GONIOSCOPY AND TONOGRAPHY IN GLAUCOMATOCYCLITIC CRISES*†

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THE syndrome of recurrent glaucomatocyclitic crises was first clearly delineated by Posner and Schlossman (1948), who defined the characteristics of this disorder as follows:

- (1) The condition is unilateral.
- (2) The eye remains white or at most only slightly congested during an attack.
- (3) Subjective symptoms are minimal and vision is usually not impaired to any degree.
- (4) Signs of cyclitis do not precede the glaucoma; posterior synechiae are never formed.
- (5) Attacks may last from a few hours to a month, but rarely more than 2 weeks.
- (6) Between attacks the eye shows no signs of glaucoma or cyclitis.
- (7) Treatment with pilocarpine or atropine does not shorten the attack.

All our patients fulfill these diagnostic criteria and their case histories will not be described in detail. Seven consecutive cases of glaucomatocyclitic crises are described in which bilateral abnormalities of the chamber angle were found on gonioscopy.

In their first report Posner and Schlossman (1948) found gonioscopically open angles in four of twelve patients and failed to comment on the angle appearances of the remaining eight. Levatin (1956), in a report of one case which showed bilateral involvement, stated that the angles were gonioscopically open. Sokolić (1966) has recently described bilateral angle anomalies in a male aged 42 who had suffered from glaucomatocyclitic attacks in the right eye for some years. According to his description the involved right eye showed

"a wide open angle, the insertion of the iris root at the ciliary body was visible; the line of insertion of the iris was irregular. Areas of stromal hypoplasia with denuded vessels and visible stratum pigmenti iridis were present at the periphery of the iris. The ciliary body band was covered with a rough sheet of greyish white tissue which was spread over the posterior trabecular border in an irregular line. The trabecular band was not translucent and showed sporadic areas of small abnormal vessels. Schwalbe's line was prominent".

The uninvolved eye which had "moderately raised intra-ocular pressure" showed "obliteration of the angle recess with iris processes. . . . These processes together with iris tissue were inserted at the posterior border of the trabecular band . . . so as to imitate insertio iridis anterior". Sokolić concluded that glaucomatocyclitic crises "might be accordingly considered as a particular form of late developmental glaucoma". The present series supports this conclusion.

Several authors have postulated that hypersecretion of aqueous is a factor in the clinical picture of the glaucomatocyclitic crisis. Sokolić in his one case report found the facility of outflow, *C*, unimpaired during an attack. Spivey and Armaly (1963), who performed outflow studies on three patients in crisis, found low *C* values and from these values

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calculated the rates of aqueous flow F which they considered were evidence of hypersecretion. The present authors' findings in three patients during crisis were similar to those of Spivey and Armaly but we do not agree with their interpretation of the data.

Case Reports

The visual acuity, applanation, and C value in our series of seven cases are given in Table I.

TABLE I
PARTICULARS OF SEVEN CASES OF GLAUCOMATOCYCLITIC CRISIS

Case No.	Age (yrs)	Visual Acuity and Correction		Eye Involved	Applanation (mm. Hg)		C value*	
		R	L		R	L	R	L
1	23	6/5 -1.5 D sph.	6/12 -1.5 D sph.	L	14	16	0.38	0.32
2	44	6/5 -0.75 D sph. +1.25 D cyl. axis 90°	6/5 -0.75 D sph. +1.0 D cyl. axis 90°	L	23	22	0.33	0.43
					21	32	0.29	0.15
3	41	6/5 +1.25 D sph. -1.0 D cyl. axis 70°	6/5 +0.25 D sph. -0.5 D cyl. axis 90°	R	10	12	0.15	0.17
4	55	6/12 +1 D sph. -1 D cyl. axis 60°	6/12 +1 D sph. -1 D cyl. axis 120°	R	14	18	0.12	0.17
5	38	6/12 +1.5 D sph. -2.75 D cyl. axis 100°	6/4 +1.0 D sph. -1.25 D cyl. axis 60°	R	14	14	0.15	0.11
				R (in crisis)	50		0.07	
6	36	Emmetrope 6/6	6/6	R R (in crisis)	14 45	14 16	0.12 Flat trace	0.12
7	21	Emmetrope 6/4	6/5	L	17	14	0.12	0.07

* C was calculated from Tables based on the 1955 calibration of Friedenwald.

Case 1, a "ladle man" aged 23 years, had had recurrent attacks of glaucomatocyclitic crisis affecting the left eye for approximately 4 years. The attacks lasted 24 to 48 hours; there were no exciting factors.

External examination was normal. The intra-ocular pressure in the left eye during an attack rose to 70 mm. Hg (Schiotz). The pupil was dilated. The pressure returned to normal in 24 hours on topical steroids.

Gonioscopy:

Right. Minimal angle recess, many fine iris processes, slight pigmentation of trabecula, no blood in Schlemm's canal, Schwalbe's line normal.

