Blind registration and glaucoma simplex

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Contributed by request and dedicated to Sir Stewart Duke-Elder

Although many aspects of glaucoma simplex have been the subject of attention in the last decade, there has been scant documentation of observations on the actual course of the disease when it leads to blindness (Frühauf, Müller, and Sismuth, 1967; Dake, 1967). A description of the glaucomatous course in such cases and the main factors influencing it form the subject of the present investigation.

Clinical material

- (a) The records of twenty patients who presented 10 years ago with glaucoma simplex were studied and their course described, with particular reference to the control of intraocular pressure.
- (b) An analysis of 34 patients registered as 'blind' or 'partially sighted' from glaucoma simplex was undertaken.

The present status of such patients was personally ascertained and facts about their presentation and course recorded.

Results

Twenty cases of chronic simple glaucoma

These patients, who were registered at this hospital during 1961 and 1962, had their subsequent course investigated in detail. They were divided into two groups (Tables IA and IB and II). The twelve patients in Tables IA and IB had intraocular pressures frequently over 25 mm. Hg by applanation despite therapy. The eight in Table II had pressures consistently below 20 mm. Hg on treatment.

Table IA shows that, of the twelve patients, eight became registrable as partially sighted or blind. None of those in Table II required registration.

An analysis of the registrable cases from Table IA was undertaken to assess the likely causes.

It can be seen that progressive glaucomatous visual loss, whilst under ophthalmic supervision, was the unequivocal dominant factor in causing 'blind' registration in two (Cases 2 and 6). Progressive glaucomatous visual loss also occurred in Cases 1, 3, 5, 7, and 8, but against a background of considerable reduction of visual reserve from trauma, cataract (Cases 1 and 3) and pre-presentation glaucomatous loss (Cases 5, 7, and 8). Late presentation significantly affected the present visual status of these three cases and also of Case 1. Little progression of glaucomatous field defect occurred in Case 4 despite uncontrolled pressures.

The total number of patients with a normal optic disc who presented in 1961-62 was five. In Cases 3, 9, and 11, the discs became pathologically cupped and there was associated

Table I Intraocular pressures frequently over 25 mm. Hg for 10 years

<i>Registrable as blind or partially sighted</i>	<i>Case no.</i>	<i>Age (yrs)</i>	<i>Status, 1961-62</i>		<i>Status, 1972</i>		<i>Comment</i>
A Yes	1	81	Anophthalmic (trauma)	6/24 Disc pale, myopic Field not charted	—	Hand movements Poor projection	Co-existence of cataract with glaucoma Progressed with miotics Also - 7D axial myopia
	2	72	6/6 Both discs cupped Full field	6/6 Arcuate loss	6/18 Arcuate loss	Hand movements	Operation refused Poor co-operation Poor medical control
	3	69	6/9 Disc cupped Arcuate loss	6/9 Disc normal Full field	6/24 Altitudinal loss	6/18 Disc cupped Large scotoma	Poor medical control Some lens opacities developed in both eyes
	4	71	Anophthalmic (trauma)	6/24 Disc not recorded Large arcuate loss	—	6/36 Poor disc view Field loss similar	Poor medical control Some lens opacity development
	5	61	6/6 Cupped disc Residual island	6/9 Cupped disc Full field	No perception of light	6/24 Altitudinal loss	Poor medical and surgical control
	6 (Negro)	49	6/12 Cupped disc Full field	6/6 Cupped disc Full field	No perception of light	6/24 Altitudinal loss	Poor medical and surgical control
	7	78	6/6 Cupped disc Residual island	6/36 Cupped disc Venous thrombosis	6/36 Island smaller	6/36	Medical control only Left eye had central venous thrombosis at presentation
	8	59	6/6 85% cupped Upper and lower arcuate scotomata both eyes	6/6 85% cupped	Perception of light	Hand movements	Poor medical and surgical control
B No	9	71	6/9 Disc normal Field full	6/6 pt Disc cupped Arcuate loss	6/9 Disc cupped Large arcuate scotoma	6/6 pt Disc cupped Large arcuate scotoma	Poor medical control Died in 1967
	10	80	6/9 Disc cupped Large arcuate scotoma	6/9 Disc cupped Large arcuate scotoma	6/12 Slightly larger scotomata	6/12 Large arcuate	Poor medical control (often 30+) Remarkably little progression
	11	79	6/6 Disc normal Field full	6/6 Disc cupped Arcuate loss	6/6 70% cupped Field full	6/9 80% cupped Arcuate loss same	Medical control only Pressures often 25 mm. Hg
	12	68	6/5 Cupped disc Arcuate scotoma	6/5 Cupped disc Field full	6/9 Arcuate scotoma same	6/6 Arcuate loss	Poor medical control High 20s usually

