SANAMED
2013; 8(1): 55–63
ISSN-1452-662X

UDK: 616.831-005.1-06 ; 616.89-008.434
Pregledni rad

POST-STROKE WRITING AND READING DISORDERS

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Primljen/Received 6. 11. 2012. god. Prihvaćen/Accepted 03. 02. 2013. god.

Abstract: The writing and reading disorders in stroke patients (alexias, agraphias and acalculias) are more frequent than verified in routine exam, not only in the less developed but also in large neurological departments.

Alexia is an acquired type of sensory aphasia where damage to the brain causes a patient to lose the ability to read. It is also called word blindness, text blindness or visual aphasia. Alexia refers to an acquired inability to read caused by brain damage and must be distinguished from dyslexia, a developmental abnormality in which the individual is unable to learn to read, and from illiteracy, which reflects a poor educational back-ground. Most aphasics are also alexic, but alexia may occur in the absence of aphasia and may occasionally be the sole disability resulting from specific brain lesions. There are different classifications of alexias. Traditionally, the alexias are divided into three categories: pure alexia with agraphia, pure alexia without agraphia, and alexia associated with aphasia ("aphasic alexia").

Agraphia is defined as the disruption of previously intact writing skills by brain damage. Writing involves several elements — language processing, spelling, visual perception, visual-spatial orientation for graphic symbols, motor planning, and motor control of writing. A disturbance of any of these processes can impair writing. Agraphia may occur by itself or as association with aphasias, alexia, agnosia and apraxia. Agraphia can also result from "peripheral" involvement of the motor act of writing. Like alexia, agraphia must be distinguished from illiteracy, where writing skills were never developed.

Acalculia is a clinical syndrome of acquired deficits in mathematical calculation, either mentally or with paper and pencil. This language disturbances can be classified differently, but there are three principal types of acalculia: acalculia associated with language disturbances, including number paraphasia, number agraphia, or number alexia; acalculia secondary to visual-spatial dysfunction with malalignment of numbers and columns, and a primary anarithmetria entailing disruption of the computation process.

Key words: alexia, agraphia, acalculia, stroke.

INTRODUCTION

The writing and reading disorders in stroke patients (alexia's and agraphias) are more frequent than verified in routine exam, not only in the less developed but also in large neurological departments (1–16). Because reading and writing are aphasic disorders, but one in which the comprehension and production of written language are affected more than spoken language modalities (3, 6).

There are a variety of theories which explain the mechanisms involved in reading and the comprehension of written language. Reading, of course, requires activation of the visual areas in the occipital and occipital and temporal lobes, so that form is perceived, thus revealing that the form is a word. In addition, letters or groups of letters must be recognized and their temporal order ascertained ("orthographic" processing); there must be semantic processing so that the meaning of the word can be derived, and there may be phonological processing so that the sound of the word may be heard within the privacy of one's head. These latter stages of linguistic analysis involve activation of Wernicke's and Broca's area, and the IPL — also demonstrated by functional imaging (3, 16). However, different regions of the brain also interact during various stages of reading, such as in consequence, abnormalities or lesions to different areas can result in different symptoms, such as an inability to recognize sentences or long words, whereas the ability to recognize letters and short words remains intact. Or, an inability to derive semantic mea-

ning from words once they are read. Or an inability to read words, although spelling ability is intact (16). For example, injuries of the left IPL can disrupt the ability to read and spell. Damage to the adjacent but more anterior-lateral supramarginal gyrus can disrupt the ability to spell by sound and to engage in phonological processing-which is one of the most common correlate of reading disability.) Injuries to Wernicke's area can disrupt reading, writing, and all aspects of linguistic comprehension (16). Lesions to the basal (middle/inferior) temporal lobe can disrupt both reading and naming; a condition referred to as phonological alexia and which is also associated with injuries of the supramarginal gyrus. Furthermore, it is showed that phonological dyslexia and dysgraphia may be produced by damage to variety of perisylvian cortical regions which is consistent with distributed network models of phonological processing (3).

