



Analysis of Visuospatial Abilities in Chiari Malformation Type I

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Published online: 8 July 2019

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Abstract

The involvement of the cerebellum in visuospatial abilities has been evidenced in numerous studies, based on the cerebellar-cortical circuitry. This domain has been evaluated in several patients with cerebellar disorders, but the assessment of visuospatial processing in Chiari malformation type I (CM-I) is scarce. The aim of this study is to analyze the visuospatial performance between CM-I adult patients and healthy controls. Participants have been tested using Block Design and Visual Puzzles subtests of the Wechsler Adult Intelligence Scale (WAIS), the Benton Judgment of Line Orientation test, and the Rey-Osterrieth Complex Figure test. The anxious-depressive symptomatology, the physical pain, and the premorbid intelligence have been controlled for, as well. The CM-I patients showed a significantly lower performance; however, after analyzing and controlling for the effect of clinical variables and psychopathological symptomatology, the main effect was maintained for visual puzzles and line orientation tasks. The findings suggest that CM-I patients show a poorer performance in tasks that require an exercise of perceptual reasoning without motor demand, accompanied by visualization and mental imagery of the stimuli. This study contributes towards the reinforcement of the evidence on the cognitive alterations associated to CM-I.

Keywords Chiari malformation · Cerebellum · Visuospatial abilities · Cognition

Introduction

In recent years, the relationship between the cerebellum and visuospatial abilities has been evidenced in numerous studies, both through neuroimaging studies and clinical observation [1–4]. In this area, Molinari et al.'s [3] study, in which the role of the cerebellum in visuospatial processing was demonstrated, is noteworthy. The study sample included patients with focal cerebellar lesions, in both left and right hemispheres, and others with idiopathic cerebellar ataxia. The study outcomes showed that cerebellar disorders may lead to worse

performance in tasks that involve mental imagery, such as mental rotation of objects, or those in which there are visuoconstructive demands. Molinari and Leggio [2] concluded that if a task does not demand the mental manipulation of the objects, individuals with cerebellar lesions could show a similar execution to that of healthy people, even though a slightly lower performance is manifested by the former. O'Halloran, Kinsella, and Storey's [5] study demonstrated that in addition to the processing of patients with cerebellar pathologies being affected, their perceptual organization and visuoconstructive ability are also reduced.

Visuospatial functioning was found to be closely related to the parietal cortex—more oriented to spatial analysis—and the frontal cortex—directed towards executive control over it—without forgetting the role of the occipito-temporal cortex in the perception and recognition of visual stimuli [6]. The cerebellar-cortical circuitry includes connecting pathways with all these areas, thus allowing for the implication of the cerebellum in different visuospatial tasks [1]. A recent study by Olivito et al. [7] found a significant correlation between the performance on visuospatial tasks and gray matter loss in different cerebellar regions (lobules VIIIB, VIIIA, Crus I, Crus II, lobule V, and vermis). Involvement of the cerebellum in the

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acquisition of spatial procedural elements is an important aspect of its role [8, 9]. Moreover, both the experimental and clinical reports indicate that the cerebellum is a significant structure involved in spatial learning and strategies, which is necessary to carry out visuospatial tasks [2, 10, 11].

Likewise, another argument in favor of the implication of the cerebellum in visuospatial processing is the “cerebellar cognitive affective syndrome” (CCAS). The CCAS presents a clinical picture whereby individuals with cerebellar lesions manifest a symptomatic profile with alterations in executive functioning, spatial cognition, and language, together with a more voluble personality [12].

