Hemispatial Neglect: Clinical Features, Assessment and Treatment

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Abstract

Hemispatial neglect is a disorder of attention which commonly follows from damage to the right side of the brain. Patients with neglect show symptoms of lateralised inattention, failing to acknowledge or report information on the left side. Neglect is a poor prognostic indicator for general functional recovery from stroke, and is associated with a range of co-morbid conditions including denial or indifference to the brain injury, hemiplegia and visual field loss. Mild to moderate cases can be over-shadowed by the more gross symptoms that accompany brain injury, however assessment and diagnosis is relatively quick and simple. Current treatment guidelines suggest that patients should be taught compensatory strategies, but these are largely ineffective. Although recent research has identified more promising treatment approaches, investigations are still preliminary. Given the prevalence and debilitating nature of neglect, there is a clear need to raise awareness and understanding of the condition amongst carers and healthcare professionals.

Introduction

Hemispatial neglect is a relatively common attentional disorder resulting from unilateral hemisphere damage, most commonly from a stroke but also from other conditions such as tumour or multiple sclerosis. Estimates of prevalence vary, but the most conservative indicate that approximately 17% of stroke patients with right brain lesions and 5% with left brain lesions will continue to show neglect 3 months post-onset (Ringman et al, 2004). Patients with neglect pay less attention to the space on the opposite side to their injury (i.e. the left side in cases of right hemisphere damage), failing to respond to objects and people and forgetting to use their limbs. Crucially, neglect is one of the strongest predictors of general functional recovery post-stroke (Nijboer et al, 2013). This may follow from the fact that many forms of neuro-rehabilitation require patient volition and active engagement, qualities that are compromised in neglect.

Clinical Presentation

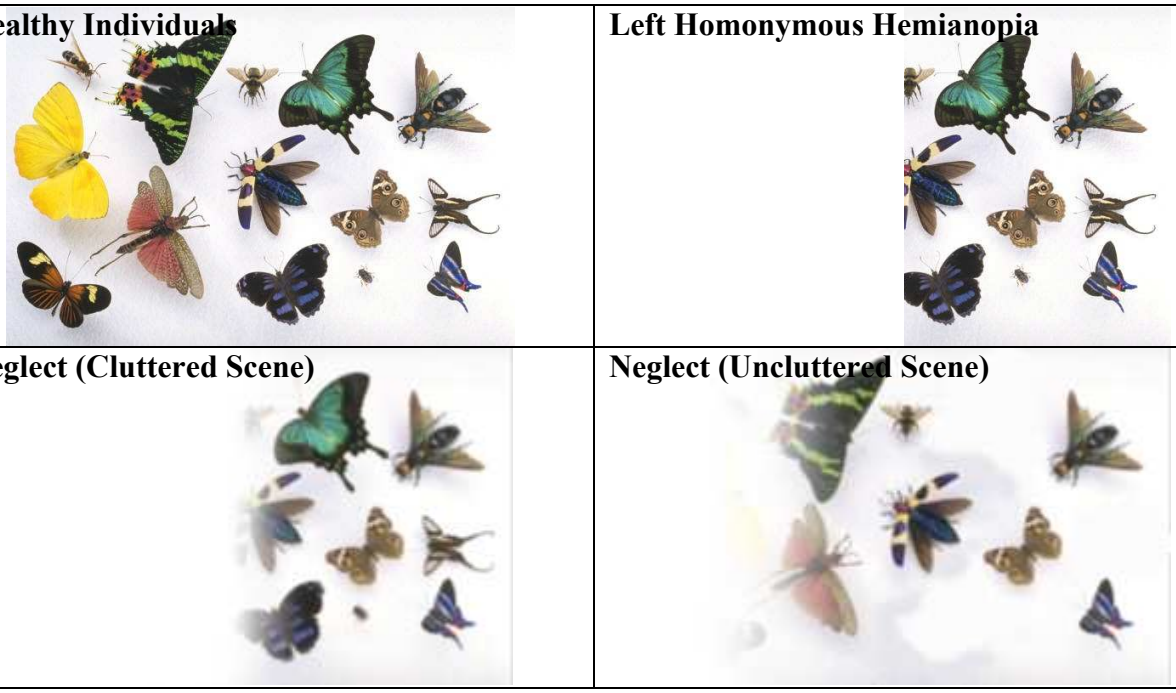
Severe cases of neglect are immediately apparent during bedside observations of behaviour; patients will turn their trunk and head to the same side as their injury, and noticeably ignore even salient left-sided events. Milder cases are less discernible, especially if the patient is bed-bound or in an unchanging, familiar environment. Sometimes the condition manifests in a relatively selective manner. With 'egocentric' neglect, patients tend not to attend to objects on the contralesional side of their environment relative to their own body, while patients with 'allocentric' neglect tend not to attend to the contralesional side of objects, regardless of their relative body position (Ting et al, 2011). Patients may also show neglect within either their personal space, leading to problems with personal care, or their peripersonal space (space near to the body), leading to problems with eating and reading. A small subset of patients may only manifest neglect toward objects located beyond reach (extrapersonal neglect), compounding the rate of collisions and navigational errors (Ting et al, 2011).

