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Cerebellum Atrophy and Development of a Peripheral Dysgraphia: A Paediatric Case

Maria Concepción Fournier del Castillo · Maria Jesus Maldonado Belmonte · Maria Luz Ruiz-Falcó Rojas · Miguel Ángel López Pino · Jordi Bernabeu Verdú · Jesús M. Suárez Rodríguez

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Abstract Two types of dysgraphia may be distinguished: the core ones, which reflect damage to the linguistic orthographic routes, and the peripheral ones, produced by alterations in the selection or execution of graphic motor patterns. We report the case of an 8-year-old male child, who consulted specialists due to difficulties in writing, with a background of acute cerebellar swelling at the age of 4. The writing pattern he has developed shows characteristic errors of a peripheral dysgraphia. The magnetic resonance imaging taken during the neuropsychological evaluation shows a mild atrophy in the cerebellum cortex. Our case is similar to previous studies of adult patients and equally supports the fact that the functional network responsible for the peripheral control of writing abilities may include the cerebellum, which not only maintains previously learnt writing processes but is also involved in the evolutionary acquisition of this ability.

Keywords Cerebellum atrophy · Language · Writing disability · Peripheral dysgraphia

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M. C. Fournier del Castillo (✉)
Psychiatry and Psychology Unit,
Niño Jesus Children's University Hospital,
Avda. Menéndez Pelayo, 65,
28009 Madrid, Spain
e-mail: cfournier.hnjs@salud.madrid.org

M. J. Maldonado Belmonte
Psychiatry Unit, Gregorio Marañón University Hospital,
Madrid, Spain

M. L. Ruiz-Falcó Rojas
Neurological Unit, Niño Jesus Children's University Hospital,
Madrid, Spain

M. Á. López Pino
Department of Radiology,
Niño Jesus Children's University Hospital,
Madrid, Spain

J. Bernabeu Verdú
Paediatric Oncology Unit, La Fe University Hospital,
Valencia, Spain

J. Bernabeu Verdú · J. M. Suárez Rodríguez
Research Methods and Diagnosis in Education,
University of Valencia,
Valencia, Spain

Introduction

The writing process has two components: linguistic and motor. The first generates the orthography of words, and the second converts abstract graphic information into motor instructions to execute script movements [1]. Dysgraphia is the term used to refer to writing disorders caused by neurological damage, two types of which may be distinguished: the core ones, which reflect damage to the linguistic orthographic routes, and the peripheral ones, produced by alterations in the selection or execution of graphic motor patterns. The peripheral writing process converts the allographic units (form of letter representation) into writing movements, which constitute the stroke sequences that specify direction, size, position and order.

The most researched peripheral type is spatial or afferent dysgraphia, initially described in adults by Lebrun [2]. It is characterised by the incorrect production of strokes and letters (omission and repetition), the overlapping of letters and words, exaggerated intervals between letters, sloping lines, tendency to write on the right side of the page and lack of deterioration of writing without vision [3]. In adults, this condition has been reported following right parietal lesions [4] and more recently in cerebellum lesions [5, 6]. Acquired cerebellum pathology is relatively less common in children; therefore, there are few studies describing alterations in cognitive functions following cerebellum damage.

Our research team has studied the case of a child with a background of acute cerebellar swelling and secondary cerebellum atrophy, who consulted specialists regarding a disorder in the writing acquisition process, without evidence of any other cerebellum alterations at the time. The writing pattern, which presents peripheral dysgraphia characteristics and is associated with a cerebellar pathology, is similar to those already described in adults.

Case Report

Clinical Description

A male patient aged 8 years and 10 months, in his third year of primary education with normal knowledge acquisition and no school failure, consulted specialists due to increasing difficulties in the writing acquisition process with respect to his school group. These writing alterations were not accompanied by any other specific learning disorder in reading or arithmetic abilities.

