Accuracy of Papilledema and Pseudopapilledema Detection: A Multispecialty Study

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Background. Present trends in medical care suggest that primary care physicians will exert increasing control over patient access to medical specialty consultation and diagnostic testing. Therefore, it is important to determine whether primary care physicians can reliably identify papilledema.

Methods. A prospective study involving 429 physicians was undertaken to assess the accuracy of papilledema and pseudopapilledema detection by five groups of physicians, family practice physicians, neurologists, neuro-ophthalmologists, neurosurgeons, and ophthalmologists.

Results. Neuro-ophthalmologists and ophthalmologists did better than family physicians, neurologists, and neurosurgeons in identifying both papilledema and pseudopapilledema ($P < .05$). Neuro-ophthalmologists more accurately identified pseudopapilledema than all other groups in the study ($P < .05$). Family physicians did as well as, or better than, neurologists and neurosurgeons in identifying all classifications of acute and chronic papilledema defined in the study. Family physicians did not perform as well as the other four groups in differentiating pseudopapilledema from papilledema ($P < .05$).

Conclusions. Although the sensitivity of detecting papilledema was high (84.5%) for family physicians, the specificity was low (59.3%). Preliminary data indicate that family physicians with prior exposure to clinical ophthalmology in medical school did better than those who had not had training. It is possible that additional exposure to clinical ophthalmology during residency training might yield improved performance.

Key words. Optic disc; papilledema; comparative study. *J Fam Pract 1991; 33:381-386.*

Primary care physicians often determine which patients are referred to other specialists and which receive neurodiagnostic studies, such as a computed tomographic scan or magnetic resonance imaging.[1–4] These physicians, therefore, have primary responsibility for identifying the early signs and symptoms of life-threatening disorders such as intracranial tumors, or vision-threatening disorders such as idiopathic intracranial hypertension, also known as pseudotumor cerebri.[5,6] Papilledema from increased intracranial pressure is a hallmark of intracranial tumors and idiopathic intracranial hypertension. Approximately 60% of all patients with intracranial tumors have papilledema when their tumors are first discovered. Nearly 75% of the patients with infratentorial tumors are found on examination to have papilledema.[7–9] Papilledema is present in all patients with uncontrolled idiopathic intracranial hypertension.[10] The accuracy of detecting papilledema is crucial for primary care physicians who oversee specialty referrals. The present study sought to compare how well various groups of medical practitioners can identify papilledema. These groups included family practice physicians, neurologists, neuro-ophthalmologists, neurosurgeons, and ophthalmologists.

Methods

The study was conducted over a 1-year period, at the following national and international conferences: the 1988 Neuro-Ophthalmology Congress (Vancouver, British Columbia); the American Neurological Association conference (Philadelphia, Pennsylvania); the American Academy of Ophthalmology conference (Las Vegas, Nevada); the American Academy of Family Physicians

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conference (Los Angeles, California); and the Congress of Neurological Surgeons (Atlanta, Georgia). A scientific booth was provided by the organizers of these conferences, and physicians attending the conferences volunteered their participation in the study. The survey of each medical group was conducted over 2 to 4 days, depending on the duration of the meeting. Statistical analysis was performed, comparing the accuracy of the responses on a given test day with the accuracy of other days' responses.

Each participant viewed one set of 23 stereophotographic slides through an illuminated hand-held stereo viewer. Although stereophotographic slides were used, participants in the study could choose at any time to view the slides monocularly (similar to an image obtained with a monocular direct ophthalmoscope) or binocularly (similar to an image obtained with an indirect ophthalmoscope). This format allowed participants the option of viewing the fundus in the manner in which they were most comfortable. To accommodate simultaneous testing of several subjects, three sets of the same 23 stereophotographic slides were used; the sequence of slide presentations were randomized so that there were three different sequences available. Statistical analysis was performed to test for differences in score accuracy due to differing slide-sequence presentation. Each slide contained a full view of an optic disc. The slides showed normal optic nerves, papilledema, pseudopapilledema from optic nerve head drusen, and other disorders that should not be confused with papilledema, such as advanced glaucoma and optic atrophy. Slides exhibiting papilledema were obtained from patients with proven increased intracranial pressure as documented by a computed tomographic scan showing an intracranial tumor with shift of the ventricles past midline or from patients who had a normal orbital and cranial computed tomographic scan but had an opening pressure on lumbar puncture of greater than 250 mm H_2O. Cases of optic nerve head drusen were documented by autofluorescence on fluorescein angiography, calcification on orbital computed tomographic scan, and in some cases, normal lumbar puncture.