Spatial cognition encompasses a broad set of systems and correlates responsibly for the localization and integration of visual elements [13]. This domain has been evaluated in patients with focal cerebellar lesions and other pathologies such as ataxia [3, 14–16]. However, when it comes to assessing visuospatial processing in adult individuals with Chiari malformation type I (CM-I), the literature is scarce. CM-I is a pathology of the craniocervical junction, characterized by a herniation greater than 3 mm of the cerebellar tonsils through the foramen magnum (see Fig. 1). Due to the ectopia, the spinal canal is invaded, and although it is not often associated to other significant brain malformations, it is commonly manifested with syringomyelia [17]. Although there are no studies aimed at evaluating this domain specifically, there are research results that suggest a deficit in the visuospatial ability associated with CM-I. For example, in García et al.’s [18] study, a lower performance of those affected by CM-I is evidenced in comparison to a group of healthy controls, both in their precision copying figures and in their visual memory ability. In a later work by the same group of authors, this finding is maintained regardless of whether patients with CM-I had undergone decompressive surgery or not [19]. In their study, Kumar et al. [20] found that 10 adults with CM-I obtained a significantly lower

performance in terms of visuospatial reasoning and visuomotor speed, compared to a group of healthy controls. In contrast, in another study reporting information on two ex-military adults with CM-I, there were no significant deficits in this domain, but authors did highlight the variety between the profiles [21]. If studies with a child population are taken into account, both Grosso et al. [22] and Haapanen [23] reported a poorer performance by those affected by CM-I.

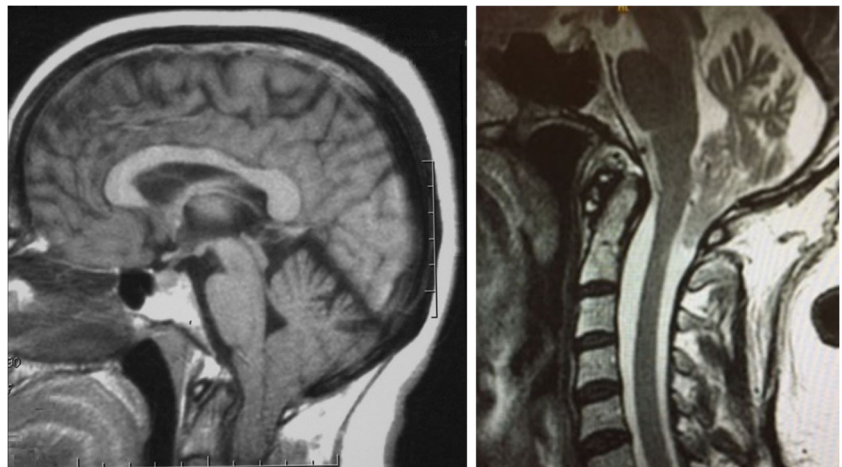
Visuospatial skills are one of the main competences that are at stake during day-to-day tasks, where skills related to orientation and spatial localization, gnosis, and praxias intervene. When there are anomalies in this regard, they become noticeable for those individuals who suffer them. The aim of this study is to compare the performance in visuospatial reasoning between a group of individuals affected by CM-I and a group of healthy controls, with a greater number of specific tests for this domain, in order to study the scope of the deficit associated to this disease. Given that this pathology presents neuropsychiatric symptoms [24–26] and physical pain [27], the possible influence of these variables on the performance of cognitive tasks has also been controlled.

Method

Participants

The clinical group was composed of 26 patients diagnosed with CM-I of congenital origin (22 females, 4 males; mean age 47.15 ± 14.61 ; mean years of education 13.50 ± 2.80). Patients were recruited through the neurology service of the Marqués de Valdecilla University Hospital (Hospital Universitario Marqués de Valdecilla, HUMV) and the Chiari and Syringomyelia Association of the Principality of Asturias (Asociación Chiari y Siringomielia del Principado de Asturias, ChySPA),

Fig. 1 Sagittal T1- and T2-weighted MRI showing tonsillar ectopia of CM-I



between 2017 and 2018. Once the contact with these organizations was established through face-to-face meetings and e-mail, the people in charge of each

organization informed the potential participants about the study and those who were interested in taking part got in touch with the researchers. The clinical

