

## Jules Dejerine and His Interpretation of Pure Alexia

DANIEL N. BUB,\*† MARTIN ARGUIN,\* AND ANDRÉ ROCH LECOURE†

*\*Montreal Neurological Institute and †Centre Hospitalier Cote-des-Neiges, Montreal, Quebec, H3W 1W5 Canada*

Dejerine's interpretation of pure alexia is routinely mentioned in all neuropsychological textbooks, yet the details of his account and the evidence on which it is based have never been subjected to a critical analysis. We provide such an evaluation in this paper, summarizing the behavioral data that Dejerine presented in his now famous case report and the theoretical framework he adopted to explain the phenomenon of alexia without agraphia. We also provide a link between Dejerine's work and current hypotheses on the nature of the syndrome. © 1993 Academic Press, Inc.

### INTRODUCTION

One hundred years ago, on the 27th of February 1892, Dejerine presented his analysis of a case of pure alexia to the Biological Society in Paris. The patient had been referred to him by a colleague—the noted ophthalmologist E. Landolt—who earlier had published some initial observations on the case in a volume dedicated to F. C. Donders on the occasion of his 50th birthday.

The interpretation of pure alexia originally proposed by Dejerine—combining as it did both a masterful neuroanatomical evaluation of the lesion responsible for the disorder (carried out during a brain autopsy of the patient) and neuropsychological evidence gathered from regular meetings with the patient over a 4-year period—was to prove highly influential. It is this account of the syndrome (or at least a version of this account popularized by Geschwind, 1965) that we see most commonly

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presented in all introductory psychology and neurology textbooks on the subject (e.g., Gletiman, 1986).

Surprisingly, given the longevity and importance of Dejerine's initial contribution and the recent revival of interest in pure alexia among cognitive neuropsychologists, there have been few attempts to uncover the details of his analysis and to relate them to the current work on the syndrome that has emerged over the last decade. For us, this obscurity has often produced a vague sense of unease when faced with the claim, now ubiquitous, that the modern taxonomic category known as pure alexia or letter-by-letter reading can be taken as equivalent to the syndrome Dejerine termed pure word blindness (*cécité verbale pure*). Are these entities, one defined by classical diagram-makers and the other by neuropsychologists relying on modern information processing architectures of the reading mechanism, really the same?

In this paper we wish to set the record straight—what was the neuropsychological evidence that Dejerine gathered on pure alexia and how sophisticated were the methods he used for behavioral testing? What is the logic that underlies his interpretation of the syndrome? Finally, is there anything that we can learn from a closer scrutiny of his work that might be used advantageously by those modern researchers who once again are attempting to unravel the complexities of pure word blindness? We begin this review (Section A) with a brief history of Dejerine, recounting his origins and professional development before turning in Section B to his classical analysis of pure alexia.

#### SECTION A: BIOGRAPHICAL SKETCH

According to Haymaker and Schiller (1970), Joseph Jules Dejerine should be considered among the founders of neurology. We can still find in medical dictionaries (for instance in Dorland<sup>1</sup>), entries which refer to "Dejerine's sign" (an increase in radiculitis induced by effort), to the "Dejerine-Lichtheim" phenomenon (the fact that an aphasic with suppression of speech resulting from a subcortical lesion can indicate the number of syllables in a word he is thinking of), to Dejerine's disease (a type of neuritis occurring in infancy), and to the Dejerine-Sottas malady (a type of progressive neuropathy), as well as no less than five "syndromes of Dejerine" (including one in which parietal lesions result in astereognosis and difficulties in recognizing differences in the intensity of somesthetic stimulation), these without including the "Dejerine-Roussy syndrome" (thalamic syndrome) and the "Landouzy-Dejerine dystrophy" (atrophy of facial and shoulder-arm muscles).

Jules Dejerine was born on August 3, 1849, in Geneva, where his father

<sup>1</sup> Dorland's Illustrated Medical Dictionary (24th ed.), Saunders, Philadelphia, 1965.

worked as a humble porter. His parents were both French from birth, although Dejerine always held a deep affection for Switzerland and made friends there that he would keep for the rest of his life.

As a schoolboy and then college student in Geneva, the young Dejerine was said to be "dissipated," "undisciplined," and "quarrelsome," preferring sports to studies that a prodigious memory made all too easy (Gauckler, 1922). This changed very quickly after he had obtained two degrees (1868 and 1870) and opted for a career in medicine rather than face conscription into the French navy. He became the student of Alfred Vulpian, who, in collaboration with Jean-Martin Charcot, began a neurology department at the Salpêtrière in 1862.

Armed with a letter of recommendation addressed to Vulpian by one of his former residents, Prévost, a Genoese doctor, Jules Dejerine left for Paris in the Spring of 1871. He was 22 years old. Vulpian received him quite warmly. Perhaps he already saw in Dejerine the ability that ultimately allowed the young man to emerge as the greatest of his students.

Dejerine passed the entrance examination for nonresident trainees in 1872 and the examination for residents in 1874.

In October 1877, a young American native of San Francisco arrived in Paris, coming from Lausanne where she has just obtained her undergraduate degree. Her name was Augusta Klumpke and she was 18 years old. Her mother had enrolled her in the Faculty of Medicine in Paris during the preceding month. She met with the Dean of the Faculty, Alfred Vulpian, who advised her to abandon any ideas of becoming a doctor. Augusta was not so easily dissuaded and Vulpian would ultimately be forced to admit that her ambition had been justified.

Meanwhile, Dejerine worked hard; he often visited "La Petite Vache—Boîte à Baptiste," located in 66 Mazarine Street, where he would debate with the Franco-Swiss intellectuals of Paris. In 1879 he received the title of Doctor of Medicine in the Faculty of Medicine in Paris. He was 30 years old. Shortly thereafter, he was appointed Head of the Clinic at the Hôpital de la Charité, in the unit of Alfred Hardy. During the next year, he met the new intern, Miss Klumpke. In letters that he regularly wrote to his mother, he began to refer to this young lady as his "little sister" (Gauckler, 1922). The little sister would eventually become his wife, but above all an indispensable and deeply committed scientific collaborator (Sorrel-Dejerine, 1980).

In July 1881, the French National Assembly authorized the creation, at the Salpêtrière, of a Clinical Chair in Diseases of the Nervous System, the first such position in the world (Signoret, 1982). Charcot, 57 years old, was the original appointee to this post in 1882. From then on, and still today, the position is referred to as Charcot's Chair. The same year (1882) also marked the successful passing of an examination by Jules

Dejerine, 33 years of age, granting him the prestigious title of "Médecin des Hôpitaux de Paris." He was doing well but the first of the really important career milestones, that of professor,<sup>2</sup> still had to be negotiated. The situation was actually complicated: Dejerine was the student of Vulpian, who got on well with Charcot (Castaigne, 1982). But Charcot did not like Dejerine. It was rumoured, at the Salpêtrière, that Dejerine shamelessly badmouthed the head of the department.

The Parisian medical world experienced, in 1886, two events. The social importance of the first was by far the greatest: for the first time, a woman was accepted as intern, and she graduated at the top of her class. Her name was Augusta Klumpke, the American (Lecours & Caplan, 1984). The second was the annual examination for candidates seeking "agrégation." Hardy was acting president of the jury, a good situation for Dejerine, but Charcot was also a member of this panel and it was known that he influenced a majority of voters. He had in fact decided to block Dejerine's candidacy. Féré, student, secretary, and librarian of Charcot, who nevertheless was on good terms with Dejerine, told him so very clearly. Dejerine reacted promptly, going directly to Charcot and openly demanding an explanation of his decision. Charcot was scarlet with rage and the encounter became very tense (Gauckler, 1922). But it finally was Charcot who yielded and promised his vote. The next day, he would say to Vulpian: "Your Dejerine! Your Dejerine is quite a character after all. He held his own against me! None of my students would have dared" (Gauckler, 1922). Charcot was true to his word at the time of the examination, but he did not promise the vote of his majority. Hardy threatened to resign on hearing this decision. Dejerine was at last accepted along with two of Charcot's students (Ballet and Brissaud).<sup>3</sup>

As a Professor, Jules Dejerine could now, at the age of 37, embark on a noteworthy career. His progress would occur in three stages, the first in Bicêtre, and the two others at the Salpêtrière.

