



Visuospatial Organization and Recall in Cerebellar Ataxia

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Abstract

Poor visuospatial skills can disrupt activities of daily living. The cerebellum has been implicated in visuospatial processing, and patients with cerebellar injury often exhibit poor visuospatial skills, as measured by impaired memory for the figure within the Rey-Osterrieth complex figure task (ROCF). Visuospatial skills are an inherent aspect of the ROCF; however, figure organization (i.e., the order in which the figure is reconstructed by the participant) can influence recall ability. The objective of this study was to examine and compare visuospatial and organization skills in people with cerebellar ataxia. We administered the ROCF to patients diagnosed with cerebellar ataxia and healthy controls. The cerebellar ataxia group included patients that carried a diagnosis of spinocerebellar ataxia (any subtype), autosomal dominant cerebellar ataxia, or cerebellar ataxia with unknown etiology. Primary outcome measures were organization and recall performance on the ROCF, with supplemental information derived from cognitive tests of visuospatial perception, working memory, processing speed, and motor function. Cerebellar ataxia patients revealed impaired figure organization relative to that of controls. Figure copy was impaired in the patients, but their subsequent recall performance was normal, suggesting compensation from initial organization and copying strategies. In controls, figure organization predicted recall performance, but this relationship was not observed in the patients. Instead, processing speed predicted patients' recall accuracy. Supplemental tasks indicated that visual perception was intact in the cerebellar ataxia group and that performance deficits were more closely tied to organization strategies than with visuospatial skills.

Keywords Cerebellum · Ataxia · Visuospatial · Memory · Organization · Cognition

Introduction

Widespread evidence points to the involvement of the cerebellum in cognition, including visuospatial abilities [1–3]. Individuals with cerebellar ataxia display impaired performance on a diverse range of visuospatial tasks, including

Benton line orientation [2, 4], mental rotation [5], and tests of visuospatial memory [6]. In healthy individuals, functional neuroimaging has shown cerebellar activation during visuospatial tasks such as line bisection judgment [7], line orientation [8], and tests of visuospatial working memory [9].

Additionally, the cerebellum may play a role in the nonvisual perception of spatial relations: for example, healthy people whose cerebellums were disrupted with TMS had difficulty gauging their body's distance from an object [10]. This function in particular may reflect the cerebellum's role in the production of feedforward models representing predicted sensory consequences of motor commands, enabling perceptual estimation and guidance of movement to be carried out online [11]. This account is consistent with subjective reports from ataxia patients, who describe having difficulty navigating their surroundings in the absence of visual feedback. For example, one patient with acute cerebellar dysfunction reported difficulty touching his nose with his eyes closed [12], while another patient recounted that his first symptom was beginning to “fall

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down in the dark” [13]. As such, disturbances of visuospatial and nonvisual spatial abilities can have a severe impact on the quality of life for people suffering from ataxia, and better understanding the nature and scope of these impairments is critically important for patient care. One study found that patients with cerebellar lesions used less efficient strategies during the multiple errands task, which measures executive function in “real-life” situations such as purchasing a soda and a card from a vending machine [14]. In other neurological disorders, cognitive impairments have been shown to impact risk of injury [15], caregiver burden [16], and overall quality of life [17].

On one classic test of visuospatial ability, the Rey-Osterrieth complex figure (ROCF) (shown in Fig. 1) [18], results have been mixed in studies of populations with cerebellar ataxia, which results from degeneration of cerebellar neurons, and other types of cerebellar injury (i.e., stroke and tumor resection). Some have found no impairment on figure copy or recall accuracy [19–22], while others have found impairment on both measures [23]. One study reported significant impairment on figure recall, but not copy, in individuals with advanced spinocerebellar ataxia type 2 [24].

In addition to assessing the accuracy of the final drawing, studies have also examined the organizational procedures that participants use to draw the complex shape [25–28]. There is evidence that patients with cerebellar lesions use less efficient figure organization to draw the ROCF [29, 30], as do people with neurological disorders such as autism [31] (“[drawing] the global outline later”) and schizophrenia [32–34], which may also involve the cerebellum [35, 36]. However, to our knowledge, no studies have looked at figure organization in cerebellar ataxia, which may represent a special case due to its progressive decline, allowing patients time to adapt and develop compensatory mechanisms in conjunction with gradual functional loss. In addition, while level of organization has been shown to correlate with copy and recall accuracy in healthy individuals [37, 38], this may not be the case in some

neurological disorders [33, 37], and no studies have investigated the nature of this relationship in cerebellar ataxia. Finally, while the mechanism behind lower figure organization in autism and schizophrenia has been explored to some degree—it has been linked to executive dysfunction [32] or a more local style of processing [31, 34, 39]—the basis of these impairments remains unexplored in cerebellar ataxia. Neuroanatomical [40–42] and resting-state functional connectivity studies [43, 44] show connections between the cerebellum and the parietal lobe, which is involved in visuospatial integration [40]. As a result, the cerebellum may contribute to figure organization by facilitating effective visuospatial integration and perception while encoding the shape.

To assess visuospatial organization ability in cerebellar ataxia, we measured figure organization and recall accuracy using the ROCF in cerebellar ataxia, hypothesizing that patients would show lower figure organization than that of controls and that this would drive lower figure accuracy. Furthermore, in order to investigate the role of gestalt perception in figure organization ability, we administered two additional perceptual tasks: a block design task based on Caron et al. [45] and a task of our own design based on bistable images (i.e., optical illusions that can be perceived in two ways). In the block design task, block patterns varied in their level of perceptual cohesiveness, where some designs fit together to form an overall “gestalt” image and others did not. Prior studies have shown that a more local style of perceptual processing gives participants an advantage on “gestalt” block design trials [46]. Accordingly, if ataxia patients are found to have an advantage on gestalt trials relative to that of controls, this would suggest a perceptual reason for lower figure organization: specifically, it would suggest that people with cerebellar ataxia have difficulty integrating details into global, “gestalt” shapes. We also developed and administered a test based on bistable images in order to examine participants’ ability to switch between multiple “gestalt” shapes embedded in a single image, similar to switching between local and global features of the

