Ischemic syndromes of the optic nerve (ischemic optic neuropathy [ION]) are classified according to (a) the location of the ischemic damage of the nerve and (b) the etiologic factor, if known, for the ischemia. Anterior ischemic optic neuropathy (AION) includes syndromes involving the optic nerve head, with visible optic disc edema. Posterior ischemic optic neuropathy (PION) incorporates those conditions involving the intraorbital, intracanalicular, or intracranial portions of the optic nerve, with no visible edema of the optic disc. While several specific etiologic factors have been identified in ION, the most critical for initial management is the vasculitis of giant cell, or temporal, arteritis (GCA); therefore, ION is typically classified as either arteritic (usually due to GCA) or nonarteritic. Nonarteritic ION is most often idiopathic, but specific etiologies such as systemic hypotension and radiation injury have been identified. Finally, several syndromes of optic disc edema with relatively mild dysfunction, including preinfarct optic disc edema and diabetic papillopathy, are presumed to represent optic nerve ischemic edema with dysfunction that is not detectable, mild, or reversible.

ANTERIOR ISCHEMIC OPTIC NEUROPATHY (AION)

AION typically presents with the rapid onset of painless unilateral visual loss developing over hours to days. An altitudinal visual field defect (typically inferior) is the most common pattern of loss (Fig. 7.1 but arcuate scotomas, cecocentral defects, and generalized depression are also frequently seen (Fig. 7.2). Visual acuity is decreased if the field defect involves fixation but may be normal if an arcuate pattern spares the central region. The pupil in the affected eye is sluggishly reactive, with a relative afferent pupillary defect present unless pre-existing or concurrent optic neuropathy in the fellow eye makes its pupillary response equally sluggish. The optic disc is edematous at onset; the edema may be pallid, but it is not uncommon to see disc hyperemia, particularly in the nonarteritic form (Fig. 7.3). The disc most often is diffusely swollen, but a segment of more prominent edema is frequently present (Fig. 7.4). Flame hemorrhages are commonly located adjacent to the disc, and peripapillary retinal arteriolar narrowing may occur (Fig. 7.5). Retinal arteriolar emboli are only rarely associated.

ARTERITIC ANTERIOR ISCHEMIC OPTIC NEUROPATHY (AAION)

Demographics

Although it often produces a severe and devastating optic neuropathy, GCA is the cause for AION in a relatively small minority (5.7%) (1) of cases, with an estimated annual incidence in the United States of 0.57 per 100,000 over age 60 (2). AION is, however, the most common cause of visual loss in GCA (3–5), accounting for 71–83% of cases (6,7), with retinal artery occlusion, choroidal ischemia, and PION less common. The mean age of onset for AAION is 70 years (1), with only rare occurrence under age 60. GCA occurs more frequently in women and with increasing age (8,9). It is most common in Caucasians and is unusual in African-
Figure 7.1. Automated quantitative perimetry in AION. Broad arcuate (altitudinal) defect (A) is most common, with central (B) and less severe arcuate (C) defects also frequently noted.
American and Hispanic patients (2). Liu et al. reviewed 121 consecutive temporal artery biopsies, assessing the rate of positive biopsies by ethnic background: 19/66 (29%) of Caucasian patients had positive biopsies, compared with 0/40 Hispanic patients and 0/6 African-Americans (10).

**Clinical Manifestations**

AAION usually occurs in association with other systemic symptoms of the disease. Headache is the most common symptom, while jaw claudication and temporal artery or scalp tenderness are the most specific for the disorder (11–13) (see also Chapter 44). The headache is often severe, constant, and disabling. True jaw claudication, with muscular cramping and fatigue progressing with continued chewing activity, has high specificity for temporal arteritis, although it has also been reported in amyloidosis (14). Claudication may also occur in muscles of the neck or tongue. The syndrome of polymyalgia rheumatica (PMR), including malaise, anorexia, weight loss, fever, proximal joint arthralgia and myalgia, is frequently reported. PMR with hematologic inflammatory markers, but without temporal artery or ocular involvement, may lead to arteritis in some cases. One series indicated that 15.8% of PMR patients eventually developed GCA with visual loss (15). In contrast, so-called occult GCA, without overt systemic symptoms and
sometimes without abnormal blood testing, may occur (16,17). In Hayreh’s recent report, 21.2% of patients with GCA and visual loss had no systemic symptoms of the disease (17).

