Principles and Practice of Clinical Electrophysiology of Vision

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Ischemic Optic Neuropathy

Graham E. Holder

Anterior ischemic optic neuropathy (ION) usually presents in the older patient with painless, often severe visual loss of sudden onset. Ophthalmoscopy reveals pallid swelling of the optic disc that may be accompanied by superficial peripapillary hemorrhages. The findings are probably caused by acute ischemia of the anterior portion of the optic nerve supplied by the posterior ciliary artery circulation.[12]

The initial report of severe visual loss in association with giant-cell arteritis appears to be that of Jennings,[18] but the term *ischemic optic neuritis* was first used by Wagener.[29] It is now usually known as ischemic optic neuropathy.[23] Recent clinical reviews identify two groups of patients: those with (arteritic) and those without (nonarteritic) giant-cell arteritis.[3, 12, 26] Many nonarteritic cases are idiopathic, but systemic hypertension and diabetes mellitus are contributory factors.[3, 26] There are reports of ION in conjunction with hypotension,[27] with migraine,[4, 22, 30] and following cataract surgery.[19, 25]

The clinical presentation of patients with ION is to some extent dependent upon the etiology. Nonarteritic patients present with visual loss in one eye, often with previous involvement of the other eye. The optic disc is swollen, and the more extensive the disc swelling, the greater the degree of visual impairment.[3] Flame hemorrhages are usually present.[3] The most common field defects are inferior altitudinal or inferior nasal; some patients have central loss. The field defect may poorly correlate with the fundus appearance, but some patients have clear superior or inferior swelling with corresponding altitudinal field loss. In one large series[12] more than 35% of the nonarteritic patients had a visual acuity of 6/36 or worse, but 30% had normal (6/9 or better) acuity.

Those patients with arteritic ION often have symptoms associated with temporal arteritis: malaise, muscle pain, scalp tenderness, etc., whereas the nonarteritic patients do not feel unwell. There is often a generalized field constriction in the affected eye. Visual acuity may be severely reduced, with 60% having an acuity of counting fingers or worse,[12] but also may be unimpaired. A percentage of both groups may have had previous transient visual dysfunction. The blood erythrocyte sedimentation rate (ESR) is usually raised in temporal arteritis, but a low ESR does not exclude the diagnosis,[12] and positive temporal artery biopsy findings are necessary for confirmation. It is important to distinguish the cases due to temporal arteritis from idiopathic cases because high-dose steroids are the treatment of choice in arteritic ION.[7] There have been reports of improvement following steroid administration in nonarteritic patients,[12] but as yet there is no satisfactory treatment.

The histological changes in arteritic ION have been reviewed by Henkind et al.[13] The orbital vessels, including the posterior ciliary arteries, the ophthalmic artery, and the intraneural central retinal artery, may be involved in the arteritic process, but involvement of the intraocular retinal or choroidal vessels is unusual. Hayreh and Baines[10, 11] suggested that the posterior ciliary arteries feed fairly well delineated areas of the choroid and nerve head and that posterior ciliary artery occlusions may infarct the optic disc and adjacent retrolaminar optic nerve. The surface of the optic disc may, however, be fed by branches of the central retinal artery, with the rest of the prelaminar and laminar portions of the optic nerve supplied by a system with contributions from the posterior ciliary arteries, the pial vessels, and choroidal arterioles.[1, 9] A case report, with-

out clinical details, of the histopathological findings in nonarteritic ION showed focal infarction 3 mm behind the lamina cribrosa that was caused by thromboembolism in three discrete pial and pial-derived arterioles.[21] The temporal aspect of the macula showed ischemic necrosis.

The first detailed report of the electrophysiological findings in ION was that of Wilson,[32] although "delays" in the pattern visual evoked potential (PVEP) had been mentioned.[2, 14] He examined both PVEP and flash VEPs (FVEPs) in a mixed group of 15 arteritic and nonarteritic patients. Both PVEPs and FVEPs showed reduced amplitude. Only 4 patients showed minimal (<10 ms) latency changes. The clinically uninvolved eye invariably had normal VEPs. These findings were contrasted with those in optic nerve demyelination where latency delays in excess of 10 ms are common and there is often subclinical involvement of the fellow eye. Other authors[5, 6, 8, 17, 31] confirmed the high incidence of reduced-amplitude, normal-latency VEPs, but Glaser and Laflamme[6] found a predominance of P100 component delays in acute cases. Harding's group[8] noted that all affected eyes showed a reduced VEP and that those that showed a delayed or triphasic response to flash had temporal arteritis. The delayed FVEP in arteritic patients was confirmed by this author.[17] It was also noted that the PVEP was more sensitive than the FVEP in nonarteritic patients. Amplitude reductions were usually relative to the uninvolved eye. Typical findings are shown in Figure 82–1. Cox et al.[5] compared the PVEPs from 24 eyes with ION with 22 eyes with optic nerve demyelination. The mean latency difference between the involved and the uninvolved eyes was 3 ms for ION but 21 ms in demyelination. Wildberger[31] found amplitude changes but also observed that patients with an inferior altitudinal defect touching the horizontal meridian show apparent latency delays that he attributed to preservation of the normal longer-latency response from the superior field.[20]

