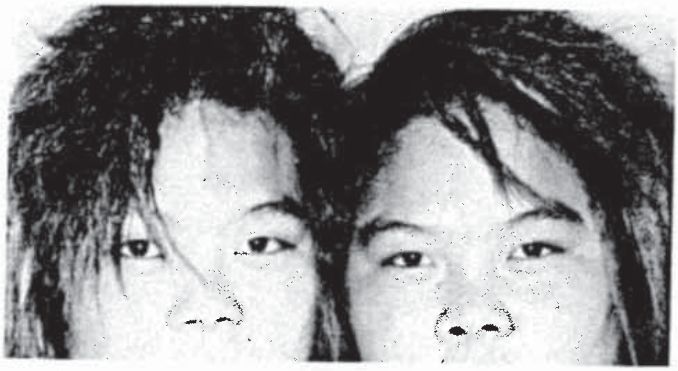

Figure (Rutzen and associates). Monozygotic twins with Vogt-Koyanagi-Harada syndrome demonstrate poliosis.

rheumatoid factor, angiotensin converting enzyme, erythrocyte sedimentation rate, and serologic tests for syphilis were normal or negative. A year earlier she had a positive tuberculin skin test and a normal chest x-ray, and had been treated with isoniazid while receiving systemic prednisone over the previous year.

Visual acuity was R.E.: 20/20 and L.E.: 20/25. Both eyes had a mild anterior chamber cellular reaction and pigmented keratic precipitates. Extensive posterior synechiae and peripheral laser iridectomies were observed bilaterally. The anterior vitreous contained a moderate number of cells, and mild optic nerve hyperemia was present in both eyes. Focal depigmented lesions were seen in the equatorial region of the fundus in both eyes.

• CASE 2: The 16-year-old monozygotic twin of the patient described in Case 1 was also examined because of a one-year history of panuveitis that had been treated with oral prednisone. She had blurred vision and mild neck stiffness. Poliosis had also developed, but she had no headaches, tinnitus, alopecia, or vitiligo. Laboratory investigations as outlined in Case 1, but also including chest x-ray and tuberculin skin testing, were all normal or negative. Visual acuity was 20/20 in both eyes. Ocular examination disclosed findings identical to those of Case 1.

Both patients fulfilled the diagnostic criteria for Vogt-Koyanagi-Harada syndrome suggested by the American Uveitis Society.[5] The HLA typing in both patients showed HLA-DR4, HLA-DR7, HLA-DQw2, and HLA-DQw3. HLA-DR4 is present in a disproportionate number of Japanese, Chinese, and Hispanic patients with the Vogt-Koyanagi-Harada syndrome when compared with controls.[1-3] The HLA-DR4 antigen is found in 56% to 100% of individuals with the Vogt-Koyanagi-Harada syndrome, compared with 20% to 40% of controls. The incidences of other HLA antigens, such as HLA-DR1, HLA-DR53, and HLA-DQ4, are also increased in patients with the Vogt-Koyanagi-Harada syndrome. Although familial cases are rare, associations with HLA antigens may indicate that genetic factors play a role in this disease.

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Early Stage of Macular Hole in a Severely Myopic Eye

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PURPOSE/METHODS: A progressive early stage of macular hole developed in a severely myopic eye of a 37-year-old woman.

RESULTS/CONCLUSIONS: The patient improved after pars plana vitrectomy, peeling of the posterior hyaloid, and gas-fluid exchange. Biomicroscopic, ultrasonographic, and intraoperative findings support the theory of cortical vitreous traction as the cause of macular hole in severely myopic eyes.

SEVERELY MYOPIC INDIVIDUALS ARE KNOWN TO BE A high-risk group for macular holes. The early stages of idiopathic macular holes have been well described, but the early stage of a macular hole in a severely

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myopic eye has not, and the origin of macular hole associated with high myopia is uncertain. The early stages of idiopathic macular hole have led to the theory of tangential cortical vitreous traction on the central fovea.[1] This case demonstrates a progressive early stage of macular hole formation in a severely myopic individual, and supports the theory of tangential cortical vitreous traction as the cause of macular hole in severely myopic eyes.

