Evaluation of cognitive and behavioral disorders in the stroke unit

Asaid Khateb,1 Jean-Marie Annoni,1 Ursula Lopez,1 Françoise Bernasconi,1 Laurent Lavanchy,1 and Julien Bogousslavsky2

1Geneva University Hospitals, Geneva, Switzerland
2Swiss Medical Network, Montreux, Switzerland

Introduction

The general aim of a clinical neuropsychological examination is to assess language, memory, attention, adaptive behavior, motivation, and emotion impairments that result from a brain dysfunction. This behavioral and cognitive evaluation can be performed during the first hours following cerebral damage either in a classical bedside approach or by means of standardized tests and may thus help establish a precise diagnostic. In the hyperacute or acute phase of stroke, neuropsychological intervention may also be necessary to establish communication with the patient (e.g. in the case of aphasia) or to set up adequate strategies to be used by healthcare providers (e.g. in the case of spatial neglect). In this context, the patient is most often tested while lying in bed and in the presence of other patients in the same room. Due to this context, as well as other disturbing factors, the evaluation of cognitive—behavioral deficits in the stroke unit is generally difficult, of relatively short duration, and must thus be repeated in order to correctly track the clinical evolution and to advise when the symptoms become more stable. Moreover, effective rehabilitation of cognitive deficits often relies on modular neuropsychological models (e.g. Pilgrim and Humphreys, 1994), which are often incompatible with clinical fluctuations and global dysfunction such as confusional states, frequently found in the acute phase. So, actual neuropsychological rehabilitation does not rely on acute evaluation.

In this chapter, we will first emphasize the difficulties in undertaking systematic neurocognitive evaluations in the stroke unit. We will also highlight the necessity of tracking the evolution of cognitive and behavioral disorders, since they constitute important predictors of stroke outcome. We will then briefly recall some common neuropsychological disorders that occur after focal cortical damage and will describe the clinical approach in the stroke unit to the disorders most frequently found following left hemisphere, right hemisphere, or posterior strokes.
Difficulty in evaluating cognitive functions in the stroke unit

Neuropsychological literature very often concentrates on post-acute or chronic cognitive impairments which persist after three months following stroke onset. Studies investigating cognitive deficits during the hyperacute or the acute phase of stroke (i.e. the first week, 1–4 days, or 1–7 days) generally refer to small cohort descriptive studies or to case reports that illustrate representative or unusual symptoms. Despite the fact that such clinical observations are of outstanding interest, detailed neuropsychological and/or behavioral evaluations remain less systematic than studies on chronic patients. At three months post-stroke, about 60% of patients are estimated to suffer from major or residual deficits in at least one cognitive domain (Pohjasvaara et al., 1998). In the stroke unit, the difficulty in obtaining systematic and reproducible evaluations during the acute phase is in part due to the concurrent intensive medical care, which does not always allow for easy and careful analysis. Due to this, but also to other reasons, cognitive disorders are often either overlooked or underestimated in acute stroke (Merino and Heilman, 2003). Of these other reasons, the following two appear as the most important to consider:

1. The attitude of the patients towards their stroke and the resulting cognitive deficits is distorted by false beliefs (Croquelois and Bogousslavsky, 2004). More than half of the patients do not initially recognize their symptoms as a stroke (Meijer et al., 2003). Acute stroke is an active injury to the “self” that makes the patient underestimate the ongoing changes in the sensory-motor interactions with both the internal and external world, and thus leads the patient to a certain degree of anosognosia (Vuilleumier, 2004), and consequently to an underestimation of cognitive impairments by the emergency care-givers. With respect to the importance of mental alterations that can occur during the hyperacute phase of the stroke, it has been observed that at least one-third of the patients have no, or have only a poor, memory of the events of the first 24 hours (Grotta and Bratina, 1995), and this independently of the affected hemisphere. Whenever therapeutical decisions must be applied, this point has to be carefully examined in order to determine whether or not the patient’s judgment and decision-making capacities are altered (Bogousslavsky, 2003; Hacke et al., 2000).

