Imaging the Macula Through a Black Occlusive Intraocular Lens

Black occlusive intraocular lens (IOL) insertion has been shown to be an effective treatment for intractable diplopia,1,2 visual confusion, and poor cosmesis resulting from leukokoria.3-5 A significant disadvantage of occlusive IOL insertion is that conventional funduscopy is not possible, preventing the detection of posterior pole disease. Here we describe a patient in whom we made the novel discovery of successful macular imaging through an occlusive IOL using an infrared light–based scanning laser ophthalmoscope/optical coherence tomography (OCT) scanner. We suggest that this report is likely to fundamentally change the current thinking on occlusive IOLs and to promote their use in the management of these patients.

Report of a Case. A 74-year-old woman had a stage 3 idiopathic full-thickness macular hole in the left eye with corrected Snellen visual acuity of 6/6 OD and 6/60 OS. She underwent pars plana vitrectomy, and revision surgery was required 3 months later for a persistent macular hole.

The patient then noted visual confusion. Best-corrected visual acuity had deteriorated to 1/60 OS. Occlusion of the left eye with a patch was the only treatment that alleviated symptoms. The patient preferred implantation with a black occlusive IOL over the options of an occlusive contact lens, corneal tattooing, or tarsorrhaphy. Phacoemulsification was performed with implantation of an 85F black occlusive IOL (Figure 1A). There was successful resolution of symptoms and the visual acuity was 6/6 OD.

The patient returned 9 months later with blurred vision in the right eye down to an uncorrected visual acuity of 6/18. We confirmed a difference in pupil color between the right and left eyes (Figure 1B and C) and that fundus visualization was not possible through the black IOL (Figure 1D). An irregular macular reflex was identified in the right eye and an OCT scan was requested. The patient then noted visual confusion. Best-corrected visual acuity had deteriorated to 1/60 OS. Occlusion of the left eye with a patch was the only treatment that alleviated symptoms. The patient preferred implantation with a black occlusive IOL over the options of an occlusive contact lens, corneal tattooing, or tarsorrhaphy. Phacoemulsification was performed with implantation of an 85F black occlusive IOL (Figure 1A). There was successful resolution of symptoms and the visual acuity was 6/6 OD.

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Comment. To our knowledge, this is the first report of fundus visualization using an infrared-based scanning laser ophthalmoscope/OCT scanner (operational at 820/870 nm) in an eye implanted with a black occlusive IOL. This observation suggests that the black polymethyl methacrylate IOL transmits sufficient infrared light to permit macular imaging. We have subsequently confirmed in a laboratory study high levels of transmission of infrared light through an occlusive IOL (I.H.Y., Stuart N. Peirson, PhD, C.K.P., unpublished data, June 2010).

Black occlusive IOLs have been implanted across a range of indications, including intractable diplopia, visual confusion, and unsightly leukokoria,\(^3,4\) and have been demonstrated to produce high levels of postoperative satisfaction in patients.\(^1,2\) However, the clinical decision to implant an occlusive IOL has, to this point, been troubled by the dogma that preventing medical and ophthalmic practitioners from visualizing posterior pole structures carries significant risk, limiting the selection of this effective treatment.

The identification of a means of visualizing the retina eliminates this risk and could allow more patients to benefit from occlusive IOL insertion. The nature of consent for patients in whom occlusive IOL is a therapeutic option needs to fundamentally change to reflect this finding.

This novel observation has significant implications for diagnosis and monitoring of medical and ophthalmic disease in current and future patients with occlusive IOLs.

Figure 2. Confocal scanning laser ophthalmoscopic image of the macula and horizontal optical coherence tomographic scan through the fovea obtained through the black occlusive intraocular lens, confirming macular hole closure in the left eye (A), and the phakic right eye showing normal macular anatomy (B).
in situ where disc and retinal assessments are essential and for preoperative evaluation if therapeutic IOL exchange to transparent media is considered.

Moreover, the observed ability of occlusive IOLs to transmit infrared light suggests the potential for development of infrared–based assessment tools such as Snellen charts for this patient group.