Bicêtre, 1887–1894—It was at Bicêtre, the proving ground of the Salpêtrière and more precisely of those who aspired to Charcot's chair, that Dejerine took over, in 1887, as the head of a clinical neurology ward. He would stay there for 8 years. And what an 8 year period!

There were the lecture theaters, of course, where Dejerine and his students examined patients with minute attention, including aphasics. There was "Siberia," the large and poorly heated room which was, according to Gauckler (1922), the first "clinical neurology museum." There was a laboratory, where a large microtome was available for sectioning (Jules Dejerine and Augusta Klumpke prepared their first classic, *L'Ana-*

<sup>2</sup> Of neurology, of course.

<sup>3</sup> The name of the fourth to be accepted is Chauffard (of the Chauffard–Still syndrome, a form of arthritic tuberculosis which is now rarely seen).

*tomie des Centres Nerveaux*, the original edition of which was published by Rueff in 1895<sup>4</sup>).

Then came the marriage, in 1889, between Dejerine and Augusta Klumpke (he was 40 and she 30). Domestic evenings in the Boulevard Saint-Germain at the Dejerine's were frequent; guests ate well, were entertained in style and, tobacco not yet having acquired its carcinogenic properties, indulged in their *patron's*<sup>5</sup> cigarettes (Gauckler, 1922).

The reputation of Dejerine's clinic developed quickly. Visitors from all over the world were received: Henschen for instance, and Hitzig, Lombroso, and Osler, and Sherrington, and Westphal, along with many others. Abroad, as in France, the "new school" of Bicêtre was compared to the "old school" of the Salpêtrière (Gauckler, 1922). So much so that Pierre Marie, former student of Broca and now student of Charcot, insultingly commented, in 1892 in the *Progrès Médical*, that Dejerine did science as some others played roulette, hedging his bets by gambling on two outcomes ("Qu'il prenne garde cependant qu'en continuant à porter ainsi à la fois sur la rouge et la noire, il risque fort de se ruiner sous peu"; Marie, 1892). Outraged, Dejerine sent representatives<sup>6</sup> to Pierre Marie, challenging him to a duel unless he published a retraction. Marie recanted, more or less, through a letter signed by his own witnesses<sup>7</sup> and published in the *Progrès Médical*: "neither the personal honor, nor the scientific integrity of Monsieur Dejerine were at issue" (Lhermitte & Signoret, 1982). The duel was avoided, at least for the time being.

Charcot died<sup>8</sup> quite suddenly on August 16, 1893, 100 years ago. Had he taken care to nominate his academic successor, Brissaud would certainly have succeeded him (Castaigne, 1982). In any event, it was the latter who gave the lectures on Tuesday during the 1893–1894 year (Guilain, 1923). But the old school was not without its appeal to Dejerine and he was definitely a potential candidate. As was appropriate, he asked for the votes of professors acting on the faculty council. Some committed themselves. Deliberations lasted for months and, in the beginning of 1894, the council elected the second appointee to Charcot's Chair. It would not be Brissaud, nor for that matter Pierre Marie, but neither was it Dejerine. He was to be elected Professor, but of history of Medicine, while Brissaud would become Professor of medical pathology (Castaigne, 1982). As for Charcot's Chair, it would go to the elder of the school, Fulgence Raymond. The status quo was thus preserved (Alajouanine, 1979).

<sup>4</sup> And the quality of which is such that it is still *reedited* nowadays.

<sup>5</sup> This word can doubtlessly be considered a *euphemism*.

<sup>6</sup> Letulle and Gley.

<sup>7</sup> Brissaud and Chauffard (!).

<sup>8</sup> At the age of 67.

La Salpêtrière, 1894–1911—Dejerine came to the Salpêtrière with Madame and their students. He took direction of the Jacquart ward, the one which, a short time before, had been headed by his mentor Vulpian. Work continued as in Bicêtre but without quite the same zest as in the good old days.

In line with the tradition of the Chair, Raymond taught at the Salpêtrière on Tuesdays; Dejerine, on his part, gave the Wednesday and Thursday lectures. In addition, overseen by a photograph of Vulpian, he forged links between his clinical and anatomopathological observations; he elaborated on syndromes. And, primarily, he worked on his second classic, *Sémiologie des Affections du Système Nerveux*, which Masson would publish for the first time in 1900.<sup>9</sup>

In 1897, it was Pierre Marie's turn to endure exile in Bicêtre. And it was shortly before the end of his decade there when, disclaiming the aphasiologic teachings of his mentors Broca and Charcot, a doctrine that he himself had brilliantly summarized and disseminated in 1888, Pierre Marie published in three consecutive issues of the *Semaine Médicale* (Marie, 1906) the authoritative texts that set forth his new views on aphasia. At the Salpêtrière, the former head of the "new school" of Bicêtre was deeply offended.

Fulgence Raymond would soon be 65 years old. The end of the interim was approaching. The potential successors were Dejerine (aged 59) and Pierre Marie (aged 54). The duel would now take place, this time, a confrontation in the form of three "Discussions sur l'aphasie" held during the Summer of 1908, at the regular meetings of the *Société de Neurologie de Paris* (Klippel, 1908). The two antagonists would maintain their positions, which were known by all, such that the best moments of the debate would feature the partisans: on Dejerine's side, Augusta Klumpke, André-Thomas, Dupré, and even, occasionally, Ballet; on Pierre Marie's side, Brissaud, Souques, and Moutier (the story and outcome of the famous debate have been recounted by Lecours & Caplan, 1984; Lecours, Chain, Ponset, Nespoulous, & Joannette, 1992; Lecours & Joannette, 1984).

Fulgence Raymond died on September 28, 1910, on the eve of his 66th birthday, before his official retirement.

La Salpêtrière, 1911–1917—Perhaps Pierre Marie tried to strike a diplomatic note when he wrote, in the *Larousse Mensuel Illustré* of December 1910, that the School of the Salpêtrière owed its world-class reputation to the research and teachings of Fulgence Raymond. Some might have found this an exaggeration. On the other hand, it would soon be 20 years since the death of Charcot and the question of the center's academic influence had undoubtedly lost some of its interest. In any event,

<sup>9</sup> And for the last time in 1926.

it was to the erstwhile student of Vulpian, to the author of the *Anatomie des Centres Nerveux* and of the *Sémiologie des Affections du Système Nerveux* that the Chair would go. That was the decision of the faculty council. Doctor Jean-Baptiste Charcot, son of Jean-Martin and illustrious captain of the good ship, the "Pourquoi pas?", escorted Dejerine on his official arrival to the chambers of the new head of the department (Gauckler, 1922; Sorrel-Dejerine, 1980).

The first 2 years of the third phase were a direct continuation of the second, with the exception that Dejerine now taught on Tuesdays as well as Fridays. In 1913, Dejerine was confined to bed for a rather lengthy time with an attack of uraemia. Then France was embroiled in a war and the composition of the patients in the Clinic of Disorders of the Nervous System changed radically. Dejerine drove himself mercilessly in spite of progressively failing health. He died of uraemia on the 24th of February 1917, at the age of 67. Pierre Marie was 63 and finally succeeded him to Charcot's Chair.

