

Figure 2. Spectral-domain optical coherence tomographic images of the right eye at the initial visit (A), 2 weeks after the initial visit (B), and 7 months after the initial visit (C). T indicates temporal; N, nasal; S, superior; and I, inferior.

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Peripheral Retinal Nonperfusion Associated With Optic Nerve Hypoplasia and Lissencephaly

Few entities exist in which optic nerve hypoplasia (ONH) is found in association with peripheral retinal nonperfusion. Among these are the congenital muscular dystrophies with abnormal glycosylation of α -dystroglycan, consisting of Walker-Warburg syndrome, muscle-eye-brain disease, and Fukuyama congenital muscular dystrophy, characterized by defective brain migration and ocular abnormalities. Posterior segment findings in these disorders have included ONH as well as retinal dysplasia.¹ Herein, we describe a full-term girl not only with ONH and bilateral peripheral retinal nonperfusion with resultant tractional retinal detachments but also with severe brain abnormalities including lissencephaly and hydrocephalus.

Report of a Case. A 3785-g girl born at 39 weeks' gestation was referred for bilateral retinal detachments. The prenatal course was uneventful, and there was no family history of eye or neurological abnormalities.

Shortly after birth, she developed seizures. Magnetic resonance imaging revealed massive dilatation of the lateral ventricles secondary to atrophy and hydrocephalus,

marked cortical dysplasia and lissencephaly, periventricular calcifications, and a thin corpus callosum (**Figure 1**). Although the calcifications were suspicious for cytomegalovirus infection, serologic and urine culture results were negative. The serum creatine kinase level, to evaluate for a muscular dystrophy, was normal.

Examination revealed normal anterior segments. Fundus photography of the right eye showed a large tractional retinal detachment involving the macula, obscuring both the macula and the optic nerve, with posterior retinal vessels dragged and distorted into a retinal fold (**Figure 2A**). Fluorescein angiography demonstrated massive leakage off the stalk and along the apex of the horseshoe-shaped retinal detachment (Figure 2B). The optic nerve of the left eye showed a double ring sign consistent with ONH as well as foveal hypoplasia (Figure 2C). The retinal vessels terminated posteriorly, especially temporally with extraretinal fibrovascular proliferation extending into the vitreous and tractional detachment inferonasally and superotemporally (Figure 2D and E).

Laser photocoagulation was applied to the avascular retinal zones. The patient subsequently underwent vitrectomy in both eyes. Three months later, the left retina was attached completely (Figure 2F) but the right retina remained detached.

Comment. There is a spectrum of disorders with ocular and neurological manifestations that overlap those of our patient. In the congenital muscular dystrophies, an underlying defect in glycosylation is thought to result in severe defects in neuronal migration, thus causing hypoplasia of various brain and eye structures.² Walker-Warburg syndrome is the most severe, with brain abnormalities including lissencephaly, hydrocephalus, cerebellar malformation, hypomyelination of the white matter, and agenesis of the corpus callosum.³ Ocular posterior segment abnormalities include retinal dysplasia as well as hypoplasia or atrophy of the optic nerve and macula. Lissencephaly can also be found in Fukuyama congenital

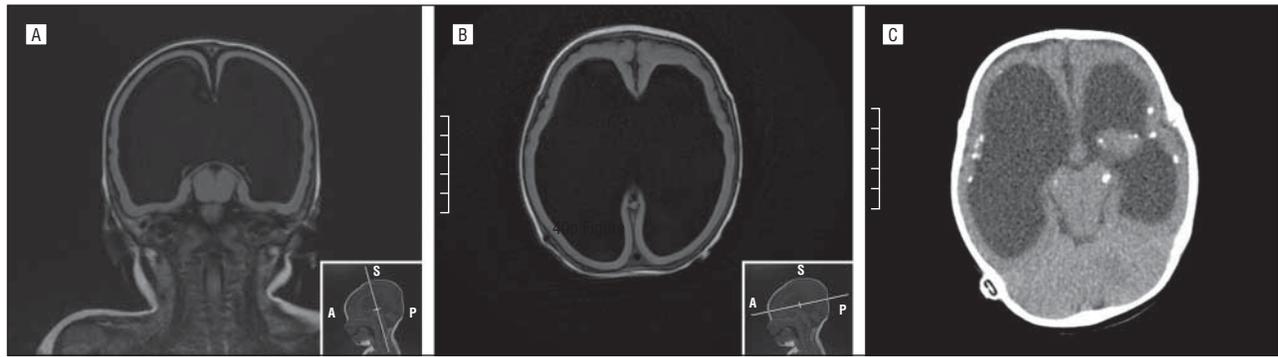


Figure 1. Coronal (A) and axial (B) T1-weighted magnetic resonance images demonstrate large dilatation of the lateral ventricles. The brain was not small but appeared smooth without gyri and sulci. The primary sylvian fissures were not seen, suggestive of developmental arrest (cortical dysplasia). A indicates anterior; P, posterior; and S, superior. C, Axial computed tomography shows dilated lateral ventricles along with paraventricular calcifications. There were small calcifications adjacent to the perimesencephalic cistern as well.

