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Acquired Alexia With Agraphia Syndrome in Childhood

ABSTRACT

The acquired alexia with agraphia syndrome is a conspicuous disorder of reading and writing in the absence of significant other language impairments that has mainly been recorded in adults. Pure cases are rare, with most patients displaying mild aphasic deficits. In children, acquired reading and writing disorders are generally reported as part of more encompassing aphasic syndromes affecting oral and written language equally, for example, Broca or Wernicke aphasia. Documented instances of predominant acquired reading and writing disorders in childhood are exceptional. We report an 11-year-old, right-handed boy who sustained a left temporoparieto-occipital hematoma following rupture of an arteriovenous malformation and who consecutively presented with the acquired alexia with agraphia syndrome associated with word-finding difficulties. Neuropsychologic and neurolinguistic data showed that there was no concomitant Gerstmann and/or angular gyrus syndrome. The recovery from the anomia was quite favorable, but recovery of written language was

more protracted and acted on the patient's further scholastic achievement. This case is reminiscent of a historical childhood case reported in 1939 and is consonant with adult cases in terms of lesion location and semiologic picture. (*J Child Neurol* 2006;21:324-330; DOI 10.2310/7010.2006.00087).

The syndrome of alexia with agraphia, first described by Dejerine in 1891,¹ is an acquired disorder of reading and writing in the absence of significant other language impairments. Pure cases of alexia with agraphia are rare because most patients display mild aphasic deficits, such as word-finding difficulties and paraphasic verbal output. In right-handers, the lesions associated with this type of acquired illiteracy are typically located at the level of the angular gyrus of the left hemisphere.² Some patients with this reading and writing disorder are believed to have a mild form of Wernicke aphasia with less severely impaired auditory than reading comprehension; they have lesions in the vicinity of the inferior parietal lobe, near the supramarginal and angular gyri.² Gerstmann syndrome and angular gyrus syndrome can be observed but are not systematically noted or necessary for the diagnosis of alexia with agraphia.^{2,3}

Alexia with agraphia has mostly been recorded in adults. In children, acquired reading and writing disorders are generally reported as part of more encompassing aphasic syndromes affecting both oral and written language.⁴⁻⁸ Yet rare instances of preponderant reading and writing disorders consecutive to brain damage have been described in children. In 1939, De Girardier and Jeannin described a 7-year-old girl who presented with fluent aphasia following a left posterior temporal skull fracture.⁹ The aphasia was characterized by severe word-finding difficulties, alexia, and agraphia. The girl was not able to name objects. Letters could not be read or written. However, repetition and auditory comprehension were not impaired. Assal and Campiche reported in a series of 18 children with acquired aphasia one exceptional case of lasting alexia with agraphia associated with word-finding difficulties, few phonemic paraphasias, and mild auditory comprehension disturbances.¹⁰ Unfortunately, the authors did not provide an in-depth analysis of this remarkable observation. Lelord and colleagues reported a 9 1/2-year-old child who presented with acquired Gerstmann syndrome and additional alexia, which both recovered within 6 months postonset of measles encephalitis.¹¹

As a result, childhood cases of acquired alexia with agraphia syndrome with or without associated word-finding difficulties have only occasionally been described. We had the opportunity to study an 11-year-old boy who presented with this peculiar clinical picture.

Case Report

Clinical History

The patient was an 11-year-old, right-handed, French-speaking boy of average intelligence without significant medical antecedents. At the time of the event, he attended primary school and was about to enter the sixth grade. He never failed examinations; however, his writing was characterized by spelling errors. Psychomotor development was normal.

While on holiday abroad, the patient felt unwell and collapsed during a bike ride. On admission, his Glasgow Coma Scale score was 10/15. His speech was incomprehensible, and his best motor response was flexion to pain. A computed tomographic (CT) scan of the brain was performed immediately. While in the scanner, he developed a brief episode of respiratory depression. Following this, his pupils were equal (size 4) and reacted to light. He was verbalizing inappropriately, and there was no eye opening. Vital signs at this stage were stable, and he was afebrile. The CT scan revealed a large left temporoparieto-occipital intracerebral hematoma with invasion of the ventricular system, edema, and midline shift toward the right. Injection of contrast showed serpiginous shadows suggesting the presence of an arteriovenous malformation as the cause of the hemorrhage. There was no definite evidence of hydrocephalus on either scan. On day 2 after admission, he was intubated and hyperventilated.