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Blue Sclera With and Without Corneal Frailty (Brittle Cornea Syndrome) in a Consanguineous Family Harboring ZNF469 Mutation (p.E1392X)

Blue sclera with corneal fragility characterizes brittle cornea syndrome (BCS) (OMIM #229200), an underrecognized predominantly ocular disorder that is often confused with Ehlers-Danlos syndrome (EDS). Brittle cornea syndrome has recently been associated with recessive mutation in ZNF469, which encodes a classic zinc-finger protein that is likely involved in synthesis and/or organization of corneal collagens. In this report, we describe the phenotype/genotype correlation for blue sclera with or without BCS in all 3 children of a consanguineous, asymptomatic Syrian couple.

Report of Cases. Case 1. Sibling 1 is a 13-year-old boy (Figure 1). He had blue sclera since birth and history of hernia repair, the details of which were not available. At age 2 years following minor trauma, he had bilateral corneal rupture and underwent multiple corneal operations that led to phthisis. It was suspected that he had congenital glaucoma. At approximately age 5 years, he had slightly decreased hearing following recurrent ear infections; this resolved with medical treatment and his hearing is now considered normal. At age 12 years, he had a right forearm dislocation following a significant fall down stairs that required surgical pinning.

Ophthalmic examination revealed visual acuity of no light perception OU. Both eyes were phthisical, had blue sclera, and were soft to palpation.

General assessment revealed a well-nourished alert boy with normal vital signs and chestnut-colored hair. Height, weight, and head circumference were age appropriate. Skin was thin, velvety, and without abnormal elasticity. Subcutaneous veins were easily appreciated. There were scattered small scars on all extremities and a large one at the right elbow where a small surgical incision had been made. There was no joint hypermobility (Beighton score 2/9) or scoliosis. Oral inspection revealed grossly normal dentition and a slightly arched palate. Extremity assessment revealed bilateral valgus foot (talipes valgus) and hallucus valgus, which were confirmed by plain-film radiography. Skeletal plain-film radiography showed normal bone densities without evidence for fractures. Complete blood cell count and electrolyte levels were within normal limits.

Case 2. Sibling 2 is an 8-year-old girl (Figure 2). She had blue sclera since birth. There was a vague history of a dislocation in the right forearm following trauma at age 1 year that did not require surgery. She was treated medically for congenital glaucoma until age 5 years, at which time she was diagnosed as having keratoglobus without evidence of glaucoma. Cycloplegic refraction at age 4 years was approximately plano in both eyes and fundus examination results were unremarkable. Protective glasses were stressed but were not used. At age 7 years, she had a minor trauma in the left eye that led to Descemet membrane detachment and associated corneal edema, for which she had surgical repositioning of the Descemet membrane via air bubble injection.

Ophthalmic examination revealed visual acuity of 20/30 OD and 20/400 OS, with no improvement by pinhole. Both eyes had keratoglobus and blue sclera and had grossly normal intraocular pressure by digital palpation. Slitlamp examination revealed thin corneas with keratoglobus and in the left eye corneal haze corresponding to the area of prior Descemet membrane detachment. At age 6 years, cycloplegic refraction was +0.75 – 1.50 × 042 OD and –11.00 – 2.50 × 090 OS with scissoring of the reflex. Fundus examination results were within normal limits.

General assessment revealed a thin alert girl with normal vital signs and chestnut-colored hair. Height, weight, and head circumference were age appropriate; however, she did have an appearance of frontal bossing and the calvaria was irregular to palpation. Skin was thin, velvety, without abnormal elasticity, and without scars. Subcutaneous veins were easily appreciated. There was significant joint hyperlaxity (Beighton score 9/9). Oral inspection revealed poor dental hygiene and an arched palate. The sternum protruded outward slightly (pectus carinatum). Scoliosis was not present but lumbar lordosis seemed exaggerated. Extremity assessment revealed bilateral talipes valgus. Skeletal plain-film radiography confirmed the irregular calvaria, exaggerated lumbar lordosis, and bilateral talipes valgus; bone densities were normal. Complete blood cell count and electrolyte levels were within normal limits.

Case 3. Sibling 3 is a 4-year-old girl (eFigure 1; http://www.archophthalmol.com). This healthy-appearing, alert girl with blue sclera since birth had normal vital signs and chestnut-colored hair. Other than blue sclera, re-