Brit. J. Ophthal. (1974) **58**, 964

Anterior ischaemic optic neuropathy

II. Fundus on ophthalmoscopy and fluorescein angiography

SOHAN SINGH HAYREH

Department of Ophthalmology, University of Iowa

The classical description of the fundus appearances in anterior ischaemic optic neuropathy (AION) is that of pale oedema of the optic disc (OD), usually accompanied by OD and peripapillary haemorrhages, and invariable optic atrophy in later stages. These are almost always considered to be the only fundus findings, although the presence of "exudates" at the posterior pole has occasionally been mentioned. The retinal vessels are usually described as being normal.

Reports of fluorescein fundus angiography (FFA) findings in AION in the literature are scanty and brief. Foulds (1968) mentioned that FFA showed incomplete filling of the capillaries in the part of the optic nerve head corresponding to the visual field defect, and might show capillary dilatation and abnormal permeability of the optic disc vessels with widespread fluorescein leakage or none. Begg, Drance, and Goldmann (1972) showed defective or delayed filling in atrophic sectors of the OD and adjacent peripapillary choroid after sectoral AION. Sanders (1971) described late choroidal filling of the peripapillary region and OD or filling defects at the disc corresponding with the field defect, dilatation of the peripapillary plexus, and hyperfluorescence of the OD.

In the present series, a detailed study has been made of the ophthalmoscopic and FFA changes in the OD, peripapillary region, retina, retinal vessels, choroid, and the rest of the fundus. The findings, which have either not been mentioned previously or have received little attention in the literature, add considerably to our knowledge not only of the pathogenesis and management of AION but also of its diagnosis.

Material and methods

(I) 25 cases of complete or partial AION were studied. All these patients had a detailed initial ophthalmic examination, and the erythrocyte sedimentation rate (ESR) was estimated by the Westergren method as an emergency. If the ESR was higher than 20 mm/1st hr, a temporal artery biopsy was performed to look for evidence of temporal (giant-cell) arteritis. A routine haematological and systemic examination was performed. Stereoscopic fundus colour photography and FFA were performed at the first attendance, or as soon as possible thereafter, in all eyes.

These patients were followed for from 3 months to 3 years, the majority for between 1 and 2½ years (mean 15 ± 9 mths). Among the review studies were included a thorough fundus examination and stereoscopic colour photography of the OD; in ten eyes serial FFA was performed at different intervals to assess the ocular circulation.

Address for reprints: Prof. S. S. Hayreh, Department of Ophthalmology, University of Iowa Hospitals, Iowa City, Iowa 52242, U.S.A.

A part of this study was supported by a grant from the British Medical Research Council.

(II) In addition to these 25 cases, I have seen more than fifty cases of AION over the years, which were not investigated as systematically and thoroughly as those mentioned above although FFA was performed in all of them. Some of the observations from these additional cases are cited in the text, to substantiate certain observations but without any statistical data.

Observations and discussion

Fundus Changes

The main ocular abnormalities in cases with AION are revealed by examination of the fundus. They may be classified as follows:

(A) OPTIC DISC (OD) CHANGES

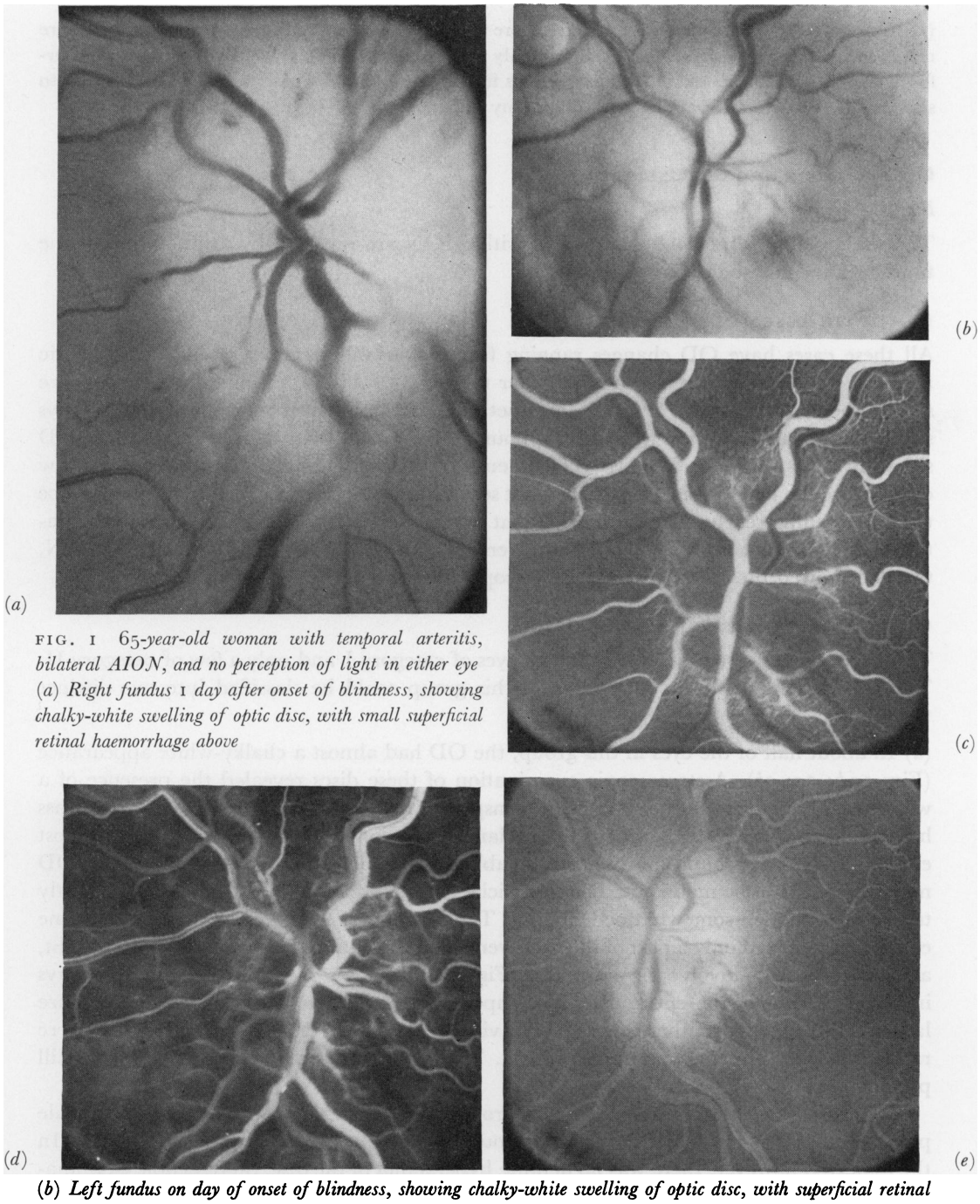
All these cases have OD changes ranging from a variable swelling of the OD to optic atrophy, depending upon the interval after the onset of AION at which the patients are seen. When seen within a few days of onset of the visual disturbance, the OD is always swollen. If a patient is seen within a few hours of the onset of visual deterioration, the OD shows swelling (Fig. 1*b*). Foulds (1968) mentioned that OD swelling may develop a few days before the loss of vision, but I have not seen any such instance so far; in my experience the OD swelling reaches its maximum about 2 to 3 days after the onset of visual deterioration. In the present study, when patients were seen during these early stages of the AION, the OD showed the following ophthalmoscopic appearances:

(1) *In AION due to temporal arteritis*

This group included eleven eyes of the 25 eyes of category I and only a few of category II. The appearances of the swollen discs in this group could be classified into two distinct types:

(*a*) In about half of the eyes in this group, the OD had almost a chalky-white appearance (Figs 1*a,b*; 2*a*; 3*b*). A stereoscopic examination of these discs revealed the presence of a white mass lying deep to the superficial transparent nerve fibre layer of the OD: the mass had the look of a white infarct (of the prelaminar region) which merged with an almost equally white zone around the disc (presumably ischaemic pigment epithelium) so that OD margins could not be made out. The superficial nerve fibre layer of the OD was frequently transparent though somewhat oedematous. The swelling of the OD and infarction in one eye at first involved only a part of the OD even when the patient had no perception of light, and later spread to involve the entire disc (Fig. 3*b*). This type of OD change almost always involved ultimately the entire disc. The superficial capillaries in the surface nerve fibre layer of the OD were neither congested nor visible. Haemorrhages on or near the OD were rare and when present were mostly slight. The central depression of the OD was still present.