Table 1 Clinical characteristics of Chiari malformation type I patients

	M (SD)/n (%)	Frequency <i>n</i> (%)			Body area <i>n</i> (%)		
		Low	High	Up/low limbs	Thorax or back	Neck	
Diagnosis data							
Age at diagnosis (years)	36.58 (13.16)						
Age of onset (years)	31.35 (14.71)						
Diagnosis delay (years)	5.23 (8.09)						
Disease duration (years)	10.81 (8.19)						
Surgical treatment							
PFD	8 (30.8%)						
Non-PFD	18 (69.2%)						
Time elapsed from surgery (months)	88.13 (56.80)						
Tonsillar ectopia (mm)	7.74 (3.95)						
Comorbidity							
Syringomyelia	7 (26.9%)						
Hydrocephalus	2 (7.7%)						
Basilar impression	1 (3.8%)						
Platybasia	1 (3.8%)						
Scoliosis	14 (53.8%)						
Other cranial malformations	1 (3.8%)						
Signs and symptoms							
Headache	25 (96.2%)	11 (44%)	14 (56%)				
Neck pain	23 (88.5%)	4 (17.4%)	19 (82.6%)				
Pain upper limbs	16 (61.5%)	6 (37.5%)	10 (62.5%)				
Pain lower limbs	12 (46.2%)	7 (58.3%)	5 (41.7%)				
Dizziness	22 (84.6%)	10 (45.5%)	12 (54.5%)				
Vertigo	10 (38.5%)	5 (50%)	5 (50%)				
Instability (balance problems)	16 (61.5%)	8 (50%)	8 (50%)				
Fatigue	13 (50%)	1 (7.7%)	12 (92.3%)				
Visual alterations	15 (57.7%)	7 (46.7%)	8 (53.3%)				
Hearing alterations	19 (73.1%)	12 (63.2%)	7 (36.8%)				
Nystagmus	1 (3.8%)	1 (100%)	–				
Swallowing problems	9 (34.6%)	6 (66.7%)	3 (33.3%)				
Sphincter problems	4 (15.4%)	4 (100%)	–				
Sleeping disturbances	13 (50%)	2 (15.4%)	11 (84.6%)				
Cognitive subjective dysfunctions	20 (76.9%)	5 (25%)	15 (75%)				
Sensory loss	13 (50%)	6 (23.1%)	6 (23.1%)	11 (84.6%)	2 (15.4%)	–	
Temperature sensitivity (hyper or hypo)	11 (42.3%)	6 (54.5%)	5 (45.5%)	9 (81.8%)	1 (9.1%)	1 (9.1%)	
Pain sensitivity (hyper or hypo)	7 (26.9%)	3 (42.9%)	4 (57.1%)	7 (100%)	–	–	
Muscle stiffness	17 (65.4%)	9 (52.9%)	8 (47.1%)	14 (82.4%)	1 (5.9%)	2 (11.7%)	
Muscle weakness	20 (76.9%)	12 (60%)	8 (40%)	19 (95%)	1 (5%)	–	

Note. The information shown in this table has been reported by Chiari patients
PFD posterior fossa decompression

characteristics are shown in Table 1. The control group consisted of 26 volunteers from outside the clinical group (22 females, 4 males; mean age 46.42 ± 13.53 ; mean years of education 14.62 ± 2.73). The control group was recruited among adult volunteers who wanted to take part in the study and who were informed of the project. There are no differences between the two groups in terms of gender ($\chi^2(1) = 0$, $p = 1$), age ($U = 317.50$, $p = 0.707$), or years of education ($U = 247.00$, $p = 0.090$).

The inclusion criteria were as follows: (i) being over 18 years of age, (ii) residing in Spain and communicating in Spanish, (iii) having received a diagnosis of CM-I according to the criteria of the ICD-10 [Q07.01], (iv) having had a magnetic resonance test that verifies the diagnosis, and (v) that a minimum of 12 months have elapsed in the case of patients who have undergone decompression surgery. The exclusion criteria of the study include (i) having any other neurological, psychological, or psychiatric diagnosis included in the ICD-10, independent of the CM-I, during their participation in the study; (ii) illiteracy; (iii) suffering sensory impairments to perform the tests; and (iv) being under pharmacological treatment susceptible of affecting cognitive performance.

All participants completed the informed consent document and voluntarily took part in the study. The project was approved by the Ethics Committee of the University of Deusto.