The patient was hospitalised at the age of 4 years and 6 months, presenting a low level of consciousness and vomiting. After 6 days, he began showing symptoms of hyperthermia with episodic headaches, accompanied by abdominal pain. During exploration, a fluctuating level of consciousness was distinguishable, with moments of stupor. He was able to adopt a sitting position although with cephalic and torso tremors. Moreover, he was able to remain on his feet for some instances, and his strolling was very clumsy, requiring help. The patient obtained a score of 27/30 in the Brief Ataxia Rating Scale. Other scores were (a) gait, 7 (walking possible only with one accompanying person); (b) knee-tibia test right, 4 (lowering jerkily with extremely long lateral movements); (c) knee-tibia test left, 4 (lowering jerkily with extremely long lateral movements); (d) finger-to-nose test right, 4 (dysmetria preventing the patient from reaching nose); (e) finger-to-nose test left, 4 (dysmetria preventing the patient from reaching nose); (f) dysarthria, 2 (moderate impairment) and (g) oculomotor abnormalities, 2 (prominently slowed pursuit, saccadic intrusions and nystagmus). There was no personal or family background related to the process.

In complementary analyses initially performed, the following results were observed: 278 cells/mm^3 (normal, <10 cells/mm^3) of cerebrospinal fluid (70% mononuclear), lactic acid level was 13.4 mg/dl (normal, <0.15 mg/dl), 0.55 g/L of glucose (normal, 0.32–0.85 g/L), 0.54 g/L of protein (normal, 0.25–0.44 g/L) and negative culture. The virological study presented formations of antibodies in opposition to the simple herpes virus. The magnetic resonance imaging (MRI) performed in the acute phase showed T2 WI and fluid-attenuated inversion-recovery hyperintensities and effacement of the cortico-subcortical differentiation in the inferomedial portions of the cerebellum and tonsils (Fig. 1). With the diagnosis of acute cerebellar swelling, a treatment with acyclovir and intravenous cefotaxime was initiated. The patient experienced a slow but progressive recovery of motor functions during treatment in hospital, with improvement of tone and decrease in tremors; however, motion difficulties, shudder, lack of movement coordination and nystagmus were present when he was discharged from hospital.

Four years later, he sought advice for writing problems. The neurological exploration was normal at this moment, his gait was normal, he was able to walk with feet in tandem, he had no abnormalities in the knee-tibia test and finger-to-nose test and he had no nystagmus: The MRI scan showed mild cerebellar cortex atrophy (Fig. 2) and prominent cortical sulci and widening of the subarachnoid space (Fig. 3). There was no abnormal finding in the supratentorial compartment.

Neuropsychological Examination

An extensive set of tests was applied (see Electronic supplementary material). Results are shown in Table 1.

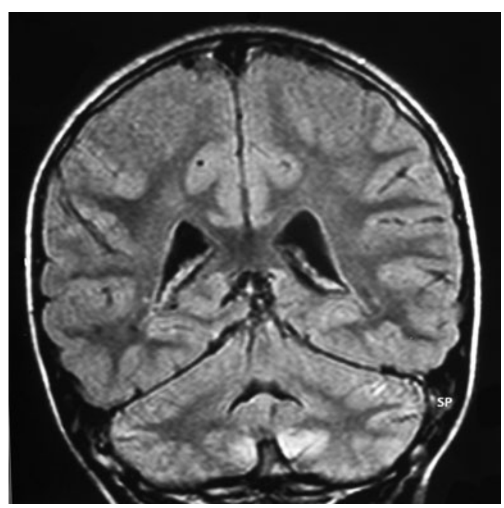


Fig. 1. Coronal fluid-attenuated inversion-recovery magnetic resonance imaging demonstrates bilateral inferomedial cerebellar hyperintensity suggesting oedema

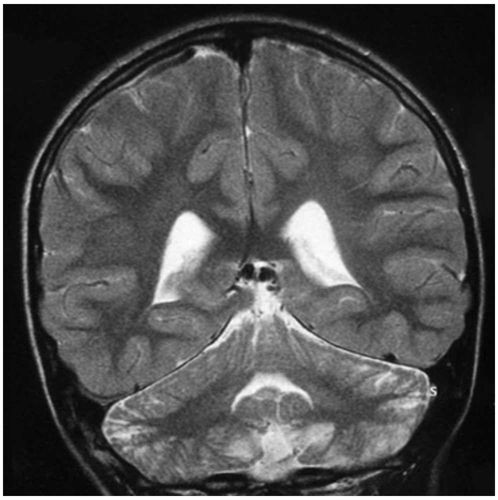


Fig. 2. Coronal FSE-T2 WI magnetic resonance imaging shows widening of the cerebellar folia due to cerebellum cortex atrophy

