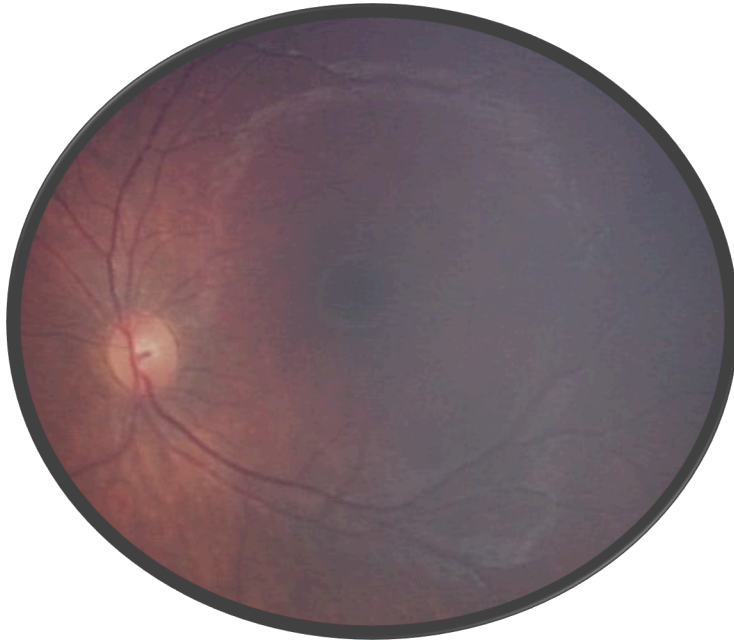


SUPPLEMENTAL APPENDIX #1

Representative images of fundus and OCT images with description of Weill Cornell Batten Severity (WCBS) score each imaging modality

Figure 1: WCBS Ophthalmic Severity Score 1

- A. Dilated fundus photograph of patient showing a normal appearing optic nerve, vessels, and fovea



- B. SD-OCT of the same patient which shows normal retinal architecture without outer retinal changes

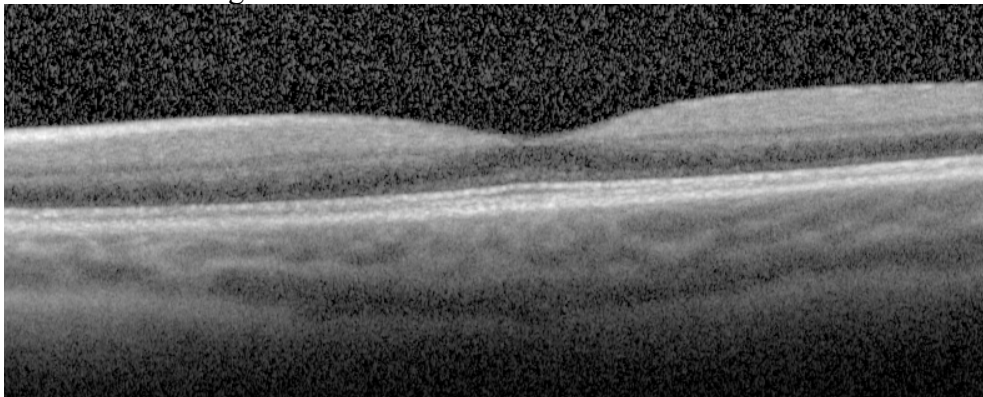
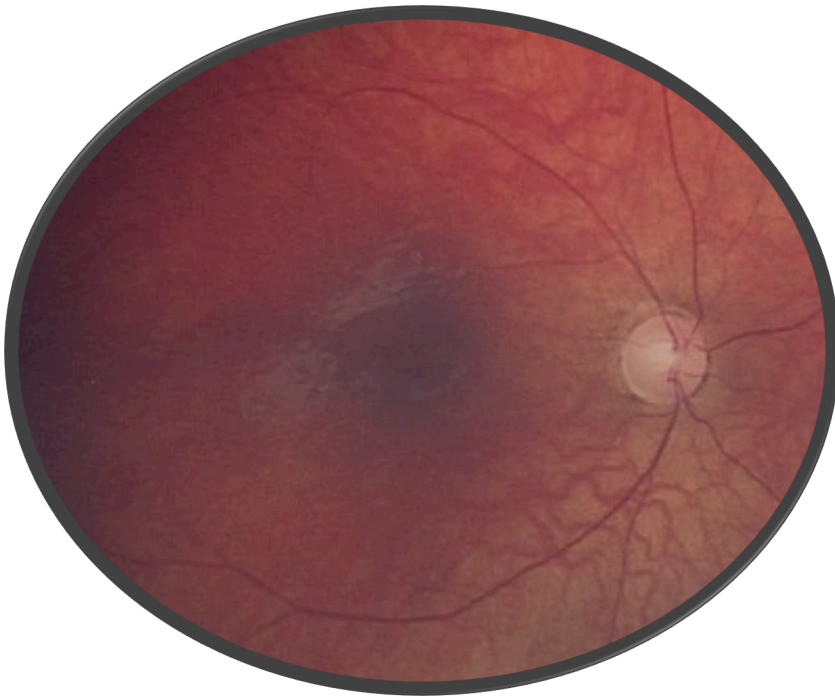