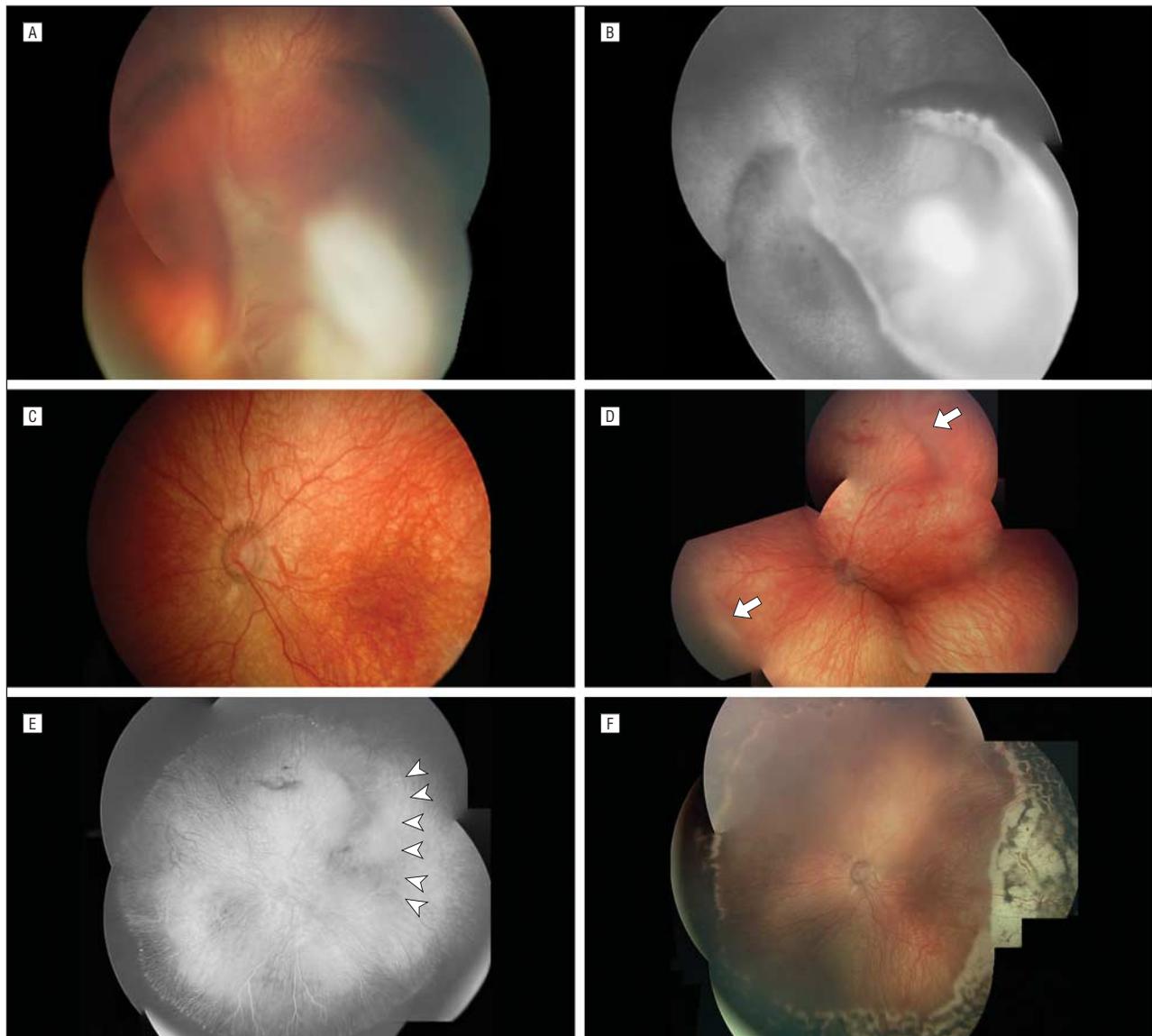


Figure 2. A montage Retcam color photograph (A) and a fluorescein angiogram (B) of the right eye show a large tractional retinal detachment involving the macula, obscuring both the macula and the optic nerve, with posterior retinal vessels dragged and distorted into a retinal fold. There was massive leakage off the stalk and along the apex of the horseshoe-shaped retinal detachment. C, A Retcam photograph of the left eye shows a double ring sign consistent with optic nerve hypoplasia. It also demonstrates an undifferentiated macular zone and foveal hypoplasia. A montage color photograph (D) and a fluorescein angiogram (E) of the left eye show the retinal vessels terminated posteriorly, especially temporally (arrowheads) (E) with extraretinal vascularization extending into the vitreous and tractional detachment inferonasally and superotemporally (arrows) (D). F, A montage color photograph of the left eye 3 months after laser treatment and pars plana vitrectomy shows a completely attached retina, peripheral laser scars, and an undifferentiated fovea.

muscular dystrophy and muscle-eye-brain disease with associated ONH and retinal hypoplasia.⁴ A case of genetically proven muscle-eye-brain disease with ONH and peripheral retinal nonperfusion with secondary fibrovascular proliferation and retinal detachment was recently described.² Our patient shares many similar features but had no evidence of a muscular dystrophy, with clinically absent hypotonia and a normal creatine kinase level. Additionally, a case of de Morsier syndrome with similar ocular findings of bilateral peripheral retinal nonperfusion and neovascularization with resultant retinal detachment was reported recently.⁵ We suggest that the abnormal neuronal development that led to lissencephaly resulted in optic nerve and retinal maldevelopment and subsequent associated retinal vascular maldevelopment. Tractional retinal detachment is an end stage of a process that starts with nonperfusion and progresses to ischemia and extraretinal fibrovascular proliferation in a variety of pediatric retinal diseases.

Although the cause of neural maldevelopment in our patient remains elusive, her case supports the idea that patients with severe neuronal migration deficits should be evaluated for ONH and abnormal retinal vasculature development, even without evidence of a muscular dystrophy.

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Author Contributions: Dr Shapiro had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

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Subclinical Photoreceptor Disruption in Response to Severe Head Trauma