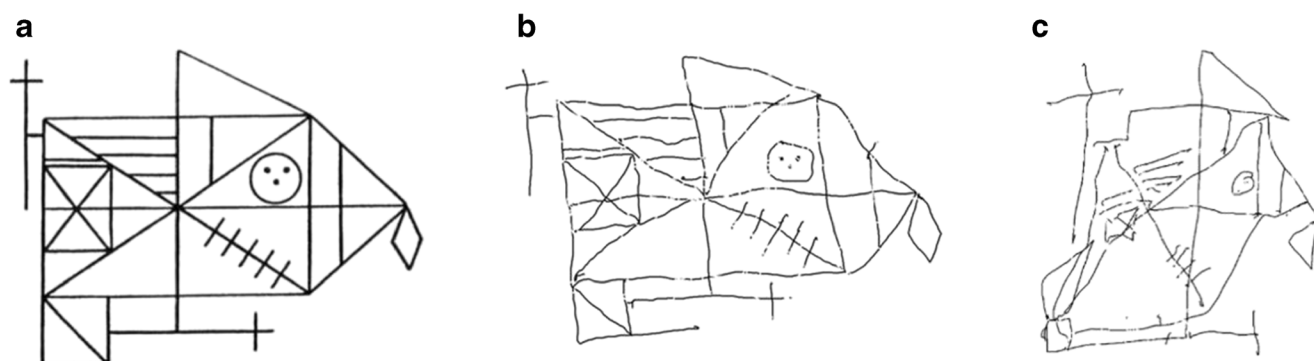


Fig. 1 The Rey-Osterrieth complex figure and participant examples. **a** The Rey-Osterrieth complex figure. **b** Figure copy by a healthy control. **c** Figure copy by an individual with cerebellar ataxia. Although scoring

ignored errors due to tremors or other motor disturbances, the ataxia patient’s drawing received a lower score than the control’s due to inaccuracies and misplacements

complex figure during encoding. Here again, lower performance would indicate that a perceptual impairment could underlie the ataxia patients' hypothesized impairment in figure organization.

Finally, in order to examine what cognitive faculties were driving performance on the ROCF in ataxia patients and controls, we administered a series of supplemental tasks to assess various cognitive, motor, and affective domains, including the Symbol Digit Modalities Test (SDMT), Digit Span, Finger Tapping, the Center of Epidemiologic Studies Depression Scale (CES-D), and the International Cooperative Ataxia Rating Scale (ICARS).

Our test administration was novel in using an electronic tablet to record complex figure drawings. Although traditional paper-and-pencil administrations of the ROCF periodically rotate colored pencils to keep track of one's drawing sequence [47], this method interrupts the task procedure and does not differentiate elements drawn within the same time window. By contrast, our administration records the precise order in which figure components were drawn.

Patients and Methods

Study Participants

In this study, 49 cerebellar ataxia patients and 66 healthy controls were recruited through the Ataxia Center at Johns Hopkins. Of these, 6 controls were excluded for being much younger than the ataxia population, which resulted in a final count of 49 cerebellar ataxia patients and 60 controls. In addition, 2 ataxia patients and 2 controls were excluded from the complex figure results due to technical issues that rendered the tablet-recorded data uninterpretable. Not all patients and controls completed all tests, as some participants were recruited specifically for some of the follow-up tests, as described below. The number of participants who completed each test is indicated in Table 1. Patients had been diagnosed according to a clinician's general assessment, which was based on genetic confirmation (if available), family history, neuroradiological readings of brain structure, overall health history, and additional clinical information to rule out other possible diagnoses. Participants were excluded if they had a history of an additional neurological disorder (any neurological disorder in controls), a history of head injury with loss of consciousness longer than 5 min, learning disability, or psychiatric disorder such as bipolar disorder, major depressive disorder, or psychosis. Patient information is listed in Table 1 and group demographics are described in Table 2.

This study was approved by the Johns Hopkins University School of Medicine Institutional Review Board. Study protocol was performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards. Informed consent was obtained from all study participants.

Rey-Osterrieth Complex Figure Task

Experimental Paradigm

Participants drew the figure at four time points with unrestricted time limits:

1. Copy: Participants were shown the ROCF image on a computer monitor, with instructions to copy the figure. The image was removed when the participant indicated he or she was done.
2. Immediate: Immediately after copy, participants were asked to draw the ROCF from memory.
3. 3-min: Three minutes after the immediate condition was completed, participants were asked to draw the ROCF from memory.
4. 30-min: Thirty minutes from copy completion, participants were asked to draw the ROCF from memory.

Digitizing Tablet

Participants used a stylus to draw the figures on a Wacom Intuos digital tablet with dimensions $48.7 \times 31.8 \times 1.2$ cm, which was connected to a Dell Optiplex 380. MovAlyzer v6.1 (Neuroscript LLC, Tempe, AZ, USA) software recorded stylus movements at a sampling rate of 133 Hz. Drawings were displayed in real time on the Dell E171FP computer monitor.

ROCF Scoring

Accuracy scoring was performed according to the criteria established by Osterrieth [25] and standardized by Meyers and Meyers [48], which assess the accuracy and placement of 18 elements in the complex figure. Each element received a score of 2 if it was accurately drawn and correctly placed, 1 if it was accurate or correctly placed but not both, 0.5 if it was neither accurate nor correctly placed but was recognizable, and 0 if it was unrecognizable or omitted. Total scores were computed by summing the scores for all 18 elements, giving a maximum possible score of 36.

