

## En Face Optical Coherence Tomographic Findings in a Patient with Bietti Crystalline Dystrophy

Shinji Makino<sup>\*1</sup>, Yuji Hosoda<sup>2</sup>, Katsuhisa Endoh<sup>3</sup>

<sup>1</sup>Department of Ophthalmology, Jichi Medical University, Shimotsuke, Tochigi 329-0498, Japan

<sup>2</sup>Hosoda Eye Clinic, Kai, Yamanashi 400-0125, Japan

<sup>3</sup>Ichijokai Clinic, Matsudo, Chiba 270-2224, Japan

\*Corresponding author: Shinji Makino  
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### Abstract

### Case Report

We investigated a patient with Bietti crystalline dystrophy (BCD) examined by *en face* optical coherence tomography (*en face* OCT). Although multiple tiny refractile yellowish crystals were detected throughout the posterior pole, their crystals were located not only at the level of the retinal pigment epithelium but also at the level of the ellipsoid zone. *En face* OCT findings may contribute to a better understanding of the location of the retinal crystals.

**Keywords:** Bietti crystalline dystrophy, *en face* optical coherence tomography.

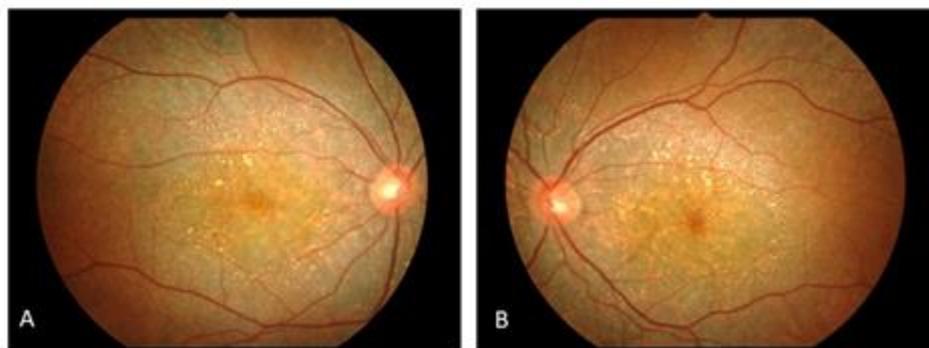
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## INTRODUCTION

Bietti crystalline dystrophy (BCD) is characterized by deposits of crystals in the marginal cornea and the paracentral and peripapillary retina [1-3]. Previously, optical coherence tomography (OCT) was shown to detect crystalline deposits in the different retinal layers and in the choroid [4-7]. However, there is few reports documented the location of retinal crystals using *en face* OCT [5-7]. To discuss the location of the retinal crystals, we investigated a patient with BCR using multimodal imaging including *en face* OCT.

## CASE REPORT

A 49-year-old woman was referred for ocular fundus abnormality by medical checkup. She had no subjective complaints. Her best-corrected visual acuity was 1.5 in the both eyes. The anterior segment was normal in both eyes with no crystals visible at the corneal limbus. Fundus examination revealed intraretinal crystals in the posterior pole and midperipheral retina associated with chorioretinal atrophy (Figure 1).

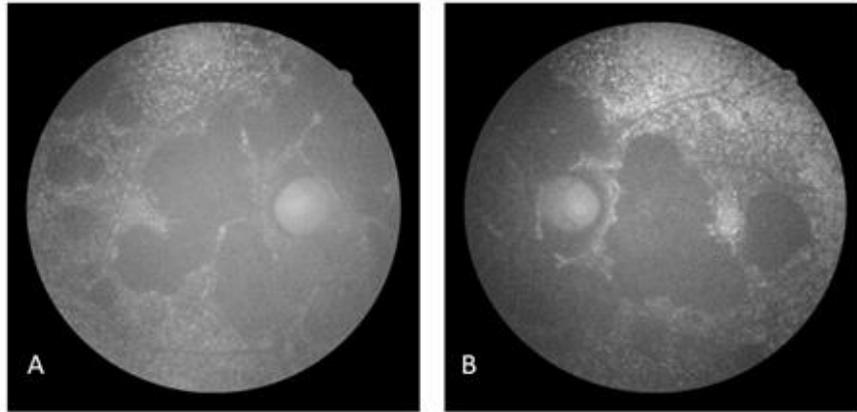


**Fig-1: Fundus photographs of the (A) right and (B) left eyes**

Note the multiple tiny refractile yellowish crystals throughout the posterior pole.

