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PRIMARY ANGLE-CLOSURE GLAUCOMA*

FAMILY HISTORIES AND ANTERIOR CHAMBER DEPTHS

BY

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THAT acute congestive glaucoma occurs in eyes with a recognizable anatomical predisposition has been known for many years (von Graefe, 1857; Priestley Smith, 1887). Shallowness of the anterior chamber appeared to be an important factor, and Rosengren (1930, 1931, 1950) presented convincing statistical evidence that this was so. The development of gonioscopy showed that a narrow angle of the anterior chamber accompanies a shallow anterior chamber (Barkan, 1938; Sugar, 1941). The members of the C.I.O.M.S. Symposium on Glaucoma (Duke-Elder, 1955) adopted the term "Closed-Angle Glaucoma" (frequently changed to "Angle-Closure Glaucoma") indicating the necessity for this form of glaucoma to be caused by apposition of the iris to the angle wall irrespective of the depth of the anterior chamber and the width of the angle. Nevertheless, in the overwhelming majority of cases of primary angle-closure glaucoma, the anterior chamber is shallow and the angle is narrow—so much so that the diagnosis should be very carefully reviewed if examination shows an anterior chamber of normal depth (and an angle of normal width in the other eye).

The importance of these anatomical factors has naturally evoked considerable curiosity as to whether they are genetically determined. Törnquist (1953) considered that one specific dominant gene was responsible for the shallow anterior chamber, although the normal variation of chamber depth from person to person could be due to polygenic factors. Kellerman and Posner (1955) examined 48 eyes of relatives of patients with acute congestive glaucoma and found that twelve (25 per cent.) had narrow angles—but no glaucoma. Paterson (1961) investigated fifty siblings of patients with definite angle-closure glaucoma and found three (6 per cent.) with definite glaucoma, 21 (42 per cent.) with narrow angles (suspect glaucoma), and 26 (52 per cent.) with no glaucoma, in the age range of 40 to 60 years. An unusually high incidence of glaucoma among siblings of patients with angle-closure glaucoma was recorded by Weekers, Gougnard-Rion, and Gougnard (1955); in eight families of subjects with closed-angle glaucoma there were 42 kinsfolk over 30 years of age and at least thirteen of these had glaucoma.

Present Investigations

During the clinical research examinations of over 300 patients who had suffered one or more forms of primary angle-closure glaucoma as described elsewhere (Lowe, 1961), 200 were questioned carefully concerning eye disease in their families.

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The angle-closure glaucoma patients were, themselves, usually elderly and information about their parents' eye diseases tended to be vague or of insufficient accuracy to be useful. However, one man aged 86 years, and his daughter aged 56 years, were examined and both were proved to have suffered attacks of acute angle-closure glaucoma.

Siblings were thought likely to provide useful information on the following grounds: acute angle-closure glaucoma is a dramatic disease that causes much distress and loss of vision from acute attacks that necessitate urgent admission to hospital (here), so that knowledge of its occurrence would be probable where family members maintained contact. Because of the possibly familial anatomical predisposition, a large number of positive family histories could be expected.

From 200 propositi, 778 siblings were listed (395 brothers and 383 sisters), but family histories of angle-closure glaucoma were found to be distinctly uncommon, only ten with glaucoma of any kind being recorded (Table I). Although many of the patients had lost touch with their brothers and sisters, the majority maintained some communication.

TABLE I

TYPES OF GLAUCOMA REVEALED IN FAMILY HISTORIES OF 200 PROPOSITI WITH PRIMARY ANGLE-CLOSURE GLAUCOMA

Type of Glaucoma	Sisters	Brothers	Total
Acute angle-closure glaucoma	3	—	3
Sub-acute angle-closure glaucoma	—	1	1
Chronic simple glaucoma with narrow angles	2	—	2
"Chronic glaucoma" no acute attack	1	—	1
"Glaucoma"—type unknown	—	3	3
Total	6	4	10

Fifty propositi were asked about consanguinity of parents, and one was positive.

Case 1, a married woman aged 52 years, had a blind left eye from neglected acute angle-closure glaucoma. Both eyes had shallow anterior chambers and typically ballooned irides that caused very narrow angles. She also suffered from advanced retinitis pigmentosa. Her parents were first cousins. She had three sisters and one brother—her brother had retinitis pigmentosa but none of her siblings was known to have glaucoma.

Discussion

The patients investigated were of European birth or descent domiciled in the State of Victoria, Australia. They were mostly public patients of the Royal Victorian Eye and Ear Hospital but some were private patients. The majority were from the lower income groups and included many "old-age" pensioners. They thus represent a selected section of the general population but the presentation of their angle-closure glaucoma and its features resembled closely cases seen elsewhere or described in the medical literature from other countries.

