Temporal visual field defects associated with nasal hypoplasia of the optic disc

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SUMMARY  Unusual stationary temporal visual field defects breaking out from the blind spot are described as they were noted in 3 patients. The nature of these field defects and the associated funduscopic findings indicate a developmental defect localised to the nasal sector of the optic disc, which we suggest be termed nasal hypoplasia of the optic disc.

Temporal, wedge-shaped visual field defects breaking out horizontally from the blind spot are a distinct perimetric rarity. This report describes such field defects occurring bilaterally in 2 patients and unilaterally in 1, with associated ophthalmoscopic signs of optic disc hypoplasia.

Case reports

CASE 1
The patient was a 40-year-old woman who consulted her ophthalmologist because of headaches. She was totally unaware of any defects in her vision. Her corrected acuity in each eye was 6/6 with -4.00 dioptre spherical lenses. Both her colour vision and her pupillary reactions to direct light stimulation were normal. An examination of the visual fields revealed symmetrical, absolute, steep-edged, wedge-shaped defects that broke out temporally from the blind spot bilaterally (Fig. 1). The optic discs were normal in colour and size. The retinal vessels emerged on the nasal side of each disc. Branches of arterioles appeared small in calibre and were sparse in number in the nasal sector of the retina. The peripapillary nerve fibre layer appeared normal in all but the nasal sector, where it was not visible. The nasal disc margins appeared abnormally sharp. The optic fundi were normal in all other respects (Fig. 2).

The intraocular pressures were normal. A general neurological examination revealed no signs of neurological deficit, and a computerised tomographic scan

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showed normal orbital and intracranial findings. Her headaches were attributed to muscle tension. A visual field examination 18 months later resulted in identical findings.

CASE 2
The patient, a 42-year-old woman, was examined by her ophthalmologist because of pain and a feeling of tightness in her right eye. She was unaware of any visual defect. Her visual acuity corrected to 6/6 in each eye with −5⋅00 dioptré spherical lenses. Her colour vision, pupillary reactions to direct light stimulation, and intraocular pressures were normal. A visual field examination revealed symmetrical, wedge-shaped, inferior temporal quadrant defects that broke out temporally from each blind spot (Fig. 3). Her optic discs were small (Fig. 4); both were normal in colour and were surrounded temporally by a pigment crescent. The retinal vessels appeared normal. The peripapillary nerve fibre layer appeared normal in all but the nasal sector, where it was not visible (Figs. 4 and 5). The optic fundi were otherwise normal. All findings from the neurological examination were normal. X-ray films of the skull and sella turcica views showed no abnormality. The discomfort round her right eye resolved spontaneously. A visual field examination performed 5 months later showed identical temporal defects.

Fig. 2 Case 1. Red-free photographs showing absence of nerve fibre striations in the nasal peripapillary retina, and a reduced amount of tissue in the nasal sectors of each disc.

Fig. 3 Case 2. Bitemporal wedge field defects.
Fig. 4  Case 2. Red-free photographs showing small optic discs.

Fig. 5  Case 2. A magnified view of the left disc showing the absence of nerve fibre striations in the nasal peripapillary retina (arrow).
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CASE 3
The patient was a 47-year-old man who consulted his ophthalmologist because of blurred vision when reading. He was completely unaware of any visual field defect. His visual acuity without correction was 6/6 in each eye; his near vision improved to N5 with +2.00 dioptre spherical lenses. His colour vision, pupillary reactions to direct light stimulation, and intraocular pressures were normal. The examination of his visual fields revealed an absolute, steep-edged, wedge-shaped defect that broke out temporally from the blind spot in the field of the right eye. The visual field on the left was normal (Fig. 6). The right optic disc was normal in colour, but its nasal margin was abnormally sharp and indented. The tissue of the disc nasally was slightly pale and reduced in amount. Branch arterioles in the nasal sector of the retina were small in calibre and decreased in number. The peripapillary nerve fibre layer was not visible from 2 to 4 o'clock (Fig. 7). The optic disc and retina of the left eye appeared normal. No signs of neurological deficit were evident from the general neurological examination. X-ray films of the skull, views of the optic foramina, and tomograms of the optic canals showed no abnormality.

Discussion
The unusual wedge-shaped, temporal field defects described in this report are the perimetric expression
of the absence of nerve fibres in the nasal sector of the retina and optic disc. In each of the involved eyes the nasal retina appeared normal except for the absence of superficial nerve fibre layer striations in the area corresponding to the field defect.

The evidence suggesting that the nasal nerve fibre deficit in these cases was developmental is: (1) the remarkable bilateral symmetry of the wedge-shaped temporal field defects in 2 patients (cases 1 and 2); (2) the small size of the discs in 1 patient (case 2); (3) the absence of sector-shaped nasal pallor in both eyes in 2 patients (cases 1 and 2); (4) the reduced amount of nasal disc tissue in the involved eyes of all 3 patients; (5) the indented nasal disc margin in 1 of the eyes of 1 patient (case 3); (6) the reduced vascularity of the affected area of nasal retina in 1 eye of 1 patient and both eyes of another (cases 1 and 3).

We believe that use of the term nasal hypoplasia of the optic disc to describe the defect illustrated by these cases is justified, if optic hypoplasia is defined as a subnormal number of optic nerve axons within the affected optic nerve, as has been suggested by Frisen and Holmegaard. Why the nasal portions of these discs are ‘hypoplastic’ is a mystery.

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