HAND-HELD SPECTRAL DOMAIN OPTICAL COHERENCE TOMOGRAPHY FINDING IN SHAKEN-BABY SYNDROME

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Purpose: The purpose of this study was to document the hand-held spectral domain optical coherence tomography (HHSD-OCT, Bioptigen, Durham, NC) findings in shaken-baby syndrome (SBS). The nonaccidental trauma in SBS has been associated with retinal findings, including hemorrhages in all layers of the retina and retinoschisis.

Methods: Three consecutive patients with presumed SBS underwent complete ocular examination, fundus photography with the RetCam (Clarity Medical Systems, Pleasanton, CA), and imaging with the HHSD-OCT. Acquisition of the HHSD-OCT images required an assistant to stabilize the head of the infant.

Results: All three patients had clinical findings consistent with SBS, including preretinal and intraretinal hemorrhages. Hand-held spectral domain optical coherence tomography documented focal posterior vitreous separation in four of the five eyes with multilayered retinoschisis in one eye, disruption of the foveal architecture and foveolar detachment in one eye, and disinsertion of the internal limiting membrane or inner retinoschisis in one eye. Hand-held spectral domain optical coherence tomography documented preretinal hemorrhages in all five eyes.

Conclusion: Hand-held spectral domain optical coherence tomography is helpful in the evaluation of patients with SBS. All patients in our series had vitreoretinal abnormalities not detected on clinical examination, including, for example, multilayered retinoschisis. Hand-held spectral domain optical coherence tomography allows high-resolution imaging of the vitreoretinal interface and retina in infants with SBS and has provided insight into the mechanism of various retinal findings.

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The nonaccidental trauma in shaken-baby syndrome (SBS) has been associated with multiple retinal findings, including retinal detachments,[1] hemorrhages in all three layers of the retina,[2,3] chorioretinal atrophy,[4] perimacular folds,[5] traumatic retinoschisis,[6] macular holes,[7,8] and epiretinal membranes.[9] Despite these well-documented retinal findings, the mechanism(s) by which retinal hemorrhages occur continues to be actively debated in the literature. Several theories have been postulated, including raised intracranial pressure, raised intrathoracic pressure, direct head trauma, and the direct effect of mechanical shaking on the globe and orbit.[10–17] Nonetheless, growing evidence suggests that multiple focal disruptions between firm vitreoretinal attachments because of the repetitive mechanical effect of shaking itself is the basis for the retinal findings in SBS.[18]

To date, the RetCam (Clarity Medical Systems, Pleasanton, CA) has been extremely helpful in the documentation of the retinal findings in patients with SBS. However, until recently, we have been unable to image the vitreoretinal interface and retinal

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microarchitecture in SBS. The hand-held spectral domain optical coherence tomography (HHSD-OCT) (Bioptigen, Durham, NC) has enabled us to obtain high-resolution in vivo images of the vitreoretinal microarchitecture to a resolution of 4.5 μm in the supine infant. This technology has allowed us to characterize the vitreoretinal changes in SBS that are likely responsible for retinal hemorrhages and the retinoschisis. The purpose of this study is to document the HHSD-OCT findings in patients with SBS.

Methods

Three consecutive infants with suspected nonaccidental trauma underwent a complete ocular examination, including slit-lamp examination, indirect ophthalmoscopy, RetCam fundus photography, and examination with the HHSD-OCT within 48 hours to 72 hours after admission. During the HHSD-OCT imaging, an assistant was required to hold the infant's head steady while the images were acquired. The focal point of the imaging handpiece was adjusted to account for the refractive error and the reference arm was manually adjusted to account for the shortened axial length in infant eyes. Reference arm adjustments were made during image acquisition to obtain the highest quality images possible in each case. Hand-held spectral domain optical coherence tomography images were acquired using a 10 × 10-mm retinal scan, which produced 100 B-scans (1,000 A-scans per B-scan). All data were collected retrospectively and reported in accordance with the Health Insurance Portability and Accountability Act.

