

Intrapapillary Hemorrhage with Adjacent Peripapillary Subretinal Hemorrhage

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Purpose: To describe the clinical features and to present results of new diagnostic methods to help define the cause of the clinical syndrome of intrapapillary hemorrhage with adjacent peripapillary subretinal hemorrhage (IHAPSH).

Design: Retrospective review of patients with IHAPSH at presentation seen in 3 centers in Hawaii and Japan.

Methods: We analyzed data including patient demographics, presenting symptoms, initial and final visual acuities, biomicroscopic findings, fundus photographs, and results of available ancillary testing, including fluorescein angiography, B-scan ultrasonography, and optical coherence tomography.

Results: There were 10 eyes of 9 patients (7 female and 2 male, 8 Asian and 1 white) aged 14 to 79 years. All patients experienced an acute onset of visual symptoms. Eight eyes had mild to severe myopia (−2.50 diopters [D] to −9.50 D), and 8 eyes had a tilted disc. Hemorrhage within the disc and adjacent subretinal hemorrhage were located nasally in 6 eyes, superiorly in 2 eyes, and temporally in 2 eyes. Vitreous hemorrhage was noted in 6 of 10 eyes. Posterior vitreous evaluation by biomicroscopy (10 eyes), by B-scan ultrasonography (4 eyes), and by optical coherence tomography (2 eyes) revealed no evidence of vitreopapillary traction, except for a follow-up optical coherence tomography in 1 eye showing localized vitreoretinal separation with residual attachment to the optic disc 10 months after presentation. Fluorescein angiography showed mild disc staining in 4 of 8 eyes. Hemorrhage spontaneously resolved within 1 to 7 months, and there were no recurrent hemorrhages with an average follow-up of 13.5 months (range, 2–31 months). Visual acuities maintained or improved to 20/25 or better in 8 eyes. The other 2 eyes had unrelated poor vision.

Conclusions: Intrapapillary hemorrhage with adjacent peripapillary subretinal hemorrhage is more common in myopic eyes and spontaneously resolves without treatment. The unique structural architecture of the elevated nasal edge of the myopic tilted disc and the choroidal blood supply of the prelaminar optic nerve may predispose patients to bleeding from the optic discs, which may be spontaneous or may be precipitated by acute disc edema, Valsalva maneuver, or vitreopapillary traction. *Ophthalmology* 2004;111:926–930 © 2004 by the American Academy of Ophthalmology.

A clinical syndrome of intrapapillary hemorrhage with adjacent peripapillary subretinal hemorrhage (IHAPSH) has been described since 1975.¹ In these cases, there is significant bleeding within the optic disc that extends into the peripapillary subretinal space and often into the vitreous. This syndrome usually resolves spontaneously with a good visual prognosis, frequently occurs in myopic eyes with tilted discs, and is associated with localized disc staining in later phases of the fluorescein angiogram.^{2–8} Many of the cases were reported in young, otherwise healthy individuals of Asian ethnicities.

The cause of this condition is uncertain, although vitreous traction on the disc has been proposed as one possible cause.^{1,2,6} The purpose of this study was to define further the clinical characteristics and course associated with IHAPSH and to report on the findings of new diagnostic testing, including diagnostic ocular ultrasonography and optical coherence tomography, which may help to evaluate the role of vitreous traction in IHAPSH.

Materials and Methods

Patients with acute bleeding within the optic disc and adjacent peripapillary subretinal hemorrhage surrounding the disc hemorrhage at presentation were identified and evaluated retrospectively from February 1994 through February 2002 in 3 centers in Hawaii and Japan. This retrospective study was reviewed and approved by the Institutional Review Board of Hawaii Pacific Health. The case histories, ocular examination results, and ancillary testing were reviewed. All 9 patients underwent slit-lamp biomicroscopy and fundus photography. Four eyes underwent B-scan ultrasonography, 8 eyes underwent fluorescein angiography, and 2 eyes underwent optical coherence tomography.