(*b*) In the other half of the eyes in this group, the OD was oedematous and had a pale pink or sometimes a nearly normal pink colour, in distinct contrast to the former group. In the majority, however, there was no definite hyperaemia of the disc. Stereoscopic examination of these discs revealed oedema of the disc, and frequently a deeper pallor; one had the impression that the prelaminar region was oedematous and somewhat pale, with the normal colour of the surface nerve fibre layers superimposed on this prelaminar oedema. The oedema extended into the immediate peripapillary region. In discs in which the AION was sectoral, the oedema was usually greatest in the involved part; the rest of the disc was not free from oedema, but it was less marked over the uninvolved part. In some of the sectoral



(a)

FIG. 1 *65-year-old woman with temporal arteritis, bilateral AION, and no perception of light in either eye*
(a) Right fundus 1 day after onset of blindness, showing chalky-white swelling of optic disc, with small superficial retinal haemorrhage above

(b)

(c)

(d)

(e)

(b) Left fundus on day of onset of blindness, showing chalky-white swelling of optic disc, with superficial retinal haemorrhage infero-temporally at the optic disc margin
(c) Fluorescein fundus angiogram of left eye. Retinal arterial phase, showing no filling of optic disc and choroid
(d) Fluorescein fundus angiogram of left eye. Retinal venous phase, showing no filling of optic disc and faint patchy filling of choroid
(e) Fluorescein fundus angiogram of left eye. Late phase about 30 minutes after (c), showing fluorescein staining of optic disc

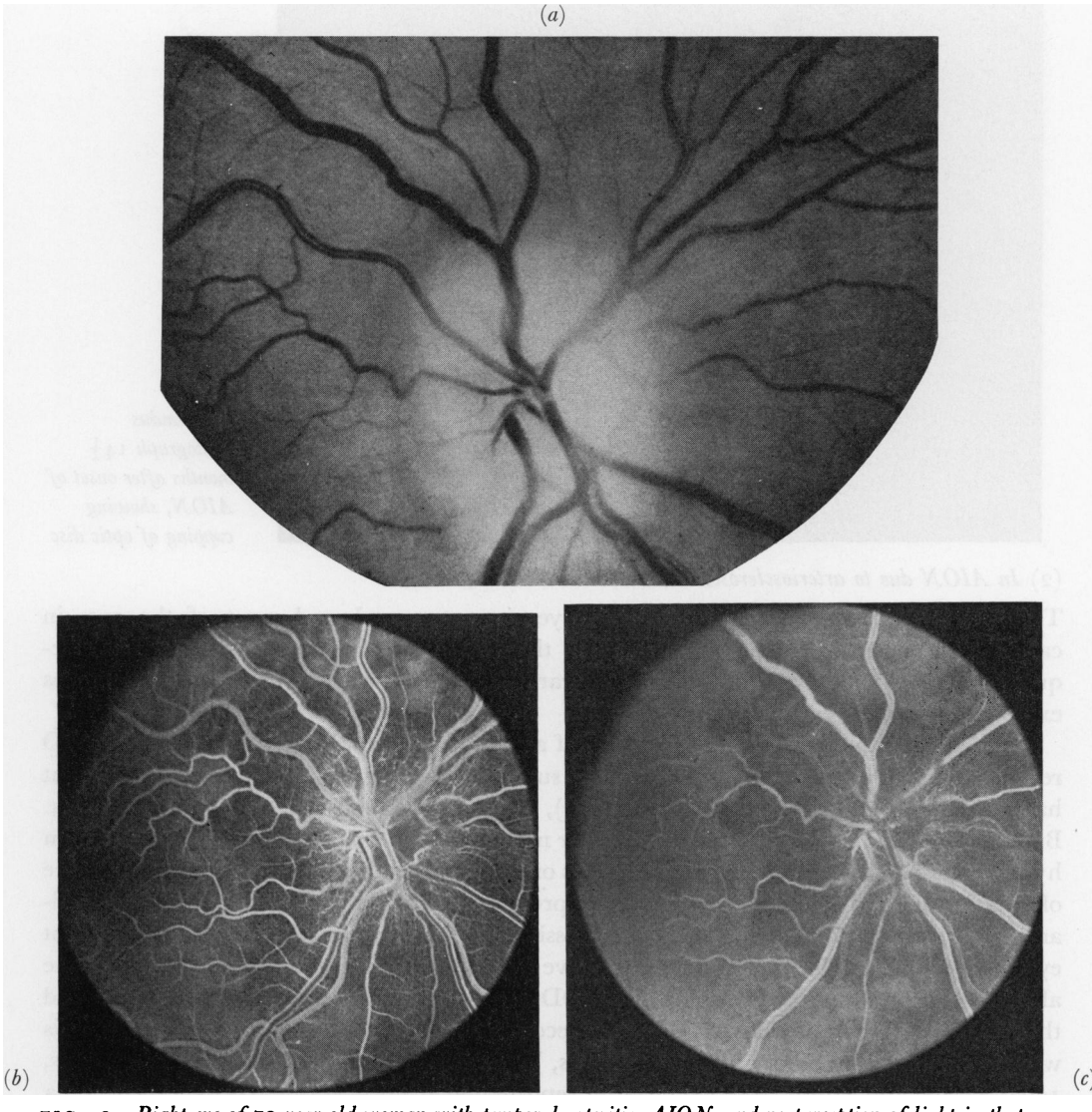
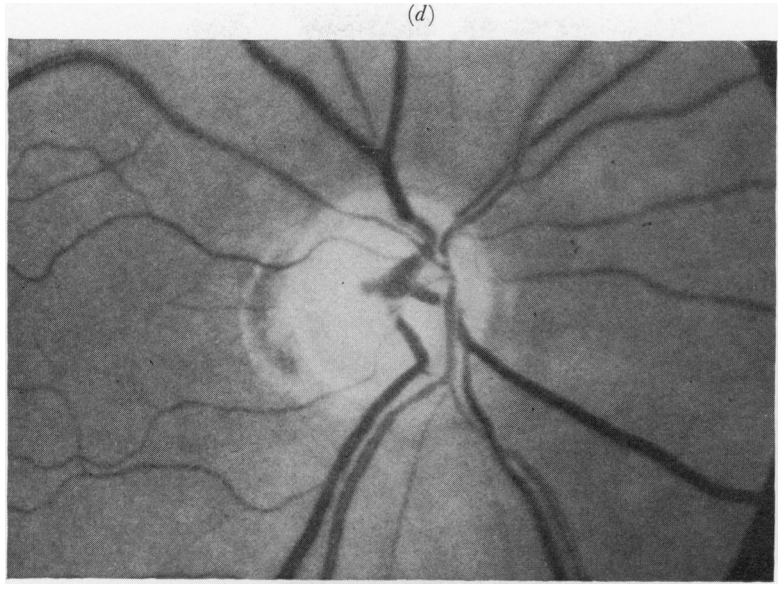


FIG. 2 *Right eye of 72-year-old woman with temporal arteritis, AION, and no perception of light in that eye (a), (b), (c) 3 days after onset of AION and on the day of development of no perception of light*
(a) Fundus photograph, showing chalky-white swelling of right optic disc, with no haemorrhages
(b) Fluorescein fundus angiogram. Retinal arterio-venous phase, showing no filling of optic disc, peripapillary choroid, and nasal choroid, with filling of temporal choroid
(c) Fluorescein fundus angiogram. Retinal venous phase, showing no filling of optic disc and peripapillary choroid, poor filling of inferior watershed zone of the choroid, and filling of rest of choroid

cases it was almost uniform all over the disc. Superficial flame-shaped haemorrhages were seen frequently, mainly along the peripapillary capillaries; they were usually slight, although one eye showed marked haemorrhages on and around the disc. The appearance of an oedematous disc of this type could be confused with oedema of the OD due to other causes, although the oedematous discs in AION frequently tended to be slightly paler than in the other types of OD oedema; also it was usually not very marked.