Results

Case 1

A 4½-month-old boy presented to the emergency department with a decreased level of consciousness. Intubation was immediately performed. Computed tomography scan of the head showed frontal subdural hemorrhage and left subarachnoid hemorrhage. An electroencephalogram showed moderate diffuse background attenuation and lack of cerebral reactivity to painful stimulus, consistent with moderate diffuse cerebral dysfunction. Seizures occurred during his hospital admission. Ocular examination showed bilateral preretinal and intraretinal hemorrhages involving the posterior pole (Figure 1A). Hand-held spectral domain optical coherence tomography of the right eye showed focal posterior vitreous separation with associated multilayered retinoschisis, disruption of the retinal architecture, and preretinal hemorrhage (**see Video, Supplemental Digital Content 1,** http://links.lww.com/IAE/A15 Figure 1, B–E). Hand-held spectral domain optical coherence tomography of the left eye also showed multiple areas of focal vitreous separation in the macula with associated retinal traction and preretinal hemorrhages (Figure 1, F and G).

Case 2

A 2-year-old boy presented to the emergency room with suspected SBS and was intubated on arrival. Computed tomography scan showed a right-sided frontal hematoma, a midline frontal hematoma of older age, a right occipital hematoma, and a left subdural hematoma with midline shift to the right. Despite being on antiepileptic medication to decrease seizure risk, multiple seizures occurred while the patient was in the hospital, and the electroencephalogram was consistent with left frontoparietal slowing and focal attenuation. Ocular examination showed a fixed and dilated right pupil with a reactive left pupil. Fundus examination showed bilateral preretinal and intraretinal hemorrhages involving the posterior pole (Figure 2, A and B). Hand-held spectral domain optical coherence tomography was only performed on the left eye to minimize examination time for this child. Hand-held spectral domain optical coherence tomography showed disruption of the foveal architecture and foveolar detachment (Figure 2, C and D).

Case 3

A 13-month-old boy presented to the emergency room with a left-sided tonic–clonic seizure, bradycardia, hypoxia, and an unstable airway, which led to intubation in the emergency room. Computed tomography and magnetic resonance imaging confirmed infarction in the left cerebral hemisphere and a left subdural hematoma. Skeletal survey showed a healing distal right radial fracture. Ocular examination showed multiple macular preretinal and intraretinal hemorrhages (Figure 3, A and B) and peripheral intraretinal hemorrhages. Hand-held spectral domain optical coherence tomography of the right eye showed small preretinal hemorrhages with associated outer retinal disruption (Figure 3C). Hand-held spectral domain optical coherence tomography of the left eye showed focal vitreous separation with associated preretinal hemorrhage and likely superficial inner retinoschisis (vs. internal limiting membrane disinsertion) (Figure 3, D–F). Ocular examination 6 weeks later showed bilateral resolution of the retinal hemorrhages.

In summary, HHSD-OCT documented focal posterior vitreous separation in four of the five eyes with multilayered retinoschisis in one eye, disruption of the foveal architecture and foveolar detachment in one eye, and disinsertion of the internal limiting membrane or inner retinoschisis in one eye. Hand-held spectral domain optical coherence tomography documented preretinal hemorrhages in all five eyes.

Discussion

We present a consecutive case series of three infants with clinical features consistent with SBS and HHSD-OCT evidence of vitreoretinal interface pathology. This includes focal posterior vitreous separation, multilayered tractional retinoschisis, disinsertion of the internal limiting membrane (or inner retinoschisis), and preretinal hemorrhages. These findings support the theory that traction at the vitreoretinal interface caused by repetitive shaking plays a major role in the mechanism of retinal hemorrhages and retinoschisis in SBS.