We scored the level of organization in people's approach to drawing the ROCF according to criteria developed by Osterrieth [25] and summarized in Lezak [49]. Osterrieth identified seven levels of organization when drawing the

Table 1 Patient information

Code	Age	Sex	Diagnosis	ICARS total	Complex figure	Block design	Bistable images
1	41	M	SCA1		X		
2	62	M	SCA1	34		X	X
3	45	F	SCA1	13		X	X
4	70	F	SCA2		X		
5	42	M	SCA2	43		X	X
6	59	F	SCA2	50		X	X
7	41	F	SCA2	54		X	X
8	58	F	SCA3	26	X		
9	43	M	SCA3		X		
10	39	F	SCA3		X		
11	45	F	SCA3		X	X	
12	53	F	SCA3			X	
13	54	F	SCA5	26		X	X
14	73	F	SCA6		X		
15	71	F	SCA6		X		
16	48	F	SCA6	2	X		
17	58	M	SCA6	23	X		
18	64	M	SCA6		X	X	
19	64	F	SCA6	58	X	X	X
20	66	F	SCA6	33		X	X
21	63	F	SCA6	38		X	X
22	41	M	SCA8		X		
23	45	F	SCA8	39	X		
24	48	M	SCA8	28	X		
25	50	F	SCA14		X		
26	62	F	ADCA	33	X		
27	67	M	ADCA	8.5	X	X	
28	53	F	ADCA	39	X	X	
29	76	F	ADCA	33	X		
30	51	M	ADCA	20	X		
31	56	F	ADCA	20	X		
32	63	M	ADCA		X	X	X
33	60	M	CAUE	34		X	X
34	58	M	CAUE		X		
35	62	M	CAUE	22	X		
36	59	M	CAUE	36	X		
37	42	F	CAUE		X		
38	73	M	CAUE		X		
39	46	F	CAUE		X		
40	72	F	CAUE	36	X		
41	53	M	CAUE	50	X		
42	82	M	CAUE	22	X		
43	56	F	CAUE	57	X	X	
44	72	M	CAUE	37		X	
45	55	M	CAUE		X	X	X
46	48	F	CAUE	30	X		X
47	36	M	CAUE	61			X
48	51	F	CAUE	28		X	X
49	59	F	CAUE	50		X	X

ICARS International Cooperative Ataxia Rating Scale. Under sex: *M* male, *F* female. Under diagnosis: *SCA* spinocerebellar ataxia, *ADCA* autosomal dominant cerebellar ataxia, *CAUE* cerebellar ataxia, unknown etiology. Under ICARS: *U* unavailable

figure, in which level 1 signified the highest level of organization, and level 7 signified the lowest. Out of 60 adults tested, Osterieth found that 55.6% of people used level 1, 26.6% used level 2, 1.6% used level 3, and 15% used level 4; only young children used levels 5 through 7 [25]. As shown in Fig. 2, figure organization can vary from highly organized (e.g., level 1) to unplanned and piecemeal (e.g., level 5). The

video supplement demonstrates a disorganized (level 5) approach to drawing the ROCF from our own data.

For maximum consistency, two raters (among SK, RB, JM, MS, SL) came to a consensus on each organization and accuracy score. Raters ignored imperfections in the ataxia patients' drawings that appeared to result from tremors or other motor disturbances related to poor coordination.

Supplementary Tasks

Participants also completed a number of supplemental cognitive and motor tests to aid interpretation of the ROCF results.

Block Design

A block design task was administered to evaluate visual perception of global and local features. Participants used blocks to recreate a series of designs, shown on a computer screen, in which each block contained two white sides, two red sides, and two bicolor sides, following the WAIS block design subtest [50]. There were eight trials with 2-min time limits, and each had its own unique design that varied by segmentation (segmented or unsegmented) and gestalt properties (maximum or minimum gestalt), with two trials of each permutation (see Fig. 3). “Segmentation” refers to whether the blocks were separated or touching, and “gestalt” properties refer to whether the blocks fit together to form a cohesive shape or “gestalt.” Per Caron et al. [45], designs with maximum gestalt are more difficult to recreate because they require participants to break up the global image into a grid of individual blocks. Performance measures included accuracy and reaction time. Errors were categorized as face errors (e.g., a red face instead of a white face) or orientation errors (e.g., a bicolor face that was rotated incorrectly). Then, error rates were computed as a percentage of the total number of blocks placed at the end of the trial, as determined by the earlier of a 2-min time limit or when the participant declared they had finished.

Bistable Images

We designed this task to measure the ability to perceive and switch between global features within a single image. Participants were asked to identify both items embedded in a series of bistable images, such as the duck/rabbit illusion (see Fig. 4). This required switching from the perception of one gestalt to another. Accuracy of image detection and response time were recorded in each 60-s trial. Additionally, if a participant did not offer a correct response within 20 s, a hint was provided, giving an opportunity for half credit. Thus, a maximum score of 2 was given if both images were perceived without assistance.

Symbol Digit Modalities Test

In the SDMT, a measure of processing speed [51, 52], participants were provided with a legend that paired unique symbols with the numbers 1–9, and this was visible as a reference throughout the test. Then, they were shown a list of symbols and asked to state the corresponding numbers aloud [53]. The experimenter recorded responses in order to minimize motor

demands that would have put the cerebellar ataxia group at a disadvantage. Scores were based on the total number of correct responses given within 2 min.

Digit Span

In the Digit Span test from the Wechsler Adult Intelligence Scale-Third Edition [54], a classic measure of verbal working memory, participants repeated a series of three, up to eight, numbers forward and backward. The number of correct trials was tallied for “forward,” “backward,” and overall conditions.

Finger Tapping

In the finger tapping task from the Halstead-Reitan Battery [55], a standard measure of motor function, participants were asked to tap their index fingers as quickly as possible for 10 s. Taps were recorded for each hand, up to 10 trials or fewer if criteria for tap consistency were met. The mean number of finger taps was computed for each hand.

CES-D

In order to control for any influence of depressive symptoms on cognitive and motor performance, participants were administered the CES-D, which measures symptoms of depression based on a series of 20 items [56]. On this questionnaire, a score of 16 or higher indicated active depression.

WRAT

Participants took the reading subsection of the Wide Range Achievement Test (3rd ed.) (WRAT-3) to measure premorbid verbal intelligence and education level [57]. The task involves reading a series of words with irregular spellings [58].

ICARS

Cerebellar ataxia patients were administered the ICARS [59], which assesses motor function across a range of different categories including posture and gait, kinetic functions, speech, and eye movements. This was administered to assess any relationship between neurological signs and test performance.