### Summary

If Dejerine should be regarded, as we have noted above, among the founders of neurology, he also was a founder of neuropsychology. Indeed, his first publication on aphasia appeared in 1879, that is in the year when he reached 30, obtained his doctorate in medicine, and met Augusta Klumpke. Twenty-three other publications on acquired disorders of spoken and written language were to follow, including those discussed below on pure word blindness, and also the first anatomoclinical description of what is today called "progressive aphasia" (Sérieux, 1893; Dejerine & Sérieux, 1897). To this should be added numerous articles on various cognitive disorders other than those mainly affecting language, major passages of the two classical monographs devoted to aphasias, to astereognosis, etc.; it is also appropriate to mention three doctoral dissertations on aphasiologic research conducted by Dejerine's students (Bernheim, 1900; Mirallié, 1896; Pélissier, 1912).

Dejerine had the reputation of being a plain-spoken man, leading to his language a direct character that some appreciated and others did not. It was said that only in one special circumstance would he abandon this manner of speaking and resort to the conversational guile of his Savoyard ancestors—when he had to convince a reluctant family to authorize an autopsy in cases where he had conducted an advance clinical investigation and wished to verify the lesion location. He could then, it was said, go so far as to talk about the vested interests of the family, alluding for instance to the possibility of hereditary illness (Gauckler, 1922). On this topic, some have commented on the story of an autopsy that he conducted alone, in town, while investigating a particularly remarkable case of pure word blindness. . . .

## SECTION B

*Theoretical Background—Dejerine's Neuropsychological Framework*

A publication in 1891 on disorders of writing in aphasia by Dejerine contains a rudimentary functional architecture of the basic word-level components he thought were needed for reading, writing, and pronunciation and provides the foundation for his subsequent work on pure alexia. He indicates that the diagram is nothing more than the schema originally proposed by Wernicke, "lightly modified" (*légèrement modifié*), and that it should be thought of only as a preliminary first step that fit well with existing data (Fig. 1).

There are a number of comments that can be made about his theoretical assumptions as formulated in the diagram. First, he incorporated the standard distinction between auditory images (*centre des images auditives des mots*) and articulatory processes (*centre moteur de l'articulation de la parole*) that was maintained in the Wernicke/Lichtheim diagram of that period. These two functional components were thought to be interactive during the perception and production of speech. According to Dejerine: "In thinking, we hear the words spoken and we are aware of the movements necessary to pronounce them. Cases in which this phenomenon does not occur are extremely rare. Thus, auditory and motor images are intimately related during the generation of inner language, and it is this union which constitutes *the idea of the word*" (*italics are Dejerine's*).

The intuition that abstract phonological word forms are derived from

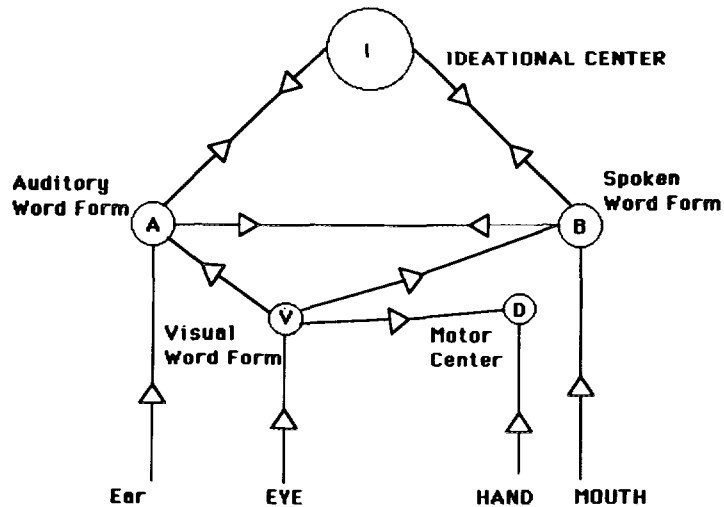


FIG. 1. Dejerine's architecture of the reading mechanism in relationship to the processing of spoken language.



the interaction between perceptual and motor processes is, of course, reinvoked by a number of contemporary theorists. Prinz (1987) aptly summarizes this position by noting that "In a sense, the listener is considered a rearticulator of the sound pattern he/she is listening to" (p. 63). A fairly recent paper by Studdert-Kennedy (1987) that explores this position concludes with a statement that is startlingly reminiscent of the one we have reproduced from Dejerine: ". . . the emergence of the phonetic segment as a perceptuomotor unit brings the entire adult lexicon, insofar as it is cognitively available, within the child's phonetic reach" (p. 79).

The Wernicke-Lichtheim model, as we have indicated, provided the framework for Dejerine's thinking on the basic components for reading and writing. He added a separate visual center for words (V in the diagram), established through perceptual training ("par l'éducation de l'oeil P"), and here it is apparent that his views departed clearly from those of Wernicke (hence, presumably, Dejerine's comment that the model had been slightly altered), who remained strongly opposed to the idea of a separate visual representation for words: "In fact, it would be tempting to assume the existence of a special cortical center for written language which is completely independent of speech functions, or rather, one which is analogous to the actual motor and sensory speech centers. The former might then be designated as the site of motor imagery involved in writing, and the latter as the site of the visual memory images for words. . . . Nevertheless, the indefensibility of this grossly schematic mode of interpretation has become more and more apparent. . . . It may be said that for most types of reading material, the act of reading occurs by means of letter-spelling, and, therefore, we accept the existence of memory images for letters only, and not for entire words. . . . (The) storage of transient letter combinations as special memory images of words would seem to be a superfluous performance of the brain, since the corresponding images of the sounds of words are already present. We may assume that such storage of words does not take place" (Wernicke, 1906; translation by Eggert, 1977, pp. 242, 243, and 244).

Wernicke's position was that reading is accomplished by deriving sound units from letter identities, and he contended that only in very unusual circumstances (in cases where no auditory language could develop, as in profoundly deaf individuals) would there be a need to postulate visual units corresponding to whole words. Dejerine, by contrast, assumed that reading normally takes place by initially contacting visual word forms and that these word-specific representations then gain access to other components of the language mechanism. The modern analogue of these opposing viewpoints can be found in the ongoing debates that emerge from recent connectionist approaches to modeling the pronunciation of written words. An influential simulation by Seidenberg and McClelland (1989), for example, is based on the assumption that: "Lexi-

cal memory does not consist of entries for individual words; there are no logogens. Knowledge of words is embedded in a set of weights on connections between processing units encoding orthographic, phonological and semantic properties of words, and the correlations between these properties" (p. 560). The rival position, adopted by many theorists of language processing (e.g., Carr & Pollatsek, 1985), is that whole words have distinct representations as perceptual units in visual and phonological lexicons.

Wernicke, in one of his last publications, took care to explicitly distinguish his view from Dejerine's: "Certainly, on the basis of Dejerine's ideas regarding the extent of the speech center, it is obvious that he feels that the optic word center, so named by him, exerts the same influence on oral language as does the acoustic word center. This view . . . we cannot accept, since in normal individuals visual images exist only for letters and not words" (Wernicke 1906; from Eggert, 1977; p. 257).

We cannot resist the speculation that the reason for the difference in standpoints between Wernicke and Dejerine has to do with the fact that German orthography, the system most familiar to Wernicke, is fairly predictable in its spelling to sound correspondence, so that it is at least plausible to infer a mechanism relying only on the pronunciation of units confined to letter groups, given that such a mechanism would often deliver the correct rendering of any word in the language even though no mapping exists at the level of whole words. French orthography, like English, is notoriously irregular, and Dejerine may have found it difficult to explain how skilled readers can easily derive the correct pronunciation of both regular and exception words (e.g., *fille* versus *ville*) without allowing for a complete representation of their orthographic form.