Figure 2: WCBS Ophthalmic Severity Score 2

- A. Dilated fundus photograph of patient showing mild pigmentary changes around the fovea. The optic nerve and fovea appear normal.



- B. SD-OCT of the same patient which shows normal retinal architecture outside of the fovea, with some disruption of the para-foveal IS/OS, yet preservation of the external limiting membrane. Subfoveally, the outer retina appears unaffected.

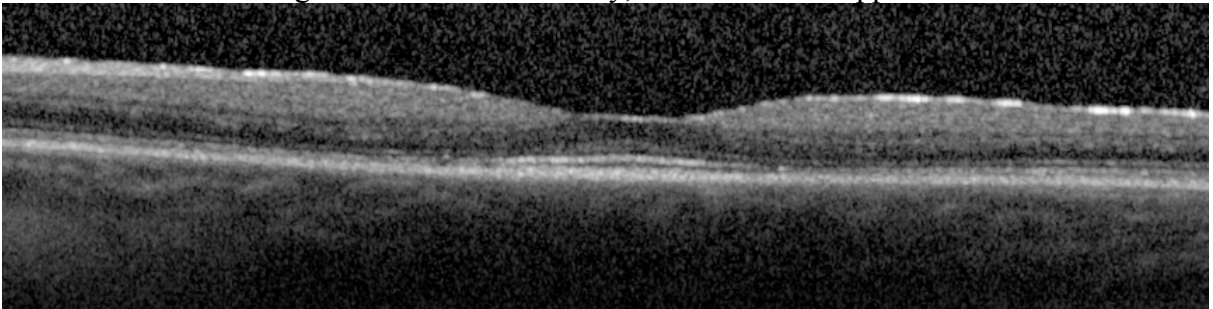
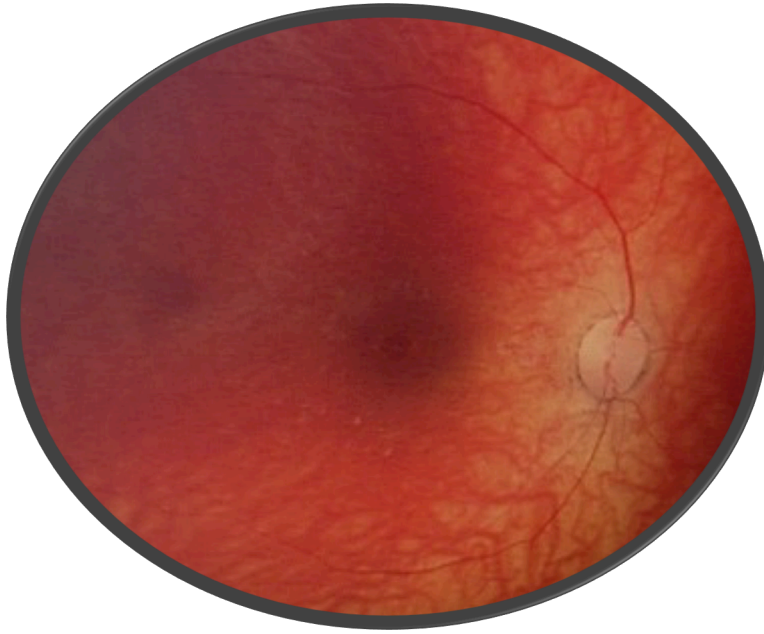