In addition to systemic symptoms, certain associated local sings may aid in the diagnosis of AAION. Induration of the temporal region, decreased or absent temporal artery pulse, and cordlike firmness or nodularity of the temporal artery all may be seen. Ischemic scalp necrosis has been documented in GCA, and it may masquerade as herpes zoster dermatitis, with facial pain and even a vesicular reaction of the affected skin (18). A wide variety of additional signs relating to vasculitic ischemia of the central nervous system have been reported; these include mental status alterations (19), brain stem and cerebellar syndromes (20), and damage to the retrochiasmal afferent visual pathways, with corresponding visual field defects (20).

AAION typically presents with severe visual loss (visual acuity less than 20/200 in 57.8% to 76.5% of cases [6,7,21,22], frequently hand motion or worse) developing rapidly over hours to days (Table 7.1). Twenty-one percent of cases in one series had no light perception (6). While the initial presentation is often unilateral, bilateral simultaneous AAION is more commonly arteritic than nonarteritic, and its occurrence raises suspicion for GCA. The persistent visual loss of AAION is preceded in 7–18% (6,7,20,23) of cases by transient visual loss similar to that of carotid artery disease, although the episodes may be of shorter duration; these are only rarely associated with the nonarteritic form of AAION and are highly suggestive of arteritis. The pallid type of optic disc edema (Fig. 7.6) is seen more frequently in the arteritic than in the nonarteritic form (24,25); chalk-white pallor with edema is seen in severe cases and is highly unusual in nonarteritic ION (NAION). Cotton-wool patches (Fig. 7.7) indicative of concurrent retinal ischemia may be seen (26). Retinal

Table 7.1
Ischemic Optic Neuropathies

<table>
<thead>
<tr>
<th>Age</th>
<th>Arteritic</th>
<th>Nonarteritic</th>
<th>Diabetic Papillopathy</th>
<th>Posterior</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean 70 yr</td>
<td>Mean 60 yr</td>
<td>Variable (&lt;50)</td>
<td>Variable</td>
<td>Variable</td>
</tr>
<tr>
<td>F &gt; M</td>
<td>F = M</td>
<td>F = M</td>
<td>F = M</td>
<td>F = M</td>
</tr>
<tr>
<td>Associated symptoms</td>
<td>Usually none</td>
<td>Usually none</td>
<td>Usually none</td>
<td>None unless arteritic or postoperative</td>
</tr>
<tr>
<td>Headache, jaw claudication, transient visual loss</td>
<td></td>
<td></td>
<td></td>
<td>Usually poor</td>
</tr>
<tr>
<td>Visual acuity</td>
<td>&lt;20/200 in &gt;60%</td>
<td>&gt;20/60 in &gt;50%</td>
<td>&gt;20/40 in &gt;75%</td>
<td>Usually poor</td>
</tr>
<tr>
<td>Disc</td>
<td>Pale swelling common</td>
<td>Pale or hyperemic</td>
<td>Hyperemia, telangiectasia</td>
<td>Usually poor</td>
</tr>
<tr>
<td>Cup normal + choroid ischemia</td>
<td>Cup small</td>
<td>Cup small</td>
<td>Cup small</td>
<td>Usually poor</td>
</tr>
<tr>
<td>ESR</td>
<td>Mean 70 mm/hr</td>
<td>Usually normal</td>
<td>Usually normal</td>
<td>Normal</td>
</tr>
<tr>
<td>FA</td>
<td>Disc delay</td>
<td>Disc delay</td>
<td>Disc delay</td>
<td>Variable</td>
</tr>
<tr>
<td>Choroid delay</td>
<td></td>
<td></td>
<td></td>
<td>Not studied</td>
</tr>
<tr>
<td>Natural history</td>
<td>Rarely improve</td>
<td>16–42.7% improve</td>
<td>Resolves 2–10 mo</td>
<td>Rarely improves</td>
</tr>
<tr>
<td>Fellow eye 54–95%</td>
<td>Fellow eye 12–19%</td>
<td>Bilateral 40%</td>
<td>Bilateral &gt;60%</td>
<td>Bilateral &gt;60%</td>
</tr>
<tr>
<td>Treatment</td>
<td>Systemic steroids</td>
<td>None proven</td>
<td>None proven</td>
<td>Steroids if arteritic</td>
</tr>
</tbody>
</table>
Figure 7.6. Bilateral simultaneous anterior ischemic optic neuropathy in a patient with GCA and no light perception in either eye. The right and left optic discs show markedly pale swelling.