Whatever the reason behind Dejerine's assumption, the notion of a visual word center raised the question for him of how this component would link up with the rest of the language mechanism. Could there be a direct connection between the visual word and its conceptual representation? This issue has been one of the central themes in the study of acquired dyslexia over the last 20 years (see for example, the widely discussed phenomenon of semantic paralexias in deep dyslexia; Coltheart, Patterson, & Marshall, 1980) and it is now widely conceded that such a route does exist, although the extent to which normal fluent reading of text relies solely on the direct mapping of visual word forms to meaning remains controversial.

Dejerine, in considering the possibility of a semantic route from the visual word center, explicitly rejects the idea. "The center V is not in direct communication with the center for ideation; it only connects with it through the centers A and B. In effect, we do not read directly, we read by activating auditory images which in turn activate the ideational center. The proof of this is that when we read even silently, we hear the

words resounding in our ears, at the same time as we are conscious of the movements necessary to pronounce them." Dejerine's argument for the importance of phonology in reading would obviously not satisfy most cognitive psychologists today, but the powerful evocation of spoken language from the processing of written text is a perception shared by all normal readers. Dejerine expresses what is reaffirmed in the following eloquent statement by a modern writer: "Ever since I . . . started to read . . . there has never been a line that I didn't hear. As my eyes followed the sentence, a voice was saying it silently to me. It isn't my mother's voice, or the voice of any person I can identify, certainly not my own. It is human, and it is inwardly that I listen to it" (Welty, 1983, p. 12).

Having constructed a preliminary architecture for reading, Dejerine then turns to the question of writing. "The matter," he remarked, "is easy to understand; the centers A and B, constituting by their union the form of a word, will activate the center V, reinvoking the corresponding visual description. The center V then acts on the motor center of the upper limb D which in turn determines the movements executed by the hand M to produce writing."

It can be seen from these comments that Dejerine did not assume a separate orthographic center for writing, so that the same representation V was used for input and output. The question of single versus dual orthographic lexicons is still hotly debated among cognitive neuropsychologists, some authors arguing for separate representations underlying reading and writing (Beauvois & Dérouesné, 1981) and others trying to marshal evidence that would support the idea of a single orthographic mechanism with separate access procedures for input and output (e.g., Behrmann & Bub, 1992).

## MONSIEUR C

### *Background*

The phenomenon of pure alexia had been reported by other theorists prior to Dejerine's contribution in 1892, but the importance and subsequent influence of the latter work is due to the details of the behavioral evidence and the force of the neuroanatomical arguments.

The patient was a 68-year-old man (referred to as Monsieur C), who was considered highly intelligent and enjoyed excellent health until his cerebrovascular accident on the 25th of October 1887. He worked as a textile merchant and had gained considerable wealth, allowing him financial independence. Both he and his wife were cultivated people and accomplished musicians, and the patient read widely and enjoyed singing either alone or with his wife. He easily could sight-read difficult musical

passages before his illness (we will return to the question of the patient's ability to read music after the onset of his word blindness at a later point).

On the 23rd of October, the patient went for a walk after experiencing some right-sided numbness of his arm and leg. He remembered clearly that he was quite capable of reading the advertisements in shop windows during that time. The next day the feeling of numbness in his arm and leg persisted and on the following day, Monsieur C found to his alarm that he was completely unable to read, even though he had no difficulty writing nor any problem communicating or understanding spoken language. Thinking that his problem was visual in nature, he visited an ophthalmologist (Landolt), who recorded the following observations: "Faced with the Snellen chart, he could not name any letters of the display, even though he affirmed that he could see them perfectly. Instinctively, he carried out the gesture corresponding to their shape without succeeding in naming them. Required to copy on paper what he saw, he managed but not without effort, to reproduce the letters line by line, as if he was dealing with a technical drawing, examining each stroke to establish the details of the pattern. In spite of all his efforts, he was incapable of naming the letters. He compared the A to an easel, the Z to a snake, the P to a buckle." Landolt confirmed that the patient had normal visual acuity with corrective lenses, but examination of his visual fields disclosed a right homonymous hemianopia and a complete hemiachromatopsia.

Dejerine first tested the patient on the 15th of November 1887. He observed that Monsieur C spoke without difficulty and that his manner of conversing indicated a highly intelligent, educated gentleman. He recognized all visual objects placed before him, naming them easily and without any hesitation. Dejerine saw no suggestion of visual agnosia, optic aphasia, or verbal deafness. The patient understood everything communicated to him.

Monsieur C wrote both spontaneously and to dictation fluently and without error. A comparison of samples of Monsieur C's handwriting before and after his illness indicated no fundamental changes, except that the letters were somewhat larger than before. Dejerine observed no spelling errors, no letter transpositions, nor any other form of agraphia.

#### *Monsieur C's Word Blindness*

Presented with the sentences that he himself had written a short time before without effort, and asked to read them. Monsieur C expressed the frustration and bewilderment shared by all patients who must reconcile themselves to the paradox inherent in pure alexia: "I still know how to write the letters, there they are; why can't I read them?"

According to Dejerine, Monsieur C's reading impairment was total:

"The patient did not recognize a single letter, not a word, except his own name on occasion." By contrast, he argued that Monsieur C had no trouble identifying other kinds of visual material, including numbers (the evidence supporting this contention will be discussed below). Dejerine's view was that Monsieur C had completely lost the significance of visual letters and words as *linguistic* codes and that they were represented for him only as meaningless perceptual forms. "In looking at an object with one eye or with both eyes, we see them with our two hemispheres; it is the same with letters; we see them with our two occipital lobes but we see them with the help of these common visual centers as designs of some sort, like we see the letters of the language which is unfamiliar to us (like I see, for example, Russian or Hebrew characters). In order for us to identify a letter, for the collection of certain letters to activate the *idea* of a word, it is necessary that these common visual cortical centers enter into a connection with the language zone" (italics are Dejerine's).

It is clear that Dejerine conceived of pure alexia as a specific deficit confined to the impaired processing of orthographic code as opposed to a more general perceptual disturbance that affects a range of visual material (both linguistic and nonverbal). The distinction is as important and controversial today as it was in Dejerine's era. There have been modern interpretations of the disorder that attempt to link it to a global disturbance in the rapid processing of multiple visual forms, regardless of their verbal or nonverbal nature (e.g., Kinsbourne & Warrington, 1962), thereby avoiding the necessity of describing the syndrome as a specific impairment in the visual identification of orthographic code. A rival hypothesis by Warrington and Shallice (1980) that is very close to the position originally advocated by Dejerine returns to the concept of a specialized visual word center that is selectively affected by cortical damage. Farah (1990) indicates precisely what is at stake given the two theoretical possibilities: "The existence of a selective impairment of reading, without concomitant impairments in other visual or language processes, implies that there is some region of the brain that is necessary for and dedicated to reading. If . . . the word form hypothesis is true, this implies that localized brain functions include functions that are evolutionarily very recent, that require extensive instructions to learn, and that relatively few individuals in the history of the species have ever possessed. While not a priori impossible, this conclusion represents a surprising departure from other functions that we know to be localized (perception, motor control, language, memory). Thus, the issue of whether pure alexia represents a selective impairment for reading per se, or whether it is a manifestation of a more general impairment, has implications beyond our understanding of reading impairments. It bears on the issue of how the functional architecture of the mind is mapped onto the physical architecture of the brain, and in particular on the distinction between the kinds

of psychological processes that make use of localized dedicated hardware and the kinds that do not" (pp. 114–115).