2. The second major reason making it difficult to conduct systematic evaluations in the stroke unit is the fact that the hyperacute and the acute phases represent unstable conditions during which symptoms can change very rapidly (Croquelois and Bogousslavsky, 2004). In this context, the major causes of fluctuations are the evolution of the ischemic penumbra in the surrounding
Evaluation of cognitive and behavioral disorders in the stroke unit

territories and the regression of the cerebral diaschisis that may involve areas distant from the infarct (e.g. Cappa et al., 1997). These unstable conditions do not allow for the adequate testing of anatomo-functional hypotheses as can be undertaken in stabilized patients, and reduce the efficiency of possible neurorehabilitation interventions.

The necessity of steadily evaluating cognitive functions in acute stroke

Despite the fact that systematic cognitive evaluations may encounter considerable difficulties in the stroke unit, there are at least two main reasons that strongly motivate such evaluation during the first days.

1. First, the effect of therapeutic intervention on cognitive deficits can be underscored when assessed by the global stroke scales. For example, Hillis et al. (2003) have shown that the National Institute of Health Stroke Scale (NIHSS) is not reliable in assessing stroke severity and outcome because of the lack of items assessing the neuropsychological aspects of the deficits. The authors showed that while perfusion indices did not correlate with NIHSS scores, these measures correlated in acute right hemisphere stroke with spatial neglect scores and with aphasia scores in left hemisphere strokes. In line with this observation, previous studies have suggested that aphasia (Wade et al., 1986) and neglect (Kalra et al., 1997) might have a direct impact on stroke outcome.

2. Second, increasing evidence in animal studies and clinical settings suggests that early therapy is associated with a better long-term outcome in both sensory-motor and cognitive functions. For instance, animal models have recently shown that motor recovery could be maximally enhanced by training, and negatively affected by limiting beam-walking experience during early recovery periods (Brown et al., 2004). These findings raise questions about the role of early neuropsychological intervention in enhancing cognitive recovery after stroke, like, for instance, motor impairment (Crisostomo et al., 1988). However, since cognitive functioning is an important predictor for the successful treatment and rehabilitation of stroke patients, it is therefore mandatory to assess cognitive status during the acute period with a clinical bedside examination (Paolucci et al., 1996). Under the practical constraints of an acute stroke unit ward, such a screening should be repeatedly performed with little technical equipment in a reasonable time, and only at a subsequent juncture can a formal assessment be undertaken to specify the persistent cognitive—behavioral deficits.
Cognitive–behavioral disorders in the stroke unit

Neuropsychological syndromes or cognitive–behavioral disorders in the stroke unit constitute the clinical manifestations of the acute dysfunction of complex and largely distributed neural systems that subserve cognition and adaptive behavior. In the acute phase, these clinical manifestations depend not only on the infarct area, but also on other transient factors such as the diaschisis, the ischemic penumbra, and intracerebral pressure that can be induced by hemorrhagic strokes. The most common cortical neuropsychological syndromes in the stroke unit are aphasia and spatial neglect that result respectively from left and right hemisphere damage (Croquelois et al., 2003; Pedersen et al., 1995). To a lesser extent amnesia, the different forms of visual agnosia (apperceptive, associative, prosopagnosia, color agnosia, and pure alexia), and Bálint’s syndrome are encountered following damage to the posterior areas (Ferro, 2001). Less localized disorders include disorientation, abulia, and delirium (or confusional syndrome) (Ferro, 2001). Delirium is particularly frequent in older patients with medical complications but also in hemorrhagic strokes where it can be associated with neglect (Caeiro et al., 2004). With respect to behavioral disorders, these most frequently include overt sadness, disinhibition, lack of adaptation, crying, anosognosia, anosodiaphoria (or lack of concern), passivity, and aggressiveness (Ghika-Schmid et al., 1999). Overt sadness and crying are more frequently associated with left hemisphere lesions. In right brain-damaged patients, anosognosia and anosodiaphoria are frequently associated with neglect. Paradoxically, anosognosia may also co-exist with severe sadness. Finally, although of a very low incidence, catastrophic reaction constitutes a particular emotional modification in the acute phase (Carota et al., 2001). Its high association with aphasia and left insular lesions suggests that this emotional disturbance is induced by damage to a specific neural module.

