

Is Posner Schlossman Syndrome Benign?

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Purpose: To determine the clinical course of patients with Posner Schlossman syndrome (PSS).

Design: Retrospective noncomparative case series.

Participants: Fifty-three eyes of 50 patients.

Methods: The case notes of all patients with PSS seen at the Uveitis Clinic of Singapore National Eye Centre were reviewed for evidence of glaucoma damage and risk factors.

Main Outcome Measures: Visual field and optic disc changes consistent with glaucoma.

Results: There were 28 men and 22 women, and their mean age at onset was 35 years. Fourteen eyes (26.4%) were diagnosed to have developed glaucoma as a result of repeated attacks of PSS. Patients with 10 years or more of PSS have a 2.8 times higher risk (95% confidence interval 1.19–6.52) of developing glaucoma compared with patients with less than 10 years duration of the disease. Nine eyes (17%) underwent glaucoma filtering surgery with antimetabolites. Their postoperative follow-up ranged from 15 to 50 months (mean, 37 months). Four eyes continued to have episodes of iritis after surgery, and one of these eyes had elevated intraocular pressure during the event.

Conclusions: A significant number of patients with PSS have glaucoma develop over time, and they need to have their optic disc appearance and visual fields carefully monitored. *Ophthalmology* 2001;108:913–918
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The Posner Schlossman syndrome (PSS) as originally described in 1948 by Posner and Schlossman is a self-limiting and benign condition characterized by unilateral, recurrent attacks of mild, nongranulomatous iritis with elevated intraocular pressures (IOP) during the acute attack, open angles, normal visual fields, and optic discs. In between the attacks, the IOP, tonography and provocative tests were within normal limits.¹ Later studies have shown that there is an association between primary open-angle glaucoma (POAG) and PSS, with as many as 45% of PSS patients having concomitant POAG.^{2,3}

Aside from the association with POAG, the development of glaucomatous damage from PSS itself has also been noted.⁴ During the many years of follow-up of our patients in the Uveitis clinic, we, too, had noticed glaucomatous loss of vision in some of our PSS patients. Because PSS normally occurs in younger individuals between the third to sixth decade of life, this potential for loss of vision from glaucoma is an important factor in the management of these patients. Hence, we reviewed our patients with PSS to

determine the risks of glaucomatous damage from PSS itself without coexisting POAG.

Methods

A retrospective review of the case notes of all patients with PSS seen at the Uveitis Clinic of the Singapore National Eye Centre was carried out. Because the Uveitis Clinic is a referral center, the case notes reviewed included both the notes from the time the patients were seen at the Uveitis Clinic and those from the referring physicians. The following data were recorded: the duration of the disease, age of the patient at onset, number of documented attacks, presence of systemic disease, highest IOP during the attack, IOP in between attacks and in the nonattack eye, optic disc appearance, visual field changes, glaucoma surgery, episodes of cyclitis, and IOP during attacks that occurred after surgery.

The diagnosis of PSS was made on the basis of the features described in Table 1. The anterior chamber inflammation is usually mild, with at most 1+ cells and trace flare. Keratic precipitates are always present. They are usually small with one to two medium-sized ones, and nonpigmented. They are found mainly on the endothelial surface of the central cornea with a few scattered inferiorly. Posterior synechiae are never present. There is never any vitreous activity, and cystoid macula edema does not occur. The attacks usually resolve by 2 to 3 weeks, sometimes spontaneously, and are recurrent. In between attacks, the eyes are normal, although a few eyes continued to have one or two small nonpigmented keratic precipitates. Other causes of acute anterior uveitis, such as idiopathic anterior uveitis, with secondary glaucoma and herpetic uveitis would have more inflammation and a longer duration of activity and do not resolve readily without treatment. IOPs were measured using Goldmann applanation tonometry. A patient was diagnosed as having glaucoma develop from attacks of PSS if:

1. The IOP in the affected eye was 21 mmHg or less in between attacks

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Table 1. Features of Posner Schlossman Syndrome

- 1 Unilateral
- 2 Recurrent episodes of mild nongranulomatous cyclitis
- 3 Symptoms of mild discomfort, halos, and slight blurring of vision
- 4 Findings of corneal edema, elevated IOP, open angles, fine keratic precipitates, few cells and minimal flare, and mydriasis in affected eye
- 5 Attacks last few hours to few weeks
- 6 Normal visual fields and optic disc
- 7 Normal IOP, tonography, and provocative tests between attacks

IOP = intraocular pressure.