Instruments

After administering a brief interview in which the sociodemographic and clinical information of the sample was collected, the test protocol was administered. All of them were administered in the corresponding version adapted for Spanish samples, exhibiting good psychometric properties.

Neuropsychological Assessment

Block Design of the Wechsler Adult Intelligence Scale-IV [28; Spanish Version—29] It is a test of visuo-perceptual reasoning, general visuospatial intelligence, and visuo-motor coordination, in which the evaluated person must reproduce an image model with the red and white cubes provided. It consists of 14 items, throughout which the task time is recorded. The test offers a score between zero and 48 (no time bonus) or zero and 66 (counting the time bonus).

Visual Puzzles of the WAIS-IV [28; Spanish Version—29] It is another indicator of visual perceptual reasoning and a test related to spatial visualization and spatial mental manipulation and integration. In a limited time, the evaluated individual must select which three pieces of the six possible options compose the figure that appears when they are united. It consists of 26 items, and the score ranges from zero to 26.

Benton's Judgment of Line Orientation [30; Spanish Version—31] It is a test that assesses visuospatial ability. The H Form was administered, which consists of five practice items and 30 trial items, from which the score is extracted, ranging from zero to 30. In each sheet, two lines with different degrees of inclination are shown, and the individual is asked to recognize them in a template with 11 lines distributed in segments of 18 degrees of inclination.

Rey-Osterrieth Complex Figure [32; Spanish Version—33] It is a test that explores perceptual organization, visual memory, and graphic visuoconstructive ability. The figure is presented in a horizontal position, and the individual is asked to copy it, and to reproduce it from memory after 3 min have lapsed. The figure consists of 18 items, each being scored between zero and two, according to its graphic accuracy and location with respect to the original, yielding a total score that oscillates between zero and 36. Copy time is also recorded.

Word Accentuation Test [34; Spanish Version—35] It is a test that evaluates premorbid intelligence. The task of the individual is to read correctly, with an appropriate phonetic intonation, the 30 words that are shown without an accent mark. The score ranges from zero to 30.

Psychopathological Assessment

Hospital Anxiety and Depression Scale [36; Spanish Version—37] It is a self-applied questionnaire consisting of a total of 14 items, seven of which assess the presence of symptomatology aimed at anxiety and the remaining seven, the trend towards depression. Each one is scored between zero and three, giving rise to two subscales whose score ranges from zero to 21 for each.

Physical Pain Assessment

Headache Disability Inventory [38; Spanish Version—39] It is a self-applied instrument that evaluates the subjective perception of how disabling the headache results in everyday life. It consists of 25 items with three response options each. The overall score ranges from zero to 100.

The Neck Disability Index [40; Spanish Version—41] It is a self-applied instrument that evaluates the subjective perception of physical pain relative to the cervical region. It is composed of 10 items, each one being scored from zero to five, the total score oscillating between zero and 50.

Oswestry Low Back Pain Disability Index [42; Spanish Version—43] It is a self-administered questionnaire that measures disability due to lumbar pain and allows to

identify how much this pain affects daily functioning. It consists of 10 items, whose score ranges from zero to 50.

Procedure

The assessment appointments were established once the participants were properly informed about the study and gave their consent. These sessions were individual and guided by the intervention of a neuropsychologist, with an approximate duration of 1 h. After completing the informed consent document, the session began with a brief interview to collect sociodemographic and clinical data. Next, the neuropsychological tests were administered in pencil and paper format, ending the session with the self-administered questionnaires on psychopathological symptomatology and physical pain. The procedure followed was identical for both the clinical and control groups.

Data Analyses

The statistical program SPSS (Statistical Package for Social Sciences) version 25.0 was used to perform the analyses. The normal distribution of the sample was tested using the Kolmogorov-Smirnov test. The raw scores were converted into z scores to carry out the statistical analyses.

