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Spatial Neglect and Anosognosia After Right Brain Stroke

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Abstract

PURPOSE OF REVIEW: Up to 80% of survivors of right brain stroke leave acute care without being diagnosed with a major invisible disability. Studies indicate that a generic cognitive neurologic evaluation does not reliably detect spatial neglect, nor does it identify unawareness of deficit after right brain stroke; this article reviews the symptoms, clinical presentation, and management of these two cognitive disorders occurring after right brain stroke.

RECENT FINDINGS: Stroke and occupational therapy practice guidelines stress a quality standard for spatial neglect assessment and treatment to reduce adverse outcomes for patients, their families, and society. Neurologists may attribute poor outcomes associated with spatial neglect to stroke severity. However, people with spatial neglect are half as likely to return to home and community, have one-third the community mobility, and require 3 times as much caregiver supervision compared with similar stroke survivors. Multiple randomized trials support a feasible first-line rehabilitation approach for spatial neglect: prism adaptation therapy; more than 20 studies reported that this treatment improves daily life independence. Evidence-based treatment of anosognosia is not as developed; however, treatment for this problem is also available.

SUMMARY: This article guides neurologists' assessment of right brain cognitive disorders and describes how to efficiently assemble and direct a treatment team to address spatial neglect and unawareness of deficit.

INTRODUCTION

Up to 80% of survivors of right brain stroke are not diagnosed with cognitive disorders of spatial cognition or awareness[1,2] during routine acute stroke care, and thus, they cannot receive a plan for personalized cognitive treatment. Worse, right brain stroke itself is underdiagnosed, so survivors of right brain stroke are at risk of undertreatment or incorrect treatment[3–5] because of the difficulty in identifying the cognitive hallmarks of right brain disorders during routine care.

This article reviews the symptoms and clinical presentation of two of the cognitive syndromes occurring after right brain stroke. They are not rare; probably about half or

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even up to 60% to 80% of survivors of right brain stroke have these symptoms for the first weeks and months after stroke (TABLE 6-1[6–15]).[11,14,16]

Cognitive symptoms after right brain stroke are important to neurologic care for two major reasons. First, they may be the only indication of a stroke event or right brain dysfunction (eg, seizure) requiring medical care. Thus, for example, detecting cognitive symptoms after right brain stroke may allow the clinician to give acute stroke treatment to more patients and improve community outcomes and quality of life. Second, by identifying cognitive symptoms of the spatial neglect syndrome, defined as asymmetric reporting, responding, or orienting to one side of space after a brain lesion, causing functional disability, one can provide management, education, and treatment.[17] Assessment of spatial neglect and rehabilitation of this cognitive disorder, which can improve daily life function and independence, are also discussed. Anosognosia, or pathologic unawareness of deficit, is also covered in this article.[18] Although both spatial neglect and anosognosia may occur after other forms of focal brain injury (eg, after brain tumor resection), they have only been studied in systematic ways in the context of right brain stroke. Thus, clinical practice guidelines for the assessment and treatment of spatial neglect are only available for stroke care.[19] People with anosognosia may be pathologically unaware of several different kinds of deficits; however, anosognosia for left body paralysis is very strongly associated with right brain injury.[20] Although rehabilitation approaches for anosognosia are not as well-researched as those for spatial neglect, evidence shows they may improve deficits and so this article describes how these treatments can be used. Neurologists need to advocate for patients with right brain syndromes to ensure quality care and good outcomes.

Right brain stroke is a major identified cause of spatial neglect.[21] Spatial neglect symptoms may occur in other disorders associated with focal brain injury, such as traumatic brain injury and brain tumor, and spatial neglect symptoms may even occur in multiple sclerosis and Parkinson disease.[22–25] However, in contrast to the high-quality evidence available supporting the use of spatial neglect assessment and treatment in quality stroke care,[26] no prospective, systematic studies of the clinical presentation and treatment of spatial neglect in these disorders have been performed. Thus, the evidence is not yet appropriate to use as a basis for planning routine clinical care in these areas. This is an important area for future research.

SPATIAL NEGLECT

Clinically identifying spatial neglect is important; a challenge exists in diagnosing this problem, and routine care processes are insufficient to identify this disorder.

Clinical Presentation

Spatial neglect is a common disorder, occurring in about half of patients with stroke during the first months after their event (TABLE 6-1). The unsafe behaviors observed as symptoms of spatial neglect can be misattributed to intellectual, reasoning, personality, or generic cognitive problems, which can be devastating to patient dignity. Misattribution of spatial neglect symptoms can also delay diagnosis of right brain stroke past the window for acute

stroke interventions; thus, clinicians should be alert to new spatial bias in patients with stroke risk factors.

SPATIAL NEGLECT SYMPTOMS.—Considerable inequity exists in management and outcomes between strokes based on which side of the brain is affected. Although cognitive symptoms caused by right brain stroke are characteristic and disabling, clinicians frequently miss them and, thus, fail to offer appropriate treatment.[27] This is probably true in other disorders that cause focal brain damage and spatial neglect (eg, brain tumor); however, formal examination of underdiagnosis of spatial neglect and its impact on the management of right versus left brain issues have not been studied in other brain disorders.

The most common reason why the symptoms are missed is that they are not assessed. An acute stroke assessment tool, the National Institutes of Health Stroke Scale, which is a quality care standard in stroke centers,[28] underestimates the severity of right brain stroke[29] because, on this 47-point scale, only 2 points (for item 11: involves testing extinction to bilateral simultaneous stimulation, see below) are specific to right brain stroke and spatial neglect, compared with 7 points total (for items 1b, 1c, and 9: answering questions, obeying a command, and aphasia testing) that are sensitive to detect aphasia after left brain stroke.

How to Assess for Spatial Neglect

This section describes the principles and testing methods used to approach accurate evaluation for spatial neglect.

NOT A VISION PROBLEM.—One of the reasons that neurologists underestimate right brain stroke symptoms is that they have often been informed that spatial neglect is a higher-order visual problem.[30] However, people with vision problems do not behave like people with spatial neglect. People with vision problems report that they cannot recognize objects or that they struggle with reading. Although people with spatial neglect may have trouble reading the left side of sentences or words (neglect dyslexia), which is disabling, they may not seem at all concerned about their partial reading. People with vision issues report they cannot see well. However, spatial neglect is characteristically accompanied by unawareness of deficit (anosognosia), and the overwhelming majority of patients with spatial neglect deny having visual issues. Unawareness of deficit (anosognosia) is reviewed later in this article.