2. The unaffected eye had normal IOP, optic discs and fields
3. The affected eye had glaucomatous optic disc and/or visual fields

All patients had at least one visual field performed on the Humphrey Field Analyzer (Humphrey Instruments, Inc, San Leandro, CA) using the 24-2 or 30-2 program. The criteria, in a reliable reproducible field with less than 15% false-negative and less than 15% false-positive responses, used for the diagnosis of glaucoma were⁵:

- A cluster of two or more central points depressed by 5 or more decibels
- A single point depressed by 10 or more decibels
- A difference of 5 or more decibels across the nasal horizontal meridian at two or more contiguous points

Optic disc criteria for glaucoma included⁶⁻⁹:

- Cup/disc ratio greater than 0.7
- Notching of neuroretinal rim or focal pit
- Narrowest remaining neuroretinal rim of 0.1 disc diameter or less
- Asymmetry in cup/disc ratio of more than 0.2
- Progressive thinning of the neuroretinal rim
- Increase in cup/disc ratio

The first four criteria are not in themselves diagnostic of glaucoma, and eyes with these changes were graded as having glaucoma only if they had concomitant field changes.

The field and disc photographs were assessed by one of the authors according to the criteria listed, and only those that had obvious changes were classified as having glaucoma damage. Not all the patients had good sequential stereoscopic disc photographs, and disc changes were given relatively low priority in the assessment.

Results

There were 53 eyes of 50 patients, with three of the patients having bilateral attacks, and these eyes were analyzed separately. Their mean age at onset was 35 years (range, 17-61 years). Twenty-two of the patients were women and 28 men. Most were Chinese (n = 45), with 4 Malays, and 1 Indian. Only three patients had diabetes mellitus. The mean duration of their disease was 7 years (range, 1-24 years). All had at least three documented attacks of PSS. None of eyes required or received any steroids or antiglaucoma therapy in between attacks.

Fourteen eyes (26.4%) had glaucomatous damage develop because of repeated attacks of PSS. Two of these eyes belonged to the same patient. All these patients were Chinese except for one Malay.

Eleven of the eyes had normal discs and fields initially, but over a mean follow-up period of 10 years (range, 2-20 years) they were observed to have glaucomatous damage develop. Ten of these eyes had both disc and field changes. In the eleventh eye, there was an increase in the cup/disc ratio from 0.3 to 0.5, with no field changes, whereas in the normal fellow eye, the cup/disc ratio remained at 0.2 throughout the period of follow-up.

Three of the eyes already had significant field and disc changes by the time that they presented to an ophthalmologist (Table 2).

The age of onset, highest IOP, duration of the disease, and number of attacks among the patients who had glaucoma develop and those who did not were compared by use of Student's *t* test.

Table 2. Characteristics of Patients with Posner Schlossman Syndrome Who Had Glaucoma Develop

Patient	Sex	Age at Onset (yrs)	Duration of Disease (yrs)	Highest Intraocular Pressure (mmHg)	Number of Attacks	Initial Cup/Disc Ratio*	Last Cup/Disc Ratio†	Visual Field	Glaucoma Surgery
1	F	49	1	50	3	0.8	0.8	Superior hemifield loss	5FUTrab‡
2	M	56	2	40	4	0.3	0.5	Arcuate scotoma	Nil
3	M	60	3	35	5	0.9	1	Central island	Nil
4	M	26	3	50	6	0.3	0.5	Paracentral scotoma	5FUTrab
5	F	42	3	58	11	0.7	0.8	Superior and inferior arcuate loss	Nil
6	F	39	5	64	4	0.4	0.7	Nasal step	Nil
7	M	36	9	44	12	0.6	0.9	Superior hemifield loss	MMCTrab§
8	M	36	10	50	9	0.5	0.8	Nasal step	5FUTrab
9	M	37	10	32	9	0.4	0.5	Superior arcuate	Nil
10	M	33	13	70	17	0.5	0.7	Nasal step	5FUTrab
11	M	33	16	60	14	0.3	0.5	Normal	5FUTrab
12 right eye	M	30	19	36	7	0.4	0.9	Superior and inferior nasal step	5FUTrab
12 left eye	M	30	19	40	9	0.6	0.8	Superior and inferior arcuate defects	5FUTrab
13	F	29	23	52	19	0.4	0.8	Nasal step	5FUTrab

