acuity was 20/20 OU with correction. She described distortion without scotoma on the Amsler grid bilaterally (Figure 1C). Examination findings were otherwise normal except for stage 2 papilledema in both eyes (Figure 2D) and moderate enlargement of the physiologic blind spots on automated perimeter in both eyes. The MRI results were normal. Lumbar puncture showed an opening pressure of 410 mm CSF, with normal indices. Results of OCT of the macula were normal. No follow-up data are available at this time.

Comment. Metamorphopsia is a visual distortion in which straight lines appear curved, and it is commonly seen in disorders that disrupt the normal orientation of the macular photoreceptors. It may remit or it may become permanent. Metamorphopsia has not specifically been mentioned in reports of IIH, but by implication it occurs in the many patients with IIH who have severe papilledema, resulting in a macular star or “fan” due to extravasation of fluid from the edematous nerve between the nerve fibers into the macula. Severe papilledema may also be associated with Paton lines (concentric retinal folds surrounding the optic nerve that may also induce metamorphopsia). A syndrome of acquired choroidal folds and hyperopia has been described with and without IIH. The incidence of metamorphopsia in patients with IIH may be underreported because of the other, more debilitating symptoms experienced by these patients, or because physicians fail to document its presence.

The cases we describe are all unusual in that the metamorphopsia was associated with very mild papilledema (stage 2 or less). None of the patients had visible distortions of the maculae, choroidal folds, macular edema, or hyperopic shifts. We speculate that the metamorphopsia was due to traction on the retina from nerve elevation, with resultant disruption of photoreceptor orientation, or from fluid within the retina or nerve fiber layer that was not apparent by ophthalmoscopy, angiography, or OCT. Although OCT is a sensitive indicator of retinal abnormalities, it probably lacks the resolution necessary to detect subtle misalignments of the photoreceptor layer. Patients with more severe metamorphopsia associated with more severe papilledema do have distortions in their retinal anatomy discernible on OCT (J.E.A.W., unpublished data, 2004).

Despite the resolution of papilledema, none of these patients had complete relief of their metamorphopsia. We speculate that these patients did not experience complete relief of their metamorphopsia because of the permanent anatomical disruption of photoreceptor alignment associated with prolonged papilledema. Three of the women had moderate myopia, and 1 had mild myopia; it is possible that the anatomy of their myopic globes and the topography of their retinas somehow predisposed them to the development of metamorphopsia. Two patients had peripapillary hemorrhages. We considered that the metamorphopsia could be a manifestation of subtle myopic degeneration or of some other condition of the peripapillary retina (such as acute zonal occult outer retinopathy [AZOOR]). However, none of these patients had refractive errors greater than −6.00, and none had ophthalmoscopic or angiographic evidence of myopic degeneration. In addition, 2 patients (cases 2 and 3) had normal photoreceptor function when evaluated by multifocal ERG. It remains to be seen whether these women harbor an occult form of macular degeneration that will become apparent in the future.

We describe 4 middle-aged women with initial complaints of metamorphopsia coinciding with the development of mild papilledema. Because no other cause of metamorphopsia was discovered and because of the temporal association between the onset of metamorphopsia and the finding of papilledema, we believe that these patients experienced metamorphopsia as an initial symptom of IIH. Physicians should consider the diagnosis of IIH in patients with metamorphopsia and optic nerve edema.

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References

Optical Coherence Tomographic Findings in X-linked Juvenile Retinoschisis

X-linked juvenile retinoschisis is a progressive bilateral disease that is probably present at birth and has been documented as early as 7 weeks of age. It was first reported by Haas in 1898 and has recently become better understood as a mutation of the XLRS1 gene on the short arm of the X chromosome (Xp22). This mutation results in an abnormal retinal protein that participates in intercellular spaces. Cystoid changes arranged in a stellate pattern with radial striae projecting from the fovea are seen in all patients, along with a peripheral schisis in 50% of cases, and variable findings are well described by Gass and others. In this study, we used optical coherence tomography (OCT) (Stratus OCT, Carl Zeiss Meditec AG, Jena, Germany) to examine the foveal areas in 2 patients with juvenile retinoschisis. Our OCT findings suggest that the foveal schisis is probably located in the outer plexiform layer, not in the nerve fiber layer (NFL), as de-
scribed previously in peripheral retinoschisis.5,6

**Report of a Case.** A 19-year-old African-American man had had poor vision in both eyes since early childhood. Family history was significant for 1 male sibling with poor vision and 2 blind male cousins. On examination, best-corrected Snellen visual acuity was 20/100− OD and 20/300 OS. Pupils were normal with no relative afferent pupillary defect. The anterior segments were unremarkable. Fundus examination showed typical stellate cystoid changes at the central macula in both eyes (**Figure 1**). In the right eye, the stellate change did not start in the central fovea, where a 400-µm area of the retina looked flat. The radial stellate change was apparent outside of this flat zone (**Figure 1A**). In the left eye, the stellate change started from the central fovea (**Figure 1B**). At the inferior temporal periphery, retinoschisis cavities with inner-layer holes were noticed in both eyes.