Figure 3: WCBS Ophthalmic Severity Score 3

- A. Dilated fundus photograph of patient showing central foveal pigmentary changes. There is some early pallor of the optic nerve with mild vessel attenuation.



- B. SD-OCT of the same patient which shows outer retinal architecture disruption, including significant subfoveal loss. There is more extensive disruption of the IS/OS junction compared with severity score 2, with external limiting membrane disruption. The width of outer retinal disruption is less than 1 disc diameter.

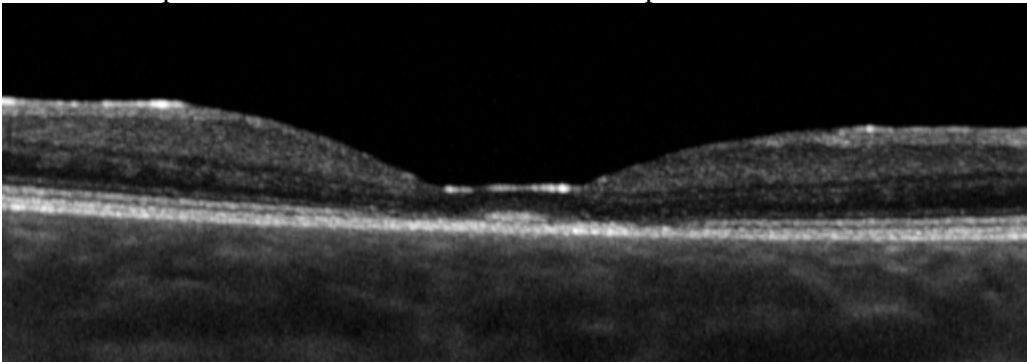
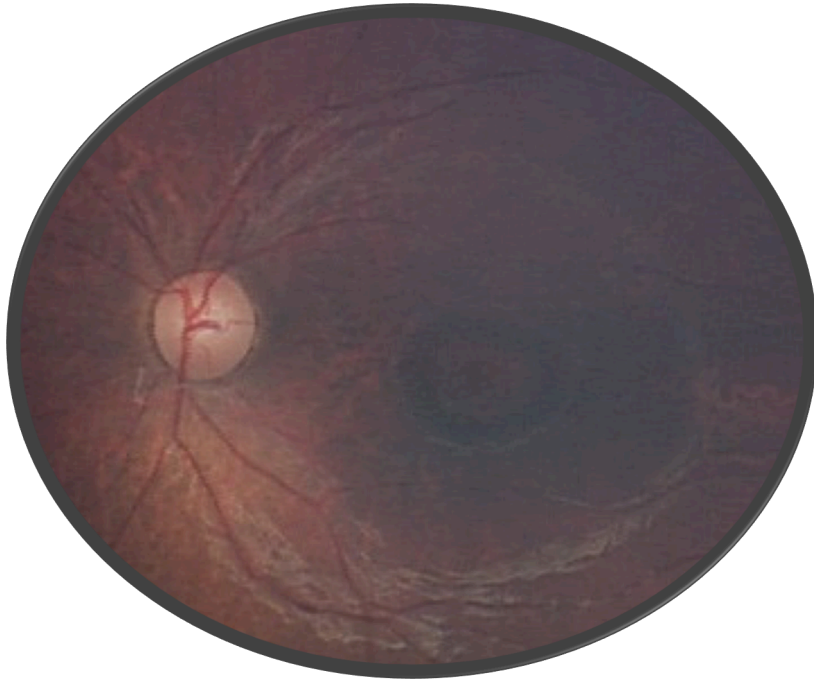


Figure 4: WCBS Ophthalmic Severity Score 4

- A. Dilated fundus photograph of patient showing central foveal pigmentary loss with a Bull's eye maculopathy appearance. Rings of pigmentary changes emanate from the fovea. There is some pallor of the optic nerve with mild vessel attenuation.