ALEXIAS

Alexia (from the Greek ἀ, privative, expressing negation, and λέξις = "word") is an acquired type of sensory aphasia where damage to the brain causes a patient to lose the ability to read. It is also called word blindness, text blindness or visual aphasia. Alexia refers to an acquired inability to read caused by brain damage and must be distinguished from dyslexia, a developmental abnormality in which the individual is unable learning to read, consequently illiteracy, reflects a poor educational back-ground (16, 17).

Most aphasics are also alexic, but alexia may occur in the absence of aphasia and may occasionally be virtually the sole disability resulting from specific brain lesions. There are different classifications of alexias. Traditionally, the alexias are divided into three categories: pure alexia with agraphia, pure alexia without agraphia, and alexia associated with aphasia ("aphasic alexia") (12).

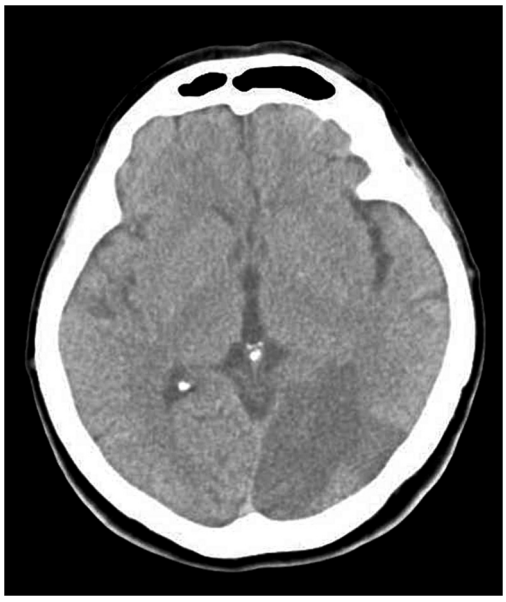

Figure 1. *Ischemic stroke in temporo — occipital lobe (by computed tomography scan) in patients with pure alexia*

More than a century ago, Dejerine described two distinct alexia syndromes — alexia with agraphia and alexia without agraphia. Alexia with agraphia occurred after left-hemisphere parietal damage; alexia without agraphia resulted from occipital damage together with damage to the splenium of the corpus callosum (Figure 1). Since then, several case reports have supported the clinical and neuro-pathological patterns of these two alexias. More recently, a third major alexia syndrome has been proposed, based on an anterior lesion in the left hemisphere; this has been called frontal alexia (19, 20, 21). It should be noted that although the symptoms of each of these alexias are relatively clear-cut, associated clinical findings vary considerably depending on the extent of the lesions and involvement of other areas of the cerebrum (22).

Pure alexia with agraphia

Synonyms for this syndrome include parietal-temporal alexia, angular alexia, central alexia, and semantic alexia. The syndrome of alexia with agraphia, described by French physician Dejerine in 1891, both reading and writing are disrupted with the writing impairment usually equal in severity to the alexia, and without significant dysfunction of other language modalities (17, 18, 19, 20, 21) (Table 1).

Patients display difficulty in comprehending written material that is read silently as well as in reading out loud. Reading of letters and words is impaired, and this difficulty extends to comprehension of numbers and musical notations. The problem with letter identification is not restricted to the visual modality; patients also have problems recognizing words when they are spelled aloud. Parietal-temporal alexia is often associated with fluent paraphasic aphasia (22, 23).

Spontaneous speech:	Fluent, often paraphasic
Naming:	Often impaired
Auditory comprehension:	Intact
Repetition:	Intact
Reading:	Impaired
Writing:	Impaired
Associated signs:	Gerstmann's syndrome, right visual filed defect
Localization:	Left inferior parietal lobule

Table 1*. Features of Pure alexia with agraphia*

Pure alexia without agraphia

Dejerine described the second acquired alexia syndrome, pure alexia without agraphia, in 1892. Synonyms for this syndrome include occipital alexia, pure alexia, posterior alexia, pure word blindness, and letter-by-letter alexia. These patients have no gross aphasia, and they can write, either spontaneously or to dictation. The hallmark of this syndrome is the paradoxical inability of the patients to read words they just wrote.

Alexia without agraphia is easily recognized because it is characterized by a disturbance of reading contrasted with relatively preserved writing skills. Patients typically cannot read what they have just finished writing. The difficulty with letter and word recognition is specific to the visual modality, and patients can spell out aloud and recognize words spelled to them by the examiner.

