

**Supplementary Table S1: Progression of retinal degeneration.**

Patient, sex	Age at last examination	BCVA at last examination	Fundus	Visual Field	SW-FAF	SD-OCT	Other
L-08031428, M	28	OU: 20/800	Waxy pallor of ONH Vascular narrowing Bull's eye macular lesion Whitish discoloration of midperipheral retina Bone spicules in nasal midperipheral retina	40° large absolute (V4e) central scotoma; preservation of peripheral V4e isopter	Large macular hypoAF round lesion with indistinct hyperAF edges Large midperipheral ring of hypoAF with indistinct hyperAF edges	Widespread disappearance of outer reflective layers (ONL, EZ, RPE) Dense irregular deposits at the level of RPE	ffERG: residual dark-adapted responses; unrecordable light-adapted responses
VV-51717, M	57	VA: 0.2 RE, 0.1 LE	Bull's eye maculopathy	Not repeated	Macula: hypoAF area, surrounded by hyperAF ring No changes in peripheral retina	Disappearance of outer layers (ONL, EZ, IS/OS, RPE) in the macula Dense irregular deposits at the level of RPE	
VV-1595021, F	20	VA: 0.2 RE, 0.25 LE	Central foveal atrophy, peripheral retina within normal limits	Not repeated	Macula: hypoAF area, surrounded by hyperAF ring No changes in peripheral retina	Disappearance of outer layers (ONL, EZ, IS/OS RPE) in the macula Dense irregular deposits at the level of RPE	
HD-OPH4200, F	53	VA : RE : 0.2, P8 LE : 0.2, P8 Eccentric viewing	Loss of foveal reflex with a few discrete hypo-pigmented spots around the fovea	Bilateral relative central scotoma	Macula: hypoAF area, surrounded by hyperAF ring No changes in peripheral retina	Disappearance of outer layers (ONL, EZ, IS/OS, RPE) in the macula	Major photophobia
HD-OPH1206, M	36	VA : RE : 0.3ff ; P4f LE : 0.3 f ; P5	Bull's eye maculopathy	Bilateral relative central scotoma	Macula: hypoAF area, surrounded by hyperAF ring No changes in peripheral retina	Disappearance of outer layers (ONL, EZ, IS/OS, RPE) in the macula Dense irregular deposits at the level of RPE	