To ensure that the slides were of good quality and represented papilledema and nonpapilledema, and to grade the severity of the papilledema, three neuro-ophthalmologists, each with 19 or more years of experience in neuro-ophthalmology (mean 24.6 years), viewed the slides without knowledge of the answers. If at least two of the three experts gave the same clinical diagnosis for a slide, then that slide was accepted as being a good representation of papilledema or nonpapilledema. The scores of the three experts were 20, 21, and 22 correct responses. For all 23 slides, at least two of the experts agreed with the actual clinical diagnosis. Thus, all 23 slides were considered good representations of papilledema or nonpapilledema.

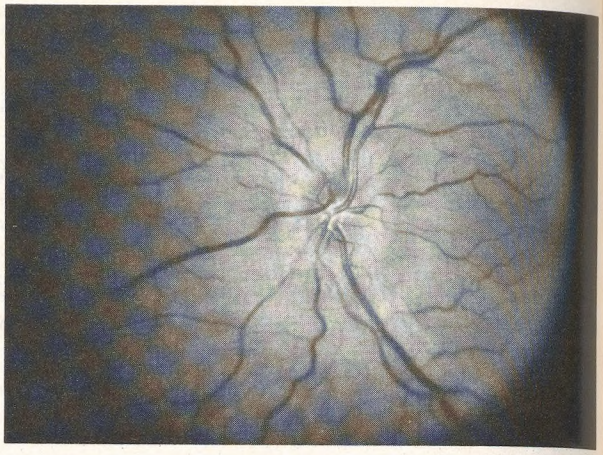

Figure 1. Mild-acute ("early") papilledema showing blurred nerve fiber layer at the superior and inferior optic disc margins, mild elevation of the optic disc, incomplete obliteration of the optic cup, and slight dilatation and tortuosity of the retinal veins.

The expert panel was asked to classify the stereophotographic slides of papilledema as acute or chronic papilledema, and to indicate whether there was mild, moderate, or marked severity of papilledema. If two or all three panel members independently arrived at the same classification of a slide, then the slide was classified with that consensual temporal (acute or chronic) and descriptive (mild, moderate, marked) classification. The descriptive and temporal classifications in the study consisted of mild-acute papilledema (Figure 1), moderate-acute papilledema (Figure 2), marked-acute papilledema (Figure 3), moderate-chronic papilledema (Figure 4), and marked-chronic papilledema (Figure 5). Mild-chronic papilledema cases were not included in the study.

Participants in the study completed a questionnaire before viewing the slides. This information was used in part to screen for test duplication. The physicians were requested to provide the last four digits of their Social Security number or another identification card number. The physicians listed their primary and secondary specialties such as family medicine, neuro-ophthalmology, neurology, neurosurgery, and ophthalmology, and indicated the number of years of residency if they were still in training. The number of years of practice in the primary and secondary specialties was ascertained. The physicians were grouped as neuro-ophthalmologists if neuro-ophthalmology was identified as the primary or secondary specialty. All other specialists were grouped according to the primary specialty indicated. The number of weeks of