EXTINCTION TO BILATERAL SIMULTANEOUS STIMULATION.—Frequently, spatial neglect occurs in patients who do not have hemianopia. Patients may be able to see single stimuli on the contralesional side of space; however, when a stimulus is presented in both visual fields (bilateral simultaneous stimulation), especially in the lower visual fields, patients with spatial neglect after right brain stroke may confidently respond that the stimulus is on the right only (ie, extinction to double simultaneous stimulation). Extinction can occur in the visual, tactile, and auditory modalities. Clinicians should bear in mind that extinction to double simultaneous stimulation is not always present in 100% of trials. In many patients, a deficit of “Where” perceptual attention causes unawareness of some stimuli on the left side, and yet not 100% of stimuli are extinguished in the presence of a right-sided

stimulus: perhaps only 40%, 60%, or 80% of left-sided stimuli are extinguished. Because extinction of the left-sided stimulus may be present only some of the time, examiners need to do enough trials (at least six or eight simultaneous stimulation trials) to capture errors; of six or eight trials, even one or two errors in which only the ipsilesional side stimulus is reported raises suspicion for spatial neglect.

Extinction to double simultaneous stimulation that is detected in two modalities is strongly suggestive of spatial neglect. To test tactile extinction, the examiner touches the patient on both hands, arms, knees, legs, or feet and asks where the stimulus was felt. If a single stimulus can be felt on the side of the body suspected to be contralesional, but the contralesional stimulus is not felt during double stimulation of both sides of the body, extinction is present. If a patient has extinction in two modalities (visual and tactile, for example), this helps clarify that the problem is spatial neglect and not a sensory issue because, clearly, a visual deficit should not, for example, cause errors in detecting a tactile stimulus.

EXTINCTION OR SPATIAL NEGLECT? OR ANOTHER TERM?—Clinicians will sometimes use three or four terms to describe spatial neglect symptoms. They may separate extinction from spatial neglect, and they may use other terms such as *hemiinattention*. Although this may be interesting when looking at laboratory-based analysis, this author discourages this approach in clinical practice. With the goal of increasing the clinical utility of spatial neglect diagnosis, this author and colleagues defined spatial neglect as pathologically asymmetric behavior caused by a brain lesion, resulting in functional disability.[31] Thus, many symptoms can all be called part of the spatial neglect syndrome. Patients who do not have a visual field cut may yet collide with a door frame; thus, with extinction to double simultaneous stimulation, they can meet criteria for spatial neglect because it appears to be the basis of functional disability (a collision and safety risk) (CASE 6-1). Here, the extinction is a feature of, or sign within, the spatial neglect syndrome. The evidence of difficulty with safety while walking should, additionally, be enough to trigger physical therapy evaluation for fall risk, as well as gait and balance training.

“AIMING” SPATIAL NEGLECT: MOTOR-RELATED SYMPTOMS.—Another reason why clinicians can miss spatial neglect if they think of it as a higher-order visual problem is that it can manifest with only maladaptive spatial movements (eg, disinclination to move in one direction with the eyes, head, arms, or whole body [TABLE 6-2]). Patients may have head and eye deviation and spontaneous rotation rightward after a right brain lesion, as in CASE 6-2 (FIGURE 6-1[34]). Even when lying in bed, they may rotate, positioning their bodies crookedly, lying unevenly and pushing their bodies farther to one side until one leg hangs off. These asymmetric movements are well-established as a primary manifestation of spatial neglect[17] and may actually be associated with a better response to spatial retraining during rehabilitation.[35] Maladaptive and spatially asymmetric movements may be limb-specific. The arm opposite the side of a stroke may not be particularly weak; however, it may not move well or may demonstrate poor persistence of movement.[36]

Asymmetric gaze and movements (eg, in ambulation) are integral components of Aiming spatial neglect as it is observed during functional performance. As illustrated in CASE 6-2

and FIGURE 6-1, Aiming spatial neglect can cause an abnormal, persistent ipsilesional turning tendency that affects the eye, head, and body. The abnormally asymmetric posture caused by Aiming spatial neglect may explain the greatly increased risk of falls in people with spatial neglect, which is at least 50% higher and may be as much as 5 times higher than in survivors of stroke who do not have spatial neglect with attendant increased risk of accidental injury.[37,38] Aiming spatial neglect also causes rightward bias in environmental movements; affected survivors of stroke with right brain stroke and Aiming spatial neglect can have a right-turning bias, as they move in the home, in the neighborhood, and even when driving (CASE 6-2).

Identifying Aiming Spatial Neglect

Unfortunately, spatial neglect has been traditionally regarded as a visual problem, and thus, no method of bedside screening has been established for neurologists to use to reliably identify Aiming spatial neglect. The abnormalities that can be caused by Aiming spatial neglect after right brain stroke do occur independently of perceptual-attentional Where spatial neglect; thus, survivors of stroke with Aiming spatial neglect may have intact awareness and ability to detect stimuli in both sides of space.

Aiming spatial neglect at the bedside can be identified when patients have marked leaning, veering, or postural rotation to the ipsilesional side (FIGURE 6-1), although contralesional leaning (“pushing”) can sometimes be observed. The “hanging eyeglasses” sign shown in FIGURE 6-2 may be seen; it is caused by hypometric leftward movement of the hand when placing the eyeglasses on the head. Key components of Aiming spatial neglect are also described in TABLE 6-2.

Visual extinction to double simultaneous stimulation may indicate the presence of Where spatial neglect because of dysfunctional perceptual-attentional spatial processing. However, paper-and-pencil tasks, such as bisecting a line (CASE 6-2) or marking all the lines scattered on a piece of paper (the Albert line cancellation task[39]), seem to require both Where and Aiming spatial skills. Therefore, the clinician can use these paper-and-pencil tests to help with screening anyone suspected of having spatial neglect.[40]

Screening for extinction to double simultaneous stimulation and for abnormal performance on paper-and-pencil tests is not sufficient to evaluate Aiming spatial neglect, however. Some survivors of stroke have only the body movement and postural abnormalities that are characteristic of Aiming spatial neglect (TABLE 6-2) without demonstrating any abnormalities on paper-and-pencil screening.[41] Because these spatial movement abnormalities are not assessed in any of the available bedside tests such as the National Institutes of Health Stroke Scale or paper-and-pencil tests and are likely responsible for motor disability, fall risk, and other limitations that affect function and freedom in spatial neglect, an assessment is needed that examines movement performance. Thus, in any survivor of stroke suspected of having spatial neglect, this author recommends referral to an occupational or physical therapist who can screen for functional performance errors with a standardized measure.