Spontaneous speech:	Normal
Naming:	Color-naming difficulty
Auditory comprehension:	Intact
Repetition:	Intact
Reading:	Impaired
Writing:	Intact
Associated signs:	Right hemianopsia; short-term memoryloss; occasionally, motor, sensory signs
Localization:	Left occipital lobe, splenium, medial temporal lobe

Table 2. *Characteristics of Pure alexia without agraphia*

Letter naming, although initially slow, improves with practice, and the patients often learn to read the individual letters of the word aloud and then decipher the words from their oral spelling (18, 19, 20, 21, 22). The features of alexia without agraphia are shown in Table 2.

Frontal alexia

In patients with frontal alexia, reading comprehension is typically limited to a few single words, usually content words. Reading comprehension of function words such as prepositions and pronouns is impaired. In contrast to their ability to recognize some words, patients are unable to read the individual letters of the word. Spelling words out loud and comprehension of words that are spelled aloud is also poor. A severe agraphia accompanies the alexia, with writing characterized by poorly formed letters, omission of letters, and grammatically incorrect sentences. Frontal alexia is typically associated with a nonfluent aphasia. Although these traditional neuro-anatomically based distinctions have provided us with a better understanding of the alexias, they do not fully explain the degree of variability seen in patients with alexia and do not permit subtypes of alexias to be distinguished. Interest has therefore shifted from the anatomical correlates of acquired reading disorders to the neurolinguistic and cognitive mechanisms underlying them. In this approach, various theoretical models of reading have been proposed to account for the performance of normal readers and to identify the components of the normal reading system which are disturbed in the alexia syndromes (23).

Aphasic alexia

Many patients with aphasia have associated alexia. Wernicke's aphasia, for example, frequent affects reading. However, in common usage, the term aphasic alexia refers alexia with global or Broca's aphasia (23).

Deep alexia

This syndrome evolves in some aphasics with severe reading impairments in which semantically related paralexias are produced in response to written stimuli. The patient may read "automobile" as "car" or "infant" as "baby". Such reading is thought to be mediated by the right hemisphere on the basis of iconic recognition (12).

Hemialexia

Alexia may occur with hemispheric lesions that produce profound unilateral neglect. The syndrome usually occurs in patients with right hemispheric lesions (Figure 2) and severe hemispatial inattention. The left

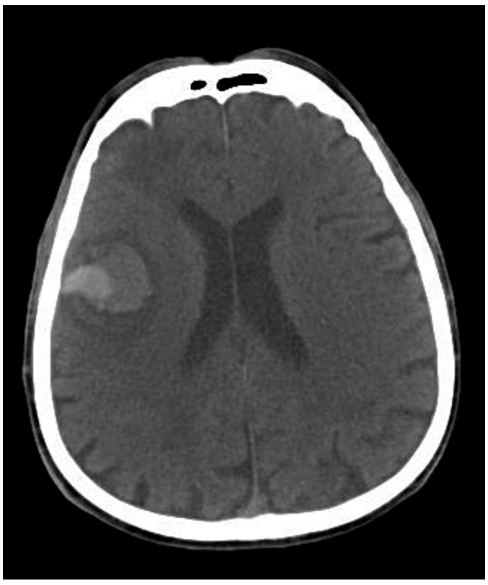

Figure 2. *Intracerebral haematoma in temporo — parietal lobe (by computed tomography scan) in patients with hemispatial alexia*

half of words is ignored so that "northwest" is read as "west" or "basketball" as "ball"; or the left half maybe misjudged so that "navigator" is read as "indicator", "match" as "hatch", or "alligator" as "narrator" (12, 24).

Assessment of reading

Although a brief screening is usually sufficient for identifying the presence of an acquired alexia, detailed assessment is necessary to delineate the precise areas of breakdown so that an effective treatment program can be developed (16, 25). A thorough evaluation includes assessment of reading comprehension to determine the level at which breakdown occurs, an analysis of single word oral reading, and consideration of associated areas of strengths and deficits including visual skills, writing, naming, and spelling (5).