Dejerine was clearly aware of the contentious aspects of his position, implying as it does a distinct representation for visual words that is different from other symbolic material (e.g., digits) and from visual objects. He wished to establish that Monsieur C had no apparent loss of object identification and that he could still identify numbers correctly. Thus, Dejerine records that the patient was able to perform fairly complex arithmetic operations; he could multiply a sequence like 2745941600 by 451, add two 8-digit numbers, and perform difficult division and subtraction. We note, however, a curious discrepancy on this fundamental point with Landolt's observations of Monsieur C. His description of the patient's ability to decipher numbers was as follows: "In carrying out arithmetic operations, he succeeded in doing a very simple addition, since he recognizes the numbers easily enough; however, he proceeded with extreme slowness. Numbers are poorly read because he could not recognize the value of several digits at the same time; presented with the number 112, he declared "It's a 1, a 1 and a 2" but it is only on writing them down that he managed to say one hundred and twelve."

It is perhaps worth considering that Dejerine's inference that Monsieur C had no constraints on his ability to read numbers could have been due to the fact that this function had recovered by the time he first saw the patient. We find this possibility quite remote, however, given the very short period between Landolt's preliminary assessment, carried out on the 9th of November, and the dates of the testing reported by Dejerine, which took place during the same month of that year. A more plausible explanation is that the way Dejerine tested the patient's ability to cope with numbers was insufficient to reveal any deficit, presumably because he was not predisposed, given his theoretical standpoint, to thinking of the disturbance as extending beyond the processing of orthography to numerical symbols. In the examples he supplies of Monsieur C's ability to carry out arithmetic operations, Dejerine indicates that the numbers were written by the patient himself (presumably to dictation), a task, according to Landolt, that would not reveal any disturbance in the processing of multiple visual elements. In addition, the actual arithmetic operations required of Monsieur C (addition, multiplication, etc.) do not require rapid visual identification of several numbers concurrently, leaving us with some doubt as to the status of the patient's reputed ability in this domain.

Landolt's remarks on Monsieur C's extreme slowness to carry out simple arithmetic and his failure to extract the identity of a sequence like 112 without writing the numbers down are strongly suggestive of a simultanagnosic disorder, reported by Kinsbourne and Warrington (1962)

to be a commonly associated perceptual disturbance underlying pure alexia. Indeed, these authors attempted to account for the reading disturbance as a direct consequence of the constraints on the processing of multiple visual forms: patients with simultanagnosia require enormously protracted exposure durations before they can identify a number of visual forms presented together, but display no such impairment when dealing with only a single object. Kinsbourne and Warrington argued that this kind of general simultanagnosic disturbance might provide an explanation for the laborious letter-by-letter approach to word reading in those cases of pure alexia who can still identify single letters. Farah (1990) has recently advocated a similar interpretation of letter-by-letter reading.

The evidence from Monsieur C's difficulty in reading numbers comprising more than one element suggests that he may well have suffered from the kind of simultanagnosic disorder that has been observed in pure alexia. However, the interpretation of his reading deficit is complicated by the fact that he clearly had a more extensive disorder that included the processing of single letters. Dejerine's initial position was that Monsieur C actually had lost the significance of letters as orthographic elements; their conceptual status in the visual modality was, on this interpretation, no different than that of unfamiliar designs to him. Regardless of the possibility of a generalized simultanagnosic disorder, evidence for a disturbance in the visual identification of letters without a corresponding deficit in the identification of single digits would be of great relevance to Dejerine's explanation of pure alexia.

To support his view, he argued that copying of words by Monsieur C was executed mechanically and segment by segment, as if the patient was attempting to duplicate an arbitrary design: "This fact is very clear and most striking when we ask him to copy successively cursive and printed text. In effect, the form of the letters is changed in the two situations. In the one case, copying of cursive handwriting, his letters are badly structured, but their general form is that of the letters he produces in writing spontaneously and to dictation. In the second case, by contrast—copying of print—the form of the letters is quite different and approximates that of the printed characters as is demonstrated by the S, and J and the C in the sample presented below."

Dejerine supplied a fragment of the patient's attempt at copying print to support his claim, which we have reproduced for inspection (Fig. 2). His interpretation was that Monsieur C was forced to carry out the task by executing a literal transcription of the shape of each letter, because he no longer could perceptually derive an abstract verbal identity of letter forms. The patient's usual style of handwriting (either from dictation or spontaneously) was therefore disrupted when he was engaged in copying print and Dejerine focusses on particular instances (e.g., the shape of the

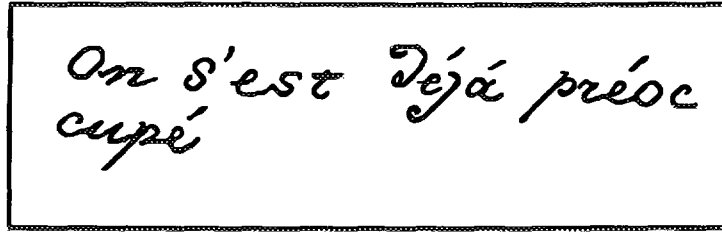


FIG. 2. A fragment of Monsieur C's copying of printed text. The phrase reads: "On s'est déjà préoccupé." The sample has been digitized from the original article and slightly enhanced for visual clarity.

letter S which now conforms to the printed as opposed to the cursive style) to bolster his claim that the patient had lost the perception of letters as abstract linguistic identities.

From even a casual glance at the written material Dejerine provides, it is apparent that he is stretching matters in claiming that Monsieur C is literally treating visual orthographic sequences as if they were unfamiliar designs. True, the patient reduplicates the printed form of certain letters. We also note the distorted aspect of the O in *préoccupé*, where the usual connecting loop on the top of the letter, readily apparent in the patient's spontaneous handwriting, is absent. However, this outcome may merely reflect the laboriousness of the copying process, with frequent pauses to take in additional letters in the word (i.e., the patient is transcribing one or two letters at a time and the fluency and style of handwriting is correspondingly affected). Certainly, we can discern many instances that violate his assumption of slavish, line by line copying—the E is typical of cursive writing, as is the A and R. The D in *deja*, although unusual for our time, has roughly the same contour as occurs in the patient's spontaneous handwriting.

Interestingly enough, Dejerine seems to have ultimately abandoned the idea that copying in pure alexia is based on a line by line transcription of the letters without any higher level interpretation of their form. Discussing the matter in a publication with Pélissier (1914), many years after the appearance of his initial report to the Parisian Biological Society, he comments: "Now, this slavish copying of print into print, so common in sensory aphasia, is not habitually observed in pure word blindness. The explanation of this is no doubt linked to the fact that in pure word blindness, the visual verbal image remains intact. The subject who spontaneously writes perfectly, who mentally sees the letter that he wishes to write will not resign himself to a slavish transcription without significance, but will search, on the contrary, for a model to recognize each letter and to write in cursive form the character that he thinks, often erroneously, he has identified."



Dejerine's modified position on the nature of copying in pure alexia may well have been based in part on a rather striking observation he made of Monsieur C while the patient struggled to name visual letters. He noted that he would on occasion arrive at the identity of a letter or even a word, *if the examiner allowed him to trace out their forms with his finger (or foot)*. This surprising and potentially informative compensatory act has not yet been discussed in the modern literature on pure alexia, presumably because it does not occur in all patients and in those who do resort to it, the maneuver may be sufficiently covert to have eluded detection. Our own experience of pure alexics is not inconsistent with Dejerine's observation: we have on occasion noticed that some patients may rapidly trace letters on the table or on the palm of their hands with their index fingers when asked to identify them. In some cases, the gesture may be confined to a subtle flicker of the thumb.

How the phenomenon of letter tracing augments visual identification has not yet been explained by contemporary theories of pure alexia. We require a better account of the mechanism underlying abnormal letter processing in the relevant cases, particularly since the patients who have been described in the modern literature (under the category of letter-by-letter reading) generally remain capable of identifying visual letters (e.g., naming or categorizing them), although their performance is much slower than normal and mislabeling does occur from time to time.