Comotio retinae is a transient opacification of the retina due to outer retinal disruption occurring in a contrecoup fashion after blunt trauma.^{1,2} Histological studies in animals and humans after ocular blunt trauma have revealed that disruption occurs at the level of the photoreceptor outer segments and retinal pigment epithelium.^{2,3} Recent reports using optical coherence tomography (OCT) have shown detectable disruption at the level of the photoreceptor inner segment/outer segment junction and retinal pigment epithelium⁴⁻⁶ and that these changes may be reversible over time with restoration of normal outer retinal architecture.⁵ However, the resolution of existing OCT technology may not be sensitive enough to detect photoreceptor disruption. Adaptive optics (AO) imaging systems enable cellular-resolution imaging of the human retina, and there is a growing number of cases where deficits have been visible on AO images but not on OCT. Herein, we report a case of subclinical photoreceptor disruption after head trauma as seen by an AO scanning ophthalmoscope (AOSO) but not apparent clinically or on spectral-domain OCT (SD-OCT).

Report of a Case. A 43-year-old man described a 5-year history of a stable, crescent-shaped purple scotoma nasal to central fixation in his right eye that developed after an industrial accident in which he sustained significant head and body trauma. A complete ophthalmic examination revealed best-corrected visual acuity of 20/20 OU and no remarkable fundus findings or abnormalities. Fluorescein angiography and SD-OCT (Spectralis SD-OCT; Heidelberg Engineering) findings were unremarkable (**Figure 1**). Humphrey visual field 10-2 testing and microperimetry revealed a small nonspecific area of functional vision loss near fixation in the right eye. Images of the photoreceptor mosaic near the fovea were acquired using a newly developed AOSO. Images were processed and registered using custom MatLab software (MathWorks). While foveal cone density was normal, the AOSO images revealed a well-defined crescent-shaped area of photoreceptor disruption just temporal to the fovea (**Figure 2A**, large arrows). Other focal areas of photoreceptor irregularities were also seen superior, temporal, and inferotemporal to the fovea (**Figure 2A**, small arrows). Both cone and rod photoreceptors were visualized with this AOSO imaging, and both cell types appeared to be disrupted (**Figure 2A and B**).

Comment. The AOSO detected photoreceptor disruption resulting from head trauma and not apparent clinically or by other standard imaging modalities, including SD-OCT. Restoration of the outer retinal appearance in SD-OCT has been reported after comotio retinae,⁵ suggesting recovery of the outer retinal structure. Our data demonstrate that photoreceptor disruption may still exist. The SD-OCT axial resolution is likely not sensitive enough to reveal the full extent of photoreceptor disruption that may occur after ocular or head trauma. The