and cognitive delays. The Joint Committee of Infant Hearing Year 2000 proposed that all infants born with hearing loss be screened by 1 month of age, diagnosed by 3 months and enrolled into early intervention by 6 months of age. This strong recommendation for early detection and intervention for infants with hearing loss cannot be overemphasized, as Yoshinaga-Itano and colleagues[7] showed that children in whom hearing loss was identified, and remediation instituted before 6 months of age had significantly higher scores on tests of vocabulary, expressive language and language comprehension than those diagnosed after 6 months. The screening tools of choice for newborns include Oto-acoustic Emission (OAE) and Auditory Brainstem Evoked Response (ABR) tests, with the latter being more reliable for the difficult to test and for very young patients.

As one of the first local institutions to acquire an Auditory Brainstem Response (ABR) machine, the Philippine Children's Medical Center (PCMC) has tested children from all over the country for the past 13 years. The study aims to present the clinical and demographic profile of subjects who have undergone Auditory Brainstem Response (ABR) test at the Philippine Children's Medical Center over a 10-year period.

RESEARCH OBJECTIVES

General Objective:

To present the clinical and demographic profile of subjects who have undergone ABR test at PCMC from January 1996 to December 2005.

Specific Objectives:

1. To present ABR patient distribution as to age, sex, working diagnosis (pertinent to ABR testing/ reason for referral), source of referral (physician's specialty);

2. To determine the incidence and degree of auditory pathway pathology for the ten most common working diagnoses/indications for ABR referral and compare them with existing international literature;

3. To rank the leading causes of referral for each age group over the 10-year period;

4. To determine the average age of auditory pathology detection by ABR among patients presenting with speech delay over the 10-year period;

5. To determine if there is a significant difference in the degree of hearing loss detected during the preschool (2-5 years old) and school age (more than 5 years old) group of patients with speech delay;

6. To determine the number of subjects 3 months and younger referred specifically for hearing screening and with no indicated risk factor/s for hearing loss; and

7. To present the most common reasons for referrals and degree of hearing loss in the neonatal age group.

Operational Definition of Terms

Hearing Loss- Hearing threshold greater than 35 dB
Mild Hearing Loss- Hearing threshold more than 35 to less than 45 dB
Moderate Hearing Loss- Hearing threshold at 45 to 65 dB
Severe Hearing Loss- Hearing threshold more than 65 to less than 85dB
Severe to Profound Hearing Loss- Hearing threshold at 85db or more

METHODOLOGY

A retrospective review was conducted on charts of all patients who underwent ABR testing at the Philippine Children's Medical Center between January 1996 and December 2005. Patients with incomplete data pertinent to the information being collected were excluded. Patient data of interest included the following:

1. Patient name and case number
2. Age
3. Sex
4. Working diagnosis pertinent to ABR testing/reason for referral.
5. Source of referral as to physician's specialty
6. ABR findings
   a. Degree of hearing loss (mild, moderate, severe, severe to profound)
   b. Laterality (unilateral/bilateral) of deficit

The subjects were sub-grouped according to age level as follows:

a. 0 to less than 24 months
b. 24 months to 5 years old
c. More than 5 to 10 years old
d. More than 10 to 15 years old
e. More than 15 to 18 years old
f. More than 18 years old

Clinical impressions of speech and language disorders were consolidated under Speech Delay. Global Developmental Delay and Psychomotor Delay were labeled under Developmental Delay while PDD (Pervasive Developmental Delay) encompassed clinical of impressions of PDD, Autism, Autistic Spectrum Disorder and Rett Syndrome. Descriptive statistics using means and proportions were applied. The z- test of association was used to analyze the difference in the hearing loss detected during the preschool (2-5 years old) from the school-aged group (>5 to 10 years old) of patients with speech delay.

The ABR machine test parameters used were the following:

Stimulus Parameters

Type:	Click
Duration:	100µsec
Rate:	15/sec
Polarity:	Alternating
Intensity:	(100) 90, 70, 50 and 30 dB
Transducer:	Elega TD 531

Acquisition Parameters

Amplification : None
Electrodes: Cz to Ipsilateral mastoid with forehead ground
Filter settings: 100-3000 Hz
Notch Filter: None
Number of Sweeps: 2000, replicated

Table 1. Gender and Age Group Distribution

	0-<24m	2-5y	>5-10y	>10-15y	>15-18y	>18y	Total
Female	343	490	177	37	2	7	**1057**
Male	480	912	290	36	2	7	**1726**
Total	**823**	**1402**	**467**	**73**	**4**	**14**	**2783**

