A Pilot Study of Fourier-Domain Optical Coherence Tomography of Retinal Dystrophy Patients

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• PURPOSE: To characterize the macular anatomy of retinal dystrophy eyes using high-speed, high-resolution, Fourier-domain optical coherence tomography (FD-OCT).

• DESIGN: Case-control study.

• METHODS: Retinal dystrophy patients and normal age- and gender-matched controls underwent FD-OCT imaging using the RTVue (Optovue Inc, Fremont, California, USA). Vertical and horizontal 8-mm scans of 1024 lines/cross-section were obtained. Based on boundaries manually drawn on computer displays of OCT cross-sections, the thicknesses of the retina, inner retinal layer (IRL), and outer retinal layer (ORL) were averaged over both 5-mm (macular) and 1.5-mm (foveal) regions centered at the fovea. The IRL was the sum of nerve fiber layer (NFL), ganglion cell layer (GCL), and inner plexiform layer (IPL) thicknesses. Total retinal thickness (RT) was measured between the internal limiting membrane (ILM) and the retinal pigment epithelium. ORL thickness was calculated by subtracting IRL thickness from RT.

• RESULTS: Fourteen patients (three retinitis pigmentosa, two cone-rod degeneration, two Stargardt disease, and seven normal controls) underwent FD-OCT imaging. Mean foveal RT was 271.3 ± 23.3 μm for controls and 158.4 ± 47.1 μm for retinal dystrophy patients ($P < .001$). Mean macular RT was 292.8 ± 8.1 μm for controls and 199.1 ± 32.6 μm for retinal dystrophy patients ($P < .001$). Mean macular ORL was 182.9 ± 4.7 μm for controls and 101.3 ± 18.7 μm for retinal dystrophy patients ($P < .001$); mean macular IRL was 109.9 ± 6.4 μm for controls and 97.9 ± 20.7 μm for retinal dystrophy patients ($P = .06$).

• CONCLUSION: Eyes with retinal dystrophy had a small (11%) decrease in macular IRL and severe (45%) decrease in macular ORL compared to normal controls. (Am J Ophthalmol 2008;146:417–426. © 2008 by Elsevier Inc. All rights reserved.)

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OPTICAL COHERENCE TOMOGRAPHY (OCT) WAS first developed as a research tool in 1991.[1] Since then, improved resolution and speed have enabled the OCT to become widely used clinically. The prior standard in OCT retinal imaging was the Stratus (also known as the OCT3; Carl Zeiss Meditec Inc, Dublin, California, USA),[2] which has an axial resolution of 9 to 10 μm. The machine acquires images at 400 axial scans (A scans) per second.[2,3] Recently, a new technology called spectral domain or Fourier-domain OCT (FD-OCT) has enabled much faster image acquisition, enabling both higher definition (more lines per image) and less motion error.[4–7] The current study used the RTVue FD-OCT system (Optovue Inc, Fremont, California, USA), which performs 26,000 A-scans per second, 65 times faster than the Stratus. This FD-OCT system also employs a higher-bandwidth light source, thus providing a finer axial resolution of 5 μm—a two-fold improvement over the Stratus. Although ultra-high-resolution (3 μm) OCT (UHR-OCT) of the retina has been previously described,[8–14] UHR-OCT requires a femtosecond laser light source, which is not practical clinically because of the high cost, bulk, and maintenance requirements. In contrast, the RTVue, as well as other FD-OCT machines, utilizes a superluminescent diode that is compact, reliable, and more economical.

Previous histopathological studies in eyes with retinal degeneration showed loss of photoreceptor segments early in the disease.[15–18] We used the FD-OCT to explore structural changes in the macular anatomy of retinal dystrophy patients. A second goal of the study was to determine whether structural findings on OCT correlated with visual acuity (VA).

PATIENTS AND METHODS

PATIENTS WITH RETINAL DYSTROPHIES WERE PROSPECTIVELY enrolled into the study. Eligible patients included those with retinitis pigmentosa (RP), cone-rod dystrophy, and Stargardt disease. Normal control subjects were selected from the database of the Advanced Imaging for Glaucoma Study (AIGS)[19] to match the age and gender of the retinal dystrophy subjects. Normal controls were defined as having both eyes free of ocular pathology after a comprehensive eye exam, intraocular pressures (IOP) less

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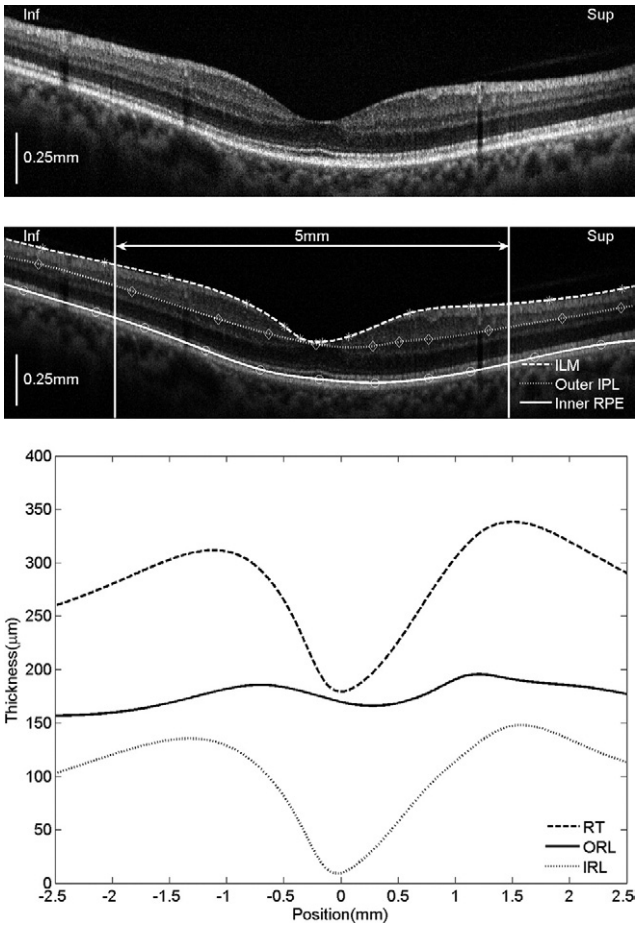