Left. Anterior insertion of iris to posterior edge of corneo-scleral meshwork obliterating the angle recess. Occasional iris processes. Minimal pigmentation. Blood in Schlemm's canal. Schwalbe's line normal.

Case 2, a housewife aged 44 years, had had recurrent attacks of glaucomatocyclitic crisis affecting the left eye for 25 years. These were frequently associated with menstruation. The attacks lasted approximately 1 week, and had been treated with topical steroids and Diamox. Her father was thought to have phakogenic glaucoma.

External examination during a remission was normal.

On July 28, 1967, she attended with a mild recurrence, and slit-lamp examination revealed a clear anterior chamber in the right eye and pale keratic precipitates, a faint flare, and a few cells in the left.

Gonioscopy:

Right. Many fine pigmented iris processes in total circumference of angle. No other abnormality detected.

Left. The iris is inserted at the posterior border of corneo-scleral meshwork. Pigmentation moderate with fine iris processes.

Case 3, a locomotive fireman and fly fisherman aged 41, had had recurrent attacks of glaucomatocyclitic crisis affecting the right eye for 13 years. He considered exciting factors to be seasonal, particularly in the spring when unaccustomed close work is demanded for tying flies. The condition was rapidly relieved by topical steroids. There was no family history of glaucoma and no allergy.

External examination and slit-lamp examination gave normal results.

Gonioscopy:

Right. Narrow entrance to angle, recess not fully visible. A few clumps of mesodermal tissue lie in the recess; trabecula obscured by thin film of greyish white tissue; Schwalbe's line prominent; blood in Schlemm's canal; no iris processes.

Left. Narrow angle as on the right. A few mesodermal remnants in angle recess. Trabecula seen clearly; Schwalbe's line normal.

Case 4, an engineer aged 55, had had recurrent attacks of glaucomatocyclitic crisis affecting the right eye for 25 years. There was no family history of glaucoma, no precipitating factors, and no allergies.

November 9, 1942.—Crisis in the right eye which was white; pupil dilated; few fine keratic precipitates. The visual acuity was 6/9 in each eye and the ocular tension was 65 mm. Hg (Schjötz) in the right eye and 25 in the left.

December 16, 1942.—Bilateral trephine.

November 4, 1943.—Further crisis in the right eye. Bilateral iris inclusion. The patient subsequently had yearly attacks involving the right eye.

June 9, 1967.—The patient was asked to attend for review (see Table I). The right eye showed no bleb, no keratic precipitates, clear anterior chamber, blue iris, normal disc. The left eye was the same but with a brown iris.

Gonioscopy:

Right. Iris inserted by broad teeth to trabecular meshwork. Peripheral iris hypoplasia with many fine vessels; Schwalbe's line prominent. No blood in Schlemm's canal.

Left. Trabecula completely obscured by fine vascularized iris processes. No blood in canal of Schlemm. Schwalbe's line normal.

Case 5, a youth leader aged 38, had had recurrent attacks of glaucomatocyclitic crisis affecting the right eye for 17 years. He considered overwork and tension to be exciting factors. There were no allergies, and no migraine, but the family history was strongly positive. His brother appears in our series as Case 6, and there was glaucoma on the mother's side. Slit-lamp examination of

the right eye showed a few keratic precipitates and cells, and some iris atrophy around the sphincter on trans-illumination. The left eye was clear.

Gonioscopy:

Right. Hypoplastic peripheral iris with fine anomalous vessels. Atrophic iris periphery inserted or continuous with trabecular membrane, which is obscured in part by a sheet of yellowish tissue. Schwalbe's line very prominent. No blood in Schlemm's canal. Goniohaemorrhage induced on gonioscopy.

Left. Angle similar to right. No haemorrhage induced.

Case 6, a paint chemist aged 36, had had recurrent attacks of glaucomatocyclitic crisis involving the right eye for about 10 years. The family history was positive on the maternal side; his brother was Case 5. There were no apparent precipitating factors, and no allergies.

The visual acuity was 6/6 unaided in each eye and the ocular tension 53 mm. Hg (Schiotz) in the right eye and 18 in the left.

September 29, 1960.—Acute attack in the right eye. The pupil was normal.

June 5, 1961.—Right peripheral iridectomy with cyclodialysis in upper temporal quadrant. Post-operative course uneventful apart from slight hyphaema.

November 11, 1961.—Right cyclodialysis at 10.30 o'clock over region of previous iridectomy.

April 18, 1962.—Right Scheie's operation.

Gonioscopy:

Right. Angle filled with gelatinous-looking tissue which is probably continuous with hypoplastic peripheral iris tissue: anterior iris root insertion.

Left. Angle very narrow, open in places; last roll of iris resembles tissue seen in right angle.

Case 7, a police officer aged 21, had had two attacks of glaucomatocyclitic crisis at yearly intervals involving the left eye. The first lasting 3 weeks, and the second one week. There were no precipitating factors, no allergies, and no relevant family history.

External examination and slit-lamp examination gave normal results.

