Residual visual function in cortical vision loss

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Introduction

It is estimated that visual field loss occurs in 46% of patients with acquired brain damage[1], with homonymous hemianopia (measured with perimetry) present in 54% of all patients with stroke-related vision loss[2]. Counter-intuitively, when asked, some of these people may be able to look[3] or point[4] toward the location of objects in their blind field, while at the same time denying that they can 'see' them in any conventional sense. Some may report an awareness of moving objects on their blind side[5]. Some may even be able to catch objects that are thrown towards them, even in cases of full field vision loss[6]. In short, although unable to report the presence of perimetric luminance stimuli, some patients are able to make correct judgements about other visual features.

The existence of these residual visual abilities may lead patients to seek an explanation from their optometrist. Acknowledgement of the phenomenon can provide some reassurance to the patient, and knowledge of the visual pathways involved can also help to understand the location(s) of cortical damage underpinning vision loss. Here, we explain what residual visual abilities may remain in patients with acquired brain damage, as well as how knowledge of the relevant neural pathways aids understanding of the phenomena demonstrated by these patients.

Visual field loss following acquired brain injury

The major visual pathway relays signals from the retina to the primary visual cortex (striate cortex / V1; situated in the occipital lobe) via the lateral geniculate nucleus (LGN) in the thalamus[7]. This pathway is known as the primary visual, *geniculocortical*, or *geniculo-striate* pathway[8] (see Figure 1). Lesions to V1, or anywhere between the retina and V1, can result in vision loss[9]. The exact area of visual field loss resulting from brain damage depends on the location of the lesion[10,11]. Understanding this visual pathway enables clinicians to approximately localise neurological damage based on perimetric data. For example, unilateral damage to V1, the optic tract (the section of the pathway that relays information from the optic chiasm to the LGN) or the LGN itself can lead to contralateral homonymous hemianopia[9] (Figure 2b), whilst unilateral damage to Meyer's loop of the

optic radiations often results in homonymous quadrantanopia (Figure 2c). Clinically, these visual defects are characterised with perimetry. However, other visual pathways exist beside the geniculocortical pathway, and if they are spared, some perceptual abilities may remain.

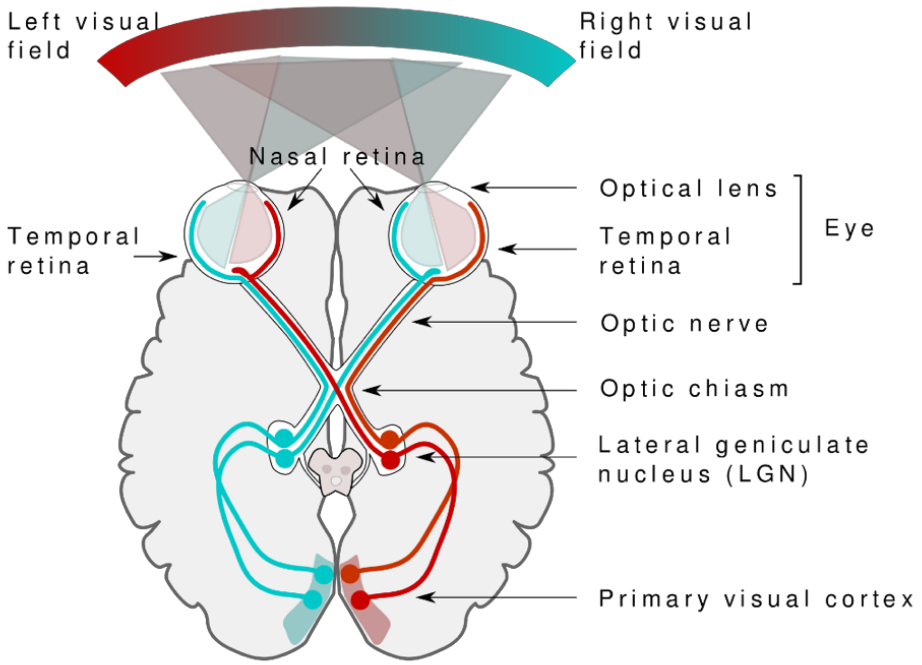

Figure 1: Illustration showing the primary visual (geniculocortical) pathway[12]. The optic radiations carry signals between the LGN and primary visual cortex.