Shortly after discontinuing sedation, his vital signs remained stable. He recognized his parents, nodded in response to questions, and moved all four limbs. His pupils were equal and reacted to light, and he opened his eyes spontaneously. Verbal responses were difficult to assess at this stage because he was still intubated.

The patient was extubated for the journey back to Brussels, which took place 3 days after admission. On arrival at the intensive care unit of Erasmus Hospital, he was conscious (Glasgow Coma Scale score 13/15). The neurologic examination further showed a rigidity of the neck, a right homonymous hemianopia, and aphasia. Cerebral angiography performed the next day demonstrated a deep temporoparietal arteriovenous malformation that appeared to be fed by the left posterior cerebral artery and the left anterior choroid artery. Clinical evolution was favorable, and the meningeal signs remained discrete. A repeat CT scan was performed 2 days after arrival in Brussels showing stabilization of the hemorrhagic lesion. The patient was transferred to the Department of Child Neurology.

Eight days after admission to Erasmus Hospital, the patient was readmitted to the intensive care unit because of a sudden alteration in consciousness, with respiratory pauses suggesting an epileptic attack without observable tonic-clonic movements or urinary loss. He rapidly recovered consciousness, and his Glasgow Coma Scale score was 15/15. He was treated with antiepileptic drugs. An electroencephalogram performed in the interictal state showed a slow generalized dysrhythmia without epileptiform activity. A repeat CT scan showed no increase in lesion size. However, there was an increased midline shift toward the right, and a beginning hydrocephalus was detected. There was a tendency toward bradycardia, probably associated with intracranial hypertension, as confirmed by the eye fundus that demonstrated bilateral papilledema.

The patient's condition improved steadily in the following days, and 4 days after admission to the intensive care unit, he returned to the child neurology ward. At that time, a neurologic examination showed a fully conscious and very cooperative child whose speech was fluent but characterized by marked word-finding difficulties. Auditory comprehension appeared to be normal. Meningeal signs were present, preventing gait from being assessed. Sensitivity and strength were normal in the upper and lower limbs, and dysmetria was absent. Myotatic reflexes were weak and symmetric. Cutaneous plantar reflexes were in flexion bilaterally. Visual field examination showed a right homonymous hemianopia. Visual acuity was 10/10. The patient presented with diplopia caused by a paresis of the VIth nerve on the right. Anisocoria was present, with the right pupil being larger than the left one. The eye fundus still demonstrated bilateral papilledema.

Magnetic resonance imaging of the brain performed 4 weeks after onset of neurologic symptoms revealed a large area of abnormal signal intensity in the left temporoparieto-occipital region on T₁-weighted images, characterized by an area of isointense signal in the middle and increased signal intensity at the periphery of the lesion (Figure 1). Proton density-weighted images confirmed the lesion as an extensive area of increased signal intensity in the left temporoparieto-occipital region surrounded by an ill-defined perilesional area of increased signal intensity corresponding to an edematous reaction (Figure 2). There was an important mass effect as demonstrated by a midline shift toward the right and a compression of the temporal and occipital horns of the left lateral ventricle.

At discharge from hospital, 8 weeks after onset of neurologic symptoms, meningeal signs were still present. No motor defects were noted. Gait was normal, except for some less fluent movements owing to the meningism. Myotatic reflexes were weak and symmetric. Abdominal reflexes were present. Sensation was normal. The right homonymous hemianopia had evolved toward an upper homonymous quadrantanopia. Diplopia associated with a paresis of the VIth cranial nerve on the right was present, as well as a decreasing papilledema. The patient spoke fluently but still experienced word-finding difficulties. Reading and writing were impaired, as was calculation. He also displayed memory disturbances. Orientation in time and space was preserved. Buccolabiolingual, ideomotor, ideational, and constructional praxes were within normal limits. Finger agnosia and left-right spatial disorientation were absent, as was visual agnosia. No abnormality of dermoxenia or two-point discrimination was observed on the hands or fingers. Stereognosis was normal. Tactile or sensory extinction was absent. The patient was still

