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On the presence of three cone mechanisms in a case of total achromatopsia

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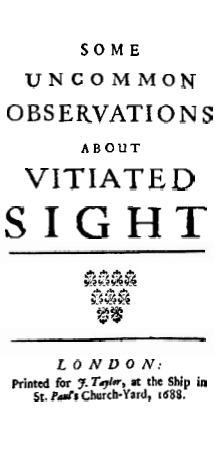
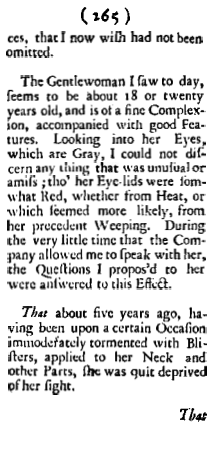
Abstract. Following a febrile illness, a young male was left totally colour blind in that he cannot match, sort or name colours; but his clinical acuity is normal and increment threshold measurements show that he retains three functional cone mechanisms. The findings suggest some dissociation in the analysis of colour and form.

1. Introduction

In 1684 there appeared in the *Philosophical Transactions* of the Royal Society the celebrated letter from Dr Dawbenry Turberville, in which the 'great and experienced Oculist' writes: 'A Maid, two or three and twenty years old, came to me from Banbury, who could see very well, but no colour beside *Black* and *White* ...' Turberville's report is slight and credulous, but a much more detailed account was given of a curiously similar case by Robert Boyle in his *Uncommon observations about Vitiated Sight* of 1688; and Boyle's account is truly brilliant. Some excerpts are given in figure 1. Although he had only a short time to examine a distressed young woman and although he probably had to


SOME
UNCOMMON
OBSERVATIONS
ABOUT
VITIATED
SIGHT

LONDON:
Printed for J. Taylor, at the Ship in St. Paul's Church-Yard, 1688.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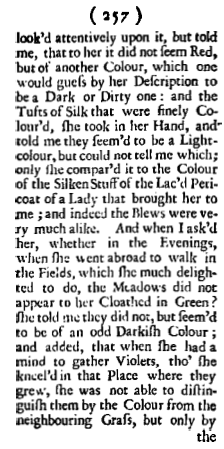
ces, that I now wiſh had not been omitted.

The Gentlewoman I ſaw to day, ſeems to be about 18 or twenty years old, and is of a fine Complexion, accompanied with good Features. Looking into her Eyes, which are Gray, I could not diſcern any thing that was unuſual or amiſs; tho' her Eye-lids were ſomwhat Red, whether from Heat, or which ſeemed more likely, from her precedent Weeping. During the very little time that the Company allowed me to ſpeak with her, the Queſtions I propos'd to her were anſwered to this Effect.

That about five years ago, having been upon a certain Occaſion immodefately tormented with Bliſters, applied to her Neck and other Parts, ſhe was quit deprived of her ſight.

That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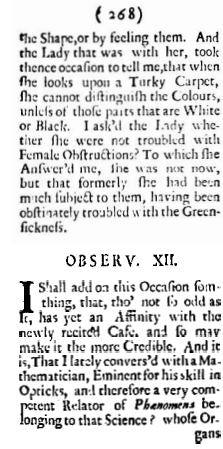
look'd attentively upon it, but told me, that to her it did not ſeem Red, but of another Colour, which one would gueſs by her Deſcription to be a Dark or Dirty one: and the Tufts of Silk that were finely Colour'd, ſhe took in her Hand, and told me they ſeem'd to be a Light-colour, but could not tell me which; only ſhe compar'd it to the Colour of the Silken Stuff of the Lac'd Petticoat of a Lady that brought her to me; and indeed the Blews were very much alike. And when I ask'd her, whether in the Evenings, when ſhe went abroad to walk in the Fields, which ſhe much delighted to do, the Meadows did not appear to her Cloathed in Green? ſhe told me they did not, but ſeem'd to be of an odd Darkiſh Colour; and added, that when ſhe had a mind to gather Violets, tho' ſhe kneel'd in that Place where they grew, ſhe was not able to diſtinguiſh them by the Colour from the neighbouring Graſs, but only by the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the Shape, or by feeling them. And the Lady that was with her, took thence occaſion to tell me, that when ſhe looks upon a Turky Carpet, ſhe cannot diſtinguiſh the Colours, unleſs of thoſe parts that are White or Black. I ask'd the Lady whether ſhe were not troubled with Female Obſtructions? To which ſhe Anſwer'd me, ſhe was not now, but that formerly ſhe had been much ſubject to them, having been obſtinately troubled with the Greenſickneſs.