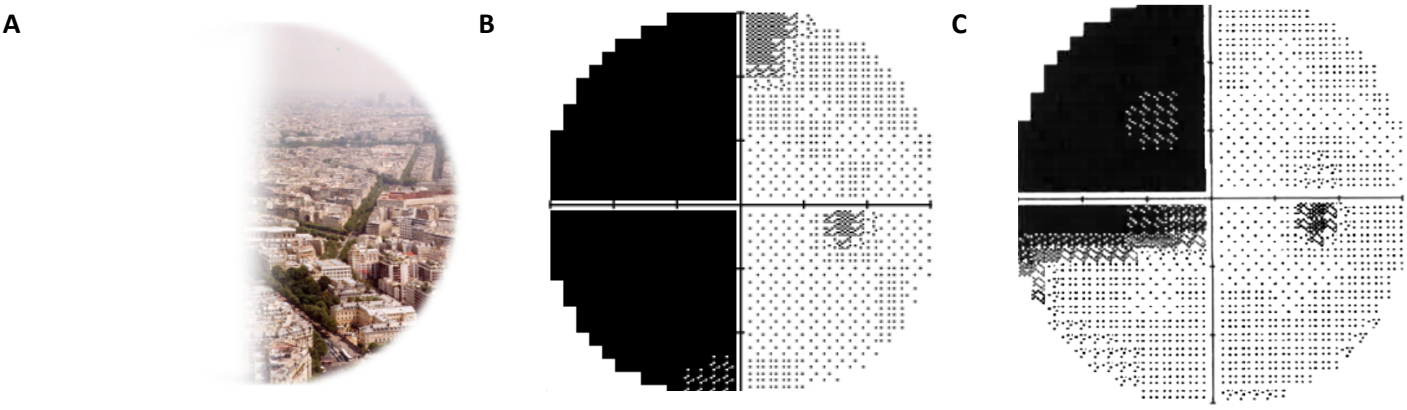

Figure 2: (A) Visual representation of a left hemianopic defect[13]. (B) Perimetric data showing a left hemianopia. (C) Perimetric data showing a left superior quadrantanopia.

Riddoch's phenomenon

The first recorded cases of residual vision following brain damage come from the early 1900s. George Riddoch noted that some soldiers with gunshot wounds affecting V1 could still perceive

visual motion in their blind field[14], despite being unable to characterise any attributes of visual stimuli, such as colour or shape. This became known as *Riddoch's phenomenon*[14]. This was later understood to be just one specific type of residual visual function displayed by those with cortical vision loss, as discussed below.

Blindsight

In the 1970s, research showed that individuals with hemianopia were able to localise the position of a visual target presented to their blind field, using a saccadic eye movement[3]. Subjects were told when a visual presentation was made, and instructed to move their eyes to look at the target location. The task initially puzzled subjects, with one asking "How can I look at something I haven't seen?" Although none of the participants reported 'seeing' a target, there was a clear relationship between gaze position and the target. These results came as a surprise to the subjects who would often insist they were simply "guessing". This phenomenon is known as *blindsight.* Below we describe two of the classic case studies.

Case studies

Much of our knowledge of the functionality of the 'blind' striate and extra-striate cortices is derived from a series of early case studies involving a patient with hemianopia known as D.B.

Case Study 1

Patient D.B. – 34 years old at the time of first publication[4]

D.B. had an arteriovenous malformation at the right occipital pole which was causing vomit-inducing headaches that could last up to 48 hours. These headaches also caused significant disruption to his vision. They were preceded by flashing lights appearing in an oval-shaped cluster to the left of his fixation; after 15 minutes these lights developed into a large oval-shaped white scotoma. After some time, the scotoma would enlarge and include coloured lights. At the age of 33, the arteriovenous malformation was surgically removed, resulting in a dense left homonymous hemianopia.