Clinical approach of right hemispheric strokes at different steps of evolution:

The example of spatial neglect

The most frequent deficit following right hemispheric strokes is the so-called “syndrome of contralesional neglect” or hemi-spatial neglect. This syndrome denotes the impaired or lost ability to react to, or to process, sensory stimuli (visual, auditory, somato-sensory, or olfactory) presented in the hemi-space opposite to the lesion side (e.g. Kerkhoff, 2001). Most frequently, spatial neglect occurs following right hemisphere (RH) damage that involves parietal, temporoparietal, frontal, limbic, and subcortical areas (Husain and Rorden, 2003; Mesulam, 1999). At the cortical level, these lesions include the angular and supramarginal gyri of the inferior parietal lobe, the temporoparietal junction,
5 Evaluation of cognitive and behavioral disorders in the stroke unit

the superior temporal gyrus, and the inferior and middle frontal gyri (Parton et al., 2004).

Although neglect is classically related to RH lesions, the occurrence of this phenomenon is more diffuse in the hyperacute phase. Actually, it has been suggested that, within the first three days following a stroke, neglect could be found with quasi-equal frequency in patients with right and left lesions (72% vs. 62%) (Stone et al., 1991). Nevertheless, it is more severe following right strokes and resolves more frequently in patients with left strokes. At two months post-stroke, visuo-spatial neglect could still be observed in 40% of right-brain-damaged patients (Robertson and Halligan, 1998).

Behavioral assessment in daily life is often the most sensitive initial evaluation of spatial neglect (Azouvi et al., 2002). In the hyper and acute phases, inability to cross the midline with the eyes or to explore left space is the typical feature. With right parietal damage, the patients may have head and eyes turned to the extreme right and never orient to the left spontaneously and thus pay attention only to items situated to their right (such as dinner or newspaper). When approached from their left, they may orient themselves to the right and respond with gaze directed away from the speaking person (Parton et al., 2004). This particular behavior can even be increased on repetitive stimulation, so that the patient tries to turn over by more than 180 degrees to the right (Ghika et al., 1995). Assessing the severity of the trouble and associated symptoms allows for tracking of the clinical evolution of the patient in the stroke unit. Clinical evaluation must thus initially rely on questions and simple tests. Psychometric testing, although not easy to undertake in acute stroke, can be conducted using “paper and pencil” tests (e.g. line bisection). Letter cancellation (Halligan et al., 1990) and star cancellation tests (Halligan et al., 1991) are some of the most sensitive tests and allow for the identification of up to 74% of neglect patients. Drawing (e.g. the sun, or a clock), copying pictures situated in near space, or describing a picture situated in far space will give other information about different modalities of neglect. Starting the description of a picture on the extreme right, as well as omitting the contralesional part of a drawing, represent positive signs of neglect. Reading sentences and composite words (i.e. rainbow, water lily, seat belt) may reveal spatial dyslexia when the left parts of words or text are omitted. In the same way, spatial dysgraphia is revealed when written words are far away from the contralesional paper’s margin. Clinical evaluation may also concern motor impairments (motor neglect) and imagined spatial scenes (representational neglect). Questions based on the patient’s or the examiner’s body parts (“whose hand is it?”) will give information on hemiasomatognosia or even on alien hand, often associated with neglect (Motomura et al., 1988). Visual extinction as well as auditory and tactile extinction must also be clinically tested (De Renzi et al., 1989).
After a few days, further assessment of neglect implies evaluation of the various aspects of spatial cognition. Hemineglect can affect the allocentric space (that refers to the spatial relationship between two stimuli separated in the space) and the egocentric space (which is related to the patient’s own body or specific body parts, see Hillis et al., 2005) differently. Neglect can also affect the diverse subdivisions of the subjective near and far space, commonly referred to as body space (personal), reaching space (peripersonal), and extrapersonal space (far space or walking space) differently. With respect to far space, representational neglect may be tested, relying on a memory description of a public place (Bisiach and Luzzatti, 1978). A memory description of the inside of a car while the patient is successively seated in the conductor’s and the passenger’s seat also allows for testing of representational neglect in the near space (Ortigue et al., 2003).