FIGURE 1. Normal retinal optical coherence tomography (OCT) image and retinal thickness (RT) profile. (Top left) Vertical OCT image, oriented inferior (left) to superior (right), of a normal patient. Note that the retinal pigment epithelium (RPE) is thicker centrally than peripherally. (Top right) Internal limiting membrane (ILM), outer inner plexiform layer (IPL), and inner RPE boundaries are indicated as dashed lines. (Bottom) OCT RT profile shows the mean OCT RT (total RT, inner RT, and outer RT) plotted as a function of eccentricity from the foveal center (μm).

than 21 mm Hg, full visual fields (VF), and normal central corneal thickness greater than 500 μm. (A detailed description of the enrollment criteria can be obtained from the AIGS Manual of Procedures [www.AIGStudy.net].) All patients signed a written informed consent form prior to entry into the study.

Both eyes of every patient were scanned three times with two scan patterns, the line scan and the cross-hairs scan. We chose the best of the three scans of each patient for analysis. These patients have poor fixation in general. The line scan consists of 1024 A-scans over an 8-mm length. The line was oriented horizontally. The cross-hairs scan consists of two line scans (vertical and horizontal). All scans are centered at fovea. To reduce the speckle noise, the study images were averaged from several scans. Up to eight frames were averaged for cross-hairs scans and up to 16 frames were averaged for line scans.

The OCT images of each patient were exported from the RTVue OCT to a computer. A computer program was used to display and draw the inner retina boundary based on image intensity on the OCT. For each boundary, points are selected and a thin spline plate fitting was applied to these points to create a smooth boundary. Three boundaries, the internal limiting membrane (ILM), outer boundary of the inner plexiform layer (IPL), and inner boundary of the retinal pigment epithelium (RPE), were drawn as shown in Figure 1. The outer plexiform layer (OPL) was excluded from boundary detection and not drawn as it could not be clearly demarcated on some retinal dystrophy patients. Total retinal thickness (RT), inner retinal layer thickness (IRL), and outer retinal layer thickness (ORL) measurements were calculated. The total RT was measured from the ILM to the inner RPE boundary, which is an accepted boundary. The IRL was measured from the ILM to the outer boundary of the IPL, and represents the sum of the nerve fiber layer (NFL), ganglion cell layer (GCL), and IPL thicknesses. ORL thickness was the value obtained from RT minus IRL thickness.

A prior study on reproducibility of macular thickness showed only small variations in measurements. In 125 normal and 112 glaucoma eyes, IRL, ORL, and RT measurements had standard deviations less than 1.7 μm and the C variance was less than 1.1% (Huang D, personal communication, January 2008).

At the foveal center, since there is no IRL, IRL thickness is zero and the foveal RT is equal to the ORL thickness. The average thickness of the central foveal point measured on horizontal and vertical scans was designated central retinal thickness (cRT). The macular IRL, ORL, and RT measurements were averaged (area-weighed) over a 5-mm region. The foveal IRL, ORL, and RT measurements were averaged over a 1.5-mm-diameter area. These thickness measurements of the retinal dystrophy cases were compared with those of the normal cases (Matlab 7.1; Mathworks Inc, Natick, Massachusetts, USA). The RPE thickness was also calculated for the normals and the retinal dystrophy patients. For each patient, an OCT RT profile (total thickness, ORL thickness, and IRL thickness) was created. On these profiles, OCT RT is plotted across various distances from the fovea. The range of normal RTs is plotted on the same graph and shaded in gray to allow for a rapid comparison of the patient's RT with the normal RTs. The *t* test was used to compare the mean RT of the retinal dystrophy group to the normal group.

Logarithm of minimal angle of resolution (logMAR) VAs were calculated from the Snellen VAs for each eye. In order to determine whether there was a correlation between VA and RTs, correlations were calculated for macular thickness and logMAR acuities and for foveal thickness and logMAR acuities.