Figure 7.7. Progressive optic atrophy and pseudoglaucomatous cupping after an attack of AAION. A. Right optic disc at the time of initial visual loss. Note pale optic disc swelling, peripapillary hemorrhage, and cotton-wool spot. B. Eight weeks after visual loss, the optic disc swelling has resolved, and both the hemorrhage and the cotton-wool spot are resolving. Note narrowing of retinal vessels. C. Four months after visual loss, the optic disc is pale and now has a large cup. The retinal vessels, especially the arteries, are markedly narrowed.
arterial occlusion may occur simultaneously with the optic neuropathy (6). In Hayreh’s series (23), cilioretinal artery occlusion occurred in 21.2%; this is a very unusual occurrence in NAION and is highly suggestive of GCA (Fig. 7.8). Peripapillary choroidal ischemia may be associated with the optic neuropathy, producing edema deep to the retina adjacent to the optic disc and exacerbating the visual loss (27,28). It may also occur in a more widespread area, with or without optic disc involvement (27,29). The optic disc of the fellow eye in AAION most frequently is of normal diameter, with a normal physiologic cup (30,31), as opposed to that in NAION, which tends to be small in diameter, with little or no physiologic cup (31) (see below).

**Pathophysiology**

Histopathologic studies in AAION demonstrate vasculitis of the short posterior ciliary vessels supplying the optic nerve head, in addition to variable involvement of the superficial temporal, ophthalmic, choroidal, and central retinal arteries (32–37). Infiltration of the short posterior ciliary arteries by chronic inflammatory cells, with segmental occlusion of multiple vessels by inflammatory thickening and thrombus (Fig. 7.9), has been documented (32,33,37). Autopsy studies have demonstrated ischemic necrosis predominantly involving the laminar and retrolaminar portions of the optic nerve supplied by these vessels and their distal tributaries (32,34,35).

Fluorescein angiographic data support involvement of the short posterior ciliary arteries in AAION. Delayed filling of the optic disc and adjacent choroid (Fig. 7.10) is consistently noted (24,28,38–41). Extremely poor or absent filling of the choroid has been suggested as one useful factor in distinguishing arteritic from nonarteritic AION. Mack et al. (29) reported mean choroid perfusion time (from first appearance to filling) of 69 seconds in patients with AAION, compared with 5.5 seconds in NAION; Siatkowski et al. (25) confirmed this finding with slightly different parameters, mean time from injection to choroid filling measuring 29.7 seconds in AAION compared with 12.9 seconds in NAION.

**Differential Diagnosis**

The acute onset of severe visual loss in the setting of headache and optic disc edema, particularly when bilateral,
requires consideration of alternate diagnoses, including acute optic neuropathy secondary to chronic papilledema (with or without intracranial mass), infiltrative optic neuropathy, and meningeal carcinomatosis involving the optic nerves. In cases of suspected AAION with negative workup or atypical course, neuroimaging should be performed to evaluate for intracranial mass or visible meningeal thickening and enhancement, and lumbar puncture should be considered to assess for evidence of elevated intracranial pressure or malignant cells.