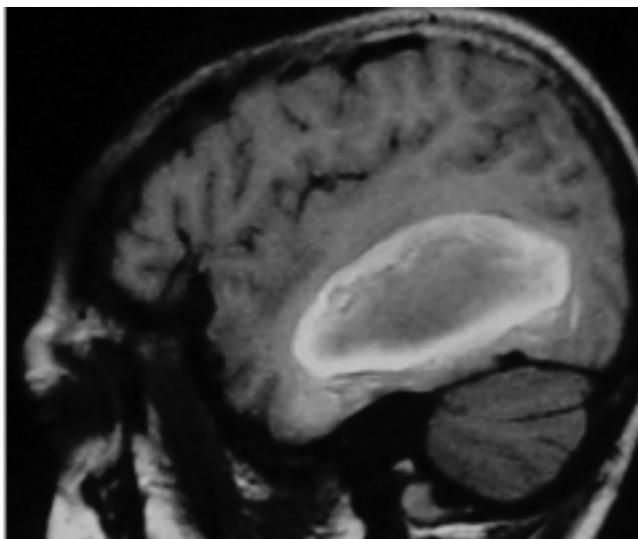


Figure 1. Sagittal T₁-weighted magnetic resonance image at 4 weeks postonset showing a large area of abnormal signal intensity in the left temporoparieto-occipital region of the left hemisphere.

on antiepileptic medication. In the following months, the patient visited the outpatient rehabilitation unit, where he received language therapy associated with individual education, physiotherapy, and occupational therapy.

Two months after discharge from hospital, the patient was readmitted for embolization of the arteriovenous malformation. Neurologic examination before and after the procedure was normal, except for persisting strabismus, right upper homonymous quadruparesis, and language problems. The child was cooperative and well oriented in time and space. One and a half months after the embolization, language therapy was recommended, with emphasis put on written language skills. Six months postonset, the patient followed reading and writing sessions twice a week for 1 hour. During these sessions, reading aloud, reading comprehension, orthography, and grammar were practiced.

Neuropsychologic Examination

The patient was first seen 4 weeks postonset. At that time, he was cooperative and oriented. Although he was slow, fatigable, and somewhat anxious about his performance, he nevertheless did his best throughout the evaluation sessions. Owing to time constraints, the aphasia, and the increased fatigability, however, the examination was only partly conducted and also made use of short clinical tests. On the Wechsler Intelligence Scale for Children-Revised (WISC-R),¹² he obtained an estimated Full-Scale IQ of 99 (estimated Verbal IQ 99; estimated Performance IQ 99). The subtests Vocabulary and Comprehension were not administered for fear that they would too severely depress the patient's score as a result of his marked naming and word-finding difficulties. Reasoning and abstract problem solving were preserved (respective scaled scores on the WISC-R Similarities, Picture Completion, and Picture Arrangement subtests: 10, 11, 11). Constructional functions were within normal limits (respective scaled scores on the WISC-R Block Design and Object Assembly subtests: 9, 9), as was also demonstrated by the copy of a complex figure (Figure 3). Short-term auditory storage capacity was normal, as revealed by the Digit Span subtest of the WISC-R (scaled score 11). The patient succeeded in performing mental arithmetic computations that did not exceed 10; when adding up and subtracting above 10, he used correct yet time-consuming written strategies. Multiplication tables were forgotten; multiplying was performed by adding up. Nevertheless, he obtained a scaled score of 10 on the Arithmetic subtest of the WISC-R. The patient presented with mild difficulty in reading numbers and made semantic and syntactic errors when writing dictated numbers (eg, "22142" for 20,142, "15018" for 1518). Clinical testing of left-right spatial orientation showed no abnormalities.