Statistical Methods

Because tests within this study included quantitative, ordinal, and categorical data, different statistical methods were employed for each data type. For quantitative data, such as figure accuracy scores, independent sample *t* tests and Pearson correlations were used. For ordinal data such as figure organization scores, linear-by-linear associations and Spearman correlations were used. For categorical data such as

Table 2 Group demographics, neurological assessments, and task scores

	Controls					Patients					<i>p</i> value
	Mean	SD	Min	Max	<i>n</i>	Mean	SD	Min	Max	<i>n</i>	
Demographics											
Gender ^a (M:F)	28:32	–	–	–	60	21:28	–	–	–	49	0.69
Handedness ^a (R:L:M)	47:1:4	–	–	–	52	37:2:0	–	–	–	39	0.15
Age ^b	58.88	9.12	37.00	77.00	60	56.22	11.08	36.00	82.00	49	0.17
Years of education ^b	16.11	2.43	12.00	23.00	60	16.38	2.78	12.00	22.00	49	0.59
Cognitive and motor measures											
WRAT ^b	50.31	3.43	42.00	56.00	59	49.63	2.84	42.00	55.00	49	0.28
CES-D ^b	7.00	7.72	0.00	28.00	31	9.45	8.53	0.00	42.00	47	0.20
ICARS total	–	–	–	–	–	33.86	14.26	2.00	61.00	32	–
Digit span forward ^b	10.55	2.25	6.00	15.00	47	10.50	2.10	6.00	15.00	38	0.91
Digit span backward ^b	7.19	2.22	4.00	13.00	47	6.61	1.62	4.00	11.00	38	0.18
SDMT ^b	61.35	13.24	33.00	107.00	51	45.29	15.27	10.00	77.00	38	<0.01*
Tapping (dominant) ^b	47.55	8.77	26.30	62.80	39	32.32	10.00	12.60	54.00	38	<0.01*
Rey-Osterrieth complex figure											
Figure organization (1–7)											
Copy ^c	2.48	1.06	1.00	4.00	42	3.20	1.21	1.00	5.00	35	<0.01*
3-min ^c	2.38	1.27	1.00	5.00	42	3.09	1.36	1.00	5.00	35	0.02*
30-min ^c	2.45	1.33	1.00	5.00	42	3.06	1.43	1.00	5.00	35	0.06
Figure accuracy (0–36)											
Copy ^b	29.90	4.42	15.50	36.00	42	25.63	8.26	6.50	36.00	35	<0.01*
3-min ^b	14.98	5.55	5.50	25.00	42	15.01	6.56	2.00	28.50	35	0.98
30-min ^b	15.01	5.55	4.00	27.00	42	15.37	6.67	4.50	29.00	35	0.80
Response time (s)											
Copy ^b	252.05	86.42	113.41	566.90	42	305.06	134.57	109.62	608.47	35	0.05*
3-min ^b	133.40	67.31	46.36	453.99	41	173.93	107.63	41.29	690.91	35	0.05*
30-min ^b	123.47	59.03	47.56	399.13	42	187.12	145.34	67.43	910.76	35	0.02*
Block design											
Error rates (%)											
Orientation ^b	0.80	1.49	0.00	6.73	29	2.67	2.93	0.00	12.63	21	0.01*
Face ^b	1.13	2.37	0.00	11.00	29	1.50	2.49	0.00	10.53	21	0.60
Timeouts ^b	0.48	0.91	0.00	3.00	29	1.00	1.05	0.00	3.00	21	0.07
Accuracy											
Seg. min-gestalt ^b	99.43	1.72	94.44	100.00	29	98.94	2.24	94.44	100.00	21	0.39
Seg. max-gestalt ^b	100.00	0.00	100.00	100.00	29	97.32	4.19	83.33	100.00	21	<0.01*
Unseg. min-gestalt ^b	98.85	3.44	83.33	100.00	29	97.35	4.16	88.89	100.00	21	0.19
Unseg. max-gestalt ^b	89.64	19.04	28.57	100.00	29	86.97	21.64	16.67	100.00	21	0.65
Response time (s)											
Seg. min-gestalt ^b	27.98	10.75	15.47	66.70	29	41.48	15.74	18.14	71.50	21	<0.01*
Seg. max-gestalt ^b	27.15	10.07	15.87	65.25	29	40.98	18.72	19.75	89.50	21	<0.01*
Unseg. min-gestalt ^b	33.99	11.89	17.74	66.87	29	52.28	17.54	24.96	79.07	21	<0.01*
Unseg. max-gestalt ^b	66.41	35.36	23.83	120.00	29	91.90	27.19	37.83	120.00	21	<0.01*
Bistable images											
Accuracy (out of 8 images)											
Total ^b	6.61	1.30	3.50	8.00	19	6.47	0.90	5.00	7.50	16	0.73
First image ^b	3.76	0.31	3.00	4.00	19	3.84	0.35	3.00	4.00	16	0.47
Second image ^b	2.84	1.09	0.00	4.00	19	2.63	0.72	1.50	3.50	16	0.50
Response time (s)											

Table 2 (continued)

	Controls					Patients					<i>p</i> value
	Mean	SD	Min	Max	<i>n</i>	Mean	SD	Min	Max	<i>n</i>	
First image RT ^b	8.34	4.60	2.50	17.00	19	7.68	4.42	2.25	18.25	16	0.67
Second image RT ^b	14.25	5.71	3.00	27.50	18	14.37	5.21	6.00	25.00	16	0.95

Figure organization scores are on a scale from 1 to 7, where lower scores indicate greater figure organization. On the other hand, accuracy scores are on a scale from 0 to 36, where 36 represents the best possible score. There are 18 total elements in the complex figure, and each element received 2 points if it was accurate and correctly placed, 1 point if it was accurate or correctly placed but not both, and 0.5 points if it was neither accurate nor correctly placed but was recognizable. All values are shown as raw scores, not scaled scores. To facilitate visualization of group comparisons, we performed direct tests between groups, as described below. However, in the “Results” section, we also report the omnibus mixed-design ANOVAs. Chi-squared tests (a) were used to compare categorical variables such as gender and handedness across groups. Independent samples *t* tests (b) were used to compare quantitative variables such as age and figure accuracy scores. Linear-by-linear associations (c) were used to compare ordinal variables in figure organization. (* = did not survive Bonferroni correction, * = significant after Bonferroni correction)

