Morphologic Characteristics of the Outer Retina in Cone Dystrophy on Spectral-domain Optical Coherence Tomography

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Purpose: To investigate the morphologic changes in the outer retina of patients with cone dystrophy, using spectral-domain optical coherence tomography (SD-OCT).

Methods: The medical records of 15 cone dystrophy patients examined from January 2007 to January 2012 were reviewed retrospectively. All patients underwent ophthalmic evaluation including best-corrected visual acuity (BCVA), color vision testing, fundus examination, full-field standard electroretinography (ERG), multifocal (mf) ERG, and SD-OCT. Qualitative and quantitative SD-OCT data and ERG responses were analyzed and compared among the patient categories and the normal control group.

Results: There were 4 major categories of SD-OCT findings, based on the status of the ellipsoid portion of the photoreceptor inner segment (ISe), outer segment (OS) contact cylinder, and retinal pigment epithelium (RPE) layer. Category 0 showed no structural abnormalities. Category 1 showed foveal ISe loss and obscurity of the border between the ISe band and the external limiting membrane (ELM). Category 2 showed foveal thinning and focal foveal ISe disruption with an intact ELM. Category 3 showed foveal thickening and perifoveal disruption of the ISe layer. Category 1 to 3 showed OS contact cylinder layer absence and RPE thickening. The patients in category 0 tended to be younger (mean, 10.0 years) than those in categories 1 to 3 (mean, 17.6 years), although this difference was not statistically significant. Category 1 to 3 patients exhibited statistically significant thinning of the central retina and outer nuclear layer and thickening of the RPE layer relative to the category 0 and normal control group. There was a significant correlation between the central foveal thickness and BCVA in the patients with cone dystrophy. ERG and mfERG responses did not differ significantly among the different cone dystrophy categories.

Conclusions: The morphologic features of cone dystrophy as revealed by SD-OCT, could be categorized as either normal or 1 of 3 different types of outer retinal changes. The presence of normal retinal structures in young cone dystrophy patients with functional impairment (category 0) indicates that electrophysiologic studies are superior to current imaging modalities for the early diagnosis of cone dystrophy. The characteristic SD-OCT findings in cone dystrophy patients may aid in differential diagnosis and be useful for future research on the pathology of cone dystrophy.

Key Words: Cone dystrophy, Electroretinography, Photoreceptor cells, Spectral-domain optical coherence tomography

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Cone dystrophy is an inherited retinal disorder characterized by the loss of cone cells- the photoreceptors responsible for central and color vision. The most common symptoms are progressive vision loss, poor color vision, and sensitivity to bright light. Full-field electroretinography (ERG), which demonstrates reduced single-flash and flicker responses in the photopic condition and a normal

response in the scotopic condition, is the gold-standard test.

The widespread adoption of high-resolution spectral-domain optical coherence tomography (SD-OCT) enabled investigators to demonstrate structural abnormalities in the retinal layers and their correlations with functional status in various retinal diseases, such as acute zonal occult outer retinopathy-complex diseases [1], epiretinal membrane [2], macular hole [3,4], and retinitis pigmentosa [5]. Several investigators have studied the retinal structural changes in cone dystrophy patients using SD-OCT. Birch et al. [6] evaluated the effects of selective rod or cone cell loss on SD-OCT findings in patients with cone dystrophy or retinitis pigmentosa and reported that the normalized product of the thickness of the outer nuclear layer (ONL) and the summed thickness of the photoreceptor outer segment (OS) and retinal pigment epithelium (RPE) was proportional to the visual field sensitivity. Hood et al. [7] studied patients with cone dystrophy or achromatopsia and showed that the intensity of the inner segment ellipsoid (ISe) band, previously called the inner segment (IS)-OS junction band, of the photoreceptor seen by SD-OCT was lower in patients with diminished cone function than in healthy controls. Each of those studies included only 6 patients with cone dystrophy, which is a relatively small number, and did not show the morphologic SD-OCT features of the outer retina in detail.

In the present study, we attempted to analyze the structural changes in the outer retinas of 15 patients with cone dystrophy using SD-OCT and related the structural changes to the functional and electrophysiologic findings.

Materials and Methods

The medical records of 15 patients diagnosed with cone dystrophy from January 2007 to January 2012 were retrospectively reviewed. The diagnostic criteria for cone dystrophy are as follows; a substantial decrease in cone cell function with relative preservation of rod cell function, which can be demonstrated by a full-field ERG, and a history of progressive visual loss, photophobia, and poor color vision (Fig. 1).