Rehabilitation of spatial neglect can be undertaken in the acute phase (Bowen et al., 2002; Pierce and Buxbaum, 2002). However, patients are usually not receptive or unable to collaborate enough for the neuropsychologist to start a full rehabilitation. Usually, clinicians have to work with the patient in order to go beyond anosognosia. Depending on the patient’s complaints, spatial representation can be slightly improved in the hyperacute or acute phase. Trying to attract the patient’s gaze from his ipsilesional to his contralesional visual space (i.e. asking the patient to describe left-cued pictures can help him to pay attention to neglected space), asking the patient to move his contralesional body parts (in the left or right mid-sagittal plan), or attracting the patient’s attention by loudly speaking in his ipsilesional and then contralesional space (by moving around the bed) can be done without use of particular materials.

Clinical approach to left hemispheric strokes: The example of aphasia

Aphasia, which refers to an acquired deficit of receptive/expressive language abilities in oral and/or written codes, is considered the most important cognitive impairment that occurs following left hemisphere strokes. This is observed in nearly 40% of all strokes, (Pedersen et al., 1995) and constitutes an important predictor of persistent disability, particularly if the initial form is global and associated with hemiplegia, hemisensory loss, hemianopia, and reduced consciousness (Heinsius et al., 1998). During the acute phase, the most important factors for aphasia recovery are the initial severity of the symptoms and the size and location of the infarct (Basso, 1992; Kertesz, 1988; Pedersen et al., 1995), but also the presence of other associated cognitive deficits (Pashek and Holland, 1988). Stationary language functions are reached after 2–10 weeks, depending on the severity of aphasia (Pedersen et al., 1995). The frequency of aphasic symptoms decreases to about 20–30% after two weeks, of which one half is considered moderate to severe (Engelter et al., 2004).
When a patient shows communication difficulties, the objective of the first clinical evaluation within the stroke unit is to help in establishing a differential diagnostic. Actually, it is important to determine if these difficulties are due to aphasia, dysarthria, confusional syndrome, locked-in syndrome, akinetic mutism, or other speech-associated disorders. This differential diagnosis is particularly important, since acute aphasia is often severe, either global or unclassified in 50% of cases (Godefroy et al., 2002). On the basis of the neurological clinical examination only, a portion of these disorders cannot be adequately diagnosed during the first days (Trapl et al., 2004). Therefore, clinical examination in the stroke unit should be rapidly accompanied by standardized simple aphasiological tests, so that every patient can be correctly treated as a function of the posited diagnostic and its severity.

Depending on the context facilities (i.e. patient in the room with others, in bed, in an armchair) and the general state of the patient, the clinical evaluation can be divided into several sessions. In severe cases, the sessions may last less than 10 minutes. The oral production will be limited to very automated spontaneous productions (answers to questions, use of “yes/no” series, singing), to the repetition of simple words or isolated phonemes and to the naming of objects belonging to different lexical fields (window, key, bottle, clock...). Word fluency tasks and propositional language are usually impossible at this level. The oral comprehension can be investigated using closed questions (i.e. that can be answered by yes or no), designation in the room, and execution of increasingly complex orders. Instead of reading, the patient can be asked to associate written words to an object on a forced-choice basis. Spelling should be tested with particular attention, since patients who have not already noticed a deficit at this level can show catastrophic reactions or refuse to take a pencil with their left hand in case of right hemiplegia.

