

Blue light hazard and aniridia

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SUMMARY The fundi of three patients with aniridia were photographed with a 470 nm illuminating light source. No apparent change in contrast was observable throughout the macular region. This would suggest an absence of the macular pigment. The likelihood of aniridics being more susceptible than normal persons to blue light damage is discussed.

In recent years the ocular hazards of non-ionising radiation have received great attention.¹⁻³ While the extent of light damage to the retina depends in the main on retinal irradiance level and exposure duration, many researchers have become increasingly aware of the role played by the wavelength of the source.^{4,5} This has been clearly demonstrated in that light of wavelengths greater than 580 nm has been shown to produce predominantly thermal injuries, while photochemical effects are encountered with visible radiation of lower wavelengths (400-580 nm⁶⁻¹¹).

The normally pigmented eye appears to be secure from possible blue light insult encountered in a natural environment because of the absorption characteristics of the cornea, crystalline lens, and macular pigment. Presumably if the situation arose such that one or more of these spectral filters were absent, the retina would be more susceptible to photochemical damage. Not surprisingly such cases do exist, for example, in the aphakic and also in the human albino (oculocutaneous and ocular), who not only lacks melanin in the pigment epithelium of the iris and the retina but also does not appear to possess any macular pigment.¹²⁻¹⁷ In addition the fovea in albinos is often reported as being hypoplastic.^{18,19}

Since poor foveal differentiation is also a frequent feature of the aniridic retina,²⁰⁻²² it was therefore the purpose of this study to ascertain if, like albinos, aniridics too lack xanthophyll pigment. As in our previous investigation, monochromatic fundus photography was employed to examine the macular area.^{17,23,24}

Subjects and methods

A fundus camera (Zeiss) was used with its aperture

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stop constantly set at 5.5 mm. A blue gelatin band-pass filter (Ilford No. 602) with a maximum transmission at 470 nm was inserted into the illuminating system in order to visualise the xanthophyll pigment. Previous studies had established that this was a suitable method to establish the presence of xanthophyll. Flash intensities of 60 and 120 w sec⁻¹ were used. Black and white ASA 125 film (Kodak Plus X) was used, and the films were commercially developed and printed with no retouching.

Three subjects with aniridia were examined. This small sample number of subjects reflects the low incidence of the condition, which is estimated to be 1 in 100 000.^{20,26} In this study two of the three subjects were brothers. An extensive family history indicated an autosomal dominant mode of inheritance. Both the brothers had monocular visual acuities of less than 6/36, bilateral peripheral corneal pannus, bilateral small central lens opacities, and no visible irides. Infrared oculography revealed that they both showed a manifest latent nystagmus with associated superimposed horizontal pendular oscillation. Our third subject had no family history of ocular disorder and monocular visual acuities of 6/18+1. A bilateral peripheral corneal pannus and a small unilateral (right eye) central lens opacity were present. No iris was visible. No ocular oscillation could be detected by infrared oculography.

Twenty-five subjects with no ocular or systemic disease acted as controls.

Results

Examination of the retina with the 470 nm illuminating wavelength revealed two distinct patterns. Firstly, a macular darkening with an increased contrast area around the fovea was found in all normal subjects regardless of their skin pigmentation (Fig. 1). The smaller central region is believed to



Fig. 1 The normal fundus examined with 470 nm illuminating wavelength to enhance the contrast of the macular pigment. Notice the marked macular darkening, indicating the presence of macular pigment.



Fig. 2 The aniridic fundus examined with a 470 nm illuminating wavelength. There is no apparent change in contrast throughout the macular region, suggesting that there is an absence of macular pigment.

correspond to the high absorption of blue light by the xanthophyll pigment. This has been described previously.^{17,23,24}

On the other hand there was no apparent change in contrast throughout the macular region in the three aniridics, suggesting an absence of macular pigment in these subjects (Fig. 2).

Discussion

The syndrome of congenital aniridia may show at least four phenotypes. The first type, to which our three patients belong, is the association of aniridia with foveal hypoplasia, nystagmus, corneal pannus, and the likelihood of secondary glaucoma. The second type is that reported by Elsas *et al.*²⁵ where visual resolution appears relatively preserved. The third and fourth phenotypes are intimately associated with mental deficiency²⁶ and Wilms's tumour²⁷ respectively.

For our subjects the retinal irradiance level is increased because of the presence of large and constant pupil aperture. This represents a substantial incremental increase in the light flux as the retinal irradiance is a function of the square of the pupil diameter. Of course additional parameters such as the transmission of the ocular media and the degree of defocus of the retinal image need to be taken into account when evaluating the absolute irradiance level.

Large fixed pupil apertures also disqualify the visual system from being optically ideal.²⁸ Yet, though the retinal image quality is degraded by optical

aberrations, the Stiles-Crawford effect, and a reduced depth of focus, the loss of visual resolution at photopic illumination levels is only about 18%²⁹—that is, a decrease in visual acuity from 6/6 to 6/7.5. Indeed Elsas and his colleagues have described a large pedigree with familial aniridia in which 61% of patients had visual acuities of 6/9 or better and only 5% had visual acuities <6/60.²⁵ None had nystagmus or corneal pannus, while only 18% of the patients had some degree of lenticular opacity. Thus the absence of iris tissue alone cannot be solely responsible for the poor acuity usually associated with aniridia.

A static pupil does, however, possess two important disadvantages. The first concerns the loss of the pupil light reflex. Constriction of the natural pupil at high light levels reduces the retinal illumination and so ensures a more rapid dark adaptation process in the retina to low light levels, as well as improving sensitivity and contrast detection during dark adaptation. Indeed experiments have shown that the dark adaptation process is speeded up by 4–10 min and the sensitivity initially increased by approximately 10 times.³⁰

Secondly, the retinal irradiance cannot be controlled. This feature has led some clinicians to propose that the lack of macular development in aniridia may be caused by excessive amounts of light entering the eye.³¹ Although nystagmus should have the effect of limiting the duration of exposure of the fovea, this would apply only to a point source, whereas most natural blue light sources are extended. Thus there is a case for fitting an aniridic patient with some form of artificial pupil (for example, an iris

print contact lens) to minimise the possible long-term effects of light damage, particularly for wavelengths of 400–500 nm.⁵ Moreover the optical portion could also be tinted, so that the absorption properties of the contact lens parallel that of xanthophyll, thus providing further protection for the retina. Similar safeguards may also be advisable for the albino individual.

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