The patient obtained a full intelligence quotient of 105, a verbal intelligence quotient of 116 and a performance intelligence quotient of 89. No alterations were registered in the sequential or simultaneous process tasks. Acquired knowledge level was concordant with normal processing and with the average expected level at his age. The child's laterality was well established, being predominantly right-handed (Oldfield questionnaire, laterality quotient=100). In tasks which required visuomotor coordination and manual speed with his dominant right hand, performance reached average values. On the contrary, slight clumsiness and slowness were registered with the non-dominant left hand. However, no alterations were registered in visual perception, whereas exploration of visuospatial abilities registered a minor visuoconstructional apraxia. The execution of the Rey complex figure revealed difficulties in locating details in space and obtaining a global form or shape. No hemineglect was observed.

No language alterations were recorded (auditory process, receptive vocabulary, comprehension of grammar structures, naming and verbal fluency). Spontaneous language was fluid and well articulated. No alterations were registered in immediate memory, learning or remembering, using verbal and visual memory tests. No attention difficulties were found (maintained attention, processing speed, divided and selective attention). Regarding the exploration of executive functions, results were average, or slightly above average. Reading decoding and comprehension were in accordance with average levels of his age and school group; the same occurs in arithmetic and problem solving, where no alterations were found, these being slightly above average in calculation procedures.

Writing Analysis

In order to evaluate the patient's writing and his orthographic errors, a writing analysis test in Spanish was employed (TALE test, see Electronic supplementary material). By comparing the test results with the patient's global age group performance and school level, mistakes in copying and dictation, especially omissions and substitutions of letters, place him below the first percentile. Considering the fact that Spanish is quite a regular language in terms of orthography, these types of mistakes are practically non-existent in the child's normative group. Figure 4 shows performance for dictation. Table 2 shows an analysis of the patient's mistakes and those corresponding to the control group, matched in age and school year, with normal school performance and no need of specific educational support. Most errors committed by the patient are characteristic of a spatial dysgraphia, such as omissions and duplications of strokes and letters, overlapped words, unnecessary gaps between letters and inclined lines; however, no spatial negligence was observed. Moreover, alterations in the