At this level and up to the stabilization of the symptoms, the decision to set up systematic speech therapy cannot be taken. However, it is inconceivable to leave the patient without any linguistic stimulation, given the importance of early training (Aichner et al., 2002). The therapist thus proposes a follow-up that aims to maximally engage the patient’s residual abilities. Informing the patient about improvement, even if very modest, leads to a better awareness and subsequently to a better adaptation to the handicap. An important part of the intervention will also be centered on the awareness of the deficits, mainly when considering that the persistence of language difficulties will constitute a major problem for professional reintegration. The therapist’s intervention with the family and the nursing personnel is also important in the acute phase, with special attention to the attitudes that care-givers and family members have to avoid. With regard to the patient’s family, the nature of the disorder has to be clearly explicated in order
to facilitate exchanges and prevent misinterpretations. Furthermore, the patient’s relatives must be informed that the difficulties are not necessarily short-lasting and that they are not related simply to tiredness or to lack of motivation.

Clinical approach to posterior strokes: The example of visual agnosia

Strokes involving the posterior cerebral artery can induce brainstem, cerebellar, thalamic, and occipital dysfunctions. Cognitive deficits may prevent resuming occupational activities, even in infratentorial strokes (Aichner et al., 2002), and thus necessitate an attentive screening. Moreover, vascular damage involving the thalamus or intrahemispheric white matter pathways can lead to severe cognitive deficits that meet the criteria of a particular form of vascular dementia (Auchus et al., 2002). However, in cortical posterior infarcts, cognitive deficits related to visual integration processes are the most frequent. Clinical bedside examination of patients with posterior strokes aims first to uncover specific disorders of visuo-cognitive functions that can be dissociated from other sensorial and cognitive disturbances, such as language or memory. Of these, visual disorders such as agnosias or dyschromatopsia appear as a difficulty to recognize respectively visual stimuli and colors in the absence of any elementary disturbance of perception or intellect. However, agnosic deficits are not always easy to demonstrate in the acute phase due to the importance of visual field defects (such as hemianopia, quadranopia, hemiachromatopsia, cortical blindness, etc.), and to the presence of other perceptive—cognitive symptoms such as visual hallucinations, neglect, dysphasia, or memory disorders (Cals et al., 2002). Acute clinical evaluation, using simple tests (asking the patient about their visual acuity, naming objects presented in the visual, tactile, and auditory modalities, copying objects, drawings, etc), should be conducted to specify the nature of the agnosia. An example of the importance of acute repeated evaluations is suggested by the difficulty in obtaining a consistent picture of Anton’s syndrome. That syndrome, the denial of blindness by patients who are clinically unable to see (Argenta and Morgan, 1998), has often been found in the very acute phase of cortical blindness after eclampsia and is accompanied by confabulations or excuses for their symptoms (“there is not enough light to see”). This syndrome is less frequent in systematic sub-acute evaluations (Argenta and Morgan, 1998), suggesting that cortical blindness and related symptoms evolve rapidly during the first days. Moreover, follow-up evaluations suggest that cortically blind patients later develop visual agnosia, or may partially recover blind sight, perception of color, movement, or selective perception of emotional stimuli (Aldrich et al., 1987; Pegna et al., 2005) but the pattern of such recovery is not clear.

Patients with apperceptive visual agnosia are generally aware of their impairment: they spontaneously complain about it and try to use hands in an
attempt to recognize what they see (Landis et al., 1982). Despite the relative preservation of elementary visual functions, such patients have difficulties in recognizing visually presented objects, whereas the same objects can be correctly identified through the other sensorial channels (e.g. touch or hearing) (Grossman et al., 1997). In contrast, patients with associative agnosia, who are generally unaware of their difficulty, will not complain about it, and will adopt a normal visual behavior without attempting to explore objects using the tactile modality. Clinically, they are able to correctly describe (or copy) a visually presented item despite their failure to recognize it.

