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Cognitive profile of a child with SOS1 mutation in Noonan syndrome☆

Perfil cognitivo de un niño con síndrome de Noonan con mutación genética SOS1

Dear Editor:

We present the neuropsychological profile of a 7-year-old boy with Noonan syndrome (NS) and *SOS1* mutation.[1] Studies on NS usually present a general, non-specific image of cognitive impairment; in our opinion, this is due to the limited number of studies and the small samples included, which do not accurately represent the variability of the condition, since they group together patients with different mutations.[2—5] The case we present is interesting due to the scarcity of literature on the differential cognitive characteristics observed in this mutation, in comparison with the disability reported in other, more frequent mutations.

We present the case of a boy who was diagnosed with NS at the age of 4 by means of the polymerase chain reaction method. The medical history suggested normal development of motor, communication, and language skills.

We assessed his performance in several cognitive processes. His general ability index was 131, and composite score (CS) was 98 on the Wechsler Intelligence Scale for Children (WISC-IV).

Attention. The patient obtained normal results in information processing speed (Conners' Continuous Performance Test II [CPT-II] version 5: CS = 61) and in cue-based tasks in WISC-IV (scaled score [SS] = 11). He scored poorly (SS = 6) in the symbol search subtest, which indicates difficulties in visual perception, confirmed by scores below the mean on the Illinois Test of Psycholinguistic Abilities (ITPA) (*T*-score = 26). Variability of response time to stimuli (CPT-II: SS = 51) and attentional vigilance (CS = 50) were normal. Results for selective and focused attention (CPT-II) were normal, but with many omissions (CS = 99), suggesting inattention. Results for response style were average (CPT-II: CS = 55).

Perception. The patient obtained average results for visual discrimination (CPT-II score = 51). In the perceptual reasoning subtests (WISC-IV), he scored highly in block design (SS = 14) and picture concepts (SS = 12), and very highly in matrix reasoning (SS = 19). Visual processing (WISC-IV) scores were average in the block design + picture completion tests (CS = 91). In the ITPA, he obtained lower scores on the visuomotor channel than on the auditory-vocal channel. Concept visual comprehension was very good (*T*-score = 43, *M* = 35) and comprehension of relationships between visual stimuli was average (*T*-score = 34). At the automatic level, he scored poorly for visual integration (*T*-score = 26).

Memory. Regarding working memory (WISC-IV), he obtained low-normal scores on the digit span (SS = 8). He obtained a slightly below-average score for visual sequential memory (ITPA: *T*-score = 32). Results in WISC-IV subtests for long-term memory (CS = 99.6) and general information (CS = 96) were excellent.

Language. Scores for verbal comprehension in the WISC-IV were high in similarities (SS = 14), very high in vocabulary (SS = 19), and average in the comprehension subtest (SS = 11). In fluid verbal reasoning, he obtained high average scores (CS = 95). Scores for lexical knowledge were high (99.6). The results on the auditory-vocal subtests in the ITPA representational level indicate an age equivalent to his age (mean = 35). Story retelling (*T*-score = 36, *M* = 35) and verbal analogies (*T*-score = 36) results were normal. He obtained average scores (STEN score = 6) in verbal expression in the ENFEN test battery. In the automatic level of the ITPA, the

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patient scored highly for the knowledge and use of grammatical structures (*T*-score = 38). He also obtained average scores (*T*-score = 40) on word production, related to phonological awareness.

Executive functions. Scores in response inhibition and motor control (CPT-II) were average (*S* = 42). Performance in perseveration (CPT-II) was good (CS = 30); performance in visual search, attention, and cognitive flexibility was normal (STEN score = 5) in basic tasks and poor in complex tasks (STEN score = 1). In the ENFEN, results were very low (STEN score = 2) in the planning test and extremely low in resistance to interference (STEN score = 1).

Motor skills. Scores for motor skills on the McCarthy Scales of Children's Abilities (MSCA) suggest that his performance is below that expected for his age (CS = 10).

Our results are relevant for the differential characterisation of NS cognitive functioning as well as for the psychological and educational approach to patients with *SOS1* mutations. Further studies on the functional variability of the different mutations associated with NS should be performed.

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A report on reliable differences in the profile of the ACE-III[☆]

Reporte de las diferencias confiables en el perfil del ACE-III

Dear Editor:

In a recent study, Matías-Guiu et al.[1] analysed the psychometric properties of Addenbrooke's Cognitive Examination III (ACE-III) for the diagnosis of dementia. These authors reported high reliability and inter-rater agreement (>0.90), good sensitivity and specificity, and a strong correlation with the Mini—Mental State Examination (MMSE). However, they focus on total ACE-III scores, disregarding subtest scores for attention, memory, fluency, language, and visuospatial abilities. These subtests provide valuable information on the patient's cognitive profile, which is essential for preparing a personalised treatment plan.

In clinical practice, ACE-III subscores vary from patient to patient; the reliability of such differences should therefore be assessed. Matías-Guiu et al. do not evaluate this factor; as a result, the extent to which an ACE-III profile is influenced by measurement error cannot be determined. A mathematical formula has been proposed to address this issue, and can be used to analyse the difference between 2 scores[2]:

$$pd = \frac{SD_1^2 + SD_2^2 - 1SD_1SD_1SD_2p_{12}}{SD_1^2 + SD_2^2 - 2SD_1SD_2p_{12}}$$

In this expression, SD_1, SD_2, ρ_1, and ρ_2 are the standard deviations (SD) and reliability coefficients (normally the α coefficient[3]) of subtests 1 and 2, respectively, and ρ_{12} is the correlation between the 2 subtests. The result ($0 \leq \rho_d \leq 1$) indicates the percentage of variability corresponding to true variance; when the latter is high, it can be concluded that the error of measurement has had no decisive impact on differences.

Matías-Guiu et al. only report SDs for each subtest in one of the tables of the study, and provide no data on their α coefficients or the correlation between subtests. Using fictitious data, below is an example of how complementary analyses may fill this gap. Firstly, to estimate the α coefficient of each subtest, the mean inter-item correlation for

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[☆☆] This article has not been presented at any meeting or congress.