All of the patients underwent full ophthalmic evaluation including best corrected visual acuity test, manifest or cycloplegic refraction examination, color vision testing, slit lamp examination, fundus examination, full-field standard ERG, multifocal ERG (VERIS II; Electro-Diagnostic Imaging Inc., San Francisco, CA, USA), and SD-OCT (Spectralis OCT; Heidelberg Engineering, Heidelberg, Germany). Some of the patients also underwent visual field testing by Humphrey or Goldmann perimetry, fluorescein angiography, and fundus autofluorescence imaging.

The retinal structural changes on the SD-OCT images were analyzed and the findings were compared between

the patients and the normal control group. The normal control group was comprised of individuals who were diagnosed as normal based on the results of ophthalmic examination including SD-OCT. The 4 hyper-reflective bands in the outer retina visible on SD-OCT images were analyzed qualitatively based on the model proposed by Spaide and Curcio [8]. The first (innermost) band corresponds to the external limiting membrane (ELM), the second (beneath the first band) to the ellipsoid portion of the inner segments (ISe), the third band (beneath the ISe band) to a structure known as the OS contact cylinder, which is an ensheathment of the cone outer segments by an apical process of the RPE, and the last, outermost band to the RPE (Fig. 2A). A retina specialist (SJW) and an ophthalmologist (SCC) thoroughly evaluated the SD-OCT images of all 30 eyes from the 15 patients.

The quantitative data included the thickness of the retinal layers as measured by SD-OCT and the ERG amplitudes. The retinal thickness profiles included the central foveal thickness (CFT) and the thickness of the ONL, photoreceptor (PR) and RPE (PR + RPE; defined as the distance between the outer margin of the RPE and the outer margin of the ELM), and RPE. The ERG data included the full-field standard ERG amplitude (scotopic maximal response, photopic response, and 30-hertz flicker response) and the multifocal ERG innermost ring amplitude.

The eyes were classified into 4 different categories based on the morphologic retinal abnormalities on the SD-OCT images, and the qualitative and quantitative data were compared between the patients and the normal control group.

The Mann-Whitney *U*-test was used for inter-group comparison of the quantitative data. Student's *t*-test was used to compare the quantitative data between the total patient group and the normal control group. Statistical analysis was performed using commercial software (SPSS ver. 18.0; SPSS Inc., Chicago, IL, USA). A *p*-value less than 0.05 was regarded to be statistically significant.

Results

Seven of the 15 patients were men and 8 were women. The mean ($\pm$ SD) age was 15.6 ± 12.7 years (range, 5 to 46 years). The best-corrected visual acuities (BCVA) of the patients' eyes ranged from 20 / 500 to 20 / 30. All patients showed ERG findings consistent with cone dystrophy. No patient had any afferent pupillary defect or visual pathway abnormality.

Of the 5 patients who had undergone Humphrey perimetry, 1 had deep central scotomas within the central 5 degrees of fixation of both eyes (case 4). Another patient had a unilateral central scotoma in the right eye and a generalized reduction of sensitivity in the left eye (case 9) (Fig. 1G and 1I). Three patients had bilateral reduction of sensitivity

Fig. 1. A 21-year-old man (case 9) with cone dystrophy. (A,B) Fundus photographs showing focal degeneration around the fovea in both eyes. (C,D) Fundus fluorescein angiography showing normal angiographic patterns in the (C) early and (D) late phases. (E,F) Optical coherence tomography showing irregular loss of the ellipsoid portion of the photoreceptor inner segment (ISE) in both eyes. (G,I) Visual field tests by Humphrey field analyzer central 24-2 threshold testing showing a deep central scotoma in the (G) right eye and generalized reduction of sensitivity in the (I) left eye. (H,J) Full-field standard electroretinography showing decreased amplitudes of the photopic (graph of the fourth row) and 30-Hz flicker response (graph of the fifth row) in both eyes. (K,L) Trace arrays of multifocal electroretinography showing decreased central retinal responses in both eyes.

in the central visual field (cases 3, 8, and 14).