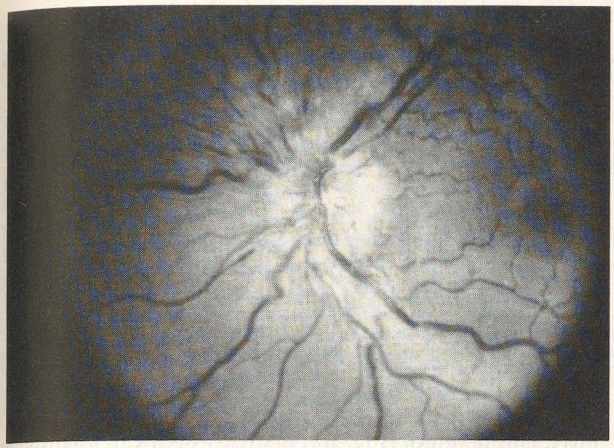

Figure 2. Moderate-acute papilledema with increased blurring of the peripapillary nasal nerve fiber layer, incomplete obscuration of retinal blood vessels at the convexity and margins of the optic disc, increased optic disc elevation, and the presence of exudates and flame-shaped hemorrhages.

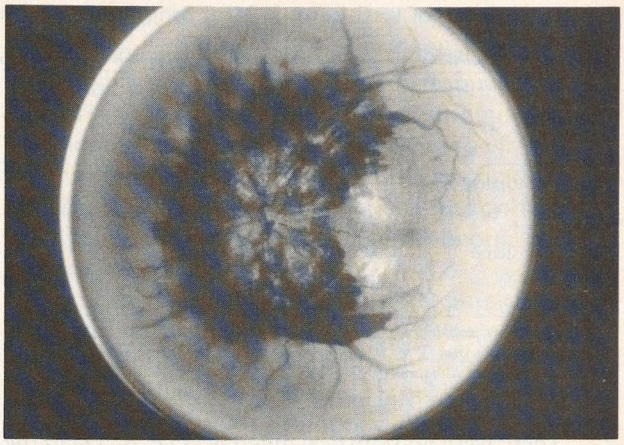

Figure 3. Marked-acute papilledema showing obscuration of all blood vessels on the optic disc convexity and margins, complete obliteration of the optic cup, marked increase in optic disc elevation, presence of intravitreal, preretinal and intraretinal hemorrhages, and macular exudates.

clinical ophthalmology that the participant had in medical school and the country in which medical school training was completed were recorded.

Analysis of variance with the F test and a Student-Newman-Keuls test were performed to assess significant differences.

Results

The test scores of a given conference day did not differ significantly ($P \geq .17$) from other days' test scores for a particular conference, indicating that differing test dates did not influence the responses. In addition, there was no statistical difference ($P = .71$) in test scores among the three different sequences of slide presentation.

A total of 429 physicians voluntarily enrolled in the study (Table 1). There were 102 family physicians (89 of whom were graduates of United States or Canadian medical schools), 69 neuro-ophthalmologists (47 graduates of United States or Canadian medical schools), 56 neurologists (41 graduates of United States or Canadian medical schools), 51 neurosurgeons (40 graduates of United States or Canadian medical schools), and 151 ophthalmologists (115 graduates of United States or Canadian medical schools).

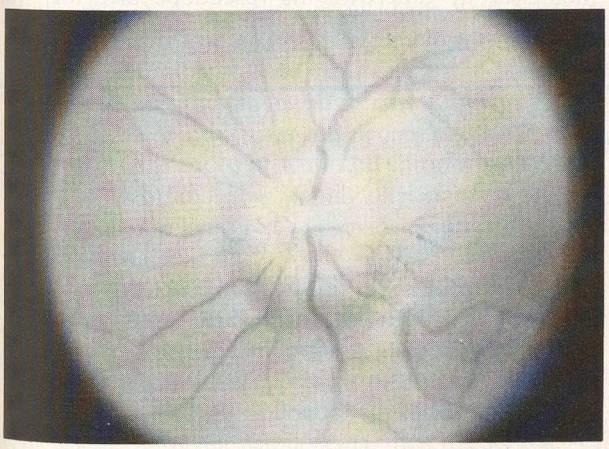

Figure 4. Moderate-chronic papilledema showing moderate optic disc elevation, peripapillary vessel sheathing, and waxy pallor of the peripheral optic disc margins.