Beyond its immediate impact on visuo-spatial ability, the appearance of neglect should alert clinical staff to the likely presence of co-morbidities. Neglect is a poor prognostic indicator for recovery from stroke (Parton et al, 2004) and is associated with a number of conditions, including depression, apraxia, limb spasticity, anosognosia ('denial of illness'), prosopagnosia, and hemianopia (Hier et al, 1983; Wilkinson et al, 2012) (*Box 1*). In one recent study, Wilkinson et al (2012) showed that individuals (n=106) with neglect are nearly one third more likely to develop limb spasticity than those without neglect (87% vs

57%), and nearly one half of those with left-sided spasticity will show neglect (44% vs 13%). Patients with neglect are more likely to have an increased length of hospital stay, are more likely to suffer from incontinence, have less functional independence and have problems with mobility. This may have major implications, such as the loss of a driving license or losing the use of an electric wheelchair (Paolucci et al, 2001).

Box 1: The difference between neglect and hemianopia.

Both neglect and hemianopia may lead patients to miss information on the left, but the two disorders have different causes and require different treatment plans. Neglect reflects an attentional deficit that is usually caused by a cortical lesion, while hemianopia reflects a cut in the visual field that is usually caused by a lesion to the geniculate striate pathway, which projects from the retina to the occipital pole of the brain. Given their common behavioural manifestation, standard visual field testing may not distinguish the two disorders as it may not be apparent whether a stimulus is missed due to a lack of attention or visual field loss. The disorders may however be disentangled by comparing individuals' responses to stimuli presented on only one side with their responses to stimuli presented on both sides simultaneously. Individuals with left hemianopia will consistently miss stimuli in the left visual field, regardless of whether they are presented alone or with competing stimuli in the opposite field. During pencil and paper tasks, patients with hemianopia will often move their head and eyes to bring left-sided stimuli into view, and show good awareness of their sensory deficit. Those with mild-to-moderate neglect will usually miss the contralesional stimulus only when it is simultaneously presented with a competing ipsilesional stimulus. They may also appear apathetic toward their neglect and, if severely affected, will turn their head and trunk away from the neglected field. They may also show neglect in the auditory and tactile domains.



The figure represents an idealised visual experience of neurologically healthy individuals compared with those with hemianopia and neglect. In hemianopia, the boundary between the intact and blind field is often perceived as a 'cliff', whereas in neglect the visual loss is more graduated with the features of the scene affecting the size of the neglected field.

Assessment

Current guidelines recommend that suspected cases of neglect be confirmed using the Behavioural Inattention Test (BIT) (Wilson et al, 1987; Intercollegiate Stroke Working Party, 2012). The BIT contains six pen-and-paper and nine behavioural assessments and is highly reliable and sensitive (Wilson et al, 1987). Pen-and-paper assessments include star cancellation, letter cancellation, line crossing, line bisection, free drawing and shape copying tasks. In the star and letter cancellation and line tasks, patients are presented with an array of target symbols that they are required to mark (*Figure 1*).