OBSERV. XII.

I Shall add on this Occaſion ſomthing, that, tho' not ſo odd as it, has yet an Affinity with the newly recited Caſe, and ſo may make it the more Credible. And it is, That I lately convers'd with a Mathematician, Eminent for his skill in Opticks, and therefore a very competent Relator of *Phænomena* belonging to that Science? whoſe Or-
gans

Figure 1. Title page and excerpts from Boyle's *Vitiated Sight*. The second and third excerpts are consecutive (the original pagination is in error). The 'Blisters, applied to her Neck and other parts' probably represent therapy rather than symptom.

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introduce for himself both the concept of a case report and the concept of colour blindness, he quickly took a case history, showed that the patient had good acuity and was not dyslexic, and tested her colour vision with materials that came to hand. In fact, although there is not enough space here to set out the evidence, Boyle's case was very probably none other than the Maid from Banbury.

Cases such as Boyle's, of acquired severe colour blindness without loss of clinically measured acuity, are very rare, but a small number of modern cases have been reviewed by Meadows (1974) and we summarise here a case that we have been able to study in detail. Such cases are of interest because their very existence bears on the question of how far the central analysis of colour is independent of the analysis of other attributes of the visual image, such as form and movement (Mollon 1977, 1979, Cowey 1979).

Case history: MS, then a police cadet and aged 22, suffered a febrile illness in 1970. The presumptive diagnosis was herpes encephalitis but a brain biopsy was not taken for fluorescent antibody studies. An EEG showed severe bilateral disturbance with the main abnormality over the right parieto-occipital area.

During the acute phase of his illness, MS suffered severe visual disturbances and probably hallucinations. Most of his visual symptoms did not persist, but he was left with achromatopsia and a left homonymous hemianopia, with macular sparing. He also has agnosia and amnesia; the latter features of this case are discussed by Newcombe and Ratcliff (1975), who give further clinical details. He is able to read with a fluency consistent with his previous educational level, he can copy drawings accurately, and has an average verbal IQ.

MS has been repeatedly tested over the period 1972–78 and his condition has been stable in that his colour vision has not improved and his clinical acuity has not deteriorated. We have been able to establish that his colour vision was normal when he entered the police force, in so far as he did not make errors on the Ishihara plates.

2. Test results

Colour discrimination. MS cannot match, sort or name colours and by conventional criteria would be classed as totally colour blind. All tests using reflective materials were presented under CIE Illuminant C. Viewing was binocular (except (ii)). (i) *Farnsworth–Munsell 100 hue test.* Error score: 1245. (ii) *Nagel anomaloscope.* MS was asked to adjust a mixture of 540 and 640 nm light to match 580 nm, the brightness of the latter having been set to a normal value by the experimenter. Settings were random, ranging widely to either side of the normal ratio. (iii) *City University test.* 5/10 errors at reading distance; 6/10 errors at 2 m (7/10 on repetition). No systematic resemblance to a congenital deficiency. (iv) *Colour sorting.* This test was introduced to determine whether MS could make the grossest colour discriminations. 7·5-cm^2 patches of Munsell papers 5PB 5/10, 5G 5/6 and 5R 5/12 were placed on a sheet of N5 paper and MS was asked to sort thirty 1 cm^2 chips into three corresponding piles. Sorting was random. (v) *Oddity.* MS made errors when asked to identify the odd one out of the following sets of Munsell papers: two 5G 5/6, one 5R 5/12; four 5G 5/6, one 5PB 5/10; four 5G 5/6, one 5R 5/12. (vi) *Naming.* MS was unable spontaneously to name the colours of 640 and 540 nm stimuli (subtending 1° 20′) presented in isolation in the anomaloscope and did not improve on

repeated randomised presentation with correction. When asked (1975) to name the colours of absent objects, he was able to name (presumably by means of verbal memory) the colours of e.g. grass, traffic lights and the union jack but made errors on other common objects (e.g. banana, pillar box).

