resembles caution. However, according to the previous postmortem analysis, the observations of this study strongly suggest that combined cSLO-SD-OCT imaging may detect structural changes within different retinal layers in vivo that were previously only identifiable by histopathology.

The striking observation of the spatial correlation of the ring of increased FAF with the transitional zone seen by SD-OCT may add to the understanding of FAF findings. It would be conceivable that the RPE cells in the transitional zone bear an increased metabolic burden. They may be unable to phagocytize the increased demand for material and compounds from severely impaired photoreceptors. This would lead to an increased accumulation of fluorophores and, subsequently, an increased FAF signal. When photoreceptor function is finally lost, the metabolic requirements for the corresponding RPE cells are reduced. The accumulated material may be partly degraded; thus, the FAF intensities would return to normal levels.

The observation of preserved FAF in retinal areas with impaired retinal sensitivity and absence of the IPRL would suggest that normal-appearing FAF intensities do not necessarily reflect an anatomically or functionally intact photoreceptor-RPE complex. The RPE might be present despite the absence of intact photoreceptors. It may be speculated that surviving RPE cells contain lipofuscin granules that are formed prior to the occurrence of outer retinal atrophy. Because it is thought that RPE cells have no means of exocytosis of such granules, a viable RPE would continue to elicit FAF phenomena. This would also indicate that constant phagocytosis of shed photoreceptor outer segment is not required for normal FAF intensities.

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Multimodal Fundus Imaging in Foveal Hypoplasia: Combined Scanning Laser Ophthalmoscope Imaging and Spectral-Domain Optical Coherence Tomography

Foveal hypoplasia is a rare disease that mostly occurs in association with other systemic or ocular diseases, such as albinism or aniridia.

Video available at www.archophthalmol.com

Diagnosis can be challenging, especially if foveal hypoplasia is an isolated finding. In recent years, optical coherence tomography (OCT) has been described as a useful tool to confirm the suspected diagnosis. However, the common asso-

Figure 3. Patient 2; left eye. Simultaneous fundus autofluorescence (FAF) and spectral domain–optical coherence tomography imaging. Arrows indicate transition zones with increased FAF. Sections *, †, and ‡ in panel B correspond to those in panel C. † In the broadened area with increased FAF (within black lines), the external limiting membrane (white lines indicate the presumed location) appears more distant from the retinal pigment epithelium layer.
cation of nystagmus with foveal hypoplasia may generate artifacts in OCT scans and may lead to off-center scans and therefore unreliable results.1 We examined 2 patients with foveal hypoplasia by using a simultaneous imaging device that combines a confocal scanning laser ophthalmoscope with a spectral domain OCT (Spectralis HRA+OCT; Heidelberg Engineering, Heidelberg, Germany). An integrated eye-tracking system ensures that the high-resolution OCT images are obtained at the exact retinal location that is selected on the stabilized fundus image. The principles of the Declaration of Helsinki were followed.

**Report of Cases.** Case 1. A 54-year-old woman had long-standing impaired visual acuity and mild photophobia. Best-corrected visual acuity was 20/32 OU. There was no nystagmus. A minor hyperopic astigmatism was present in her right eye (+1.0 diopter sphere/−0.75 cylinder × 30°) and a myopic astigmatism in her left eye (−0.5 diopter sphere/−0.75 cylinder × 125°). Color vision was normal on examination with Ishihara plates and panel D-15 testing, as was the anterior segment, aside from a faint posterior subcapsular cataract secondary to long-standing steroid therapy for Crohn disease. The iris was gray with small patches of pigmentation on the anterior stroma and did not transilluminate. The fundus had a normal pigmentation, the macula showed slightly mottled pigment irregularities, and there was no foveolar or macular reflex on funduscopy.

Case 2. An otherwise healthy 9-year-old boy was referred for unexplained bilateral low visual acuity that had been unchanged for years. Visual acuity was 20/63 OD and 20/50 OS, and a slow rotating nystagmus was present. The anterior segment of the eye revealed a posterior embryotoxon. The gray iris showed patches of pigmentation and was without transillumination. A deep anterior chamber, despite hyperopia of +6 diopters, was caused by a large radius of the cornea as assessed by corneal topography. Funduscopically, there was no foveolar reflex and no oval macular reflex. The peripheral fundus showed an
overall light pigmentation with visible large choroidal vessels.

Retinal vessels at the posterior pole extended close to the presumed foveal area in both patients and were tortuous in patient 2. Some small vessels crossed the horizontal meridian. Fundus autofluorescence imaging did not show the typical foveal darkening due to absorption of the excitation light by macular pigment (Figure, A). This finding was more pronounced in patient 2. Confoveal blue reflectance imaging revealed a concentric wrinkling of the inner retinal surface surrounding the posterior pole in patient 2 (not shown).

Three-dimensional OCT volume scans consisting of a narrow sequence of single horizontal scans detected neither a foveal pit nor a clivus or anticlivos at the central retina (Figure, B) (video of a volume scan through the central retina of the right eye in patient 1 is available at http://www.archophthalmol.com). Single scans through the presumed foveal area showed continuity of all neurosensory retinal layers (Figure, C). The retinal layer representing the photoreceptors was broader at the presumed foveal center compared with other retinal areas in patient 1.

Comment. In both cases, 3-dimensional imaging of the central retina confirmed the suspected diagnosis of foveal hypoplasia. Moreover, high-resolution OCT allowed a reliable diagnosis of subtle anatomic changes of the macula in spatial correlation with the confocal fundus image. The thickening of the photoreceptor layer found in patient 1 at the presumed foveal center is also present in healthy eyes. This thickening is thought to be due to the elongated cone photoreceptors at the central fovea and may be a sign of lesser anatomic alterations in a subset of patients with foveal hypoplasia, as it was not present in patient 2, who had worse visual function than patient 1.

Analyzing fluorescein angiographs in patients with foveal hypoplasia, Oliver and coworkers observed a similar fluorescence at the macular area compared with more peripheral parts of the fundus. They concluded that this phenomenon may be related to the amount of macular pigment present. This suggestion is supported by our finding that the usual foveal attenuation of fundus autofluorescence by macular pigment is reduced (patient 1) or almost absent (patient 2). Therefore, an intact foveal anatomy appears to be related to a physiological macular pigment storage in the neurosensory retina. Because this finding was more pronounced in patient 2, who had worse visual acuity, it may be speculated that macular pigment density correlates with the anatomical and functional integrity of the fovea in patients with foveal hypoplasia.

The integrated eye-tracking system that continuously compensates for eye movements during the examination allowed recordings in all imaging modes, even in patient 2, who had a slow rotating nystagmus. Therefore, even in the presence of a nystagmus, the system may be of diagnostic value in unexplained low visual acuity to rule out otherwise occult macular pathologies.

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