(*d*) *Fundus photograph* $14\frac{1}{2}$ *months after onset of AION, showing cupping of optic disc*

(2) *In AION due to arteriosclerosis*

This group includes fourteen of the 25 eyes in category I and most of the eyes in category II. Most of these discs resembled those described in Type 1*b* (Figs 4*a*; 5) frequently with minor or no pallor, and only rarely those in Type 1*a* (in two of twelve eyes examined at a very early stage).

Thus, during the early stages of AION, if an ophthalmoscopic examination of the OD reveals a chalky-white swollen disc with no superficial congestion of capillaries and slight haemorrhage (Fig. 1*a*,*b*) or none (Figs 2*a*; 3*b*), it is most probably due to temporal arteritis. But when the disc shows oedema with a near normal, slightly pale colour (Fig. 4*a*) or even hyperaemia (Fig. 5) and some haemorrhages on and around the disc, there is a high chance of its being due not to temporal arteritis, but probably to arteriosclerosis. However, no hard-and-fast rules can be made on this basis. Possibly the chalky-white swollen discs represent eyes with massive infarction of the optic nerve head and retrolaminar optic nerve, because all of them showed marked cupping of the OD on resolution, generally much more marked than in Type 1*b*. A partial AION may become total after several days (Fig. 3*b*); this was also reported by other authors (François, Verriest, Neetens, De Rouck, and Hanssens, 1962; Saraux and Murat, 1967). The latter reported, in arteriosclerotic AION, OD oedema with some hyperaemia on the first day, a pale OD the second day, and a pink OD several days later. I have not observed such a pattern.

Bonamour, Bonnet, Brégeat, and Juge (1968) reported some cases which they considered to be cases of AION, but their clinical description is that of OD vasculitis Type II which I have already described (Hayreh, 1972b).

The swelling of the OD usually starts to subside about 7 to 10 days after the onset, and after about a month or more a pale atrophic disc is seen, usually with well-defined margins (Figs 2*d*; 3*d*; 4*b*; 6). The interval between the onset of AION and the development of optic atrophy has been given as a few weeks (Meadows, 1968), 4 weeks to 4 months (Lasco, 1961), several weeks (Saraux and Murat, 1967), 2 to 3 months (Bonamour, 1966), and from the 15th day (Bonamour and others, 1968). In some of the bilateral cases, if AION develops in one eye when the fellow eye is already atrophic, the condition may be misdiagnosed as the Foster–Kennedy syndrome (Larmande, 1948; Saraux and Murat, 1967).

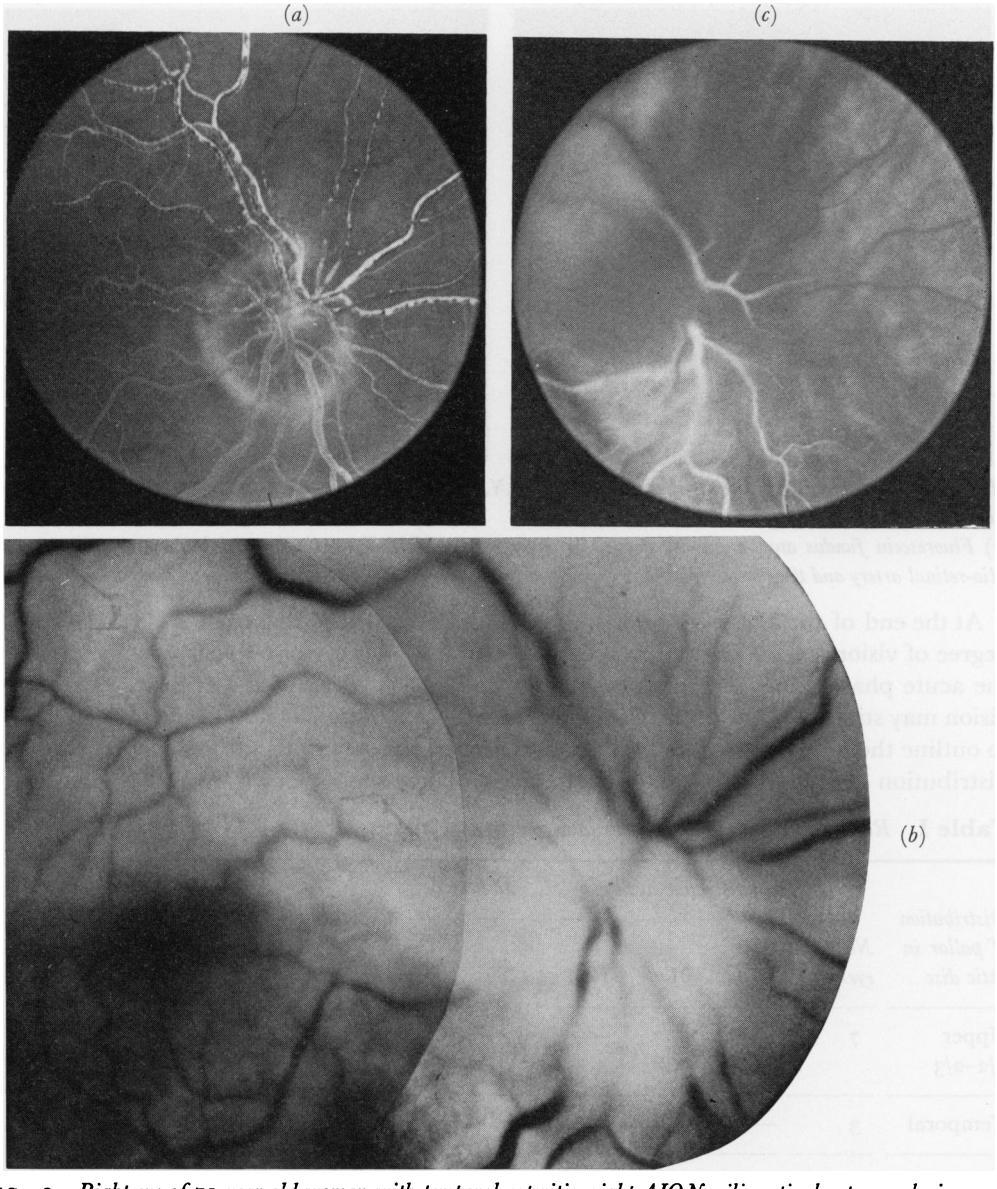
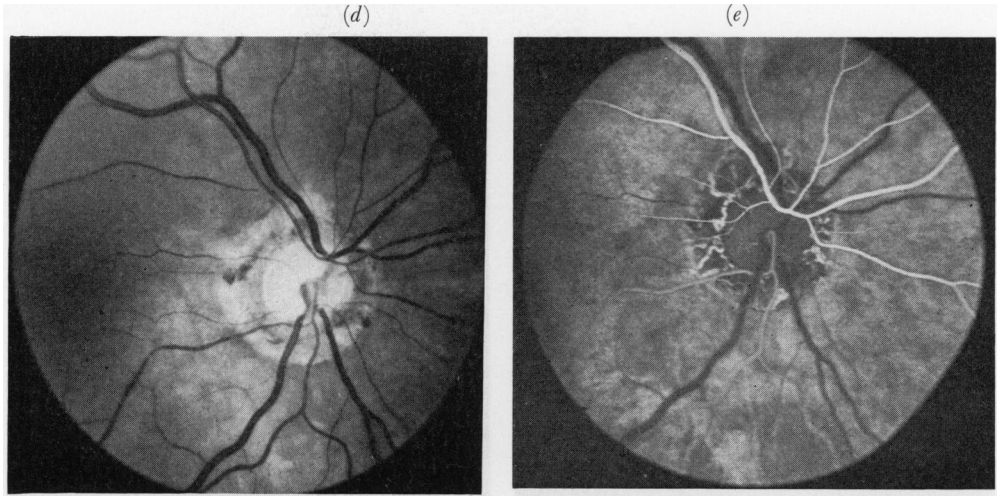


