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Visual neglect: should we attend to it?

TRACEY L. SHIPMAN PGC BSc (Hons) DBO(D)

Orthoptic Department, Sheffield Teaching Hospitals NHS Foundation Trust, Royal Hallamshire Hospital, Sheffield

Abstract

Aim: **To review the literature on unilateral visuo-spatial inattention/neglect following stroke, concentrating on the areas of the brain involved, methods of assessment and therapeutic options.**
Methods: **A literature-based survey was conducted using relevant articles and practical experience to elicit current theories of inattention and to discover whether any evidence-based treatment strategies exist. As there are numerous testing approaches for inattention, evidence was also sought for the most appropriate and accurate assessment tool(s).**
Results: **The star cancellation, line bisection and random shape cancellation tests appeared to be the most sensitive for detecting neglect; however, a range of tests is necessary in order to detect severity. Therapeutic options may include scanning techniques and visuo-motor cueing into the affected side, prism adaptation and limb activation to increase awareness of the affected side and reduce inhibition by the healthy hemisphere. Motor and functional recovery of stroke patients with neglect seems to be improved by targeted treatment.**
Conclusion: **Neglect is an important predictor of poor functional recovery and therefore treatment remains a high priority. However, more research is needed to better define which treatment options are the most effective.**

Key words: Neglect, Scanning training, Visual inattention, Visuo-motor cueing

Introduction

Unilateral visual inattention (VI) or neglect is an umbrella term to describe perceptual, attentional, motor and sensory defects. VI forms part of this deficit and refers to behavioural symptoms whereby a patient fails to report, respond to or orientate to meaningful stimuli presented to the side opposite a cerebral lesion. Motor neglect is a failure to move in the contralesional space and can involve the eyes, head, limbs or trunk. Sensory neglect is a lack of or decreased awareness of sensory stimulation in the contralesional space.[1]

Correspondence and offprint requests to: Tracey Shipman, Orthoptic Department, Royal Hallamshire Hospital, Sheffield Teaching Hospitals NHS Foundation Trust, Glossop Road, Sheffield, S10 2JF. e-mail: Tracey.Shipman@sth.nhs.uk

The characteristics of VI vary from complete unawareness of anything or anyone situated on one side, to omitting the occasional word from part of a line of text.[2] The left side is more commonly affected than the right. Typical signs include patients bumping into objects on their left side, failing to eat food on the left side of the plate, or dressing only one side of their body. Unfortunately, many patients are totally unaware of or deny any ill-effects of their stroke. This is termed anosognosia and is common following dominant parietal lobe lesions with hemi-paresis. This results in a belief that their perception of space is entirely normal, and making patients aware of the deficit can be extremely difficult.

The neglected space can be far space (extra-personal), reaching space (peri-personal) or body space (personal).[2] There may be some or all of these present in combination. Most formal tests of VI are tests of the peri-personal space; visual acuity testing at distance can be considered a test of extra-personal space. Neglect of the body is not always associated with neglect of peri-personal space, so that a patient may not attend to their left arm or apply makeup to the left side of the face but can reach out for an object on the left side. What exactly constitutes the neglected side is also debatable – the reference point for the neglected side does not necessarily start at the midline of the body but refers to the left half of the information wherever it appears in the visual field, and vice versa in right neglect.[2]

Neurophysiology

VI and spatial neglect is a common deficit that may occur following stroke.[3] It occurs as a result of damage to various disparate areas within the brain, namely the posterior parietal cortex, frontal lobe, cingulate gyrus, striatum and thalamus;[1] however, the inferior parietal cortex is the structure that is nearly always involved in the insult.[4] This has been corroborated in patients with middle cerebral artery stroke, where the critical area involved was the angular gyrus of the inferior parietal lobe. In posterior cerebral artery stroke all patients with neglect had lesions involving the parahippocampal region on the medial surface of the temporal lobe.[4] Posterior parietal lobe damage results in a decreased ability to shift attention covertly and reduces the ability to disengage from an attentional focus to a contralateral target.