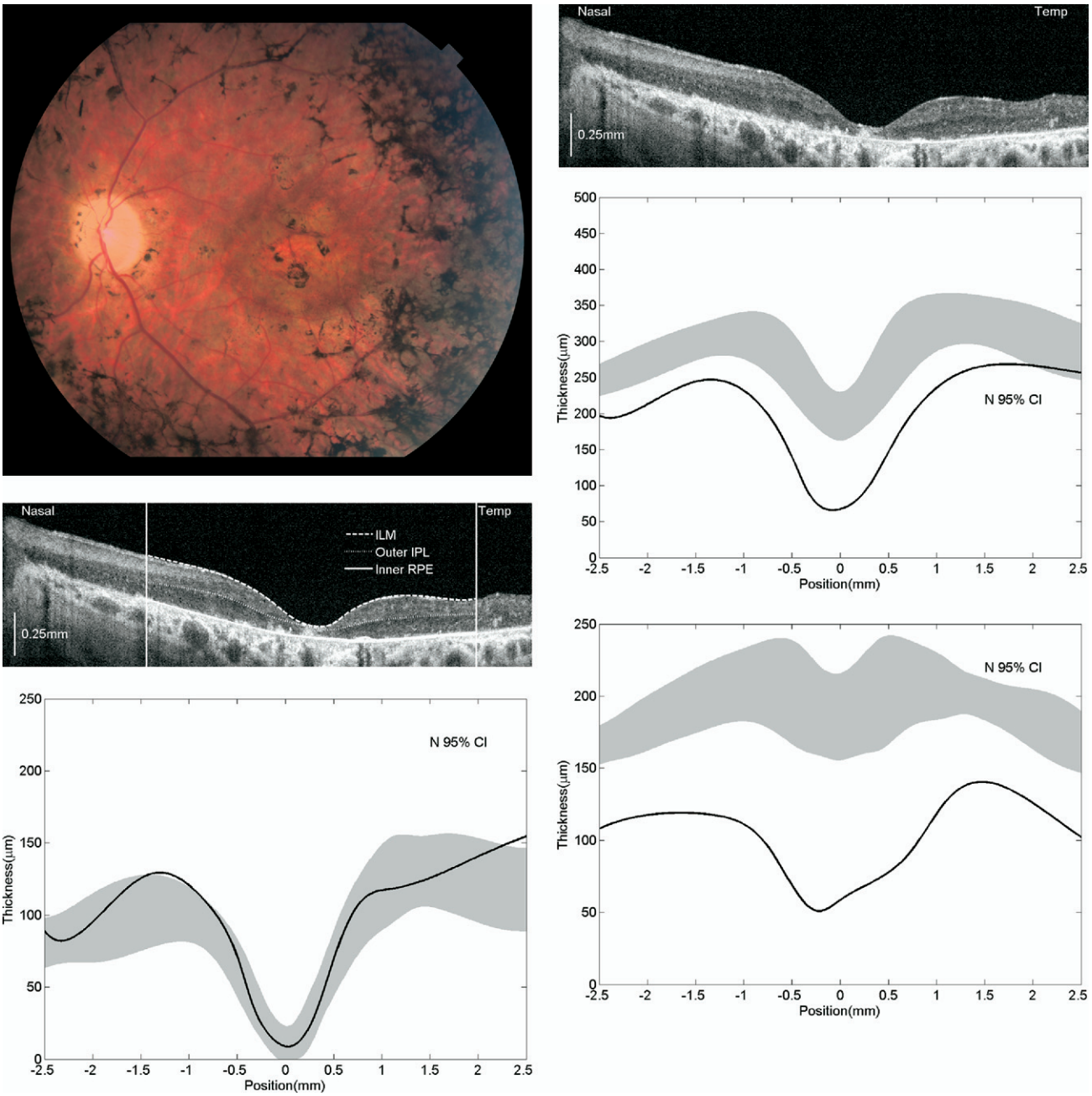

FIGURE 2. Representative retinal dystrophy patient OCT and RT profiles. (Top left) Retinal dystrophy patient fundus photograph. (Top right) Retinal dystrophy patient's OCT image, oriented nasal (left) to temporal (right). Note the generalized thinning of the RPE layer. OCT shows lack of clear demarcation of the outer plexiform layer OPL. (Middle left) Retinal dystrophy patient's OCT image with retinal boundary overlaid. (Middle right) Total RT profile in the retinal dystrophy patient is shown with the normal range. (Bottom left) IRL thickness profile in the retinal dystrophy patient is shown with the normal IRL range with 95% confidence interval (CI) shown in gray. (Bottom right) ORL thickness profile in the retinal dystrophy patient is shown with the normal ORL range with 95% CI shown in gray.

RESULTS

FOURTEEN EYES OF SEVEN RETINAL DYSTROPHY PATIENTS and 14 eyes of seven normal patients underwent RTVue imaging. The 14 eyes of the retinal degeneration patients included diagnoses of RP (six eyes), cone-rod dystrophy (four eyes), and Stargardt disease (four eyes). Patients ranged in age from 33 to 84 years old. There were three male and four female patients, aged 51 ± 14 years, in the retinal dystrophy group. There were three male and

TABLE 1. Retinal Thickness Measurements of Dystrophy Patients

Patient	Site	Diagnosis	Macular IRL (μm)	Macular ORL (μm)	Macular RT (μm)	Foveal IRL (μm)	Foveal ORL (μm)	Foveal RT (μm)	cRT (μm)	logMAR
1	OD	RP	132	114	246	70	112	182	108	0.70
1	OS	RP	121	112	233	66	78	144	99	1.30
2	OD	RP	116	120	237	85	129	214	131	0.30
2	OS	RP	102	103	205	54	101	155	91	0.18
3	OD	RP	121	96	216	85	142	227	148	0.00
3	OS	RP	112	98	210	83	143	226	150	0.00
4	OD	Cone Rod	90	99	188	52	76	128	74	2.00
5	OD	Cone Rod	79	94	173	77	92	169	131	1.00
5	OS	Cone Rod	89	94	182	8495	103	187	155	1.00
6	OD	Stargardt	78	51	129	58	39	97	57	2.00
6	OS	Stargardt	65	94	159	52	83	135	90	2.00
7	OD	Stargardt	86	119	205	45	53	98	41	0.54
7	OS	Stargardt	81	123	204	51	46	98	36	0.70

Cone Rod = cone-rod dystrophy; cRT = central retinal thickness; IRL = inner retinal layer; logMAR = logarithm of minimal angle of resolution; OD = right eye; ORL = outer retinal layer; OS = left eye; RP = retinitis pigmentosa; RT = retinal thickness; Stargardt = Stargardt macular dystrophy.

four female patients, aged 54 ± 10 years, in the normal group.