FIG. 3 *Right eye of 71-year-old woman with temporal arteritis, right AION, cilio-retinal artery occlusion, and no perception of light in that eye*

(a) Fluorescein fundus angiogram 2 days after onset of blindness. Late phase, showing sludging of fluorescein in cilio-retinal artery and accompanying retinal vein (in upper half of the retina), no fluorescence of the disc. Central retinal artery (in lower half of the retina) filled normally with no filling of cilio-retinal artery and optic disc during transit of the dye

(b) Fundus photograph, 4 days after onset of blindness, showing chalky-white swelling of entire optic disc with oedema of upper half of retina. 2 days after the onset of blindness, the optic disc showed white swelling of the lower part only, with normal colour of the upper part, and retinal oedema of the upper half of the retina

(c) Fluorescein fundus angiogram 4 days after onset of blindness. Retinal arterial phase, showing filling of the central retinal artery but the cilio-retinal artery has just started to fill slowly. There is no filling of the optic disc, peripapillary choroid, and superior watershed zone in the choroid, but there is faint filling of the rest of the choroid



(d) Fundus photograph 8½ months after onset of AION, showing atrophy and cupping of optic disc with peri-papillary chorio-retinal degeneration
(e) Fluorescein fundus angiogram 8½ months after onset of AION. Pre-retinal arterial phase showing that the cilio-retinal artery and the choroid fill before the central artery (compare with Fig. 3c), with no filling of the optic disc

At the end of the follow-up period, 21 of the 25 eyes of my category I still retained some degree of vision, indicating the survival of a variable number of nerve fibres. Even during the acute phase, though there is frequently a diffuse swelling of the disc, some degree of vision may still exist. The presence of a diffuse oedema in the OD may make it very difficult to outline the infarcted sector of the disc. In my series, when optic atrophy supervened, the distribution of the pallor of the disc and visual acuity was as shown in Table I.

Table I *Relationship of optic atrophy and final visual acuity*

Distribution of pallor in optic disc	*No. of eyes*	*Final visual acuity*								
		NPL	PL	HM	CF	6/60	6/36	6/12	6/9	6/6 or better
Upper 1/2–2/3	7	—	—	—	1	—	—	1	2	3
Temporal	3	—	—	—	2	—	1	—	—	—
Inferior temporal	1	—	—	—	1	—	—	—	—	—
Diffuse	19	9	2	3	2	1	1	—	—	1

The patient who could see 6/5 with diffuse optic atrophy in that eye reported that she was seeing as if through holes in a lace curtain; the involvement of the nerve fibres scattered in patches all over the OD must have been responsible for these symptoms and the diffuse optic atrophy.

(3) Cupping of the optic disc

In previous reports of AION there is hardly any mention of the incidence of cupping of

the OD in these cases. This is somewhat surprising. Begg, Drance and Sweeney (1970, 1971) reported the presence of notching of the neuro-retinal rim in patients with sectoral AION in chronic simple glaucoma, which occurred some 2 to 3 months after the original haemorrhage had disappeared. Drance (1972) commented that, after the usual AION, the optic nerve becomes atrophic but rarely cupped. Miller (1972) mentioned the occurrence of cupping of the OD without exception in AION due to temporal arteritis but gave no other details.

In my series cupping was present in thirteen eyes (Figs 2*d*; 3*d*; 6). The cupping usually developed about 2 to 3 months after the onset of AION, sometimes in as little as 6 weeks. There was a rapid progress in cupping, so that after 3 to 4 months it was at its maximum and thereafter increased only minimally in eyes followed-up for 12 to 20 months (Fig. 3*d*). The subject of cupping of the OD in AION and its pathogenesis is discussed elsewhere (Hayreh, 1974b). The relationship of the cupping of the OD to temporal arteritis, optic atrophy, and final visual acuity is shown in Table II.

Table II *Correlation of optic atrophy, cupping of the optic disc, temporal arteritis, and final visual acuity*

Optic atrophy	*No. of eyes*	*Cupping of optic disc*	*Temporal arteritis*		*Final visual acuity*								
			Present	*Absent*	NPL	PL	HM	CF	6/60	6/36	6/12	6/9	6/6
Diffuse	10	Present	9	1†	8*	0	1	0	1	0	0	0	0
	7	Absent	0	7	0	0	2	1	0	2	0	0	2
Sectoral	3	Present‡	2	1	0	0	0	2§	0	0	0	1	0
	7	Absent	0	7	0	0	0	3	0	0	1	0	3

* In one of these, optic atrophy was more marked in the temporal than the nasal part, and so was the cupping
† This eye had a very shallow saucer-shaped cupping as compared to the other eyes with cupping
‡ Atrophy and cupping involved upper one-half to two-thirds of the optic disc in two eyes
§ One eye also had central retinal artery occlusion

(B) RETINAL CHANGES

In the present study, in about half of the eyes, a variable number of small superficial flame-shaped retinal haemorrhages was seen at the margins of the OD, sometimes even extending on to the adjacent retina (Figs 1*b*; 4*a*; 5). Congestion of the radial peripapillary capillaries was occasionally seen, but was in no way as frequent or extensive as that seen in OD oedema due to intracranial hypertension. In marked cases, the retina near the margin of the OD showed patchy oedema and haziness which partially or completely masked the retinal vessels in the localized area (Figs. 1*a,b*; 2*a*; 3*b*). The nerve fibre layer over the OD was usually transparent though somewhat oedematous. Rarely, a small cotton-wool spot was seen near the margin of the OD. Whenever a cilio-retinal artery was present in these cases, a localized area of retinal oedema (infarction) in the region of supply of the artery was seen (Fig. 3*b*); this has also been found by other authors (Cüppers, 1951; Siegert, 1952; Simmons and Cogan, 1962; François and others, 1962). Since AION is due to occlusion of the posterior ciliary arteries, it is natural that a cilio-retinal artery will also be occluded. Such retinal infarcts were described by Cullen (1968) as "exudates" in AION due to temporal arteritis.