Scott et al[19] were the first to document HHSD-OCT findings in infants. This article describes the use of HHSD-OCT in two children with SBS. The children

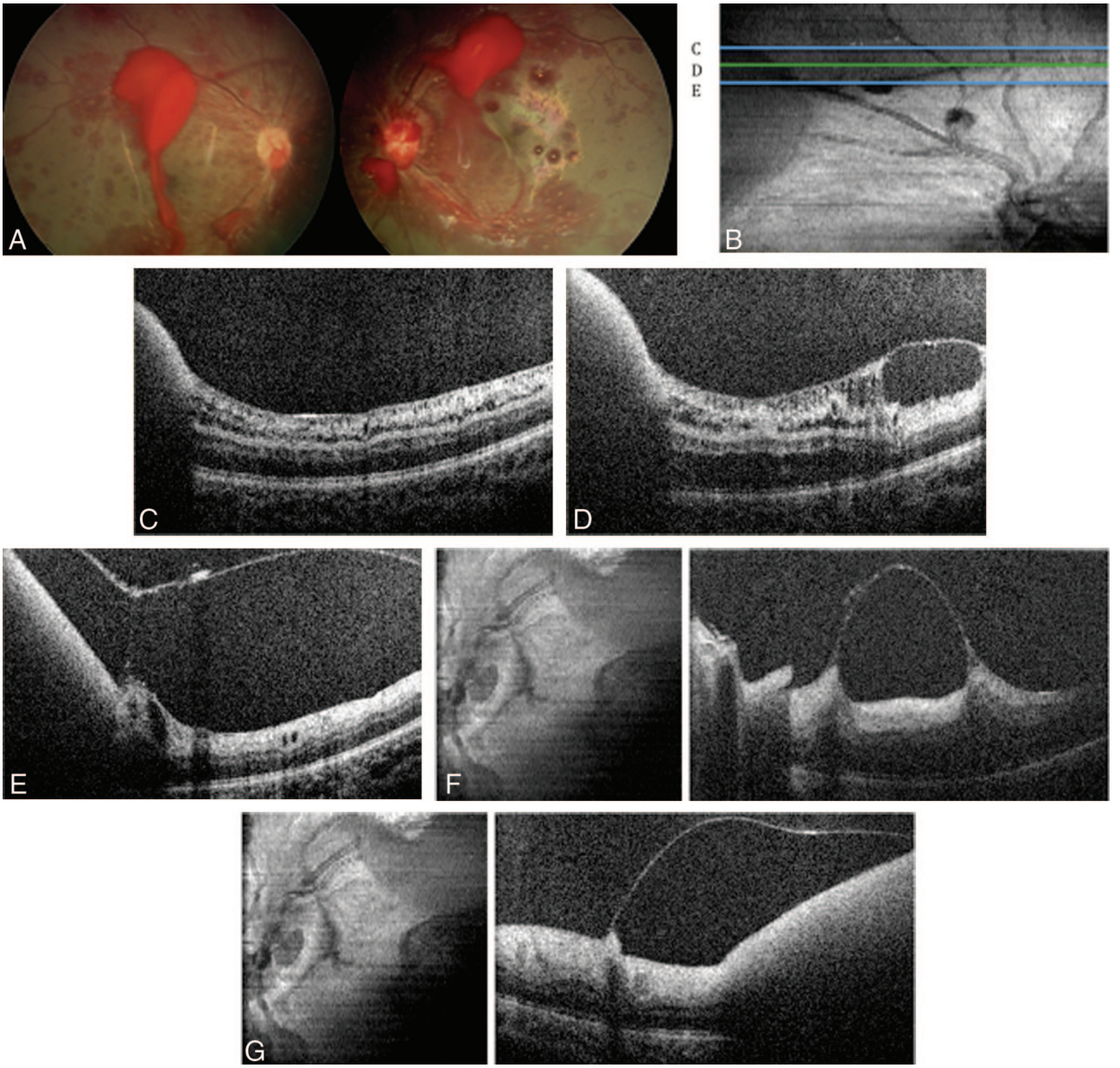

Fig. 1. A. Bilateral subhyaloid and intraretinal hemorrhages involving the macula. **B.** Composite image of the right fundus indicating the location of the cuts for Figures **C**–**E**. **C.** Disruption of the retinal architecture and lamellar retinoschisis is evident in a location where the hyaloid is attached just anterior to the focal vitreous separation. **D.** Multilayered retinoschisis at the exact location of the posterior vitreous separation. **E.** Preretinal hemorrhage evident within the area of separated vitreous. **F.** Hand-held spectral domain optical coherence tomography of the left macula showing focal posterior vitreous separation with retinal traction. **G.** Hand-held spectral domain optical coherence tomography of the left macula shows focal vitreous separation with subhyaloid hemorrhage.

in their study were imaged 1 month to 8 months after the suspected trauma. One of their patients had a macular hole with perimacular folds in the right eye and a lamellar hole with partial separation of the posterior hyaloid in the left eye. Their other patient had a double-layered epiretinal membrane. The authors concluded that the HHSD-OCT is a useful tool in aiding in the differentiation of retinal disease processes and that it should be used as an adjunct to RetCam photography in the assessment of patients with SBS.

Sturm et al[20] recently published a case series of 3 eyes in 3 infants with SBS who were imaged with the Zeiss Stratus OCT3 (Carl Zeiss Meditech, Dublin, CA) in the pediatric intensive care unit while intubated. Infants were placed in an anti-Trendelenburg position in front of an OCT machine being held by a second person. Optical coherence tomography identi-