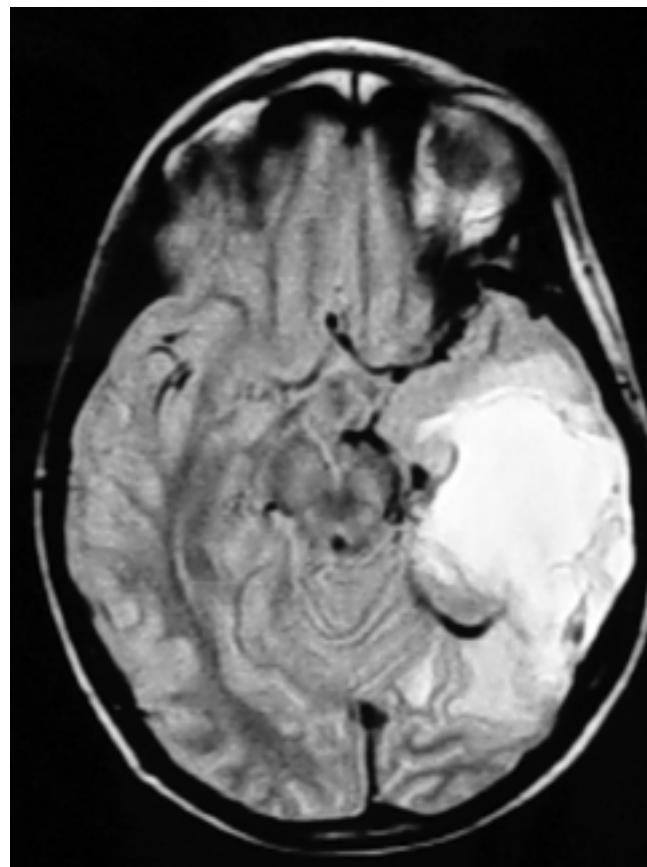


Figure 2. Axial proton density-weighted magnetic resonance image at 4 weeks postonset showing an extensive area of increased signal intensity in the left temporal lobe surrounded by an ill-defined perilesional hyperintense signal area.

Fifteen months after the initial neuropsychologic examination, the patient was partially reassessed. He had resumed school activities and had entered the first grade of secondary school. Reading was slow and hesitant, but reading comprehension was normal. Writing showed persistent spelling errors corresponding to a primary school fifth grade level. Neuropsychologic assessment demonstrated long-term auditory memory difficulties, along with poor verbal learning abilities, which both most probably acted on his potential to master the orthographic rules and to learn foreign languages (Dutch and Latin).

Neurolinguistic Examination

Neurolinguistic assessments were carried out in agreement with a three-epoch time frame model of vascular aphasia,¹³ that is, in the peracute phase (5 weeks postonset), the lesion phase (4 months postonset), and the late phase (10 months postonset). Oral language was investigated by means of the shortened version of the Token Test,¹⁴ Pierre Marie's "Three Paper Test,"¹⁵ the Bachy picture naming test,¹⁶ and subtests of the French version¹⁷ of the Boston Diagnostic Aphasia Examination.¹⁸ Written language was investigated by means of the Boston Diagnostic Aphasia Examination, as well as clinical and experimental tasks making use of regular and irregular words, homophones and pseudohomophones, and pronounceable nonwords. Table 1 shows the patient's scores on the expressive and receptive language tests at the initial and follow-up assessments.

Five Weeks Postonset

Syntactic auditory comprehension disturbances were not observed. However, the patient experienced mild lexical difficulties, as shown by a picture-pointing and a body-part identification task (Boston Diagnostic Aphasia



Figure 3. Copying of a complex figure at 4 weeks postonset (patient's copy is on the right).

Examination) (eg, he could not point at his wrist and forehead and mistook his hip for his thigh, his eyebrow for his eyelashes, and his middle finger for his index finger). Spontaneous language was fluent and characterized by severe word-finding difficulties, multiple pauses, and semantic paraphasias. Phonemic paraphasias, neologisms, and syntactic errors were absent. Picture naming tasks, a responsive naming task, and body-part naming were all impaired; however, phonemic cueing helped in almost all instances. Errors consisted of adequate circumlocutions, nonresponses, and semantic paraphasias (see Table 2 for examples). Definitions of orally presented verbs and nouns were adequate, except for word-finding difficulties.

Semantic evocation (animal naming) was poor. Oral repetition of words and sentences was normal, as were automatized sequences.