Definite latency delays have been reported,[24] but stimulus and recording parameters were not given, and a recent study[28] emphasized the difficulties in accurate component identification with a single midline recording channel (see also Chapter 18 for a discussion of normal PVEP components and their distribution). Thompson et al.[28] occasionally used a 15-

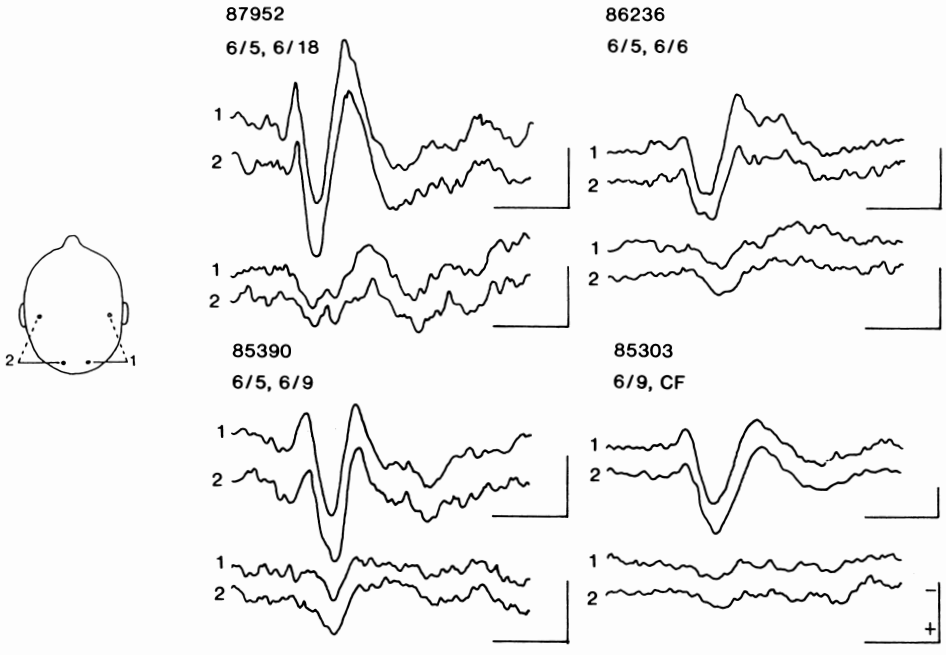

FIG 82–1.

PVEP findings (11-degree full field, 26-minute checks, 1.95 reversals per second) in four patients with nonarteritic ION. The *upper pair* of tracings from each patient represents the uninvolved eye and in each patient falls within the normal range. Note the marked amplitude reduction in all affected eyes. One case (85390) shows a purely P100 amplitude reduction, one case (87952) shows a broadening of P100 with an increased-latency N135 component, one case (86236) shows a mild P100 latency increase, and in the fourth case (85303) the PVEP is probably extinguished (calibration, 5 μV, 80 ms; positivity downward).