A representative OCT of a normal patient is shown in Figure 1 with the retinal boundaries drawn. Note that the RPE is thicker subfoveally than more eccentrically from the foveal center. Figure 2 shows a representative example of a retinal dystrophy patient (Figure 2, Top left) with the retinal boundaries drawn (Figure 2, Top right). The RPE is diffusely thinned as compared to the normal patient's RPE. The patient's OCT RT profile shows that the total RT of the retinal dystrophy patient is significantly thinner than the average normal RT range (shaded gray). The IRL RT is within the normal range (Figure 2, Bottom left). The ORL RT profile shows that the ORL thickness is markedly thinned as compared to normal (Figure 2, Bottom right). Thus, the ORL thinning accounts for the overall thinner retina as compared to normals.

Table 1 enumerates the actual RT measurements for each of the retinal dystrophy patients. Representative case studies are given for RP, cone-rod degeneration, and Stargardt disease.

• **CASE 1:** A 31-year-old man with a lifelong history of nyctalopia was diagnosed five years ago with RP. He has no family history of RP. VAs were 20/20 bilaterally. Retinal examination revealed bilateral optic disc pallor, peripheral bone spicules, vascular attenuation, and mild epiretinal membranes (Figure 3, Top left). There were also drusen in the right macular region. An electroretinogram (ERG) showed a barely detectable rod response and a significantly delayed photopic response. Dark adaptation was delayed in both eyes. Goldmann VF testing showed bilateral mild concentric VF contraction with superior field depression. OCT scans (Figure 3, Top right) showed a mildly atrophic retina with some perifoveal cysts present. The RPE was more thinned peripherally than centrally. On the ORL thickness profile, these perifoveal cystic areas appeared as peaks. The remainder of the ORL thickness profile showed the retina was thinner as compared to the normal range (Figure 3, Bottom left). The IRL profile showed the IRL thickness was within the normal range (Figure 3, Bottom right).

• **CASE 2:** A 69-year-old man with a history of amblyopia in his right eye complained of bilateral photopsias that developed six years ago with progressive bilateral VA loss. In March 2005, VAs were 20/80-2 right eye (OD) and 20/60 left eye (OS). Ishihara color plate testing showed only one correct response in each eye. One year later, VAs were 20/200 in each eye. Dilated ophthalmoscopy revealed a bull's-eye lesion in the OD, an atrophic macula in the OS (Figure 4, Top left), and bilateral peripheral pigmentary changes. ERG testing showed decreased photopic and scotopic function consistent with cone-rod dystrophy. Fluorescein angiography (FA) showed a bull's-eye maculopathy. OCT scans (Figure 4, Top right) showed marked choriocapillaris attenuation and central macular thinning. The RPE was more thinned centrally than peripherally. The ORL thickness profile showed generalized thinning as compared with normals (Figure 4, Bottom left). The IRL thickness profile showed some thinning, but much less than the ORL thinning, as compared to controls (Figure 4, Bottom right).