Figure 1. Age Group Distribution

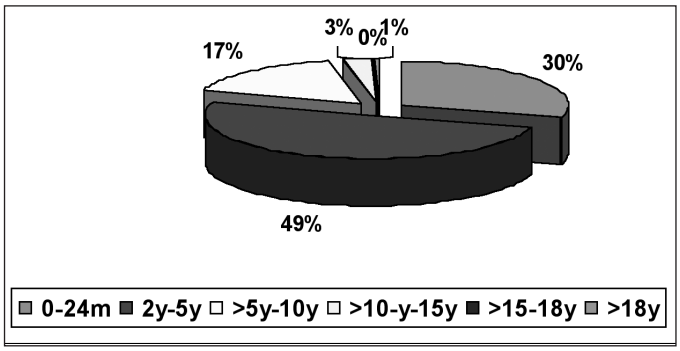
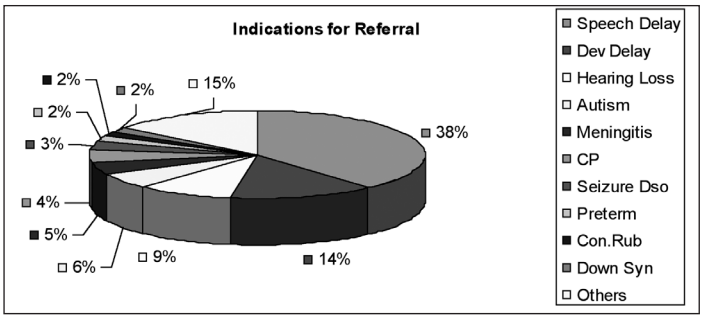


Figure 2. Percent Distribution of ABR Indications



The average age of auditory pathology detection was determined by taking the total age in months of the affected patients at the time of detection divided by the total number of affected patients.

RESULTS

A total of 2783 patients were included in the study with an overall female to male ratio of 1:1.63 *(Table 1)*. The 2-to-5 year-old age group was largest, making up 49% *(Figure I)*. There were 111 total listed indications for referral. Close to 85% of the total cases were shared by the top 10 clinical entities; of these, 38% were represented by the Speech Delay Disorders *(Appendix I, Figure 2)*. The remaining 15 % were shared by 101 other indications. Sixty-one percent of the referring physicians were residents and fellows, child neurology consultants contributed sixteen percent 16% followed by neurodevelopmental pediatricians and otolaryngologists with 8 and 4%, respectively *(Figure3)*.

The incidence and degree of pathologic ABR results for the 10 leading indications are listed in *table 2*. *Table 3* shows comparative results with selected foreign studies on these 10 indications of referral. No direct comparison could be found in published studies regarding ABR results of patients with seizure disorder per se and patients primarily suspected for hearing loss. *Table 4* shows the most common causes for ABR referral for each age group. Global developmental delay was the primary reason for referrals in subjects under 2 years old, followed by speech delay. Bacterial meningitis was also a notable cause in this age group. Speech delay was the leading indication for ages 2-10 years old while hearing loss dominated the older age group.

The overall average age of auditory pathology detection in speech-delayed patients was 46.48 months, with no trend observed in the yearly average age of auditory pathology detection except for a peak in 2002 (*Appendix 2, Figure 4*). For all degrees of hearing loss, there was a slight increase in the >5-10-year-old age group compared to the 2-5-year-old age group *(Table 5)*. These differences were not statistically significant using the z- test of association *(Appendix 3a, 3b)*.

Table 2: Incidence and Degree of Auditory Pathology of Top 10 Ranked ABR Referrals

