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1 **Residual visual function in cortical vision loss**

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18 [in-cortical-vision-loss/article](https://www.aop.org.uk/ot/CET/2019/02/12/residual-visual-function-in-cortical-vision-loss/article)

## 19 Introduction

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

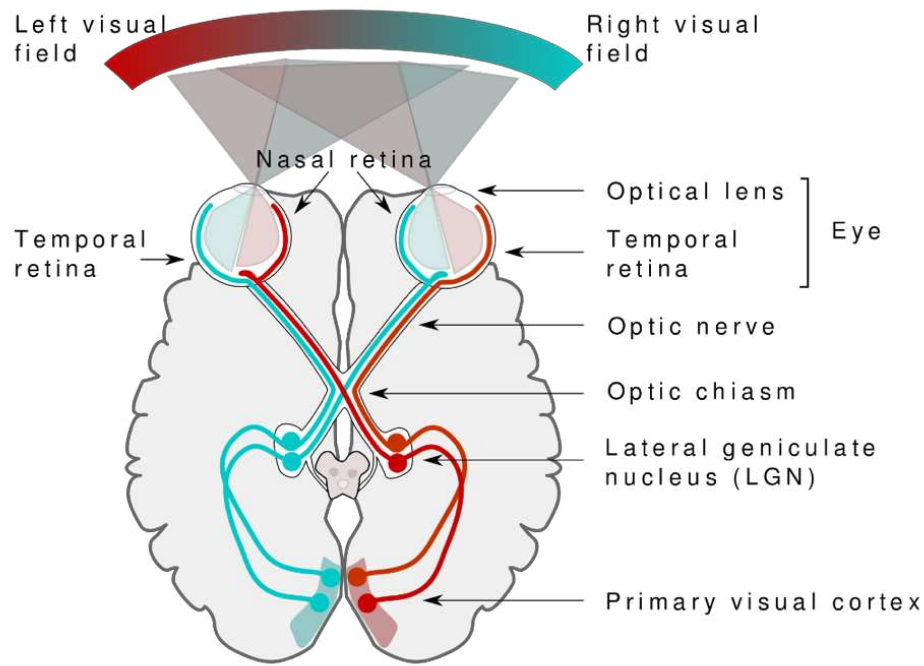
28 The existence of these residual visual abilities may lead patients to seek an explanation from their  
29 optometrist. Acknowledgement of the phenomenon can provide some reassurance to the patient,  
30 and knowledge of the visual pathways involved can also help to understand the location(s) of  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

## 34 Visual field loss following acquired brain injury

35 The major visual pathway relays signals from the retina to the primary visual cortex (striate cortex /  
36 V1; situated in the occipital lobe) via the lateral geniculate nucleus (LGN) in the thalamus<sup>7</sup>. This  
37 pathway is known as the primary visual, *geniculocortical*, or *geniculo-striate* pathway<sup>8</sup> (see Figure 1).  
38 Lesions to V1, or anywhere between the retina and V1, can result in vision loss<sup>9</sup>. The exact area of  
39 visual field loss resulting from brain damage depends on the location of the lesion<sup>10,11</sup>.

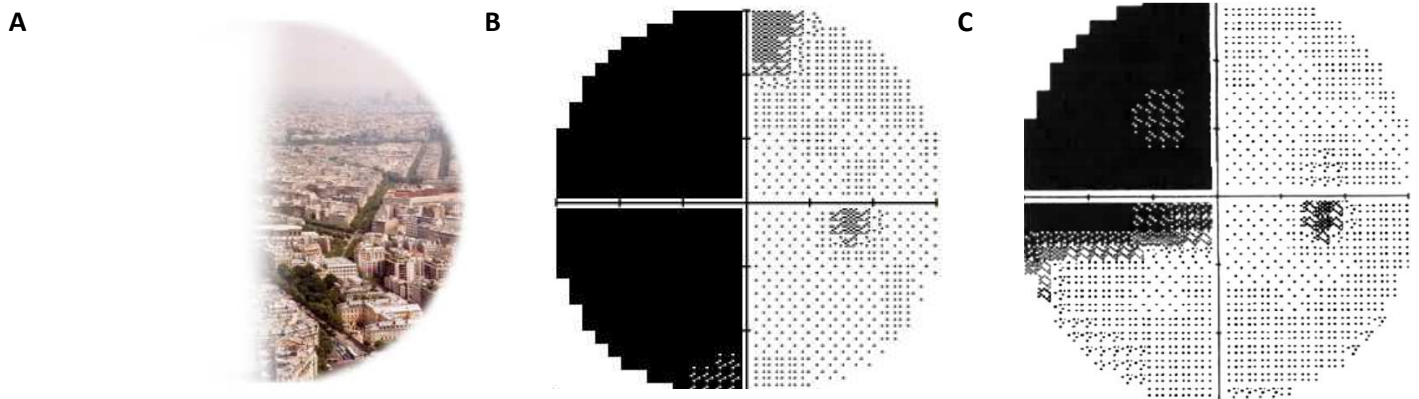
40 Understanding this visual pathway enables clinicians to approximately localise neurological damage  
41 based on perimetric data. For example, unilateral damage to V1, the optic tract (the section of the  
42 pathway that relays information from the optic chiasm to the LGN) or the LGN itself can lead to  
43 contralateral homonymous hemianopia<sup>9</sup> (Figure 2b), whilst unilateral damage to Meyer's loop of the