Increment thresholds. Even though a subject can report nothing about colour, it is nevertheless possible, by measuring increment thresholds, to establish whether he has access to signals from more than one class of photoreceptor. In the two-colour method of Stiles (1978), the subject is asked only whether he can detect a monochromatic flash on a monochromatic background. Thus Gibson (1962) reported that it was possible to demonstrate three π mechanisms in a congenital monochromat studied previously by Weale.

To determine whether MS retained more than one functional cone system in the red–green range, we used the minimum possible version of Stiles' procedure, measuring the thresholds for 640 and 510 nm flashes on 640 and 510 nm fields. The intensities of the fields were chosen (on the basis of Stiles' measurements) to raise thresholds for the normal observer by approximately 1 log unit in the homochromatic cases. If only one cone system, obeying the Principle of Univariance, were present, the relative efficiency of the two fields in altering sensitivity should be the same whether the thresholds were measured with red or with green light. This procedure closely resembles the measurement of the 'heterochromatic threshold reduction factor' of Boynton and Wagner (1961). Using a Maxwellian-view stimulator under computer control (see Mollon and Polden 1977 for details of apparatus, calibrations and experimental procedures), we presented 200-ms flashes of 1° of visual angle every 7 s. Thresholds were measured by an automated staircase procedure and each data point in the figures corresponds to 50 trials. MS proved a good psychophysical observer and the four independent determinations of each threshold were in good agreement.

Results are shown in figure 2 for MS and for two normal observers. It is clear that MS must have two functional cone mechanisms in the red–green range, since the

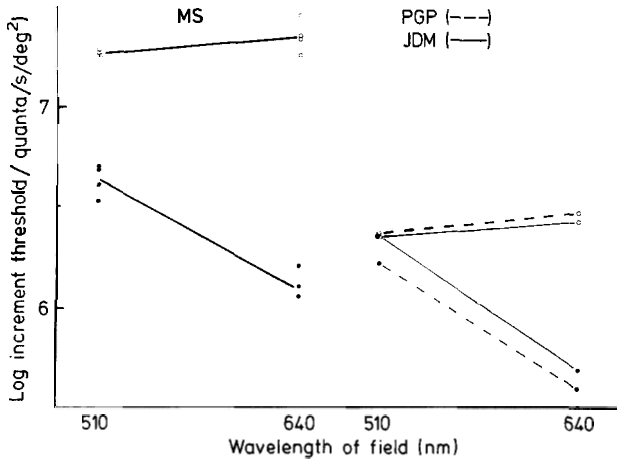

Figure 2. Incremental thresholds for 510 nm (•) and 640 nm (○) test flashes presented on 510 and 640 nm fields of $10^{8\cdot1}$ and $10^{8\cdot7}$ quanta/s/deg^2, respectively.

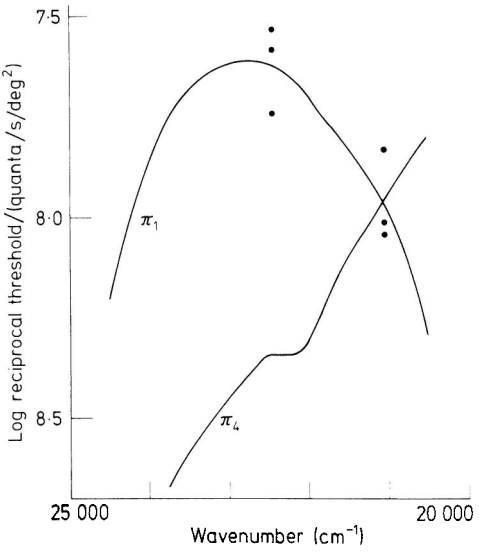

Figure 3. Test thresholds for 445 and 475 nm targets on 580 nm field of $10^{10\cdot3}$ quanta/s/deg^2. Each threshold was measured three times, test wavelengths being alternated. The full lines represent the sensitivities of π_1 and π_4 (Stiles 1978), adjusted vertically so that they pass through the mean threshold for 475 nm.

effectiveness of a given field depends on the wavelength of the test flash used to measure the threshold. Although this method does not directly give relative field sensitivities for the two mechanisms, the similarity of the slopes for the three observers suggests that MS has red and green mechanisms with spectral sensitivities close to normal.