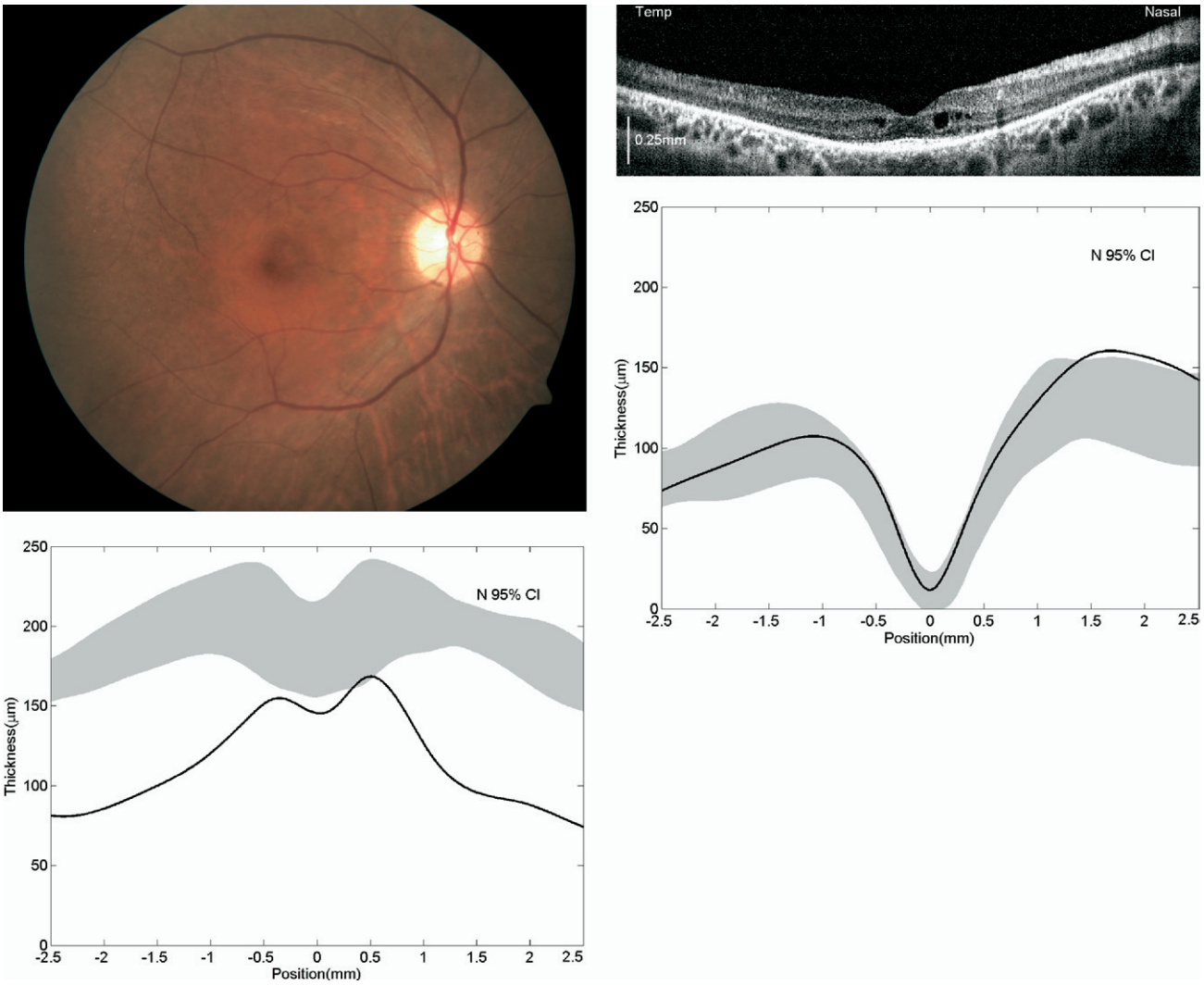

FIGURE 3. Early stage of retinitis pigmentosa (RP) in a 31-year-old patient with bilateral 20/20 visual acuities (VA). (Top left) Color fundus photograph shows a mild macular bull's-eye lesion with arterial attenuation but no bone spicules. (Top right) Horizontal OCT scan, oriented temporal (left) to nasal (right), shows macular thinning and perifoveal cysts. The RPE is thinned peripherally and relatively intact centrally. (Bottom left) Horizontal OCT ORL thickness profile shows that the mean ORL thickness measurements of this patient with early RP are below the normal average ORL thickness measurements. The normal ORL thickness with 95% CIs is shown as a solid gray band. (Bottom right) Horizontal OCT IRL thickness profile shows that the IRL is within the normal IRL range (mean with 95% CI shown in gray).

• **CASE 3:** A 39-year-old woman had a history of metamorphopsia for three years with glare but no nyctalopia. VAs were 20/70 in the OD and 20/100 in the OS. Ishihara color vision testing showed 12 of 14 correct responses in the OD and 11 of 14 correct responses in the OS. Ophthalmoscopy showed pisciform flecks and crystalline retinal deposits (Figure 5, Top left). A FA showed a dark choroid consistent with Stargardt disease. OCT scans revealed central macular thinning with loss of perifoveal structures but with maintained thickness further from the foveal center. The RPE was thinned centrally and more intact eccentrically from the foveal center (Figure 5, Top right). The ORL thickness profile showed markedly abnormal thinning within 1.5 mm of the fovea and less thinning further (more eccentric) from that area (Figure 5, Bottom left). The IRL thickness profile also showed mildly abnormal thinning (Figure 5, Bottom right).

• **CONTROLS:** The seven age-matched (43 to 73 years old) controls had VAs ranging from 20/20 to 20/25. Except for one patient, who had an early nuclear sclerotic cataract in one eye and was pseudophakic in the OS, the eye exams were normal.

• **QUANTITATIVE RESULTS:** In Tables 2 and 3, the averages of the macular RT measurements for the retinal dystrophy patients are presented next to the values for the normal control patients. Except for macular and foveal

