Neurol Sci (2011) 32 (Suppl 3):S307–S309
DOI 10.1007/s10072-011-0779-x

CLINICS

Can Chiari malformation negatively affect higher mental functioning in developmental age?

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Published online: 8 October 2011

Abstract Numerous studies on adults have confirmed that the cerebellum has a role in processing higher brain functions, and evidence of this role has emerged more recently in developmental age as well. Various types of congenital lesion are associated with neuropsychological impairments and behavioral changes that can sometimes even give rise to a picture of autism. Acquired cerebellar lesions (especially tumors and stroke) in children of normal intelligence have enabled different neuropsychological profiles to be identified, depending on the cerebellar site involved. In Chiari malformation, the cerebellar structures are squeezed and crowded inside the posterior fossa and along the time this could generate various kinds of cognitive and behavioral disorders. Currently available data remain inconclusive, however, and prospective longitudinal studies on sizable series will be needed to ascertain whether and to what degree Chiari malformations may negatively affect mental functioning in developmental age.

Keywords Chiari malformation · Cognition · Development

The cerebellum was once believed to be almost exclusively involved in coordinating voluntary movement, but its perceived role has changed in the last two decades as a result of emerging findings in the spheres of neuroanatomy, neuroimaging and clinical neuropsychology, which have shown that the cerebellum is involved in intelligence, language and higher social functions. More and more clinical and neuroimaging studies support the cerebellum's important role in perceptual, language-related, cognitive and affective functions [1].

The cerebellum was often associated with various neurodegenerative and psychiatric diseases, but specific patterns of neuropsychological malfunctioning have been described more recently in patients with focal discrete lesions. Impairments relating to mental imagery, anticipatory planning, the appropriate use of temporal cues, visuospatial functions and language production have also come to light [2]. Such recent additions to our understanding prompted Schmahmann's clinical studies and the formal identification of a whole array of cognitive and emotional symptoms in adult patients with various kinds of neurological disease classifiable as *cerebellar cognitive affective syndrome* (CCAS) [3]. Lesions of the posterior cerebellar lobe are reportedly responsible for more severe cognitive disorders than those affecting the anterior lobe; and lesions involving the vermis coincide with more severe affective and relational disorders. Neuropsychological and behavioral disorders have been identified in children with congenital cerebellar diseases [4] and acquired conditions [5–8]. The studies on children with neoplastic diseases involving the cerebellum have confirmed the existence of related cognitive affective syndromes and topographical issues, i.e. the posterolateral regions process cognitive and neuropsychological functions, while the vermis is involved in processing emotions and complex social behavior.

Now that the role of the cerebellum in modulating higher mental functioning has become clear, attention has rightly focused on Chiari malformation, since compression of the structures in the posterior fossa could negatively affect how these cerebellar structures function.

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Novegno et al. [9] described ten children (eight of them with a normal IQ and two borderline), five of whom had slowed speech, poor fluency and word finding difficulties, while language comprehension and production were adequate. In four of those with language difficulties, the tonsils had descended more than 10 mm. Two children were hyperactive, but there were no cases of autism or relational disorders.

Our sample comprises 35 children (mean age 78 months, SD 53 months, range 5–196 months), 15 of them pure cases (with no associated neurological conditions, and particularly no neurogenetic syndromes that might include Chiari malformations as part of their symptom picture) and 20 with associated conditions and syndromes. The former 15 cases had an IQ in the range of 72–122 (median 105), while for the latter 15 it was 29–114 (median 64) (unpublished data).

The preliminary analysis of these data already shows a marked variability and, even among the pure cases, cognitive functioning depends on numerous other factors as well as the Chiari malformation. The heterogeneity of behavioral and cognitive phenotype is greater among cases with associated conditions and syndromes, especially for children with normal functioning despite they have the Chiari malformation and associated conditions.

To give an example, we describe two cases of children with Chiari malformations type I who underwent decompression and were administered an extensive battery of neuropsychologicical tests, among which only those concerning general intelligence, language and behavior are of interest here. One case was a 5-year-old boy with a history of language delay, tonsillar ectopia and flow alterations, who underwent decompression at the age of 3 years and 3 months. At 29 months of age, he had Griffiths Mental Development Scales GQ of 95 and language subQ of 77. Specific language assessment showed an equivalent age corresponding to 16–17 months in comprehension task, and an equivalent age of 21 months in lexical production. The Child Behavior Checklist returned pathological scores in all total, internalizing and externalizing scales. After surgery at the age of 41 months, the boy had a Full IQ of 88, a Verbal IQ of 95 and a Performance IQ of 84. Consistent with the increase in the Verbal IQ, the Test di Valutazione del Linguaggio [10] showed an equivalent age of 44½ months both in comprehension and production, meaning a significant improvement respect to the situation before surgery. His behavioral issues included behavioral difficulties, attentional and motor instability, which became worse than before and were pathological for both internalizing and externalizing scores. In short, this child's language improved considerably, but his behavior continued to deteriorate.

The second case was a 15-year-old girl with an arachnoid cyst in the left temporal region and a Chiari malformation, with tonsillar ectopia and a disrupted flow, who was decompressed in the atlanto-occipital site at 11 years. At 3 years, she had Griffiths Mental Development Scales GQ of 93 and language subQ of 94. Assessed again at the age of 13 years and 8 months, she had a Full IQ of 80, a Verbal IQ of 70 and a Performance IQ of 94. As for the factorial indexes, she obtained a verbal comprehension index of 76 confirming low scores in all verbal items. Language testing showed an equivalent age of 10.7–11.2 years in comprehension and of 10.3 years in lexical production, a 1-year delay in grammatical skills and a markedly deficient semantic and phonological fluency. Before surgery, her behavior had been characterized by mild hyperactivity and she was easily distracted, but after decompression she behaved normally. In short, this child's language skills became worse after surgery, while her attentional instability improved considerably.

These two cases clearly exemplify the complexity of the problem, since they present completely contrasting pictures, although both had Chiari malformation with flow disruptions. Their age at the time of decompression differed considerably and a more prolonged compression would presumably have more persistent, less reversible effects. It is worth noting that the second case had an arachnoid cyst in the left temporal region (an area crucial to language comprehension), where it was bound to influence the girl's language processing and development. It is also important to bear in mind the variability due to the genetic constitution and the complex plasticity-related mechanisms of cognitive reorganization. Different cases can thus present an extremely varied picture both between and within individuals. In addition, the cerebellum's impact on mental functioning is by no means exclusive, but takes effect in cooperation with other factors, including genetics and individual constitutional characteristics.

For a thorough assessment of the impact of Chiari malformation on neurocognitive and behavioral functioning, we must therefore design prospective, preferably longitudinal studies on large series so as to correlate findings with different variables, or consider smaller samples already selected a priori in terms of confounding variables.

Conflict of interest The authors declare that there is no actual or potential conflict of interest in relation to this article.

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