We were especially interested to know whether MS retained a short-wavelength mechanism, since it has been supposed that signals from the violet-sensitive cones do not have access to achromatic channels (e.g. Guth *et al* 1968, Boynton 1971, Mollon and Krauskopf 1973). Figure 3 shows test sensitivities for 445 and 475 nm targets presented on a fixed 580 nm field of $10^{10\cdot3}$ quanta/s/deg^2: sensitivity is consistently higher at 445 nm, a result suggesting that detection is by a mechanism resembling π_1 rather than π_4. When asked what the flashes looked like at threshold, MS spoke of slight changes in 'lightness' or 'density'.

Pseudoisochromatic plates. These require report of a figure rather than explicit colour discrimination and so are treated separately.

If the Ishihara plates are presented in random order and at reading distance, MS typically fails all plates except the first (which is intended to detect malingerers).

A remarkable feature of this patient is that he can read the plates correctly when they are presented at 2 m. We are satisfied that his failure at reading distance does not arise from refractive error or because his colour field is small. Rather we believe that the character of the Ishihara test changes at a distance: the luminance contours of individual discs are no longer resolved and the dominant contour is the hue boundary between figure and ground. The latter contour, extracted by peripheral mechanisms, may be available centrally as a contour even though hue information is not available (cf De Valois and De Valois 1975, p142).

Colour-specificity of movement after-effect. The after-effect of seen movement has been reported to be strongest when adapting and test stimuli are of the same colour (Lovegrove *et al* 1972). This test was chosen because it offers another instance where the

normal observer's response is dependent on colour but no explicit report of colour is required. After adaptation for 30 s to a rotating (3 rpm) textured disc in red light, the mean duration of the after-effect for MS when tested in green light of similar brightness was only 53% of its value when he was tested in red light. Each test condition was repeated eight times, conditions being alternated, and the difference was highly significant (p = 0·003; Mann–Whitney U-test). In the light of a recently reported failure to obtain colour specificity of the movement after-effect even in normals (Day and Wade 1979), we draw attention to the low rate of rotation that we used; colour-specific channels may be especially sensitive to low velocities.

Spatial vision. Some have insisted that dyschromatopsias are invariably found to be associated with some impairment of spatial vision, provided only the examiner is sufficiently persistent (e.g. Teuber *et al* 1960). Tested on a variety of clinical charts (Snellen letters; Landolt C's) between 1971 and 1978, MS consistently demonstrated a visual acuity of 6/6 or better. However, he failed to resolve a printed square-wave grating of 20 c/deg; and formal measurements for vertical and horizontal sine-wave gratings revealed a depressed CSF with a peak at 1·25 c/deg. We do not understand this contradiction. Testing on the Keystone telebinocular showed no macular suppression, no phoria and macular stereopsis of 60% or better.

3. Conclusions

MS retains three functional cone mechanisms and each can independently control his verbal response in a detection task. Yet he apparently lacks the machinery to compare their outputs. In earlier days, MS would almost certainly have been classed as a case of total achromatopsia with normal acuity. This would still be a fair description, but we have had to introduce qualifications and in particular the meaning of 'colour blindness' has to be considered more analytically in cases of central achromatopsia. MS is totally unable to discriminate hue but we find several senses in which his behavioural response can depend on wavelength.

In so far as hue discrimination is lost and certain spatial functions are unimpaired, we have evidence for some independence of processing. It is possible that MS and similar cases have lost the homologue of the specialised colour area V4 described by Zeki (1978) in the pre-striate cortex of the rhesus monkey. An equally interesting possibility is that such patients have selectively lost one class of fibre in the visual system. At any level of the visual system there is a wide variation in the size of cell bodies and in the diameters of axons; and there is now extensive electrophysiological evidence that different classes of fibre carry different kinds of information. It is plausible that one class of cell may be more susceptible than another to a disease or toxin.

Acknowledgments

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