The Catherine Bergego Scale,[42] which has been validated in many medical and rehabilitation settings, is one measure used by those professionals that is strongly recommended to detect spatial movement abnormalities that affect daily life function and safety. The test captures asymmetric functional body movements and predicts daily life disability related to spatial neglect.[6] Because of the time required to administer the scale and its ease in combining with a functional assessment in therapy care, a therapist rather than a neurologist is most likely to use this scale. If performed as a completely separate evaluation, the Catherine Bergego Scale takes about 15 to 20 minutes. If it is combined with a clinically standard therapy assessment of activities of daily living, the Catherine Bergego Scale takes only about 5 minutes of documentation time in addition to the therapy assessment. It also requires the examiner to be reliability-trained at observing and scoring spatial performance in patients while they actually perform daily life tasks.

"WHERE" REPRESENTATIONAL FUNCTION IN SPATIAL NEGLECT.—Spatial cognition entails an intermediate information processing stage between Where perceptual-attention and Aiming motor-intention in which explicit and implicit spatial representations (ie, knowledge in the form of imagery, maps, and internal descriptions) are required. These can be visual-spatial, auditory-spatial, and even spatial-somesthetic representations, and any of these may be impaired in spatial neglect. Although disordered spatial representations may sound like a theoretical problem, this issue has functional consequences. In the modern world, we need internal maps while we are working, completing our social activities, and even interacting with groups and navigating crowded environments. The survivors of stroke who have an isolated deficit of Where spatial neglect affecting representational function may have trouble drawing or interpreting maps or charts and may be perceived as confused, intellectually impaired, or perseverative when the problem is a loss of spatial knowledge.

At the bedside, one sign of representational spatial neglect is right-sided bias affecting the way we use a mental number line.[43] Mathematical operations rely on our ability to spatially map numeric information in our minds; the left side of these operations may be distorted when people have Where representational spatial neglect. To test this ability, the examiner can ask the patient to "tell me what number is halfway between 11 and 55." Giving two numbers that require little analysis to calculate a midpoint (eg, 10 and 30) should be avoided. When administering this test to a survivor of stroke whose educational level and general ability to perform intellectually demanding tasks are appropriate for this testing of mental mathematics (eg, a college professor), the examiner may be surprised by a "rightward" biased response (eg, 50).

Summary of Spatial Neglect Subtypes

Although the daily life performance errors made by people with spatial neglect are often interpreted as being exclusively related to perceptual or attentional problems, many functional impairments and limitations on daily life activities and participation also result from Where, representational problems, or Aiming spatial neglect. For example, because people with directional hypokinesia have difficulty initiating leftward movement, they may find themselves unable to dress on the left or may steer a wheelchair rightward just as the patient in CASE 6-2 steered his truck with exclusively right turns. TABLE

6-3[44–46] summarizes some of the potential relationships between spatial neglect subtypes and observed daily life limitations.

Trends

This section describes treatment that is now established to be effective in reducing daily life disability for people with spatial neglect. These protocols have been demonstrated to be effective in stroke; further research in spatial neglect associated with other disorders is needed.

REHABILITATION OF SPATIAL NEGLECT.—Although the Centers for Disease Control and Prevention reported that 30% to 35% of survivors of stroke receive rehabilitation after leaving acute hospital care in many states,[47] the National Institute of Neurological Disorders and Stroke estimates that about twice as many would benefit from rehabilitation.[48] Neurologists frequently find that survivors of stroke who received meticulous attention to highest-quality standards of care in the first hours and days after their stroke received little or no rehabilitation once they left acute settings. This treatment gap needs to be closed for all patients with spatial neglect, and, in particular, for survivors of stroke with the spatial neglect syndrome.

At least 10% of people who have spatial neglect acutely continue to have chronic symptoms.[49] As discussed earlier, ipsilesional bias can be observed in people with spatial neglect. It is not uncommon to observe that, years after stroke, a patient may have trouble eating from the left side of a plate (CASE 6-3). This is not only interesting to observe, it is pragmatically important. The Intercollegiate Stroke Working Party[50] in the United Kingdom warns that people with stroke and spatial neglect should "be monitored to ensure that they do not eat too little through missing food on one side of the plate." Patients with spatial neglect after right brain damage may fail to dress the left side of their body or may favor right space when navigating a wheelchair. This limits their independence, but it also has a devastating effect on their dignity.

The American Heart Association[19] and American Occupational Therapy Association,[51] as well as stroke organizations worldwide, recommend spatial neglect treatment as part of quality stroke care. Neurologists cannot usually administer spatial neglect treatment in their offices; however, the neurologists' efforts are very important in assembling and directing a treatment team to administer rehabilitation. Evidence on which professional guidelines and recommendations are based is not yet available for other neurologic disorders associated with spatial neglect. Further research in this area is needed.

PRACTICE-BASED IMPROVEMENT.—Guideline-based early rehabilitation protocols for spatial neglect are available. Keeping this information for reference is an area for practice-based improvement.

Practice-based improvement for spatial neglect also requires the neurologist to digest information about personalized care (spatial neglect subtypes, noted earlier) because spatial neglect is undiagnosed in patients who receive no formal assessment or who have Aiming spatial neglect symptoms. However, a second reason, beyond the need to diagnose the

presence of spatial neglect, is that personalized care is important. Just as different kinds of cardiac disease (eg, arrhythmia, heart failure, ischemia) should be prioritized and addressed differently in a treatment plan, patients with Aiming spatial neglect symptoms[35] may be especially likely to improve after early spatial retraining (see TABLE 6-4[52–57] for treatment options).

Aiming spatial neglect may predict an excellent response to treatment as compared with Where spatial neglect.[58] The brain mechanisms explaining this observation are under research; however, this may be related to frontal lobe disconnection in spatial function. Different neuroanatomic regions might play different roles in the networks supporting Where versus Aiming spatial cognitive processing, with posterior, temporal-parietal cortical sensory association networks more critical in supporting information input (Where spatial neglect) or storage (Where, representational spatial neglect[59]) whereas anterior or motor-related subcortical networks are more critical to output cognition (Aiming spatial neglect[17]).

Treatment

Because it is supported by both the American Heart Association and American Occupational Therapy Association and because it is a low-risk, 10-day protocol that therapists can learn to use in a few days, prism adaptation therapy is appropriate as a first-line treatment. This author recommends that neurologists refer survivors of stroke with spatial neglect, especially if Aiming spatial neglect is suspected, for this treatment protocol. FIGURE 6-3[60] illustrates improvement in daily life function after prism adaptation therapy, in multiple studies; although the impact of treatment varied in different studies, better-designed studies showed stronger benefit, supporting the value of this treatment approach.

During prism adaptation therapy for left-sided spatial neglect,[61] patients wear left-based, yoked optical prisms, which shift what they see rightward. They then make multiple goal-directed hand movements for a prescribed session of about 20 to 30 minutes. Although in randomized clinical trials of this approach some parameters of the treatment varied, in one standard regimen, patients wore 20-diopter wedge prisms and completed 10 sessions over 14 days.[58] Patients wear prisms only during treatment sessions; at other times, they may engage as usual in other activities or rehabilitation.