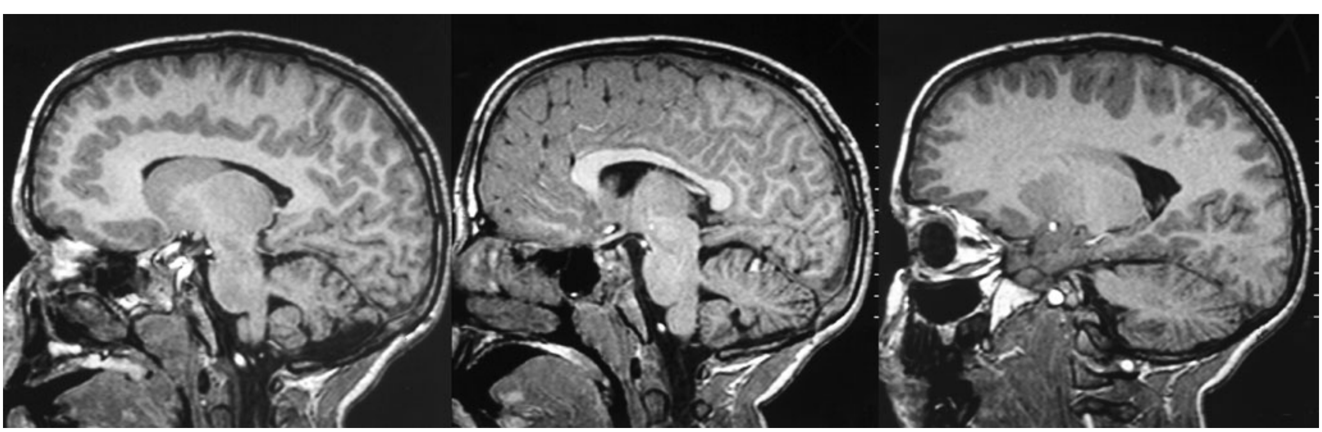


Fig. 3. Sagittal SE-T1 magnetic resonance imaging shows the cerebellar with prominent cortical sulci and mild widening of the subarachnoid space

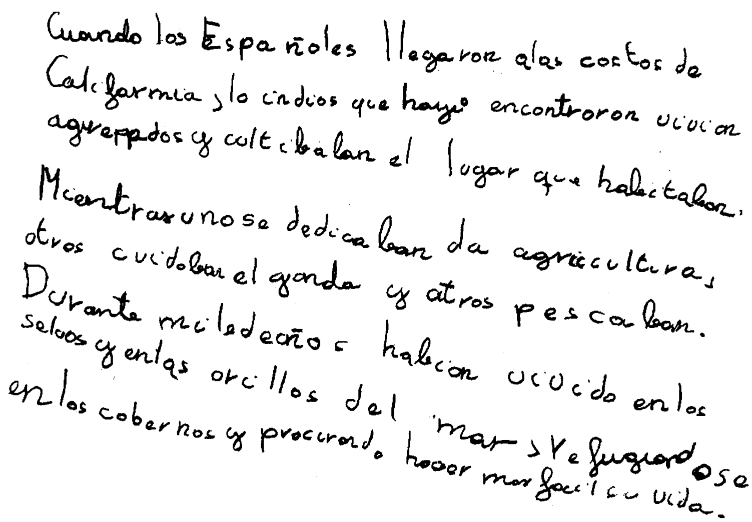
Table 1 Results of the neuropsychological evaluation

	Score	Mean	SD	*Z* scores
Verbal IQ	116	100	15	1.06
Performance IQ	89	100	15	−0.73
Full scale IQ	105	100	15	0.33
Sequential processing	105	100	15	0.33
Simultaneous processing	87	100	15	−0.86
Mental processing composite	93	100	15	−0.46
Achievement	108	100	15	0.53
Manual speed/preferred hand	38	38	9.05	0
Manual speed/non-preferred hand	61	41	14.6	−1.43
Visuomotor coordination	−3	−3.41	2.77	0.14
Visual attention	10	10	3	0
Gestalt closure	5	10	3	−1.66
Line orientation	17	19	4.3	−0.46
Face recognition	41	37.00	3.00	1.33
Visuo-constructional praxis	13.5	19.63	5.25	−1.16
Nonverbal reasoning	9	10	3	−0.33
Verbal reasoning	11	10	3	0.33
Auditory processing	117	100	15	1.13
Recognition vocabulary	116	100	15	1.06
Verbal comprehension	21	16.84	2.45	1.55
Naming	127	100	15	1.8
Word fluency/phonetic association	5	5.21	2.1	−0.09
Word fluency/semantic association	16	12.31	2.7	1.25
Memory for sentences	111	100	15	0.73
Verbal learning	99	100	15	−0.06
Verbal delayed recall	99	100	15	−0.06
Spatial memory	13	10	3	1
Visual learning	11	10	3	0.33
Visual delayed recall	11	10	3	0.33
Sustained attention/speed of processing	11	10	3	0.33
Divided attention	44	50	11	−0.54
Selective attention	90	100	15	−0.66
Executive functions/operative memory	11	10	3	0.33
Executive functions/motor sequencing	13	10	3	1
Executive functions/concept formation	125	100	15	1.66
Executive functions/planning	11	10	3	0.33
Executive functions/interference	45.11	2.71	9.05	−0.42
Executive functions/nonverbal fluency	29	33.1	11.46	−0.35
Reading/decoding	107	100	15	0.46
Reading/understanding	105	100	15	0.33
Writing/copy	−13	−2.75	3.21	−3.19
Writing/dictation	−28	−0.97	1.3	−20.79
Phonetic analysis	111	100	15	0.73
Arithmetic/calculation	129	100	15	1.66
Arithmetic/applied problems	97	100	15	−0.2

