

Supplementary Note

More detailed description of MM's subjective experiences can be found on Mike May's Perceptions Home Page, <http://www.senderogroup.com/perception.htm>

Psychophysics: The spatial CSF was measured using vertically oriented sinusoidal gratings within a rectangular aperture of 50×37 degrees of visual angle at a distance of 15 inches. Gratings had a mean luminance of 19.7 cd/m² and were temporally modulated by a 1 Hz square wave. Each grating was presented in a fixed order, and was presented between four and eight times during each session. Both forced choice and adjustment techniques were used. During adjustment procedures the contrast of each grating was adjusted using a trackball mouse until the orientation was just visible. The variance of MM's settings was the same or less than that of normal observers. Two control observers were tested in addition to MM.

Non-grating stimuli in the behavioral experiments were presented on a high contrast monitor at a viewing distance of at least 30 cm, and were sized so as to subtend about 30 degrees of visual angle. Stimuli presented to control observers (inside and outside the scanner) were blurred using a filter designed to match MM's spatial resolution. Filtering was carried out by convolving the stimulus image with a filter with a high pass cutoff of 1c.p.d. (slightly less than MM's psychophysically measured resolution limit). Behavioral responses were collected from three control observers for most of the stimuli presented to MM. In a few cases the task was trivially simple for visually normal observers using both blurred and unblurred stimuli and formal testing was omitted.

MM was always given 2–10 practice trials (sometimes with longer presentation durations than during the actual test) where the correct answer was given and the instructions were clarified before responses were recorded. MM was then presented with a series of at least four stimuli on at least two separate testing sessions. 4-8 repetitions were carried for tasks g–j and n. 20 or more repetitions were carried out on the remaining tasks. A variety of experimental procedures were used, including forced choice (FC) yes/no (b-d, l, m), adjustment, identification (e, h, i, n, o) tasks and FC identification tasks (where MM was asked to choose among a limited set of possible responses; f, g, j, k). In many of the tasks MM found the task trivial and was correct 100% of the time. In the illusory contour and perspective tasks (Fig. 2e,i) MM completely failed to interpret the stimulus and was very reluctant to give a response. In tasks where consistency of judgments could be measured (such as measuring contrast sensitivity) MM's responses were at least as consistent as those of control observers. In face and object discrimination tasks high contrast images of faces and objects were used (as in earlier studies¹). Images subtended about 30 degrees of visual angle and were presented for 2 seconds. Control observers were presented with the same set of (blurred) faces. Observers were asked to judge the expression and the gender of each face. Two sets of standard face stimuli, and two sets of standard object stimuli were used similar to those used in earlier fMRI studies¹. In the full set of faces available to us there were several

faces for which observers were inconsistent in discriminating happy v. neutral or sad vs. neutral, and these faces were excluded from our analysis. After measuring identification performance for objects we asked MM whether he had visually experienced that object post-operatively, and excluded objects that he might never have seen from our analysis.

FMRI Imaging: Functional images were acquired with a spiral k-space sampling sequence on a 3T GE Signa system. Data were analyzed using in-house software²⁻⁵. FMRI response was quantified as the phase and amplitude of the sinusoid that best fit the average time-series of voxel responses averaged across a given cortical region of interest and projected onto a unit vector with an angle representing the hemodynamic delay⁶. We attempted to define MM's visual areas using standard retinotopic mapping^{3,7} and cortical flattening techniques⁸, carried out during six separate retinotopy sessions on six different days. Every session contained both rotating wedges and expanding ring stimuli, with duty cycles of 45, 90 or 180 degrees and a field size of diameter 32 degrees of visual angle (23 coronal slices, TR, 3s; voxel size, 1.2 x 1.2 x 3 mm). Eccentricity mapping (responses to expanding rings) showed a broadly similar pattern to that obtained in large numbers of visually normal observers in our laboratory using similar or identical procedures^{3,8,9}. However MM's activation covered a much smaller region of cortex. Angular responses (rotating wedges) also activated a smaller region than those activated in visually normal observers, and also showed abnormal mapping of polar angle. Because MM's pattern of phase reversals within the occipital pole consistently did not match the pattern usually found for such stimuli our definition of MM's V1 is based on both anatomical¹⁰ and functional criteria.