The right hemisphere is usually thought to be responsible for many unconscious processes and orientates attention to the left or right.[5] The left hemisphere,

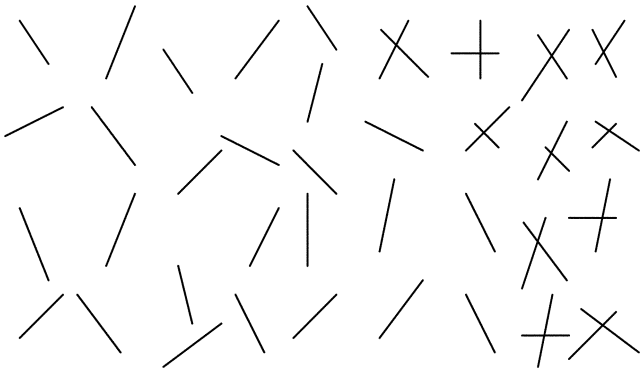

Fig. 1. Albert's test: patient responses showing left-sided visual neglect.

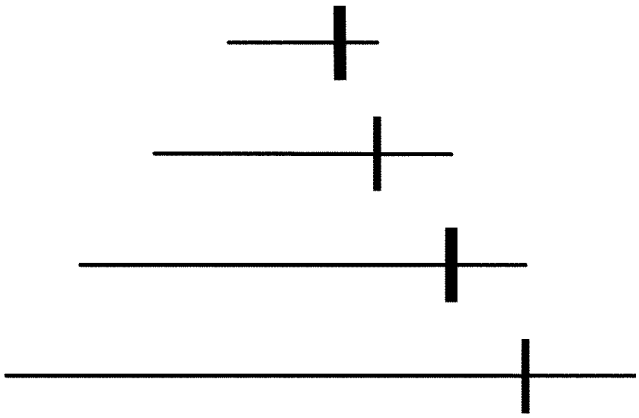

Fig. 2. Line bisection test.

however, orientates attention exclusively to the right; therefore following right hemispheric damage these patients may be left without any left-orientating facility. This leads to the theory that right hemispheric damage more likely results in the development of left neglect than vice versa.[6]

Brain arousal is mainly controlled by the right hemisphere.[5] Cognitive attentional problems are a significant predictor of persisting *in*attention, so it appears that these two systems (attention and inattention) are linked.[1] Patients who have been trained to improve sustained attention also demonstrated improvements in inattention.[7] There are several theories of the attentional system. The posterior parietal lobe, frontal eye fields, superior colliculus, pre-motor cortex and limbic system all play a role. These systems have extensive inter-connections with each other and with a larger neural network. The limbic system forms a representation of the external environment; it targets specific stimuli and then searches these stimuli through visual scanning.[8] As the parietal lobe is largely responsible for creating this sensory representation and targeting stimuli then an interruption to any of its connections, rather than a lesion in a specific anatomical area, may result in damage to the attentional system and cause neglect.[8]

Incidence

VI is a frequent sequela of right-hemisphere damage[9] although it has been reported in left hemisphere damage.[10–12] The incidence varies but some degree of neglect has been reported in up to 80% of cases.[11,13] The effects of VI can persist for at least 18 months after a stroke[14] and may sometimes be permanent. The neglect syndrome rather than overall stroke severity is an important predictor of poor functional recovery,[15,16] and therefore therapy treatment for neglect remains a high priority.