In order to compare the clinical group with the control group with respect to sociodemographic data and cognitive and clinical variables, the Mann-Whitney U and chi-squared tests were used for quantitative and categorical variables, respectively. The effect size was calculated according to Pearson's correlation coefficient (r) and Kramer's V , as appropriate. To analyze the correlation between different variables, Spearman's Rho statistic was used.

The multivariate analysis of covariance (MANCOVA) was used to analyze the influence of psychopathological variables on the difference in cognitive performance between the clinical and control groups. The partial η^2_p was established as an indicator of the effect size.

A multiple regression analysis was used to analyze the relationship between clinical variables and cognitive performance. The level of significance was established at a p value of 0.05.

Results

Regarding the neuropsychological assessment, the findings indicate a differentiated performance between the clinical group and the control group. The CM-I patients showed a significantly lower performance in the block design test (both with time bonus (TB) and without it), in the visual puzzles task, in the Judgment of Line Orientation (JLO) test, and in visual memory assessed through the Rey-Osterrieth

Complex Figure (ROCF), as well as in the copy model. The latter shows how the group with CM-I performed a predominantly disorganized copy model, which correlated negatively with its execution both in the copy ($r = -0.454, p = 0.020$) and in the recall after the 3-min lapse ($r = -0.506, p = 0.008$). These data indicate that the greater the disorganization in the copy of the figure, the worse the score that is obtained both in the copy accuracy and in the recall. Despite these findings, no differences were observed between both groups in the accuracy and copy time of the ROCF, nor in the level of premorbid intelligence assessed through the Word Accentuation Test (TAP). These results are shown in Table 2.

Regarding the examination of the psychopathological variables, the Hospital Anxiety and Depression Scale (HADS) results indicate statistically significant differences between the clinical group and the control group, the former being the one that manifested a greater presence of symptomatology with tendency towards anxiety and depression (see Table 2).

The state of physical pain was assessed from three characteristic dimensions of CM-I, in which headaches, neck pain, and lower back pain are included. In all of them, the clinical group showed higher scores than the control group, this being indicative of a greater presence of pain. The differences were statistically significant and are detailed in Table 2.

Given the differences found in the perceived levels of anxiety and depression, a MANCOVA analysis was carried out in order to control the effect of this symptomatology on cognitive performance. Table 3 shows how the differences in the visual memory variable were eliminated once the influence of anxiety-depressive symptomatology was ruled out ($F = 3.41, p = 0.071$), while for block design, visual puzzles, and JLO tasks, the differences between the groups were maintained.

In order to examine the influence of clinical variables on the visuospatial performance of the CM-I group, a multiple regression analysis was carried out. The three dimensions that examined physical pain (headache, cervical, and lumbar pain) together with the millimeters of tonsillar ectopia, the years of coexistence with the diagnosis of CM-I, and the surgical status were included as possible predictor variables. The results show significant regressions for the block design task without TB ($F = 3.53, R^2 = 0.527, p = 0.016$) and block design with TB ($F = 4.07, R^2 = 0.562, p = 0.009$). However, only cervical pain (block design task without TB: $\beta = -0.977, p = 0.027$; block design with TB: $\beta = -0.934, p = 0.028$) and years of coexistence with the disease (block design task without TB: $\beta = -0.410, p = 0.019$; block design with TB: $\beta = -0.434, p = 0.011$) were found to be statistically significant predictors. The model was not met for visual puzzles ($F = 1.30, R^2 = 0.290, p = 0.306$), nor for the JLO task ($F = 1.59, R^2 = 0.335, p = 0.203$).

These results indicate that at least for the visual puzzles task and the JLO task (both of which demand imagery and mental manipulation of the stimuli), the differences found between

Table 2 Differences between cognitive, psychopathological, and physical statuses of clinical and control groups