Score, mean and standard deviation (SD) are shown. *Z* scores are also calculated (by average definition 0 and standard deviation 1) for comparison between different tests. In the tasks in which errors are registered, direct scores and means are presented in negative. Since the 45 dimensions of the test battery are compared to a cut-off, a multiple-comparison correction was introduced in the dataset using the Bonferroni method to determine the cut-off points for the identification of problems

shape of letters regarding cursive and print writing were also recorded, which is characteristic of allographic dysgraphia. In spite of the difficulties in writing, the patient is capable of spelling and separating words within a sentence without difficulty and without making any of the mistakes detected when writing. When asked to write words without visual support, the errors were similar to those made when writing with vision.

Fig. 4. Patient's handwriting in a dictation task (above) with the correct text in Spanish and English, and samples of patient's written words (below). Duplication, omission and overlapping errors are shown (*arrows*). Correct written words in Spanish and English (*in brackets*) are shown below the patient's written words



"Cuando los españoles llegaron a las costas de California, los indios que allí encontraron vivían agrupados y cultivaban el lugar que habitaban.
Mientras unos se dedicaban a la agricultura, otros cuidaban el ganado y otros pescaban.
Durante miles de años habían vivido en las selvas y en las orillas del mar, refugiándose en las cavernas y procurando hacer más fácil su vida."

"When the Spaniards arrived at the coasts of California, the native indians they found there lived in groups and cultivated their own inhabited areas.
While some dedicated their time to agriculture, others took care of the cattle and others would fish.
For thousands of years they had lived in jungles and on sea shores, taking refuge in caves and trying to make their life easier".

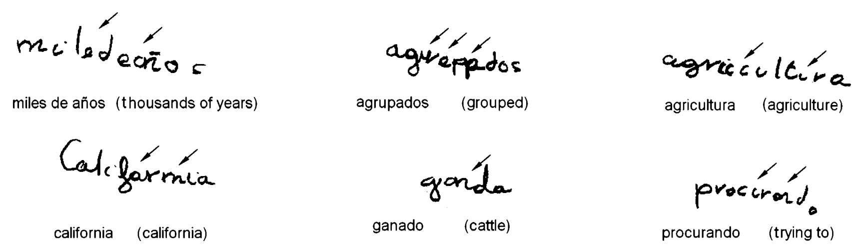


Table 2 Error classification from dictation writing, when comparing the patient's errors with those of the control group, for which mean and standard deviation are shown

Type of errors	Dictation (words=62)		
	Patient	Group control (N=15)	
		Mean	SD
Stroke omission	15	0.0667	0.2582
Stroke repetition	3	0.0667	0.2582
Letter omission	4	0.5333	0.8338
Letter repetition	1	0.0000	0.0000
Unnecessary gaps (>2 mm)	23	0.2000	0.7746
Overlapping letters	0	0.0000	0.0000
Overlapping words	5	0.0000	0.0000
Sloping lines (>10 mm)	+	+ in 6	
Left neglect	–	–	
Alterations in the shape of cursive/print letters	+	–	