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The criteria for inclusion in the study were ophthalmoscopic evidence of both intrapapillary and adjacent peripapillary subretinal hemorrhage. Exclusion criteria included disc hemorrhage only without adjacent peripapillary subretinal hemorrhage, subretinal hemorrhage only without associated intrapapillary hemorrhage, optic nerve head drusen, peripapillary subretinal neovascularization, ischemic optic neuropathy, Terson's syndrome, and Leber's idiopathic stellate neuroretinitis. Follow-up duration ranged from 2 months to 31 months after the initial presentation. Patients were followed up until the complete resolution of the hemorrhage.

The following data were reviewed: age, gender, ethnicity, presenting symptoms, possible precipitating event, visual acuity, pupillary reflexes, and visual field, which was tested by confrontation, tangent screen, and automated or Goldmann perimetry. Stereoscopic fundus photographs were reviewed for disc anatomy, location of hemorrhage in and around the disc, other retinal findings, and findings in the fellow eye. Diagnostic testing included fluorescein angiography, B-scan ultrasonography, and optical coherence tomography in selected cases.

Particular attention was given to examination of the status of the vitreous in relation to the optic disc to evaluate the role of vitreopapillary traction as a possible cause of this syndrome.^{1,2,6} In this study, using slit-lamp biomicroscopy, the presence of a visible, freely moving Weiss' ring was defined as a posterior vitreous detachment (PVD),⁹ and the presence of a fibrous glial ring attached to the disc was defined as a partial PVD. B-scan ultrasonography provides an extremely accurate method for determining the vitreoretinal interface^{10,11} and was performed in 4 eyes using a standard contact method on the globe to assess the vitreoretinal and vitreopapillary relationships using kinetic evaluation of the vitreous with high gain.^{10,11}

Results

The clinical features and course on 10 eyes of 9 patients were evaluated in the study. Patients' ages ranged from 14 to 79 years (average, 47 years). There were 7 females and 2 males. All patients were Asian (7 Japanese, 1 Korean), except for one white person (patient 9). All patients had an acute onset of visual symptoms at presentation, including blurred vision, floaters without flashes, black spots, smudge in the visual field, and decreased visual acuity. None had a history of diabetes, vasculitis, or bleeding diathesis. In 2 instances, patients had a preceding history of Valsalva maneuvers, including a vigorous sneeze and lifting a heavy trash bag. Two patients had long-standing hypertension without hypertensive retinopathy. Two patients underwent recent chemotherapy for breast cancer. One patient chronically was taking anti-inflammatory medication.

There were 3 right eyes and 7 left eyes, with 1 patient having bilateral involvement. Eight of 10 eyes were myopic, including 2 mildly myopic (≤ -3 diopters [D]), 4 moderately myopic (-3 D to -6 D), and 2 severely myopic (≥ -6 D). One had mild hyperopia ($+1.00$ D). Presenting best-corrected Snellen visual acuities were 20/40 or better in 7 eyes, 20/50 to 20/100 in 1 eye, and 20/200 or worse in 2 eyes. For the 2 eyes with visual acuity less than 20/200, both had unrelated preexisting conditions, including a full-thickness macular hole in 1 eye and myopic macular degeneration and posterior staphyloma in the other eye. Mild retinal pigment epithelial atrophic changes associated with myopic macular degeneration were noted in 3 eyes without effect on visual acuity.

A tilted disc and localized nasal disc edema were noted in 8 of 10 eyes (Fig 1A), all of which had mild to severe myopia and subretinal hemorrhage on the nasal or superior edge. Two eyes

with normal disc structure had peripapillary subretinal hemorrhage on the temporal edge (Fig 2). The optic nerve head appeared nondysplastic, without disc drusen, and pink in all cases. Cup-to-disc ratios were observed at the time of presentation or after the resolution of disc edema and ranged from 0.2 to 0.45 in 8 eyes and <0.2 in 1 eye. Of the 8 eyes with tilted disc in the involved eye with IHAPSH, tilted discs were seen in 5 fellow eyes. One of these fellow eyes with a tilted disc went on to have IHAPSH 12 months after the initial episode in the other eye.