**Clinical Course**

If untreated, AAION results in severe damage of the affected optic nerve. Recovery of useful vision after initial involvement is unusual, even with prompt therapy. In cases with unilateral presentation, estimates for development of second eye involvement range from 54% to 95% (6,42–44). Time to second eye involvement varies greatly, but it may occur within hours to days. The optic disc edema typically resolves over 4–8 weeks, with resultant optic atrophy and generalized attenuation of retinal arterioles. Excavation of the optic disc (Fig. 7.7) occurs frequently after AAION (40,45–49) but is unusual in NAION. Danesh-Meyer et al. (48) reported optic disc cupping in 92% of 92 patients with AAION versus 2% of 102 patients with NAION. The excavation may be severe, mimicking glaucomatous damage. Hayreh and Jonas (49) noted lack of increase in peripapillary atrophy associated with the cupping after AAION, in contrast to glaucoma. The severity of the optic atrophy seen after AAION also differentiates it from glaucomatous excavation.

**Diagnostic Confirmation**

The most important initial step in the management of AION is the assessment for evidence of GCA. A tentative diagnosis may be made on the basis of advanced age and typical clinical symptoms in conjunction with elevation of the erythrocyte sedimentation rate (ESR). Most cases of active GCA show markedly elevated ESRs (mean 70 mm/hr, often above 100 mm/hr). When the level is not extremely high, however, interpretation of the value becomes more difficult, as normative data are imprecise. As a rule, we recommend the clinically useful guideline that the upper limit of normal, in mm/hr, is calculated by dividing patient age by 2 in males and patient age plus 10 by 2 in females (50). However, by these criteria, the level may be normal in up to 22% of patients with GCA (5,51). Conversely, the ESR rises with age, and levels above the listed upper limit of normal for the laboratory are common (up to 40% over 60 mm/hr) in patients over 70 without arteritis (52). In the Ischemic Optic Neuropathy Decompression Trial (IONDT), 9% of patients with NAION had ESR levels over 40 mm/hr, with a range of 0–115 (53). Moreover, the test is nonspecific, elevation confirming only the presence of any active inflammatory process or other disorder affecting red cell aggregation. In studies of temporal artery biopsy-negative cases with such elevation, the most common associated diseases have been occult malignancy (most frequently lymphoma) in 18–22%, other inflammatory disease in 17–21%, and diabetes in 15–20% (54,55). In these cases initially suspected of being GCA, internal medicine consultation to rule out other systemic diseases is essential.

Additional blood abnormalities are common in GCA and may have prognostic value. Measurement of C-reactive protein, levels of which do not rise with age or anemia, may increase diagnostic accuracy and are currently recommended in conjunction with the ESR (56). Hayreh et al. (69) reported that a specificity of 97% for GCA was attained for AION patients with ESR levels above 47 mm/hr and C-reactive protein above 2.45 mg/dL (normal less than 0.5) from their laboratory. We currently recommend simultaneous measurement of both parameters in suspected cases of AAION. Fibrinogen is commonly elevated and may supplement C-reactive protein levels in increasing accuracy over the use of ESR alone (57,58). Thrombocytosis is seen in up to 50% of patients with GCA; its presence has been shown to be a marker for positive temporal artery biopsy (59) and may be a predictor of visual loss (60,61). Liozon et al. (61) assessed 174 patients with GCA, correlating thrombocytosis with the development of permanent visual loss. Of 20 patients with permanent visual loss due to AAION, 13 (65%) had thrombocytosis. This feature may have implications for therapy (see below).