A 37-year-old myopic woman had a two-day history of a focal central blurred spot in the vision in her right eye. Her visual acuity was R.E.: 20/25 (−7.25 +0.50 × 168) and L.E.: 20/20 (−7.75 +1.25 × 175). Contact lens biomicroscopy disclosed no clinical evidence of a posterior vitreous detachment, but disclosed fine macular cystic changes with loss of the foveolar light reflex. Over six weeks her visual acuity worsened to 20/50 with enlargement of the area of metamorphopsia. A 500-μm yellow foveal ring with thinning, elevation of the central fovea, and a focal 50-μm hole developed (Fig. 1). B-scan ultrasonographic examination showed no evidence of posterior vitreous detachment, but showed focal insertion of cortical vitreous fibers into the foveal region, and a localized foveal elevation (Fig. 2). Axial length measurement was 26.5 mm in both eyes.

After the patient gave careful informed consent, a pars plana vitrectomy, peeling of the posterior hyaloid, and gas-fluid exchange with 18% C_3F_8 were performed. Intraoperative findings included a posterior staphyloma with tilted disk, and intraoperative development of a posterior vitreous detachment with a Weiss's ring and a 50-μm operculum attached to the hyaloid over the macula. After 14 months, visual acuity in the right eye improved to 20/20 with slight residual metamorphopsia. Slit-lamp examination showed no evidence of cataract, and ophthalmoscopic examination showed loss of the foveolar light reflex, but did not show macular cysts or hole. Mild symptomatic blurring developed in the left eye, but visual acuity remained 20/20. Fine petalloid cystic changes were noted in the central fovea, which did not progress over five months.

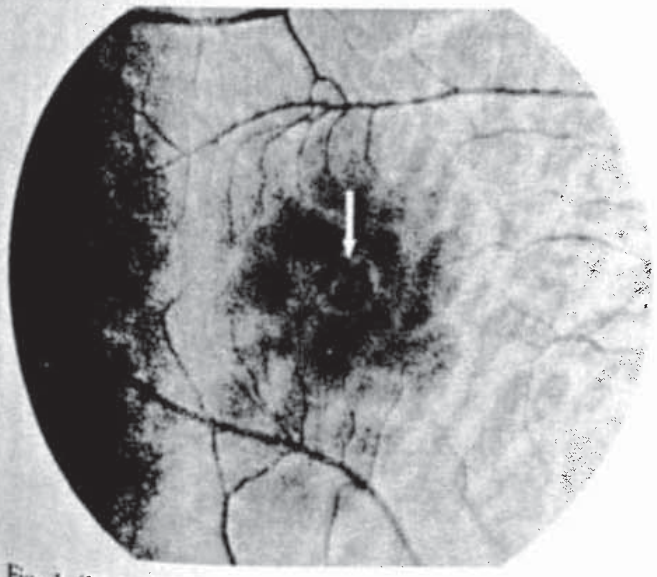

Fig. 1 (Kokame). Fundus photograph of right macula shows yellow ring with tractional striae and tractional foveal elevation. Note the 50-μm hole just within the superior edge of the yellow ring (arrow).

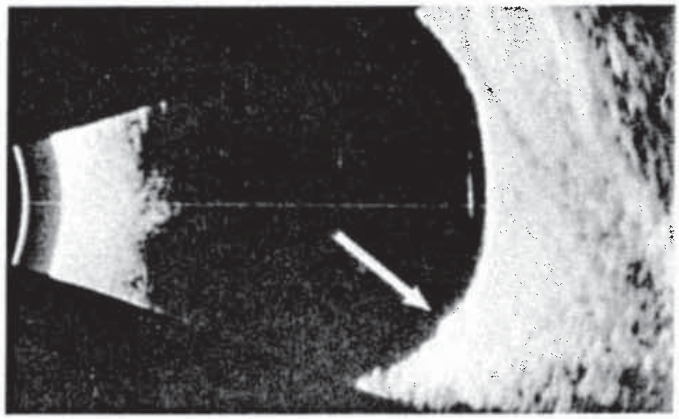

Fig. 2 (Kokame). B-scan ultrasonographic longitudinal scan of the macula at reduced sensitivity demonstrates localized foveal elevation (arrow). Note the large size of the globe and insertion of optic nerve posterior to the foveal elevation.