Cognitive tasks	Clinical Group M (SD) ^a n (%)	Control Group M (SD) n (%)	<i>U/χ</i> ²	<i>p</i>	Effect size <i>r/V</i>
Block Design					
No Time Bonus	32.15 (9.09)	40.77 (7.42)	163.00	0.001	0.46
Time Bonus	34.42 (12.37)	45.69 (10.41)	157.00	0.001	0.44
Visual Puzzles	10.46 (3.30)	16.69 (4.45)	99.50	< 0.001	0.05
Judgment of Line Orientation	21.65 (5.06)	26.08 (2.74)	156.50	0.001	0.001
ROCF					
Copy accuracy	32.87 (4.17)	33.94 (2.17)	312.50	0.631	–
Copy time	158.00 (47.91)	157.85 (74.03)	291.50	0.395	–
Recall 3-min	16.46 (6.43)	19.90 (4.34)	211.00	0.020	0.30
Copy model					
I	7 (26.9%)	17 (65.4%)			
II	8 (30.8%)	8 (30.8%)	12.83	0.005	0.50
III	5 (19.2%)	1 (3.8%)			
IV	6 (23.1%)	0 (0%)			
WAT	26.54 (1.70)	26.92 (2.07)	283.50	0.312	–
Psychopathological and physical status					
HADS					
Total score	17.08 (8.04)	10.12 (6.33)	152.50	0.001	0.43
Anxiety	10.81 (4.79)	7.08 (3.53)	177.50	0.003	0.43
Depression	6.27 (3.82)	3.04 (3.12)	164.00	0.001	0.42
HDI					
Total score	45.88 (25.71)	0.08 (0.39)	0.00	< 0.001	0.78
Functional	25.19 (12.62)	0.08 (0.39)	0.00	< 0.001	0.81
Emotional	20.69 (13.86)	0.00 (0.00)	26.00	< 0.001	0.73
NDI	17.12 (9.01)	1.65 (2.86)	19.00	< 0.001	0.76
OLBPDQ	11.31 (7.17)	2.85 (6.71)	89.00	< 0.001	0.52

ROCF: Rey-Osterrieth Complex Figure Test; Copy model: I: construction on the central armature, II: details within the armature, III: overall contour, IV: juxtaposition of details; WAT: Word Accentuation Test; HADS: Hospital Anxiety and Depression Scale; HDI: Headache Disability Index; NDI: Neck Disability Index; OLBPDQ: Oswestry Low Back Pain Disability Questionnaire; *U*: Mann-Whitney U test; χ^2 : Chi-squared test; *r*: Pearson's correlation coefficient; *V*: Kramer's V.

^aData are shown in raw scores

the clinical group and the control group were independent of the presence of psychopathological symptomatology and of the clinical variables included in the regression model. The performance in the block design task was influenced by cervical pain and by the years of coexistence with the CM-I; therefore, the differences found between groups cannot be attributed solely to a deficit in the visuospatial abilities of the clinical group.

Discussion

In this study, the visuospatial abilities of a sample of 26 patients with CM-I were examined, comparing their performance with a group of 26 healthy controls. In a first analysis, the clinical group showed a generalized lower visuospatial performance compared to the control group, except for the accuracy and time of the ROCF copy. The results also reported that there is no difference in the level of premorbid

intelligence between both groups. By controlling the effect of anxious-depressive symptomatology, the differences in visual memory between both groups were also eliminated. Once the influence of the clinical variables was analyzed, it was observed that cervical disability and years of coexistence with the disease influenced the block design task with and without TB. However, the performance differences in the visual puzzles and JLO tasks remained independent of the physical and psychopathological state.

Compared to previous studies in which cognitive performance was assessed in visuospatial tasks, heterogeneous findings were observed. In García et al.'s [18, 19] publications, the cognitive performance of adults with CM-I, both with decompression surgery and without surgery, were examined in comparison to that of healthy controls. Through a protocol of tests that included the ROCF as a measure of spatial cognition, it was observed that the group with CM-I showed a lower performance in both the accuracy of copying and visual memory. This result remained statistically significant even after

Table 3 MANCOVA for cognitive performance after controlling for the effect of anxious-depressive symptomatology