SD-OCT demonstrated morphologic changes in the outer retinal layers. Of the 30 eyes of the 15 patients, 22 eyes (of 11 patients) showed changes in the outer retinal layers in both eyes. Another 8 eyes (in 4 patients) showed no definite structural changes on OCT. There were 4 major categories of SD-OCT findings, based on the status of the ISe band, OS contact cylinder layer, and RPE layer. Both eyes for each individual showed symmetric categorization. The first category (category 0: cases 5, 6, 10, and 13) (Fig. 2B) showed normal SD-OCT findings. The second category (category 1: cases 1, 2, 8, and 9) (Fig. 2C) showed irregular foveal loss of the ISe band and obscurity of the border between the ISe band and the ELM due to the accumulation of reflective material between the 2 layers. The third category (category 2: cases 3, 7, 11, 12, 14, and 15) (Fig. 2D) showed retinal thinning around the fovea and segmental

(or irregular) foveal loss of the ISe band. The last category (category 3: case 4) (Fig. 2E) showed ISe band thickening around the fovea and irregular perifoveal loss of the ISe band. In category 1 to 3, the OS contact cylinder layer was not discernible and the RPE layer was thickened (Fig. 2).

The demographic, quantitative, and qualitative SD-OCT data for the patients and the normal control group are shown in Tables 1 and 2, and Fig. 3. The patients in category 0 tended to be younger (mean, 10.0 years) than those in categories 1 to 3 (mean, 17.6 years), although this difference was not statistically significant (Mann Whitney *U*-test, $p = 0.317$). The CFT, ONL, PR + RPE and PRE thicknesses did not differ significantly between the normal control group and category 0 patients. CFT, ONL, and PR+RPE thicknesses were significantly lower in category 1 to 3 patients than in the normal control group or the cone dystrophy category 0 patients. It is noteworthy that

the RPE (the outermost high signal band) thickness was greater in category 1 to 3 patients than in the normal con-

trol group or the category 0 patients. The CFT was significantly lower in category 2 patients than in category 1 or 3

Fig. 2. Spectral-domain optical coherence tomography (SD-OCT) horizontal scan images of a normal control eye (A) and representative cases of each category of cone dystrophy patients (B–E). (A) SD-OCT of normal eyes showed 4 distinct high-signal bands in the outer retinal layer OS. (B) Category 0, OS, case 6, with normal SD-OCT findings. (C) Category 1, OD, case 8, with irregular foveal loss of the ellipsoid portion of the inner segment (ISe) band and obscurity of the border between the ISe and external limiting membrane. (D) Category 2, OS, case 3, with central retinal thinning and segmental foveal loss of the ISe band. (E) Category 3, OD, case 4, with central foveal thickening of the ISe band and irregular perifoveal loss of the ISe band. Loss of the outer segment contact cylinder layer and the thickening of the retinal pigment epithelium layer were found in category 1 to 3 patients.

Table 1. Patient demographic and ophthalmic examination data

Case	Age (yr)	Sex	Symptom duration (yr)	Familial history	BCVA		OCT findings category		Center foveal thickness (μ m)		Photopic response amplitude (μ V)		30-Hz flicker response amplitude (μ V)	
					OD	OS	OD	OS	OD	OS	OD	OS	OD	OS
1	8	F	4	None	20 / 50	20 / 50	1	1	216	219	25.0	-6.5	14.7	17.8
2	11	M	11	None	20 / 125	20 / 200	1	1	174	163	27.3	15.7	15.0	17.0
3	17	F	5	None	20 / 200	20 / 200	2	2	143	140	9.1	9.1	9.1	9.1
4	18	M	3	Yes*	20 / 100	20 / 30	3	3	188	193	13.8	19.3	12.6	12.4
5	11	F	7	AR	20 / 50	20 / 80	0	0	228	231	11.8	14.7	8.8	11.8
6	13	F	6.5	AR	20 / 80	20 / 50	0	0	242	227	22.4	31.0	16.7	8.5
7	5	F	3	None	20 / 250	20 / 500	2	2	133	128	10.0	10.0	5.9	4.1
8	44	M	NA	AR	20 / 200	20 / 200	1	1	179	180	15.0	10.0	12.5	10.0
9	21	M	15	Yes*	20 / 100	20 / 100	1	1	175	173	11.8	11.8	10.0	5.9
10	7	F	3.5	None	20 / 125	20 / 50	0	0	209	209	11.7	15.3	11.8	5.9
11	46	M	NA	None	20 / 200	20 / 125	2	2	97	111	16.5	6.6	3.9	4.7
12	5	M	2	XR	20 / 100	20 / 100	2	2	160	145	6.1	21.8	7.1	8.8
13	9	F	3.5	None	20 / 63	20 / 50	0	0	210	213	58.7	59.1	27.6	32.5
14	14	F	5	None	20 / 125	20 / 125	2	2	80	69	12.9	12.5	12.5	20.0
15	5	M	1.5	None	20 / 50	20 / 80	2	2	163	170	15.9	4.3	20.7	17.8

BCVA = best-corrected visual acuity; OCT = optical coherence tomography; AR = autosomal recessive; NA = not available; XR = X-linked recessive.