*Initial C/D = cup/disc ratio at diagnosis.

†Last C/D ratio = cup/disc ratio at last visit.

‡5FUTrab = trabeculectomy with intraoperative application of 50 mg/ml 5-fluorouracil.

§MMCTrab = trabeculectomy with intraoperative application of 0.3 mg/ml mitomycin-C.

Table 3. Factors in Development of Glaucoma in Patients with Posner Schlossman Syndrome

Factor	Glaucoma	No Glaucoma	P
Mean age at onset (yrs)	38	34	ns
Mean duration of disease (yrs)	10	5	0.04
Mean highest IOP (mmHg)	49	46	ns
Mean number of attacks	9	8	ns

IOP = intraocular pressure; ns = not statistically significant at 95% level.

The only factor, that was predictive of the development of glaucoma was the duration of the disease (Table 3). The risk of glaucoma developing after 10 or more years of exposure is 2.8 times higher (95% confidence interval, 1.19–6.52) than in those with less than 10 years of the disease.

Nine of the eyes (17%) had progressive damage and underwent glaucoma filtering surgery. One eye received a trabeculectomy with mitomycin-C and the other eight received a trabeculectomy with intraoperative application of 5-fluorouracil (50 mg/ml applied for 5 minutes). Their mean postoperative follow-up was 37 months (range, 15–50 months). Their mean IOP at the last visit was 10 mmHg (range, 6–15 mmHg). One of the eyes required two antiglaucoma drops; the rest of the eyes were on no medications.

In the year before surgery, these nine eyes had three to four attacks of iritis each, but in the year after surgery, six eyes had no further attacks of iritis. In the three eyes that continued to have episodes of iritis after surgery, two eyes had one attack each with no elevation of IOP during the attack and one eye had 15 attacks with elevated IOP during the attacks. This has resulted in failure of the initially functioning bleb with progression of the glaucoma damage. The other eight eyes had avascular functioning blebs at the last visit, and there has been no further glaucomatous damage in these eyes.

Fifty of the eyes had 6/9 or better vision at the last visit, two eyes had 6/12 vision because of cataract, and one eye had lost light perception because of progression of his glaucoma.

Case Reports

Patient 12—Right and Left Eyes

This Chinese man was first seen in 1979 at the age of 30 with evidence of PSS in both eyes. He continued to have nonsimultaneous acute attacks about once a year. The highest IOP in his right eye was 36 mmHg and in his left eye was 40 mmHg. In 1994 his cup/disc ratio was noted to be 0.8 in the right eye and 0.6 in the left eye. Visual field testing at that time showed severe superior arcuate and inferior nasal loss in the right eye and a small nasal step in the left eye (Fig 1A,B). He received a right trabeculectomy with 5-fluorouracil in January 1995, and the field in the right eye has remained stable since then.

In the meantime, he continued to have repeated attacks of cyclitis, accompanied by IOP spikes in the left eye, and he underwent trabeculectomy with 5-fluorouracil in March 1995 in that eye. The visual field of the left eye in January 1996 showed extensive superior and inferior arcuate loss. Because his IOPs had remained between 8 and 10 mmHg during this time, the damage presumably occurred as a result of the preoperative episodes of cyclitis. The visual field in March 1998, 5 months after cataract surgery, showed essentially the same pattern of loss (Fig 1C–E). Postop-

eratively, his IOP ranged from 7 to 14 mmHg in both eyes with no antiglaucoma medication. He had one episode of mild iritis in both eyes, but there was no spike of IOP.