Judging from Dejerine's comments, he assumed that the mechanisms responsible for visual letter identification and production of their written form could interact, and the retraction of his previous statement on the quality of copying in the syndrome amounted only to the concession that any patient who retained the abstract conceptual description of a letter (the visual verbal image) would not generally be reduced to duplicating the target as a meaningless design, because the knowledge of the movements necessary to produce the form would partly govern the patient's response.

His final position on pure word blindness, summarized in the paper coauthored by Pélissier, remained in essence unchanged from his original conceptualization of the syndrome. The disorder represented, according to Dejerine, a failure of the perceptual forms of letters to activate the corresponding word as a *linguistic* unit. The stored visual description of the word (*la notion graphique du mot*; Centre V in the diagram) was intact because "the subject writes as easily as before and as a result, he appears capable of "mentally seeing" the word at the moment of writing it" (Dejerine & Pélissier, 1914). Centre V, however, requires access to the phonological components (the coactivation of Centres A and B) of the language mechanism for interpretation as a verbal entity, and it was this connection directly from the visual pathways that was no longer possible in cases of pure alexia. From our contemporary viewpoint, we

could say that Dejerine thought of the visual word form system in terms of a device mediating access to (and retrieval from) the lexicon that did not yield a conscious word percept until additional phonological and semantic levels of representation were contacted. The idea is closest to a type of model known as activation-verification (AV) in the now extensive literature on word recognition. Paap, McDonald, Schvaneveldt, and Noel (1987) summarize the basic assumption underlying this conceptual approach as follows: "The AV model consists of encoding, verification and decision operations. Encoding is the process that leads to the unconscious activation of learned units in memory. . . . Verification follows encoding and usually leads to the conscious recognition of a single lexical entry from the list of candidates. Verification should be viewed as an independent, top-down analysis of the stimulus that is guided by a stored representation of the word" (p. 222).

If visual words and letters no longer made contact with the rest of the language system in pure alexia and therefore could not be "verified," according to Dejerine, they nevertheless retained whatever other kinds of learned associations they held for the patient. For example, the processing of visual forms as numerical (i.e., nonlinguistic) concepts should be largely intact (Dejerine remained adamant on this point in all his discussions of pure alexia), because they could "act as symbols and do not call into play the psychological processes for reading." In evaluating Monsieur C's ability to identify letters, Dejerine reported that the retired businessman still retained his knowledge of a code he had devised to conceal the true price of material from the gaze of potential customers (a widely used subterfuge by merchants of that time, apparently) by replacing each number between 0 and 9 with a corresponding letter. Monsieur C, when asked, could indicate the "mark of the house" (la marque de la maison), although the example given in the text only shows that he could write down the sequence he had used in the conventional order (C toi fameux = 1234567890), even though he was subsequently incapable of reading the names of any of the letters he had produced. We do not know for certain that Monsieur C would have been able to translate any arbitrary string of letters taken from the code into the corresponding number, but it is apparent from Dejerine's interpretation that he was in little doubt that the patient retained the perception of letters as numerical values despite the fact that he had completely lost their visual identities as orthographic units.

### *Discussion*

These, then, were the facts that Dejerine had gleaned from Monsieur C's performance and that he marshalled to defend his basic interpretation of pure alexia. The patient had no apparent difficulty in identifying visual

objects or faces, he spelled quite normally, and he could identify single digits correctly, although he appeared to have more trouble with several digits together, an impairment that probably reflected a general simultanagnosic disorder often cooccurring with pure alexia. He had no language deficits of any kind. His reading disturbance was profound and irreversible; Dejerine indicates that Monsieur C remained forever cut off from the letters on the posters and signs in shop windows viewed daily on his walks along the boulevard Montmartre until his death 4 years after the onset of his word blindness.

The main features of the patient's reading are rather complex, more so in fact than the already complicated pattern that has been the subject of a number of reports in the modern literature on pure alexia. Monsieur C was not capable of the usual letter-by-letter naming strategy that is taken as an important diagnostic feature of the syndrome currently being actively researched by cognitive neuropsychologists. If he did rely on a compensatory strategy in an attempt to reconstruct the word, it appeared to involve feedback generated by the activity of the writing mechanism. While Monsieur C was unable to derive the name of a letter directly from vision, he occasionally managed to do so by tracing the surface form with his finger or other extremity.

We remain somewhat unclear on the precise nature of the information that the patient actually could obtain from the visual analysis of letters, an issue that is of considerable interest given Monsieur C's ability to label other visual symbols like digits. Dejerine's contention was that letters were the perceptual equivalent of meaningless designs to the patient but the assumption, at least in its most basic form, is called into question by the fact that copying of print often yielded the cursive form of the letter (despite Dejerine's initial remarks to the contrary). On the other hand, it is not the case that the patient's difficulty can simply be reduced to letter anomia (i.e., the failure to derive the name of a letter after its identity has been established). Dejerine was clearly aware of this possibility and deals with it explicitly in his later discussion of the syndrome with Pélissier. He notes in milder cases, identification of single letters or even syllables may be relatively spared, and that when this capacity remains available and the names of the letters are derived, laborious deciphering of the word may occur. He describes one patient (MC) who was capable of this approach to reading. Faced with the written phrase "le siège d'Andrinople" this patient proceeded as follows: "Le, yes, that's it . . . L . . . E . . . le, after that an S . . . E, siècle, I have perceived, Le siècle . . . D . . . E . . . that's an E, let's see . . . and after that . . . what's that? . . ." (etc.).

Here, then, is the kind of strategy that has come to be known as letter-by-letter reading in the modern literature, a phenomenon that occurs, in Dejerine's view, when "letters and syllables are still identified, but

recognition is so slow and laborious that the patient will not succeed in assembling these word fragments into a single term."

Even when explicit naming of letters is not possible, Dejerine noted, it may still be the case that they are fully identified, although less direct measures than overt labeling by the patient must be used to demonstrate this capacity. Thus, he argues, intact spelling by means of letter cubes (a skill that was fully retained by patient MC) or the ability to point to one letter from a number of choices would indicate some preservation of letter identities. Wernicke (1906), in his discussions of this issue, is perhaps even more sophisticated, mentioning tasks that require judgements of the equivalence between upper- and lowercase forms of the same letter, a test that is now widely utilized to assess letter identification in pure alexia (e.g., Reuter-Lorenz & Brunn, 1990).

In the light of these remarks, we would accept Dejerine's conclusion that Monsieur C was unable to identify letters, in the sense that we assume he would have been unable to perform cross-case matching (i.e., judging that A is equivalent to a) or to choose a letter from a set of alternatives if given the name, without resorting to the artificial strategy of tracing the shape of the letters with his finger. Yet, we cannot agree that Monsieur C had no visual access whatsoever to the stored representation of letters as graphic elements, given that he would produce cursive forms when asked to copy printed text. A further tantalizing claim is that Monsieur C was able to derive a number from the letter code he used for marking the price of materials, although we find Dejerine's observation on this point somewhat ambiguous.