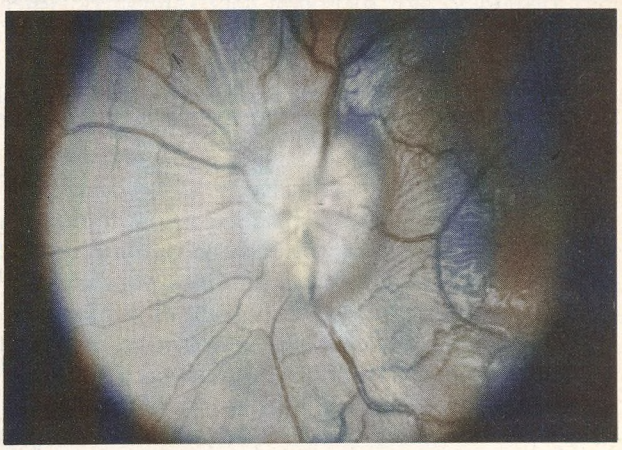

Figure 5. Marked-chronic papilledema showing marked optic disc elevation, peripapillary vessel sheathing, diffuse waxy pallor of the optic nerve, and drusen-like deposits within the optic nerve head.

Table 1. Accuracy of Participants in Identifying Papilledema and Pseudopapilledema Based on Country of Medical School Training

Specialty	United States/Canada No.	Mean Score*	SD	Other Countries No.	Mean Score	SD	Total No.	Mean Score
Neuro-ophthalmology	47	19.5	1.90	22	18.7	2.78	69	19.2
Ophthalmology	115	18.8	2.13	36	18.3	2.38	151	18.7
Neurology†	41	17.3	2.43	15	19.1	2.26	56	17.8
Family medicine	89	17.2	2.07	13	16.8	2.30	102	17.2
Neurosurgery	40	16.9	3.11	11	17.2	1.99	51	17.0
Total	332			97			429	

**The highest score possible was 23 correct responses.*
†A significant difference (P = .01) in performance was noted only between United States and Canadian neurologists compared with neurologists from other countries.
SD denotes standard deviation.

There were no statistical differences ($P \geq .23$) in the performances of United States and Canadian neuro-ophthalmologists, ophthalmologists, neurosurgeons, or family physicians when compared with their respective groups from other countries. There was a significant difference ($P = .01$), however, in the performance of the neurologists, such that the United States and Canadian neurologists did not score as well as neurologists from other countries (Table 1). Since the study attempted to address the performance of physicians who had their medical school training in the United States and Canada, no further comparisons were made with physicians from other countries.

There was no significant difference ($P \geq .18$) in the performance of the residents in training when compared with physicians in practice for the respective medical groups. Thus, the scores for physicians in practice and for residents in training were grouped together.

There was no statistical difference ($P > .05$) between the performance of the neuro-ophthalmologists and the ophthalmologists. The neuro-ophthalmologists and ophthalmologists scored better than the neurologists, family physicians, and neurosurgeons in identifying both papilledema and nonpapilledema ($P < .05$). When papilledema was further classified into the various temporal and descriptive groups (Table 2), no statistical differences ($P \geq .14$) were observed among the five groups in identifying mild-acute and marked-acute papilledema. The neuro-ophthalmologists and ophthalmologists did better than the neurologists and neurosurgeons ($P < .05$), but no better than the family physicians, in identifying moderate-acute and moderate-chronic papilledema. The neuro-ophthalmologists, ophthalmologists, and family physicians did better than the neurosurgeons ($P < .05$), and the family physicians did better than the neurologists ($P < .05$), in identifying marked-chronic papilledema. The family physicians did less well than the other four groups in differentiating pseudopapilledema (optic disc drusen) from papilledema ($P < .05$). The neuro-ophthalmologists more accurately identified pseudopapilledema than all other groups in the study ($P < .05$). There was no difference in the performance of the neurologists, neurosurgeons, and ophthalmologists in differentiating pseudopapilledema. The