Fundus autofluorescence (FAF) showed well defined sharply demarcated patches of hypoautofluorescence around the macular area involving the peripapillary zone in both eyes (Figure 2).

The area outside of these patches of hypoautofluorescence showed granular hyperfluorescence.

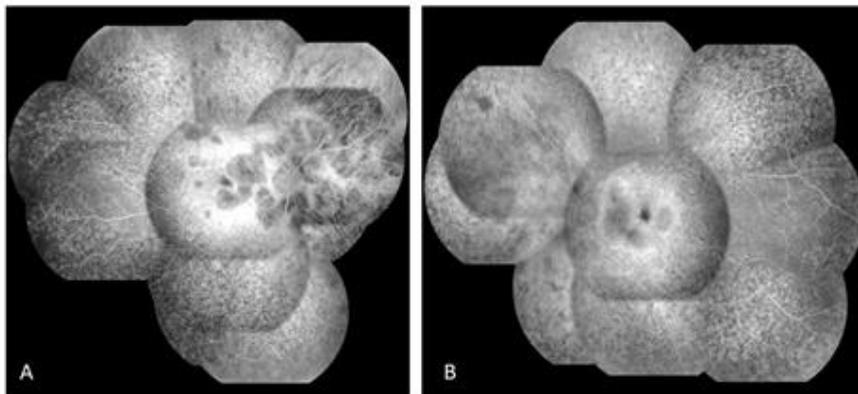


**Fig-2: Fundus autofluorescent (FAF) imagings of the (A) right and (B) left eyes**

Note the well-defined hypoautofluorescence and the granular hyperautofluorescence areas.

Fluorescein angiography (FA) revealed a transmission defect with granular hyperfluorescence

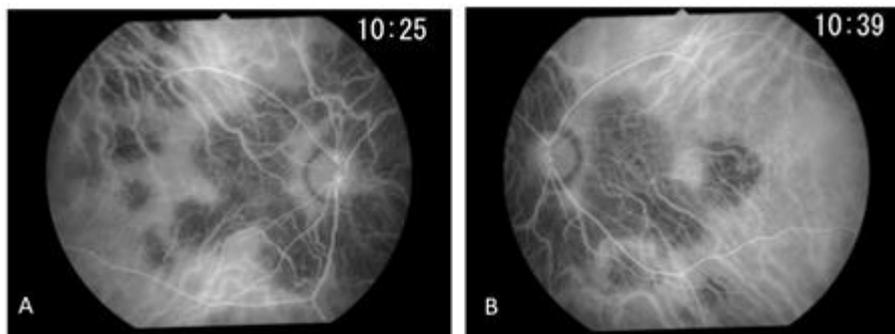
and hypofluorescent areas corresponding to atrophy or loss of choriocapillaris in both eyes (Figure 3).



**Fig-3: Fluorescein angiography of the (A) right and (B) left eyes**

Note the granular hyperfluorescence and hypofluorescent areas.

Indocyanine green angiography demonstrated hypofluorescent areas corresponding to atrophy or loss of choriocapillaris in both eyes (Figure 4).



**Fig-4: Indocyanine green angiography of the (A) right and (B) left eyes**

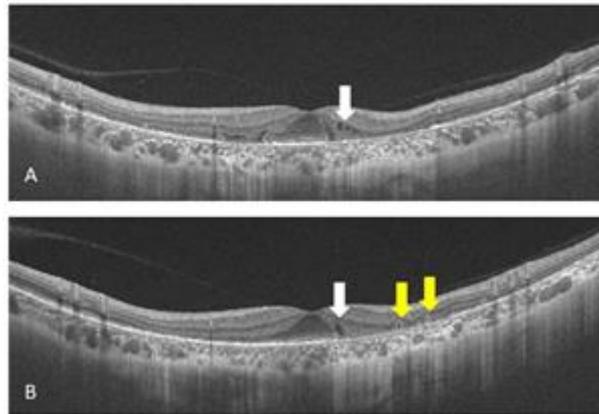
Note the well-defined hypofluorescence area

OCT scan through the fovea of both eyes showed relatively preserved foveal contour and retinal

layers. Although the ellipsoid zone (EZ) was not disrupted, pseudocyst and outer retinal tabulation were

detected (Figure 5). In addition, the crystals were appreciable as small faintly hyperreflective dots above

the level of the retinal pigment epithelium (RPE).