The evolution of these visual deficits can take the form of more specific agnosic impairment. Prosopagnosia refers to the failure to recognize previously known faces and occurs more frequently after right hemisphere posterior stroke. Since some patients are aware of the deficit and can spontaneously describe their troubles, the easiest way to identify the problem consists in asking them whether or not they have difficulties in recognizing familiar people. The crucial test consists of confronting the patient with several people (which should be very similar for various criterions, such as stature, dressing . . . ), one of who ought to be known by them (Rentschler et al., 1994).

The counterpart of prosopagnosia but following left posterior lesions is pure alexia or agnostic alexia that concerns the visual and not the language system per se (Grüsser and Landis, 1991). This reading impairment, which occurs without agraphia or other language impairments, is due to a lesion in the inferior occipito-temporal cortex (Déjerine, 1892). Pure alexia has been thought to result from damage that affects the visual word-form (Cohen et al., 2000; Warrington and Shallice, 1980). Clinically, in the most severe forms the patients are not able to recognize isolated letters and indicate whether the letters “a” and “A” represent the same grapheme (Miozzo and Caramazza, 1998). In moderate cases, recognition errors may depend on the configuration complexity of the letters (e.g. I and O vs. G and H), on the difficulty in discriminating between the graphic features of the letters (O vs. Q), and of the phonemic similarity between the letters (M vs. N, or P vs. B).

Cognitive evaluation of color vision following posterior strokes aims at specifying whether a given deficit is related to achromatopsia, color agnosia, or color anomia. Since these different deficits can result from impairment at different levels of color processing, clinical screening should be undertaken according to three successive steps (Gil, 1996). The first step consists of exploring the perceptive stage with, for example, a matching color tokens test. The second step should evaluate the associative level using tests such as coloring objects, matching color/objects (i.e. red/cherry). In the third step, visuo-verbal tests, like naming the color of objects, can simply be used. According to this schema, it is
possible to decide if color impairment is due to achromatopsia (impairment of the first stage), color agnosia (deficit at the second and third stage), or color anomia (impairment of the third stage only). Finally, akinetopsia or motion blindness is an often-misdiagnosed symptom in the acute phase (Blanke et al., 2003). All these specific syndromes are difficult, but important to detect in a stroke unit, to improve assessment, follow-up, rehabilitation, and information to the family.

There are few data about acute intervention in visual impairment. A cognitive–behavioral assessment in the acute phase is, however, mandatory as cognitive impairment is a predictor of poor functional outcome in stroke survivors. Patients with deficits should then have neuropsychological support in order, for example, to reduce their anosognosia, and later on to fully specify their residual deficits (for example, it is important in the case of visual agnosia to determine which visual processes are impaired). Later, after an extensive neuropsychological investigation, therapy targeting the impaired processes will be designed (Burns, 2004).

Conclusion

This short introduction has emphasized the importance of cognitive assessment in acute stroke, despite ongoing modifications to the clinical picture and the difficulty in obtaining reliable and clear responses. Patients with cognitive deficits should first benefit from neuropsychological support (if no contraindications are detected such as acute confusional state), and later from a more extensive assessment with a neuropsychological rehabilitation program oriented to their specific deficits. The evaluation, clinical and bedside in a first attempt, and inspired by cognitive models in a second step, is important in determining severity, evolution, and prognosis in the stroke unit. Ultimately, the questions that have to be answered by a cognitive evaluation in the acute phase are:

1. Is the clinical picture due to a single or multiple infarcts?
2. Is there a specific or a global cognitive impairment or a confusional state?
3. What is the pattern of stabilization or recovery during the first week?
4. Which therapy programs should be conducted at a given time, specific or more global?

REFERENCES