	Clinical group M (SD) ^a	Control group M (SD)	<i>F</i>	<i>p</i>	η^2_p
Block Design					
No time bonus	−0.45 (0.19)	0.45 (0.19)	10.45	0.002	0.176
Time bonus	−0.42 (0.19)	0.42 (0.19)	8.72	0.005	0.151
Visual Puzzles	−0.62 (0.16)	0.62 (0.16)	25.48	<0.001	0.342
Judgment of Line Orientation	−0.46 (0.19)	0.46 (0.19)	11.06	0.002	0.184
ROCF					
Recall 3 min	−0.28 (0.20)	0.28 (0.20)	3.41	0.071	0.065

ROCF Rey–Osterrieth Complex Figure test, *M* mean, *SD* standard deviation, *F* MANCOVA, η^2_p partial eta squared (effect size)

^aData are shown in *z* scores

controlling the effect of the psychopathological variables and the perceived physical pain. This aspect does not coincide with the results of the present study, as the performance between the groups in the ROCF became similar once the effect of the anxious-depressive symptomatology was controlled for. However, both studies coincide in that there were no differences between groups regarding the time of copying. It seems possible that these results are due to methodological issues, as the sample in the present study was smaller. The conclusions were also more limited as only one test was performed for this domain, the ROCF. Regarding the analysis of the influence of decompressive surgery on cognitive performance, García et al. [19] reported that there is no difference between both groups of patients, concluding that their cognitive profile was homogeneous. This assessment agrees with the present study as the surgical status was included as a possible predictor of visuospatial performance, and it was observed that the execution of the CM-I was independent of the status.

Another prominent study in which visuospatial ability was assessed in a group of adults with CM-I is that of Kumar et al. [20]. The findings of these authors agree with those of the present study, as they also found a lower performance of the group with CM-I compared to that of healthy controls. In addition, some of the tests used in its protocol also coincide with those used in the present study, which leads to the suggestion that performance in those tasks that require mental integration of objects is more deficient in those affected by CM-I. However, the difference between both works is that Kumar et al. [20] also correlated these data with measurements obtained through diffusion tensor imaging (DTI), finding microstructural abnormalities in some brain regions. Despite being speculative, as those types of measures were not used in the present study, it is possible to compare both studies based on the data obtained through the neuropsychological tests. Considering the results, it is likely that the worse performance of patients with CM-I is due to other anomalies beyond the tonsillar ectopia itself, and the compression that it entails.

Only a few studies have analyzed the performance of visuospatial tasks in the adult population with CM-I. However, with other key cerebellar pathologies such as ataxia, more specific works can be found [14, 44]. A recent study by Slapik et al. [15] assessed the organization and visuospatial memory in a group of 49 patients with cerebellar ataxia of different etiology and a group of 60 healthy controls. After a very detailed analysis of the performance in the different tests, the authors pointed out a marked disorganization on behalf of the clinical group, attributing this phenomenon to difficulties in planning and sequencing, which could have been compensated by other mechanisms in the correct performance of perceptual tasks. Previous experimental and clinical studies about procedural learning have reported that cerebellar networks are involved in the acquisition of sequences, which is impaired in patients with cerebellar lesions [45]. This aspect is consistent with the present study, as the type of copy in the ROCF was also significantly more dysfunctional in patients with CM-I. Moreover, Slapik et al. [15] concluded that the low performance of the group with ataxia was probably due to deficient cortico-cerebellar connectivity. Despite the common factor of being two cerebellar pathologies, this interpretation must be examined carefully given the particularities of each disorder.

In the present study, patients with CM-I performed lower in visual puzzles and JLO tasks, even after controlling for the clinical variables. Both tasks require an exercise of perceptual reasoning without motor demand, accompanied by visualization and mental manipulation of the stimuli, with this being more demanding in the visual puzzles test [46]. According to the literature, it is precisely when the task demands mental imagery that those affected by cerebellar damage show a worse performance and do not necessarily show a deficient processing of visual information [1]. This finding is in agreement with Molinari's et al. [3] findings which showed that the worst performance of patients with cerebellar lesions is in mental rotation tasks. This phenomenon is consistent also with what was found in the present study, as those affected by CM-I had a lower performance compared to healthy controls in tests

that require mental integration of the stimuli (planning or organization of the ROCF copy, visual puzzles, and JLO). Meanwhile, performance was similar in tasks in which the visuospatial demand was accompanied by physical manipulation of objects and movement (block design with and without TB, ROCF copy), which may indicate that motor exercise serves as a compensatory mechanism and allows for a visuospatial performance similar to that of healthy controls, this being a remarkable strength. The findings regarding these variables in which no inter-group differences were observed are also relevant, as they shed light on the specific visuospatial deficits that may be associated with CM-I and on which direction future treatments should follow.

