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Abnormal visual field maps in human cortex: A mini-review and a case report

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ABSTRACT

Human visual cortex contains maps of the visual field. Much research has been dedicated to answering whether and when these visual field maps change if critical components of the visual circuitry are damaged. Here, we first provide a focused mini-review of the functional magnetic resonance imaging (fMRI) studies that have evaluated the human cortical visual field maps in the face of retinal lesions, brain injury, and atypical retinocortical projections. We find that there is a fair body of research that has found abnormal fMRI activity, but also that this abnormal activity does not necessarily stem from cortical remapping. The abnormal fMRI activity can often be explained in terms of task effects and/or the uncovering of normally hidden system dynamics. We then present the case of a 16-year-old patient who lost the entire left cerebral hemisphere at age three for treatment of chronic focal encephalitis (Rasmussen syndrome) and intractable epilepsy. Using an fMRI retinotopic mapping procedure and population receptive field (pRF) modeling, we found that (1) despite the long period since the hemispherectomy, the retinotopic organization of early visual cortex remained unaffected by the removal of an entire cerebral hemisphere, and (2) the intact lateral occipital cortex contained an exceptionally large representation of the center of the visual field. The same method also indicates that the neuronal receptive fields in these lateral occipital brain regions are extraordinarily small. These features are clearly abnormal, but again they do not necessarily stem from cortical remapping. For example, the abnormal features can also be explained by the notion that the hemispherectomy took place during a

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critical period in the development of the lateral occipital cortex and therefore arrested its normal development. Thus, caution should be exercised when interpreting abnormal fMRI activity as a marker of cortical remapping; there are often other explanations.

1. Introduction

Human visual cortex contains several maps of the visual field (Wandell et al., 2005, 2007). Within each of these visual field maps, cortical neurons respond to stimuli in a limited region of the visual field, the neuronal receptive field, and neighboring neurons respond to neighboring regions of visual space. If critical components of the visual system are damaged, some visual neurons may change their receptive field profile (Kaas et al., 1990, 2002; Heinen and Skavenski, 1991; Chino et al., 1992; Gilbert and Wiesel, 1992; Darian-Smith and Gilbert, 1995; Calford et al., 2005; Giannikopoulos and Eysel, 2006). These neuronal receptive field changes result in abnormal visual field maps. A method that has gained increased popularity over the past decade to infer the presence or absence of an abnormal visual field map is functional magnetic resonance imaging (fMRI). This mini-review therefore concentrates on the fMRI studies of abnormal visual field maps in humans. By stimulating specific parts of the visual field one can evaluate whether stimulus-locked fMRI activity is present or absent at an expected cortical location. For example, when the center of the retina is damaged, a normal visual field map should no longer respond to stimuli presented at the center of the visual field. Such a silent zone is usually referred to as the lesion projection zone or LPZ. Another way to infer the presence or absence of an abnormal visual field map is by explicitly evaluating the layout of the map using an experimental paradigm called retinotopic mapping (Engel et al., 1994, 1997; DeYoe et al., 1994, 1996; Sereno et al., 1995; Wandell and Winawer, 2011; Engel, 2011).

2. Abnormal visual field maps following retinal lesions

Let us start with the abnormal visual field maps that were found in patients with retinal lesions due to inherited photoreceptor abnormalities (Baseler et al., 2002). In a condition called rod-monochromacy, a genetic deficit causes the malfunctioning of the cone photoreceptor cells in the retina. Because the most central portion of the human retina contains only these cone cells, the patient is left with a small but permanent scotoma (blind spot) at the center of his or her visual field. In addition, the cortical regions that would normally be driven by this central portion of the retina, the LPZ, are now deprived from inputs. Interestingly, however, the LPZ was not entirely unresponsive, as would be expected if the visual field maps in these rod-monochromats were normal. Instead, it was found that the LPZ was now driven by those parts of the visual field directly surrounding the scotoma, suggesting that the receptive fields of the cortical neurons inside the LPZ had shifted away from their original location. Importantly, these abnormal responses could not be detected in normal control subjects in whom the retinal lesions were simulated by performing the same experiment but stimulating only their rod photoreceptor cells. It is therefore, that the abnormal visual field maps in rod-monochromats are taken as evidence for cortical reorganization.

Abnormal LPZ responses can also be recorded in patients who acquired bilateral scotomas later in life, for example as a result of the eye-disease macular degeneration (MD). However, there is substantial controversy as to whether these responses can also be taken as evidence for cortical reorganization. Abnormal responses in MD patients were first presented by Baker et al. (2005). In this study, the authors presented a series of face, object and scene images to the intact, peripheral portions of the patients' visual field while requiring them to indicate whether the current image was the same as the one presented immediately before (one-back task). Interestingly, Baker et al. found that the activity in response to these peripherally presented stimuli was not only located in the expected regions of cortex that normally represent the periphery of the visual field, but also inside the patients' LPZ. Like the abnormal responses found in rod-monochromats, these abnormal responses were not present in normal controls, which is why the abnormal responses in the LPZ were taken to "demonstrate large-scale reorganization of visual processing in humans with MD" (Baker et al., 2005, p. 614).

Given their experimental paradigm, however, there are also mechanisms other than reorganization that could potentially explain the abnormal LPZ responses reported by Baker et al. (2005). For example, it could not be ruled out that the abnormal LPZ activations were caused by the task rather than the stimulus. This in turn opened up the possibility that the differences between patients and controls were due to differences in the strength of the top–down feedback signals from higher-order visual cortex. To address this issue, Masuda et al. (2008) examined the abnormal LPZ responses in more detail, testing whether these responses were also present when MD patients performed a task that was not directed to the face stimuli, or when there was no task at all. In addition, the authors asked whether the abnormal responses could be due to the fact that the stimuli were meaningful by repeating the experiment using a simple checkerboard pattern instead of faces. The authors found that the abnormal responses observed by Baker et al. (2005) were indeed a consequence of the task rather than the stimulus or how meaningful they were. Similar task-dependent LPZ responses were also found some years later, in patients with another eye-disease, retinitis pigmentosa (Masuda et al., 2010).

Because the Masuda studies questioned the idea that cortical reorganization is responsible for the abnormal