FINDING A TREATMENT TEAM.—A neurologist may be tempted to delegate all of the tasks of evaluation and treatment for rehabilitation to the therapist with whom care is being shared. However, as evidence-based protocols of treatment emerge, it is very helpful for a neurologist to share the responsibility with the therapy clinicians to ensure that patients receive quality care. When neurologists work with a partner organization to whom they send stroke therapy referrals, it is very helpful to meet with this partner, project the need for quality standard care, estimate the potential volume of referrals, and describe the benefit in improved outcomes, providing information about how therapists may need to be trained or what equipment may be needed. In the experience of this author, many therapists or therapy business administrators have busy day-to-day experiences that have prevented them from researching new treatment options. However, they are often willing to consider how their

organization can offer a new treatment protocol, which can distinguish their therapists as experts, drive referrals, and help engage their staff and enhance staff retention.

DIRECTING SPATIAL NEGLECT TREATMENT PROTOCOLS.—In the past, allied health professionals received little support from neurologists about how to plan or execute rehabilitation care. Fortunately, most therapy clinicians are very resourceful and can make treatment decisions independently. However, focus group studies indicate, consistent with this author's personal experience, that evidence-based treatments recommended by professional associations are not being selected by therapists in this kind of independent consultation partnership.[62] Therapists may have learned about spatial neglect during training and may feel more comfortable with continuing to use other treatment approaches (eg, self-reminders to "look left," or prism exposure, in which patients wear prisms without making visually guided movements).

To be successful in leading a spatial neglect treatment team, the neurologist should take a collaborative, decisive leadership role. Unlike the isolated practitioner of the past, the modern neurologist "leads and facilitates clinical teams, builds tools to implement new therapies, and communicates the vision and purpose of team-based care."[63] Because most neurologists do not directly employ or supervise speech-language pathologists or occupational or physical therapists, this means that the neurologist should use interpersonal leadership skills (persuasion and influence) to motivate and engage the treatment team and help the team examine and continuously improve performance. In a 2018 article, this author and colleagues in the American Academy of Neurology Transforming Leaders Program[63] described this vision for the 21st century: neurologist-led team-care pathways are "essential to achieving the triple aim … improving functional outcomes, cost-effectiveness, and the patient experience. The care pathway from diagnosis to treatment for spatial neglect, using prism adaptation, traverses this new territory…"

UNAWARENESS OF DEFICIT (ANOSOGNOSIA) AFTER RIGHT BRAIN STROKE

Anosognosia means, literally, "without knowledge of disease." People with neurologic disorders can demonstrate reduced awareness, or complete disavowal, of disabling consequences of brain dysfunction; notably, after right brain stroke, patients can appear unaware of left hemiparesis.[64,65]

Clinical Presentation

Anosognosia for hemiparesis has long been identified as a sign of right brain stroke,[66] especially when it is moderate to severe.[67] However, people with anosognosia after right brain stroke can also be unaware of visual and other sensory disturbance[67] and other problems such as memory and cognitive deficits and gait disorder.[18]

Unawareness of deficit is a major barrier to receiving health care, although formal studies of this relationship are not available. Numerous anecdotal reports suggest that unawareness of deficits after right brain stroke prevents patients from presenting promptly

for stroke evaluation. Many patients continue to drive under unsafe circumstances after having a stroke, unaware of their signs and symptoms. For example, a patient may have a stroke caused by the effect of recreational stimulant use on preexisting hypertension and cerebrovascular disease, lose control of the car while driving, and have an accident. Anosognosia after right brain stroke also interferes with reporting other medical symptoms that require immediate attention. In one report, a patient who was previously able to report angina reliably lost awareness of this left-body symptom after a stroke.[68]

The inability to recognize functional limitations poses a significant safety risk, is associated with falls,[69] and is known to be a major barrier to effective rehabilitation.[70] Anosognosia can take several forms, with different degrees of disavowal of the neurologic deficit (TABLE 6-5[71]). It can be associated with a loss of the feeling of ownership of an impaired body part (asomatognosia) or a distorted experience of an impaired body part (somatoparaphrenia) and, at the far end of the continuum, a dislike or hatred of a dysfunctional body part (misoplegia).[72] Although misoplegia is uncommon, it can cause dramatic behavioral problems, such as patients throwing themselves from their beds “in order to get rid of that awful leg.” Although classic studies support an association of right brain dysfunction with anosognosia for hemiparesis,[65] awareness networks for motor and cognitive function are complex and still being investigated.[73]

How to Assess for Anosognosia

Formal assessment of awareness of deficit is very important because of its potential impact on the outcomes of survivors of stroke and their caregivers.[74] It is also easy to miss anosognosia for hemiparesis when patients have an amotivational state, are saying little, or have depression or decreased engagement. When a survivor of stroke is aware of other symptoms, the neurologist may incorrectly assume that the patient is aware of hemiparesis. Thus, this author recommends that neurologists always assess awareness of hemiparesis as part of acute care of stroke or right brain injury. People with right brain stroke and anosognosia for hemiparesis experience the same range of emotions as control subjects.[75] In many of these people, awareness of the disabling consequences of their stroke and hemiparesis may be implicit (unconscious). This means they may not be able to articulate the awareness or their distress.[76] Thus, they may even be at higher risk of depression than other patients with right brain stroke. It also means that we should not assume survivors of stroke are aware of deficits just because they have dysthymia or depression.

Formal screening for anosognosia for hemiparesis is described in CASE 6-4 and can be completed in just a few seconds. Although with this quick formal screening it is not possible to identify anosognosia that causes relatively small-magnitude overestimation of self-performance, large-magnitude inconsistencies between self-rating and performance are regularly identified by using this method.

Some patients “know what to say” when asked about their deficits after stroke; however, they do not seem concerned about this information (anosodiaphoria) (TABLE 6-5)[71]; this represents a form of anosognosia. They may report, for example, “Everyone tells me that my arm is weak.”

It is very helpful to communicate with other clinicians (nurses, therapists, social workers) and with caregivers about the anosognosia assessment because many people confuse this disorder with psychological denial or think that it is protective from the emotional impact of having a brain disorder. The vertical line rating is a convenient way of introducing the message that this problem is a reality distortion caused by the brain injury. Neurologists can, for example, show the vertical line as in CASE 6-4 and communicate that the patient actually experiences left arm strength as being normal. Researchers specifically examining whether anosognosia protects patients from depression found no evidence that it does.[78] In the past, when members of the care team assumed that anosognosia was protective to the patient, efforts to provide cognitive rehabilitation were reduced, and this would be expected to increase disability.