Methods of assessment

VI is multidimensional and clinical testing can be challenging. During an orthoptic assessment of the stroke patient it often becomes apparent if the VI is moderate or marked. Patients often miss or omit letters from one side of the vision chart. Those patients with marked neglect often turn their head and body to the unaffected side; that is, in left neglect they turn towards the right and may demonstrate a large abnormal head posture of face turn right with their eyes deviated in extreme right gaze, simulating a gaze palsy. In fact, a marked spontaneous horizontal deviation of the eyes and the head observed in the very early days following a stroke is a specific sign of spatial neglect.[17]

Stroke can selectively impair reading and this can have a profound impact upon daily activities;[18] a reading assessment is therefore a vital component of an orthoptic examination. When assessing reading skills patients may miss either the first word on the left side or only read the very last word on the right side (in left neglect), depending upon the extent of the VI. They often continue to read in this way, somehow making sense of nonsense. Frequently, patients 'fill-in' omitted words to compensate,[2] or comment that they are unable to read, as the words are confusing, but they deny that any are missing from their view.

VI can usually be tested quite simply but formally at the bedside using paper and pencil tests such as line cancellation tests, drawing and copying objects. The most sensitive objects or shapes for detecting VI are a clock face, a butterfly and the human body.[19] When asked to draw the numbers on a clock face patients typically fit all numbers 1–12 on the right-hand side of the clock, or omit numbers 7–12 when asked to copy a clock face.[2] Cancellation tests require the patient to initially search for, and then score through, certain target symbols such as stars,[20] shapes,[21] bells[22] and lines.

Albert's test[23] (Fig. 1) consists of seven columns of black lines. Three of the seven columns (= 12 targets) are on the left side of a horizontally orientated 21.0 × 29.7 cm sheet of paper, one column, containing 5 lines, is in the middle, and three columns (= 12 targets) are on the right side. Patients are asked to cancel or score through all the targets.

The line bisection test (Fig. 2) generally consists of 4 horizontal lines of differing lengths. The test page should be presented directly in front of the patient so that its centre is lined up with the patient's mid-sagittal plane. Patients are asked to mark the centre of each line so that the lengths of the line on each side of the mark are equal. Patients with neglect typically place this mark to the right of centre (in left neglect), demonstrating that their

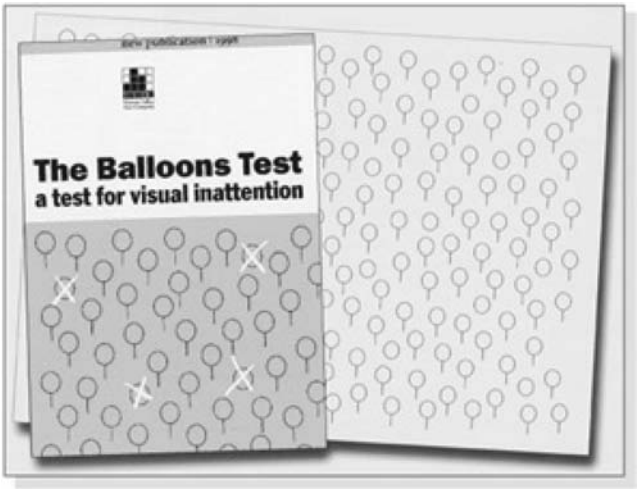

Fig. 3. The balloons test.[25] (Thames Valley Test Company, Bury St Edmunds.)

perception of centre is markedly skewed, as shown in Fig. 2. It has been reported that the extent of the deviation from centre is proportional to the line length, so that the longer the line the greater the deviation from centre.[24] This suggests that there is some type of brain processing of the whole line before the patient makes their decision on where to bisect the line, rather than part of the line simply being neglected. It seems that for testing purposes and obtaining meaningful results, longer lines should therefore be used rather than shorter ones.[24]

The balloons test[25] (Fig. 3) is another paper and pencil test and consists of two similar assessment sheets. On one is a series of small circles, a few of which have a small vertical line extending from the bottom – so-called balloons, or 'pop-out' stimuli. The balloons are easier to locate than the circles, and the patient is instructed to cross through the balloons only and not the circles. The second assessment sheet has the reverse situation, where most of the shapes are balloons rather than circles. Here the patient is asked to cross through the circles only. This is a more difficult test as there are no 'pop-outs', and it requires more of a functioning search strategy. This test can also help distinguish between neglect and a visual field defect as the prime cause of missing objects to one side. If a patient makes significantly more errors on the second, harder test than the first then it can be said that this is due to neglect rather than visual field loss, as the tests are visually similar.[2]

