who had clear lenses and clear media. All sources of artifact such as pupil size, refraction, etc., were carefully controlled so that errors of this type did not enter. The effect on the upper field is unlikely to be a lid effect since the test is done on a circular area and is only 15° from fixation. It is evident from these findings that, with the decline in sensitivity in the Bjerrum area, particularly superiorly, baring of the blind spot may occur with physiological aging.

The patient with ocular hypertension shows reduced sensitivity in the Bjerrum area in comparison to normal subjects of his age group. This is particularly notable in the upper field. This result agrees with the observed effects of artificially raised intraocular pressure on the visual fields, and leads to the hypothesis that patients with ocular hypertension show field changes of aging in advance of their chronological age. The accelerated aging change could result in blind spot baring at an earlier age than it may occur physiologically.

There was no increase in scotoma detection in the Bjerrum area with this technique.

The value of this screening technique in the diagnosis of glaucoma will become apparent only with sequential testing of this group of suspects. One will expect to see the normal aging pattern develop and the normal versus pathological rates for this will have to be established. Specific changes due to the glaucomatous process will be expected to be superimposed on the physiological ones. Currently decisions on individuals cannot be made until standards are established and more experience is obtained.

The author would like to thank Dr. John Milligan of the Department of Physiology, Queens University, for his advice and help with the computer analysis of the data, Mrs. Carol Reifel for providing the invaluable perimetry, and Mrs. Marilyn Pyne for secretarial services.

REFERENCE

Differential diagnosis of the arcuate scotoma

David O. Harrington

Any discussion of the etiology and differential diagnosis of the arcuate scotoma or nerve fiber bundle defect in the visual field must start with the assumption that this is the typical visual field defect of established open angle glaucoma with optic nerve damage. It must also be recognized that the scotoma, as seen in glaucomatous eyes, can assume many forms. These variations in morphology, however, retain certain characteristics which make them recognizable as nerve fiber bundle defects long after they have advanced beyond the scotomatous stage to involve large areas of the peripheral and central visual field. These characteristics have been enumerated in articles and textbooks of perimetry and need not be discussed here.

It should be pointed out that the pres-
ence of an arcuate scotoma in the visual field is presumptive evidence of glaucomatous nerve damage until proved otherwise. But such a scotoma must be associated with other evidence of glaucoma and is not of itself diagnostic of the disease any more than the isolated finding of elevated intraocular pressure will establish such a diagnosis.

It may be very difficult to prove that a well-formed arcuate scotoma is not due to glaucoma. The not infrequent diagnosis of "low tension glaucoma" attests to this problem.

Before discussing those conditions which may give rise to an arcuate scotoma and which are definitely not glaucoma, this problem of "low tension glaucoma" should receive attention.

It has been established that artificial elevation of intraocular pressure by means of dynamometry and steroid-induced glaucoma will produce defects in the Bjerrum area of the visual field in selected and susceptible individuals. Such persons do not have glaucoma when judged by the usual diagnostic criteria but the transient scotoma resulting from such an elevation of intraocular pressure has all the characteristics of the true glaucomatous field defect.

Fig. 1 illustrates such a visual field defect produced by artificially raising the intraocular pressure from a pretest level of 19 to 44 mm. Hg for an interval of four minutes. This patient's pretest visual field was normal in the same isopters, optic discs were minimally cupped, retinal diastolic pulse was obtained with 50 grams of pressure, and the facility of outflow was 0.22. The visual field defect occupied the superior Bjerrum area and arched over fixation into a nasal step. It disappeared after restoration of normal pressure.

The same type of visual field defects have been reported in steroid-induced glaucoma in eyes which had been previously considered normal.

It may be argued, with some justification, that such eyes are susceptible to optic nerve damage and can therefore be considered as "preglaucomatous" eyes.

When glaucoma of many years' duration has produced extensive cupping and atrophy of the optic nerve, resulting in dense arcuate scotomas, the intraocular pressure may gradually decrease through lack of aqueous production, the visual

Fig. 1. Nerve fiber bundle defect in visual field produced by artificial elevation of intraocular pressure by ophthalmodynamometry.
field defects may remain static for long periods of time, and for all practical purposes the disease may be considered cured. If such a patient is seen for the first time he may be diagnosed as having "low tension glaucoma." If he has been under continuous observation throughout the development of his optic atrophy and scotomas the true nature of his glaucoma is evident. Such patients must have had an elevated intraocular pressure at one time and they always have an extremely poor outflow facility.