Table 2. Accuracy of Identifying Papilledema and Pseudopapilledema

Classification	Neuro-ophthalmologists	Ophthalmologists	Neurologists	Family Physicians	Neurosurgeons
Acute Papilledema					
Mild (3)*	2.21	2.09	2.05	2.26	1.95
Moderate (3)†	2.89	2.85	2.54	2.71	2.52
Marked (1)	0.91	0.87	0.88	0.78	0.90
Chronic papilledema					
Moderate (5)†	4.26	4.10	3.63	4.19	3.48
Marked (2)†	1.66	1.70	1.54	1.89	1.40
Pseudopapilledema					
Optic disc drusen (5)†	3.89	3.41	3.12	1.91	3.05
Other					
Nonpapilledema (4)	3.79	3.83	3.56	3.43	3.58

**The number in parentheses in column 1 is the total possible score for that classification.*
†Indicates significant intergroup differences (P < .05) for that classification.

specificity of detecting papilledema for the neuro-ophthalmologists was 85.3% and the sensitivity was 85.2%, for the ophthalmologists, 80.4% and 82.9%; for the neurologists, 74.2% and 76.0%; for the neurosurgeons, 73.7% and 73.2%; and for the family physicians, 59.3% and 84.5%.

A significant difference in test performance correlating with the number of weeks of ophthalmology training in medical school was noted for the family physicians. The family physicians who did not have a clinical ophthalmology rotation during medical school did not do as well as those who received some clinical training ($P < .05$). There was no correlation between test performance and the number of weeks of ophthalmology training in medical school for neurologists and neurosurgeons.

The years in specialty practice had no effect on the performance of any group except for neurologists. Neurologists in practice for less than 5 years or in practice for 10 or more years did better than those in practice for 5 to 10 years ($P < .05$).

Discussion

The detection and management of vision-threatening disorders such as idiopathic intracranial hypertension and life-threatening disorders such as intracranial tumors still need the continued involvement of such specialists as neuro-ophthalmologists, ophthalmologists, neurologists, and neurosurgeons.[11] Nevertheless, since significant disease morbidity and mortality can be reduced with early disease detection, it is important that primary care physicians be able to identify early stages of papilledema. The study confirmed that the family physicians were able to identify mild-acute ("early") papilledema with accuracy similar to that of the neuro-ophthalmologists, ophthalmologists, neurologists, and neurosurgeons. The study also showed that the family physicians did as well as or better than the neurologists and neurosurgeons in identifying all classifications of acute and chronic papilledema defined in the study. Thus, family physicians within this study population appeared to provide accurate diagnosis of papilledema.

Although family physicians appeared to have done well in diagnosing papilledema, this may be related to the study design. Participants in the study had to determine whether papilledema was present or absent. If the participants identified all abnormal-appearing or elevated optic discs as showing papilledema, then the accuracy of detecting papilledema would be high. This would, however, result in poor discrimination between true papilledema and pseudopapilledema. Indeed, the results indicated that the neuro-ophthalmologists more accurately identified pseudopapilledema than all other groups in the study, while the family physicians performed poorly in this area. The family physicians often misdiagnosed pseudopapilledema as true papilledema. The family physicians did correctly identify all normal optic discs. The results indicate that family physicians tend to overdiagnose papilledema when evaluating an abnormal-appearing or elevated optic disc.