Patient 7

This Chinese man was seen in April 1989 at the age of 36, with misty vision of the right eye, headache, and ocular discomfort. His vision was 6/6 in both eyes. There was mild epithelial edema of the cornea with a few keratic precipitates and cells seen on slit-lamp examination of the right eye. The IOP was 36 mmHg. Gonioscopy showed open angle. The left eye was normal. He had nine episodes of PSS crises over the next 6 years, with a highest IOP of 44 mmHg. In 1995 his cup/disc ratio was noted to be 0.6 in the right eye and 0.3 in the left eye. Visual fields in January 1996 showed a superior arcuate defect (Fig 2A). He had another three attacks after that, and in December 1997 the field loss in the right eye had progressed to a dense superior hemifield defect (Fig 2B). He underwent a trabeculectomy with mitomycin-C in January 1998, and the IOP has been less than 18 mmHg on no medications. He has had one attack since, with no elevation in IOP.

Discussion

One fourth of the eyes in our series of PSS patients had glaucomatous damage. None of these eyes had evidence of coexisting POAG. The major determinant of occurrence of glaucoma was the duration of the disease. Because this is a retrospective study, it was not possible to accurately determine the duration of elevated IOP during each episode and the number of attacks that the patients had had. Although these patients were followed up at 6 month intervals and asked to return earlier if they had an attack, some of the patients may not have sought treatment for every attack, because they were short-lived. Hence, we could not conclusively exclude the possibility that the duration of the attack and number of attacks were also factors in causing glaucoma in these patients.

Although these patients were from the Uveitis Clinic, they constitute more than 90% of all PSS patients seen at the Singapore National Eye Centre and therefore are fairly representative of the group.

Our longitudinal study shows that PSS in our population is not as benign as was previously thought. This was also observed in another study, which again involved a predominantly Chinese population.⁴ Although this has not been described so far among PSS patients of other races, we recommend that all patients with PSS need to be carefully examined for evidence of glaucoma even in the absence of associated POAG.

This study also shows that glaucoma filtering surgery with antimetabolites is successful in preventing IOP spikes during the cyclitic attacks in most cases (80%). There also seems to be a reduction in the number of attacks that these individuals have after surgery. This decrease in the severity of the uveitis has also been noted by others,^{10,11} and we suggest that it is possible that a functioning filtering bleb may allow egress of inflammatory cells from the eye, and this may in turn protect the bleb from failure. This would be in keeping with the observation by Hill et al¹² that in patients with chronic uveitis who had received Molteno

