

Necessity of Revising the Evaluation Protocols of Syndromes with Hearing Loss

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During a research in 2015, author of this paper found a patient with Noonan syndrome (NS) with a problem in middle ear but with delay in speech despite this fact that he was 5 years old (according to his profile). There was an inconsistency between the results of audiological tests and this fact that the patient had a delay in speech. For this purpose, the patient invited and set an appointment for him on 2nd July 2015. The patient referred to audiology clinic and audiological tests were done for him after written informed consent by parents.

It was the first time that I could test a patient with NS. Before the day of appointment, I read his profile very carefully and later I searched about the results of audiological and auditory electrophysiological tests but surprisingly I could not find anything in literatures. That is, after 1962 the year which NS identified by Jacqueline Anne Noonan, all of the researches about NS focused on many features of this syndrome and unfortunately hearing loss has not been considered. There are some reports in articles [1-6] about the presence of hearing loss and malformations in hearing system of patients with NS. In addition, there was not any paper or even one paragraph in the literatures regarding the analysis of the results of electrophysiological tests in NS.

By the way, audiological tests were done and the results showed a conductive hearing loss (CHL) but the results of auditory brainstem response (ABR) was atypical [7]. Here should be mentioned that CHL in this patient occurred at low frequencies that might not affect the ABR waveforms with clicks stimulation [8]. By the way, analysis of the ABR results has suggested me that some deformities/malfunctions in his CANS that has not been discussed before. Although, ABR recordings showed a good morphology but surprisingly, the IPL of III-V was abnormally shorter in NS child than in the normal subject, meanwhile the near-normal IPLs of I-V is contributed by the slight delayed IPLs of I-II and even more prolonged IPLs of II-III (Table 1).

Ear	Polarity	Absolute latency V (ms)	Interpeak latency I - V (ms)	Interpeak latency I - III (ms)		Interpeak latency III - V (ms)
				I - II (ms)	II - III (ms)	
Right	Rare	5.60 (5.59)	4.13 (3.98)	1.13 (0.96)	1.50 (1.13)	1.50 (1.89)
	Cond	5.54	4.00	1.13	1.25	1.62
	Alt	5.54 (5.51)	4.00 (3.85)	1.06 (0.94)	1.38 (1.08)	1.56 (1.83)
Left	Rare	5.67 (5.64)	4.25 (3.93)	1.06 (0.9)	1.19 (1.12)	2.00 (1.91)
	Cond	5.67	4.25	1.13	1.25	1.87
	Alt	5.60 (5.58)	4.19 (3.86)	1.12 (0.96)	1.38 (1.06)	1.69 (1.84)

Table 1: Absolute latency and interpeak latencies of ABR peaks for the NS child and the respective normal child (in bracket) in left and right ears.

Here it should be mentioned that in addition to ABR, there are at least 4 or 5 other auditory electrophysiological tests (such as auditory middle and late responses and auditory event related potentials) that could be done for this child. For instance, the authors have recorded the speech evoked-ABR (sABR) test for this NS child and comparing the results with normal male children sABR (Figure 1) indicated that atypical changes in morphology and amplitudes of the peaks especially in frequency following response (FFR) component.

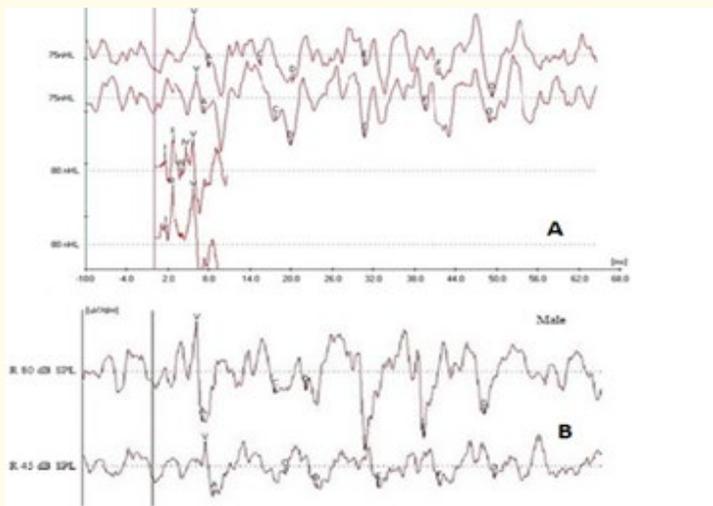


Figure 1: Comparison of the results of sABR in the patient with NS (A) with a sABR recording of a child with normal hearing (B).

Recall that there are also many syndromic disorders like NS that hearing loss is one of the signs of those syndromes and still many of the aspects in relation with their malformations/malfunctions of CANS in the patients with those syndromes have not yet discovered. In the author's opinion, attention to some aspects of syndromes should not lead to a lack of attention to other aspects. Certainly there is the right of these patients to be able to use the abilities of perceptive and expressive language, but before that we need to fully understand the functioning of the CANS of these patients and their defects. It has been documented that delay in speech can be resultant of a delay in the maturation of the nervous system as well [9]. Therefore, it is suggested that the Evaluation Protocol for the patients with these syndromes should be revised, that is, performing auditory electrophysiological tests is required for these patients that they can benefit from all-inclusive services.

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