CES-D Center for Epidemiologic Studies Depression Scale, *WRAT* Wide Range Achievement Test, *ICARS* International Cooperative Ataxia Rating Scale, *SDMT* Symbol Digit Modalities Test. Under gender: *M* male, *F* female. Under handedness: *R* right, *L* left, *M* mixed. For block design: *seg* segmented, *unseg* unsegmented, *min-gestalt* minimum gestalt features, *max-gestalt* maximum gestalt features

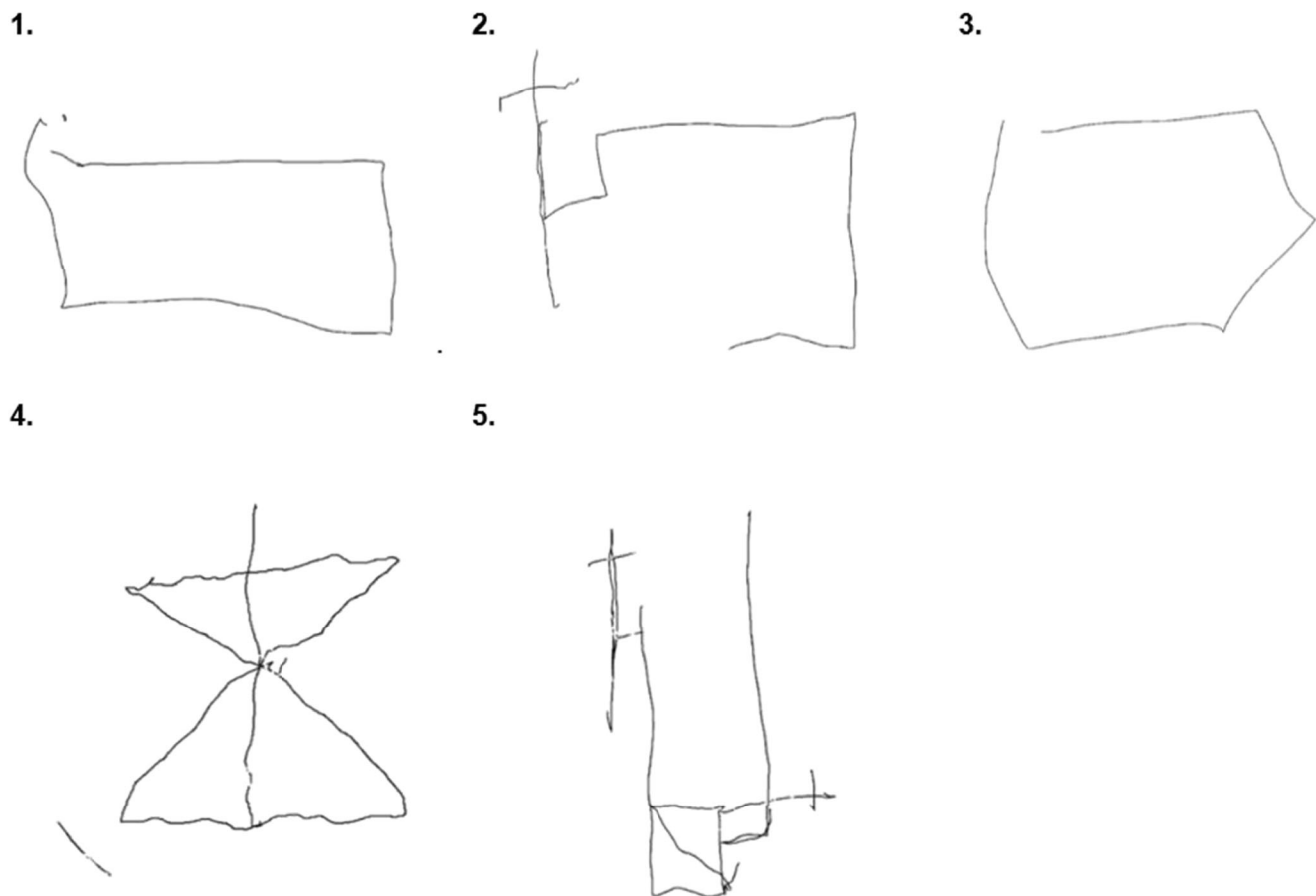


Fig. 2 Levels of organization in the Rey-Osterrieth complex figure task. Organization score = 1: the participant begins by drawing the large rectangle. Organization score = 2: the participant begins by drawing a detail attached to the large rectangle and then proceeds to the rectangle. Organization score = 3: the participant begins by drawing the overall contour of the shape and then adds internal details. Organization score = 4: the participant draws details one by one without an organizing

structure. Organization score = 5: the participant copies discrete parts of the figure with no semblance of organization (find a full time lapse of this drawing in the [video supplement](#)). Organization score = 6 (no example): the participant substitutes a similar object, such as a boat or a house. Organization score = 7 (no example): the participant creates an unrecognizable scrawl

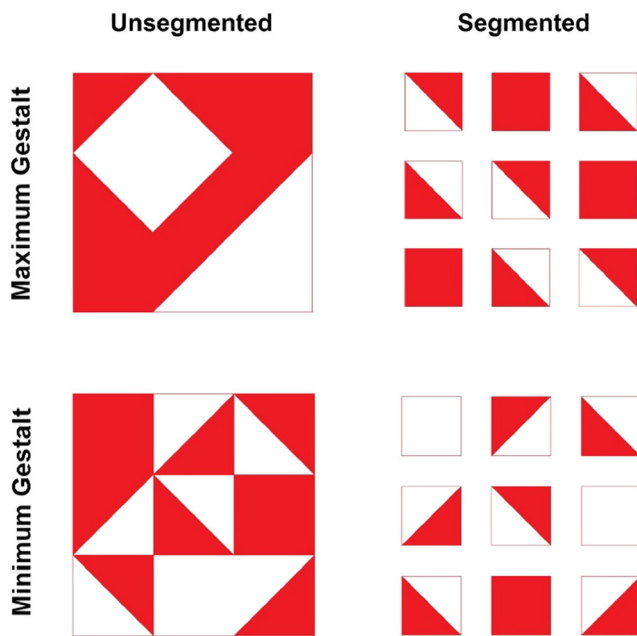


Fig. 3 Block design. Block designs varied by perceptual cohesiveness and segmentation. Segmentation refers to whether the blocks were separated or touching, whereas gestalt refers to whether they fit together to form a larger image

handedness and gender, Pearson chi-square tests were used. Significant interactions revealed by ANOVAs were followed up with post hoc *t* tests. Aside from post hoc *t* tests, all other *t* tests and correlations were Bonferroni corrected.