Indication for Referral	Total		Results (Ears tested)										Total Incidence	
	Patients Tested	Ears Tested	Intact		Mild HL		Moderate HL		Severe HL		Severe to Profound HL		N	%
			N	%	N	%	N	%	N	%	N	%		
Speech Delay	**1073**	**2146**	**1350**	**62.91**	**90**	4.19	**142**	6.62	**119**	5.54	**445**	20.74	**796**	37.09
GDD	**391**	**782**	**530**	**67.77**	**45**	5.75	**66**	8.44	**26**	3.32	**115**	14.71	**252**	32.23
HL	**259**	**518**	**139**	**26.83**	**10**	1.93	**20**	3.86	**34**	6.56	**315**	60.81	**379**	73.17
Autism	**163**	**326**	**303**	**92.94**	**10**	3.30	**6**	1.98	**0**	0	**9**	2.97	**25**	7.67
Meningitis	**128**	**256**	**165**	**64.45**	**19**	7.42	**20**	7.81	**7**	2.73	**45**	17.58	**91**	35.55
CP	**124**	**248**	**137**	**55.24**	**6**	2.42	**16**	6.45	**2**	0.81	**87**	35.08	**111**	44.76
Seizure	**92**	**184**	**152**	**82.61**	**7**	3.8	**8**	4.35	**3**	1.63	**14**	7.61	**32**	17.39
PT	**55**	**110**	**61**	**55.45**	**6**	5.54	**9**	8.18	**6**	5.54	**28**	25.45	**49**	44.55
Con. Rubella	**48**	**96**	**9**	**9.38**	**5**	5.20	**5**	5.20	**12**	12.50	**65**	67.71	**87**	90.62
Down	**47**	**94**	**44**	**46.81**	**9**	9.57	**22**	23.40	**4**	4.26	**15**	15.96	**50**	53.19

There were 57 (2%) patients aged 3 months and younger in the study, 4 of these were well babies, specifically referred for hearing screening. It is interesting to note that in patients 6 months and under, 7 were referred as follow ups to a "refer" result of a previous otoacoustic emission test.

Only 6 (0.22%) neonates were referred, 2 for aural atresia and 1 each for bacterial meningitis, prematurity, cerebral palsy and sepsis.

DISCUSSION

Speech and developmental delays were the leading causes for ABR referral across age groups with most (almost 50%) belonging to the 2-to-5-year-old age group. Because intelligible speech is expected from a child of this age, verbal delay prompts parents and guardians to consult. Abnormal ABR results for speech delay in our study were more than twice those of a European study.[8] Congenital Rubella syndrome presented with the highest incidence of abnormal ABR results at 90.62 %, slightly higher than that published by Roizen in 1999[9] while Niedzielska[10] and Sadijhi, J. *et al*[11] reported a much lower incidence. Our results for hearing loss in children with global developmental delay fell within the wide range of other studies exemplified by Haggard[12] and Rupa.[13] Our incidences for autism were 5.28% and 2.97% for mild to moderate, and severe to profound hearing loss, respectively, lower than the 7.9% and 3.5% reported by Rosenhall[14] in a European population. Taylor[15] similarly showed a higher incidence in his study considering only autistic children without associated features.

Bacterial meningitis was the most common reason among the infectious causes for ABR referral. The incidence of hearing loss as a sequel of bacterial meningitis were reported at 10% by Tarlow[16] and 7% by Koomen,[17] much lower than our results which were similar to those conducted at a children's hospital in Nepal.[18] Our results of more than 50% abnormal ABR in Down syndrome support the consistently high incidence of hearing impairment in this clinical entity as diagnosed by ABR.[19,20] We had a 44.55% incidence of abnormal ABR in cerebral palsy patients, twice higher than the findings of Robinson[21] and Zafeiriou[22] and much higher than the 2-6% average incidence of hearing loss in the preterm infant population reported elsewhere.[23] It is possible that co-morbid conditions in a majority of our patients with cerebral palsy may have increased the incidence of abnormal ABR results. While the incidence of abnormal ABR results for developmental delays, post bacterial meningitis, Down syndrome and congenital Rubella were at par with foreign statistics, our incidence for autism was slightly lower. The myriad etiologies for, and different classifications of seizure disorders precluded making any direct comparisons with the literature.

No trend was noted on the average age of hearing loss detected in speech-delayed patients with respect to time; the peak average in year 2000 was attributed to an adult patient tested. Our results showed that we are 21 to 34 months behind the world's average age of detecting hearing loss in the pediatric population using speech delay as the presenting symptom. Reasons for late consultation may include a lack of awareness of the early signs of hearing loss on the part of parents and guardians. On the other hand, significant observations of concerned parents may have been disregarded by well-meaning health care givers

Table 3. Comparative Table on the Incidence of Patholigic Results of Top 10 Ranked ABR Referrals