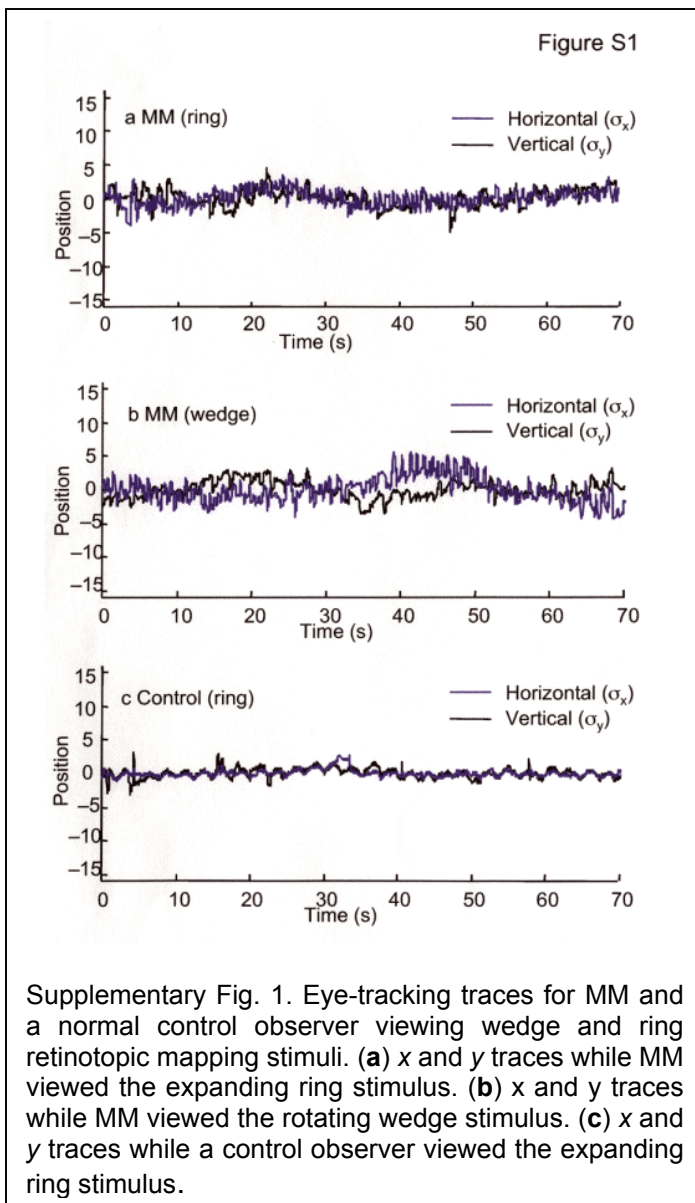
Areas V2, and V3 could not be identified, suggesting either weak responsivity or disrupted retinotopic organization in occipital areas beyond V1. MM also showed little foveal activity, but this may have been due to unstable fixation (see below). Activity patterns were reproducible across multiple sessions. The mean correlation coefficient between scans for eccentricity values along the left calcarine sulcus were MM=0.59, control=0.69. For angular values along a contour of constant eccentricity in the left hemisphere correlation coefficients were MM=0.63, control=0.81. MT+ was identified by measuring fMRI responses to 0.5 deg radius white dots (field size, 32 degrees) that alternated between moving (8 deg/sec radially inward and outward) and stationary on a black background¹¹. We noted evidence of retinotopic phase map encoding in MT/MST, as has been found in visually normal observers¹².