If we were forced to draw any further conclusions regarding the nature of Monsieur C's extraordinary reading deficit, we would begin by noting that his performance with respect to the identification of orthography reminds us of the oddly paradoxical and unusual dissociations between language and vision that occur in patients with optic aphasia (Lhermitte & Beauvois, 1973; Beauvois, 1982; Beauvois & Saillant, 1985). Thus, in these so-called "visual-verbal" disconnections, patients will appear to have completely misidentified an object from vision, both in their labeling response and, depending on the circumstances, in some tests of nonverbal classification. However, further more subtle investigation will reveal that at least partial access to the visual identity of the target has taken place (e.g., the patient may provide a gesture that demonstrates he has accessed knowledge regarding the use of the object, or he may accurately distinguish the object from plausible but nonexistent ones), despite the fact that he does not indicate correct understanding in his verbal remarks to the examiner (nor for that matter, in the way he uses language to represent the situation to himself; cf. Beauvois & Saillant, 1985). Monsieur C, by analogy, seemed to have lost the significance of visual letters (but not objects or even numbers, apparently) when asked to verbally

interpret their form, yet we have evidence that he retained some access to their perceptual identity from the nature of his copying performance. In their discussion of optic aphasia, Riddoch and Humphreys (1987) argue for the possibility that manual gestures associated with an object may be directly activated from a representation of its visual structure, without full access to the object's semantic description. We may speculate that Monsieur C's copying of printed text was driven by a similar direct translation of letter shapes into abstract motoric schemas.

One further analogy might be drawn between Monsieur C's word blindness (or in this particular instance, letter blindness) and optic aphasia. A patient with optic aphasia will immediately identify and label an object encountered through a nonvisual channel (e.g., an object that he is allowed to touch). Monsieur C likewise could easily arrive at the identity of a letter if he was allowed to access its spatial description by manually tracing over its contour.

Our best guess, then, is that Dejerine's case departed from the conventional aspects of pure alexia—in which the identification and labeling of single letters is generally intact—because the patient had a more extreme deficit, confined to the visual modality, that affected the mapping between individual letter shapes and their corresponding representation as lexical forms.

#### *Some Brief Comments on the Neuroanatomy of the Lesion*

Monsieur C suffered another stroke on January 5, 1892 that left him with impaired spoken language (many paraphasic errors) and a total agraphia. He produced only a few repetitive scratches on the page when asked to produce a written sentence. He died a short time after this second episode, and Dejerine was subsequently given permission by the family to carry out an autopsy. The damage produced by the more recent cerebrovascular event was confined to the posterior and inferior part of the left parietal lobe, the angular gyrus, and the area between the second and third left temporal gyri and the first occipital gyrus.

The extent of the lesion that caused the pure alexia was as follows: In the posterior half of the occipitotemporal boundary was an area of damage measuring 5.5 cm in length and 1.5 cm across at its widest point, which tapered posteriorly toward the extremity of the occipital lobe. A second focus, measuring 2.5 cm in length, occupied the posterior extremity of the calcarine fissure and involved the superior surface of the lingual gyrus and the internal surface of the cuneus. A third small area of damage (1 cm in length and 5 mm wide) involved the cuneus and the internal surface of the gyrus that borders the interhemispheric fissure. There was a fourth section of necrotic tissue, connecting the posterior extremities of the first two, that began at the tip of the occipital lobe and extended to

the lateral surface. A small additional lesion was recorded on the inferior part of the splenium of the corpus callosum, 1 cm long and 3 mm wide.

Dejerine's reasoning on the functional consequences of the lesion began with the assumption that the left angular gyrus was the critical region involved in representing the visual word form. In a previous publication (Dejerine, 1891), he had documented a patient with alexia *and* a severe writing disorder, where the lesion (established at autopsy) was found to be "the size of a five-franc piece, occupying the inferior three-quarters of the angular gyrus and terminating at the external vertical fissure." The details of the patient's reading and writing/spelling are scanty, but Dejerine's conclusion was that both impairments "are linked to one and the same cause, the loss of visual images for letters."

The anatomical explanation of Monsieur C's word blindness and preserved spelling followed directly from this initial position: the damage must be located in such a way as to disrupt the connections of both occipital lobes to the angular gyrus. As a result: "He saw letters as designs of some sort and copied them as such, but they had no meaning for him, because the connections between his two visual centers and the visual word form (left angular gyrus) were disrupted." We reproduce in Fig. 3 the schematic that Dejerine used to illustrate his reasoning (for a sophisticated contemporary analysis of the possible neuroanatomical correlates of pure alexia, see Damasio & Damasio, 1983, who include in their paper, a discussion of the neuroanatomical observations recorded by Dejerine).

He explicitly stated that he did not think the tiny lesion of the splenium of the corpus callosum should be considered to play an important functional role in the syndrome. The damage to the white matter tract from the left occipital lobe was sufficiently great to disrupt the communication of both visual cortices to the angular gyrus without requiring any additional speculation on the effect of the splenial damage.

The emphasis of the splenium as a major factor in pure alexia has generally been attributed to Foix and Hillemand (1925) and in the modern literature to Geschwind (1965) but it was the great Wernicke in 1906 who probably was the first to anticipate this argument. He did not concur with Dejerine, as we have seen, on the existence of a specialized visual word form center that could be localized in one hemisphere. Rather, the only representation that need be postulated for written language over and above the components for auditory language was, in his opinion, the visual form of individual letters. Wernicke doubted that this stored information was restricted to the left hemisphere and he argued that if letters were represented bilaterally, the occurrence of pure alexia may require a callosal disconnection. "It is obvious, therefore, that the question of unilaterality of a visual word, or more accurately, letter center is very important for the entire doctrine of localization. If it is rejected—which

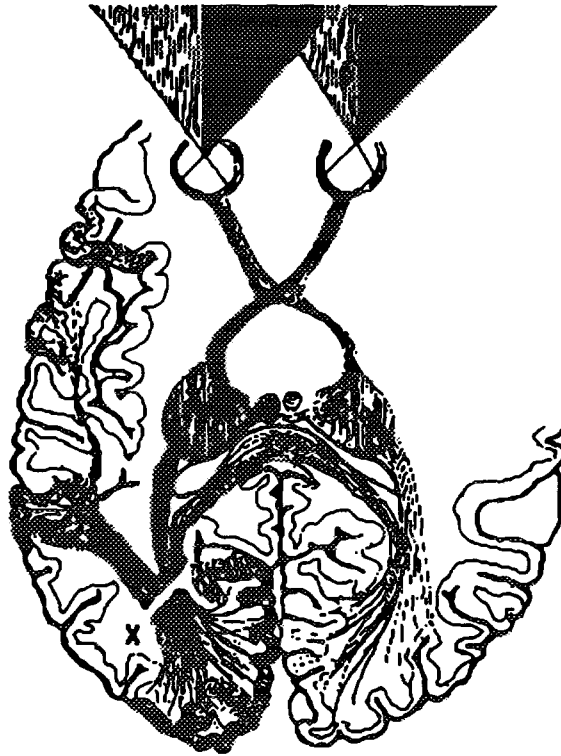


FIG. 3. The disconnecting lesion, marked with an X, hypothesized by Dejerine to produce word blindness. The drawing has been digitized from the original article and computer enhanced.

in my opinion must happen—the door is open for investigation of a pathology of the callosum in causing a subcortical alexia or pure word blindness” (from Eggert, 1977; p. 260).

#### CONCLUDING REMARKS: A POSTMORTEM ON DEJERINE'S HYPOTHESIS

Pure alexia—at least the variant known today as letter-by-letter reading—has been the focus of a number of recent articles by cognitive neuropsychologists. Unfortunately, despite this considerable effort, it is fair to say that an adequate explanation of the syndrome in functional terms remains as elusive today as it did a century ago. One modern account holds that the patient's impaired reading is due to a generalized disturbance in the perception of multiple forms (i) because of a low level impairment in the extraction of visual features from a horizontal array (Rapp & Caramazza, 1991), (ii) because the system operates with a greatly in-

creased refractory period between the identification of each element (Kinsbourne & Warrington, 1962), or (iii) because of a general inefficiency in the processing of a single visual element that becomes magnified when many elements (i.e., letters) must be identified at once (e.g., Farah, 1990; Friedman & Alexander, 1984).