poor control of intraocular pressure. In Cases 13 and 17, where previously abnormal pressures were controlled, pathological cupping did not occur.

The only patient to become registrable in 10 years, who presented with a normal disc, was Case 3. Although there was definite cupping after 10 years of uncontrolled pressures, the visual defect leading to registration was partly due to lens opacity.

The average age at presentation of patients who became registrable as "blind" or "partially sighted" in 10 years, was 67 years (Table IA). The average age of those who did not progress to registration was 63 years (Tables II and IB). This difference is not statistically significant, so that age at presentation does not significantly affect the 10-year prognosis.

Any slight tendency for the non-registrable patients to be younger at presentation than those progressing to registration, is probably the result of younger patients adhering more closely to therapeutic regimes. Hence the average age of patients with pressure con-

trolled below 20 mm. Hg was 55 years (Table II) whilst that of those whose pressures were not controlled below 25 mm. Hg was 70 years (Tables IA and IB).

Table II *Intraocular pressures always below 20 mm. Hg for 10 years*

Case no.	Age (yrs)	Status, 1961-62		Status, 1972		General comment
13	56	6/5 50% cupped Full field	6/18 90% cupped Altitudinal loss	6/5 50% cupped Full field	Perception of light	Sudden postoperative loss of vision left eye Medical therapy only right eye
14	?	6/24 90% cupped Large arcuate scotoma	6/6 80% cupped Small arcuate scotoma	6/60 Discs same Field not charted	6/9 Field loss same	Bilateral trephines
15	49	6/5 Disc cupped Arcuate loss	Hand movements Disc cupped Field not charted	6/6 Field same	Hand movements Field not charted	Pressures around 12 mm. Hg after surgery (large blebs)
16	69	6/6 Cupped disc Full field	6/9 Cupped disc Full field	6/12 Full field	6/12 "Contracted" field	Bilateral drainage operations Left eye had central venous occlusion on presentation
17	56	6/9 Normal disc Full field	6/9 80% cupped Full field	6/9 Normal disc Full field	6/9 80% cupped Arcuate loss	Presented at glaucoma "sibling" clinic Control: right pressure below 20 mm. Hg; left peaked to mid-20s
18	39	6/9 Cupped disc Full field	6/5 Cupped disc Full field	6/6 Discs same Isolated scotomata both eyes	6/12	Medical control Thrombotic episode in left eye
19	56	6/6 Cupped disc Arcuate loss	6/9 Cupped disc Arcuate loss	6/9 pt Some increase arcuate scotomata both eyes	6/9	Good surgical control right Fair surgical control left (low 20s often)
20	64	6/6 Cupped disc Arcuate loss	6/6 Cupped disc Arcuate loss	6/9 Fields same	6/6 pt	Surgical control both eyes

Five patients had surgery and three were observed on medical therapy

34 patients registered "blind" or "partially sighted" from glaucoma simplex

All patients registered blind or partially sighted from glaucoma simplex in the last 3 years at Moorfields Hospital, High Holborn, were reviewed.

Although in many of the 34 cases of glaucoma simplex there were multiple causative factors leading to blindness, it was possible to single out the major cause in each case:

- (1) Late presentation (The criteria for this was extensive cupping and field loss in both eyes when first seen) 14 cases
- (2) Progressive field loss from glaucoma simplex whilst under ophthalmic supervision 7 cases
- (3) The formation of cataract or its sequelae 9 cases
- (4) The occurrence of vascular occlusive episodes 4 cases

(1) LATE PRESENTATION (14 cases)

Fourteen patients were registrable as "partially sighted" or "blind" on presentation.