RANK	INDICATION	PCMC	FOREIGN STUDIES
1	Speech Delay	37.09	13.3 (Psarommatis I.M. et al, 2001)
2	GDD	32.23	18 (Haggard M. 1992) 91 (Rupa V. 1995)
3	HL	73.17	**
4	Autism	7.96	9.5 (Rosenthal et al, 1999) 18.75 (Taylor et al, 1982)
5	Meningitis	35.55	10 (Tarlow, 1997) 36 (Kanti Children's Hospital, 1984) 7 (Koomen I., Grobbee et. al 2003)
6	CP	44.76	20 (Robinson, 1983) 22.7 (Zafeiriou, 1999)
7	Seizure	17.39	*
8	PT	44.55	2-6 (JCIH, 1994)
9	Down	53.19	66 (Roizen, 1997) >75 (Cunningham & McArthur 1981)
10	Con Rubella	90.62	50 (Neidzieska, 1999) 90 (Roizen, 1999) 12 (Sadijhi,J. et al, 2004)

Figure 3. Distribution of Referral Source

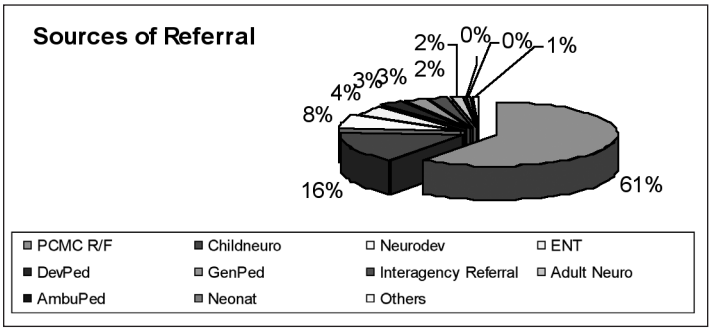


Figure 4. Yearly Average Age of Auditory Pathology Detection in Patients Presenting with Speech Delay

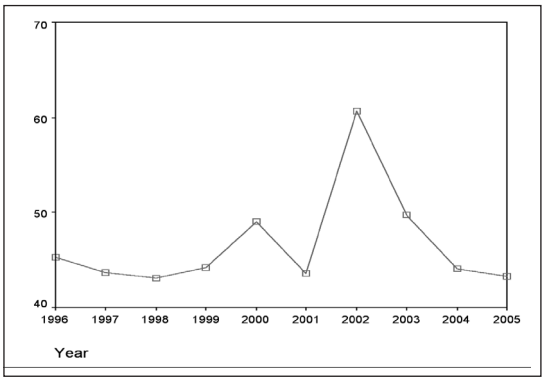


Table 4. Ranked Leading Causes of Referral for Each Age Group

	0-<24 Mos		2-5 y/o		>5-10 y/o		>10-15 y/o		>15-18 y/o		>18 y/o	
Rank	CoR	N	CoR	N	CoR	N	CoR	N	CoR	N	CoR	N
1	GDD	153	SD	754	SD	183	HL	16	HL	2	HL	3
2	SD	118	GDD	173	HL	67	SD	15	-	-	-	-
3	Men	83	PDD	125	PDD	61	GDD	8	-	-	-	-
4	CP	69	HL	113	GDD	56	MR	8	-	-	-	-
5	HL	59	CP	47	MR	27	Men	4	-	-	-	-
6	Sz	33	Sz	37	Sz	18	PDD	3	-	-	-	-
7	PT	31	Men	33	Men	11	OM	3	-	-	-	-
8	ConRu	29	CoRu	15	CP	6	Sz	3	-	-	-	-
9	DoSy	29	Dosy	14	HyBil	5	-	-	-	-	-	-
10	HyBil	27	HyBil	9	CoRu	3	-	-	-	-	-	-
					HCP	3						
					OM	3						

Legend:

CoR	Cause of Referral	Sz	Seizure Disorder
GDD	Global Developmental Delay	PT	Prematurity
SD	Speech Delay	CoRu	Congenital Rubella
Men	Meningitis	DoSy	Down Syndrome
CP	Cerebral Palsy	HyBil	Hyperbilirubinemia
HCP	Hydrocephalus	OM	Otitis Media
HL	Hearing Loss	MR	Mental Retardation

PDD-Pervasive Developmental Delay

Table 5. Degree of Hearing Loss in Preschool (2-5 y/o) and School Aged (>5-10 y/o) Group with Speech Delay

Age Grp	Patients Tested	Ears Tested	Result (Ears Tested)									Total	%
			Inact	Abnormal Auditory Pathway									
				Mild		Mod		Severe		Sev-Pro			
			N	N	%	N	%	N	%	N	%		
2-5yo	760	1520	1006	59	3.88	87	5.72	89	5.86	279	18.36	514	33.82
>5-10yo	183	366	200	20	5.46	31	8.47	23	6.28	92	25.14	166	45.36
P value				>.05		>.05		>.05		>.05			