Trends

This section reviews the need for rehabilitation of anosognosia after right brain stroke and approaches to this rehabilitation.

REHABILITATION OF ANOSOGNOSIA AFTER RIGHT BRAIN STROKE.—It is, first, extremely important for the neurologist to understand that studies very clearly demonstrate that patients with right brain disorders and anosognosia still benefit from intensive multidisciplinary rehabilitation, such as inpatient rehabilitation hospitalization. Second, specific approaches have been developed to improve anosognosia in stroke and traumatic brain injury. Even though randomized controlled trials of these treatments to improve anosognosia for hemiparesis after stroke are not yet available, several treatment approaches have been associated with improved rehabilitation outcome benefits in open-label studies, including self-awareness training,[79] spatial neglect interventions such as vestibular stimulation,[80] and self-observation via video feedback.[81] As described earlier, many therapy clinicians delivering stand-alone care may not implement specific anosognosia treatment because they are familiar with using techniques based on expert wisdom, such as reminders, caregiver counseling, or management for safety. However, for best-quality care, neurologists may want to ensure that their patients receive specific treatment for anosognosia, which may often require an interprofessional program that administers particular behavioral protocols. This is yet another justification for considering inpatient rehabilitation if a survivor of stroke is otherwise eligible.

Such a therapy team includes neuropsychology, speech-language pathology, and occupational and physical therapy, and clinicians may have certification as “brain injury specialists.” Frequently, clinicians cross-apply approaches to remediate awareness, approaches that were developed in traumatic brain injury settings, to survivors of stroke or people with brain tumors or other focal brain disorders who are enrolled in therapy.

CONCLUSION

In this article, two major cognitive syndromes resulting from right brain injury, spatial neglect and anosognosia, were reviewed. As stroke centers begin to evaluate cognitive assessment with standardized measures during inpatient stroke care, it is likely that more and more people with these disorders after stroke will be diagnosed and that neurologists

will play a greater role in leading a treatment team to address these important invisible disabilities. Unfortunately, not much evidence is available at present to guide assessment and treatment of spatial neglect due to other focal brain disorders, and this is an appropriate topic for future research.

The diagnosis and treatment of spatial neglect and the feasibility and utility of using prism adaptation treatment as a first-line therapy with a rehabilitation team were also reviewed in this article. Although randomized controlled trials and professional guideline recommendations strongly support specific identification and treatment of poststroke spatial neglect, neurologists can help this move forward.

For unawareness of deficit, neurologists can play a vital role in counseling caregivers and other clinicians about the crucial distinction between anosognosia and psychological denial and steering the patient to an interdisciplinary setting where specific protocols of care for anosognosia are used. Both stroke and traumatic brain injury are associated with anosognosia to left hemiparesis. In a broader sense, this author is concerned that anosognosia presents a public health barrier to allocation of resources to people with right brain dysfunction. People who have experienced a right brain injury may be unable to report their significant disability, leading to falsely inflated reports of good quality of life or well-being. It is also concerning how anosognosia affects access to health care and rehabilitation. In recent years, patients with unawareness of deficit have been systematically limited from admission to inpatient rehabilitation or other intensive treatments. However, if patient-reported outcomes become the main path of entry to rehabilitation care, it is up to the neurology community to assure access to inpatient rehabilitation and other forms of intensive therapy for our patients who cannot self-advocate.[82]

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CASE 6-1

An 80-year-old woman who resided in an assisted living facility reported having heartburn one morning. The symptoms were gone a few hours later; however, she got out of bed and started moving around the facility, going into the rooms of three other residents and insisting they were in her space and needed to leave. The nurses could not redirect her; she kept breaking away and going into other resident rooms. The geriatrician on staff found her only mildly tachycardic, possibly dehydrated, and unable to tell him the day of the week or date, although she knew the month and year. She insisted she was in her room, although she was in the cafeteria lounge. About an hour after her symptoms started, the geriatrician diagnosed delirium, calling the patient's behavior "impulsive," but no medical condition could be identified on screening laboratory tests, ECG, and urine evaluation. Her past medical history was notable for hypertension, high cholesterol, and a possible past episode of atrial fibrillation.

Her family insisted on an assessment from a neurologist 2 days later. The neurologist found her irritable and saying "I'm fine. I want to go home." Her gaze drifted to the right when not stimulated (right gaze preference), although she easily looked leftward when the examiner wiggled fingers on the left or told her to look leftward. She had no visual field cut to single stimuli; however, she made errors about half the time when tested with double simultaneous stimulation in the left and right lower visual fields, responding "right." She also sometimes responded before the examiner showed a stimulus or when the examiner was not actually showing her anything, reporting she saw a stimulus on the left. The examiner gave her a pamphlet to read, and she started in the middle of the page, read the right side of several sentences in the first paragraph, then tossed the pamphlet aside, angrily saying "This is stupid." She had no weakness; however, when drift was tested, her left palm rose, and she veered when walking. Sometimes she veered leftward, and she collided with the door frame as she left the examination room; however, within the examination room and when trying to find a chair, she moved in a tight, rightward circle, looking for the seat that was directly in front of her as she entered: she finally found it by nearly tripping over it. The neurologist ordered a brain MRI, which revealed a right parietal stroke; the geriatrician was much surprised, saying "she had no symptoms, just delirium."

COMMENT

Spatial neglect symptoms can be misclassified or misattributed to problems with concentration or motivation. Losing orientation in space, in a patient who does not have an obvious hemiparesis, can be mistaken for a problem of continuous attention. A patient can have both problems; spatial neglect is a strong risk factor for developing delirium after stroke,[32] either because cognitive problems increase delirium vulnerability or because right brain systems play a role in delirium development.[33]

Further, this patient not only failed to report a left-sided visual stimulus when one was simultaneously present on the right side, she also sometimes falsely reported a left-sided stimulus when none was there. Higher visual processing that occurs after stimuli is processed in the occipital lobes occurs in two parallel association cortex pathways in

which dysfunctions can lead to agnosias. The dorsal pathway extends to the parietal lobe and processes location and motion and is therefore known as the *Where pathway*. The ventral pathway extends to the temporal lobe and processes shape, form, and color and is therefore known as the *What pathway*. This case is consistent with Where spatial neglect, in which distorted perceptual-attentional input lowers the resolution of sensory perception and thus reduces the accuracy of simple stimulus detection. Patients will make false-positive as well as false-negative (omission) errors.

CASE 6-2

A 45-year-old man was brought to the emergency department by his employer because the employer noted that the patient suddenly “can’t turn left.” His employer found that the patient, a food service delivery driver, had made only four deliveries after a full day of driving and was behaving strangely. The global positioning system (GPS) records in his vehicle revealed he had been making deliveries by making exclusively right turns. The patient showed how he operated the steering wheel using only his right hand, and when asked why he was not using his left hand, he said it “is lazy” and “won’t work right.”