The classical "silent" loss of sight in glaucoma simplex may well be the major culprit in this group. However, other notable features were observed:

(a) Neglect of symptoms (6 cases)

One patient had symptoms for 8 years, three for 2 years, one for 1 year, and one for 4 months, all before seeking medical attention.

(b) Delayed referral to an ophthalmologist (4 cases)

Presentation to a general practitioner, referral to an optician, return to the general prac-

tioner, and thence to a hospital department with a waiting-list for outpatient appointments resulted in delays. There was a delay of 6 months in two cases and of 10 months in one case, and one case was not referred to hospital by his general practitioner for 2 years.

(c) *Mistaken or missed diagnosis* (4 cases)

After presentation to an ophthalmologist, the diagnosis was missed for 14 months in one case, 9 months in one case, and 6 months in two cases. It is notable that in these cases the disease was advanced when eventually diagnosed.

Seven patients in this group were over 60 years of age and had had repeated refraction for several years before hospital attendance.

(2) **PROGRESSIVE FIELD LOSS** (Table III) (7 cases)

These seven patients with glaucoma simplex became sufficiently visually handicapped to merit registration because of progressive glaucomatous field loss whilst under ophthalmic supervision.

(a) Three of them failed to return for clinical examination (Cases 7 and 10) and one refused surgery (Case 8).

(b) In four patients (Cases 7, 8, 10, and 24) progression of field loss was associated with raised intraocular pressure. In two (Cases 9 and 33) the intraocular pressures were usually below 20 mm. Hg but rose to peaks of 23 and 30 mm. respectively occasionally.

(c) Three patients (Cases 7, 24, and 33) had a normal disc on presentation and yet progressed to registration in 5½, 7, and 9 years respectively. The intraocular pressures in Cases 7 and 24 were grossly out of control. Case 33 had progressive cupping and field loss whilst pressures were usually under

Table III *Progressive field loss group*

Case no.	Age (yrs)	Presentation status		Registration status		Interval before registration (yrs)	Comment
7	50	6/6 "New vessels" on disc Full field	6/6 Disc normal Full field	6/9 Cup: disc ratio 0.9 both eyes Bilateral tiny central fields	6/9	5½	Negro Relapsed from clinic 1½ yrs Presenting tensions 50 mm. Hg Low I.Q. No surgery
8	62	Not available		6/18 Fully cupped discs Tiny central fields	6/18	10½	Operation refused Vague, poor witness Pressures not controlled medically
9	73	Hand movements Cupped disc Field loss + + +	6/12 Cupped disc Full field	No perception of light	6/12 Altitudinal field loss	5	Pressures usually under 20 mm. Hg peaking sometimes to 23 No surgery
10	50	6/12 Cupped disc Full field	6/6 Cupped disc Full field	No perception of light	6/24 Altitudinal field loss	9	Negro Relapsed clinic 5 yrs Pressures 30+ after surgical and medical therapy
24	72	6/12 Discs normal Full field	6/9 Full field	6/60 Discs fully cupped Field loss involving fixation	6/60	7	Presenting pressures 35 mm. Hg—always 25+ No surgery
32	73	Hand movements 80% cupped Gross field loss	6/12 80% cupped Full field	Perception of light Large arcuate scotomata Splitting maculae	6/12	5	Medical therapy Pressure control not available
33	52	6/9 Disc normal Field full	6/12 Cupped disc Venous thrombosis	6/12 Small central field Disc cupped (left thrombotic glaucoma)	Enucleation	9	Pressures mostly 20 mm. Hg but fluctuated between 14 and 30 No operation

20 mm. Hg, but medical therapy was considered less risky than surgery in this uniocular patient, who can still see 6/12 9 years after presentation.

(3) CATARACT AND GLAUCOMA SIMPLEX REGISTRATION

(a) Nine patients in this group qualified for "blind" or "partially sighted" registration because of cataract in addition to glaucoma. In three cases cataract extraction was advised after this investigation. In other cases the lens opacities are reducing vision to 6/18–6/24 in one or both eyes and co-exist with extensive glaucomatous field loss in one eye only. These patients are considered to be coping adequately visually at present and not to merit cataract extraction. One registered patient prefers uncorrected aphakic vision to 6/9 with a very high cylinder.