The patient's reading ability was severely impaired. He had problems reading letters: only 12 of 26 letters were immediately and correctly read, and 4 letters (g, l, r, x) were either guessed or spontaneously corrected. Discrimination of upper- and lowercase letters (Boston Diagnostic Aphasia Examination Symbol and Word Discrimination subtest) was normal. Oral reading of words and sentences was slow and painstaking, with the patient using a letter-by-letter decoding strategy yet failing to read or spell the words because of letter misidentifications. He performed at chance level when matching written words to real objects. Performance was equally poor on an experimental task made up of five regular and high-frequency words, five regular and low-frequency words, five irregular and high-frequency words, five irregular and low-frequency words, five pronounceable nonwords, and five pseudohomophones. Again, letter misidentifications hindered the patient's letter-by-letter decoding strategy. Only 7 of 30 words were correctly read (ie, 2 regular and high-frequency words, 2 regular and low-frequency words, 2 irregular and high-frequency words, 1 irregular and low-frequency word); nonwords and pseudohomophones could not be decoded. However, identification of real words spelled aloud by the examiner was not altered, but responses were characterized by latency (Boston Diagnostic Aphasia Examination Comprehension of Oral Spelling subtest). Similarly, the patient was able to select from a multiple choice of five written words the one spoken by the examiner (Boston Diagnostic Aphasia Examination Word Recognition subtest). Reading comprehension of sentences and texts was defective according to the Boston Diagnostic Aphasia Examination.

Table 1. Neurolinguistic Test Data for Initial and Follow-Up Assessments*

Language Modality	Test	Time Postonset			Cutoff Score
		5 wk	4 mo	10 mo	
Auditory comprehension	Word Discrimination, BDAE	61.5/72 (85.4)	70/72 (97.2)		67/72
	Body-Part Identification, BDAE	15/20 (75.0)	15/20 (75.0)		18/20
	Complex Ideational Material, BDAE	12/12 (100.0)			8/12
	Three Paper Test	3/3 (100.0)			
	Token Test	31/36 (86.1)	34.5/36 (95.8)		29/36
	Verbal Agility, BDAE	13/14 (92.9)			11/14
	Automatized Sequences, BDAE	9/9 (100.0)			7/9
	Word Repetition, BDAE	9.5/10 (95.0)			9/10
	Sentence Repetition, BDAE	15/16 (93.8)			12/16
	Visual Confrontation Naming, BDAE	59/105 (56.2)	93/105 (88.6)		105/105
Oral expression	Bachy picture naming test	8/36 (22.2)	19/36 (52.8)	33/36 (91.7)	33/36
	Body-Part Naming, BDAE	16/30 (53.3)	27/30 (90.0)		24/30
	Responsive Naming, BDAE	23/30 (76.7)	28/30 (93.3)		27/30
	Noun and Verb Definitions	22/23 (95.7)			
	Animal Naming, BDAE	8	12		12
	Letters	16/26 (61.6)	25/26 (96.2)		
	Symbol and Word Discrimination, BDAE	10/10 (100.0)	10/10 (100.0)	10/10 (100.0)	10/10
	Oral Word Reading, BDAE	12/30 (40.0)	20/30 (66.7)	30/30 (100.0)	30/30
	30 Words: Oral Reading (Experimental Task)	7/30 (23.3)	30/30 (100.0)		
	Oral Sentence Reading, BDAE	0/10 (0.0)	10/10 (100.0)	10/10 (100.0)	8/10
Reading	Word-Object Matching	3/6 (50.0)			
	Word-Picture Matching, BDAE		10/10 (100.0)	10/10 (100.0)	10/10
	Comprehension of Oral Spelling, BDAE	8/8 (100.0)	7/8 (87.5)	8/8 (100.0)	6/8
	Word Recognition, BDAE	7/8 (87.5)	8/8 (100.0)	8/8 (100.0)	6/8
	Sentence and Text Comprehension, BDAE	0/10 (0.0)	5/10 (50.0)	9/10 (90.0)	7/10
	Mechanics of Writing, BDAE	4/5	5/5	5/5	5/5
	Serial Writing, BDAE	44/47 (93.6)	47/47 (100.0)	47/47 (100.0)	47/47
	Letter Dictation (uppercase)	22/26 (84.6)			
	Letter Dictation (lowercase)	20/26 (76.9)			
	30 Words: Dictation (Experimental Task)	16/30 (53.3)	17/30 (56.7)	20/30 (66.7)	
Writing	Primer-Level Dictation, BDAE		15/15 (100.0)	15/15 (100.0)	15/15
	Spelling to Dictation, BDAE		4/10 (40.0)	5/10 (50.0)	6/10
	Sentences to Dictation, BDAE	4/12 (33.3)	6/12 (50.0)	8/12 (66.7)	10/12
	Written Confrontation Naming, BDAE	5/10 (50.0)	7/10 (70.0)	10/10 (100.0)	7/10
	Narrative Writing BDAE	3/5 (60.0)	4/5 (80.0)	4/5 (80.0)	2/5
	30 Words: Copying (Experimental Task)	26/30 (86.7)	27/30 (90.0)		

BDAE = Boston Diagnostic Aphasia Examination.