Fig. 2 illustrates such a case of "burnt out glaucoma" in a 75-year-old man. Visual field defects in the form of dense double arcuate scotomas were first noted in the left eye in 1955. The right field was normal. Intraocular pressure varied between 14 and 27 mm. Hg. The right optic disc was moderately cupped but the left disc was markedly excavated. The retinal vessels were narrowed and sclerotic. Facility of outflow was 0.10 in the right eye and 0.05 in the left. During the next 13 years the optic nerve cupping increased, the visual field defect in the left eye increased slightly, and a well-developed arcuate scotoma appeared in the right visual field. Initial therapy consisted of pilocarpine, epinephrine, and acetazolamide but, as the intraocular pressure level decreased, therapy was decreased and finally discontinued. For the past two years intraocular pressure has varied between 9 and 12 mm. Hg in each eye. Out-
flow facility remains very low and both optic discs show advanced excavation and atrophy.

There is no doubt that this patient has or has had glaucoma. If, however, he had been examined for the first time in 1968 such a diagnosis might be difficult to justify and the arcuate scotomata in the visual fields might well be attributed to a nonglaucomatous optic nerve atrophy.

The sudden development of a dense arcuate scotoma in the visual field of a patient with well-controlled glaucoma and circulatory insufficiency has been well documented.\(^5,6\) Borderline elevations of intraocular pressure in the range of 22 to 26 mm. Hg will produce gradually increasing optic nerve cupping and, eventually, a typical arcuate scotoma. If, however, the retinal arterial pressure is maintained at fairly high levels in patients with significant vascular hypertension, the high ratio of ophthalmic artery pressure to intraocular pressure will prevent the optic nerve ischemia essential to the development of optic atrophy and nerve fiber bundle defects in the visual field. Anything which upsets this ratio, whether it be marked increase in intraocular pressure or marked decrease in intravascular pressure (and consequently decreased blood flow), will encourage the development of optic nerve damage and the rapid onset of an arcuate scotoma in the visual field.

Such changes in the ratio of intraocular to intra-arterial pressure may be accomplished by spontaneous decrease in systemic and retinal arterial blood pressure associated with coronary artery occlusion or with cerebrovascular accidents or, iatrogenically, by rapid and significant decrease in blood pressure through too vigorous use of antihypertensive medication.

Fig. 3 illustrates the result of too drastic a drop in blood pressure on an eye with only minimally elevated intraocular pressure. The patient had moderately large optic cups and an intraocular pressure of 24 mm. Hg (Schiötz) in 1949. There was no visual field defect. Moderate arterial hypertension was noted. In 1961 the optic cups had enlarged but intraocular pressure measured 20 mm. Hg and visual fields were normal. In 1966 the patient had severe arterial hypertension with a blood pressure of 230/120. In 1967 the patient suffered a mild cerebrovascular accident with considerable drop in both systolic and diastolic blood pressure. At the same time antihypertensive medication was started with a resulting drop in blood pressure to 140/70. There was definite bradycardia. Very shortly after this blood pressure decrease the patient noted some visual disturbance in the left eye and was found to have a fairly dense and broad superior arcuate scotoma. Intraocular pressure was 15 mm. Hg. The optic disc atrophy and excavation was increased.

That this patient had borderline glaucoma there can be little doubt. Such a diagnosis is justified by the slightly elevated intraocular pressure readings in the past and by the slowly increasing optic disc atrophy and excavation. But the arcuate scotoma in the visual field is not entirely glaucomatous in origin. Rather it is the result of optic nerve ischemia secondary to decreased ophthalmic artery pressure and an abnormal ratio of intraocular to intra-arterial pressure.

In addition to the above illustrations of typical glaucomatous type arcuate scotomas in atypically glaucomatous eyes, there are a variety of conditions which can and do produce arcuate scotomas in eyes which definitely do not have glaucoma. Most of these have been described in a previous paper.\(^1\)

In general it may be said that any lesion which interrupts a nerve fiber bundle at or near the disc margin or within the optic nerve, even as far back as the chiasm, may produce an arcuate scotoma or nerve fiber bundle defect in the visual field.