Giant cell arteritis is confirmed by a positive temporal artery biopsy, which is strongly recommended in any case of suspected arteritic AION. The certainty of biopsy-proven GCA provides support for long-term systemic corticosteroid therapy, which often is required for up to a year and may be associated with severe systemic complications. It also makes later decisions regarding the risk/benefit ratio of prolonged therapy more clearcut. A negative biopsy, however, does not rule out GCA. A false-negative biopsy may result from (a) discontinuous arterial involvement (“skip lesions”) (62–65), undetected in 4–5% due to insufficient

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**Figure 7.10.** Fluorescein angiography in AAION. The optic disc and adjacent choroid show marked filling delay.
length (minimum 3–6 cm recommended) of specimen or insufficiently detailed step-sectioning; (b) unilateral involvement with biopsy of the uninvolved temporal artery; (c) improper handling of specimens; or (d) review by a pathologist inexperienced in the diagnosis of acute and healed arteritis.

The false-negative error rate for unilateral temporal artery biopsy has previously been estimated at 5–11% (66–68). Hayreh et al. (69) more recently prospectively studied 76 patients who underwent contralateral biopsy due to high clinical suspicion after initially negative biopsy; 7 (9.2%) had evidence of active inflammation in the second biopsy. Boyev et al. (70) reviewed bilateral biopsy results for concordance in 186 cases of suspected GCA. Identical diagnoses were obtained in 176, an overall concordance rate of 97%. Six of the remaining 10 cases had useful biopsy results from each side, all demonstrating discordance (3%); 5 of the 6 were eventually diagnosed with GCA based on clinical evidence. Most (150) of the cases involved bilateral simultaneous biopsies, but 36 were sequential, more directly addressing the issue of the value of a second (contralateral) biopsy after an initially negative one; of these, only 1 (2.8%) showed discordance. Danesh-Meyer et al. (71) performed a similar retrospective study of 91 bilateral biopsies, the majority simultaneous, finding concordance in 90 (99%); it is unclear whether the discordant case involved sequential biopsies. They performed a meta-analysis of previous reports that yielded an overall discordance rate of 4%. Pless et al. (72) reviewed 60 bilateral biopsies, 31 of which were simultaneous, finding discordance in 13.4% overall but only 5% in sequential cases in which the initial result was negative. Hall et al. (73) found a similar rate.

While the overall discordance rate for bilateral biopsies is low (1–3% in the largest series), there is substantial variation, even in major academic institutions with experienced pathologists, with rates as high as 13.4%. In the critical subset of patients who undergo contralateral biopsy after an initially negative result (a sample biased toward the positive based on clinical parameters raising suspicion for GCA), studies with the greatest number of patients suggest a slightly higher discordance (2.8–9.2%), but the difference is small. The data all suggest some degree of increased accuracy from bilateral biopsies, and considering the severe consequences of missed diagnosis, the relatively low risk of procedural complications, and the benefit of biopsy proof in the long-term management of these patients, we consider bilateral biopsy in all cases. Expert opinion varies, however, as to preference for bilateral simultaneous biopsies versus immediate sequential biopsies (with negative initial frozen-section analysis) versus delayed sequential biopsies (with negative initial permanent section analysis) if clinical suspicion for GCA is high.

**Therapy**

If GCA is suspected, therapy should be instituted immediately, as patients are at high risk for further visual loss in the affected eye, involvement of the fellow eye, and systemic complications of vasculitis, including stroke and myocardial infarction. Initial treatment should not await diagnostic confirmation by temporal artery biopsy. Although chronic systemic corticosteroid therapy has been reported to reduce positive biopsy results in GCA from 60% to 20% (74), delay of 7–10 days has no significant effect on the results, and in some cases active inflammation may be detected after longer therapy (75). High-dose intravenous methylprednisolone at 1 g/day for the first 3–5 days is most often recommended, particularly when the patient is seen in the acute phase, since this mode of therapy produces higher blood levels of medication more rapidly than oral therapy (76,77). As these patients are often elderly with multiple and complex medical problems, we routinely provide inpatient therapy under the supervision of an internist.