Percent correct responses in parentheses. Blanks indicate data not available.

Table 2. Examples of Errors on Picture Naming Tests^{16,17} at 5 Weeks and 4 Months Postonset

Time Postonset	Target Word	Response	Error Type
5 wk (periacute phase)	Seau (bucket)	On met de l'eau dedans (One puts water in it)	Circumlocution
	Paon (peacock)	Ils savent bouger leur queue et la mettre en rond derrière (They can move their tail and put it in a circle behind)	Circumlocution
	Cactus (cactus)	Ça pique... (It's prickly...)	Circumlocution
	Trèfle (clover)	C'est dans un jeu de cartes (It's in a card game)	Circumlocution
	Domino (domino)	Une carte (a playing card)	Semantic paraphasia
	Moto (motorbike)	Camion...non, c'est pas une voiture (Truck... no, it's not a car)	Semantic paraphasia
	Fenêtre (window)	Porte (door)	Semantic paraphasia
	Echelle (ladder)	Escalier (stairs)	Semantic paraphasia
4 mo (lesion phase)	Bouilloire (kettle)	On met de l'eau dedans et on met ça sur le feu (One puts water in it and one puts it on the fire)	Circumlocution
	Loupe (magnifying glass)	C'est pour voir plus gros (It's to see bigger)	Circumlocution
	Evier (sink)	Pour faire la vaisselle...un lavabo (to do the dishes... a washbasin)	Circumlocution then semantic paraphasia
	Aquarium (aquarium)	Un bocal (a jar)	Semantic paraphasia
	Domino (domino)	Un dé (a die)	Semantic paraphasia

Writing was also impaired, either at elementary (alphabet: omissions and self-corrections) or more elaborated levels. Copying, writing to dictation, narrative writing, and written naming of visually presented items (Boston Diagnostic Aphasia Examination) were characterized by spelling errors and literal paraphasias (see Figure 4 for an example). The 30 words of the experimental reading task were also dictated. Only 16 of 30 words were correctly written (ie, 4 regular and high-frequency words, 4 regular and low-frequency words, 1 irregular and high-frequency word, 2 irregular and low-frequency words, 5 pronounceable nonwords).

Four Months Postonset

Auditory comprehension for lexical items had improved except for body-part identification. Spontaneous language still embodied word-finding difficulties, although semantic paraphasias were less prominent. Responsive naming and body-part naming tasks had normalized. Picture naming tasks had improved but were still below normal because of prolonged response latencies, circumlocutions, and semantic errors (see Table 2). Semantic evocation equaled the cutoff score.

Reading and writing skills had also evolved favorably. Reading letters showed one error ("r" for "p"). Discriminating upper- and lowercase letters and matching written words to pictures were normal, although deciphering the words was time-consuming. Oral reading of words and sentences occurred slowly and analytically using a letter-by-letter or syllable-by-syllable strategy, which often resulted in self-corrections. Reading comprehension of sentences and texts had improved but remained inferior to normal. Word recognition and comprehension of orally spelled words were normal. The production of automatized written sequences, copying, and elementary dictation (Boston Diagnostic Aphasia Examination) were normal. Writing to dictation at a more elaborated level, however, was still defective, as demonstrated by the Boston Diagnostic Aphasia Examination and the experimental 30-word tasks. Most errors consisted of phonetic script and regularization of irregular words. Written naming of visually presented items (Boston Diagnostic Aphasia Examination) still revealed literal errors, along with a tendency to regularize irregular words (eg, "disse oit" for *dix-huit* [eighteen]). Narrative writing (Boston Diagnostic Aphasia Examination) comprised a coherent account with literal paraphasias and spelling errors (Figure 5).

Ten Months Postonset

Language therapy sessions twice a week that had started 6 months postonset revealed a steady progress in lexical and reading abilities and a more mitigated improvement in writing. Spontaneous language was fluent and contained word-finding difficulties only on rare occasions. The Bachy picture

naming task showed a word-finding problem just for three infrequent items: *perroquet* (parrot), *corbillard* (hearse), and *balcon* (balcony).