Results

Complex Figure

Figure Organization

Figure organization scores were compared between the two groups. For simplicity, we will focus on the 3- and 30-min

Fig. 4 Examples of bistable images. There were four bistable images, each of which could be perceived in two distinct ways. Here, image 1 can be seen as a young woman or an old woman, while image 2 can be seen as a duck or a rabbit



Image 1

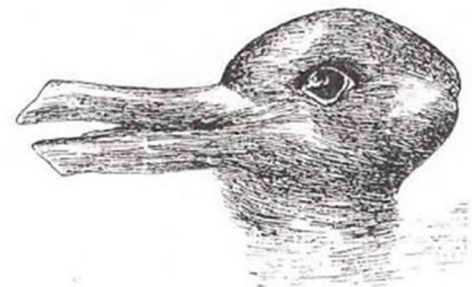


Image 2

conditions because the immediate condition generalized to these later time points. A 3×2 mixed-design ANOVA with condition (3: copy, 3-min, 30-min) as a within-subjects factor and group (2: ataxia, controls) as a between-subjects factor revealed a main effect of group, $F(1, 75) = 6.69$, $p < 0.05$, indicating that ataxia patients showed less efficient figure organization than did controls; see Table 2 for group means at each condition and Fig. 5 for frequencies of different levels of organization in controls and ataxia patients. There was no main effect of condition or interaction of group by condition (all p values > 0.05), suggesting that when a participant employed a level of organization at copy, they generally maintained it at subsequent recall time points, consistent with the results of Osterrieth [25]. According to chi-square linear-by-linear associations, the frequencies of organization levels differed significantly between groups at copy, $\chi^2(1, N = 77) = 7.17$, $p < 0.01$ and at 3-min: $\chi^2(1, N = 77) = 5.22$, $p < 0.05$, and trended significance at 30-min: $\chi^2(1, N = 77) = 3.56$, $p = 0.06$.

In addition, we observed that controls showed a correlation between figure organization and recall accuracy, which did not survive Bonferroni correction. However, this relationship was entirely absent in ataxia patients (see Table 3).

Figure Accuracy

Surprisingly, despite their propensity toward lower figure organization, ataxia patients were not impaired on recall accuracy. A 2×2 mixed-design ANOVA with condition (2: 3-min, 30-min) as a within-subjects factor and group (2: ataxia, control) as a between-subjects factor showed no main effect of group, condition, or interaction of group by condition (all p values > 0.05). The copy condition was excluded from this analysis because it did not rely on recall ability. Ataxia patients were, however, significantly less accurate on the copy condition ($p < 0.01$, see Table 2).

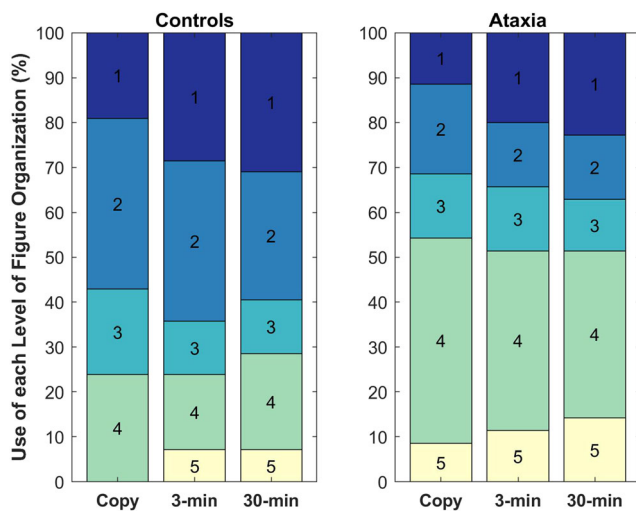


Fig. 5 Figure organization in controls and ataxia patients across organization conditions. Ataxia patients showed less efficient figure organization across all conditions collapsed together. Assessing each time point separately with chi-square associations, frequencies of figure organization scores were significantly different between controls and ataxia patients at copy ($p < 0.01^*$) and 3-min ($p = 0.02^*$) and trended significance at 30-min ($p = 0.06$)

Figure Response Time

Total response times for participants to complete the figure differed between groups. A 3×2 mixed-design ANOVA with task condition (3: copy, 3-min, 30-min) as a within-subjects factor and group (2: ataxia, control) as a between-subjects factor revealed a main effect of group, $F(1, 74) = 6.61$, $p < 0.05$, in which ataxia patients generally took longer to complete the figure, and a main effect of condition $F(2, 148) = 98.34$, $p < 0.001$, in which the copy condition took significantly longer to complete overall (see Table 2). There was no interaction of group by condition.

Block Design

In order to investigate why ataxia patients generated less figure organization than did controls, we administered a supplemental block design task that focused on visual perception abilities. A $2 \times 2 \times 2$ mixed-design ANOVA with segmentation (2: segmented or unsegmented) and gestalt (2: maximum and minimum gestalt) as within-subject factors and group (2: patients or controls) as a between-subject factor revealed main effects of segmentation $F(1, 48) = 14.08$, $p < 0.001$ and gestalt $F(1, 48) = 14.06$, $p < 0.001$, and an interaction of gestalt by segmentation $F(1, 48) = 10.15$, $p < 0.005$, but no effect or interaction by group (all p values > 0.05). However, ataxia patients made significantly more orientation errors than did controls ($p < 0.05$), and this survived Bonferroni correction. In terms of response times, ataxia patients were slower than were controls when completing the block design, but patients were not disproportionately slower on any specific condition. A $2 \times$

2×2 mixed-design ANOVA with segmentation (2: segmented or unsegmented) and gestalt (2: maximum and minimum gestalt) as within-subject factors and group (2: patients or controls) as a between-subject factor revealed main effects of group $F(1, 48) = 15.63$, $p < 0.001$, segmentation $F(1, 48) = 106.01$, $p < 0.001$, and gestalt $F(1, 48) = 97.98$, $p < 0.001$, and an interaction of gestalt by segmentation $F(1, 48) = 106.17$, $p < 0.001$, but no interactions by group (all p values > 0.05).