This case demonstrates a progressive early stage of macular hole in a severely myopic eye identical in appearance to a stage II idiopathic macular hole in the classification system of Gass.[1] Additionally, there was no biomicroscopic, ultrasonographic, or intraoperative evidence of posterior vitreous detachment or vitreofoveal separation. Vitreous attachments to the central fovea could thus be responsible for tangential traction and the localized foveal elevation (Fig. 2), as has been theorized in idiopathic macular holes.[1] Severely myopic eyes may be at increased risk for macular holes because of an increased incidence of vitreomacular adherence and relatively early syneretic changes in the vitreous, resulting in traction on the

central fovea at an earlier age than idiopathic macular holes.[2] If cortical vitreous traction is responsible for macular hole in severely myopic eyes, then pars plana vitrectomy, with peeling of the posterior hyaloid, may be a reasonable therapeutic alternative in such patients. Surgical intervention in idiopathic macular holes with vitreoretinal surgical techniques may improve vision,[3,4] and is presently being studied in a randomized multicentered study.[5] Further surgical experience with macular holes in severely myopic individuals is necessary before surgical intervention can be generally recommended.

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Ophthalmoscopic Visualization of the Inferior Ophthalmic Vein Through a Chorioretinal Coloboma

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PURPOSE/METHODS: In a patient with a coloboma of the optic nerve, inferior retina and choroid, and deep portions of the sclera, a retrobulbar vessel moved irregularly relative to the retinal vessels, which moved contiguously with the posterior globe.

RESULTS/CONCLUSION: Fluorescein angiography showed the retrobulbar vessel filling slightly later than overlying retinal vessels. We conclude that the retrobulbar vessel was a vein situated inferiorly to the optic nerve.

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TYPICAL OR ATYPICAL COLOBOMAS OF THE IRIS, ciliary body, and choroid can occur as an isolated dysembryogenesis, or can be a component of more complex malformations of known or unknown origin. Typical colobomas occur as a result of faulty closure of the embryonic fissure anywhere from the optic disk to the pupil in the inferonasal meridian.[1-3]

A 34-year-old man came to our clinic because of diminishing vision in his left eye. He had had a history of right exotropia since early childhood. Spontaneous shrinkage of the right globe occurred at the age of 8 years. The right eye had been enucleated at the age of 18 years. There was no history of trauma. General health was good and there was no family history of coloboma. Visual acuity in the left eye was 20/25. Biomicroscopy showed an inferior coloboma of the iris and ciliary body as well as a lens coloboma at the 6 o'clock meridian with an adjacent early cortical cataract. Although pupillary dilatation was poor, the pupil reacted well to light. The intraocular pressure was 18 mm Hg. On ophthalmoscopy we saw a large coloboma of the optic nerve, inferior retina and choroid, and deep portions of the sclera. The margin of the coloboma was highlighted by hyperplastic retinal pigment epithelium. A thin diaphanous membrane, continuous with the retina and containing retinal vessels, extended over the defect. In the depth of the coloboma, retinal pigment epithelium and choroid were absent, and sclera was attenuated.

Behind the intercalary membrane, another vascular structure suspended in the orbital fat was visible in the retrobulbar space. During ocular movement, the overlying retinal vessels moved contiguously with the posterior globe, but the retrobulbar vessel showed irregular movements in an opposite direction relative to the retinal vessels, with an aftermotion once the eye movement had ceased. Fluorescein angiography showed the retrobulbar vessel filling slightly later than the overlying retinal vessels, and demonstrated the countermotion as well as the aftermotion of this vessel in regard to movements of the globe and the retinal vessels (Figure). B-scan ultrasonography confirmed the coloboma inferior to the optic disk, with