The most plausible explanation for this phenomenon is related to cortico-cerebellar connectivity. Experts do not doubt the implication of the cerebellum in cognitive processes [47], as well as in perceptual processes [48]. According to Leggio et al. [11], the role of this structure could be based on developing the strategy during a spatial task and connecting it to the prefrontal cortex. In fact, there are studies that, through neuroimaging tests, have shown how cerebellar regions are activated during the execution of purely visuospatial tests such as Landmark's task [49]. An example of this can be found in the work of Stoodley et al. [50], in which the specific regions of the cerebellum that intervene in spatial tasks including mental rotation, copy, and organization of the ROCF and JLO are detailed with graphic representations. In addition, the cerebral parietal and frontal cortex, closely related to visuospatial abilities, also have a strong system of cortico-cerebellar connective pathways [51]. This frontoparietal network occupies the largest volume in the cerebellum, superimposing the one it represents in the cortex comparatively [52]. Given that the present study does not have neuroimaging tests that could directly support the results, caution is required when contemplating this explanatory hypothesis. Another observed difficulty is the notable lack of studies in the literature on visuospatial skills related to CM-I, which conditions the interpretation of the results.

Regarding the limitations of the study, the first to be highlighted is the small sample size, which limits the representativeness and generalization of the results, as well as the design, as it does not provide longitudinal information. Similarly, the distribution of gender across the sample is not equal since the number of women is higher, which could be expected, as it is considered that CM-I is more common among women. Moreover, the variety across the symptomatology of the clinical group must be mentioned, in addition to the fact that the sample has both decompressed patients and patients who have not undergone decompression surgery, which further contributes to this heterogeneity. However, this is an aspect that also reflects reality, as both the course and the clinical manifestation of the disease are very variable among cases. Additionally, according to the literature [19], the

cognitive profile manifested in patients with CM-I is independent of the surgical status of the patients. The fact that the recruitment took place in two different centers can also be considered a bias since the treatments and the intervention have been carried out by different medical professionals. Another important limitation is the source of information, as the patients themselves have provided the information about their clinical history. There was no direct access to all medical records. Finally, as already noted above, the study does not have neuroimaging tests that could support the neuropsychological findings. In future research, all these issues should be addressed in order to eliminate the possible biases of the study, and thus reinforce the conclusions.

Conclusion

The group with CM-I presents a generalized lower performance in the proposed visuospatial tasks compared to the group of healthy controls. After controlling the effect of anxious-depressive symptomatology and analyzing the influence of clinical variables on the performance of cognitive tests, the main effect is maintained for visual puzzles and JLO tasks. Both tasks demand an exercise of mental manipulation of the stimuli, which coincides with what the literature states about alterations in cerebellar disorders. This study contributes towards the reinforcement of the evidence on the cognitive alterations associated to CM-I, and specifically, on the analysis of visuospatial abilities. Given that these are a necessary competence in everyday life and in light of the results found, it is necessary to include this domain in the assessment and treatment of the diagnosis of CM-I.

Acknowledgments We thank ChySPA, and all of participants for their involvement in the study and their effort.

Funding This study was funded by a grant from the Education Department of the Basque Government's "Programa Predoctoral de Formación de Personal Investigador No Doctor" (PRE_2016_1_0099 to Maitane García).

Conflict of Interest The authors declare that they have no conflict of interest.

Ethical Approval and Informed Consent All procedures performed in this study were developed in accordance with the ethical standards and with the 1964 Helsinki declaration and its later amendments. Informed consent was obtained from all individual participants included in the study.

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