Reading comprehension and level of breakdown

Reading comprehension tasks provide information about the ability to access semantic meaning from print. All general tests of aphasia have a variety of subtests that assess reading comprehension. These subtests are typically hierarchically organized, beginning with recognition of letters, matching letters written in different forms (e.g., uppercase, lower case, script), and letter naming. Testing then progresses to the single word level (e.g., recognizing highly familiar words such as name, name of town, country; matching single words to pictures), the sentence level (e.g., following written commands), and finally to more complex paragraphs (e.g., answering questions about a paragraph that has just been read). However, reading subtests on aphasia batteries such as the Western Aphasia Battery (28) and the Boston Diagnostic Aphasia Examination (29) may have too few items on some tasks and may be insufficient to detect milder problems (30).

A specific test of reading comprehension that has been developed for individuals with aphasia is the Reading Comprehension Battery for Aphasia (2nd Ed.) (RCBA-2) (31). Test includes single word comprehension tasks in which a single picture must be matched to one of three words that are orthographically, phonologically, or semantically similar. Sentence comprehension is assessed by having the patient select one of three pictures that correspond to a written sentence. One subtest assesses specifically morphosyntactic reading and another subtest assesses short paragraph comprehension. Longer paragraph comprehension is evaluated with factual and inferential questions (26).

Functional reading of labels and signs is also included. This second edition of the RCBA also includes a lexical decision task in which patients choose a real word from a triad of two pseudowords and one real one. Several tests of reading comprehension provide grade levels and age equivalencies and may be appropriate for individuals with acquired alexia. Such tests include the Gates-MacGinitie Reading Tests (27), the Woodcock Reading Mastery Tests (28), and the Nelson-Denny Reading Test (29).

Treatment

Traditional approaches to the treatment of acquired reading problems typically begin at the level of breakdown, that is, at the grapheme, word, phrase, sentence, or paragraph level, and patients practice tasks that are arranged hierarchically. Some commonly used treatment tasks include: letter matching; word–picture matching; word––word matching in which the target may be the category name, an antonym, or a synonym; word–definition matching; phrase or sentence completion; following written commands; and answering yes/no or "way" questions about a sentence or paragraph. Difficulty is modified by a change in various parameters such as the degree of similarity between the target and distracters; the number of distracters in the field; the frequency, grammatical class, concreteness of the words; or the complexity of the grammatical structure.

Most commercially available workbooks for aphasia have numerous pages of pencil and paper exercises, and more recently a number of computer programs also have been developed to provide practice on these types of reading activities. However, despite their clinical widespread use, these reading tasks and programs for aphasia have not been carefully evaluated and there is little evidence supporting their efficacy.

One randomized clinical trial evaluated the efficacy of computerized treatment in general by using hierarchically organized reading activities consistent with the traditional approach (30). Fifty-five patients with chronic aphasia were randomly assigned to one of three conditions: computer reading treatment that consisted of visual matching and reading comprehension tasks, computer stimulation such as nonverbal games and cognitive rehabilitation tasks, or no treatment. Patients in the computer groups used the computers 3 hours a week for 26 weeks. The results suggested that computerized reading treatment was efficacious with improvements generalizing to non-computer language performance. It was also shown that these improvements resulted from the language content of the software and not from the stimulation provided by the computer (21).

AGRAPHIAS

Agraphia is defined as the disruption of previously intact writing skills by brain damage. Writing involves several elements — language processing, spell-

ing, visual perception, visual-spatial orientation for graphic symbols, motor planning, and motor control of writing. A disturbance of any of these processes can impair writing. Agraphia may occur by itself or in association with aphasias, alexia, agnosia and apraxia. Agraphia can also result from "peripheral" involvement of the motor act of writing (17, 23). Like alexia, agraphia must be distinguished from illiteracy, where writing skills were never developed (31).

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There is several classification of agraphia. First, writing disorders can be classified by the underlying cognitive deficits: aphasic agraphia, apraxic agraphia, and spatial agraphia. In addition, "pure agraphia" indicates the absence of any other language or cognitive disorder.