Intrapapillary hemorrhage was observed superonasally in 4 eyes, inferotemporally in 3 eyes, and for 360° in 3 eyes, extending beyond the disc margin as an intraretinal hemorrhage in 4 eyes. Peripapillary subretinal hemorrhage was seen adjacent to the intrapapillary hemorrhages in all cases. Crescent-shaped peripapillary subretinal hemorrhage extended from 0.25 to 2.5 disc diameters from the disc edge. The extent of peripapillary subretinal hemorrhages ranged from 1 to 12 clock hours. In 2 eyes with normal disc architecture, the peripapillary hemorrhages appeared on the temporal side, 1 of which extended into the fovea (Fig 2). Among the other 8 eyes with nasal or diffuse disc edema, peripapillary subretinal hemorrhages were seen nasally in 6 eyes (Fig 1A) and superiorly in 2 eyes. Microscopic vitreous hemorrhage was limited to the region over the disc in most cases, but inferior vitreous hemorrhage was noted in 6 of 10 eyes.

At presentation, posterior vitreous evaluation by biomicroscopy demonstrated a complete PVD in 2 eyes. However, there was no evidence of complete PVD, partial PVD, or vitreopapillary traction in the other 8 eyes. In 4 eyes, B-scan ultrasonography was performed specifically looking at the vitreopapillary relationships. There was no evidence of partially detached posterior vitreous cortex inserting into the optic nerve, using axial and longitudinal scans with high gain and scans perpendicular to the optic nerve to allow the best potential views of partially detached vitreous attached to the optic nerve (Fig 1B).^{10,11} Optical coherence tomography was performed in 2 eyes. The examination in 1 eye at presentation showed marked disc edema but did not show evidence of vitreopapillary traction (Fig 1C). In the same eye at the 10-month follow-up, insertion of the partially detached vitreous cortex to the superotemporal aspect of the disc was evident (Fig 1D). In another eye, the test was performed at the 17-month follow-up only and the eye did not show evidence of vitreopapillary traction.

Three female patients from the center in Japan underwent computed tomography and magnetic resonance imaging of the brain, the results of which were normal. In those 3 subjects, serum diagnostic test results, including complete blood count, fibrinogen, prothrombin time, partial thromboplastin time, and fibrin degradation products, also were normal.

Fluorescein angiography was performed in 8 eyes. Seven eyes with superior or nasal subretinal hemorrhage were evaluated at the time of presentation, and 4 of those 7 eyes were studied with repeat angiography. Initial angiograms all showed blocked hypofluorescence from intrapapillary and peripapillary subretinal hemorrhage (Fig 1E). Five eyes had mild unilateral localized disc staining (Fig 1E). In 3 eyes, we were unable to assess disc staining because of a large area of blockage from extensive superficial hemorrhage and lack of follow-up fluorescein angiography after the resolution of hemorrhage. One eye with temporal subretinal hemorrhage underwent angiography only 2 months after presentation, which showed no blockage or disc staining. None of the studies showed disc drusen; abnormal intraretinal, intrapapillary, or choroidal vasculature; or subretinal neovascularization after the resolution of the hemorrhage.

Hemorrhage spontaneously resolved within 1 to 7 months (mean, 2 months) without treatment. Visual acuities maintained or improved to 20/25 or better in all eyes except the 2 with preexisting macular disorders as noted above. Visual acuity of the eye