Despite having homonymous hemianopia, D.B. could make accurate saccadic eye movements toward 'unseen' targets (as also shown in other patients[3]). D.B. was also able to locate visual stimuli in the blind field by reaching with his finger[4], with an average error of only 3.8°. It is important to note that D.B. had no awareness of these stimuli but was forced to guess. This series of tasks was the first robust and explicit measure of residual visual abilities in the absence of conscious awareness.

Case Study 2

Patient G.Y. – aged 22 years old at the time of the first publication (1980)[15]

G.Y. was involved in a road traffic accident at the age of eight, resulting in significant trauma to the left hemisphere. The damage rendered G.Y. with a dense right homonymous hemianopia with macular sparing. The region of spared vision extended 3° into his blind side. MRI showed almost total destruction of V1 with little-to-no damage to extrastriate areas.

Patient G.Y. offers further insights into residual visual function. Interestingly, despite not being consciously aware of videos of faces presented to his blind hemifield, G.Y. was able to discriminate between the different emotions in the faces shown (happy, sad, angry, fearful)[16]. It is worth noting

that the faces were shown in this study as videos; therefore it is possible that motion cues could have contributed to the perception of emotion.

Classification of blindsight

Weiskrantz – one of the pioneers of blindsight research – originally separated blindsight into two categories[17]. 'Type I' blindsight was defined as lacking any conscious awareness, while 'type II' was more akin to Riddoch's phenomenon, i.e. some awareness is present. More recently, Danckert and Rossetti proposed a new taxonomy of blindsight, suggesting three distinct sub-groups; *action-blindsight*, *attention-blindsight* and *agnosopsia* (see Table 1)[18].

Table 1: Summary of the sub-types of blindsight and associated responses

	Type I blindsight	**Type II blindsight**	
Sub-type	**Agnosopsia**	**Action-blindsight**	**Attention-blindsight**
Observable behaviour	Form and wavelength discrimination	Action based responses, saccades, motor responses, grasping	Motion detection, higher-level discrimination
Responses	Reflexive, forced-choice guessing	Direct responses toward stimulus	Implicit, explicit, forced-choice guessing
Level of awareness of stimulus	None	Low	Moderate

Patients who are able to accurately point or make an eye movement toward an object, but are unable to describe or distinguish any other visual characteristics of that object, can be considered to have *action-blindsight*, i.e. they can generate an action in response to a stimulus, with very little conscious awareness of what that stimulus is. However, if the patient can detect the direction of motion, or discriminate between two stimuli presented to their blind field, they are considered to have *attention-blindsight*. These patients are consciously aware of stimuli, unlike those with action-blindsight. It is essential to note that although attention-blindsight implies some conscious awareness or visual sensation in response to stimulus presentation, it is quantifiably distinct from a normal state of unimpaired conscious vision[18] (which is known as *gnosopsia*). The third sub-type of blindsight is one that lacks all conscious perception of blind field stimulation, known as *agnosopsia*,

which means “not knowing what one sees”[5]. Residual visual function in these patients can only be assessed through reflexive responses and/or forced-choice paradigms, and the patient will never experience or report seeing a stimulus in their blind field. The patient with *agnosopsia* will not be able to direct an action towards a stimulus, nor will they be able to describe any visual characteristics such as form or motion. They simply make visual judgements with above-chance accuracy.

Alternative visual pathways

Advances in magnetic resonance imaging (MRI) are beginning to tease apart the neural underpinning of some of these residual visual abilities. A very recent study demonstrated, in patients capable of discriminating motion in a ‘blind’ hemifield, connectivity between the LGN and hMT+; the area of the cortex implicated in motion processing in humans[32]. Residual motion perception in the absence of an ability to characterise shape or form might, therefore, be expected if the hMT+ region is spared in an individual with damage confined to V1.