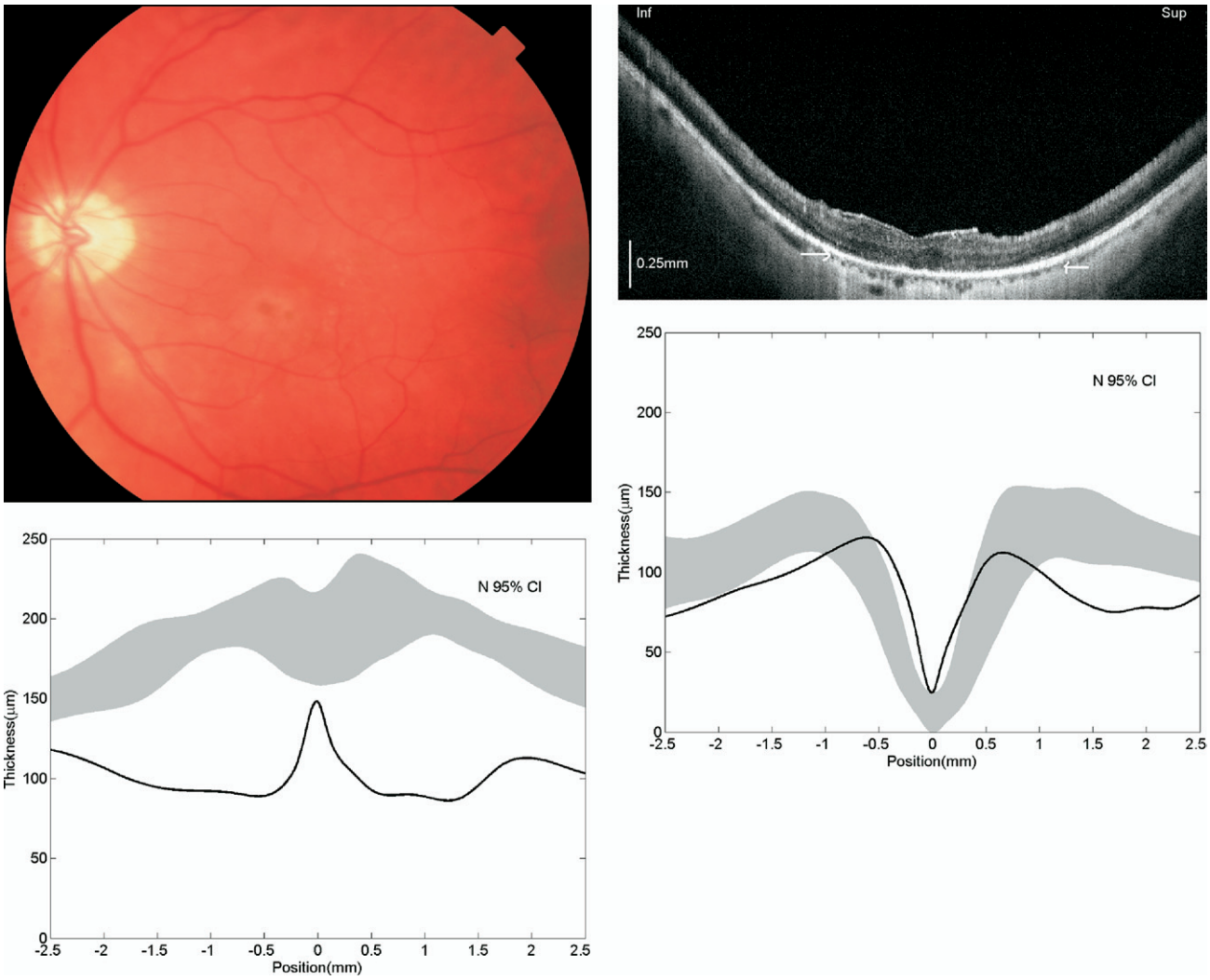

FIGURE 4. Cone-rod dystrophy in a 69-year-old man with VA of 20/80-2 right eye (OD) and 20/60 in the left eye (OS). (Top left) Color fundus photograph shows macular atrophy, vascular attenuation, and absence of RPE pigment spicules. (Top right) Vertical OCT scan, oriented inferior (left) to superior (right), shows macular thinning and marked loss of the choriocapillaris. The RPE is relatively more thinned centrally than peripherally. An overlying epiretinal membrane is visible. (Bottom left) ORL OCT thickness profile shows marked thinning of the macular area ORL thickness as compared with normal ORL thickness measurements (solid gray band). (Bottom right) IRL thickness profile shows this patient's IRL is close to the normal IRL range (normal mean IRL thickness with 95% CI shown in gray).

IRL, average RT measurements (macular ORL, macular RT, foveal cRT, foveal ORL, and foveal RT) of retinal dystrophy patients were significantly thinner than the corresponding measurements in normal patients (*t* test, $P < .001$). For the retinal dystrophy group, macular ORL was decreased 45% compared with normals. For the retinal dystrophy group, macular IRL was decreased 11% compared with normals.

We also calculated the fractional deviation (FD) and standardized deviation for these RT measurements in retinal dystrophy compared with normal controls (Tables 2 and 3). FD was the deviation of the retinal dystrophy group value from the normal controls group value, expressed as a fraction of the average normal controls value. Standardized deviation was defined as the deviation of the retinal dystrophy value from the normal controls value divided by the standard deviation in the normal controls group. FD for cRT, foveal ORL, foveal RT, macular ORL, and macular RT yielded statistically significant results for retinal dystrophy patients compared with the normal controls. Of all the parameters we evaluated, standardized deviation of the macular ORL showed the most statistically significant difference in retinal dystrophy compared with normal controls.

The correlation coefficient values (R^2) between logMAR VAs and RT measurements are given in Table 4. The R^2 were modest and most significant for macular RT and foveal ORL.