Bistable Images

We administered bistable images to assess gestalt perception abilities specifically. With respect to image detection accuracy, 2×2 ANOVA with image number perceived (2: first vs. second) as a within-subject factor and group (2: ataxia or control) as a between-subject factor revealed an effect of image number $F(1, 33) = 56.78$, $p < 0.001$, but not a main effect of group or interaction of group by image number (all p values > 0.05), suggesting that the groups accurately detected images at similar frequencies. With respect to response time, 2×2 ANOVA with image number (2: first vs. second) as a within-subject factor and group (2: ataxia or control) as a between-subject factor revealed an effect of image number $F(1, 32) = 30.05$, $p < 0.001$, but no main effect of group or interaction of group by image, suggesting that the groups detected images with similar response times.

Correlations with Supplemental Tests

To further explore the basis of ROCF performance, we took a closer look at the relationship between ROCF scores and other cognitive and motor factors (see Table 3). We observed that ataxia patients displayed a strong positive correlation between figure recall accuracy and scores on the SDMT, a measure of processing speed, whereas this relationship was notably absent in controls. The processing speed and figure accuracy correlation was not likely affected by eye movement disorders (e.g., nystagmus) because oculomotor scores from the ICARS did not correlate with figure recall (Table 3) or the SDMT (Table 4). Similarly, although the speech subsection of the ICARS correlated with SDMT scores for ataxia patients, overt speech was not an element in the complex figure test and not a likely confound. A partial correlation was upheld between SDMT and figure recall accuracy while controlling for ICARS total and dominant hand finger tapping (see Table 4, all p values < 0.05), reducing the role of motor impairments as a confound. Finally, a partial correlation was upheld between figure recall accuracy and SDMT while controlling for working memory (as measured by digits forward and backward) in ataxia patients (see Table 4, all p values < 0.05). Thus, the

Table 3 Recall accuracy correlations with figure organization and supplemental tasks

Correlation Coefficient -.75 0 .75			Cognitive Assessments			Motor Assessments		
			Figure Organization ^a	Digits Backwards ^b	SDMT ^b	Tapping: Dominant ^b	ICARS: Total ^b	ICARS: Oculomotor ^b
Figure Accuracy	Controls	Copy	0.00	0.23	0.40 ^(*)	0.41*	---	---
		3-min	-0.33 ^(*)	0.02	0.17	0.14	---	---
		30-min	-0.34 ^(*)	0.12	0.19	0.23	---	---
	Ataxia	Copy	-0.14	0.41 ^(*)	0.62*	0.34 ^(*)	-0.34	0.00
		3-min	-0.16	0.22	0.53*	0.10	-0.12	-0.09
		30-min	-0.19	0.16	0.56*	0.14	-0.14	0.05

Under figure organization, correlations with accuracy (copy, 3-min, or 30-min) used the respective figure organization time point and are shown as Spearman correlations, indicated by the superscript “a.” All other values are shown as Pearson correlation *r* values, indicated by the superscript “b.” Additionally, all values used for these correlations were raw scores, not scaled scores. ^(*) = did not survive Bonferroni correction, * = significant after Bonferroni correction

SDMT Symbol Digit Modalities Test, ICARS International Cooperative Ataxia Rating Scale

relation between processing speed and figure accuracy recall was strong in the ataxia patient group.

Interestingly, ataxia patients displayed an inverse relationship between figure organization and the kinetic subscore in the ICARS: at copy, $r(19) = -0.48, p = 0.04^*$; at 3-min, $r(19) = -0.53, p = 0.02^*$; and at 30-min, $r(19) = -0.50, p = 0.03^*$, meaning that higher severity of motor impairments was associated with better figure organization. This relationship was significant at all time points, but did not survive Bonferroni correction.

Discussion

In this study, we investigated visuospatial organization and recall in cerebellar ataxia and found that ataxia patients were less organized than controls in their approach to copying and recalling a complex figure. Secondary tests of perceptual processing, however, did not find any evidence of problems with gestalt perception, indicating that their organizational approach to figure drawing was not related to an inability to perceive the overall figure image construct. Despite their less

Table 4 SDMT correlations with recall accuracy and supplemental tests

Correlation Coefficient -0.75 0 .75		Figure Accuracy			Motor Ability					Working Memory		
		Copy	3-min	30-min	Tapping (Dominant)	ICARS				Digits Forward	Digits Backward	
						Total	Posture / Gait	Kinetic	Speech			Oculo-motor
Controls	SDMT	0.40*	0.17	0.19	0.07	---	---	---	---	---	0.32 ^(*)	0.43*
	SDMT (Controlling for Motor Skills)	0.41 ^(*)	0.16	0.18	---	---	---	---	---	---	0.32	0.45*
	SDMT (Controlling for Working Memory)	0.34 ^(*)	0.20	0.16	0.14	---	---	---	---	---	---	---
Ataxia	SDMT	0.62*	0.53*	0.56*	0.42 ^(*)	-0.71*	-0.72*	-0.52 ^(*)	-0.61*	-0.22	0.36 ^(*)	0.43*
	SDMT (Controlling for Motor Skills)	0.62*	0.65*	0.68*	---	---	-0.50 ^(*)	0.48	-0.16	0.11	0.30	-0.01
	SDMT (Controlling for Working Memory)	0.55*	0.47*	0.53*	0.41 ^(*)	-0.67*	-0.67*	-0.43	-0.63*	-0.15	---	---

SDMT controlling for motor adjusts for dominant hand tapping in controls and both dominant hand tapping and ICARS total scores in ataxia patients. SDMT controlling for working memory adjusts for digits forward and backward. All values used for these correlations were raw scores, not scaled scores. Also all values are shown as Pearson correlation *r* values, ^(*) = did not survive Bonferroni correction, * = significant after Bonferroni correction ICARS International Cooperative Ataxia Rating Scale, SDMT Symbol Digit Modalities Test

efficient organization strategies, ataxia patients managed to achieve a normal recall accuracy relative to that of controls, which goes against prior studies (and control performance in this study) that has associated recall and organization abilities [25, 37]. This indicates that ataxia patients employed a compensatory mechanism to achieve normal performance.