(b) There were 14 cases altogether in which cataract and glaucoma co-existed. In nine of these, cataracts co-existed with glaucoma on presentation and increased after medical or surgical therapy. In the other five cases cataracts developed during the period of medical or surgical supervision.

(4) VASCULAR EPISODES (4 cases)

These did not form a sufficiently large group to merit further analysis.

Discussion

Several observations have led clinicians to question the role of raised intraocular pressure in the causation of glaucomatous visual field loss. Thus progressive field loss and cupping of optic discs have been observed frequently in the "low tension" glaucomas, when intraocular pressures have consistently been below 21 mm. Hg. Glaucomatous field loss causing blindness and gross cupping of the optic discs have also been observed frequently, with intraocular pressures never higher than the mid-twenties. Such pressures are known to occur in the normal physiological range without causing any change at the optic disc, or any functional loss. Finally, the rate of progress of field loss in glaucoma simplex patients does not always correlate well with the degree of control of intraocular pressures.

Many of these features are seen in 1 year's presentation of patients with glaucoma simplex at this hospital (Tables I and II). Cases 4, 10, 11, and 12 showed relatively little progression of field loss over 10 years despite obviously uncontrolled intraocular pressures. With the exception of Case 11 (right eye), all the other discs were pathologically cupped initially and this has been thought to increase susceptibility to field loss. Conversely, Cases 18 and 19 showed a small increase in field loss despite pressures being well controlled. Case 33 (Table III) had a normal disc with a full field which progressed to gross cupping and a central island of vision, when the intraocular pressures were usually 12 to 14 mm. Hg.

A non-causative association of raised intraocular pressure with disc cupping and field loss in glaucoma, has been postulated by several authors (Duke-Elder, 1969). Armaly (1969) wrote "The relationship between ocular pressure level and its dynamics on the one hand and the development of glaucomatous visual function loss on the other, continues to head the list of unresolved questions in open-angle glaucoma".

It is possible that a single pathological process, such as occlusive vascular disease, affects both the aqueous outflow mechanisms and the optic disc. This would result in an incidental relationship between raised intraocular pressure and glaucomatous sequelae. The evidence from Tables I and II does not refute this possibility but it does strongly suggest that the intraocular pressure, when allowed to remain high, increases the rate of disc damage. Thus, despite the individual exceptions, a detailed comparison of Table I and II shows that the "pressure controlled" group fared much better.

In Table IA are cases in which pressures were uncontrolled and eight progressed to registration as partially sighted or blind. This gives an erroneous emphasis on uncontrolled

pressures, as two of these patients were registrable at presentation, and in four others the progressive glaucomatous loss occurred against a background of diminished visual reserve from trauma, cataract, and pre-presentation glaucomatous loss. However, despite the presence of these factors exaggerating the different progress in the two groups, comparison does show the important part played by intraocular pressure. Table II shows that, by lowering the intraocular pressure effectively, the progression of glaucomatous visual loss is at least slowed and can even be arrested.

A relationship between cupping of the disc and raised intraocular pressure is also evident. The three discs which were normal on presentation (Table I: Cases, 3, 9, and 11) became pathologically cupped when associated with uncontrolled intraocular pressure. In Cases 13 and 17 (Table II), where previously abnormal pressures were controlled, cupping did not occur.

It may be that, in the eyes with pathologically cupped discs, poor control of intraocular pressure, and yet little progression of field loss in 10 years (Table I: Cases 4, 10, 11, and 12), it is the actual degree of lowering of intraocular pressure achieved that is important. The blood supply to the discs in such cases may withstand some degree of elevation of intraocular pressure. In these eyes, the outflow mechanism is probably the major site of primary abnormality, not the disc. In some of the low tension glaucomas, and in cases such as 14, 17, 18, and 19 (Table II), where field loss has progressed with low pressures, the blood supply to the disc is probably the major site of primary abnormality. In many cases of glaucoma both sites could be pathologically involved, and raised pressure would then act as an exacerbating but not as an isolated causative factor. The important practical consideration is that lowering of intraocular pressure will increase disc perfusion in all cases, and the greater the fall in intraocular pressure the larger the increase in perfusion. It may therefore be more beneficial to reduce a pressure of 40 mm. Hg to 25 mm. Hg than to reduce a pressure of 20 mm. Hg to 15 mm. Hg. As it is more difficult to obtain a large reduction in intraocular pressure if the initial pressure is low, the possibility of improving the disc perfusion is much less.