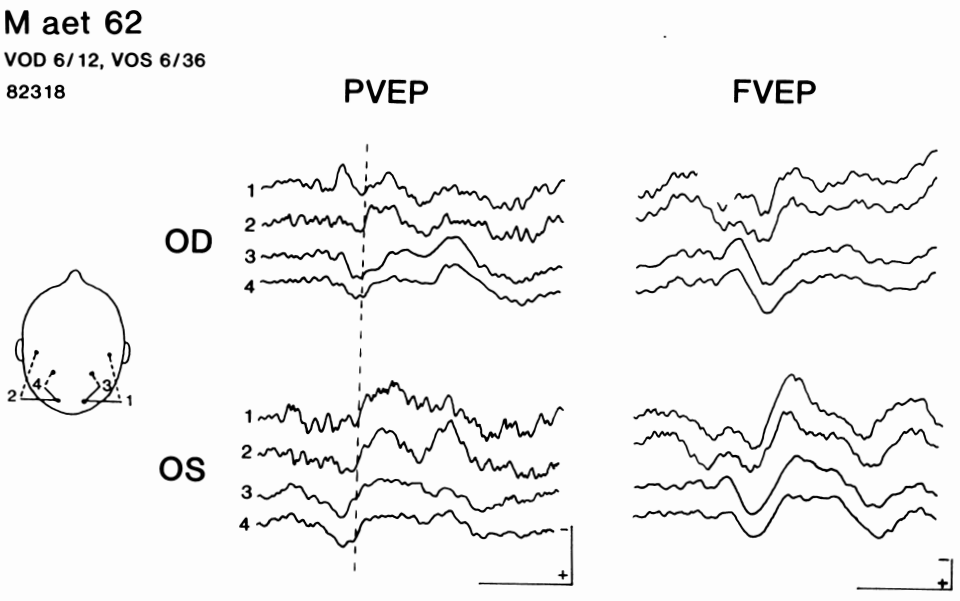

FIG 82–2.
PVEP (11-degree full field, 26-minute checks, 1.95 reversals per second) and FVEP findings in a patient with pseudo–Foster-Kennedy syndrome due to acute ION in the right eye (marked disc swelling) and an old ION in the left eye (atrophic disc). The PVEPs from both eyes are of subnormal amplitude with no definite latency change. The *dotted vertical line* represents the upper limit of normal latency (mean +3 SD) (calibration, 2.5 μV, 100 ms).

degree radius, 50-minute check stimulus and found "delays" that in some cases could be explained by complete or partial substitution of the paramacular P135 subcomponent for the usually dominant, macular-derived P100 component. It should be remembered that this interpretation can only apply with a large field (see Chapter 18). Most of their patients had single-channel recordings with central field stimulation. PVEPs were often extinguished, but delays were observed. Follow-up studies suggested that the abnormalities remained essentially unchanged.

This author[15] recently reported PERG abnormalities in seven cases, five with involvement of the P50 component and two with an abnormality confined to the N95 component. The histopathological observations[21] of macular necrosis in ION may be relevant because the PERG P50 component is reduced in macular disease.[16]

The finding of a normal-latency, reduced-ampli-

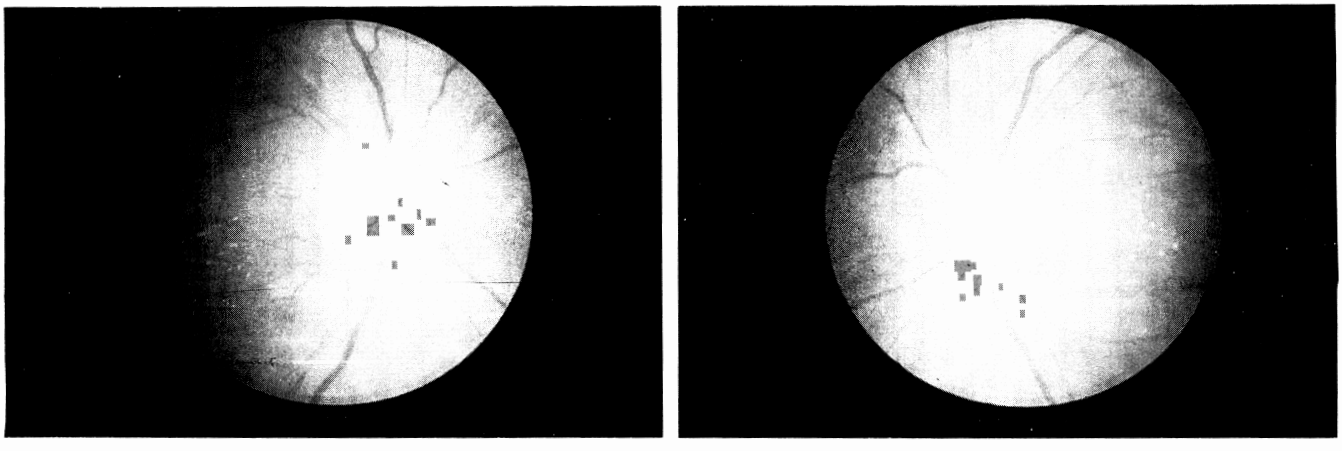

FIG 82–3.
Fundus photographs (see Fig 82–2). The PVEPs from both eyes are of subnormal amplitude with no definite latency change. **(See also Color Plate 12.)**

tude PVEP suggests ION in a patient with sudden, painless loss of vision and a swollen optic disc. Delayed PVEPs can be observed but the delays are less marked than those in demyelination. If there has been a previous episode in the fellow eye with resultant disc pallor, the appearances may be mistaken for the Foster-Kennedy syndrome (Fig 82–2, 82–3, Plate 12). An abnormal VEP is not a feature of papilledema per se, and electrophysiology should resolve any diagnostic difficulties in such cases. The findings from clinically uninvolved eyes are normal. There may be associated systemic symptoms and elevation of the blood ESR with ION and temporal arteritis.

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