- B. SD-OCT of the same patient demonstrating significant outer retinal atrophy with build-up of a hyper-reflective material at the level of the retinal pigment epithelium subfoveally (and occasionally these collections form a ring parafoveally). Typically the outer retinal loss extends less than 2 disc diameters with normal retinal architecture beyond this region.

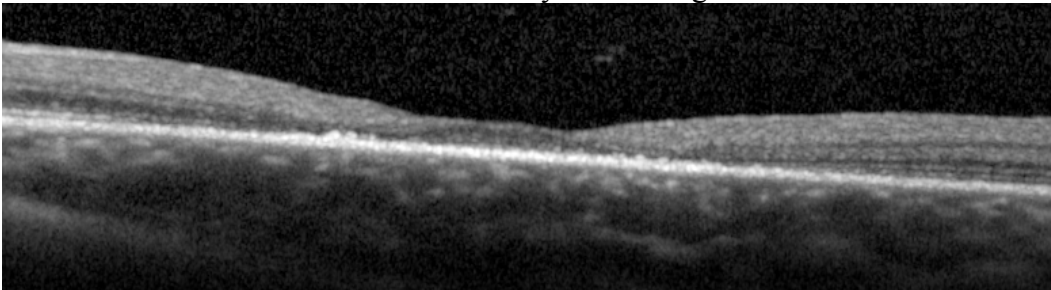
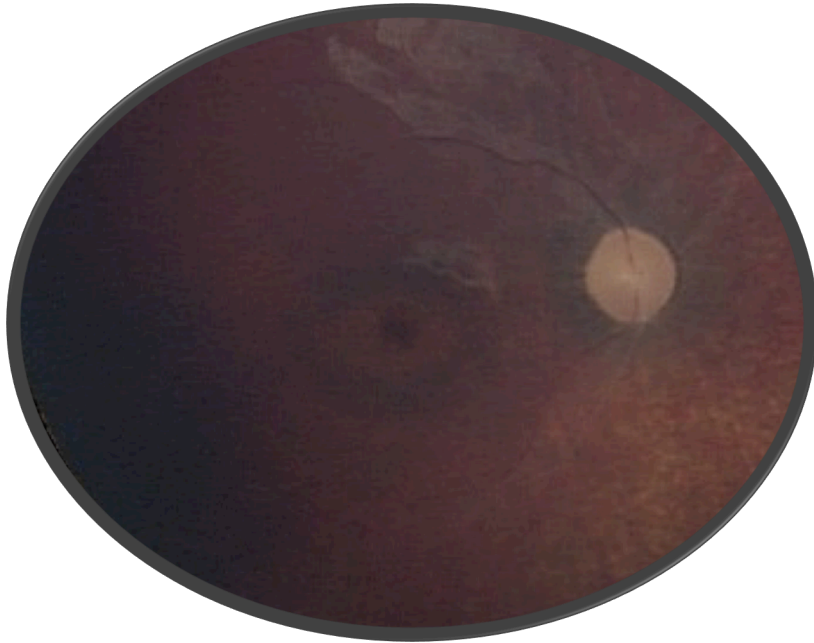


Figure 5: WCBS Ophthalmic Severity Score 5

- A. Dilated fundus photograph of patient showing central foveal pigmentary loss with a Bull's eye maculopathy appearance. There are concentric rings of pigmentary changes centered on the fovea. There is significant pallor of the optic nerve with severe retinal vessel attenuation.



B. SD-OCT of the same patient demonstrating significant outer retinal atrophy with build-up of a hyper-reflective material at the level of the retinal pigment epithelium subfoveally (and occasionally these collections form a ring parafoveally). The outer retinal changes extend throughout all visualized areas of the retina with no normal tissue visible.