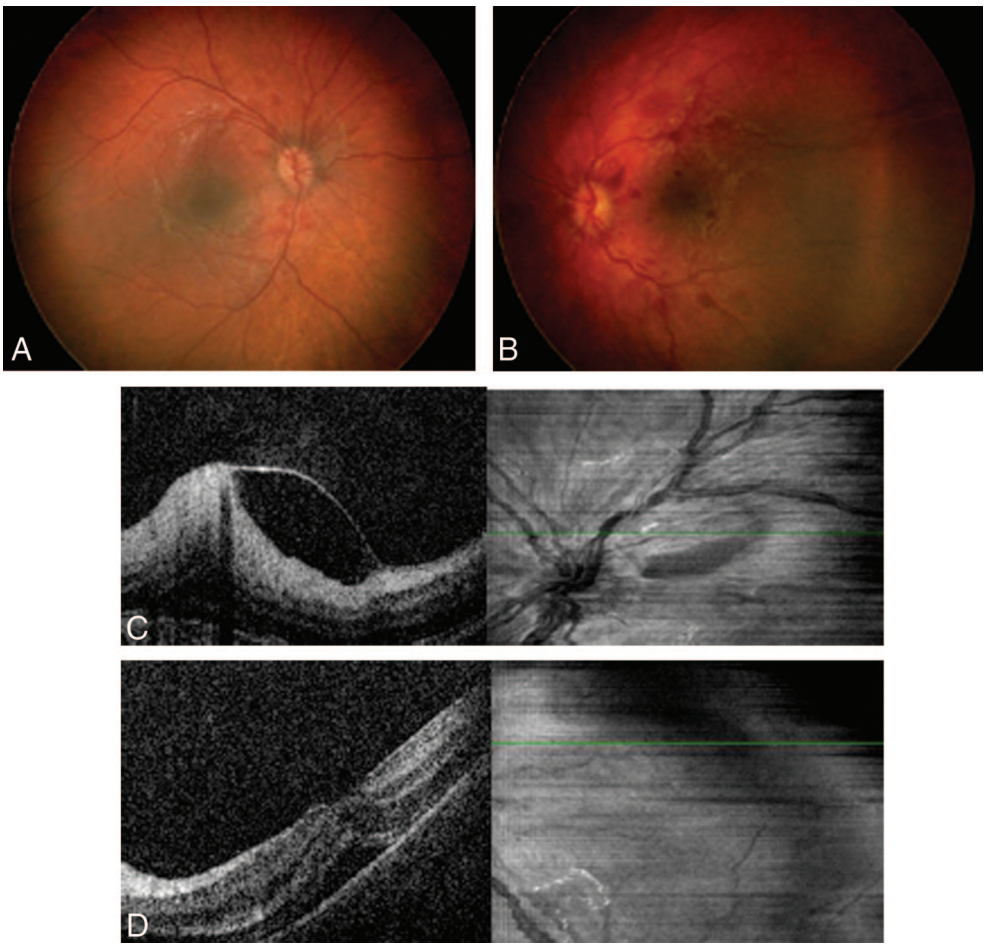

Fig. 2. **A** and **B.** RetCam photographs of the right and left eyes showing intraretinal hemorrhages on the right and preretinal and intraretinal hemorrhages involving the macula on the left. **C.** Focal vitreous separation with associated preretinal hemorrhage in the left eye. **D.** Disruption of the foveal architecture and foveolar detachment in the left eye.

fied perimacular vitreoretinal traction in all three eyes with possible macular retinoschisis in one case and perimacular folds in another. On the basis of their OCT findings, Sturm et al postulated that it is the shaking itself that induces shearing forces at the vitreoretinal interface as a primary factor in the generation of retinal hemorrhages and the possible retinoschisis. In his editorial of the article by Sturm et al, Forbes[21] also supported this theory.

On the basis of our HHSD-OCT findings, we agree with Sturm et al and the editorial by Forbes that the retinal hemorrhages and retinoschisis seen in SBS are primarily caused by the shaking per se, which induces shearing forces at the vitreoretinal interface. In our first case, we document significant disruption of the retinal microarchitecture with multilayered tractional retinoschisis intricately associated with focal posterior vitreous separation. This is informative because we are able to image the retinal morphology anterior to, at, and posterior to the exact location of the vitreous separation. Anteriorly, there is multilayered lamellar retinoschisis in an area of attached hyaloid. At the point of vitreous separation, there is significant disorganization of the retina with worsening retinoschisis, and posterior to the separation, we see the large preretinal hemorrhage. These findings together with the disruption of the foveal architecture and foveolar detachment in Case 2 and inner retinoschisis or disinsertion of the internal limiting membrane in Case 3 support the notion that vitreoretinal traction is playing a major role in the retinal findings of SBS.

Although clinical examination and RetCam fundus photography have been essential in our ability to diagnose and document cases of SBS, it has been inadequate in the examination of the vitreoretinal interface. This retrospective consecutive case series of high-resolution HHSD-OCT findings in three patients with SBS support the prevailing theory that vitreous traction is involved in the mechanism of retinal hemorrhages and retinoschisis in SBS. We agree with Scott et al that HHSD-OCT should be considered as a useful adjunct to RetCam fundus photography in the diagnosis of retinal pathology associated with SBS, especially in cases in which the diagnosis is less clear.