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



47

48 *Figure 1: Illustration showing the primary visual (geniculocortical) pathway<sup>12</sup>. The optic radiations carry signals*  
 49 *between the LGN and primary visual cortex.*



50 *Figure 2: (A) Visual representation of a left hemianopic defect<sup>13</sup>. (B) Perimetric data showing a left hemianopia.*  
 51 *(C) Perimetric data showing a left superior quadrantanopia.*

52 **Riddoch's phenomenon**

53 The first recorded cases of residual vision following brain damage come from the early 1900s.

54 George Riddoch noted that some soldiers with gunshot wounds affecting V1 could still perceive

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

## 59 Blindsight

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

## 68 Case studies

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

**Patient D.B. – 34 years old at the time of first publication<sup>4</sup>**

**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.**

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

**Patient G.Y. – aged 22 years old at the time of the first publication (1980)<sup>15</sup>**

**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.**

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



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

#### 84 Classification of blindsight

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

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

Sub-type	Type I blindsight	Type II blindsight	
	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

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

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

### 107 [Alternative visual pathways](#)

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

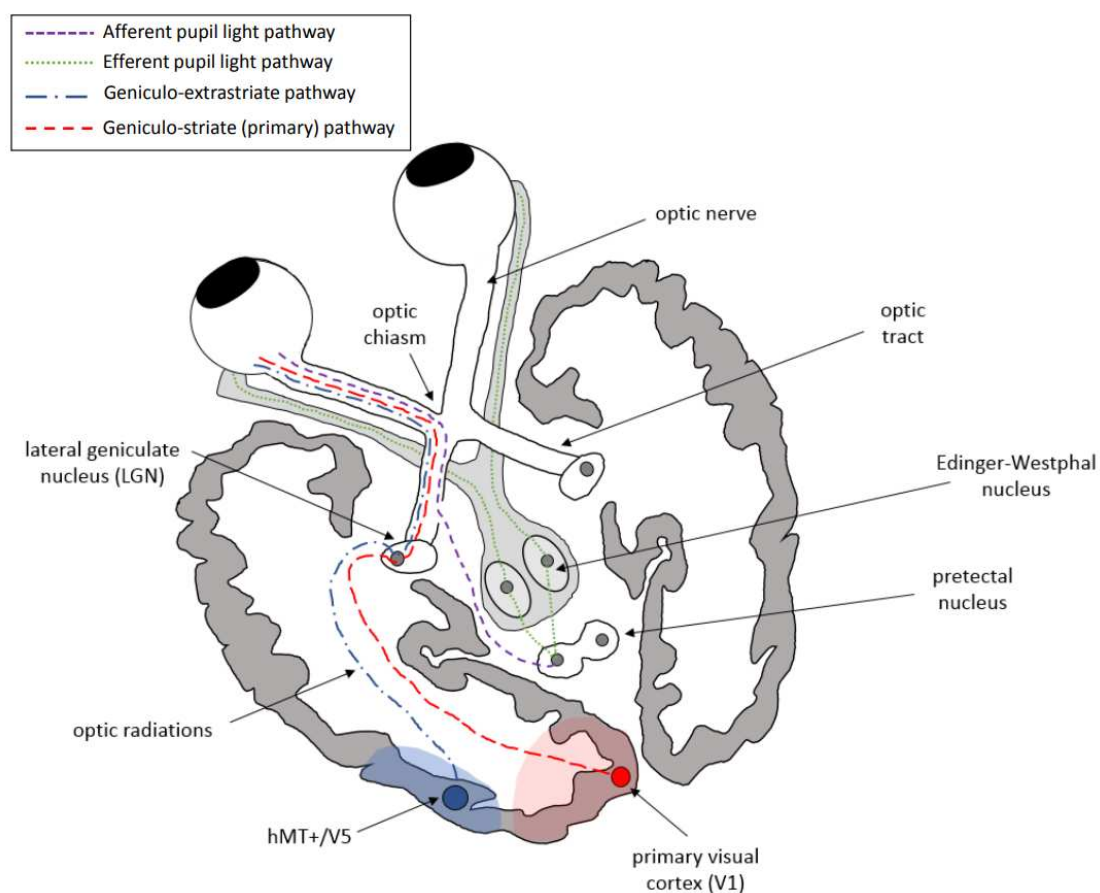
114 One promising study has shown that the existence of blindsight in patients with cortical vision loss  
115 can often be predicted by observing subtle pupil size changes (as measured using a pupillometer) in  
116 response to the presentation of isoluminant gratings in the blind field<sup>19</sup>.