One promising study has shown that the existence of blindsight in patients with cortical vision loss can often be predicted by observing subtle pupil size changes (as measured using a pupillometer) in response to the presentation of isoluminant gratings in the blind field[19].

Investigation of various aspects of visual function may help approximate the site of damage, as anomalies of only one function suggest localised damage, whereas anomalies of multiple functions might suggest more widespread damage. Findings would also help to provide some explanation to the patient regarding his/her symptoms, and aid in any referral for further neurological assessment.

As the pupil pathway is considered to be non-cortical (Figure 3), post-geniculate damage should not, in theory, affect the pupillary light reflex. However, the pupil response is both slowed and reduced in patients with hemianopia due to optic tract damage[20] and in those with homonymous hemianopia as

a result of stroke affecting the occipital lobe[21]. This observation challenges the classic view that the pupillary light reflex is a purely subcortical pathway[22].

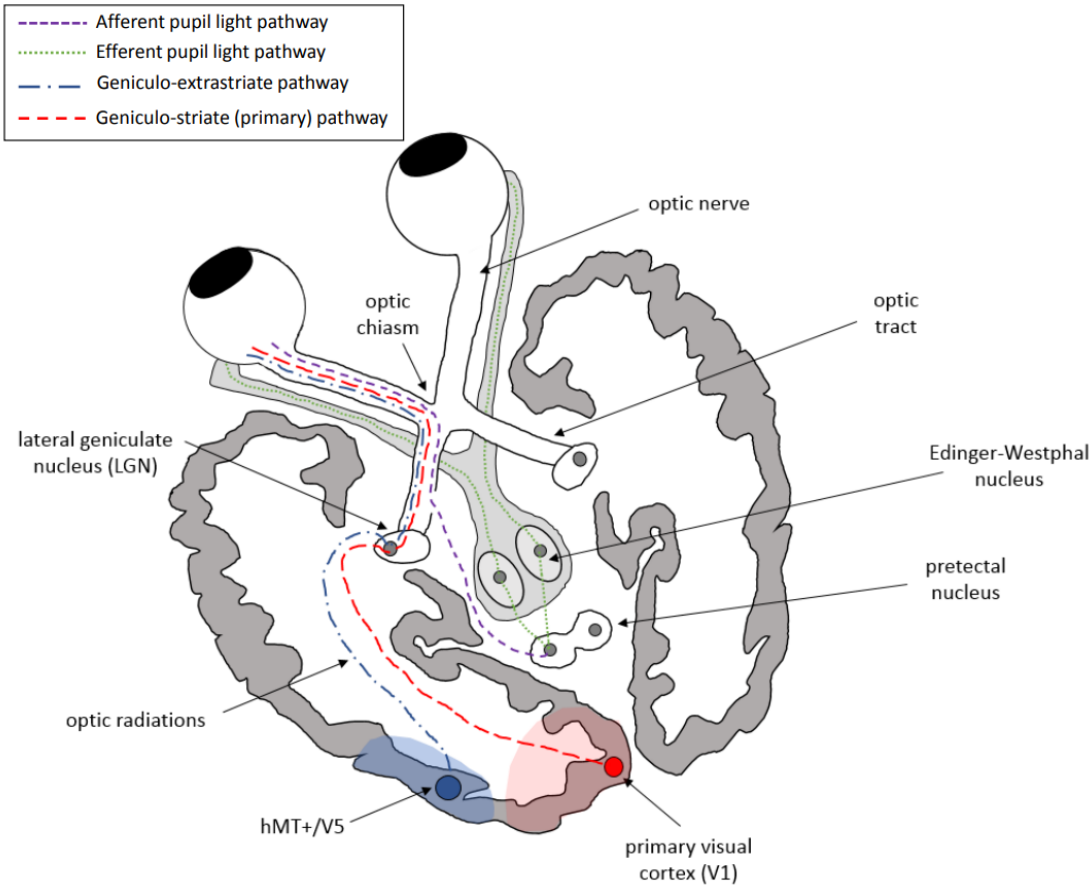

Figure 3: An illustration of the pupillary light reflex pathway, as well as the geniculo-striate pathway and geniculo-extrastriate pathway to hMT+. The afferent pupil signal (purple dashes) travels from the retina to the pretectal nucleus and then to both Edinger-Westphal nuclei. Green dots (efferent pathway) show the projection from the Edinger-Westphal nuclei to the ciliary ganglia via the oculomotor nerve. The ciliary ganglia innervate the sphincter pupillae muscles, resulting in pupillary constriction.

Training blindsight for rehabilitation

It is not clear how useful residual vision is in everyday visual activities. However, it has long been known that visual perception can be enhanced through repeated exposure to particular visual stimuli; a process known as 'perceptual learning'. Researchers have demonstrated that residual visual function can be similarly enhanced through training. For example, patients with unilateral post-geniculate lesions are better able to detect flickering grating stimuli in their blind field after training[23]. Patients have also been shown to recover some ability to discriminate the direction of visual motion[24]. This research has led to the development of formalised rehabilitation

programmes, based on the premise that increased visual sensitivity to moving or flickering stimuli should translate into improvements in everyday visual function.

Summary