Appendix 1. Percent Distribution of ABR Referral Indications

Cases	SD	DD	HL	PDD	Men	CP	Sz D	PT	CR	DS	OI	Total
Female	349	185	113	32	51	59	35	20	25	21	166	1056 (38%)
Male	724	206	146	131	77	65	57	35	22	26	240	1729 (62%)
Total *(%)*	1073 *38.53*	391 *14.04*	259 *9.30*	163 *5.85*	128 *4.60*	124 *4.45*	92 *3.30*	55 *1.97*	47 *1.69*	47 *1.69*	406 *14.58*	**2785**

Legend:

SD	Speech delay	Sz D	Seizure Disorder
DD	Developmental Delay	PT	Prematurity
HL	Hearing Loss	CR	Congenital Rubella
PDD	Pervasive Developmental Delay	DS	Down Syndrome
Men	Meningitis	CP	Cerebral Palsy
OI	Other Indications		

who downplayed the possibility of hearing loss. Costs associated with hearing screening may also contribute to delay. Although it may be posted that a more severe degree of hearing loss in younger patients prompts earlier referral than among school-aged children in whom it was usually an incidental finding, this was not the case in our study. Further, there was no statistically significant difference in the degree of hearing loss between the preschool and school age groups with speech delay.

Less than 1% of the overall indications for referral were for follow-up of failed hearing screening and only 2% of the subjects were 3 months old and younger. These values may reflect the minimal usage of ABR for hearing screening in our institution as well as a lack of awareness of the importance of newborn hearing screening in general. This may be exacerbated by the absence of a mandate to screen all newborns for potential hearing loss, and not just the high risk group as traditionally done. An important study among patients with congenital hearing loss showed that 50% did not have any risk factors.[24] Apart from delayed cognitive complications of hearing loss, economic and societal repercussions are not to be disregarded. It is estimated in an American study that households lose $122 billion in lost income and reduced federal tax revenues by another $18.4 billion.[25]

ABR in hearing screening of neonates and children constitutes only a small fraction of the total indications for ABR Testing at the Philippine Children's Medical Center. Detection of hearing loss at an earlier age may reveal the true burden of illness and facilitate earlier intervention. Finally, to improve on the poor average age of hearing loss detection so that subsequent remedial measures are administered, it is strongly recommended that universal hearing screening be performed for all newborns and not just for high risk infants.

Appendix 2. 10-year average age (months) of speech delay diagnosis

	1996	1997	1998	1999	2000	2001	2002	2003	2004	2005	Overall
Cases	112	87	166	73	100	93	94	127	122	99	**1073**
Average	45.28	43.63	43.1	44.15	49.05	43.56	60.66	49.69	44.03	43.31	**46.48**

Appendix 3a. Cross tabulation of preschool and school-aged group with speech delay

			Ears Tested (Right and Left)				Total
			Mild	Mod	S	S-P	
AGE GROUP	2-5 y/o	Count	59	87	89	279	514
		% within AGEGROUP	11.5%	16.9%	17.3%	27.15%	100.0%
		% within Ears Tested (Right and Left)	74.7%	73.7%	79.5%	75.4%	75.6%
	>5-10 y/o	Count	20	31	23	46	166
		% within AGEGROUP	12.0%	18.7%	13.9%	27.7%	100.0%
		% within Ears Tested (Right and Left)	25.3%	26.3%	20.5%	24.6%	24.4%
Total		Count	79	118	112	371	680
		% within AGEGROUP	11.6%	17.4%	16.5%	27.25%	100.00%
		% within Ears Tested (Right and Left)	100.0%	100.0%	100.0%	100.0%	100.0%

Appendix 3b. p Value using z-test of association

	Mild	Mod	S-P	SP
2-5 y/o	11.47859922	16.92607004	17.3151751	22.95719844
>5-10 y/o	12.04819277	18.6746988	13.85542169	21.68674699
Difference	-0.56959355	-1.748628756	3.459753411	1.270451456
p value	>.05	>.05	>.05	>.05
Conclusion	not significant	not significant	not significant	not significant

Appendix 4. Patient Data Collection Form

YEAR									
Case No	Name	Age	Sex	Indication	Right Ear		Left Ear		Referring Physician (Specialty)
					Threshold (dB)	Final Reading	Threshold (dB)	Final Reading	

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