dense deposits containing interspersed collagen fibrils, extracellular empty spaces (likely clefts formed from lipid removal during processing), and persistently activated keratocytes are similar to these animal findings. The extracellular lipid collections, which likely correspond to the crystalline deposits seen clinically, probably arise from chronic mechanical irritation to keratocytes that continually strive to heal the stromal wound.

The breaks in the Bowman layer could correspond to predisposition to keratoconus and ectasia. Although the reinforced stromal bed was not strong enough to prevent further ectasia from developing in this case, longitudinal studies have not yet addressed the overall long-term success or stability of ICR segments for corneal ectasia beyond 1 year postoperatively. These studies are necessary to address whether the natural history of the ectasia changes after ICR segment implantation.

Marc J. Spirn, MD  
Daniel G. Dawson, MD  
Roy S. Rubinfeld, MD  
Christine Burris, OD  
Jonathan Talamo, MD  
Henry F. Edelhauser, PhD  
Hans E. Grossniklaus, MD

Correspondence: Dr Grossniklaus, BT428 Emory Eye Center, 1365 Clifton Rd NE, Atlanta, GA 30322 (ophtheg@emory.edu).

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**Schisis in Sickle Cell Retinopathy**

Retinal schisis is a rare but potentially serious complication of sickle cell retinopathy. It is related to chronic low-grade ischemia of the inner nuclear layer, which houses the Muellerian glia, the structural backbone of the retina. Schisis as part of proliferative sickle cell retinopathy is characterized clinically by a concave tractional retinal elevation, retinal nonperfusion, inner-layer breaks, absorption of laser by the outer layer, and a split pattern on optical coherence tomography. Two cases of retinal schisis are described herein, both featuring the conjunctival sickle sign and both eventually complicated by outer-layer breaks and retinal detachment that possibly might have been prevented by timely laser treatment.

**Report of Cases. Case 1.** A 44-year-old African American woman complained that in recent months the visual acuity in her left eye had been decreasing as a result of what she described as “a moving veil.” She had been diagnosed with sickle cell disease, type SS, 17 years earlier. She had a history of multiple pulmonary emboli complicated by pulmonary hypertension, which had necessitated placement of an aortic umbrella 2 years earlier. She took warfarin sodium and folic acid. Her visual acu-

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**Figure 1.** Funduscopy, angiography, and optical coherence tomography results in patient 1. A. Drawing of the fundus showing the extent of schisis (hatched lines). B. Ocular coherence tomogram of the arteriovenous phase focused on the fovea (E) and vessels of retinal elevation (F).
ity with glasses was 20/30 OD and 20/120 OS; near vision was 20/65 OD and 20/200 OS. Slitlamp examination showed the sickle comma sign in conjunctival arterioles. Intraocular pressure was normal.

Funduscopy of the right eye disclosed tractional schisis from the 9- to 7-o'clock meridian within the inferotemporal arcades. Small inner-layer breaks were noted posterior to equatorial fibrovascular proliferation. The peripheral retina appeared nonperfused; the macula had a glial sheen. Funduscopy of the left eye showed equatorial fibrovascular proliferations. The temporal retina, including the fovea, was elevated from 12- to 6-o'clock (Figure 1A). The inferotemporal fovea was thrown into striae related to a point of vitreous adhesion just inside the arcades. An ovoid macular vitreous condensation was present (Figure 1C and D).

Visual field testing showed dense scotomas corresponding to the areas of nonperfusion and schisis. Angiography confirmed peripheral retinal nonperfusion beyond the near periphery; all areas of schisis received little or no perfusion (Figure 1E and F). Optical coherence tomography of the right macula showed schisis in the inner layer and, in the left macula, schisis and Muellerian pillars toward the temporal periphery (Figure 1B).

After photocoagulation of the nonperfused attached retina, laser burns were noted in the areas of retinal elevation, corroborating schisis. Four years of follow-up showed the area of schisis to be stable. Vision was 20/50 OD and 20/120 OS. A rhegmatogenous retinal detachment occurred in the left eye and was repaired, improving acuity to 20/50. After 5 years, visual acuity in the eye operated on was 20/80 following cataract extraction and repeat capsulotomy.

Case 2. A 56-year-old African American man with sickle cell trait had had repeat vitreous hemorrhages in his right eye. Years earlier, a closed-funnel retinal detachment had caused blindness in his left eye. Visual acuity was 20/40 OD; near vision was J2 at 14 in. There was light perception in the left eye. The previous winter, the patient had had pneumonia. He had no other medical problems and was taking no medications.

Slitlamp examination of the bulbar conjunctiva showed the comma sign of sickling. Indirect ophthalmoscopy of the right eye showed the disc and macula through vitreous hemorrhage. Large fibrovascular proliferations were noted from 11- to 1-o'clock and from 5- to 8-o'clock.
Just anterior to them, the retina was concave and elevated and had large inner-layer breaks (Figure 2A).

The patient was told that he had retinal schisis and was at risk of retinal detachment but that his hemorrhage was likely to clear. He was lost to follow-up for 5 months and then was seen with 5/200 OD vision. Ophthalmoscopy showed a mild vitreous hemorrhage and a retinal elevation that was shallow superiorly and more prominent inferiorly and temporally. Peripheral nonperfusion was demonstrated angiographically (Figure 2B and C). The retina was elevated from the disc to the ora serrata. Its configuration was convex, indicating rhegmatogenous detachment. The detachment was repaired without a scleral buckle using lensectomy, vitrectomy, endolaser, and, because the patient was one eyed, silicone oil. More laser burns were applied postoperatively to a total number of 3500. Six months after surgery, visual acuity had improved to 20/150 OD. Cystoid foveal edema was noted angiographically. The oil was subsequently removed, improving visual acuity to 20/25 OD and decreasing angiographic foveal edema. In the 6 o’clock meridian, almost equatorially, the causative large break in the outer layer of the retinal schisis cavity was readily apparent (Figure 2D). Vision remained stable after 5 years’ follow-up.

Comment. In a series of 500 patients, Raichand et al described a patient who was incidentally found to have peripheral retinal schisis in a distribution similar to that in our second patient. This patient’s left eye also was phthisical, suggesting a high risk of retinal detachment in such patients. In both of our patients, nonperfusion, which was prominent outside the distribution of the radial peripapillary net but most prominent in the area of the temporal raphe, probably occurred slowly over decades.

Sickle type SS produces systemic complications more often than ocular ones but certainly leads to low-grade ischemia. In both of our patients, chronic low-grade ischemia had affected the inner nuclear layer, leading to schisis, epiretinal proliferation, and eventually traction on the inner layer. In all likelihood, it was the chronicity of ischemia that allowed schisis to occur. Rapidly progressive proliferative sickle retinopathy would have been characterized by traction detachments because there would not have been enough weakening of the intraretinal cohesive forces to permit schisis. The pathomechanism of schisis in patients such as ours may be similar to that of proliferative diabetic retinopathy, in which schisis is found after 20 years of disease accounting for half of all tractional elevations of the retina.

One can only speculate about the value of timely laser treatment to nonperfused retina in cases such as those reported. Laser scars may make the retina more cohesive and reduce the mediated stimulus for proliferation and traction on the inner layer. Therefore, all patients with sickle cell, including those with the sickle trait and particularly those with conjunctival sickle signs, should be followed up for extraretinal proliferations to prevent schisis-related retinal detachment, a rare but vision-threatening complication of proliferative sickle cell retinopathy.

Hermann D. Schubert, MD

Correspondence: Dr Schubert, Columbia University, 635 W 165th St, New York, NY 10032 (hds@columbia.edu).

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