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

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



124 a result of stroke affecting the occipital lobe<sup>21</sup>. This observation challenges the classic view that the  
125 pupillary light reflex is a purely subcortical pathway<sup>22</sup>.



126  
127 *Figure 3: An illustration of the pupillary light reflex pathway, as well as the geniculo-striate pathway and*  
128 *geniculo-extrastriate pathway to hMT+. The afferent pupil signal (purple dashes) travels from the retina to the*  
129 *pretectal nucleus and then to both Edinger-Westphal nuclei. Green dots (efferent pathway) show the projection*  
130 *from the Edinger-Westphal nuclei to the ciliary ganglia via the oculomotor nerve. The ciliary ganglia innervate*  
131 *the sphincter pupillae muscles, resulting in pupillary constriction.*

### 132 Training blindsight for rehabilitation

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

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

## 142 Summary

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

### The Neurological Vision Loss (NVL) Panel

**Researchers at Cardiff University are currently seeking to recruit research participants for studies of neurological vision loss – in particular, people with homonymous hemianopia, to further clinical understanding of residual vision. Further information for anyone interested in taking part in this research can be found at [psych.cf.ac.uk/home2/nvl](https://psych.cf.ac.uk/home2/nvl).**

149

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- 200

201 **Multiple choice questions**

- 202 1. Unilateral damage to *Meyer's loop* often results in...
- 203 **a. homonymous quadrantanopia**
- 204 b. bitemporal hemianopia
- 205 c. homonymous hemianopia
- 206 d. complete cortical blindness
- 207 2. Which of the following terms is **not** used to refer to the major visual pathway
- 208 involving the LGN and V1?
- 209 a. Primary visual pathway
- 210 b. Geniculocortical pathway
- 211 c. Geniculo-striate pathway
- 212 **d. Retinotectal pathway**
- 213 3. The pupillary light reflex signal travels from the pretectal nucleus to...
- 214 a. the ipsilateral Edinger-Westphal nucleus only
- 215 b. the contralateral Edinger-Westphal nucleus only
- 216 **c. both Edinger-Westphal nuclei**
- 217 d. the ipsilateral hMT+ only
- 218 4. If a patient can accurately make a saccade to a visual stimulus presented in their
- 219 blind field but cannot discriminate any characteristics of the stimulus (such as
- 220 shape or colour) they can be considered to have...
- 221 **a. action-blindsight**
- 222 b. attention-blindsight
- 223 c. agnosopsia
- 224 d. gnosopsia
- 225 5. Which of the following best approximates the geniculocortical pathway?
- 226 a. Retina → optic tract → superior colliculus → extrastriate cortex
- 227 b. Retina → optic tract → LGN → extrastriate cortex
- 228 c. Retina → optic tract → hMT+ → V1
- 229 **d. Retina → optic tract → LGN → V1**
- 230 6. *Riddoch's phenomenon* refers to the ability to...
- 231 a. discriminate the emotional expression of faces presented to the blind field
- 232 **b. detect the presence of a moving stimulus in the blind field**

- 233                   c. detect the presence of a static stimulus in the blind field
- 234                   d. discriminate the orientation of lines presented to the blind field
- 235       7. A patient with homonymous hemianopia shows an above-chance ability to
- 236       discriminate the *direction* of visual motion in their blind field. They are
- 237       displaying...
- 238                   a. Riddoch's phenomenon
- 239                   **b. attention-blindsight**
- 240                   c. action-blindsight
- 241                   d. type 1 blindsight
- 242       8. What is the name given to the normal state of unimpaired vision, in which
- 243       individuals are consciously aware of, and able to make discriminations between,
- 244       visual stimuli?
- 245                   a. Anopsia
- 246                   b. Gnosanopsia
- 247                   c. Agnosopsia
- 248                   **d. Gnosopsia**
- 249