Cerebrospinal Fluid Research



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Neuropsychological assessment of cerebellar malformation in Spina Bifida

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Background

It is widely agreed that the typical cognitive profile of children with spina bifida is influenced by medical variables such as hydrocephalus, the height of the lesion and other physical complications [1]. However, very little is known about the specific contribution of the Arnold-Chiari malformation (ACM) in the information processing of these children. The aim of this study is to delineate in the cognitive profile the specific contribution of the ACM by comparing within a group of children with spina bifida among those with and without ACM.

Materials and Methods

Fifty-five children between 6 and 14 years of age were recruited from the spina bifida team of the Radboud University Nijmegen, Medical Centre. Of the whole group of children, those with a well documented presence or absence of ACM were included for further analysis (complete group). To eliminate the confounding influence of IQ a selection was made on the criteria of a total IQ of above 70 (non-retarded group). Table 1 presents the patient characteristics of the different subgroups. All children underwent a neuropsychological assessment which consisted of tests on a wide range of cognitive functions, comprising verbal, performal, and general intelligence, visuo-motor processing, selective and sustained attention, simultaneous and sequential memory, word fluency, and

speed of information processing. To address the cerebellar cognitive functions, some tasks were included which are expected to measure cerebellar information processing [2,3].

Results

Results reveal significant differences in information processing among the children with and those without ACM. In the complete group, children with ACM performed significantly worse on the following cognitive functions: perception, visual-motor integration, verbal functioning, sequential memory, (non)verbal speed, and arithmetics. For the non-retarded group the data reveals a different cognitive profile. In this group, children with ACM showed impairments on tasks which require perception, imprinting and verbal memory, and verbal fluency.

Discussion

In the complete group, a cognitive profile of strengths and weaknesses was found that resembles the one associated with hydrocephalus and spina bifida as presented in the literature. However, this typical cognitive profile was not found for the non-retarded group. The non-retarded ACM group showed impairments on cognitive tasks which are hypothesized to be mediated by the cerebellum and can be compared to cognitive deficits related to cerebellar pathology.

Table 1: Patient characteristics

		n	Mean age	Age range	TIQ	VIQ	PIQ
Complete group	ACM-	10	10.5	6.8–14.1	89.6	93.3	87.5
	ACM+	23	10.4	6.5-13.3	72.7	79.8	69. I
Non-retarded group (TIQ>70)	ACM-	9	10.4	7.6-12.5	91.9	95.8	89.2
	ACM+	13	10.2	6.5-13.3	83.7	90. I	79.9

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