*Indicates cases with present familial history, but limited information about the inherited pattern.

Table 2. Comparison of OCT and ERG data between cone dystrophy categories and normal controls

			Cone dystrophy				Normal Controls	<i>p</i> -value (patient vs. normal)	
			Categorized group						
			0	1	2	3			Total
Basic characteristics	Eyes		8 (26.7)	8 (26.7)	12 (40)	2 (6.7)	30 (100)	40	
	Age (yr)		10.0 ± 2.4 [*]	21.0 ± 15.1	15.3 ± 15.2 [*]	18	15.6 ± 12.7 [*]	29.2 ± 10.3	<0.001
	BCVA (logMAR)		0.50 ± 0.17 [*]	0.75 ± 0.25 [*]	0.85 ± 0.27 [*]	0.44 ± 0.37	0.70 ± 0.28 [*]	0.10 ± 0.16	<0.001
SD-OCT outer retinal structural findings	ISe		Normal	Irregular foveal loss + obscure ELM border	Segmental (or irregular) loss + foveal thinning	Irregular perifoveal loss + foveal thickening		Normal	
	Contact cylinder layer		Normal	Absence	Absence	Absence		Normal	
	RPE		Normal	Thickening	Thickening	Thickening		Normal	
SD-OCT thickness data	CFT		221.1 ± 12.5	184.9 ± 20.8 [*]	128.3 ± 32.7 [*]	190.5 ± 3.5 [*]	172.3 ± 45.5 [*]	228.6 ± 13.0	<0.001
	ONL		91.0 ± 7.8	70.0 ± 19.5 [*]	38.8 ± 17.6 [*]	67.5 ± 3.5 [*]	63.0 ± 26.4 [*]	99.1 ± 12.5	<0.001
	PR + RPE		96.6 ± 5.5	64.9 ± 4.9 [*]	63.8 ± 17.5 [*]	78.5 ± 9.2 [*]	73.8 ± 18.5 [*]	101.2 ± 5.9	<0.001
	RPE		22.9 ± 1.7	32.6 ± 2.3 [*]	32.0 ± 2.3 [*]	29.5 ± 2.1 [*]	29.6 ± 4.7 [*]	24.1 ± 1.6	<0.001
ERG amplitude	Standard ERG	Scotopic maximal	327.9 ± 118.2	281.6 ± 83.6	274.3 ± 56.7	233.0 ± 25.5	287.8 ± 83.9	343.9 ± 125.3	0.066
		Photopic	28.1 ± 20.1 [*]	13.8 ± 10.3 [*]	11.2 ± 5.0 [*]	16.6 ± 3.9 [*]	17.9 ± 13.8 [*]	72.4 ± 24.7	<0.001
		30-Hz flicker	15.5 ± 9.6 [*]	12.9 ± 4.0 [*]	10.3 ± 6.1 [*]	12.5 ± 0.1 [*]	12.5 ± 6.7 [*]	63.8 ± 24.5	<0.001
		Multifocal ERG	64.2 ± 38.1 [*]	86.0 ± 100.4	94.2 ± 94.4	63.2 ± 6.8	79.6 ± 74.4	121.9 ± 25.9	0.129

Values are presented as number (%) or mean ± SD.

SD-OCT = spectral-domain optical coherence tomography; ERG = electroretinography; logMAR = logarithm of the minimum angle of resolution; ISe = ellipsoid portion of photoreceptor inner segment; ELM = external limiting membrane; CFT = central foveal thickness; ONL = outer nuclear layer; PR = photoreceptor; RPE = retinal pigment epithelium.

*Indicates a statistically significant difference compared with the normal control group ($p < 0.05$).

patients ($p < 0.001$ and $p = 0.022$, respectively).

To investigate the structure-function relationship in cone dystrophy patients, we compared the BCVA among the patient categories (Fig. 4). The mean BCVAs of category 1 and 2 patients were significantly worse than that of category 0 patients ($p = 0.031$ and $p = 0.006$, respectively). CFT and BCVA correlated significantly in cone dystrophy patients (Spearman's correlation coefficient = 0.648, $p < 0.001$).