(C) RETINAL VASCULAR CHANGES

The presence of arteriosclerosis, often marked in the retinal arteries, is a common finding in

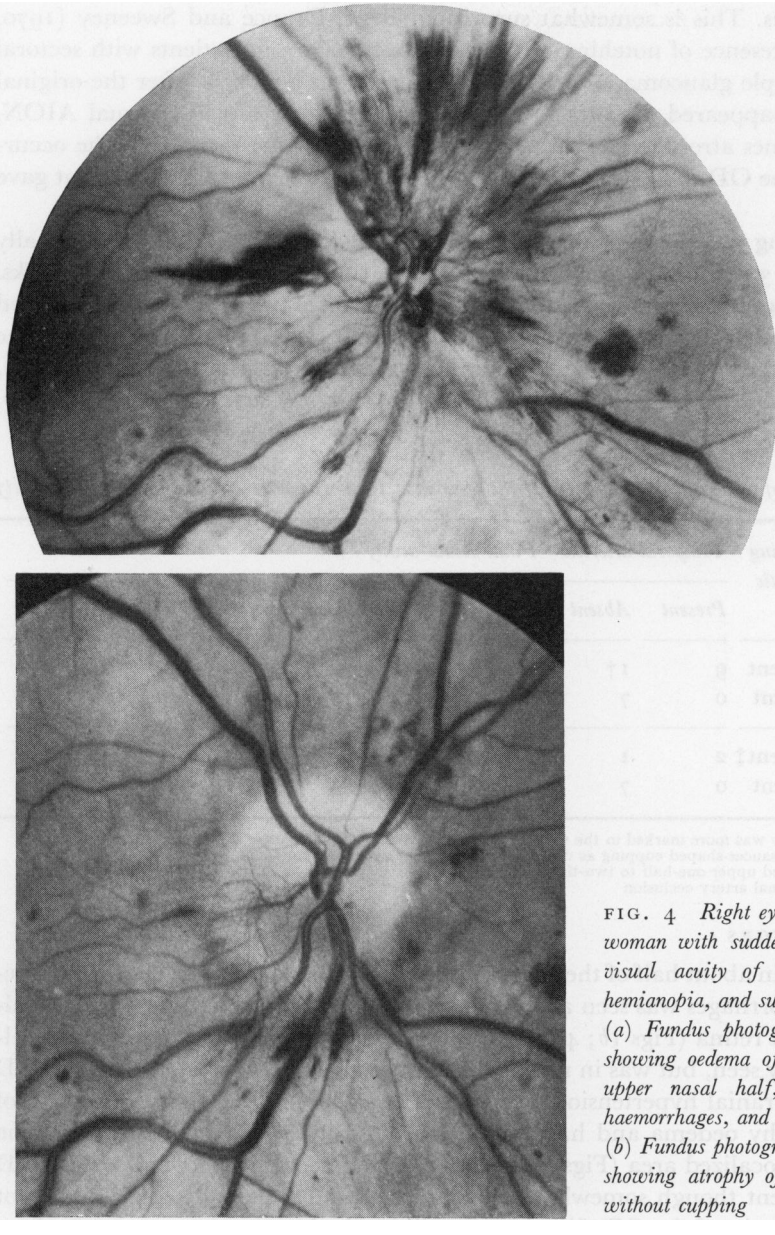


FIG. 4 *Right eye of a 60-year-old diabetic woman with sudden onset of blurred vision, visual acuity of 6/12, inferior altitudinal hemianopia, and superior sectoral AION*
(a) Fundus photograph 16 days after onset, showing oedema of optic disc, greatest in the upper nasal half, with superficial retinal haemorrhages, and diabetic retinopathy
(b) Fundus photograph 3½ months after onset, showing atrophy of upper half of optic disc without cupping

these elderly patients, but is not necessarily indicative of functional change. In the present series it was interesting to observe that in some cases arteriosclerotic changes were significantly more advanced in the eye with AION than in the other, normal, eye. Three patients in this series showed evidence of associated retinal arterial occlusion:

(*i*) Occlusion of a large cilio-retinal artery supplying the upper half of the retina (Fig. 3);
(*ii*) Occlusion of the central retinal artery associated with a sectoral involvement of the OD by AION (Fig. 7);
(*iii*) In a patient seen a few weeks after onset of AION, electroretinogram revealed evidence of an old central retinal artery occlusion (Fig. 6).

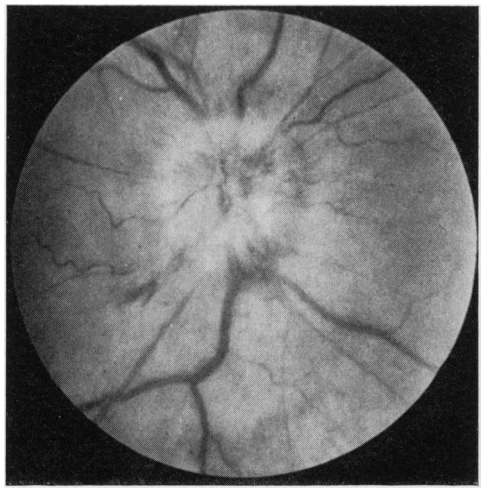


FIG. 5 *Right eye of 74-year-old man with sudden onset of inferior altitudinal hemianopia, superior sector AION, no temporal arteritis, and visual acuity 6/7.5. He had similar trouble in the left eye 10 years previously and has residual left inferior altitudinal hemianopia and optic atrophy of upper half of the left disc*

Fundus photograph of right eye 10 days after onset, showing oedema of the optic disc with hyperaemia and superficial haemorrhages

In another patient (not included in this series) a small cilio-retinal artery was seen in both eyes which on development of bilateral AION resulted in a localized retinal infarction in each eye (Hayreh, 1969a,b).

No doubt the central retinal artery can be involved by temporal arteritis independently of AION and I have seen such cases. However, the central retinal artery occlusion in temporal arteritis can be a part of the AION process in many cases and the latter has been missed in the past on simple ophthalmoscopic examination. Simultaneous presence of central retinal artery occlusion with AION can easily be explained if one considers the modes of origin of the central retinal artery.

In my early studies (Singh and Dass, 1960a), I found that the central artery of the retina

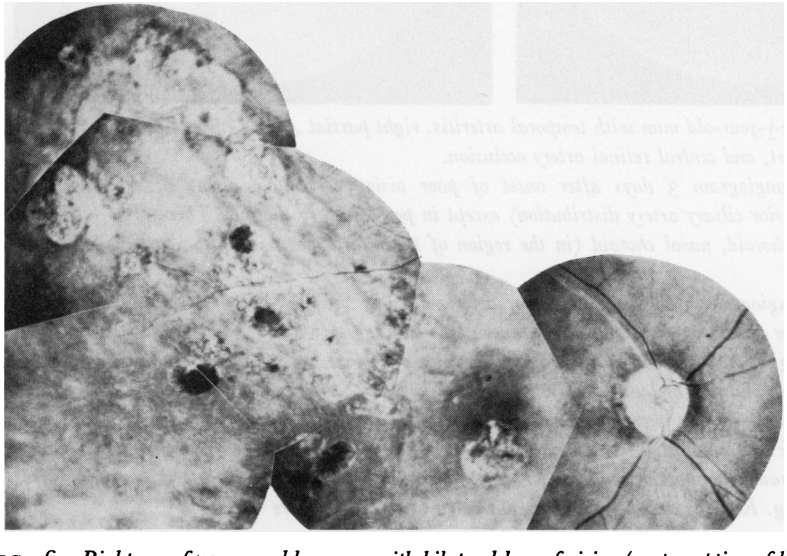


FIG. 6 *Right eye of 72-year-old woman with bilateral loss of vision (no perception of light in right eye and hand movements in left eye) 9 months previously, with temporal arteritis and bilateral AION*

Fundus photograph, showing pale, atrophic, and cupped optic disc, with sheathing of right retinal arteries and multiple chorio-retinal degenerative patches. These were also seen in the left eye