Dubois-Poulsen and Magis\(^7\) believed that the lesion which gives rise to the scotoma is extraocular in location.
Hoyt has determined the position and confirmation of the superior and inferior arcuate nerve fiber bundles in the optic nerve and chiasm and has reported examples of arcuate scotomas produced by lesions within the optic nerve and chiasm.

Kearns and Rucker and I have reported examples of pituitary adenoma which gave rise to arcuate scotomas in the visual field.

Fig. 4 illustrates another such case in a young individual with no evidence of glaucoma. The arcuate scotoma is confined to the superior temporal field and has a sharply demarcated hemianopic border.

Two cases of opticochiasmatic arachnoiditis with adhesions between the chiasm and adjacent vessels and the development of arcuate scotomas in the visual field have been previously reported. One of these has been recently re-examined 20 years after the initial diagnosis was confirmed by surgical exploration of the chiasmal area.

Fig. 5 illustrates the static, dense, atypical arcuate scotoma in the left field which has remained unchanged through the years. The upper nasal quadrant defect in the right field is much less extensive than that found on initial examination years ago.

Drusen or hyaline verrucosities in the optic nerve or on the papilla have been shown by me and a number of others to produce fairly typical arcuate scotomas indistinguishable from those seen in advanced glaucoma.

Fig. 6 illustrates another such case. The defect is dense and slowly progressive, with steep borders and peripheral breakthrough. Both optic discs have visible hyaline bodies but the left visual field is normal.

Hoyt has reported an arcuate scotoma in a patient with a barely visible optic nerve pit. I have seen two such cases, one of which is illustrated in Fig. 7. In this instance the pit was large and located at the inferior edge of a moderately large and deep optic cup. The field defect was steep margined and dense.

Another patient with an optic nerve pit developed a cuneate type of scotoma in the upper nasal field. Neither of these eyes showed any evidence of glaucoma.

Visual fields have been carefully studied in other patients with optic nerve pits with negative results.

One glaucomatous eye, with both a deeply excavated optic cup and a very small optic nerve pit at the extreme lower border of the cup on the margin of the disc, has a well-established and quite dense superior arcuate scotoma. It is impossible to be certain which lesion produced the scotoma but it is probably due to the glaucoma. The other eye also shows a field defect consistent with the cupping of the disc and all of the criteria for the diagnosis of glaucoma are satisfied in both eyes.

Two highly myopic eyes with a very wide conus around each optic nerve head developed symmetrical inferior arcuate scotomas. Neither eye revealed any evidence of glaucoma after exhaustive study.

Fig. 8 illustrates what may be termed “refraction scotomas” in two eyes of the same patient, due, apparently, to posterior staphyloma around the optic disc with marked difference in refraction between this area and the macula. When the field was examined on the two-meter tangent screen with lenses correcting the refractive error in the macular region, the scotomas were readily demonstrated. When the refractive error on the optic disc was corrected, the arcuate scotoma in the right eye disappeared and the one in the left eye became smaller and less dense. Such “refraction scotomas” have also been reported by Schmidt and Enoksson.

Injury to the optic nerve within the orbit has given rise in one case in my personal experience to a narrow arcuate type of scotoma extending from the blind spot and passing immediately above fixation to end in a very small nasal step. The nerve fibers involved were in the
lower portion of the papillomacular bundle.

In another case, a very small meningioma at the superior lip of the intracranial end of the optic foramen gave rise to a gradually expanding inferior arcuate scotoma. The defect started in the Bjerrum area just below fixation and expanded laterally to connect up with the blind spot and nasally to end in a broad nasal step.

All of the above lesions giving rise to "mechanical" pressure on the nerve fiber bundles probably interfere with their vascular supply as well.

More frequent, and certainly more significant, are those frankly vascular lesions in nonglaucomatous eyes which give rise to arcuate scotomas in the visual field which are identical with those seen in advanced optic nerve damage from open angle glaucoma. Some of these have been reported elsewhere and other authors have noted their occurrence.