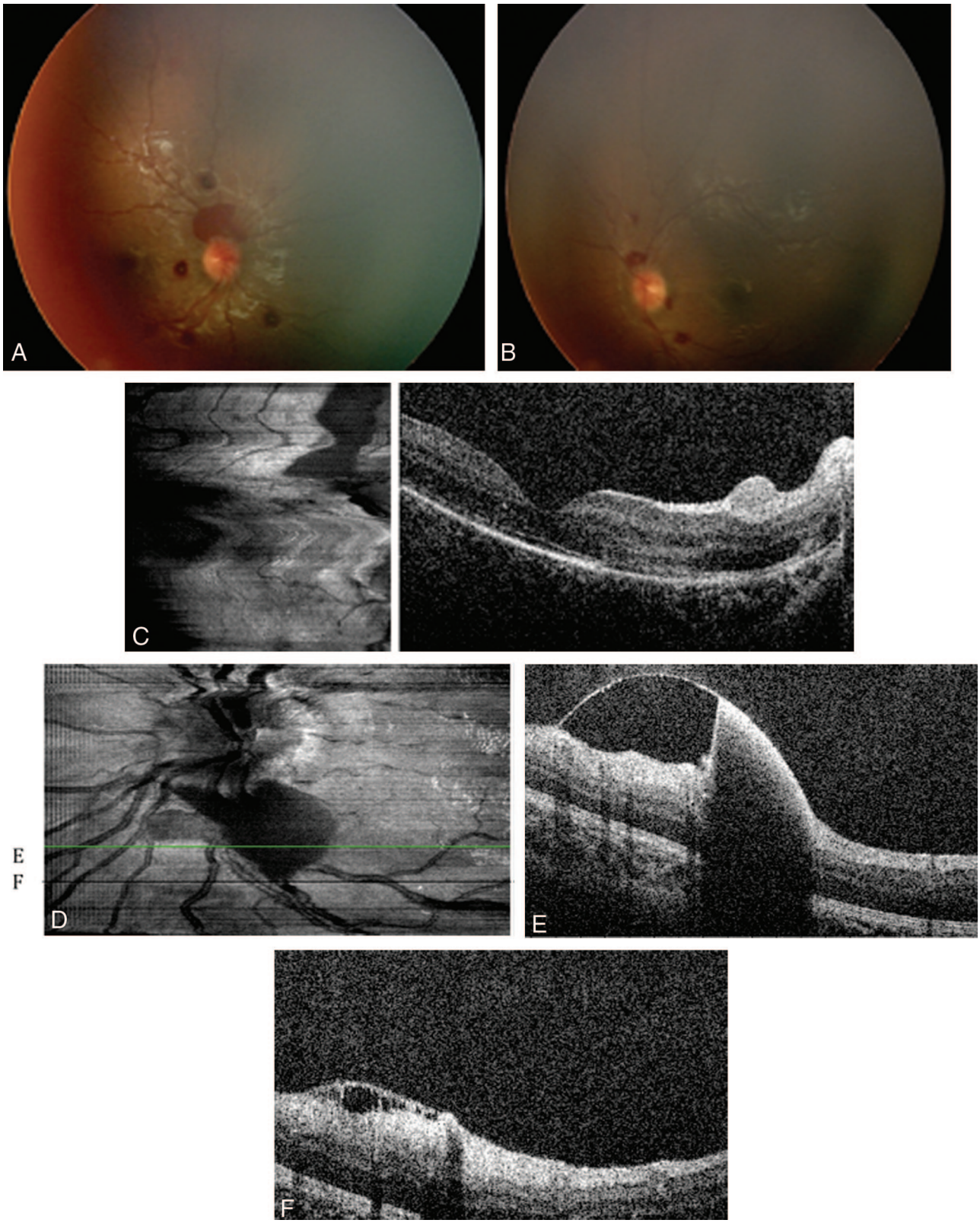

Fig. 3. A and **B.** RetCam photographs of right and left eyes showing preretinal and intraretinal hemorrhages involving the posterior pole. **C.** Hand-held spectral domain optical coherence tomography of the right eye showing small preretinal hemorrhages with associated outer retinal disruption. **D.** Composite image of the left fundus indicating the location of the cuts for Figures **E** and **F**. **E.** Focal vitreous separation with associated preretinal hemorrhage and (**F**) superficial inner retinoschisis (versus internal limiting membrane disinsertion) at the edge of the focal vitreous separation.

Conclusion

The HHSD-OCT allows high-resolution imaging of the vitreoretinal interface and retina in infants with SBS and has provided insight into the mechanism of retinal hemorrhages and retinoschisis in infants with this condition.

Key words: handheld SD-OCT, OCT, retinal hemorrhages, retinoschisis, shaken-baby syndrome, focal vitreous separation.

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