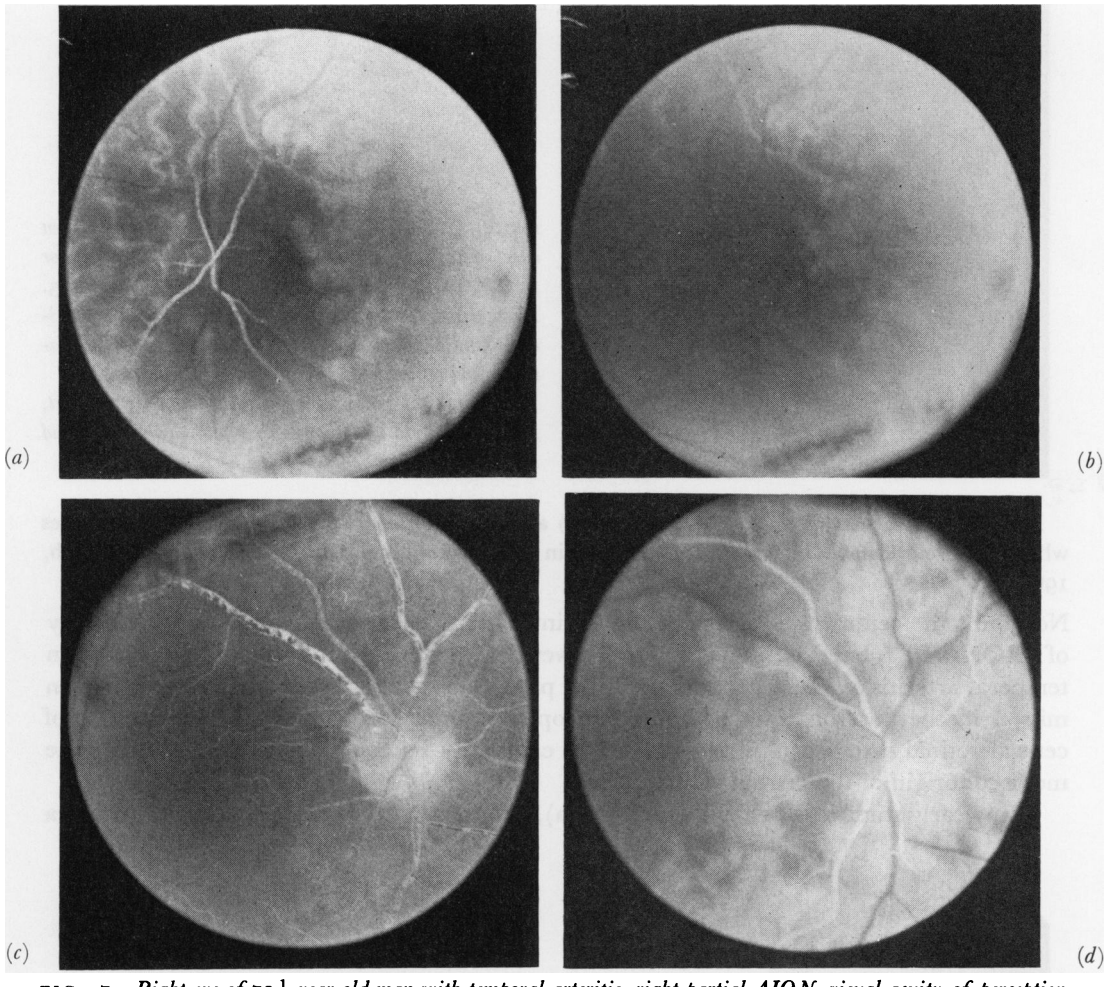


FIG. 7 *Right eye of 72½-year-old man with temporal arteritis, right partial AION, visual acuity of perception of light in upper nasal part, and central retinal artery occlusion.*

(a) Fluorescein fundus angiogram 3 days after onset of poor vision. Adequate filling of temporal choroid (in region of lateral posterior ciliary artery distribution) except in peripapillary choroid. There is no filling of the optic disc, peripapillary choroid, nasal choroid (in the region of the medial posterior ciliary artery distribution), and central retinal artery

(b) Fluorescein fundus angiogram 3 days after onset of deterioration of vision. Start of filling of central retinal artery and medial posterior ciliary artery almost simultaneously and sluggishly 4 to 5 seconds after (a)

(c) Fluorescein fundus angiogram 3 days after onset of deterioration of vision. Late phase. Sludging of circulation in retinal veins and fluorescein staining of lower half of optic disc

(d) Fluorescein fundus angiogram 11 days after onset. Retinal arterial phase, showing a much improved retinal, nasal choroidal, and optic disc circulation compared to a, b, and c. However, filling defects in the temporal and superior peripapillary choroid and superior watershed zone of the choroid still persist till late. Upper two-thirds of optic disc show poor filling. Retinal circulation still slow though better than before

and the medial posterior ciliary artery (PCA) arise by one common trunk from the ophthalmic artery in 40·4 per cent., and divide into the central retinal artery and medial PCA at some distance from the ophthalmic artery. Similarly, the central retinal artery may arise in common with the lateral PCA (in 12·5 per cent.) or with the medial PCA + lateral PCA

(in 6·7 per cent.). If the occlusion takes place in the common trunk near its origin from the ophthalmic artery, it will occlude both the central retinal artery and one or both PCAs. Thus it is no surprise that central retinal artery occlusion is seen in AION. The fact that AION is not as commonly associated with central retinal artery occlusion, as is suggested by the mode of origin of these arteries, indicates two possibilities:

(*1*) The lesion producing occlusion of the PCAs involves the arteries at some distance from their origin and is not an extension of the process from the ophthalmic artery. Thus, PCAs may have a selective vulnerability to involvement in temporal arteritis and in the arteriosclerotic process which is not shared to the same extent by the central retinal artery, although the presence of arteriosclerotic changes more marked in the eye with AION than in its fellow eye (as mentioned above) would indicate that the central retinal artery is equally or only slightly less involved by arteriosclerosis than the PCA.

(*2*) When the perfusion pressure in the intraocular arteries falls, and an imbalance occurs between the intraocular pressure and the perfusion pressure, the intraocular distribution of the PCAs, particularly to the optic nerve head and peripapillary choroid, is much more susceptible to obliteration than the central retinal artery (Hayreh, Revie, and Edwards, 1970; Hayreh, 1972a). This has been well demonstrated in patients with AION. A complete occlusion of the PCAs is thus not essential to the production of AION (Hayreh, 1974a). With partial occlusion of the parent trunk of both the PCA and central retinal artery and on a fall of its diastolic perfusion pressure to below the intraocular pressure, AION can be produced although the retinal circulation may still be intact. Thus, all elderly patients with central retinal artery occlusion and pale-looking ODs should be investigated for temporal arteritis (by erythrocyte sedimentation rate estimation and by FFA). FFA will show a normal filling of the small vessels in the OD in cases with central retinal artery occlusion unassociated with PCA occlusion, and no filling of the disc, peripapillary choroid, and central retinal artery in AION cases with central retinal artery occlusion (Fig. 7*a*). This precaution will prevent the development of AION in the other eye if the presence of temporal arteritis is detected early and treated with corticosteroids. Even if the central retinal artery occlusion is unassociated with AION and is due to temporal arteritis, the discovery of the temporal arteritis and the use of early and intensive corticosteroid therapy can prevent involvement of the other eye. Every patient over 60 years suffering from central retina artery occlusion should therefore be investigated for temporal arteritis.

(D) MACULAR AND PERIPAPILLARY CHANGES

Eyes with AION frequently show a variable degree of senile macular degenerative changes compatible with their age. Similarly, a variable degree of peripapillary degenerative halo—complete or partial—is not uncommon. During the first 2 to 3 weeks after the onset of acute AION, a whitish lesion continuous with the infarct of the optic nerve head is seen, involving the peripapillary region for a variable distance (Figs 1*a*,*b*; 2*a*; 3*b*); this is in all probability degenerate pigment epithelium in this region caused by the ischaemia of the underlying peripapillary choroid. The occurrence of similar whitish lesions, due to ischaemic necrosis of the pigment epithelium and outer layers of the overlying retina in other parts of the fundus in rhesus monkeys, on experimental occlusion of the PCAs, has been demonstrated both ophthalmoscopically and histopathologically (Hayreh and Baines, 1972; Hayreh, 1973). Like these experimental lesions, this peripapillary whitish lesion later resolves, leaving a variable amount of chorio-retinal degeneration of the involved area (Figs 2*d*; 3*d*).