Liu et al. (6) reported improvement of visual loss in AAION in 39% of those treated initially with intravenous therapy, compared with 28% in those treated orally alone. Hayreh et al. (78) similarly suggested a better visual outcome with intravenous administration, although Cornblath et al. (79) have raised questions regarding the advantages of this mode of therapy. Oral prednisone at doses of at least 1 mg/kg/day is recommended after intravenous therapy (or initially if the intravenous route is not used) and is tapered slowly, monitoring for control of systemic symptoms and ESR level; therapy is usually maintained for at least 4–6 months, often up to a year. Systemic symptoms typically subside within a week, a response so characteristic that if it is absent, an alternate disease process should be strongly considered. Alternate-day corticosteroid therapy is inadequate for GCA. Hunder et al. (66) compared alternate-day to daily therapy in GCA, finding that both systemic symptoms and ESR were incompletely suppressed on alternate-day therapy; one patient in this treatment group suffered recurrent transient visual loss, while none in the daily groups had similar symptoms.

Some degree of visual recovery in the affected eye may be obtained on therapy but is not generally anticipated. Reports of visual improvement on corticosteroids vary widely with regard to delay to therapy, dosage, and parameters for visual recovery. Aiello et al. (7) reported improvement of vision in 5 of 34 patients (15%), while Liu et al. (6) noted it in 14 of 41 (34%). Foroozan et al. (80) reported improvement in visual acuity in 5 of 39 eyes (13%), although visual fields remained severely constricted. Hayreh et al. (78) studied 114 eyes in 84 patients for evidence of visual improvement, finding only 5 eyes (4%) with both improved visual acuity and central visual field after therapy.

The major goal of therapy, other than prevention of systemic vascular complications, is to prevent contralateral visual loss. Untreated, fellow eye involvement occurs in 50–95% (42,81), often within weeks. With therapy, Aiello et al. (7) found that 2/24 (6.3%) patients with AAION developed such involvement, at 4 days and 1 month after initiation of therapy; Liu et al. (6) detected fellow-eye AAION on therapy in 3 cases. While corticosteroid therapy reduces the risk of further visual loss, it is not uniformly effective (82); breakthrough involvement of an affected eye while on therapy occurred in 9–17% (6,7). Liu et al. (6) found six cases developing AAION while on systemic corticosteroid therapy for systemic symptoms alone, without previous visual loss. The risk of recurrent or contralateral optic nerve involvement with tapering of medications has been reported at 7% (6).
Thrombocytosis in GCA and its possible predisposition to visual loss suggests the possibility that antplatelet therapy may be of benefit in conjunction with corticosteroids. Nesher et al. (83) reviewed 175 cases with GCA for evidence of cranial ischemic complications (predominantly AION and stroke), comparing those with and without low-dose aspirin therapy for other conditions. At presentation, 3 (8%) of the aspirin-treated cases had cranial ischemic complications, compared with 20 (29%) of the non-aspirin-treated cases ($P = 0.01$). During follow-up of at least 3 months on steroid therapy, cranial ischemic complications developed in 3% of the aspirin-treated cases versus 13% on steroids alone ($P = 0.02$). Further study may confirm a benefit for low-dose aspirin or other antplatelet therapy in GCA.

Other Vasculitides
Arteritic conditions other than GCA may cause AION, including herpes zoster (84,85), relapsing polychondritis (86,87), rheumatoid arthritis (88,89), Takayasu’s arteritis (90), and connective tissue disorders such as periarteritis nodosa, systemic lupus erythematosus (91,92), and allergic granulomatosis (Churg-Strauss angiitis) (93,94). The clinical picture is typically that of sudden unilateral or bilateral visual loss, often central, and sometimes accompanied by pain, with minimal recovery. Fibrinoid necrosis of the arterioles and secondary myelin and axonal loss has been demonstrated in multiple cases (91). Crompton et al. (89) documented necrotizing lymphocytic vasculitis and pervasculitis affecting the posterior ciliary arteries, in a case of AION in rheumatoid arthritis.

