CORRESPONDENCE

Comment on: ‘Sector retinitis pigmentosa caused by mutations of the RHO gene’

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To the Editor:

In their recent article, Xiao et al. report five cases of sector retinitis pigmentosa (RP) owing to three distinct missense mutations in the rhodopsin (*RHO*) gene in unrelated Chinese probands [1]. Two of these pathogenic mutations (p.T17M and p.G106A) have been described previously whilst the third (p.L31Q) is novel. Recognition of cases of sector RP is important as this phenotype has potential implications for patient management which we wish to highlight.

Typically, the inferior retina is more severely affected in sector RP. This suggests a disease-exacerbating role for incident light to which inferior regions of the retina are more intensely exposed. Indeed, complete light restriction has been found to reduce the severity of outer retinal degeneration in animal models of rhodopsin-related RP [2], and these retinas also appear to display a greatly reduced threshold for light-induced damage [2, 3]. Further, exacerbation of disease has been reported in patients harbouring certain rhodopsin missense mutations (e.g. p.P180A) as a result of occupational exposure to high intensities of light [4]. We recently investigated the effect of filtration of short-wavelength light using red optical filters on disease progression in the rhodopsin p.P23H knock-in mouse model of sector RP [5]. We hypothesised that reducing exposure of rod photoreceptors to frequencies of light to which they are maximally sensitive might achieve a treatment effect whilst still allowing for useful vision. We observed a significant

Fig. 1 Filtration of short-wavelength light results in a relative preservation of the outer nuclear layer (ONL) in an animal model of sector RP. Representative spectral domain ocular coherence tomography (SD-OCT) scans are shown from mice heterozygous for the Rho p.P23H mutation housed in (a) standard clear plastic cages and (b) red-tinted plastic cages. Images were taken at equivalent retinal loci from littermates aged 3 months. GCL ganglion cell layer, IPL inner plexiform layer, INL inner nuclear layer, OPL outer plexiform layer, ONL outer nuclear layer, RPE retinal pigment epithelium

structural and functional benefit as a result of this intervention (Fig. 1) [5].

Although clinical trial data in support of light restriction or filtration as a treatment strategy for sector RP is lacking, we feel it appropriate to counsel patients with this phenotype on the importance of avoiding retinal exposure to bright light wherever possible. We typically recommend that such individuals wear highly attenuating sunglasses or red-tinted spectacles when outdoors in high luminance light conditions. We also avoid serial blue autofluorescence imaging in patients with sector RP for whom the intense short-wavelength light involved in its acquisition might be retinotoxic.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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