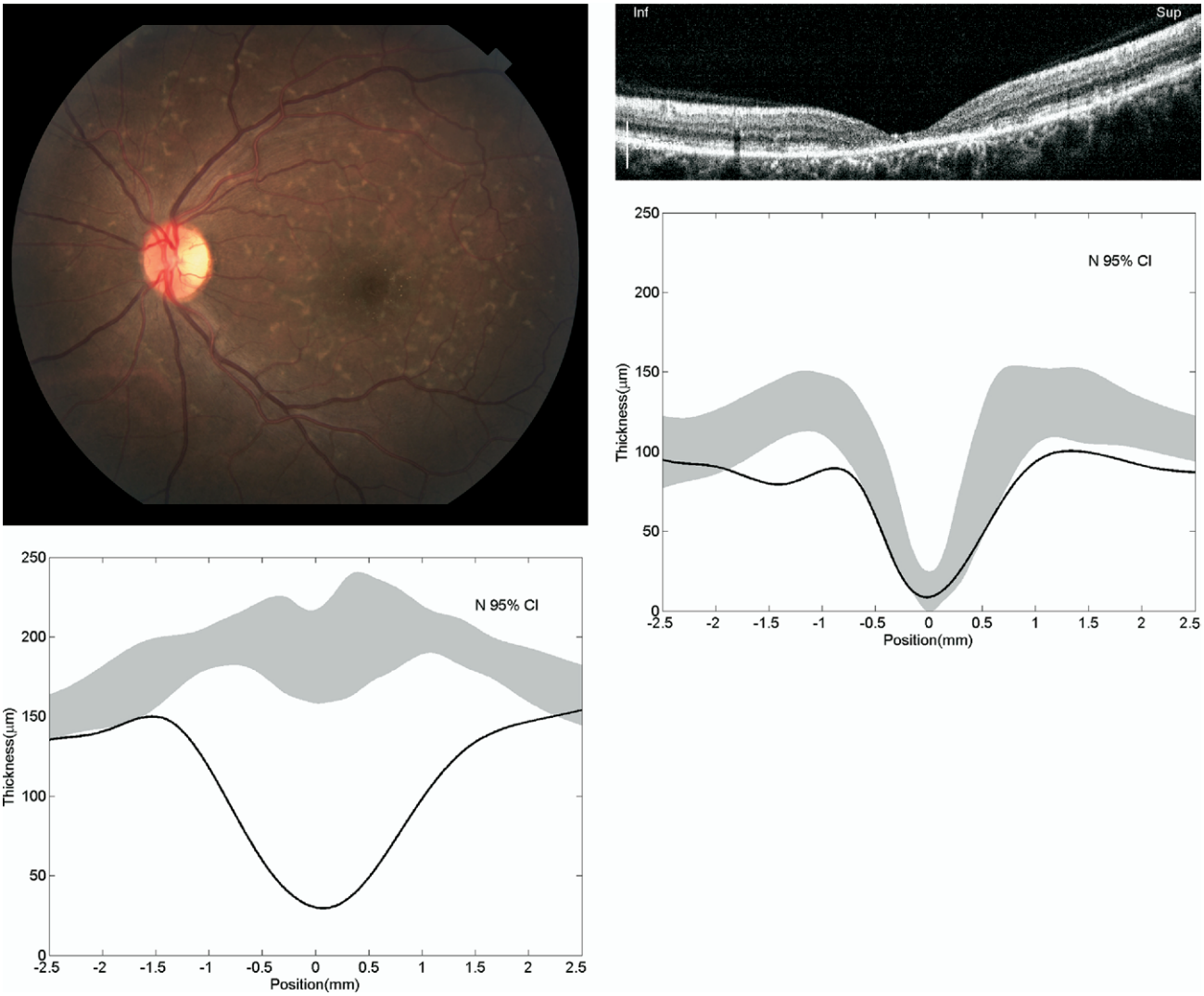

FIGURE 5. Stargardt disease in a 39-year-old woman with VAs of 20/70 in the OD and 20/100 in the OS. (Top left) Color fundus photograph shows numerous pisciform yellow flecks, normal-appearing RPE, and absence of atrophy. (Top right) Vertical OCT scan, oriented inferior (left) to superior (right), reveals a thinned perifoveal retina with normal adjacent RT. The RPE also appears more thinned centrally than peripherally. (Bottom left) ORL thickness profile shows this marked ORL thinning centrally (deep central dip) as compared with normal controls (gray band). (Bottom right) IRL thickness profile shows more decreased central than more peripheral IRL thickness from normals (normal IRL range with 95% CI shown in gray).

The RPE thickness was also significantly different for the dystrophy vs the normal eyes in the foveal area ($P < .001$) (Table 5). The correlation between RPE thickness and logMAR was weak ($R^2 < .04$) and the slope was not significant ($P > .5$).

DISCUSSION

OPTICAL COHERENCE TOMOGRAPHY IMAGING HAS ENabled in vivo cross-sectional analysis of retinal pathology in various diseases. In this pilot study, FD-OCT with its higher resolution and definition facilitated measurements of the thickness of retinal sublayers, allowing us to precisely map the loss of retinal sublayers in retinal degeneration patients as compared with normal patients. We used the line scans for analysis in this study because line scans can be more precisely averaged and are amenable to manual segmentation. In the case of retinal degenerations, the OCT findings parallel those seen on histopathology. Milam and associates showed that the earliest change noted is shortening of the outer segments (OS) of rods and cones.[16] Our finding of a 45% decrease of macular ORL in retinal degeneration patients vs an 11% decrease in IRL is consistent with histopathology results of retinal degeneration patients that show more loss of ORLs than IRLs.[16,17] The presence of significant macular ORL thinning in eyes with good central acuity and very early RP disease mimics

TABLE 2. Comparison of Average Macular Retinal Thickness of Dystrophy Patients With Normals[a]

Group Name	Macular IRL (μm)	Macular ORL (μm)	Macular RT (μm)
Normal	109.9 ± 6.4	182.9 ± 4.7	292.8 ± 8.1
Dystrophy	97.9 ± 20.7	101.3 ± 18.7	199.1 ± 32.6
P value (*t* test)	.064	<.001	<.001
95% CI of difference in means	−29.8, 5.8	−97.6, −65.6	−121.6, −65.7
FD (95% CI)	−0.11 (−0.27, 0.05)	−0.45 (−0.53, −0.36)	−0.32 (−0.42, −0.22)
Standardized deviation (95% CI)	−1.88 (−4.67, 0.90)	−17.25 (−20.63, −13.87)	−11.51 (−14.94, −8.08)

CI = confidence interval; FD = fractional deviation; IRL = inner retinal layer; ORL = outer retinal layer; RT = retinal thickness.
[a]Macular averages are computed in the central 5 mm diameter area.