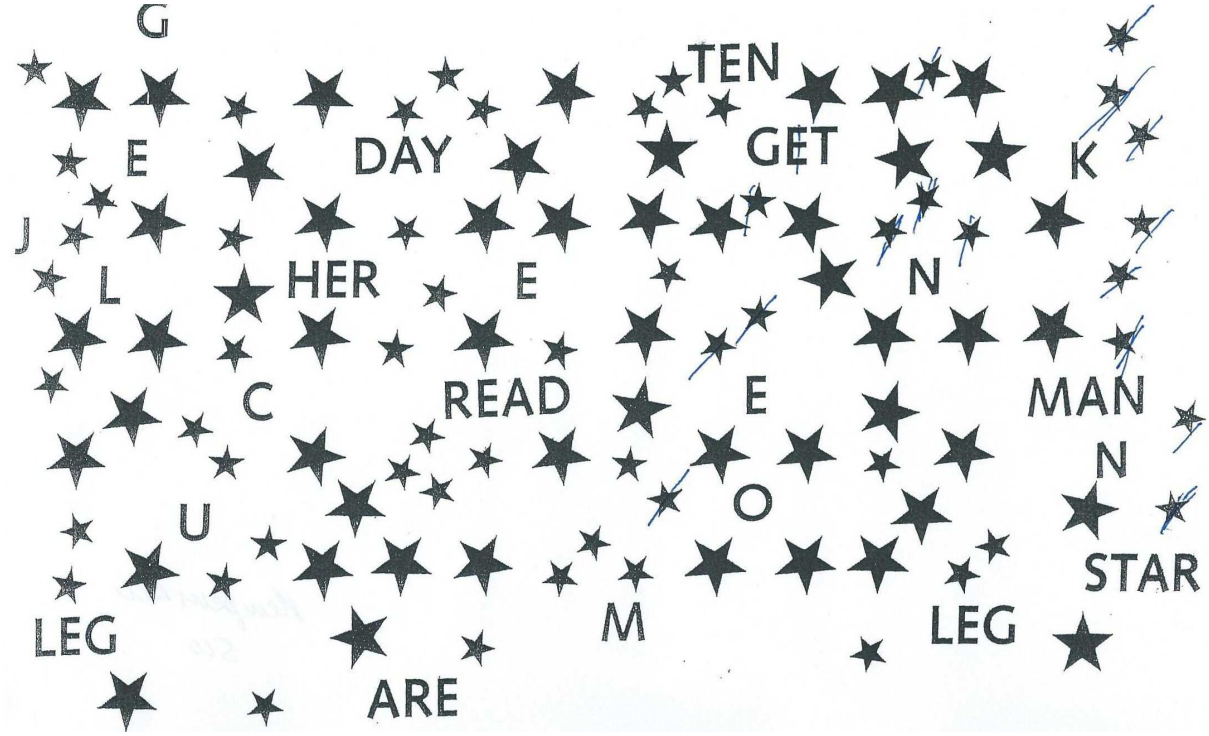


Figure 1. Star cancellation performance of a neglect patient showing the characteristic failure to cross out left-sided, small stars

Patients with neglect are likely to miss targets toward the contralesional side and often begin to search from the ipsilesional side of the page. In the line bisection task (*Figure 2*), patients are asked to mark the mid-point of several horizontal lines. Those with left neglect are likely to mark the mid-point further to the ipsilesional side than healthy control subjects.