The most typical example of such a lesion has been previously reported and is illustrated by Figs. 9, 10, 11, and 12.

The patient entered my office with the statement that he had lost the vision of his left eye in the previous ten minutes. He had noted visual loss in the same eye of one or two minutes' duration a week before. He was totally blind in the left eye.

Fig. 6. Dense arcuate scotoma with peripheral nasal breakthrough produced by drusen in the optic disk.

Fig. 7. Optic nerve pit giving rise to a typical nerve fiber bundle defect in the field.

Fig. 8. Bilateral "refraction scotoma" in the form of grossly enlarged blind spots with extension of the defect into the Bjerrum area.

Fig. 9. Altitudinal type field defect resulting from visible transient embolus in the superior branch of the central retinal artery.
Fig. 10. Conversion of altitudinal field defect seen in Fig. 9 into a nerve fiber bundle defect after movement of the arterial embolus.

Fig. 11. Conversion of the inferior complete nerve fiber bundle defect seen in Fig. 10 into a typical arcuate scotoma after further clearing of the arterial embolus.

Fig. 12. Small residual arcuate scotoma three days after the field defect seen in Fig. 11. The defect eventually cleared completely.

Fig. 13. Atypical arcuate scotoma involving a part of the papillomacular nerve fiber bundle from an arterial plaque on the optic disk.

with a fixed and dilated left pupil, and the fundus picture of a central retinal artery occlusion. Immediate therapy consisted of retrobulbar injection of 2 per cent lidocaine, intravenous injection of 20,000 units of aqueous sodium heparin, 150 mg. of nicotinic acid, and massage of the globe. Within about three minutes the patient noted a return of some vision in his superior visual field and examination of the optic disc showed an arterial occlusion with a creamy white embolus, now involving the superior trunk of the central retinal artery and extending into both nasal and temporal branches with "box car" segmentation and sludging. The visual field showed a complete inferior altitudinal defect with macular splitting. Within the next 10 to 15 minutes there was marked visual improvement with regression of the arterial occlusion and the visual field examination now showed an area of macular sparing and a nerve fiber bundle type of defect ending in a nasal step. At the end of 1½ hours the embolus had moved into the superior temporal ar-
tery to a new bifurcation and blood flow was more even and rapid. The visual field defect was now a typical broad dense arcuate scotoma with macular sparing and a wide nasal step. Vision had returned to 20/20. At the end of three days the retinal arterial occlusion had disappeared but the visual field still showed a small inferior arcuate scotoma, detectable only with small stimuli and not connected with the blind spot. One month later the fundus appeared normal and there was no defect in the visual field. In the past four years there has been no visual complaint. No evidence of glaucoma has been found in either eye and results of clinical and arteriographic studies of the vascular system have been negative.

Three eyes with arcuate scotomas produced by an ophthalmoscopically visible creamy white embolus in a branch of the central retinal artery on the face of the optic disc have been previously reported and one such case is illustrated in Fig. 13. In addition to the arterial plaque there was segmental atrophy (ischemic infarct) in the optic nerve.

A number of patients with arcuate scotomas associated with carotid artery insufficiency or occlusion have been seen and some reported in a previous publication.

Fig. 14 illustrates a new and interesting example of such a case. The right eye had been injured twelve years before and had been "blind" since then. The left eye had had periodic attacks of amaurosis fugax with "blackout" of thirty seconds' to several minutes' duration during the previous year. Constant visual disturbance had been present for about a month. Visual acuity in the right eye was reduced to hand movements in all areas except the lower temporal quadrant, which was totally blind. Left visual acuity was 20/25.

The right eye had a subluxated lens with a hypermature cataract, but with a dilated pupil some retinal details could be seen in the peripheral fundus. No previous attempt had been made to examine the visual field in this allegedly "blind" eye. The left retina showed marked narrowing of some of the retinal arteries, especially at the inferior disc margin where they were almost obliterated. There was some asteroid hyalitis overlying the optic disk.

Visual field studies, using sufficiently large stimuli to be perceived by the "blind" right eye, showed a complete congruous right inferior homonymous quadrantanopsia. In the left field there was also a superior arcuate scotoma which broke through into the superior peripheral field and connected with the right inferior homonymous defect.