Acquired brain damage directly affecting V1 can cause a phenomenon in which conscious vision is affected, but other aspects of function, processed via separate pathways, may be preserved. This can lead to the ability to make correct judgements about some aspects of a visual scene, despite lacking conscious visual awareness. An understanding of these phenomena and the pathways involved in processing visual stimuli will enable clinicians to provide a tentative explanation of symptoms to patients and determine the most appropriate management.

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Multiple choice questions

1. Unilateral damage to *Meyer's loop* often results in…
    - **a. homonymous quadrantanopia**
    - b. bitemporal hemianopia
    - c. homonymous hemianopia
    - d. complete cortical blindness
2. Which of the following terms is **not** used to refer to the major visual pathway involving the LGN and V1?
    - a. Primary visual pathway
    - b. Geniculocortical pathway
    - c. Geniculo-striate pathway
    - **d. Retinotectal pathway**
3. The pupillary light reflex signal travels from the pretectal nucleus to…
    - a. the ipsilateral Edinger-Westphal nucleus only
    - b. the contralateral Edinger-Westphal nucleus only
    - **c. both Edinger-Westphal nuclei**
    - d. the ipsilateral hMT+ only
4. If a patient can accurately make a saccade to a visual stimulus presented in their blind field but cannot discriminate any characteristics of the stimulus (such as shape or colour) they can be considered to have…
    - **a. action-blindsight**
    - b. attention-blindsight
    - c. agnosopsia
    - d. gnosopsia
5. Which of the following best approximates the geniculocortical pathway?
    - a. Retina → optic tract → superior colliculus → extrastriate cortex
    - b. Retina → optic tract → LGN → extrastriate cortex
    - c. Retina → optic tract → hMT+ → V1
    - **d. Retina → optic tract → LGN → V1**
6. *Riddoch's phenomenon* refers to the ability to…
    - a. discriminate the emotional expression of faces presented to the blind field
    - **b. detect the presence of a moving stimulus in the blind field**

c. detect the presence of a static stimulus in the blind field
d. discriminate the orientation of lines presented to the blind field

7. A patient with homonymous hemianopia shows an above-chance ability to discriminate the *direction* of visual motion in their blind field. They are displaying…
    a. Riddoch's phenomenon
    b. attention-blindsight
    c. action-blindsight
    d. type 1 blindsight
8. What is the name given to the normal state of unimpaired vision, in which individuals are consciously aware of, and able to make discriminations between, visual stimuli?
    a. Anopsia
    b. Gnosanopsia
    c. Agnosopsia
    d. Gnosopsia