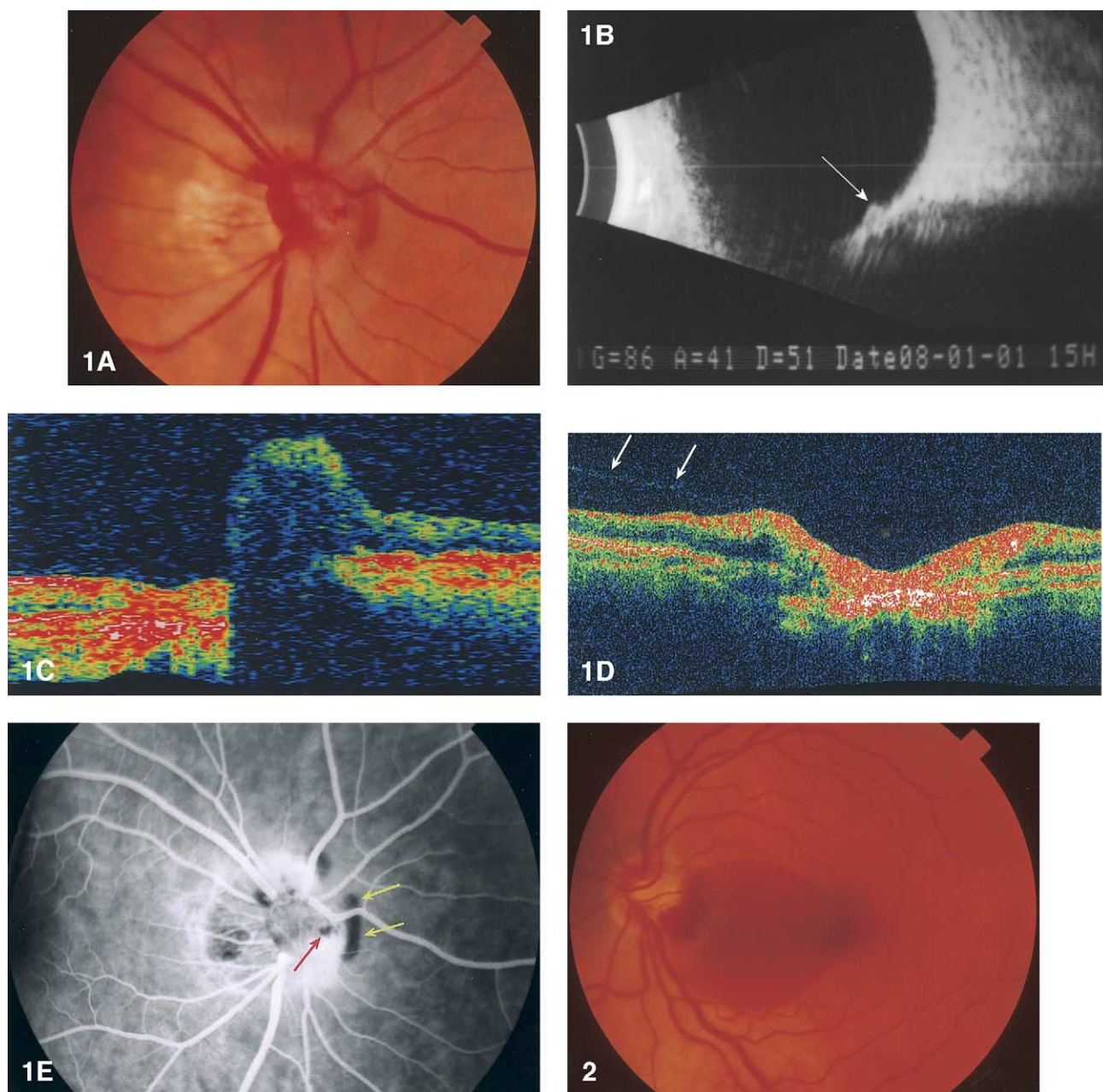


Figure 1. **A**, Fundus photograph of the right eye showing nasal intrapapillary hemorrhage at the 3-o'clock position and adjacent nasal peripapillary subretinal hemorrhage (patient 1). Note disc edema of the nasal disc margin and severe tilting of the optic disc. **B**, Diagnostic B-scan ultrasonography of patient 1 with high gain perpendicular to the optic nerve showing elevated optic disc (white arrow). Note the absence of evidence of vitreopapillary traction without partially detached posterior vitreous cortex inserting into the optic nerve. **C**, Optical coherence tomography of patient 1 (horizontal scan of optic nerve) at presentation demonstrating marked nasal edema of the optic disc without evidence of vitreopapillary traction. Note the absence of vitreous cortex inserting into the optic disc. **D**, Optical coherence tomography (vertical scan of the optic nerve) demonstrating localized vitreoretinal separation with residual attachment to the superior margin of the optic disc (white arrows) 10 months after initial presentation. **E**, Late-phase fluorescein angiogram demonstrating focal disc staining on the nasal aspect of the optic disc and a blocking defect resulting from intrapapillary hemorrhage (red arrow) and peripapillary subretinal hemorrhage (yellow arrows). Note marked disc edema of the nasal edge and severe tilting of the optic disc.

Figure 2. Fundus photograph showing intrapapillary hemorrhage involving the temporal margin of disc and adjacent peripapillary subretinal hemorrhage extending into the central fovea. Note the lack of a tilted disc (patient 9).

with a macular hole remained 20/200, and the eye with myopic macular degeneration improved from 20/200 to 20/70. There have been no recurrences of intrapapillary hemorrhage in the same eye during the follow-up period ranging from 2 to 31 months (average, 12 months).

Discussion

Intrapapillary hemorrhage with adjacent peripapillary subretinal hemorrhage (IHAPSH) most commonly occurs in

eyes with a tilted disc architecture associated with myopia and has a benign clinical course with an excellent visual prognosis. The hemorrhage in the disc and in the peripapillary subretinal space resolves spontaneously without visible damage to the nerve or retina. In our series and that of previously published cases, there have been no reported cases of recurrence in the same eye. Although