TABLE 3. Comparison of Foveal Thickness Measurements for Dystrophy Patients With Normals[a]

Group Name	Foveal IRL (μm)	Foveal ORL (μm)	Foveal RT (μm)	cRT (μm)
Normal	71.8 ± 10.2	199.6 ± 14.1	271.3 ± 23.3	195.2 ± 17.5
Dystrophy	66.2 ± 15.0	92.2 ± 34.2	158.4 ± 47.1	100.9 ± 41.0
P value (*t* test)	.28	<.001	<.001	<.001
95% CI of difference in means	−19.8, 8.7	−137.4, −77.3	−155.0, −70.8	−130.4, −58.4
FD (95% CI)	−0.08 (−0.28, 0.12)	−0.54 (−0.69, −0.39)	−0.42 (−0.57, −0.26)	−0.48 (−0.67, −0.30)
Standardized deviation (95% CI)	−0.54 (−1.93, 0.84)	−7.61 (−9.74, −5.48)	−4.84 (−6.65, −3.04)	−5.39 (−7.45, −3.33)

CI = confidence interval; FD = fractional deviation; IRL = inner retinal layer; ORL = outer retinal layer; RT = retinal thickness.
[a]Foveal averages are computed in the central 1.5 mm diameter area.

TABLE 4. Correlation Coefficients of Retinal Thickness Measurements vs Logarithm of Minimal Angle of Resolution Visual Acuity

	R^2	Slope	*P* value
Macular IRL	0.34	−16.37	.04
Macular ORL	0.24	−12.43	.08
Macular RT	0.42	−28.93	.02
Foveal IRL	0.22	−9.53	.11
Foveal ORL	0.40	−29.25	.02
Foveal RT	0.37	−38.80	.03
cRT	0.16	−22.49	.17

cRT = central retinal thickness; IRL = inner retinal layer; ORL = outer retinal layer; RT = retinal thickness.

the histopathology, where thinning and loss of photoreceptors is found before VA is significantly affected.[16]

The finding of reduced macular ORL and foveal ORL thicknesses is similar to the reduced foveal photoreceptor outer segment/pigment epithelial thickness (FOSPET) found using the UHR-OCT.[8] In that UHR-OCT Study, although central foveal thickness was not significantly different between RP patients and normal subjects (P = .103), the difference in FOSPET was significant (52.8 ± 18.3 RP vs 78.6 ± 5.1 normals; P = .003). The authors noted that FOSPET was a way to quantify photoreceptor loss. However, the inner segment (IS)/OS junction is often obliterated in advanced cases of retinal dystrophy. Thus measurement of FOSPET may be problematic in retinal dystrophy patients. The IS/OS junction could also not be detected in these patients in our study. In our study, both macular ORL and foveal ORL were significantly different between the retinal dystrophy and normal patients. Macular ORL and foveal ORL may be useful to help quantify photoreceptor loss in retinal dystrophy patients. Both measurements can be obtained using the RTVue and do not require UHR-OCT.

Although the 5-μm resolution of the RTVue system is slightly lower than the 3-μm resolution of the UHR-OCT systems,[12] 5-μm resolution enabled clear identification of retinal layers and resulted in statistically significant RT differences between retinal dystrophy and normals. The software allowed us to determine IRL, ORL, and RTs at various locations using either automated computer map output or computer-aided caliper spot measurements.

In Witkin and associates' study,[8] VA showed a fair correlation with cRT (Pearson $r = -0.43$, r (2) = 0.187; P = .245) and an excellent correlation with FOSPET (Pearson $r = -0.942$, r (2) = 0.887; $P < .0001$). FOSPET was statistically thinner in RP patients than in normal control eyes and showed correlation with logMAR VA. In our study, logMAR VA correlated better with foveal ORL (1.5 mm) than with macular ORL (5 mm) or with cRT.

TABLE 5. Comparison of Overall Macular Area Retinal Pigment Epithelium Thickness With the Foveal Area Retinal Pigment Epithelium Thickness

Group Name	Macular RPE[a]	Foveal RPE[b]
Normal	20.7 ± 2.2	21.7 ± 2.7
Dystrophy	17.5 ± 5.0	15.8 ± 5.1
P value	.049	.001
95% CI of difference in means	−19.8, 8.7	−10.5, −1.4
FD (95% CI)	−0.08 (−0.28, 0.12)	−0.28 (−0.49, −0.06)
Standardized deviation (95% CI)	−0.54 (−1.93, 0.84)	−2.20 (−3.88, −0.52)

CI = confidence interval; FD = fractional deviation; RPE = retinal pigment epithelium.

The R^2 between RPE and logarithm of minimal angle of resolution is weak ($R^2 < .04$) and the slope is not significant ($P > .5$).

[a]Average RPE thickness in the central 5 mm of the macula.

[b]Average RPE thickness in the central 1.5 mm of the foveal area.

Ergun and associates have found central FT correlated with VA in eyes with Stargardt disease.[13] Transverse photoreceptor atrophy did not correlate with central FT. It remains to be determined whether these transverse areas of atrophy, which were also seen in our present study, are predictive of future progression of the disease.

The ORL thickness profiles were helpful in comparing the location of retinal thinning of retinal dystrophy patients with normals. In RP patients, the ORL thickness profiles show more thinning in macular ORL than foveal ORL early in the disease process. This finding is perhaps consistent with the VF losses beginning peripherally and concentrically contracting in RP. A different pattern is seen in cone-rod and Stargardt disease. Early loss of foveal ORL is seen in cone-rod and Stargardt disease with relative preservation of the macular ORL. This pattern is consistent with the early central visual loss in these diseases. We also noted thinning of the macular choriocapillaris in the cone-rod dystrophy patients as compared to the RP patients and controls. Further work is needed to determine the implications of our OCT findings as prognostic indicators for visual progression in patients with retinal dystrophy.

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Biosketch

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Biosketch

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