Reading had improved to such an extent that all words and sentences could adequately be decoded, notwithstanding many pauses and a marked slowness. Comprehension of sentences and texts had normalized. Reading numbers was errorless. However, writing remained effortful, with persistent difficulty in converting phonemes to graphemes. There was an obvious tendency to regularize irregularly spelled words that was most conspicuous during dictational and narrative writing.

Discussion

The patient's semiologic picture in the periacute and lesion phases was characterized by a frank reading and writing disorder associated with anomia in the absence of significant auditory comprehension disorders, paragrammatic verbal output, or nonfluent language features. This clinical description is congruous with that of acquired alexia with agraphia syndrome.^{2,3} This syndrome is only rarely found in its pure form, with most patients exhibiting some degree of anomia.³ That is why Lecours and Lhermitte consider it to be a third type of Wernicke aphasia.¹⁹ These authors refer to classic Wernicke aphasia as Wernicke aphasia type I, whereas Wernicke aphasia type II represents transcortical sensory aphasia. Wernicke aphasia type III, then, encompasses two linguistic dissociations: (1) a defect in oral language mainly restricted to word-finding difficulties as opposed to a more general alteration of written language (agraphia) and (2) near-normal auditory comprehension abilities contrasting with an inability to comprehend written language (alexia).¹⁹ Given the presence of an anomia associated with an alexia and agraphia, the term Wernicke aphasia type III might alternatively be applied to denote our patient's language disturbance.

In the periacute phase, naming was mainly characterized by adequate circumlocutions, nonresponses, and semantic paraphasias. Phonemic paraphasias and neologisms were not observed. Pointing at body parts and depicted objects named by the examiner was only mildly disturbed. The patient benefited from phonemic cueing when experiencing word-finding difficulties. In addition, he was able to extract the meaning of verbs and nouns spoken by the examiner as demonstrated by his adequate performance on the definition task. These word-finding characteristics suggest that the problem was related to impaired access to the semantic lexicon rather than to a loss of semantic knowledge.²⁰

The rate of recovery of the patient's word-finding difficulties was quite favorable because naming had normalized in the late phase. This positive course is in accordance with the optimistic views on the recovery from acquired childhood aphasia as propagated by the standard doctrine on its clinical picture.²¹ However, such a favorable outcome in naming ability is by no means the general rule, with some anomic children displaying per-



Figure 4. Copying of single words at 5 weeks postonset with requested letter case transposition (ordinateur [computer], éléphant [elephant], envoyer [to send]).

sistent word-finding difficulties in the long term even when the age and etiology are similar to those of our patient.²² As a matter of fact, current knowledge about the outcome of acquired aphasia in children points to the presence of persisting linguistic deficits impeding further academic achievement even of those children considered to have clinically recovered from aphasia.^{4,23,24} As naming, reading, and writing progressively improved in the course of our patient's recovery, it became increasingly evident that his pre-morbid spelling difficulties gradually came to the fore again. Whereas scores on reading tasks had normalized in the late phase—although reading remained globally slow—dictational and narrative writing still showed dysorthographia and frequent self-corrections. However, a comparison with our patient's classroom notebooks revealed that in the late phase, his writing had not yet improved to premorbid levels, thus indicating that recovery of written language was more protracted than that of oral language.

Few children with acquired alexia with agraphia have been documented. One of the first detailed accounts that unfortunately remained unnoticed in the Anglo-Saxon literature was presented by De Girardier and Jeannin.²⁵ Following a brief annotation published in 1938,²⁶ these authors described in 1939 a 7-year-old girl who, following a fall from a step, sustained a left temporal depressed skull fracture without loss of consciousness.⁹ Three days later, she developed aphasia as the sole neurologic symptom. It was characterized by manifest word-finding difficulties, alexia, and agraphia in the absence of auditory comprehension or repetition disorders. The girl had not been mute for one moment. Although she had always been able to speak fluently, she never exhibited paraphasic jargon. There was no word deafness. The girl was operated on 10 days after the fall. Bone splinters and subdural blood clots located up to the level of the second temporal gyrus were removed. Because of the late onset of the