Agraphia with fluent aphasia
Agraphia with nonfluent aphasia
Alexia with agraphia
Gertsmann's syndrome agraphia
Pure agraphia
Agraphia in confusional states
Deep agraphia
Disconnection agraphia
Praxic agraphia

Table 3. *Aphasic agraphias*

Another way of classifying agraphias is to divide writing into its component psycholinguistic steps and to analyze writing disorders according to the specific step that is disrupted, as in the classification of agraphias, we first distinguish between "central" agraphia, resulting from disorders of central language processing, versus "peripheral" agraphia, resulting from disorders of the motor aspect of writing. Central agraphias thus affect lexical (word choice), semantic (word meaning), and phonological processes, after which a "graphemic" (written) version of the word is generated. Peripheral portion of writing involves selection of the proper letter string and the motor output to write it (23).

From practical point of view agraphias can be divided into two categories: aphasic and non-aphasic agraphias. Aphasic agraphias include: agraphia with fluent aphasia; agraphia with non-fluent aphasia; alexia with agraphia, Gertsmann's syndrome agraphia; pure agraphia (Figure 3); agraphia in confusional states; deep agraphia; disconnection agraphia; apraxic agraphia (Table 3). Non-aphasic agraphias are: motor agraphia; reiterative agraphia; visuospatial agraphia and hysterical agraphia (Table 4) (12).

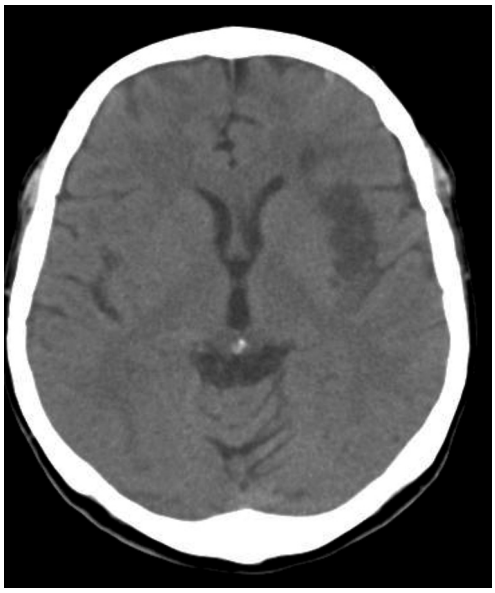

Figure 3. *Ischemic stroke in fronto-temporal lobe (by computed tomography scan) in patients with pure agraphia*

Aphasic agraphias

For aphasic patients, writing is often most severely impaired language modality. In aphasic patients, written language typically mirrors spoken language expression, and in some cases the language abnormalities may be more marked in written than spoken language. Thus, in non-fluent aphasias such as Broca's, writing resembles speech: brief, effortful, and lacking in

<u>Motor agraphia</u>
Paretic agraphia
Hypokinetic agraphia:
Micrographia with parkinsonism
Hyperkinetic agraphia:
Tremor
Chorea, athetosis, tics
Dystonia (writer's cramp)
<u>Reiterative agraphia</u>
Perservation
Paligraphia
Echographia
Coprographia
<u>Visuospatial agraphia</u>
<u>Hysterical agraphia</u>

Table 4. *Nonaphasic agraphias*

syntax; there is sparse graphic output, with clumsy calligraphy, agrammatism, and poor spelling. The fluent aphasias, especially Werincke's aphasia, also produce fluent errors in writing (have a normal quantity of well-formed letters, but with a lack of substantive words and insertion of literal, verbal, or neologistic paragraphias similar to oral paraphasias), and spelling errors are sensitive of mild deficits (12, 23).

Deep agraphia

Deep agraphia parallels deep alexia involving writing rather than reading. A patients have difficulty spelling non-words, deficits in spelling certain classes of words; can write words with concrete, imageable meanings better than those with abstract meanings and semantic words (nouns and verbs) better than syntatic words (propositions and conjunctions). Errors may involve semantically related words, such as "chair" for "desk". Lesions generally involve the left parietal region, often including the supramarginal gyrus or insula but sparing the angular gyrus (23).

Gertsmann's syndrome and angular gyrus syndrome

In 1924, Josef Gertsman described a syndrome occurring with discrete left angular gyrus lesions and consisting of tetrad of clinical findings including agraphia, finger agnosia, inability to distinguish left from right, and aclaculia (12). In 1940, Gertsmann reviewed the considerable literature that had evolved concerning the syndrome and concluded that the finding had clinical validity and localizing value (32).