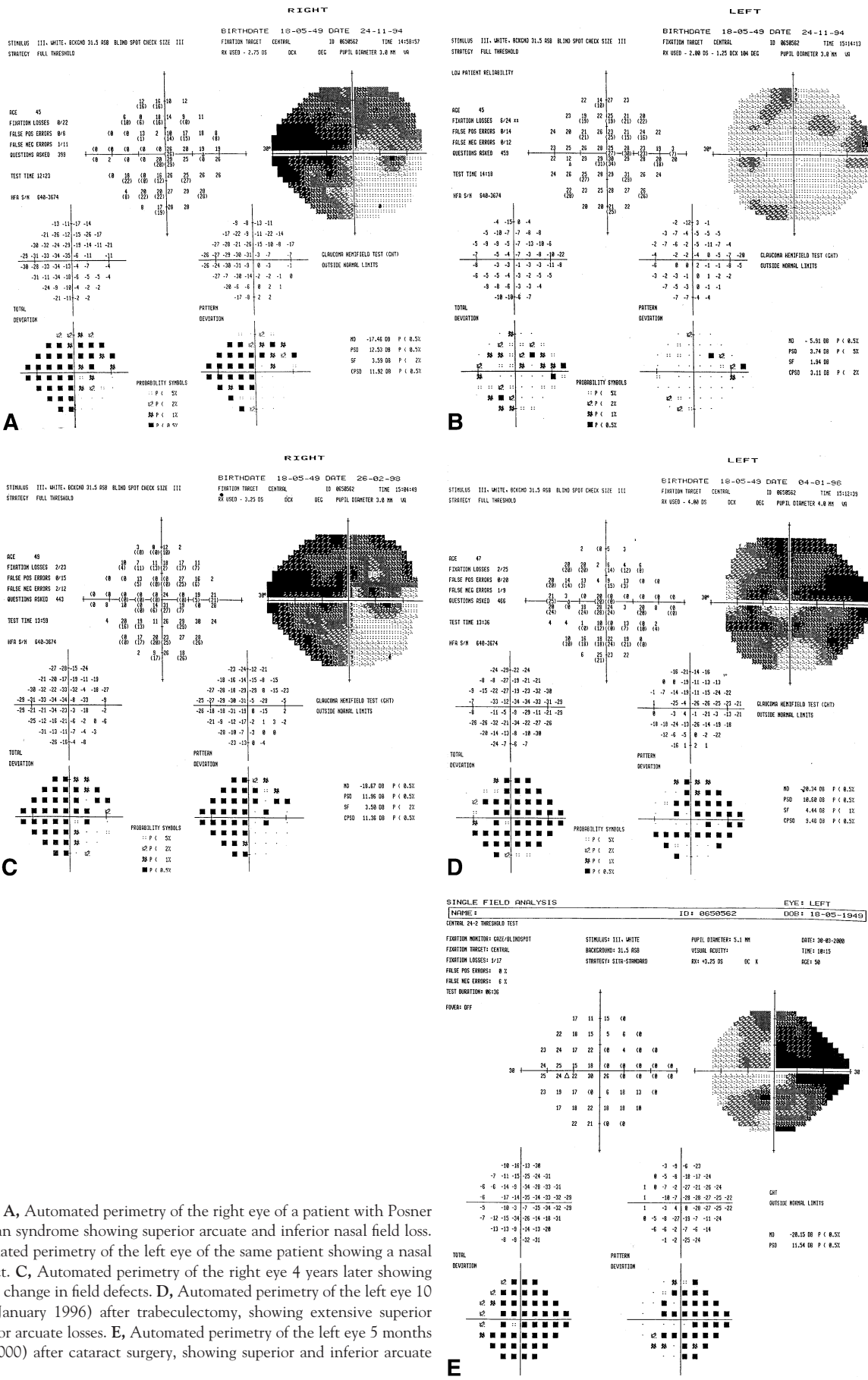


Figure 1. A, Automated perimetry of the right eye of a patient with Posner Schlossman syndrome showing superior arcuate and inferior nasal field loss. B, Automated perimetry of the left eye of the same patient showing a nasal field defect. C, Automated perimetry of the right eye 4 years later showing no further change in field defects. D, Automated perimetry of the left eye 10 months (January 1996) after trabeculectomy, showing extensive superior and inferior arcuate losses. E, Automated perimetry of the left eye 5 months (March 2000) after cataract surgery, showing superior and inferior arcuate losses.