The star cancellation, line bisection and random shape cancellation tests appear to be the most sensitive tests for detecting visuo-spatial neglect.[26,27] A high score on the line cancellation test has been associated with recovery of VI and discharge home.[26] The best tests of visuo-spatial neglect for predicting outcome are also the star and line cancellation tests. Cancellation tests, however, appear to have greater test–retest reliability than line bisection tests.[28] A range of tests is likely to be required to cover various aspects of detection and severity of neglect.[29] Also, patients may perform better on one test but have marked VI on another;[29] this may be a reflection of the type of neglect or their attentional status at the time or the stage of the assessment.

It may be useful for the therapist to record how these tests are completed by the patient, noting their executive functioning in performing the test, such as planning, the start and finish points,[30] and the scanning pattern. In normal subjects, searching usually starts on the left and progresses systematically either horizontally or vertically,[22] whereas in patients with attentional defects, searching is disorganised and unmethodical.[22,31] The spatial location of the starting point has been noted to be the most sensitive measure[13] rather than the number of omissions.[22] If line cancellation tests are fully completed but in a disorganised manner, then subtle attentional deficits may go unnoticed if only the final result is assessed.[30]

Neglect or hemianopia?

The differential diagnosis of VI from hemianopia can be difficult, as the two deficits often coexist.[32] The differences are highlighted in Table 1.

When assessing eye movements, patients with VI often demonstrate few searching saccades across the midline and once the target has disappeared there is no search for it. Patients are usually unable to predict the

Table 1. Hemianopia or neglect?

Visual neglect	Hemianopia
Visual loss is commonly defined in terms of the patient's mid-sagittal plane	Visual field loss is retinopic
Attentional disorder	Sensory disorder
May involve the whole hemisphere	May be restricted to a quadrantanopia only
Little insight into the deficit	The patient usually demonstrates insight into the deficit and may complain of bumping into objects, or failing to notice people on the 'blind side'
During formal/informal assessments the patient fails to demonstrate compensatory strategies and may fail to cancel targets or bisect lines on the contralesional side	During formal/informal assessments the patient often compensates by scanning; they are usually able to cancel targets or bisect lines on the contralesional side
Inconsistent responses dependent upon level of attention	Consistent responses
May be evidence of visual processing on the side of the deficit	No evidence of visual processing on the side of the deficit
With head turn information is still unseen	With head turn information is often seen
Neglect may be present in other modalities: when asked to recall a known street the patient may fail to describe one half of the street; or may fail to wash/dress one side of their body even with the eyes closed	Hemianopia is purely visual and does not extend to other modalities
Extinction often present	Extinction often absent

position of a target once it has been removed from sight[2] and there is often insufficient smooth pursuit movement towards the side of neglect. Conversely, in hemianopia, patients can usually pursue a target into the hemianopic side. Their saccades are more ordered, spontaneously searching towards the hemianopic side and crossing the midline. Once a target is removed patients often start searching for it immediately. They can usually predict the position of an object placed in their hemianopia after a few successful search movements.[2] When reading, patients with both VI and hemianopia may omit the first few words on a line. However, the hemianopic patient will usually begin to make step-wise saccades back towards the start of the line to make sense of the written word.

Therapeutic options

Current therapy for VI focuses on stimulating the inattentive side using a range of strategies such as teaching visual scanning, using visual cues and providing non-specific visual stimulation into the affected side, such as seating friends and relatives on this side, along with the TV, books, puzzles, etc. This is generally referred to as visuo-spatial rehabilitation. Other therapists involved in the care of stroke patients can also integrate some of these strategies into their therapy sessions, combining motor and sensory stimulation with visual stimulation. The use of temporary prisms to shift the visual field into a more central position and utilising mirrors are further strategies. Other, more complex treatments have included using computer-based training programmes that generate left-sided visual cues[33] or other electronic devices; however, there is a lack of quality evidence on the efficacy of these devices and many are not widely available for use in the National Health Service and therefore will not be considered further.

