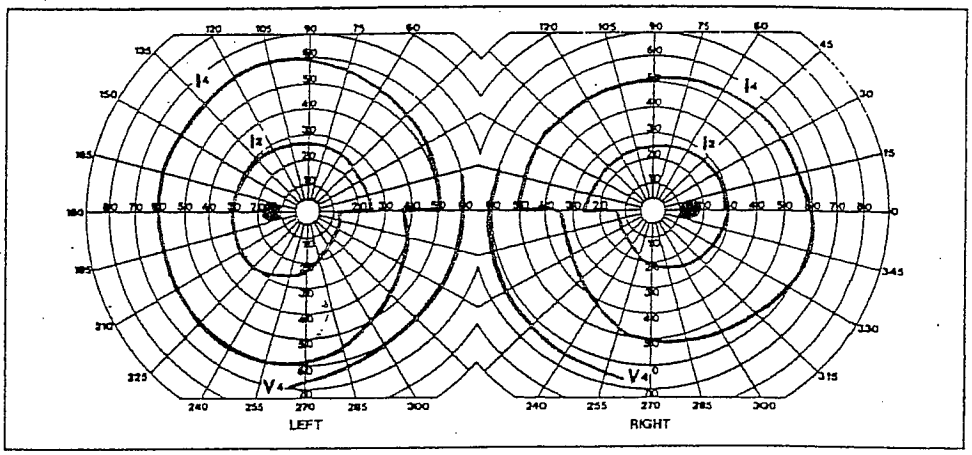
Pseudopapilledema

A 12-year-old boy was referred because of headaches and elevated optic disks. The headaches, which were often associated with nausea, were described as bifrontal and throbbing, occurring about once a week, lasting 1 to 4 hours, and generally relieved by sleep. He had no visual complaints. His medical history was otherwise unremarkable.

Neuro-Ophthalmic Examination

	OD	*OS*
Visual acuity	20/20	20/20
Color plates correct	10 of 10	10 of 10
Pupils	Normal	Normal
Motility, lids, orbits	Normal	Normal
Fundi (see Plate 12)	Elevated disk	Elevated disk



Results of a neurologic examination were normal.

Summary

A 12-year-old boy with headaches was found to have elevated optic disks and visual field defects but otherwise normal examination results.

Differential Diagnosis

In a case such as this, the clinician must determine whether the patient has true disk edema, which probably would be secondary to increased intracranial pressure, or whether the disk appearance is that of pseudopapilledema. Obviously the distinction is a crucial one since the diagnosis of true papilledema will indicate the urgent need for additional studies. In most cases the differentiation can be made with a high degree of certainty on clinical grounds.

If papilledema is well developed, the diagnosis is usually obvious. It is early papilledema that is difficult to distinguish from pseudopapilledema. Some of the distinguishing ophthalmoscopic features are summarized below.

Feature	*Early papilledema*	*Pseudopapilledema*
Disk color	Hyperemic	Pink, yellowish pink
Disk margins	Indistinct early at superior and inferior poles, later entire margin	Irregularly blurred, may be lumpy
Disk elevation	Minimal	Minimal to marked, center of disk most elevated
Vessels	Normal distribution, slight fullness, spontaneous venous pulsations absent	Emanate from center, frequent anomalous pattern, spontaneous venous pulsations present or absent
Nerve fiber layer	Dull owing to edema, which may obscure blood vessels	No edema, may glisten with circumpapillary halo of feathery light reflections
Hemorrhages	Splinter	Subretinal, retinal, vitreous

The appearance of the nerve fiber layer on the disk and at its margin is generally the one most useful distinguishing feature. In true papilledema the edema in the nerve fiber layer will generally obscure to some degree the underlying blood vessels, whereas in pseudopapilledema the vessels can be seen clearly. Venous distention and hemorrhages are possible in both and thus are not differentiating features. The presence of spontaneous venous pulsations would suggest pseudopapilledema, but as mentioned in Chapter 6 intracranial pressure can fluctuate between elevated and normal levels and thus the observation of spontaneous venous pulsations does not rule out an intracranial process.

In this case it can be seen in the photographs that although the disk margins were blurred, there was no obscuration of the underlying blood vessels and the nerve fiber layer was clear. Also the elevation of the disks was most prominent nasally in both eyes, particularly in the left. These

features all suggest a diagnosis of pseudopapilledema. The headaches were probably migraine and were presumed to be unrelated to the disk anomaly.

Clinical Diagnosis: Pseudopapilledema

Pseudopapilledema is most commonly produced by optic disk drusen, although the drusen may be "buried" within the disk and not visible on the surface. High hyperopia with a small scleral canal and heaping up of the nerve fibers is another possible cause. Occasionally remnants of the embryologic hyaloid or myelination of the nerve fibers overlying the disk may be confused with disk edema.

Optic disk drusen are dominantly inherited with variable penetrance. The incidence is 20 in 1000, and they are present in whites much more commonly than in blacks. The pathogenesis of drusen has been postulated to be related to axonal degeneration from altered axoplasmic flow. The presence of drusen is usually an incidental finding, and not infrequently the observation of disk elevation is made in a patient being examined for a cause of headaches (as in this case). When on the surface of the disk, the drusen may be observed to be whitish and glistening. When buried they are not obvious, but the clinical features described above help to differentiate them from true papilledema. In children drusen tend to be buried, whereas in adults they are more often on the surface. The progression from buried to surface drusen in a given individual has been documented.

Optic disk drusen generally do not produce any visual symptoms although rarely a patient may experience transient visual obscurations similar to those with increased intracranial pressure. However, visual field defects are common, occurring in about 70 percent of eyes with visible drusen and 35 percent with pseudopapilledema but no visible drusen. The visual field defects are generally in a nerve fiber bundle distribution (see Chapter 1), with inferior nasal defects being most common. An increase in the blind spot and generalized field constriction are also possible. Progression of visual field defects is well documented. It is rare for visual acuity to be decreased from drusen, and if decreased visual acuity is noted in a patient with drusen, an evaluation for an alternative cause should be performed.

Visual loss can occur with drusen for reasons other than an effect on the optic nerve fibers: hemorrhage, retinal degeneration, or ischemic optic neuropathy. Hemorrhages with drusen are usually subretinal in a peripapillary distribution. Retinal and vitreous hemorrhages are less common. Retinitis pigmentosa occurs in association with drusen in about 2 percent of cases. Ischemic optic neuropathy occurs with a higher than expected frequency, especially in young adults.

Additional Testing and Management

If a clinical diagnosis of pseudopapilledema can be made with certainty, additional testing is not necessary. Fluorescein angiography is generally not helpful as there may be staining of the disk in both early papilledema and drusen. Ultrasound may be useful to identify buried drusen. If a diagnosis of pseudopapilledema is probable but not definite and the patient has no neurologic symptoms or signs, it is reasonable to photograph the disks and reexamine the patient in 1 to 2 months. If true papilledema is present some changes in the interim would be expected, whereas with pseudopapilledema the appearance should be identical.

If the diagnosis is in doubt, it is prudent to perform computed tomography to rule out an intracranial mass and, if the scan is normal, a lumbar puncture to measure intracranial pressure.