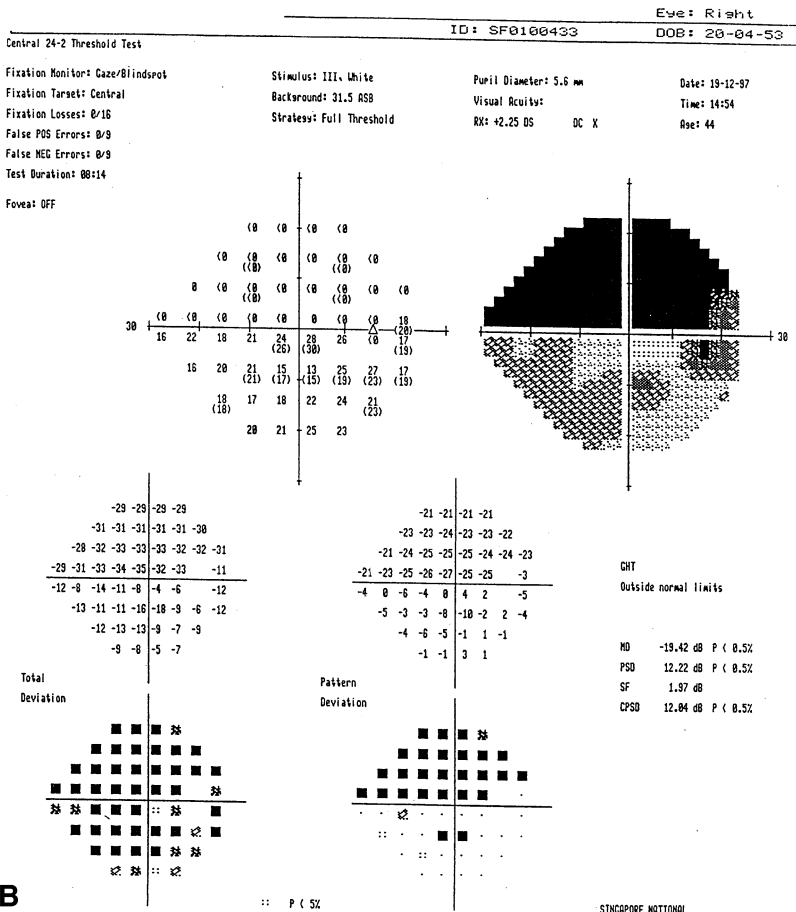
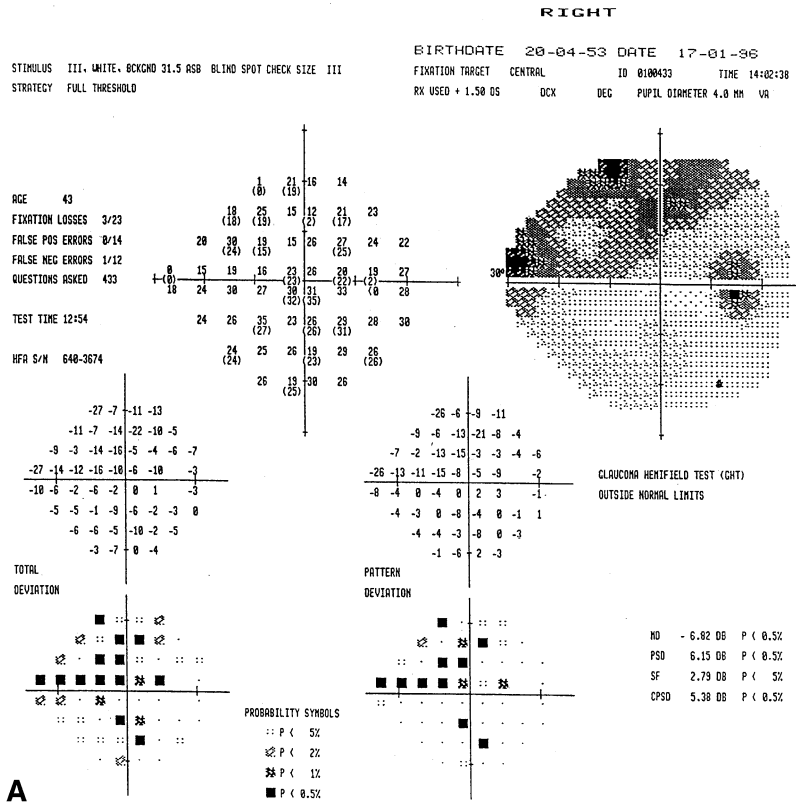


Figure 2. A, Automated perimetry of the right eye of a patient with Posner Schlossman syndrome showing a superior arcuate field defect. B, Automated perimetry of the same eye 2 years later showing worsening of field loss with a dense superior hemifield defect.

implants, the drainage tubes removed the inflammatory cells from the anterior chamber. However, again in a retrospective study, there may have been missed episodes, because these patients may now have less severe symptoms and therefore not seek treatment.

Furthermore, glaucoma surgery should be advised as soon as glaucoma change has occurred, because the rate of progression can be quite rapid in these individuals.

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