Neural contrast response functions were measured using stimuli alternating between vertical 1 Hz contrast reversing sinusoidal gratings and a blank field of the same mean luminance in an on-off 32 second block design (23 coronal slices, TR, 3s; voxel size, 1 x 1 x 3 mm). The visual field was 32 degrees of visual angle. Cortical areas sensitive to images of faces and objects were identified by measuring fMRI responses to stimuli alternating between images of faces vs. phase scrambled faces, objects vs. phase scrambled objects, faces vs. objects and faces vs. blank. Each image was presented for 2 seconds in an on-off 32-second block design (23 coronal slices, TR, 3s; voxel size, 1.2 x 1.2 x 6 mm). Faces and objects were the same as were used for the psychophysical judgments. Two control observers were tested in addition to MM. We found little or no significant activation within MM's lingual and fusiform gyri in response to the face images and only small amounts of activation to the object images. Nor

did MM show activity in lingual or fusiform gyri to either the face vs. blank condition or other on-off stimulus conditions, suggesting that fusiform and lingual regions were unresponsive rather than unspecialized.

Eyetracking: We measured MM's ability to fixate while viewing the retinotopy stimulus to confirm that unstable fixation was not responsible for MM's anomalous data.

Eyetracking was performed outside the magnet in a "simulator" where the viewing distance and stimulus display was designed to mimic the MRI environment. Fig. S1 a-c shows example fixation eye-traces for MM and a visually normal control during presentation of rotating wedge and expanding ring stimuli. As in the scanner, the stimulus subtended 32 degrees of visual angle. The standard deviations of MM's horizontal and vertical eye position when asked to fixate centrally were $\sigma_x=1.90$ and $\sigma_y=1.30$ degrees



during presentation of the rotating wedge stimulus, and $\sigma_x=1.10$ and $\sigma_y=1.24$ degrees during presentation of the ring stimulus. In comparison, the standard deviations of our control (IF) were fairly typical: $\sigma_x=0.46$ and $\sigma_y=0.52$ degrees for the wedge stimulus, and $\sigma_x=0.55$ and $\sigma_y=0.68$ degrees for the ring stimulus. While MM's fixation was less stable than visually normal observers, his anomalous retinotopic mapping cannot be explained by fixation instabilities or an offset in fixation. Fixation instabilities may have disrupted measurements of retinotopy in the parafovea (where retinotopy is generally poor), but not within the periphery. Measurements of cortical magnification made in our laboratory¹³ predict that while 1 degree of visual angle corresponds to approximately 7mm of cortex at 1.5 degrees eccentricity, at 6 degrees eccentricity 1 degree of visual angle only corresponds to 2.5mm of cortex. By 12 degrees eccentricity, 1 degree of visual angle corresponds to about 1 mm in the cortex.

ERG: We carried out standard clinical electroretinograms (ERGs) to measure responses in the rod pathways (after 20 minutes of dark adaptation) and cone pathways (under normal conditions of light adaptation). For medical reasons these ERGs were carried out using skin electrodes rather than the standard corneal electrodes, so the obtained amplitudes were much lower than those obtained using corneal electrodes. A Nicolet Bravo system, referenced to FpZ, was employed in conjunction with skin electrodes and a Ganzfield bowl was used for stimulus presentation. Testing was performed with the pupils dilated. An experienced physician who was naïve as to the purpose of the test carried out the analysis of the ERG results. The pattern-reversed ERG under both scotopic and photopic conditions produced reproducible responses within normal limits. Under scotopic conditions the amplitude of the B-waves was 58 microvolts to blue flashes, 20 microvolts to red flashes and 102 microvolts to white flashes. The flicker response was present. While MM's electroretinogram responses were within normal limits¹⁴ it should be noted that a normal ERG response can be obtained even when a considerable proportion (more than 50%) of the retina is disturbed, so obtaining a normal ERG for MM cannot completely exclude the possibility of retinal degeneration. It should also be noted that his retina was clearly visible through an ophthalmoscope, with no visual indications of retinal degeneration as a consequence of deprivation¹⁵.

VEP: A standard clinical visual evoked potential (VEP) was performed using high contrast flickering checkerboard stimulus with check sizes of 0.45, 0.9 and 1.48 degrees of visual angle. Standard skin electrodes were used. An experienced physician who was naïve as to the purpose of the test carried out the analysis of the VEP results. The largest checks produced reproducible positive peaks with a latency of 90 milliseconds and amplitude of 1.68 microvolts. P100 responses to the smaller checks were poorly reproducible and of low amplitude, suggesting little or no neural response.

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