IHAPSH is associated most commonly with a tilted disc and subretinal hemorrhage located superiorly or nasally, as noted in 80% of the cases in this series and in 2 additional previously published cases,³ this syndrome also can occur with temporal subretinal hemorrhage, which can extend into the macula and even the central fovea. In our series based in Hawaii and Japan with large Asian populations, IHAPSH most commonly occurred in Asian patients, which is consistent with previously reported cases being predominantly Asian.^{1,2,4-8} However, 1 of our patients was white and did not have a tilted disc, and cases in non-Asian persons rarely have been reported.² Although some investigators suggested a predilection for younger patients,^{2,4,5,8} the age of our patients ranged from 14 years to as old as 79 years, with an average age of 47 years. A female predominance was noted (78%) in our series.

The acute onset of IHAPSH could be spontaneous, but could also be the result of an acute event stimulating optic disc bleeding in these eyes. Any theory of pathogenesis with regard to this syndrome of IHAPSH should explain the common features noted in these cases: (1) bleeding from the disc, (2) increased frequency in tilted discs of myopic eyes, (3) more common involvement of superior and nasal aspects of the optic disc, (4) acute onset with recovery of good vision, and (5) lack of recurrence.

The vascular anatomy of the prelaminar portion of the optic nerve head has unique architecture.¹²⁻¹⁴ The arterial supply is derived from peripapillary choroid and posterior short ciliary arteries. The venous system of the prelaminar portion almost exclusively drains to the central retinal vein with minor contribution to peripapillary choroidal veins.^{12,14} Bleeding may originate from capillaries of choroidal origin, where they penetrate the nerve tissues in the prelaminar portion of the optic disc, as suggested by Watanabe et al⁵ and Hirotsuji et al.⁷

In myopic eyes, the discs appear tilted because of the oblique insertion of the nerve into the sclera of the posterior staphyloma. This tilting results in elevation of the superior and nasal margin of the disc. This elevated superior and nasal margin of the tilted disc results in dragging of the retinal and choroidal tissues over and around the elevated edge.^{15,16} Japanese authors^{5,7} have suggested the dragging puts capillaries at risk for bleeding.