The four elements of the Gertsmann's syndrome do not necessarily all occur together; any combination of three items would indicate a left inferior parietal lesion, and other, related deficits including alexia and mild aphasia may be combined (32).

Nonaphasic agraphia

Writing depends on a complex array of motor and visuospatial skills in addition to language abilities.

Motor agraphias. Disruption of any aspect of the motor system-peripheral, corticospinal, extrapyramidal, cerebellar-will produce agraphia, and in each case the muscles, peripheral nerves, or corticospinal tracts produce a clumsy, uncoordinated agraphia secondary to limb paralysis. Micrographia is a common manifestation of Parkinsonism, and is characterized by a progressive diminution in the size of the letters, often accompained by increased crowding. Action tremors of either the cerebral or postural type produce disturbances in writing an may make written productions unintelligable. Chorea, athetosis and tics are hyperkinetic movement disorders that influence writing in the same way that they affect other voluntary motor activity. In severe cases writing is impossible, and even in mild cases the output will be visibly distorted. "Writer's cramp" is among the most well-known and most misunderstood of all agraphias. The syndrome of progressive cramping of the hand and forearm among individuals in professions demanding fine finger movements, including writers, telegraphers, pianists, and violinists, was treated as a neurotic disorder in past but today it is well-known focal dystonia.

Reiterative agraphias refer to the abnormal repetition of letters, words, or phrases in writing. Perseveration is a continuation of activity after the appropriate stimulus has stopped. Paligraphia is the rewriting of phrases generated by the patient. Echographia is the rewriting of phrases produced by the examiner.

Visuospatial agraphia is manifested by a tendency to neglect one portion of the writing page, slanting of the lines upward or downward, and abnormal spacing between letters, syllables or words.

Hysterical agraphya. Agraphia may occasionally occur as a hysterical conversion symptom. The agraphia is usually part of a monoparesis in which the limb is weak throughout with slightly diminished tone and normal muscle stretch reflexes. Sensation may or may not be affected (12).

ACALCULIAS

Acalculia is a clinical syndrome of acquired deficits in mathematical calculation, either mentally or with paper and pencil (12, 24, 34). This language disturbances can be classified differently, but there are three principal types of acaculia: acalculia ssociated wirh laguage disturbances, including number paraphasia, number agraphia, or number alexia; acalculia secondary to visuospatial dysfunction with malalinment of numbers an columns, and a primary anarithmetria entailing disruption of the computation process (Table 5) (12, 35, 36).

Aphasia-related disturbances of calculation include paraphasic errors in which the patient makes a

Aphasia-related acalculias
Number paraphasia
Number alexia
Number agraphia
Visuospatial acalculia
Primary anarithmetria
Symbol agnosia

Table 5. *Classification of the acalculia*

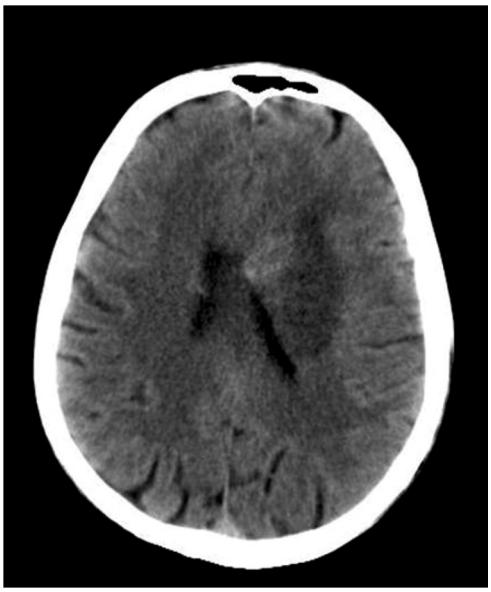

Figure 4. *Ischemic stroke in parietal lobe (by computed tomography scan) in patients with transcortical motor aphasia, alexia, agraphia and acalculia*

verbal paraphasic error, substituting one number for another. Number alexa and number agraphia may also occur and, in some cases, may be disproportionately greater than letter reading and writing disturbances. Acalculia occurs with nearly all aphasias but is more severe in patients with lesions of the posterior aspect of the left hemisphere involving the parietal cortex (Figure 4) (12).