This series shows also that the glaucomatous course can be benign when the patient presents with considerable visual reserve and especially when intraocular pressures are effectively reduced (Table II). Dake (1967) emphasized the benign course of many cases of glaucoma simplex, especially when one disc was not pathologically cupped. Cases 1, 11, 14, 20, and 21 in his series of 29 cases are notable in this respect.

Frühauf and others (1967), in a large series of 920 cases followed over 16 years, found that visual deterioration was much less in cases presenting early. Only 3 per cent. showed deterioration in 5 years. In the patients with full fields, however, their criteria for diagnosis of glaucoma simplex is suspect and it is surprising that they could exclude cataract formation in so many cases during such a long period of observation.

Severe visual incapacity can follow in 5, 7, or 9 years (Table III: Cases 7, 24, and 33 respectively), even when the discs are normal on presentation. However, for different reasons, these patients did not receive the fullest possible glaucoma therapy.

Late presentation was the dominant aetiological factor in the registration of fourteen cases out of the 34 studied. These patients were registrable as partially sighted or blind at presentation. To this must be added the finding that late presentation enhances subsequent progress to visual disability (Frühauf and others, 1967).

It is this pre-presentation "silent" loss of vision which has stimulated attempts to devise adequate glaucoma screening techniques in the last decade. Of the 53 patients studied here, 49 could have been diagnosed from disc appearances alone. Only Case 24 (Table III)

had normal discs. Data for Case 8 (Table III) was not available, and Case 1 (Table I) was difficult to evaluate because of myopia and lens opacity.

It is remarkable that in only seven out of 34 patients registered as "blind" was progressive field loss the dominant factor (Table III). Uncontrolled pressures have been shown to play a prominent part in this group. These cases suggest that registrable blindness from isolated glaucomatous damage to eyes, once under supervision, accounts for only about one-fifth of cases. Progressive glaucomatous visual loss may occur in most cases, especially if pressures are not controlled (Table I), but this usually leads to registrable levels only if visual reserves have already been seriously reduced at presentation.

A surprising finding is the prominent part played by cataracts in the development of visual incapacity in glaucoma patients. This was noted by Roberts (1970). The co-existence of the two conditions is well known, as they occur in the same age group (Becker and Shaffer, 1965).

Axelsson (1969) could find no evidence coupling the degree of glaucoma in the two eyes with the degree of cataract. Nine out of our 34 cases of glaucoma already had cataracts affecting visual acuity on presentation. With the important age factor controlling the incidence of cataract in non-glaucomatous and glaucomatous populations, these figures are too small to be of any significance in trying to establish a more than incidental relationship. However, miotics are known to cause, and to worsen, pre-existing cataracts (Axelsson and Holmberg, 1966; Shaffer and Hetherington, 1966). Likewise, all forms of glaucoma surgery result in a higher incidence of cataracts (Leydecker, 1966; Shaffer and Hetherington, 1968).

The summation of all three factors explains the prominence of cataracts in causing visual incapacity in glaucoma patients who are registered as blind.

Summary

- (i) Control of intraocular pressure has been found to exert a beneficial influence in cases of glaucoma simplex over 10 years.
- (ii) Late presentation was responsible for nearly half of the registrations of "blind glaucoma" in the cases studied.
- (iii) There is a good prognosis for patients who present with significant visual reserve, and in whom the ocular tension is controlled. There is a bad prognosis for patients with chronic simple glaucoma who are indifferently controlled, particularly if their visual reserve is poor.
- (iv) Senile cataracts and cataracts secondary to medical therapy or surgery contribute significantly to visual morbidity in glaucoma patients.

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