In routine clinical practice, family physicians, neurologists, and neurosurgeons generally do not examine the optic disc in stereoscopic projection through a dilated pupil. Thus, these clinicians were provided with a better than customary view of the optic disc, which may account for the fairly good performance (≥73.9% correct) of all groups in the study. Although nonstereoscopic viewing of the optic disc through an undilated pupil, as with the use of direct ophthalmoscopy, would mimic routine nonophthalmologic clinical practice, we wanted the best possible score for family physicians, neurologists, and neurosurgeons. Despite this advantage, the neuro-ophthalmologists and ophthalmologists did statistically better than the family physicians, neurologists, and neurosurgeons in identifying both papilledema and pseudopapilledema.

Papilledema may be associated with several conditions including intracranial tumors, idiopathic intracranial hypertension (pseudotumor cerebri), shunt obstruction, subarachnoid hemorrhage, subdural hematoma, and intracranial inflammation.[12] Similarly, many causes exist for optic disc elevation that mimic papilledema such as refractive error, optic disc hypoplasia, myelinated optic nerve, optic disc drusen, primary and metastatic optic nerve tumors, optic neuritis, sarcoidosis, and retinal vascular occlusion.[12] One important role of primary care physicians is to screen for abnormalities. Hence, the detection of any abnormality of the optic nerve head by family physicians would be beneficial for patient management. Some of these conditions require only periodic ophthalmologic evaluations, however, while others require more extensive evaluations with computed tomography or magnetic resonance imaging. Although the sensitivity of detecting papilledema was high (84.5%) for the family physicians, the specificity was low (59.3%) as compared with the neuro-ophthalmologists, ophthalmologists, neurologists, and neurosurgeons. These data suggest that substantial savings in the cost of medical care possibly could be obtained if patients diagnosed with papilledema by family physicians were evaluated by one of the above specialists before obtaining neuroradiologic studies.

The study indicated that the number of years in practice did not influence the performance of the family

physicians in the study. However, those family physicians who received training in clinical ophthalmology as medical students did better than those who had no training. It appears that exposure to clinical ophthalmology as medical students is important in improving the clinical diagnostic ability to differentiate papilledema from pseudopapilledema, at least for family physicians. The study did not assess the differences in performance of family physicians who did or did not receive training in clinical ophthalmology during family medicine residency. Future studies could address this issue.

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References

1. Clancy CM, Hillner BE. Physicians as gatekeepers—the impact of financial incentives. Arch Intern Med 1989; 149:917–20.
2. Caldwell JR. Is the gatekeeper concept legitimate or another mirage on the cost containment trip? J Fla Med Assoc 1988; 75:109–11.
3. Reagan MD. Physicians as gatekeepers—a complex challenge. N Engl J Med 1987; 317:1731–4.
4. Powe NR, Eisenberg JM. Studying and teaching the gatekeeper. J Gen Intern Med 1986; 1:197–8.
5. Corbett JJ, Savino PJ, Thompson HS, et al. Visual loss in pseudotumor cerebri: follow-up of 57 patients from five to 41 years and a profile of 14 patients with permanent severe visual loss. Arch Neurol 1982; 39:461–74.
6. Wall M, George D. Visual loss in pseudotumor cerebri—incidence and defects related to visual field strategy. Arch Neurol 1987; 44:170–5.
7. Petrohelos MA, Henderson JW. The ocular findings in intracranial tumor. Am J Ophthalmol 1951; 34:1387–94.
8. Raimondi AJ, Tomita T. Hydrocephalus and infratentorial tumors—incidence, clinical picture, and treatment. J Neurosurg 1981; 55:174–82.
9. Raimondi AJ, Yashon D, Matsumoto S, Reyes CA. Increased intracranial pressure without lateralizing signs: the midline syndrome. Neurochirurgia 1967; 10:197–209.
10. Durcan FJ, Corbett JJ, Wall M. The incidence of pseudotumor cerebri—population studies in Iowa and Louisiana. Arch Neurol 1988; 45:875–7.
11. Corbett JJ, Thompson HS. The rational management of idiopathic intracranial hypertension. Arch Neurol 1989; 46:1049–51.
12. Whiting AS, Johnson LN. Papilledema: clinical clues and differential diagnosis. Am Fam Physician. In press.