Discussion

Spatial dysgraphia has always been associated with injuries located in the right parietal lobe and more recently to cerebellum damage, both diffused by atrophy [5] as well as left focal lesions due to vascular damage in adults [6]. Our case presents similar characteristics, with the exception that the cerebellar damage was produced during writing skills acquisition; however, cerebellar development entailed a few characteristic errors of spatial dysgraphia associated with cerebellum atrophy, which still persisted at the time of the study. Our case shows proportionally more mistakes than those described in adults, maybe because of our patient's age. It is also worth noticing that in Spanish, acquisition of orthography is facilitated by a direct correspondence from phonemes to graphemes, and therefore, this kind of errors are very uncommon. As is the case in the most recent study in the literature concerning adults, our patient's writing does not worsen when he is asked to write blindfold. This indicates that feedback from vision is not completely efficient and does not make up for proprioceptive deficit [6]. The cited studies do not mention whether copying tests were performed to assess writing in addition to dictation. It should be noted that mistakes in copying are comparatively less than those in dictation. This discrepancy is associated with a deficit in allographic representations which could affect tasks in which the shape of the letter is not provided, such as in copying [7]; alterations in the shape of cursive or print letters written by the patient can be equally interpreted. Mistakes made by our patient do not include substitutions of letters with similar shape, except when the letter “a” is replaced by “o”, which may be due to a missing stroke or also to a substitution of letters, these being characteristic of allographic dysgraphia. On the other hand, the patient shows strong deficit and mistakes in the execution of graphic motor patterns, which affect the formation and order of letters, word separation and the use of straight lines characteristic of spatial dysgraphia. The absence of mental imagery tests for children using allographs has made it impossible to better characterise peripheral dysgraphia observed in our patient.

Research among the paediatric population with cerebellum congenital pathologies often shows a severe cognitive incapacity, suggesting that the cerebellum is indispensable in obtaining an appropriate cognitive development [8]. In contrast, lesion studies in children and adolescents with focal cerebellar lesions are insufficient to prove the cerebellar involvement in cognition [9]. Language alterations and sequential functions have been described in children after cerebellitis [10] Children with infratentorial tumours show visuospatial, language sequencing and memory problems. Some studies show that right cerebellar damage is related with a plateau of linguistic skills, and left cerebellar damage can be connected to impaired visuospatial skills [11–13]. However, studies with larger series [14] that make a neuropsychological profile comparison of children after tumour resection has not found a location-related profile. In adults, one crossed cerebellum–cerebral diaschisis was informed. Recent studies in patients with cerebellar lesions [15, 16] and with functional neuroimaging using single photon emission computed tomography [17, 18] and functional MRI [19] show a lateralised functional organisation of the cerebellum, relating linguistic functions to right cerebellum hemisphere and visuospatial functions to left cerebellum hemisphere.

The role of the cerebellum in visuospatial and visuoconstructive processes through associated cerebellum–parietal connections has been extensively analysed in adults [20]. The cerebellum is involved in the representation of temporal information not only in motor control but also in perception, which optimises sensory quality to coordinate processes. Thus, visuospatial skills depend on cerebellar participation [21]. Moreover, apart from dysgraphia, our case also presents a mild visuoconstructive apraxia, an association equally registered in a previous case of an adult [6]. It is important to notice that difficulties in writing are more serious that the symptoms of visuoconstructive apraxia (see Table 2), so the writing problems cannot be explained by apraxia in our case. The patient's handwriting does not show apraxic agraphia characteristics (distorted, incomplete, hesitant and imprecise letter formation), and words are not strings of illegible scrawls, which have been also associated with right cerebellar damage [18].

The cerebellum compares motor orders for intentional movements with the afferent feedback of the current movement, which permits error detection and adjustment of required movements. The graphic motor patterns, which constitute writing, require constant monitoring through sensorial, visual and proprioceptive feedback processes, which analyse if the strokes and letters have been correctly produced [22]. Cerebellum injuries such as the ones presented by our patient could produce discoordination between the planning of graphic patterns generated by supratentorial structures and the proprioceptive afferents during the execution of ongoing writing movements [6, 7]. The functional network responsible for the peripheral control of writing may include the cerebellum and supratentorial structures, in addition to the classic observations of peripheral dysgraphia, not only in adults but also in children.

Current investigation points out the idea that the cerebellum has a relevant role in the diverse execution stages of cognitive processes [23, 24], both once they have been acquired, and also in their development, and could cause specific deficiency similar to those caused in adults after acquired cerebellum damage [25]. This case is an illustration, and as Mariën [26] says, functional topographic organisation has not been cleared up yet, although there is some evidence of a certain lateralised functional organisation in the cognitive modulation of the cerebellum.

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Conflict of interest None.

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