(E) PERIPHERAL CHORIO-RETINAL DEGENERATIVE PATCHES

In patients with AION the peripheral part of the fundus may show patches of chorio-retinal degeneration. These were seen in 30 per cent. of the eyes in this series and the most extensive example is shown in Fig. 6. The size, shape, and distribution of the patches varied considerably.

Of the nine eyes with such patches, the visual acuity was no perception of light in five eyes, nasal hemianopia with the border passing through the blind spot in one eye (Fig. 13 in Hayreh, 1970: the patches were only in the temporal part of the fundus, *i.e.* the region with lateral PCA occlusion), hand motion to counting fingers in two eyes, and 6/12 in one eye. The development of such patches and their ophthalmoscopical and histopathological evolution has been demonstrated in experimental PCA occlusion in rhesus monkeys (Hayreh and Baines, 1972; Hayreh, 1973). If occlusion of the PCA produces such chorio-retinal degenerative patches, it must be asked why these are seen in only 30 per cent. of eyes with AION when AION is due to occlusion of the PCAs? This rarity of chorio-retinal lesions in patients can be explained by the fact that, in almost all the patients with AION, FFA showed the presence of some choroidal circulation from the PCAs by the venous phase of the retinal circulation (*e.g.* Fig. 2*c*) in spite of there being no filling of the peripapillary choroid and OD all along the transit of the dye (Fig. 2*b*,*c*). This is because, in all cases, complete occlusion of the PCAs is not essential to produce AION (p. 959; and Hayreh, 1974a). Thus, in these cases, the OD suffers most, while the poor and delayed circulation in the rest of the choroid in these eyes is sufficient to prevent the development of the classical fundus lesions seen during experimental complete occlusion of the PCAs. This survival of the pigment epithelium even in the presence of a poor choroidal circulation was observed in our experimental studies (Hayreh and Baines, 1972; Hayreh, 1973).

Intravenous Fluorescein Fundus Angiography (FFA)

The angiographic pattern varied not only with the degree of involvement of the OD but also with the interval between the onset of AION and the angiographic examination.

(A) OPTIC DISC CHANGES

(*1*) *In AION involving the entire OD*

(*i*) During the first week after the onset of AION, no fluorescence of the OD was seen either during the transit of the dye or during the late phases (Figs 1*b*,*c*; 2*b*,*c*; 3*a*,*c*) although very occasionally very late staining was seen (Fig. 1*e*).

(*ii*) During the second week, although no fluorescence of the OD was present during the transit of the dye, the disc usually stained with fluorescein during the late phases.

(*iii*) From the third week onwards, the OD showed fluorescence during either the retinal arterial or the arterio-venous phase, and later it showed staining with blurred margins.

(*iv*) After about 2 months, when oedema of the OD had subsided and optic atrophy was established, the OD usually showed very faint fluorescence during both the transit of the dye and the late phases (Fig. 3*e*) or none at all.

(*2*) *In sectoral AION*

The normal part of the OD filled normally while the ischaemic part either did not fill (Fig. 7*c*,*d*) or filled very late during the transit of the dye depending upon the age of the lesion. During the late phases, the pattern was as follows:

(*i*) During the first week, it was usually only the the normal part that stained with fluorescein (Fig. 7*c*).

(*ii*) Towards the end of the second week and thereafter, the ischaemic part started to stain more than the normal part and had a blurred border, with a transitional period when the whole disc stained equally well.
(*iii*) When optic atrophy was established and there was no oedema, only the normal part was fluorescent during the transit and later.

In eyes with sectoral AION, the filling defects in the OD shown during the transit of the fluorescein were significantly correlated with the visual field defects found in these eyes. There was also a significant correlation with the filling defects in the OD and those in the peripapillary choroid during the transit of the dye when seen during the first two weeks after onset; after that period, once the choroidal circulation was restored, such a correlation was usually not seen.

(B) CHOROIDAL CIRCULATION

(*1*) *In AION involving the entire OD*

(*i*) During the first week after the onset of AION, there was markedly delayed and poor filling of the choroid by the PCAs (Figs 1*c*,*d*; 2*b*,*c*). The earlier a patient was seen the more marked it was; so much so that within a few hours of the onset of AION practically no choroidal filling was detected during the retinal transit of the dye (Fig. 1*c*,*d*). One PCA might fill before the other (Figs 2*b*; 3*c*), but all were slow to fill. The choroid filled either in the retinal venous phase or more commonly afterwards, and only rarely did the PCAs start to fill during the retinal arterio-venous phase. The peripapillary choroid was much more delayed than the rest of the choroid—in fact, during the first 3 days, it frequently did not show any filling (Figs 1*c*,*d*; 2*b*,*c*) or did not fill till the late retinal venous phase or later (Fig. 3*c*). The filling of the choroid and peripapillary choroid, particularly of the latter, was frequently patchy, and they filled completely only very slowly. In some eyes the filling of the choroid deteriorated towards the second half of the week before it improved.

(*ii*) During the second week, the PCA circulation started to improve, usually slowly, so that the choroidal and peripapillary filling could be seen in the late retinal arterio-venous phase. The improvement was slower in the peripapillary choroid than in the rest of the choroid; the filling there might not be complete till the retinal venous phase. This filling gradually improved as time passed, although the choroidal circulation might still not fill until the retinal arterio-venous phase by the end of the third week.

The watershed areas between the lateral and medial PCAs were usually the last to fill. The superior watershed zone generally showed the filling defect more frequently and longer than the inferior watershed zone (Fig. 3*c*).

(*2*) *In sectoral AION*

FFA in some of these eyes showed a filling defect in the choroid and peripapillary choroid, localized to the sector corresponding to the location of the AION, or more extensive; however, this was noticed in only a few eyes (Fig. 7*a*,*b*,*d*).

After about 2 to 3 months, in both (*1*) and (*2*) above, the choroidal circulation was usually restored to normal (Fig. 3*e*). Delay in filling persisted slightly longer in the peripapillary choroid than in the rest of the choroid.

The presence of a choroidal filling defect during the transit of the dye, when angiography was performed soon after the onset of AION, was highly correlated with ischaemia of the OD. Once the choroidal circulation had improved, this correlation disappeared. The choroidal circulation improved significantly within a week or even less, so that the presence

of occlusion might escape the notice of an investigator who did not perform FFA within the first few days after onset. There was a very high correlation between the OD ischaemia and peripapillary choroid filling defects during the transit of the dye in the initial stages of the AION. As the peripapillary choroid filling improved, this correlation also disappeared. The peripapillary choroid filling defects lasted longer than those in the choroid.

(c) RETINAL CIRCULATION

The retinal circulation was generally perfectly normal on angiography in these cases. It was involved in the following two eyes in this series:

(1) *Cilio-retinal artery occlusion*

In a patient with right AION, there was occlusion of a large cilio-retinal artery, supplying the upper half of the retina; in the central retinal artery, supplying the lower half of the retina, the circulation was normal (Fig. 3*a*). The filling of the cilio-retinal artery and the choroid improved significantly by the 4th day (Fig. 3*c*) and a perfectly normal choroid and cilio-retinal artery circulation was present when the patient was seen 8½ months later (Fig. 3*e*).

I have recently seen another similar patient (not included in this series), with a cilio-retinal artery supplying the lower half of the retina, who developed AION of the lower half of the optic nerve head and retinal infarction of the lower half of the retina, with an associated superior altitudinal field defect. His choroidal and cilio-retinal artery circulation was back to normal when he was seen 27 days after the onset of AION; however, the phenomenon of "hibernation of the blood in the retina" (Hayreh, 1971) in the distribution of the cilio-retinal artery was seen, *i.e.* the retinal arterio-venous phase was missing. In this latter patient the AION was of an arteriosclerotic nature, while in the first patient the AION was due to temporal arteritis.