Visuospatial acalculia may occur with lesions of either hemisphere but is most common with right paretal dysfunction. Spacing of multidignit numbers, place-holding values, and column alignment are disrupted.

Primary anarithmetria occurs mainly in the context of Gerstmann's syndrome with lesion in the region of dominant angular gyrus, but it may occasionally be seen as an isolated abnormality with disturbances of the same region. It is acquired neurological deficit in mathematical thinking, which not involve problem in reading and writing of numbers or the spatial arrangement of numbers (12).

CONCLUSION

Post-stroke language disorders are very complex in their clinical phenomenology and in the classifications. Speech disorders of aphasic type, alexia, agraphia and acalculia, in neurology, first of all in acute phase of stroke are more frequent disorders then it verified in routine exam, not only in the less developed but also in large neurological departments. The most important aspect of language disorders is the necessity of testing these functions to detect deficits. Language deficits are important in practical localization of brain lesions and they impair patient's ability to function in the world. Regardless of the fact that a significant number of these disorders spontaneously improves, it is necessary to start the treatment as soon as possible. Early treatment of language disorders is important not only because of speech recovery, which is crucial for everyday communication, but also because of the entire rehabilitation of patients with neurological symptoms accompanying speech impairment. Rehabilitation of motoric impairments is more complex and slower in persons with these syndromes, especially if the patient is not treated by parallel rehabilitation of neurological deficit and speech impairments.

Sažetak

JEZIČKI POREMEĆAJI NAKON MOŽDANOG UDARA

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Jezički poremećaji nakon moždanog udara su česti i uključuju afaziju, aleksiju, agrafiju i akalkuliju.

Poremećaji pisanja i čitanja kod pacijenata sa moždanim udarom (aleksija, agrafija i akalkulija) su mnogo češći nego što se to u rutinskom neurološkom pregledu konstatuje, ne samo u manje razvijenim nego i u velikim neurološkim odjelima, u svojoj suštini su to afazički poremećaji i neretko su više oštećeni nego govorni jezik.

Aleksija je stečeni tip senzorne afazije gde oštećenje mozga uzrokuje gubitak mogućnosti, od ranije naučenog čitanja. Naziva se još slepoća za reči, slepoća za tekst ili vizualna afazija. Treba je razlikovati od disleksije, razvojnog poremećaja gde osoba nije u stanju da nauči čitati, te od nepismenosti koja reflektuje siromašnu obrazovnu pozadinu. Većina afazičara su aleksični, ali aleksija može biti prisutna i u odsustvu afazije, a može biti prisutna u nekim slučajevima nakon speci-

fičnog oštećenja mozga i kao izolovani poremećaj. Postoje različite klasifikacije aleksija. Tradicionalno, aleksije se dele u tri kategorije: čista aleksija sa agrafijom, čista aleksija bez agrafije i aleksije udružene sa afazijama ("afazičke aleksije").

Agrafija se definiše kao oštećenje ranije naučene veštine pisanja, nastalo nakon oštećenja mozga. Pisanje uključuje nekoliko elemenata — jezičko procesuiranje, speling, vizualnu percepciju, vizualno-prostornu orijentaciju za grafičke simbole, motorno planiranje i motornu kontrolu pisanja. Oštećenje bilo kojeg od ovih procesa može oštetiti pisanje. Agrafija se može pojaviti kao izolovani poremećaj ili udružena sa afazijom, aleksijom, agnozijom i apraksijom. Može takođe biti i posledica zahvatanja "perifernog" akta pisanja. Slično aleksiji, agrafija se mora razlikovati od nepismenosti kada veština pisanja nije ni razvijena.

Akalkulija je klinički sindrom stečenog deficita matematičke kalkulacije, ili na mentalnom planu ili sa „papirom i olovkom". Ova jezička oštećenja mogu biti različito klasifikovana, ali u biti postoje tri glavna tipa akalkulija: akalkulija udružena sa jezičkim poremećajima uključujući parafaziju, agrafiju brojeva, aleksiju brojeva; akalkuliju sekundarnu na vizuoprostornu disfunkciju sa nepravilnim položajem brojeva ili kolumni i primarnu anaritmetriju sa „čistim" poremećajem računanja.

Ključne reči: aleksija, agrafija, akalkulija, moždani udar.

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