Supplemental tests were suggestive of alternative mechanisms used by the ataxia patients for figure recall. Specifically, patients showed a strong correlation between recall accuracy and performance on the SDMT, considered a measure of processing speed [51, 60]. This test also might incorporate elements of working memory and eye movements, but lack of correlations in our data argued against the influence of these factors (see Tables 3 and 4) [61, 62]. Because processing speed is broadly defined as “the rate at which people process information” [63], it is difficult to discern the specific approach ataxia patients might employ. Still, processing speed may track general cognitive facilities and reflect a patient’s ability to devise one of many possible alternative approaches to figure recall, which may have varied from patient to patient. If specific methods of figure recall varied, this would explain why figure recall correlated with processing speed in the ataxia cohort but not with more specific variables, such as working memory or motor skills (see Table 3).

Because the secondary tests of perceptual processing, as measured by block design and bistable image tests, did not reveal deficits in gestalt perception in cerebellar ataxia, results suggest that the patients displayed less figure organization as a result of difficulties with another aspect of complex figure drawing, such as efficient planning [64–66] or action sequencing [67]. In the same way that the cerebellum uses internal models to coordinate and adapt movement sequences, it may contribute to the coordination and adaptation of action sequences in complex cognitive-motor tasks such as playing chess [68], implicit sequence learning [69–71], and puzzle assembly [67]. There is evidence for a cerebellar role in generating and adapting effective action sequences in spatial tasks: for example, rats whose cerebellums were surgically removed showed specific difficulty learning and adapting effective search procedures in a series of spatial navigation tasks [72–74]. According to this interpretation, the ataxia patients perceived and encoded the complex figure normally but struggled to generate effective procedures for sequentially reproducing the shape in an efficient way. From the start of the drawing, they were less likely to draw the global outline first (level 1 figure organization), or to draw the global outline after entering one or two details (level 2 figure organization). This interpretation is further supported by ataxia patients’ orientation errors (rotation errors) in the absence of perceptual deficits in the block design task. Therefore, further research should aim to clarify how action sequencing contributes to performance on visuospatial tasks in cerebellar ataxia. This would include real versus imagined tool (e.g., stylus) use.

Evidence suggests that motor simulation plays a role in reproducing the Rey-Osterrieth complex figure [75], and the cerebellum may maintain separate internal models for real and imagined tool use [76, 77].

Motor impairment could be proposed as an explanation of why ataxia patients have lower figure organization. One well-documented motor symptom of cerebellar ataxia is decomposition of movement, measured by the finger to nose test, the knee tibia test, and Archimedes’ spiral in the kinetic section of the ICARS. This symptom could make it more difficult for ataxia patients to draw the larger shapes of the figure continuously, such that they have to resort to a more piecemeal approach. However, the segmentation subscores of the ICARS did not correlate with figure organization in the cerebellar ataxia group (all p values > 0.05). Moreover, ataxia patients showed an inverse correlation between motor skills and figure organization, as measured by the kinetic subscores in the ICARS, in which lower motor skills (somewhat counter-intuitively) tracked with more efficient figure organization. Therefore, in ataxia, poor figure organization was not a result of frank motor impairment.

Another possible explanation for the present findings is that ataxia patients struggle with motor simulation, which may play a role in developing an organized procedure for drawing the figure. For example, De Lucia et al. [75] found evidence that motor simulation contributes to effective encoding of the figure. De Lucia conducted a study in which participants memorized the Rey-Osterrieth complex figure with one hand behind their back. Here, people who hid their dominant hand behind their back recalled the shape with less accuracy than people who hid their nondominant hand, suggesting that motor simulation played a role in encoding the figure. If motor simulation is impaired in cerebellar ataxia, this could help explain their less organized approach to the ROCF task. At the same time, their ability to motor simulate may not directly track their motor skills. For example, Imamizu and Kawato [76] and Higuchi et al. [77] showed that the cerebellum maintains separate internal models for real and imagined tools. As a result, future research should investigate how motor simulation contributes to visuospatial abilities in motor disorders like cerebellar ataxia.

One limitation of this study was that patients had heterogeneous cerebellar ataxia etiologies, differing slightly in modes of inheritance, clinical symptoms, and overall brain pathology possibly extending outside the cerebellum. To minimize the heterogeneity of the sample, all ataxia patients included in this study had either been genetically confirmed with a known cerebellar ataxia subtype or diagnosed according to a clinician’s general assessment, based on a variety of factors including family history, neuroradiological readings of brain structure, symptom progression, and overall health history. Even though we cannot rule out how brain pathology outside the cerebellum might have contributed to cognitive impairments

in some of the patients [78], all diagnoses were characterized by cerebellar dysfunction. As a result, the most likely explanation for ataxia patients' performance was cerebellar degeneration and disruption of cerebro-cerebellar networks.

A second limitation was that the use of a stylus and tablet may have been more challenging than the use of typical paper-and-pencil recordings, possibly putting the ataxia patients at a disadvantage. However, controls achieved similar copy and recall scores to paper-and-pencil norms [79], and within our study, controls and ataxia patients exhibited similar recall scores. If difficulty using the device had disproportionately affected the ataxia group, then we would have expected a group difference on drawing accuracy in addition to figure organization.

Conclusion

In summary, our findings indicate that cerebellar ataxia patients exhibit impairments of visuospatial organization in the absence of visuospatial recall. Patients seem to be able to compensate for lack of organization abilities, a skill that is predicted by processing speed. Many tasks in everyday life involve an organizational component—from planning a route to the grocery store to preparing a meal. As a result, two methods of clinical therapy warrant further research: improving a patient's ability to develop and use organized procedures and bolstering processing speed, the alternative mechanism ataxia patients already seem to employ.

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Compliance with Ethical Standards

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

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