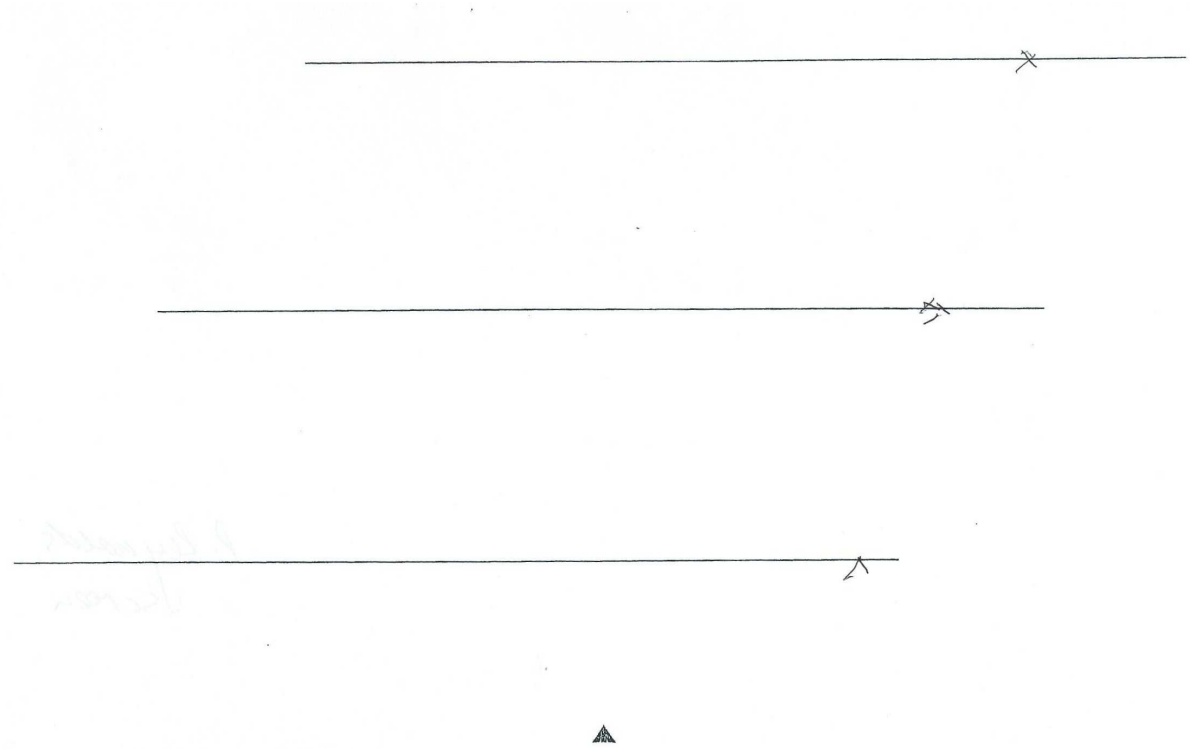


Figure 2. Line bisection performance of a neglect patient, showing characteristic right-sided midpoint estimation

In shape copying and free drawing tasks patients are asked to reproduce simple geometric shapes or everyday objects, such as a clock face or flower (*Figure 3*). Those with neglect are more likely to miss the contralesional side of these images when drawing.