In the standard full-field ERG data, the scotopic maximal response did not differ significantly between cone dystrophy patients and the normal control group (Fig. 5A). The photopic and 30-Hz flicker responses were lower in the cone dystrophy category 0 to 3 patients than in the normal control group ($p < 0.001$) (Fig. 5B and 5C). The scotopic maximal, photopic, and 30-Hz flicker responses did not differ among patient categories (category 0 to 3). CFT and photopic response correlated significantly in cone dystrophy patients (Spearman's correlation coefficient = 0.371, $p = 0.043$). Although the correlation between CFT and 30-Hz flicker response was not statistically significant ($p = 0.094$), ONL thickness and 30-Hz flicker response correlated significantly in cone dystrophy patients (Spearman's correlation coefficient = 0.375, $p = 0.041$).

The multifocal ERG innermost ring amplitude was sig-

nificantly lower in category 0 patients than in the normal control group ($p = 0.005$) but did not differ significantly among the patient categories (Fig. 5D).

Discussion

This study described in depth and categorized the outer retinal morphologic abnormalities in cone dystrophy patients using SD-OCT. To our knowledge, this is the first such report. In our study, structural abnormalities of the outer retinal layers on SD-OCT images were found in 73.3% (11 / 15) of the cone dystrophy patients, and the morphologic features on SD-OCT could be categorized into 4 distinct patterns. With the exception of category 0, which demonstrated no structural changes on SD-OCT, absence of the OS contact cylinder layer and thickening of the RPE layer were found in all of the patient categories, indicating that the reflectivity of the 2 layers was merged due to structural damage to the corresponding layers. This finding also suggests that the retinal pathology of cone dystrophy occurs mainly between the photoreceptor outer segment and RPE layer.

Eyes with cone dystrophy showed decreased CFT due to thinning of the outer retinal layers. In addition, thinning of the central retinal layers correlated with visual acuity and

Fig. 3. Optical coherence tomography thickness in the normal control and cone dystrophy grouped categories. (A,B,C) Central foveal thickness (CFT), outer nuclear layer (ONL), and photoreceptor (PR) + retinal pigment epithelium (RPE) thicknesses were significantly lower in category 1 to 3 eyes than in normal control or category 0 eyes. (D) RPE thickness was greater in category 1 to 3 eyes than in normal control or category 0 eyes. The open circles reveal the means of the thicknesses, and the vertical error bars represent the standard deviation. The asterisk indicates a statistically significant difference in thickness ($p < 0.05$).

electrophysiologic function in eyes with cone dystrophy. Lim et al. [9] demonstrated that eyes with retinal dystrophy (cone-rod dystrophy, retinitis pigmentosa, and cone-rod degeneration) had a small (11%) decrease in the thickness of the macular inner retinal layer (inner plexiform layer, ganglion cell layer, and nerve fiber layer) and a severe (45%) decrease in the thickness of the macular outer retinal layer, which is consistent with our results. Choi et al. [10] used an adaptive optics fundus camera to demonstrate extensive space between cone cells in patients with retinal dystrophy (rod-cone dystrophy, cone-rod dystrophy, and juvenile macular dystrophy), which is also consistent with our finding that the photoreceptor layer is the major site of structural impairment on SD-OCT.

Although category 0 patients showed no retinal structural abnormalities on SD-OCT, they exhibited electrophysiologic dysfunction comparable to that of patients in the other categories of cone dystrophy. The young age of this group implies that their structural abnormality can not be visualized on SD-OCT in this stage despite the significant functional expression. This finding indicates that ERG is more sensitive and can be used to diagnose cone dystrophy earlier than current SD-OCT techniques. Further research is necessary, as future imaging modalities such as the adaptive optics technique may be able to detect structural abnormalities in category 0 patients.

Category 1 patients showed irregular foveal ISe band disruption and obscurity of the border between the ISe

layer and the ELM. Kim et al. [11] reported a similar structural abnormality in a 13-year-old girl with cone-rod dystrophy 6 with mutations in the retinal guanylate cyclase gene (GUCY2D). They demonstrated similar irregular photoreceptor loss at the IS-OS junction (ISe) and in the OS (OS contact cylinder) layer, and showed that the photoreceptor abnormalities on SD-OCT corresponded to the perifoveal ring seen by autofluorescence imaging of the patient. Querques et al. [12] also showed similar disruption and focal loss of the IS-OS junction in the case of a 47-year-old woman with fundus albipunctatus associated with cone dystrophy.