Gonioscopy:

Right and Left. Very narrow entrance to angle with absence of angle recess. Iris inserted at posterior border of trabecula. Schwalbe's line not prominent.

Discussion

The seven patients described in the present report have all shown bilateral angle anomalies similar to those described by Gorin (1964) as typical of the angle appearances in developmental glaucoma. The outstanding feature of all the angles described was the anterior insertion of the iris with consequent absence of a normal angle recess. In addition, the presence of fine iris processes, cellophane tissue obscuring the trabecular meshwork, and fine irregular vessels coursing across the angle have been noted. The angle anomalies are often more marked on the side prone to the crisis, but this was not so in Case 2.

In the presence of such a macroscopic angle anomaly it is not unreasonable to postulate a microscopic defect involving the trabecular meshwork, and this is to some extent supported by the low normal values for the facility of outflow found during remission in all patients except Case 2 (see Table I).

The mechanism of the rise in intra-ocular pressure during a crisis is disputed. Mills (1966) and Mansheim (1953) have recorded a reduction in the facility of aqueous outflow

during crisis consistent with the increase in intra-ocular pressure. Evidence to the contrary has been published by Sokolić (1966) and Spivey and Armaly (1963). Sokolić (1966) found the facility of outflow normal and unaltered during a crisis in one case. Spivey and Armaly (1963) presented three patients who had tonography performed during a crisis; from this they calculated the rate of aqueous flow— F —from the formula $F = (Po - 10)C$. Their results are summarized in Table II. These authors propose that their results support the hypothesis that the increased intra-ocular pressure is due, in part at least, to a hypersecretion of aqueous. Their data do not support this conclusion for three reasons:

TABLE II
RESULTS REPORTED BY SPIVEY AND ARMALY
(1963)

Case No.	In Crisis		
	Po	C	F
1	46.9	0.07	2.58
2	37.7	0.08	2.22
3	38.7	0.10	2.87

(1) The calculated F values are within the normal limits of 1.1 to 5.3 cu. mm./min. as defined by Grant (1951), who used the formula $F = C(Po - 4)$.

(2) The formula used by Spivey and Armaly (1963) to calculate the rate of aqueous flow gives disproportionately low F values at normal intra-ocular pressures and so inevitably an eye with a high intra-ocular pressure will appear to be hypersecreting in comparison with the normal eye.

(3) Tonography is notoriously unreliable at high intra-ocular pressures, and a small error in this measurement will give a large error in the calculated rate of aqueous flow. Table III shows the results of tonography during and between crises in our Cases 2, 5, and 6. There is no evidence of hypersecretion at any time. We consider that the marked reduction in the facility of outflow together with the angle anomaly is sufficient explanation for the marked elevation of intra-ocular pressure found during a crisis.

TABLE III

Case No.	Eye	Normal			Crisis		
		Po	C	F	Po	C	F
2	Left	22	0.43	5.2	32	0.15	3.3
5	Right	14	0.15	6.0	50	0.07	2.8
6	Right	14	0.12	4.8	53	flat trace	

$$F = (Po - 10) C$$

In all our patients the severity of the cyclitis has been found to bear no relationship to the rise in intra-ocular pressure, a finding noted by previous authors (Posner and Schlossman, 1948; Theodore, 1952). Posner and Schlossman emphasized the mildness of the inflammatory signs and in their original paper presented one case in which none was seen. It is the opinion of the present authors that the inflammatory signs are due to the acute rise in intra-ocular pressure, but one cannot exclude the possibility that the primary cause of the syndrome is an oedema or low grade inflammation of the ciliary body.

The precipitating factors of an attack remain unknown; all the patients in the series were closely questioned regarding a history of allergy or migraine—a relationship noted by Posner and Schlossman and by Theodore. Case 2 has noted that the attacks occur at the onset of menstruation; Case 4's attacks have a seasonal incidence.

Two of our patients, Cases 5 and 6, are brothers with a family history of glaucoma on the maternal side. In such a rare disease this is strong evidence for recessive inheritance such as one finds in developmental glaucoma.

Two patients in the series of twelve described by Posner and Schlossman were noted to have a positive family history of glaucoma. In a pair of identical twins, one of whom was described by Levatin (1956) as having glaucomatocyclitic involvement, the fellow twin failed to show the condition.

The mechanisms of the acute rise in intra-ocular pressure remain unknown, although in view of the gonioscopic and tonographic results obtained in the present series there would appear to be a sufficient embarrassment of outflow to account for the glaucoma.

Summary

Seven typical cases of recurrent glaucomatocyclitic crises are presented. All patients showed abnormalities of the drainage angles of the type seen in developmental glaucoma. Measurements of the facility of outflow in crisis and in remission suggest that the glaucoma is due to obstruction of the drainage channels. It is the authors' opinion that glaucomatocyclitic crises should be considered as one of the developmental glaucomas, although retaining its peculiar individuality in view of its clinical manifestations and benign prognosis.

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