On examination, he had head and eye deviation to the right; when the examiner moved the patient’s head to the right, his eyes moved about 5 degrees conjugately leftward, supporting a cortical cause of his gaze deviation. He had left pronator drift and reduced left grip strength; however, during most of the examination, his left arm hung motionless, as if it were plegic. When asked to bisect a long line drawn on a piece of printer paper, he made a mark about 7.6 cm (3 in) to the right of the actual line center. While he was sitting on the gurney in the emergency department, he leaned to the right, and although when urged, he could sit straight again, his posture quickly returned to an asymmetrical lean; it looked like this postural bias could cause him to slip off the gurney. A nurse found him on the floor about 1 hour later. When the team reviewed the patient’s brain imaging, they found he had a right-sided putaminal ischemic stroke with some extension into subcortical white matter.

COMMENT

Accurate line bisection cannot rule out spatial neglect, but a survivor of stroke who errs more than 1 cm (0.4 in) rightward in bisecting a horizontal line longer than 22 cm (8.7 in) is highly likely to have spatial neglect.

CASE 6-3

A 62-year-old man presented to a neurologist after his wife noticed that he ate only from the right side of his plate and was falling frequently. He had a right middle cerebral artery ischemic stroke 1 year before.

On examination, he had a mild left hemiparesis. Screening for extinction in the visual and tactile modalities and errors on line bisection and cancellation tasks revealed chronic spatial neglect. The patient had received acute stroke care at a comprehensive stroke center and had sought care from tertiary-care specialists at the local academic medical center for his cardiology and internal medicine needs. He had also received nursing and some kind of therapy (unspecified in his records; possibly physical therapy) through home health for the first few weeks after the stroke to address left-sided weakness; however, since the home health therapy ended, the patient and his wife reported that he received no specific outpatient therapy and no therapy targeted to improve spatial neglect.

COMMENT

The classic teaching to clinicians was that spatial neglect invariably resolves spontaneously. However, 10% or more of survivors of stroke have chronic spatial neglect years after a stroke.[49]

The Centers for Disease Control and Prevention reports that more than 60% of survivors of stroke receive absolutely no outpatient rehabilitation.[47] Referring patients with spatial neglect for spatial retraining, as endorsed by professional organizations, helps them recover to greater independence and quality of life.[19]

CASE 6-4

A 27-year-old man was seen in neurologic consultation on a brain injury unit at a rehabilitation hospital after admission subsequent to a car accident that had occurred while he was intoxicated with stimulants and cocaine. He reportedly experienced a "hemorrhagic brain injury," a pelvic fracture, and multiple other injuries. Medical record review revealed elevated blood pressure since admission (systolic blood pressure approximately 150 mm Hg to 170 mm Hg).

On examination, his memory was normal for orientation and three-object recall and calculations were intact. He demonstrated psychomotor slowing; however, all of his reporting about his history and prior work experience was consistent with the chart and with normal reasoning ability. His apparent mood was euthymic. Spontaneously, he appeared to have left hemiparesis affecting his face, arm, and leg, with the greatest weakness in his arm and hand. Effort was decreased on confrontation strength testing; the examiner had to reposition the patient's left arm several times when testing pronator drift because, although he was strong enough to elevate his hand, he kept dropping it, as if tired. At the end of the interview, the examiner drew a vertical line on a piece of paper, wrote "normal strength" at the top of the line and "cannot move" at the bottom of the line, and told the patient "I want you to rate the strength you showed on the testing just now for the left side of your body and your left arm. You might mark here [gesturing to the top of line] if your strength is normal; you might mark here [gesturing to the bottom of line] if you cannot move your left body at all. Or, you might mark your strength somewhere in between [gesturing to show the whole line]." The patient took the pen and, without hesitation, made a mark at the very top of the vertical line. The examiner asked, "Does this mean you don't feel weak on the left side?" The patient looked at the examiner mildly and said, "Everyone says I am weak, but I don't feel weak at all." The examiner was subsequently able to view the patient's brain image from the acute care hospital; it revealed a right putaminal hemorrhage.

COMMENT

A classic teaching about anosognosia is that only patients with global cognitive impairment have unawareness of deficit.[77] However, many patients, like the one described in this case, have relatively normal memory and yet are unaware of their neurological deficits. Testing for unawareness of deficit (anosognosia) with a visual analog scale (the line the patient marked) can be very helpful and useful to show to staff or caregivers so that the neurologist can discuss anosognosia as a neurologic deficit independent of motivation, psychological denial, or willingness to recover.

KEY POINTS

- The unsafe behaviors observed as symptoms of spatial neglect can be misattributed to intellectual, reasoning, personality, or generic cognitive problems, which can be devastating to patient dignity.
- Misattribution of spatial neglect symptoms can delay diagnosis of right brain stroke past the window for acute stroke interventions; thus, clinicians should be alert to new spatial bias in patients with stroke risk factors.
- Aiming spatial neglect causes several different movement-related problems. Spatial neglect can cause an ipsilesional turning tendency and disinclination to move in the direction opposite the brain lesion. This symptom is disabling beyond the effects of hemiparesis or awareness problems.
- In-hospital fall riskcan be more than 5 times higher in people with spatial neglect than in those without it. Further, active movements that are posturally biased means that the best supervision and guarding by nursing assistants and other personnel may not be effective.
- Line bisection is a simple spatial neglect screening test. Accurate line bisection cannot rule out spatial neglect, but more than 1 cm (0.4 in) of rightward error in bisecting a horizontal line longer than 22 cm (8.7 in) is highly likely to indicate that a patient has spatial neglect.
- Many survivors of stroke benefit from continuing rehabilitation, but most receive no outpatient rehabilitation. Referring patients with spatial neglect for spatial retraining, as endorsed by professional organizations, helps them recover to greater independence and quality of life.
- Ten percent or more of survivors of stroke have chronic spatial neglect years later.
- The degree of unawareness of deficit can be varied; patients with anosognosia may “know what to say,” about their deficits and yet they may not believe they are disabled.

FIGURE 6-1. Photo of a patient with a right brain stroke and left spatial neglect who maintained an abnormal, asymmetric posture. His rightward head and eye turning (apparent rotation of his torso) caused severe neck and back pain.