Several studies have shown that motor interventions, in the form of small movements of the left hand in left space, reduced left inattention notably,[34–36] but this was less apparent when the left hand crossed into right space. It appears that when mutually facilitatory attentional circuits are activated, such as motor and sensory systems, and when this is combined with visual stimulation, then inhibition by the healthy hemisphere is overcome.

If limbs are activated bilaterally then the effects of left limb activation in left space are eliminated. In rehabilitation, therefore, it is important to stimulate the affected hemisphere alone without competition from the undamaged hemisphere. Extinction describes this phenomenon well and explains cases of left neglect where patients are able to attend to their left but when the stimulus is bilateral they always favour the right side.[2] However, in subtle neglect disorders it is important to stimulate both sides to confirm the diagnosis. When assessing and treating suspected neglect it is important to try to reduce or eliminate visual (and auditory) stimuli as far as possible, so that attention is not distracted from the required task. A busy out-patient department would make this almost impossible; a quiet room on a ward might be more suitable.

There is evidence to support visuo-spatial rehabilitation for deficits associated with VI after right hemisphere stroke.[37,38] Visual scanning training[39] has been shown to deliver improvements in VI. When advising the patient on visuo-motor cueing and scanning exercises to increase their awareness of the neglected side, it is important to stimulate as many pathways as possible, i.e. motor, sensory and proprioceptive.[2] It may be possible to ask the patient to place their left hand in left space as a target for scanning[2,40] and/or to move their hand/fingers or for the therapist to start passive hand/finger movements or to use tactile stimulation. Gradually moving the target further into the neglected side and scanning towards this[2] can be a simple bedside and home exercise programme that can be done with other professionals or with carers. Other visual stimuli can be used to increase attention and reduce neglect, such as using coloured vertical line guides[2] of high contrast and visibility on the left of the page when reading that give the patient a cue as to where to start reading from and are also a useful reminder. Using a typoscope can help focus the patient's attention towards the required part of the page and exclude unwanted visual 'noise' from the surrounding area.[41]

Other studies have used coloured stickers as a visual cue and reported a reduction in visual neglect.[42] Using a combination of scanning and cueing methods has been found to reduce visual neglect and this was transferable to some activities of daily living.[39,43] Contralesional limb activation or scanning and cueing methods resulted in a positive effect of reducing aspects of unilateral VI in some subjects, relative to subjects not treated.[40]

Prisms have been advocated for therapeutic use in both VI and hemianopia.[44,45] From the literature it is difficult to extract results from these treatments as both conditions are often included together in the research (possibly because they often coexist); also improvements on visual perception tests are not always transferable to activities of daily living.[46] However, prisms may still be useful, especially in patients with no insight into their difficulty, as they do not require voluntary orientation of attention to the affected side.[47] A short period of visuo-motor adaptation to a right prismatic shift was effective in improving VI.[44] Furthermore, there is evidence to suggest that long-term improvements can be maintained after treatment across a variety of visuo-spatial tasks,[46] and that the adaptation to prisms affects spatial representation in the brain,[44] although this was demonstrated in trials with small numbers. However, other studies have not confirmed the positive effects of prism adaptation on spatial neglect.[48]

The therapeutic use of mirrors has also been proposed. Placing a mirror in a sagittal plane to the patient's right (in left neglect) enables the left hemisphere to be visible to the patient. Some patients with VI, when asked to reach for an object placed on the left and reflected in the mirror to their right, grab the reflection or try to reach behind or through the mirror[49] – so-called mirror agnosia; however, when a mirror is presented in 'real space' patients recognise the object as a mirror. Normals rarely confuse a mirror image with the real image.[50] Some patients, however, are able to locate a reflected

object in left space,[51,52] and this has led to the theory that repeated exercises using mirrors to enable patients to locate objects to their left might be a useful therapeutic option for VI in those patients with mirror agnosia.[53] However, trials are based on small numbers or case studies and firm conclusions cannot be drawn.