One proposed theory of IHAPSH is acute vitreopapillary traction.^{1,2,6} Cibis et al¹ initially postulated posterior vitreous detachment based on the diagnosis of posterior vitreous detachment by biomicroscopy. Katz and Hoyt² theorized partial posterior vitreous detachment based on a glial tag on the optic disc and reported findings of partial attachment of the vitreous to the optic disc by B-scan ultrasonography. Conversely, biomicroscopic and ultrasonographic evidence of vitreopapillary traction was not confirmed in 2 previously published cases by Kokame³ and was not noted biomicro-

scopically in any of the cases in this series of 10 eyes. Ultrasonography^{3,10,11,17,18} and optical coherence tomography¹⁹⁻²¹ are diagnostic methods that allow careful assessment of vitreoretinal relationships, which cannot be well visualized on biomicroscopic examination. These techniques have been useful in delineating subtle vitreomacular traction in macular hole and other vitreomacular traction disorders.^{10,20,21} None of 4 additional eyes evaluated with ocular ultrasonography in this series had evidence of partial posterior vitreous detachment from the nerve. Optical coherence tomography also did not demonstrate vitreopapillary traction at the time of acute bleeding (Fig 1C). Although vitreous traction could be an inciting factor in some cases, these results suggest there may be other acute triggers to initiate bleeding.

Acute disc edema is another possible acute event that could be responsible for this syndrome of IHAPSH. Disc swelling was seen clinically in 80% of the cases in this series. Disc staining on fluorescein angiography at the time of initial presentation was demonstrated in 2 prior published IHAPSH cases³ and in 4 of 7 eyes in this current series. The other 3 eyes in this series showed a large blocking defect preventing assessment for disc edema. Marked disc swelling also was confirmed on ultrasonography (Fig 1B) and optical coherence tomography (Fig 1C). We previously reported 2 cases of Leber's idiopathic stellate neuroretinitis that had associated intrapapillary and peripapillary subretinal hemorrhage.³ Inflammatory disc edema thus can result in a similar appearance of intrapapillary hemorrhage with adjacent peripapillary subretinal hemorrhage. Ischemic optic neuropathy, which also can result in disc edema, has also been shown to result in this finding of intrapapillary hemorrhage with associated peripapillary subretinal hemorrhage.^{22,23} Although none of the cases in this series had findings consistent with Leber's idiopathic stellate neuroretinitis or ischemic optic neuropathy, acute disc swelling may be a common etiologic factor in these cases. In addition, these cases of intrapapillary hemorrhage and peripapillary subretinal hemorrhage associated with neuroretinitis and ischemic optic neuropathy further demonstrate that this type of bleeding can occur unassociated with vitreopapillary traction.

Valsalva maneuvers are another possible acute precipitating event that may affect the vascular system of the optic disc in susceptible eyes. Two of the cases in this series had a history of Valsalva maneuver. Satomi and Ohara⁸ reported a similar case of a 17-year-old male with myopia who had an enlarged blind spot at presentation immediately after lifting a 40-kg barbell in a gym. Valsalva maneuvers may put a shearing force on the vulnerable capillaries of choroidal origin on the nasal edge of the disc and may initiate bleeding. Andreoli et al²⁴ described a patient who repeatedly demonstrated transient decreased perfusion to the central retinal artery after sneezing, as shown with color Doppler imaging. This evidence suggests acute vascular hemodynamic changes resulting from a Valsalva maneuver occur in the prelaminar area and may play a role in bleeding from the optic disc in this syndrome.

In conclusion, this benign syndrome of intrapapillary hemorrhage with adjacent peripapillary subretinal hemor-

rhage affects patients of all ages and both genders. It most commonly affects myopic eyes with tilted discs and spontaneously resolves without treatment, with an excellent prognosis for vision recovery and without recurrence. Intrapapillary and peripapillary hemorrhage usually appears on the nasal or superior aspect of tilted discs, but may also occur in nonmyopic eyes in which the adjacent subretinal hemorrhage extends temporally from the disc to the macula. Especially in these eyes, it is important to note the intrapapillary hemorrhage as the source of bleeding.

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