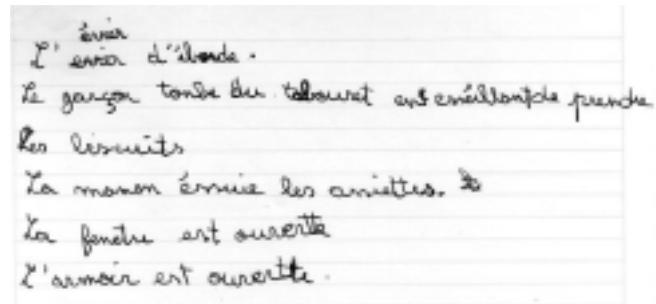


Figure 5. Narrative writing at 4 months postonset (Cookie Theft picture from the Boston Diagnostic Aphasia Examination). Correct spelling with English translation is as follows: L'évier déborde (The sink is overflowing); Le garçon tombe du tabouret en essayant de prendre les biscuits (The boy is falling from the stool while trying to take the cookies); La maman éssie les assiettes (The mother is washing the plates); La fenêtre est ouverte (The window is open); L'armoire est ouverte (The cupboard is open).

language disorder and the rapid postoperative remission of all aphasic symptoms, the authors concluded that the aphasia had resulted from the progressive cerebral compression caused by the subdural hematoma. They stated that the type of aphasia did not fit the Broca-like symptomatology but anomic aphasia ("aphasie amnésique de Pitres") with alexia and agraphia. Consequently, some 40 years prior to Woods and Teuber's seminal article on acquired childhood aphasia,²⁷ a description was published of a child who definitively did not fit within the standard doctrine²¹ because she presented with "a dissociated form of amnestic aphasia associated with alexia and agraphia that is related to Wernicke's aphasia."⁹ Thus, the semilogic resemblance to our case description is all the more striking.

In contrast to De Girardier and Jeannin's patient, the girl with anomic aphasia documented by Hynd and colleagues did not exhibit alexia and agraphia.²² She initially evidenced fluent and circumlocutory spontaneous language and obvious word-finding difficulties. In accordance with adult reports, there was marked impairment in naming characterized by frequent phonemic and semantic paraphasias. These authors concluded that their case was consistent with adult anomic cases in terms of lesion location and rate of recovery, as well as in relation to the severe defects in confrontational naming. The same year, a 13-year-old boy was described with anomic aphasia caused by simple partial status epilepticus as a consequence of the cranial irradiation and intrathecal methotrexate he had received as treatment for acute lymphoblastic leukemia at the age of 9 years.²⁸ Unfortunately, the authors did not provide details about the boy's language characteristics. Finally, anomic aphasia was also reported in a 5-year-old boy who, following herpes simplex encephalitis, presented with fluent and well-articulated but circumlocutory and empty language, semantic paraphasias, and word-finding latencies.²⁹ According to the authors, the child showed great similarity with adult cases in terms of naming abilities and lesion location.

The association of the acquired alexia with agraphia syndrome with anomia has frequently been documented in adults.^{2,3} Naming difficulties in adult patients with anomic aphasia have often been linked to lesions affecting the inferior parietal lobe (supramarginal and angular gyri) or the inferior temporal lobe (middle and inferior temporal gyri).²⁰ As these posterior brain regions are also crucial for reading and writing,^{30,31} it is by no means surprising that patients presenting with acquired alexia with agraphia syndrome—also termed parietal-temporal alexia³—show some degree of anomia. The case of alexia with agraphia we describe is consonant with adult cases in terms of lesion location in the left parietotemporal area and in relation to the severe reading and writing disorder in combination with a marked naming deficit. As we could not evidence finger agnosia, left-right disorientation, or constructional apraxia in our patient, this case description further demonstrates that, in concordance with the literature,^{2,3} the presence of concomitant Gerstmann and/or angular gyrus syndrome is not compulsory for the diagnosis of acquired alexia with agraphia.

Dejerine originally reported a patient with the syndrome of alexia with agraphia in 1891¹ and 1 year later another patient suffering from the syndrome of alexia without agraphia.³² Since Woods and Teuber's landmark publication,²⁷ it has come to the fore that acquired aphasia in children is not uniformly non-fluent irrespective of lesion location, as claimed by traditional neurologic tenets,³ but that all aphasic syndromes recorded in adults can also be encountered in children. Thus, after having reported a child presenting with an acquired alexia without agraphia syndrome some years ago,³³ we now have the opportunity to document a rare childhood case of acquired alexia with agraphia syndrome.

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