sue in concentric layers, and (5) external loose connective tissue with fibroadipose elements, muscle, and blood vessels. Our patient had each of these elements.

Conservative treatment includes observation with aspiration as needed. In recalcitrant cases not amenable to aspiration or if considerable facial dysmorphism exists, excision is conventionally undertaken.

In summary, congenital cystic eye is exceedingly rare. Diagnosis historically was based on physical and histopathological findings. However, newer imaging modalities are revealing characteristic findings of the condition at or even prior to birth, as in our case. Given the frequent association with intracranial abnormalities, including the possibility of septo-optic dysplasia, neuroimaging is warranted to screen for such aberrations. Finally, although not always required, treatment conventionally involves excision.

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Development of a Premacular Vitreous Pocket

The premacular vitreous pocket (PVP), or vitreoschisis cavity, is a liquefied vitreous cavity in front of the posterior retina that is characteristic of various macular diseases, including macular holes and diabetic maculopathy.1 The reason for the development of PVPs is unknown because of the difficulty observing the formed vitreous in vivo. India ink and the fluorescein staining technique have delineated the structure of the PVP in the vitreous cavity in human eyes at autopsy; however, the technique is limited because of the presence of artifacts during fixation of the fragile and mobile vitreous and postmortem changes. Optical coherence tomography has facilitated observation of the vitreous structures in vivo. Herein, we describe the development and fine details of PVPs in real time.

Methods | We retrospectively analyzed the posterior vitreous, retinas, and optic discs of 56 healthy eyes (39 patients; age range, 1-54 years) using swept-source optical coherence tomography (Topcon), which provides detailed images of the fine ocular structures. The scanning protocol used in this study was a single-line scan with 96 overlapping images and a radial scan with 32 overlapping images. Each line has a 12-mm transverse scanning length with 1024-pixel resolution. Eyes that appeared healthy were excluded if the patient had a family history of a hereditary vitreoretinal disease.

Results | A PVP (Figure 1C-F) was detected in all eyes of patients older than 10 years and in no eyes of patients younger than 2 years (Figure 1A). A crack in the formed vitreous (Figure 1B), considered to be a primitive structure of the PVP, developed first in eyes around age 2 years. Between ages 3 and
9 years, a PVP was present in 16 eyes (49%) and a crack in 20 eyes (61%). Nine eyes (56%) with a PVP also had a crack. Among eyes with both a PVP and a crack, 16 eyes (86%) had cracks connected to the PVP. Twenty-eight eyes (80%) with PVPs had a liquefied connection between the PVP and the Cloquet canal (Figure 1C-F). The connection to the Cloquet canal was identified in both the PVP and the crack (Figure 1B). In younger eyes, the PVP was wider horizontally than vertically, and all detectable cracks were wider horizontally than vertically. During the early phase of PVP development, several eyes had multifocal PVPs and cracks (Figure 2A-C) in the premacular vitreous. A high-density structure, which appeared to be a remnant of regressed hyaloid vessels and was connected to the Bergmeister papilla, was present temporally along the crack and wall of the PVP in several eyes (Figure 2D).

Discussion | Kishi and Shimizu originally identified PVPs in eyes at autopsy and implied that development began with slight separation of the vitreous at about age 2 years, although the PVPs might include postmortem changes. The current in vivo study showed that a PVP is often absent at birth and is often present by about age 3 years. Interestingly, the crack in the formed vitreous also was observed as an initial change around age 2 years.

The PVP and posterior Cloquet canal, which are separated by a dishlike wall of vitreous, were connected in most eyes of the current patients, even in eyes with a crack at an initial stage. Aqueous humor from the posterior Cloquet canal may play a role in formation of the crack and PVP.

Almost all primary PVPs and cracks that occasionally developed multifocally and coexisted with remnants of hyaloid vessels were wider horizontally than vertically. Because ocular movement is usually dominant horizontally, horizontal shear stress might generate cracks in horizontally layered premacular vitreous, in which remnants of hyaloid vessels may be related to the friability of the premacular vitreous. The vitreous and hyaloid vessels are symmetric along the anteroposterior axis during early development and become asymmetric after dominant growth of the temporoposterior region. Since the remnant, cracks, and PVPs were observed only temporally in the premacular vitreous, the asymmetric vitreous growth may contribute to the asymmetric location of these structures. Further study is needed to confirm our preliminary findings.

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