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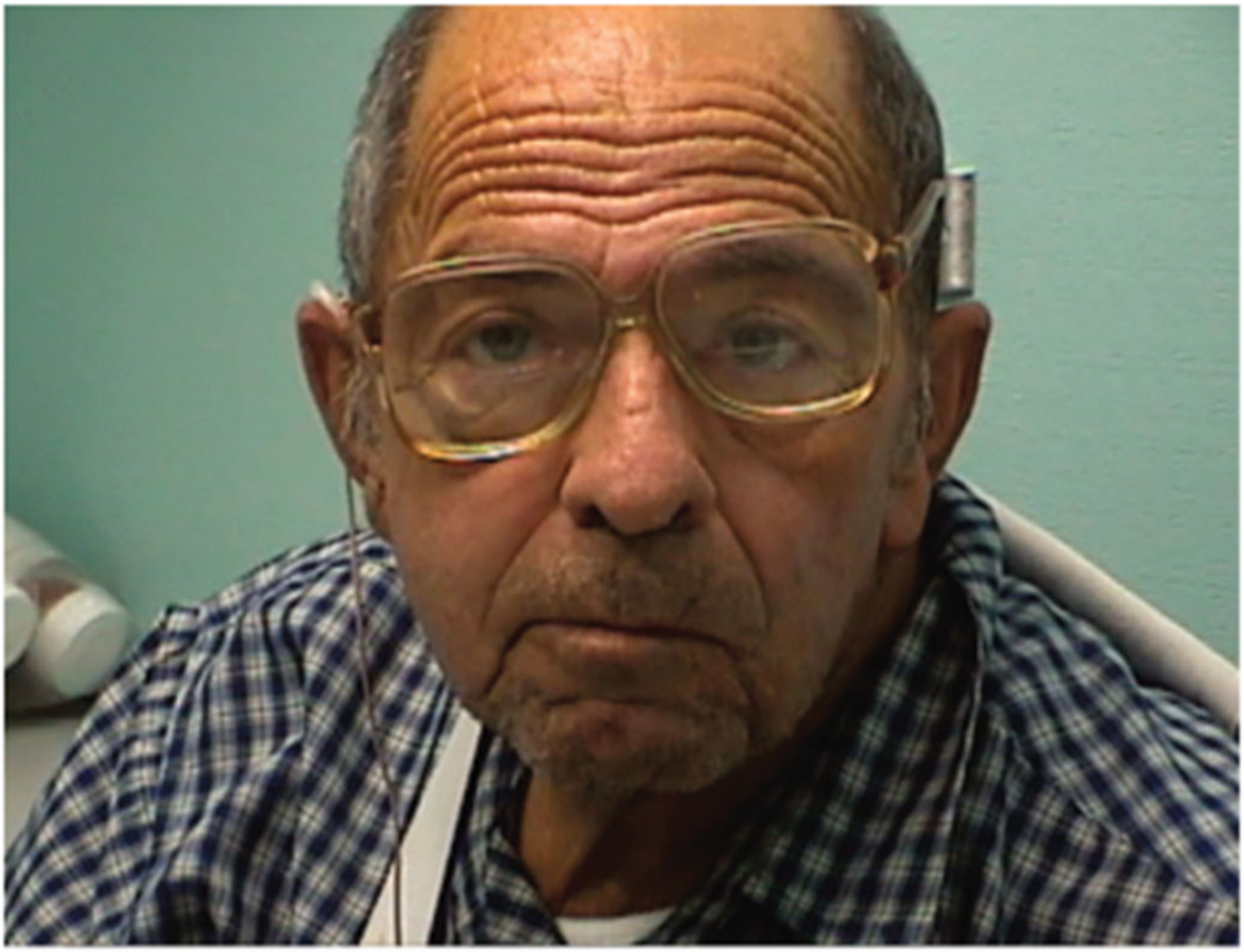

FIGURE 6-2. Photo of a man with spatial neglect after right brain stroke manifesting the hanging eyeglasses sign, which is evidence of asymmetric movement. The left temple of the eyeglasses rests on the side of the head, above the ear, because the leftward movement of the hand is too small to seat the eyeglasses on the ear on the left side.

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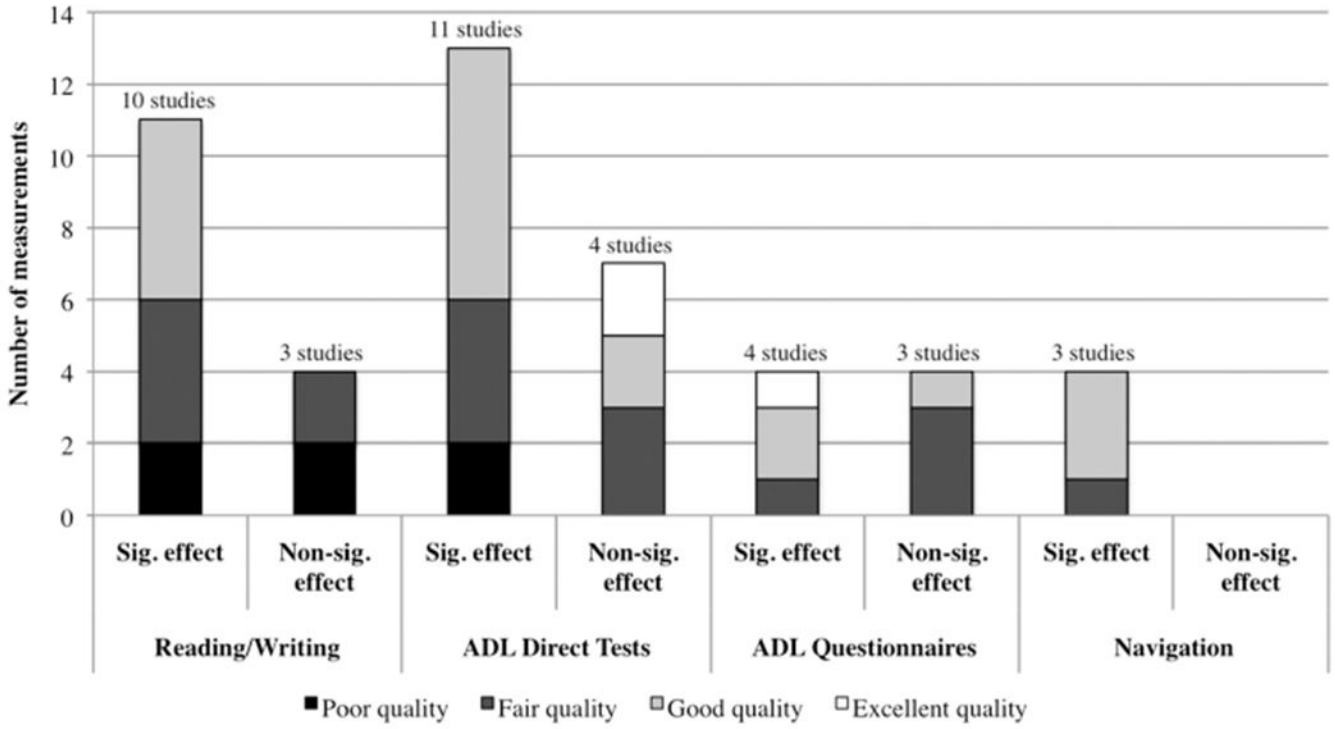

FIGURE 6-3. Evidence of prism adaptation treatment effects on daily living skills: reading/writing, activities of daily living (ADL) direct tests, ADL questionnaires, and environmental navigation tests.