The OCT scans showed that in the right fovea (**Figure 2A**), the inner nuclear layer and ganglion cell layer were condensed into a single layer. The outer nuclear layer was not of uniform thickness and showed atrophic change. The outer plexiform layer appeared as an uneven slit between the outer nuclear layer and the inner retina. The retinal pigment epithelium showed focal atrophic changes (**Figure 2A**). In the left fovea (**Figure 2B**), large foveal cystoid spaces were noticed that were localized between the inner and outer retina, specifically in the outer plexiform layer. These cystoid spaces were of relatively uniform thickness, and there were multiple bridging strands crossing in the cystoid spaces. These strands were vertically oriented and, in some areas, very evenly spaced, resembling a ladder resting on its side. In other areas, a “rung” was missing, suggesting the possible coalescence of smaller uniform spaces. There were also small areas of early retinal pigment epithelial atrophy. The most notable finding, however, was that the cystoid pockets appeared to be located deeper in the retina than the highly reflective NFL. They were localized in the outer plexiform layer, as seen in typical pseudophakic cystoid macular edema.4

The patient’s 12-year-old brother was also examined, and his visual acuity was counting fingers at 3 ft OD and 20/60 OS. The fundus photographs of the right eye were nearly identical to those of his 19-year-old brother’s left eye: prominent stellate change present in the foveal center. His left eye showed no stellate change but only atrophic retinal pigment epithelial mottling (photograph not shown). The OCT of the 12-year-old showed that in the right fovea, cystoid spaces were already formed in the foveal center, where the foveal pit was still recognizable (**Figure 3A**). In the parafoveal area, small cystoid changes were noticed as well. These cystoid spaces were clearly localized in the outer plexiform layer. Occasional tiny pockets of cystoid changes were seen in the inner retina, probably in the inner plexiform layer. In the left eye, small cystoid changes had begun to develop on the temporal side of the fovea (**Figure 3B**), but not on the nasal side. The foveal pit was easily identified. Thus, retinoschisis appeared to start in the foveal center and spread toward the parafoveal area.
Comment. Yanoff et al7 and Manschot6 described the retinoschisis in this disease as occurring in the NFL based on histopathologic findings, further postulating that the primary defect might then involve the Muller cells. The OCT findings in our cases suggest that the foveal cystoid separation is not located in the NFL, as described in peripheral retinoschisis,5,8 but is in the outer plexiform layer. Trese and Foos7 reported a series of premature infants with infantile cystoid maculopathy resembling X-linked juvenile retinoschisis, in whom gross examination showed cystoid pockets at various retinal layers including deep to the NFL. Azzolini et al8 reported OCT findings in 3 cases of X-linked juvenile retinoschisis showing a macular cleavage plane in the outer retinal layers as well as in the NFL. Optical coherence tomography provides an in vivo correlation to previous investigations of the histopathologic features of the disease. The OCT findings in this report suggest that the primary abnormality of the fovea in patients with juvenile retinoschisis is actually in the outer retina, specifically in the outer plexiform layer, unlike the peripheral retina, where the schisis is located in the NFL. These findings suggest different developmental mechanisms of retinoschisis in the fovea and peripheral retina. A larger series of OCT imaging in this interesting disease is desirable.

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Intraocular Surgery After Treatment of Germline Retinoblastoma

Germline retinoblastoma results from a somatic mutation involving loss or inactivation of the tumor suppressor gene located at 13q14.1.1 Absence of gene activity predisposes to retinoblastoma and other tumor development.1,2 The introduction of chemotherapy protocols with focal consolidation therapy has enhanced our ability to treat germline retinoblastoma while salvaging many eyes that would have been lost previously.3,4 A secondary benefit has been that we are now preserving eyes with useful vision. However, because of the aggressive, multimodal therapy involved, patients often develop intraocular complications. We evaluated a group of patients who had undergone enucleation for retinoblastoma in one eye with salvage of the other eye and who had maintained tumor quiescence for a period of 12 months or longer in that eye. Our purpose was to determine how these patients fared following intraocular surgery of the salvaged eye.

Patients and Methods. We performed a retrospective review of all germline retinoblastoma cases from January 1, 1985, until December 31, 2000, to identify all patients who had undergone unilateral enucleation with salvage of the other eye. Cases were drawn from the records of the University of Tennessee, Memphis, Department of Ophthalmology, Memphis; the records of the Ophthalmic Oncology Service at St Jude Children’s Research Hospital, Memphis; and the private records of Retina Associates of Florida, Tampa. Cases were enrolled only if the salvaged eye achieved a 12-month period of tumor quiescence and then subsequently underwent intraocular surgery for non–tumor-control reasons. Tumor quiescence was defined as lack of documented tumor growth, lack of vitreous or subretinal seeding, lack of anterior chamber seeding, and lack of metastases. Eligible intraocular surgery included cataract extraction, barrier laser for retinal break, scleral buckle procedure, pars plana vitrectomy, and Nd:YAG laser capsulotomy. Original treatment of the retinoblastoma was recorded, as was the Reese-Ellsworth classification. The 3 primary outcomes were tumor activity, visual acuity, and development of complications. Tumor activity was classified as quiescent, recurrent, and...