Monocular occlusion is another strategy that has been used to treat VI. Retinal input from the eye is strongest to the contralateral superior colliculus.[54] Saccades to the left are generated by visual stimuli to the right superior colliculus and vice versa;[55] therefore if the right eye is occluded then, in theory, the remaining visual stimuli generate saccades to the left because of the remaining intact pathway to the now dominant right superior colliculus. This theory follows the belief that inhibition of the dominant side facilitates direction of attention to the remaining weaker side.[12] However, long-lasting effects of monocular occlusion are apparently limited to the time when the occlusion is worn.[56]

As VI often coexists with hemianopia, the aim in these patients is to improve neglect of the inattentive side and increase awareness of the limitations of their visual field defect. If successful, reading speeds and other activities of daily living can be enhanced, resulting in an overall improved outcome.

Recovery and therapy programmes

VI can adversely affect recovery of most activities of daily living, such as reading, dressing, navigation, walking, locating objects and drawing, and also may prevent the acquisition of any new motor skills.[2] VI can also limit the efficacy of rehabilitation programmes, often to a larger extent than more apparent deficits such as speech and motor deficits.[19] The role of the orthoptist in the assessment and management of these patients is therefore vital in the both early and long-term care. Establishing good communication links and liaison with other health professionals who manage patients with VI is fundamental for an optimum outcome.

Recovery of VI frequently occurs in the early post-acute stage and is greatest in the first month after a stroke.[26] Although some recovery does occur in most patients, a significant proportion will be left with persisting neglect.[14,26] This has been documented to be as high as 31.5%.[26] Specific data are lacking as to the extent and speed of recovery, although there is a greater recovery of patients with left brain damage than those with right damage.[2,21]

Treatment should be initiated as soon as is practically possible, in the early post-acute phase. However, there are no recommended guidelines on the length of a therapy programme, how long the average patient will take to improve, or which patients will benefit the most from any treatment. Selecting patients for treatment is, therefore, difficult. Patients with severe associated stroke-related cognitive deficit tend not to do well, whereas those with some insight into their difficulties, who can follow simple instructions, may be a more suitable choice for therapy. Patients who are to be discharged home or into the community with supportive carers, and who can continue home exercise programmes on a frequent basis, might do better.

Spatio-motor cueing and early emphasis on function can improve outcome and reduce the use of resources in patients with visual neglect.[57] Motor and functional recovery of stroke patients with neglect seems to be significantly improved by the simultaneous presence of a treatment specifically focused on neglect.[58] Patients with neglect require more assistance at discharge and have been found to need longer in-patient care and rehabilitation than those without neglect.[59–61] However, it is generally recognised that more research is needed to better define which treatment techniques are the most beneficial[62] and which patients will benefit the most from them.

In summary, unilateral visuo-spatial neglect or inattention is a common deficit following stroke that hinders rehabilitation and prolongs in-patient stay. It is defined as the failure to respond or attend to objects or people to one side of personal space. The right posterior cerebral cortex is frequently involved, resulting in the failure of left-orientating facility. Methods of assessment are largely based on drawing, copying or line cancellation tests, and are easily performed at the bedside. The experienced orthoptist will often suspect visual inattention on routine orthoptic testing before pencil and paper methods are employed. Performing a variety of tests for VI will give the most reliable results. Inexpensive therapeutic options are available with some success. These include scanning techniques and visuo-motor cueing into the affected side, prism adaptation, and limb activation in an attempt to increase awareness of the affected side and reduce inhibition by the healthy hemisphere. The recovery of stroke patients with VI is notably enhanced by treatment specifically targeted at neglect, although at present there is insufficient evidence to identify the most efficacious method. The neglect syndrome as a whole is an important predictor of poor functional outcome and, therefore, therapy focused on VI remains a priority for service providers.

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