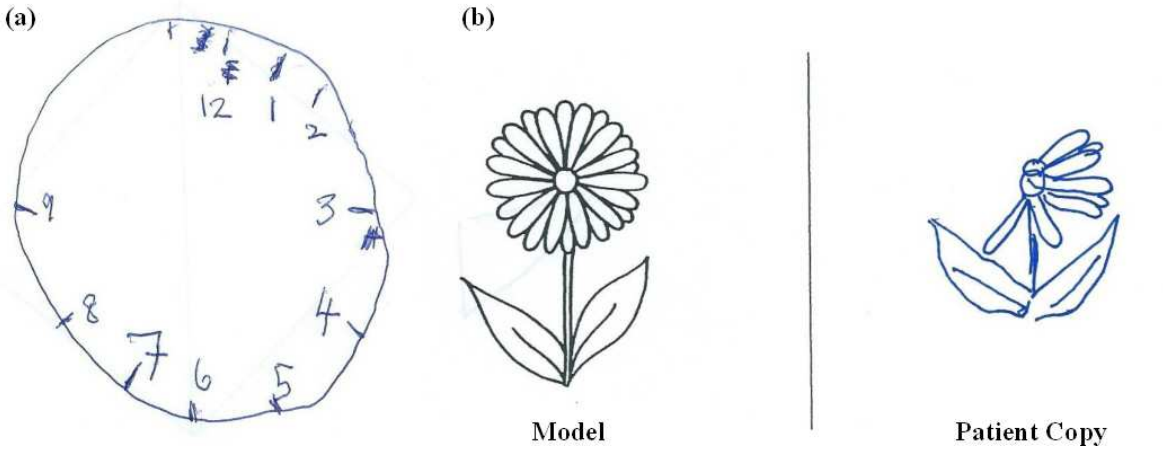


Figure 3. Examples of a patient with neglect drawing from memory (A) and copying a figure (B)

Although studies have found that the cancellation tasks are the most sensitive of all pen-and-paper assessments (Ferber and Karnath, 2001), the combination of several subtests most effectively detects neglect. Although the pen-and-paper tasks tend to reliably capture the visual and spatial elements of neglect, they are time-consuming and do not map simply to

the problems encountered during daily living (Azouvi et al, 1996). To address this shortcoming, the BIT includes the additional assessments of menu and article reading, setting and telling the time, map navigation, card sorting, picture scanning, telephone dialling, coin sorting and sentence copying. These assessments do not, however, significantly increase the sensitivity of the battery, so they are rarely administered for diagnostic purposes.

One scale that does attempt to capture how neglect affects activities of daily living is the Catherine Bergego Scale (CBS) (Azouvi et al, 1996). The CBS consists of 10 items related to everyday functioning, including grooming, dressing, eating and navigation. Each item is assessed on a four point scale, where a score of 0 is indicative of no neglect and a score of 3 indicates severe neglect. One form is completed by the therapist and another by the patient to help assess his/her anosognosia. Although the CBS can provide a more detailed image of how neglect affects daily living, it lacks the large-scale validation of the BIT and relies on subjective rather than objective assessment.

Anatomy

Although a range of aetiologies may lead to neglect, it is most commonly observed following a cerebral haemorrhage or infarction within the territory of the middle cerebral artery (Kerkoff, 2001) (*Figure 4*). Often the resulting lesion centres on the inferior parietal cortex, but damage to the brain is typically widespread and involves a number of brain regions involved in attention, perception and memory. This underlying anatomical variability may partly explain the heterogeneous presentation of the condition, whereby different lesion locations give rise to different behavioural subtypes (Karnath and Rorden, 2012).

Figure 4. Axial computerised tomography scan of a neglect inducing lesion (circled) within the right temporal-parietal region

Although no critical brain region has been identified for neglect, a recent meta-analysis (Molenberghs et al., 2012) found nine brain regions which are commonly associated with the condition. These regions included the right superior longitudinal fasciculus; right posterior middle temporal gyrus/right angular gyrus; right inferior parietal lobule; right caudate nucleus; right anterior horizontal intraparietal sulcus/postcentral sulcus; right precuneus; right superior temporal gyrus/superior temporal sulcus; right posterior insula; and right middle occipital gyrus. This widespread network of brain regions may thus account for the fact that neglect is relatively common following right hemisphere damage.

Treatment

Current guidelines for the treatment of neglect recommend teaching the patient compensatory strategies that may be incorporated into physiotherapy and occupational therapy sessions. The most widespread technique is visual scanning therapy (VST; Pizzamiglio et al, 1992). This technique involves retraining patients to look toward the contralesional side via visual search, reading and copying exercises. Although several studies have shown that VST can benefit patients (Luukkainen-Markkula et al, 2009; Kerkhoff and Schenk, 2012), often the treatment is time-consuming (requiring approximately 40 hours of therapy) and only targets the visual aspects of neglect.

