# Bilateral Non-Arteritic Anterior Ischemic Optic Neuropathy in a Young Male

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**Summary**

*A 43 year old diabetic male presented with complaints of blurring of peripheral vision, in the left more than right eye, since a couple of months. Best corrected visual acuity was 6/9 in the right eye and 6/24 in the left eye. Dilated fundus examination showed a slightly edematous disc superiorly with a cup:disc ratio of 0.1:1 in the right eye and nasal disc pallor with a cup:disc ratio of 0.1:1 in the left eye. Visual fields examination was suggestive of a partial inferior altitudinal defect in the left eye. OCT showed nerve fibre layer thinning and cup:disc ratio of 0.14:1 in the left eye. A diagnosis of bilateral NAION was made. Serum cholesterol levels were raised. VEP was advised which was suggestive of bilateral axonopathic optic neuropathy. The patient was started on a combination of Syndopa-Carbidopa, Aspirin and Atorvastatin. On follow-up, visual acuity improved to 6/12 in the left eye. Field defect was reduced slightly compared to the previous examination. The pathogenesis of NAION remains unclear but may involve ischemia to the optic nerve head from insufficient perfusion of the short posterior ciliary arteries. The most important structural risk factor is a small and crowded disc. The presentation in young patients is similar to elderly patients having classical AION. Prompt diagnosis and management gives a better chance of visual improvement in younger patients.*

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## Introduction

Anterior ischaemic optic neuropathy (AION) is caused by diminished blood supply to the optic disc and anterior portion of the optic nerve by the posterior ciliary arteries. Sudden vision loss in elderly patients with vasculopathic risk factors is typical of this condition. AION may be arteritic (AAION) and nonarteritic (NAION) in origin. AAION occurs in patients with temporal arteritis (giant cell arteritis). Risk factors for NAION are diabetes and/or hypertension and particularly a "disc at risk" i.e., a small, crowded optic nerve head. Bilateral simultaneous nonarteritic anterior ischaemic optic neuropathy (NAION) is extremely rare. There is no known effective therapy for NAION but a number of medical and surgical therapies have been proposed.

## Case Report

A 43 year old male presented to our centre with chief complaints of blurring of vision in the left eye more than the right eye since two months which was gradual in onset, progressive and painless in nature. The blurring of vision was peripheral and more towards the temporal side. There were no aggravating or relieving factors. The patient gave no history of using spectacles. He was a known diabetic since five years, on regular medication. There was no history of any other systemic illness. On examination, the visual acuity (unaided) was 6/9 and 6/24 in the right and left eye respectively. Colour vision was normal. Anterior segment examination was unremarkable. Pupils were bilaterally equal, reacting to light. RAPD was absent. Intraocular pressure on Perkin's applanation tonometry was 10 and 12 mm Hg respectively.

Dilated fundus examination showed a slightly edematous hyperemic disc superiorly with a cup:disc ratio of 0.1:1 in the right eye (Figure 1) and nasal disc pallor with a cup:disc ratio of 0.1:1 in the left eye (Figure 2). There were no peripapillary