Goldstein and Wexler (95) reported the histopathologic findings in a patient with periarteritis nodosa who had sudden bilateral visual loss and pale optic discs. They described mononuclear cell infiltration of the posterior ciliary arteries, necrosis and infiltration of some choroidal arteries, and infiltration and atrophy of the optic nerves. Although the patient was not examined at the time of visual loss, it is likely that the process was a bilateral simultaneous AION. AION has also been described in two patients with Behçet’s disease (96) and in a patient with Crohn’s disease (97), presumably from posterior ciliary arteritis.

The term “autoimmune optic neuropathy” (98,99) has been used to describe patients with nonspecific symptoms of connective tissue disease, frequently insufficient to confirm a specific diagnosis such as lupus, who develop acute optic neuropathy, either anterior or posterior (retrobulbar). The syndrome is atypical for optic neuritis in that pain is not a consistent feature, and visual recovery is variable. Steroid dependence, with improvement on therapy but recurrence on tapering, is common. Immunosuppressive therapy may be required. Histopathologic studies have not been documented, but it has been postulated that vasculitis of the ciliary arteries results in optic nerve ischemia in these cases.

NONARTERITIC ANTERIOR ISCHEMIC OPTIC NEUROPATHY (NAION)

Demographics
Most (94.7%) cases of AION are nonarteritic (1) NAION is the most common acute optic neuropathy in patients over 50 years of age, with an estimated annual incidence in the United States of 2.3–10.2 per 100,000 population (2,100), accounting for at least 6,000 new cases annually. The prevalence of NAION in the Medicare Database has been reported at 0.30% (101). The disease occurs with significantly higher frequency in the white population than in black or Hispanic individuals (2). There is no gender predisposition (1,21,22,53,102). The mean age at onset in most studies ranges from 57–65 years (1,21,22,102). In the IONDT, mean age was 66 years, probably somewhat biased by the exclusion criterion for patients under 50 (53).

Clinical Presentation
NAION presents with loss of vision occurring over hours to days, often described as blurring, dimness, or cloudiness in the affected region of the visual field, most often inferiorly. Although Hayreh et al. (103) reported that visual loss was most frequently reported upon awakening, this feature was not confirmed in the IONDT (53). NAION typically presents without pain, although some form of pericentral discomfort has been reported in 8–12% (21,104,105); in the IONDT, 10% reported minor ocular discomfort (53). In contrast to patients with optic neuritis, those with NAION usually do not report pain with eye movement. Headache and other symptoms associated with GCA are absent. Episodes of transient visual loss as seen in GCA are rare, but vague intermittent symptoms of blurring, shadows, or spots were reported in 5% of patients in the IONDT (53). The initial course may be static, with little or no fluctuation of visual level after initial loss, or progressive, with either episodic, stepwise decrements or steady decline of vision over weeks prior to eventual stabilization. The progressive form has been reported in 22–37% of nonarteritic cases (21,104,106,107), 29% in a limited group of patients with visual acuity better than 20/64 in the IONDT (53).

NAION usually presents with less severe visual loss than AION, with visual acuity better than 20/200 in 58–61.2% of patients (21,22,104) (Table 7.1). In the IONDT, 49% had initial visual acuity of at least 20/64, 66% better than 20/200 (53). Color vision loss in NAION tends to parallel visual acuity loss, as opposed to that in optic neuritis, in which color loss is often disproportionately greater than visual acuity loss. Visual field defects in NAION may follow any pattern related to optic nerve damage, but altitudinal loss, usually inferior, occurs in the majority, ranging from 55–80% of reported cases (21,22,104,108,109).

The optic disc edema in NAION may be diffuse or segmental, hyperemic or pale, but pallor occurs less frequently than in the arteritic form. A focal region of more severe swelling is often seen and may display an altitudinal distribution, but it does not consistently correlate with the sector of visual field loss (110). In the IONDT, 25% of patients demonstrated sectoral disc edema (53). Diffuse or focal telangiectasia of the edematous disc (Fig. 7.11) may be present, occasionally prominent enough to resemble a vascular mass (pseudoangiomatoma) (111). Peripapillary retinal hemorrhages are common, seen in 72% in the IONDT (53). Retinal exudates are unusual, but both soft and hard exudates may