These studies indicate that the so-called branch retinal arterial occlusion described with AION by various authors is in all probability an occlusion of a cilio-retinal artery and not a true branch retinal arterial occlusion (p. 975).

(2) *Central retinal artery occlusion*

AION involved the upper half of the right OD in a patient with temporal arteritis, and was associated with central retinal artery occlusion (Fig. 7). FFA on the third day after onset of visual disorder showed a normal filling of the lateral PCA in this eye (Fig. 7*a*), but the onset of filling of both the medial PCA and central retinal artery was delayed by 5 seconds (Fig. 7*b*), and their complete filling took more than one minute, with a retinal arterio-venous gap of 23 seconds (in the normal left eye of this patient the central retinal artery filled in 12 seconds with a retinal arterio-venous gap of 3 seconds). With the passage of time both the central retinal artery and medial PCA circulation improved (Fig. 7*d*). The mechanism of the involvement of the central retinal artery with the medial PCA in occlusive disorders has already been discussed (p. 975).

Within the first few days after the onset of the AION, the retinal venules over the swollen OD sometimes showed fluorescein leakage and some engorgement of the radial peripapillary capillaries.

These FFA findings in the OD, choroid, and retina in AION have no significant resemblance to the true oedema of the OD seen in intracranial hypertension and other conditions. FFA can thus help to differentiate AION from other types of swelling of the OD. As has been shown, the FFA pattern keeps altering as the interval between onset of AION and examination changes. This is an important factor to be borne in mind while interpreting these angiograms, to prevent unnecessary confusion.

(d) CHORIO-RETINAL DEGENERATIVE LESIONS IN AION

FFA of the lesions shown in Fig. 6 revealed degeneration of the pigment epithelium resulting in unmasking and masking of the choroidal fluorescence in the depigmented areas and

pigmented spots respectively, without late staining of the lesion. In AION patients, I have not so far seen any acute chorio-retinal ischaemic lesion of the type produced by us experimentally and studied by FFA (Hayreh and Baines, 1972).

Summary

In patients with anterior ischaemic optic neuropathy (AION), a detailed study has been made of the ophthalmoscopic and fluorescein angiographic (FFA) changes in the fundus. The main ocular abnormality on examination is in the optic disc (OD). In temporal arteritic AION, half of the patients show chalky-white swelling of the OD with a rare haemorrhage, while the other half show pink or pale-pink oedema of the OD with frequent flame-shaped haemorrhages. In the non-arteritic type of AION, it is rare to see chalky-white swelling of the disc, but the pale-pink or even hyperaemic oedema with haemorrhages is more common. The oedema of the OD usually starts to subside in about 7 to 10 days and optic atrophy develops after about a month or two. The oedema and atrophy may involve the entire disc or only a sector of it. The vast majority of ODs with AION due to temporal arteritis develop cupping of the disc; this is uncommon in cases without temporal arteritis. Rarely there may be associated infarction of a sector or of the entire retina because of additional involvement of the cilio-retinal or central retinal arteries respectively. The mechanism of this retinal arterial occlusion in AION is discussed. About a third of the cases with AION may show peripheral chorio-retinal degenerative patches. The pathogenesis of these patches is discussed.

On FFA, in complete AION, the OD shows no filling of vessels arising from the posterior ciliary artery (PCA) during the first and second weeks after the onset, or in the atrophic stage; some filling is seen during the intermediate period. In sectoral AION, staining is seen in only the normal part of the disc during the first week but in the whole disc towards the end of the second week, till the ischaemic part becomes atrophic and shows no fluorescence. Markedly delayed and poor filling of the choroid is seen during the first week, the filling defect being very pronounced in the peripapillary choroid; in sectoral AION this choroidal and peripapillary choroid filling defect is localized to the corresponding sector. After a few weeks or even earlier, the choroidal circulation is restored to normal. In the event of retinal arterial occlusion, angiography shows evidence of occlusion during the early stages.

I am grateful to many ophthalmologists and colleagues for referring these patients to me and for their invaluable help in this study; it would be invidious to make a special mention of any one of them. I also wish to thank my wife Shelagh for her help in the preparation of this manuscript and to Mrs. Maria Warbasse for her secretarial help.

References

BEGG, I. S., DRANCE, S. M., and GOLDMANN, H. (1972) *Canad. J. Ophthal.*, **7**, 68

————, ————, and SWEENEY, V. P. (1970) *Ibid.*, **5**, 321

————, ————, ———— (1971) *Brit. J. Ophthal.*, **55**, 73

BONAMOUR, M. G. (1966) *Bull. Soc. Ophtal. Fr.*, **79**, p. 846

————, BONNET, M., BREGEAT, P., and JUGE, P. (1968) 'La papille optique', p. 264. Masson, Paris

CULLEN, J. F. (1968) "Proc. Wm. Mackenzie Centenary Symp: The Ocular Circulation in Health and Disease", ed. J. S. Cant, p. 142. Kimpton, London

CÜPPERS, C. (1951) *Klin. Mbl. Augenheilk.*, **118**, 645

DRANCE, S. M. (1972) "Proc. 2nd Wm. Mackenzie Memorial Symp: The Optic Nerve", ed. J. S. Cant, p. 339. Kimpton, London

FOULDS, W. S. (1968) "Proc. Wm. Mackenzie Centenary Symp: The Ocular Circulation in Health and Disease", ed. J. S. Cant, p. 136. Kimpton, London

FRANÇOIS, J., VERRIEST, G., NEETENS, A., DE ROUCK, A., and HANSSENS, A. (1962) *Ann. Oculist. (Paris)*, **195,** 830

HAYREH, S. S. (1969a) *Brit. J. Ophthal.*, **53,** 721

———— (1969b) "Proc. Int. Symp. Fluorescein Angiograpy—Albi", ed. P. Amalric, p. 510. Karger, Basel

———— (1970) *Brit. J. Ophthal.*, **54,** 289

———— (1971) *Amer. J. Ophthal.*, **72,** 998

———— (1972a) *Brit. J. Ophthal.*, **56,** 175

———— (1972b) *Ibid.*, **56,** 652

———— (1973) *Trans. Amer. Acad. Ophthal. Otolaryng.*, **77,** 300

———— (1974a) *Brit. J. Ophthal.*, **58,** 955

———— (1974b) *Ibid.*, **58,** 863

———— and BAINES, J. A. B. (1972) *Ibid.*, **56,** 736

————, REVIE, I. H. S., and EDWARDS, J. (1970) *Ibid.*, **54,** 461

LARMANDE, A. M. (1948) *Rev. oto-neuro-ophtal.*, **20,** 483

LASCO, F. (1961) *Ophthalmologica (Basel)*, **142,** 429

MEADOWS, S. P. (1968) "Neuro-Ophthal. Symp. Univ. Miami and Bascom Palmer Eye Institute. IV.", ed. J. L. Smith, p. 148. Mosby, St. Louis

MILLER, S. J. H. (1972) *Trans. ophthal. Soc. U.K.*, **92,** 563

SANDERS, M. D. (1971) *Ibid.*, **91,** 369

SARAUX, H., and MURAT, J. P. (1967) *Ann. Oculist. (Paris)*, **200,** 1

SIEGERT, P. (1952) *Klin. Mbl. Augenheilk.*, **120,** 254

SIMMONS, R. J., and COGAN, D. G. (1962) *Arch. Ophthal. (Chicago)*, **68,** 8

SINGH, S., and DASS, R. (1960) *Brit. J. Ophthal.*, **44,** 193