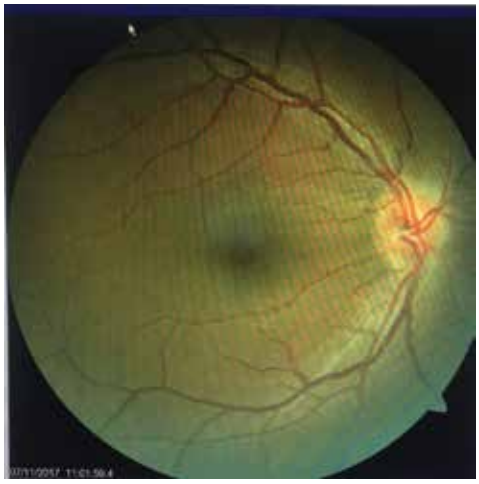

**Figure 1:** *Fundus of the Right Eye*

**Figure 2:** *Fundus of the Left Eye*

hemorrhages. There was no evidence of diabetic retinopathy changes. Visual field examination was suggestive of a partial inferior altitudinal defect in the left eye. Optical Coherence Tomography was done, which showed nerve fibre layer thinning and cup:disc ratio of 0.14:1 in the left eye. The nerve fibre layer thickness was normal in the right eye with a cup:disc ratio of <0.1:1. A diagnosis of bilateral NAION was made. Blood tests including blood sugar levels, serum homocysteine, ESR and C-reactive protein were normal. Serum cholesterol levels were raised (238 mg/dl). VEP was advised, which was suggestive of bilateral axonopathic optic neuropathy, more in the left eye. The patient was started on a combination of oral Syndopa-Carbidopa (110 mg once daily), Aspirin (150 mg once daily) and Atorvastatin (20 mg once daily). He was advised to keep strict blood sugar control and was counseled to maintain a healthy diet and moderate exercise routine. On subsequent follow-ups, visual acuity gradually improved to 6/12 in the left eye by the end of one month. Superior optic disc edema in the right eye resolved. However, field defect was only slightly reduced compared to the previous examination.

## Discussion

Nonarteritic anterior ischemic optic neuropathy (NAION) most frequently occurs in patients aged 50 years or older.[1] It can cause sudden visual loss and visual field defects. It is often associated with various systemic disorders such as diabetes, atherosclerosis and systemic connective tissue diseases. The disorder has been characterized by the following distinctive features:[2]

1. Acute or subacute onset of visual loss, usually monocular at onset, involving the inferior visual field (altitudinal loss) most commonly, but any pattern of field loss consistent with optic nerve origin may occur
2. Single episode of visual loss in the majority, with a minority progressively worsening over weeks before stabilizing
3. Absence of pain or mild nonspecific pain (without eye movement) associated
4. Occurrence in "crowded discs" (small diameter and small cup-disc ratio as measured in the fellow eye)
5. Optic disc edema at onset and probably prior to onset of visual field loss
6. Diffuse or segmental pattern of either superior or inferior localized edema, sometimes with a sharply demarcated horizontal linear border
7. Occasional prominent optic disc surface vascular dilation
8. Superimposed development of optic disc pallor over weeks, with the appearance of "pale swelling" replacing hyperemic edema, later evolving into optic atrophy
9. Optic atrophy, which may be diffuse or may show a sharply segmental or altitudinal configuration
10. Visual loss, which is usually permanent and stable, but with a substantial spontaneous improvement rate for visual acuity (42.7% improved by ≥3 lines Snellen acuity at 6 months in the Ischemic Optic Neuropathy Decompression Trial (IONDT)
11. Unusual recurrence in the same eye
12. Fellow eye involvement in a minority

Although it is traditionally thought to be a condition of patients older than the age of 50, NAION has increasingly been recognized in patients younger than age 50 in whom hypercholesterolemia has been identified as a risk factor.[3]

In AION, there is insufficient blood circulation of the posterior ciliary arteries in the prelaminar, laminar, and retrolaminar regions of the optic nerve.[4] In younger patients with NAION, it is essential to rule out optic nerve inflammation, either idiopathic demyelination or other causes, such as vasculitis or infection. Crowded disc (small diameter with a cup-disc ratio <0.2) i.e., optic discs lacking biological cups, has been identified as the most important underlying risk factor for developing AION.[5]

Optic disc ischemia in patients under 50 years of age encompasses a spectrum ranging from optic disc edema without optic nerve dysfunction to overt infarction (NAION) with permanent visual field loss; the ischemia may be identified in all forms by the characteristic optic disc early filling delay seen on FA. Diabetes is the single factor, consistently identified in all age-groups, and associated with a younger age of onset in the overall NAION population.

Our patient was a known diabetic who developed bilateral NAION at a young age and showed significant improvement in visual acuity on treatment but did not show much improvement in visual fields.

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