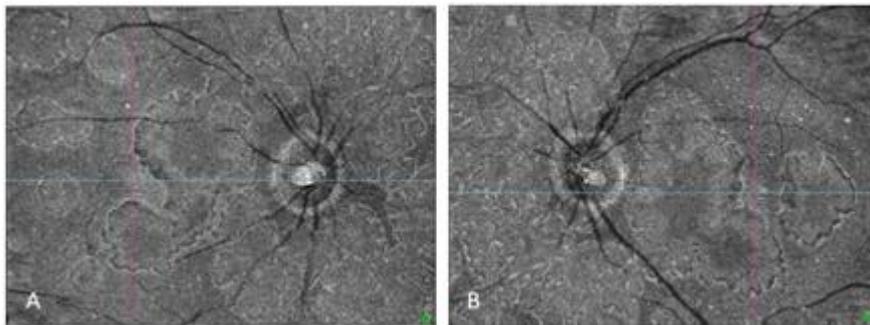


**Fig-5: Optical coherence tomographic images of the (A) right and (B) left eyes**

Note pseudocyst (white arrow) and outer retinal tabulation (yellow arrows)

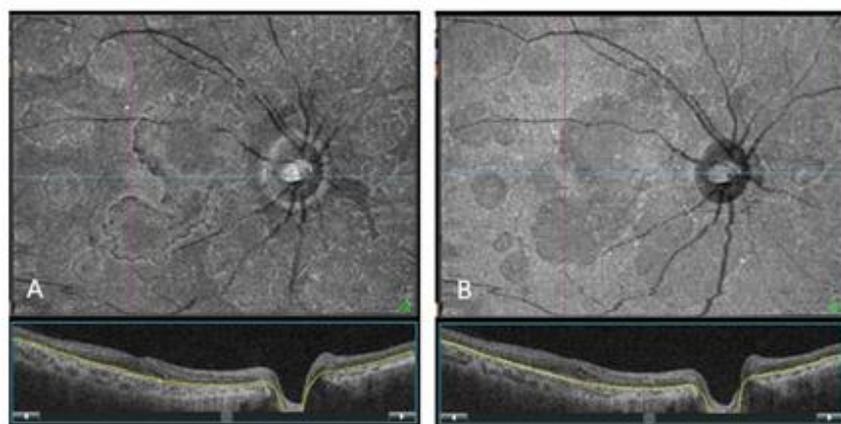
*En face* OCT images at the level of RPE showed hyperreflective crystals in both eyes (Figure 6). In addition, the crystals were detected more

prominently at the level of the EZ than at the level of the RPE (Figure 7).



**Fig-6: *En face* optical coherence tomographic images of the (A) right and (B) left eyes**

Note hyperreflective crystals at the level of the RPE



**Fig-7: *En face* optical coherence tomographic images at the level of the retinal pigment epithelium (A) and at the level of the ellipsoid zone (B) of the right eye**

## DISCUSSION

In BCD, deposition of crystals made of cholesterol is seen in corneal limbal region and in the retina. The crystals decrease in number with

progression of RPE atrophy, which is reflected by fewer crystals in the areas of RPE atrophy in this case. In general, the affected areas on OCT initially have loss of the outer retinal layers, which is followed by retinal and choroidal thinning, however, relatively preserved foveal

contour and retinal layers were detected in this case. Although reports have documented crystals as hyperreflective dots on conventional OCT scans, the crystals were better evident on *en face* OCT [5-7]. Furthermore, crystals at the level of the choroid have been reported [5], however, the crystals were not seen at the level of choroid in our case.

## CONCLUSIONS

Although our report is based solely on a patient with BCD, *en face* OCT findings may contribute to a better understanding of the location of the retinal crystals.

### Disclosure

The author declares no conflict of interest.

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