More recent experimental treatments have focused on targeting the underlying causes of neglect rather than bypassing or minimising the behavioural loss. Pharmacological treatments have been developed with varying degrees of success. Given that dopamine modulates attention and working memory, several studies have tested whether dopaminergic drugs can reduce lateralised attentional bias. A recent study (Gorgoraptis et al, 2012) found that the administration of rotigotine improved performance on the cancellation task, and another found that treatment with carbidopa–levodopa significantly improved BIT scores in three of four patients (Mukand et al, 2001). Although the results appear promising, other studies have not replicated these findings (Buxbaum et al, 2007), highlighting the need for further clinical trials (Sivan et al, 2010). The efficacy of pharmacological interventions in neglect is also often hampered by patients' lack of insight, self-monitoring and motivation, all of which lower compliance.

Recent years have witnessed a proliferation in noninvasive neuro-stimulation therapies for brain injury, including vestibular stimulation, transcranial magnetic stimulation and transcranial direct current stimulation (Müri et al, 2013). These techniques are believed

to facilitate neuroplastic change within and around the damaged brain regions, through various physiological mechanisms. The most longstanding method of non-invasive brain stimulation is vestibular stimulation (Bárány, 1914). The vestibular system, also known as the balance system, conveys information about head movement from the inner ear to the brain, which in turn increases blood flow to those regions typically damaged in neglect patients. Until recently the procedure was not easily tolerated by patients, but advances in biomedical engineering have overcome this shortcoming and produced safe, cheap stimulators suitable for home-based use (Utz et al, 2010; Kerkhoff and Schenk, 2012; Zubko et al, 2013). As with other neglect therapies (*Box 2*), the efficacy shown in early-stage studies now needs to be replicated in larger randomised controlled trials (Bowen et al, 2013). At present the range of interventions available to a particular patient depends largely on the local opportunities that happen to be available (Ting et al, 2011; Wilkinson et al, 2011).

Box 2: Current and experimental treatments for neglect.

There are a number of potential treatments for neglect, ranging from compensatory and training strategies to those targeting the underlying deficit. Although some have been widely researched, others are still under investigation. All lack validation from large-scale trials (Bowen et al, 2007).

- Optokinetic stimulation
    - Patients watch stimuli on a computer screen moving coherently to the left side. Repeated sessions have been shown to help normalise attentional orienting.
- Neck muscle vibration
    - Vibration over the left neck muscles induces a perception of continuous movement to the right. As with optokinetic stimulation, this perception causes patients to compensate for this movement and shift attention to the left.
- Prism adaptation
    - Patients are asked to point to visual targets while wearing lenses that shift the visual field to the right. This field shift is believed to induce a visuo-motor recalibration that encourages leftward movement and orienting.
- Transcranial magnetic stimulation (TMS)
    - Magnetic pulses are applied to the intact side of the brain, potentially disrupting the neural activity and thus reducing the ipsilesional bias. In addition, TMS may, like other brain stimulation techniques, induce neuroplasticity.
- Eye-patching
    - Patches are applied to the normal, ipsilesional (i.e. right) visual field which prevents visual information from reaching the intact hemisphere. This inhibition appears to ‘release' visual processes within the damaged hemisphere and help restore neglect.

Conclusion

Hemispatial neglect is a common condition following unilateral brain damage and can profoundly affect many aspects of daily routine. Neglect is a poor prognostic indicator for recovery from stroke (Paolucci et al, 2001) and is often accompanied by a range of comorbid conditions (Wilkinson et al, 2012). Early identification and awareness may facilitate recovery and improve wellbeing by minimising impact on functional tasks. Diagnosis is relatively simple and accurate, yet clinical time is often directed toward more grossly observable conditions that also follow from acquired brain injury, such as pain, aphasia and hemiplegia. Although many treatments for neglect hold promise, at present they are experimental and not widely available. However, with increased awareness among health practitioners and an openness to participate in trials, the impact of neglect need not be so vast.

Key Words: Hemispatial neglect, stroke, brain injury, neuropsychological assessment, attention

Key Points:

- Hemispatial neglect is a common attentional disorder following unilateral brain damage
- Neglect is one of the strongest predictors of general functional recovery from stroke
- Identification of patients with neglect is important, as it can alert clinicians to co-morbid conditions and aid staff with day-to-day patient care
- Potentially effective treatments are in development but these lack large-scale, trials validation.

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