Ophthalmodynamometry of the left eye revealed a very low systolic and diastolic retinal arterial pressure in relation to the patient's brachial blood pressure. The arterial pressure in the right eye could not be measured. There was clinical and arteriographic evidence of severe left inter-
nal carotid artery stenosis with probable left ophthalmic artery and posterior cerebral artery occlusion.

Finally there are a variety of inflammatory lesions which may give rise to arcuate scotomas, the best known of which is juxtapapillary choroiditis.

While the typical visual field loss in optic neuritis is not an arcuate scotoma, this defect occurs often enough to make it important in differential diagnosis. The early lesion, with edema of the nerve and segmental vascular changes on the disc, is not difficult to differentiate from glaucoma. The late segmental atrophy secondary to localized ischemic infarct may present a picture in both the optic disc and the visual field which is very similar to that seen in long-standing glaucoma. I have seen several such cases and they have been reported by others.8, 14, 15

Temporal and cerebral arteritis have been previously reported1, 3 as causes of arcuate scotoma in the visual field. I have recently seen a patient in whom a complete loss of vision in one eye from temporal arteritis was preceded by the appearance of an altitudinal field defect with loss of the entire lower field but with sparing of a small portion of the papillomacular bundle and with a typical nasal step.

Fig. 15 illustrates still another case of temporal arteritis with incipient central retinal artery closure and the development of a double arcuate scotoma in the visual field. Clinical symptoms and signs of left temporal arteritis were severe pain in the left temple brow, and orbit with palpable and very tender arteries. Sedimentation rate was considerably elevated. There was dramatic response to corticosteroid therapy with cessation of pain and disappearance of the nerve fiber bundle defects in the visual field. There is no evidence of glaucoma in either eye.

Summary

The arcuate scotoma is the characteristic visual field defect in open angle glaucoma with optic nerve damage. Its morphology is varied. It occurs most typically in eyes with long-standing increase in intraocular pressure but it may be produced in certain susceptible eyes by artificially increasing the intraocular pressure by means of topical application of corticosteroids or by ophthalmodynamometry. It may appear suddenly in eyes with normal intraocular pressure but with rapid reduction of arterial pressure and blood flow in the arteries of the optic nerve. It is a common finding in “burnt out” glaucoma with low intraocular pressure.

The arcuate scotoma is not, however, pathognomonic of glaucoma. It may result from a variety of lesions located in the retina, on the optic disc, in the retrobulbar optic nerve, and in the chiasm, and the defect may be indistinguishable from the classical nerve fiber bundle defect of glaucoma.

The most common nonglaucomatous etiologic factors producing the arcuate scotoma are vascular lesions. These, in effect, produce ischemic infarcts in the optic nerve in locations which interrupt nerve fiber bundle conduction in the retina. Other lesions, such as drusen, tumor, and fibrous adhesions, may give rise to arcuate scotomas either by direct compression of nerve fiber bundles or by interference with their blood supply.

The finding of such typically glaucomatous visual field defects in nonglaucomatous eyes lends support to the thesis that the defect as seen in glaucomatous eyes is due not to increased intraocular pressure per se but to the effect of such an increase in pressure on the extraocular vasculature in the optic nerve.

REFERENCES

The visual field defect and ocular pressure level in open angle glaucoma

M. F. Armaly

When we inquire about the effect of ocular pressure level on the course of the visual function loss in glaucoma, we are in general offered two extreme though simple hypotheses. The first contends that the entire course can be explained by variation in ocular pressure level, and the second proclaims that the course of the visual function loss is independent of the level of ocular pressure. Despite their attractive simplicity, which accounts in a major part for their survival, neither one of these hypotheses alone can describe the reality of clinical experience.

There seems to be a general acceptance of the notion that ocular pressure level exerts its effect on visual function by compromising the blood supply to the nerve fibers sufficiently to interfere with their function or survival. Thus it follows that the effect of a given pressure level at a certain time depends upon the prevailing characteristics of the vascular bed and upon the ability of the neuronal elements to withstand the resulting deprivation. Hence the magnitude of reduction in blood supply that results from a certain level of ocular pressure will depend upon such factors as the arterial pressure of the feeding vessels and the number and caliber of the vessels comprising the vas-