These arguments have been contested by evidence suggesting that some patients who are letter-by-letter readers may nevertheless show reasonably good perception of random letters strings (Warrington & Shallice, 1980), good enough to raise serious doubts about the plausibility of an account that attempts to reduce the syndrome entirely to a generalized perceptual deficit. While the dissociation reported by Warrington and Shallice has been questioned (e.g., Farah, 1990), we are sympathetic to their claim. We have recently had the opportunity of testing a young letter-by-letter reader (age 26; etiology a left hemorrhagic infarct of the occipital region) whose perception of briefly displayed letters and letter strings now approximates that of age-matched controls but who nevertheless requires between 2 and 3 sec to identify a common four-letter word.

The alternative interpretations of pure alexia that have been proposed in the modern literature bear some resemblance to Dejerine's hypothesis. Patterson and Kay (1982) have opted for a disconnection account, by assuming that the mapping of letter identities onto visual word units has been altered from a fast parallel process to a slow sequential one. Warrington and Shallice (1980) favor a more central deficit to the word form system itself: they suggest that the activation of this component has become seriously impaired, so that words as unitized percepts cannot be derived from their constituent letters. To account for the fact that letter-by-letter readers spell correctly, Warrington and Shallice infer the existence of separate orthographic mechanisms for input and output; thus, the defective perceptual synthesis of visual word forms does not translate into a corresponding failure to retrieve the spelling pattern for written production.

There are now almost as many explanations of pure alexia as there are authors, it would seem, a diversity that has recently prompted the suggestion that there may be many different underlying causes of the phenomenon (Kay & Hanley, 1991) and that to seek a unified account of the syndrome may be futile. We do not share this somewhat depressing assumption, however, if only because none of the contemporary hypotheses is as yet based on anything more substantial than observed co-occurrences between letter-by-letter reading and a variety of other impairments (e.g., slow feature extraction over a horizontal array, reduced visual span, or abnormal object identification) that may well have no direct bearing on the fundamental components of the disorder. We lack a sufficiently detailed theoretical bridge between the striking fact of the disorder—that the patient seems to have lost awareness of even the sim-



plest visual word as a unitized percept—and the underlying processing mechanisms (either general or specific) that are responsible for the transformation of letters into a higher level orthographic or phonological representation.

What can we learn from Dejerine's work in attempting to elucidate the question of pure alexia? For us, the dramatic case of Monsieur C emphasizes a number of issues. For one, he reminds us of the dispiriting fact that we do not fully understand the nature of the constraints on the visual processing of single letters in pure alexia that either render patients unable to identify a letter, in extreme cases like Monsieur C, or more generally increase the time for identification to yield the enormously slow performance that is characteristic of letter-by-letter reading. It is usually assumed that pure alexics who can respond to the identity of a letter—correctly indicate the equivalence between different tokens ( $A = a$ ) of the same letter type—must be doing so by means of the normal visual procedures operating only with a reduction in speed and efficiency. We should be careful about taking this assumption for granted, however, in view of Monsieur C's reported failure and laborious compensatory efforts when attempting the task of identifying letters. Are other pure alexics who do succeed in classifying letters treating them as abstract orthographic elements or have we missed the possibility that the patients have a deficit that fundamentally limits the nature of the representation that is derived from letter forms? It could be the case, for example, that pure alexics must generally treat visual letters as specific tokens linked to a particular shape and that they do not perceive letters in terms of more abstract type identities. If so, a task like matching upper- and lowercase letters, while trivial and direct for the normal reader, would pose considerable difficulty for the pure alexic, although it could still be accomplished if the shape token for a letter were mapped onto a corresponding pronunciation. The equivalence between  $A$  and  $a$  would then be determined only indirectly on the basis of their phonological equivalence (literally, that  $A$  and  $a$  are judged to be equivalent only because the patient can determine they are homophones) and not on the more natural basis of their abstract graphemic unity. In our view, this hypothesis is worth pursuing; we feel it is important to determine exactly how the pure alexic deals with letters as perceptual forms, letters as abstract identities, and letters as words. In the limit, this question boils down to one that we believe Dejerine would have relished given his own theoretical standpoint: How exactly does the pure alexic process the letter  $A$  as a familiar shape having a particular arbitrary name, as an abstract orthographic unit that has more than one form, and as a high-frequency word that represents the indefinite article "a"?

A second question that Dejerine's observations would prompt concerns the issue of letter identification as a specific function dissociable from the

identification of other material like visual objects and more controversially, numerical symbols. The evidence on this point from Monsieur C is rather ambiguous, and the problem has not been adequately dealt with in the modern literature. Is pure alexia category-specific? Given the startling demonstrations of category-specific impairments at the level of object classification and labeling in recent years (e.g., Warrington & Shallice, 1980), we cannot immediately dismiss the possibility that letters have a functional status that is neurologically distinct from other kinds of symbolic material. Curiously enough, Dejerine, for all his insistence that pure word blindness did not affect the perception of numbers, vacillated on the degree of specificity underlying the disorder. Monsieur C forever lost the ability to read music along with his alexia for words and letters. Dejerine appeared quite happy inferring that the relationship was causal, but he made no attempt to explain why numbers but not musical notation should fractionate from letters. In the paper with Pélissier, he indicates that another of his cases with good musical training lost the ability to sight read along with her ability to read words and that after a period of recovery, music that was written in the key of *sol*, but not in the key of *fah*, could be deciphered. His interpretation of this strange dissociation was that the identification of more familiar symbolic forms would recover before less frequent ones, although he again avoids the central theoretical question concerning the range of visual material affected by the disorder.

Finally, what of Dejerine's anatomical explanation for pure alexia, an issue that is particularly relevant in the light of the recent developments in brain imaging techniques like Positron Emission Tomography (PET) and the now burgeoning interest in the neurological bases of cognitive processes? The association between pure alexia and left occipitotemporal damage involving the paraventricular white matter cannot be doubted, although a number of variants on this theme have been uncovered. As for Dejerine's contention that the angular gyrus maintains the visual form of words, we find little convincing support, particularly since it is now clear that many different kinds of damage to the orthographic mechanism (and correspondingly, damage to many different brain regions) may produce a co-occurrence of alexia and agraphia. A widely cited paper by Posner and colleagues (1988), who used PET to measure regional changes in blood flow, finds an area in the left extrastriate cortex that is specifically activated when normal readers passively view single written words. Modern neuropsychological studies that include lesion data further complicate matters, however. The syndrome (or a particular subtype of the syndrome) that would be the best bet, in our opinion, for inferring a central loss of orthographic knowledge (i.e., Dejerine's notion of a visual word center), is now termed surface dyslexia, an acquired or developmental disorder characterized by an approach to reading and spelling that yields many errors when words are encountered that depart from the

conventional mapping between orthography and phonology (Patterson, Marshall, & Coltheart, 1985). A word like STEAK is misread as "steek" based on the usual rendering of EAK (freak, weak, teak, leak, etc.) and written as stake or staik to dictation. It is as if the patient has lost the exact orthography of the word and must resort to generalizations using subword units to approximate its form. A wide range of lesions can produce this reading and spelling impairment, but the most general location is thought to be in the left temporal lobe (Vanier & Caplan, 1989; McCarthy & Warrington, 1990).

Here we end our commentary on Dejerine. We have subjected his interpretation of pure alexia to a rather close scrutiny—it is only fair to wonder what *his* thoughts would have been, if he had been allowed to reflect on the modern work that has been accomplished on the syndrome. Would the current degree of understanding have greatly impressed him or left him somewhat disappointed? We cannot speak for Dejerine, of course, but we do know our present grasp of pure alexia is sufficiently imperfect that we would probably do best to wait a few more years before we speculate on the answer.

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