Sig. = significant; non-sig. = non-significant.

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TABLE 6-1

Prevalence of Spatial Neglect[a]

Study	Total study, N	Left brain stroke, %	Right brain stroke, %	Setting	Country
Gainotti et al, 1972 [7]	222	31	42	Outpatient clinic	Italy
Denes et al, 1982 [8]	48	21	33	Geriatric hospital	Italy
Fullerton et al, 1986 [9]	205	25	49	General hospital	Ireland
Stone et al, 1993 [10]	171	65	82	General hospital	United Kingdom
McGlone et al, 1997 [11]	138	31	62	General hospital	Canada
Kalra et al, 1997 [12]	145	21	43	General hospital	United Kingdom
Ringman et al, 2004 [13]	750	20	43	Acute care hospital	United States
Chen et al, 2015 [14]	121	47	76	Inpatient rehabilitation	United States
Hammerbeck et al, 2019 [15,b]	~90,000	37	41.5	Acute care hospital	England, Wales, Northern Ireland

[a] Modified with permission from Chen P, et al, Top Stroke Rehabil.[6] © 2012 Taylor & Francis Ltd.

[b] Side of stroke assigned based on side of arm weakness on the National Institutes of Health Stroke Scale.

TABLE 6-2

Key Components of Aiming Spatial Neglect

Symptom	Abnormality (after right brain damage)	Finding (after right brain damage)
Motor extinction	Difficulty moving both sides of the body at the same time, with the left body failing to move properly when the right body is activated	Patient raises only the right arm when asked to raise both arms; however, strength tested to confrontation is good in both arms
Directional hypokinesia	Problems moving leftward with the eyes, head, limbs, or axial body; not accounted for by paralysis alone	Patient sits, stands, and moves with rightward rotation; veering while ambulating can cause collisions
Hemispatial hypokinesia	Smaller or weaker movements in left space as compared with right space	The patient's grip with either hand is weaker to the left of the body than it is with the hand positioned in the right body space
Limb hypokinesia	Smaller or weaker movements by the left hand, arm, and even leg compared with the right limbs; not accounted for by paralysis alone	Similar to motor extinction, except that spontaneous left arm movements are weak or small even when that limb moves in isolation; however, strength tested to confrontation is good in both arms; patient may "forget" arm and leave it in an unsafe position
Defective motor response inhibition	Stimulus-evoked responses in a leftward direction or with the left body; cannot be inhibited by goal-oriented, conscious intention although the right body can be inhibited	Patient cannot inhibit leftward glances or cannot inhibit grasp or reach (leftward or with left hand or arm) while walking, during transfers, or during complex activities (eg, using power equipment); can interfere with safety

TABLE 6-3

Spatial Neglect Subtypes and Potential Associated Deficits[a]

Spatial neglect subtype	Key behaviors demonstrating spatial neglect	Frequently observed functional impairments[b]
Where, perceptual-attentional awareness [45]	Extinction to double simultaneous stimulation, decreased vigilance in left body space	Accidents due to failure to respond to left-sided events; failure to respond in social interaction; difficulty in eating entire meal
Where, representational imagery [46]	Visual imagery incomplete on left, neglect dyslexia, right bias on mental number line	Poor environmental navigation; illusions, difficulty recognizing objects especially under unfamiliar conditions; difficulty reading or completing mathematical operations accurately for work, financial activities
Aiming, motor intention	Abnormalities listed in TABLE 6-2	Postural imbalance, veering while ambulating while walking or in a wheelchair, augmented hemiparesis (weakness out of proportion to motor dysfunction)

[a] Modified with permission from Barrett AM, et al, Handb Clin Neurol.[44] © 2019 Elsevier Ltd.

[b] Proposed, further systematic research demonstration is needed.

TABLE 6-4

Professional Guidelines for Spatial Neglect Treatment[a]

Professional organization endorsement	Spatial neglect treatment for any patient	Targeted treatment to improve Aiming spatial neglect and arousal/activation	How might this treatment work to improve outcomes?[b]
American Heart Association (AHA),[19] **American Occupational Therapy Association,**[51] **Intercollegiate Stroke Working Party**[50]	Prism adaptation therapy[17]	Prism adaptation therapy	May reduce Aiming spatial neglect, making leftward and left-body movements more adaptive
US Department of Veterans Affairs,[52] **Intercollegiate Stroke Working Party**	Compensatory, self-mediated cuing strategies[53]	Not applicable	Not a treatment, rather a management technique that may reduce the possibilities for error
AHA, Intercollegiate Stroke Working Party	Visual scanning training[54]	Not applicable	Neuropsychological mechanisms uncertain
AHA, Intercollegiate Stroke Working Party	Limb activation[55,56]	Limb activation	Proposed to stimulate body-based spatial systems, alter interhemispheric interaction; may also improve limb hypokinesia and Aiming spatial neglect
Canadian Partnership for Stroke Recovery [57]	Medications (acetylcholinergic, nicotine, noradrenergic)	Medications (dopaminergic, noradrenergic)	May improve attention, alertness, arousal, generative behavior, persistence, memory; paradoxical worsening can occur with dopaminergic treatment

[a] Modified with permission from Barrett AM, et al, Handb Clin Neurol.[44]

[b] Proposed, further research is needed.

TABLE 6-5

Different Forms of Anosognosia That Can Manifest After Right Brain Stroke[a]

Term	Anosognosia type
Anosodiaphoria	Lack of emotional concern for deficits that are verbally acknowledged
Anosognosia with causal attribution abnormality	Acknowledgment of the deficit without linking the problem to a neurologic issue (eg, a patient saying, "I don't feel like moving my left arm right now."); inconsistent verbal acknowledgment (patient reports that others believe the deficit is present)
Anosognosia with implicit awareness	Conscious disavowal of deficits that are acknowledged through behavior (eg, patient claims the ability to walk but never actually tries to get out of bed)
Anosognosia with modality specificity	Acknowledgment of one deficit but not others (eg, patient is aware of a visual field cut but not aware of hemiplegia)
Anosognosia without implicit awareness	Combined explicit and implicit disavowal of deficits (both behavioral and verbal reports) without acknowledgment of deficits

[a]Data from Ofrei MD, et al.[71]