Category 2 patients revealed distinct foveal thinning and segmental loss or irregular disruption of the ISe layer. The central foveal thickness was lower in this category than in any of the other patient categories. Segmental ISe layer defects were previously reported by Hood et al. [7] in a study of cone dystrophy and achromatopsia. Emfietzoglou et al. [13] also reported a similar total absence of the IS-OS junction layer (ISe layer) and foveal thinning in the case of a 25-year-old man with cone-rod dystrophy.

Category 3 patients demonstrated foveal ISe layer thickening. This type of morphologic abnormality was found only in case 4. The accumulation of material consequent to cone cell dysfunction could be the reason for the ISe thickening. This type of structural change was reported by Duncan et al. [14] in a study of cone-rod dystrophy patients with peripherin/RDS mutations.

We showed that the outer retinal SD-OCT findings of cone dystrophy patients are heterogeneous; this diverse pathology may stem from genetic diversity. Several research-

ers have suggested that cone dystrophy arises from diverse genetic sources with considerable variability in clinical consequences [15]. It is noteworthy that patients in different categories of morphologic abnormality showed similar electrophysiologic responses. This important result indicates that the morphologic characteristics demonstrated by SD-OCT could possibly distinguish patients with different genotypes, while the electrophysiologic study could not. Further genetic study must be performed to test this hypothesis.

In addition to cone dystrophy, some diseases such as acute zonal occult outer retinopathy (AZOOR) complex and occult macular dystrophy (OMD) also show abnormalities on OCT and electrophysiologic test. AZOOR complex is a disease that includes acute idiopathic blind-spot enlargement syndrome, multiple evanescent white dot syndrome, punctate inner choroidopathy, multifocal choroiditis and panuveitis, and acute macular neuroretinopathy. AZOOR complex is similar to cone dystrophy in that the IS/OS boundary defect is found in the OCT. (In a study by Spaide et al. [1] for the 18 eyes with AZOOR complex, patients with blind spot enlargement had outer retinal damage surrounding the optic nerve head while those without blind spot enlargement did not.) Unlike in cone dystrophy, the main symptoms in AZOOR complex are acute loss of one or more zones of visual field and photopsia. In cone dystrophy, only cone response diminishes in the standard full-field ERG while diverse ERG findings are observed in AZOOR complex cases (In a study by Gass et al. [16] for the 90 eyes with AZOOR, both scotopic and photopic function were abnormal in 55 eyes, only the photopic function was abnormal in 16 eyes, and only the scotopic function was abnormal in 16 eyes.) OMD is similar to cone dystrophy in that disruption of the IS/OS junction is observed in the OCT [17]. However, OMD is different from cone dystrophy in that while the standard full-field ERG is normal, the foveal amplitude of the multifocal ERG is decreased.

The limitation of our study is the lack of genotypic data in the enrolled patients. As stated above, the distinct patterns of structural abnormality may represent different patient genotypes. We recently began a genetic study on cone dystrophy patients, and we hope that the genotype-phenotype relationship will be revealed in the near future.

In conclusion, the structural findings on SD-OCT images in cone dystrophy could be classified into 4 major categories based on the morphologic changes in the retinal outer layer. The presence of normal retinal structures in young patients with functional impairment indicates that electrophysiologic studies are superior to current imaging modalities for the early diagnosis of cone dystrophy. The characteristic SD-OCT findings in cone dystrophy patients may aid in differential diagnosis and be useful for future research on the pathology of cone dystrophy.

Fig. 4. Best-corrected visual acuity (BCVA) in the cone dystrophy category. The open circles reveal the means of the BCVA, and the vertical error bars represent the standard deviation. The asterisk indicates a statistically significant difference in BCVA ($p < 0.05$). logMAR = logarithm of the minimum angle of resolution.

Fig. 5. Full-field and multifocal electroretinography (mfERG) amplitudes in the normal control and cone dystrophy grouped categories. (A) Scotopic maximal response. (B) Photopic response. (C) 30-Hz flicker response. (D) mfERG ring 1 amplitude. The open circles reveal the means of the amplitudes and the vertical error bars represent the standard deviation. The asterisk indicates a statistically significant difference in amplitude ($p < 0.05$).

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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