

Table 2 Continued

	Family 6				Family 7				Family 8		Family 9		Family 10	
Pedigree number	II:2		III:2		II:1		II:2		II:1		II:1		III:1	
Age at examination (y)	3.5		4.75		50		58		31		58		44	
Eye	RE	LE	RE	LE	RE	LE	RE	LE	RE	LE	RE	LE	RE	LE
BCVA	0.125	0.125	0.8	0.8	1.0	1.0	1.0	1.0	1.0	1.0*	1.0	1.0	0.15	0.2
Refractive error	-0.25	-0.25	+1.50/ -2.75x20°	+2.75/ -2.5x160°	+0.5/ -1.5x4°	+1.75/ -3.35x163°	+0.5	+0.25	+0.75/ -3.25x22°	+3.0/ -6.0x125°	+1.25	+1.25/ -0.25x95°	-2.5	-2.5/ -0.5x90°
Age at cataract surgery (y)	-	-	-	-	47	43	-	-	-	-	52	52	-	-
Iris hypoplasia/atrophy	NA	NA	NA	NA	+	+	-	-	-	-	+	+	+	+
Arcus	-	-	-	-	+	+	+	+	+	+	+	+	-	-
Shagreen	-	-	-	-	+	+	+	+	-	-	+	+	+	+
ACD (mm)	NA	NA	NA	NA	6.01	6.31	NA	NA	4.66	5.09	5.89	5.88	4.78	4.75
WTW (mm)	13.5	13.5	15	15	15.7	15.5	15	15	15.9	15.7	14.5	14.5	14.29	14.29
AL (mm)	22.33	21.8	NA	NA	26.93	26.49	NA	NA	24.58	25.14	25.06	24.94	24.64	24.54
CCT (µm)*	NA	NA	NA	NA	446	436	390	392	418	411	450	456	408	401
ECD (cells/mm ²)	NA	NA	NA	NA	1713 (at 44 y)	2010 (at 44 y)	NA	NA	NA	NA	NA	NA	-	-
Glaucoma	-	-	-	-	S	S	-	-	-	-	-	+ (UGH)	-	-
Other	-	-	-	-	PD, PCVs	PD, FHI	-	-	PD	KC (TKC 3-4), PD, XT	-	-	-	-

ACD = anterior chamber depth (normal values under 97.5 percentiles ≤ 3.8 mm [12], AL = axial length, BCVA = best corrected visual acuity, CCT = central corneal thickness (normal values normal above 2.5 percentiles ≥ 481 µm) [12], ECD = endothelial lens density, FHI = Fuchs heterochromic iridocyclitis, KC = keratoconus, LE = left eye, NA = not available data, PCVs = posterior corneal vesicles, PD = pigment dispersion, RE = right eye, S = suspect, TKC = Topographic Keratoconus Classification derived from Pentacam, UGH = Uveitis-Glaucoma-Hyphaema syndrome, WTW = horizontal white to white, XT = exotropia, y = years, - = not present, + = present,

*Measurements were obtained by spectral domain optical coherence tomography