Törnquist (1953) examined 59 siblings of 49 patients who had developed acute glaucoma (and who had shallow anterior chambers). Of the 59 siblings, three had also suffered from acute glaucoma, but only two of the 56 unaffected siblings had

anterior chambers more shallow than their corresponding propositi. Only six of the 49 propositi compared with 52 of the siblings had anterior chambers deeper than 2 mm.

Rosengren (1953), using the figures given by Nelander (1933), and Törnquist (1956), from his own examinations, showed that the risk of acute glaucoma increased greatly as the anterior chamber depth diminished (Table II).

TABLE II

CALCULATIONS OF RISK OF DEVELOPING ACUTE GLAUCOMA, BY ANTERIOR CHAMBER DEPTH

Anterior Chamber Depth (mm.)	Risk	Author	Date
>2·53	1:32,573	Rosengren	1953
<2·53	1:152		
2·5–2·0	1:180	Törnquist	1956
2·0–1·5	1:10		
1·5–1·0	52:1		

Although the precipitation of an attack of angle-closure glaucoma depends upon various circumstances that appear to be almost fortuitous in their occurrence (Lowe, 1961), the most important predisposing factors are undoubtedly a shallow anterior chamber and a narrow angle.

The depth of the anterior chamber of normal eyes follows a normal frequency distribution curve (Stenström, 1946: Rosengren, 1953; Törnquist, 1953). Likewise, persons who have developed primary angle-closure glaucoma and their siblings have a range of anterior chamber depths, but their whole range is approximately two-thirds that of the normal depths, and it is usually only those with the most shallow anterior chambers who actually develop angle-closure glaucoma.

Törnquist (1956) considered there was "no crucial threshold value below which the risk of acute glaucoma appears". Nevertheless, an anterior chamber less than 2 mm. deep appears to carry such a high risk of angle-closure that prophylactic surgery becomes a reasonable proposition. Such cases for potential surgery are rare, however, since most of the siblings of patients with angle-closure glaucoma have anterior chambers deeper than 2 mm., so that they would escape the prophylactic surgery (and the disease).

In this respect the eyes of siblings differ greatly from the fellow (uninvolved) eyes of patients who have suffered an attack of primary acute angle-closure glaucoma. Here, the risk of acute angle-closure glaucoma in the second eye (with no treatment or with miotics) approaches 3:1; this not only justifies prophylactic surgery but makes it desirable to permit the patient to lead a relatively normal life (Lowe, 1962). For these patients the usual provocative tests, such as the dark-room test or the mydriatic test (with or without tonography), are not necessary because they bear little relationship to the provocations that induce "natural" angle-closure glaucoma, and if negative are liable to cause procrastination of the valuable prophylactic surgery. Such tests are more useful indications of ocular instability in persons who have not developed any form of angle-closure glaucoma but have shallow anterior chambers.

The provocative tests can be readily performed and their results have been adequately evaluated, whereas the accurate measurement of anterior chamber depths has not yet been done often enough for the results to be adequately assessed.

The following case illustrates the need for careful examination:

Case 2, a married woman born in 1890, attended in 1952 when aged 62 years, with bilateral acute angle-closure glaucoma, and bilateral iridencleises were done. The right eye fared badly but she recovered 6/6 vision in the left eye although with the 5/2000 target the left visual field was greatly reduced and showed a lower arcuate scotoma breaking through to the periphery. Her visual acuity slowly deteriorated but she retained useful vision until she died in 1961.

Case 3, her sister, a married woman aged 59 years, attended for new glasses in 1963. She had no other symptoms but reported that her sister had required operations for acute glaucoma. Refraction was performed under homatropine and cocaine drops and while the pupils were dilated the ocular tension in each eye was found to be 28 mm. Hg (1955 scale). She had shallow anterior chambers, normal optic discs, and normal visual fields to 2/2000 white targets. She was considered to have angle-closure glaucoma. She was treated with eserine and pilocarpine drops and advised that surgery (peripheral iridectomies) would probably be necessary. At the glaucoma unit and after discontinuing miotics, the tests showed:

Applanation	C	Po/C
27|25	·18|·17	150|137

The ocular tensions and outflow coefficients were not affected by the darkroom-tonography test or the mydriatic test. When the pupils were dilated with homatropine and cocaine the angles were noted to be open as they were before this test. The raised ocular tensions and reduced outflow coefficients were therefore not due to angle-closure but to early chronic simple glaucoma. The angles were narrow but not unduly so (Shaffer Grade II) and the irides were very little ballooned, so that for the time being, treatment with miotics was recommended.

Comment

These two sisters had shallow anterior chambers and narrow angles, only one (so far) developed angle-closure glaucoma, but both had chronic simple glaucoma. In the writer's experience, family histories of chronic simple glaucoma are much more frequent than those of angle-closure glaucoma and even though a person has suffered from angle-closure glaucoma, if close relatives have glaucoma it is more likely to be the chronic simple type than the angle-closure form.

Conclusions

Now that reasonably accurate devices for measuring the anterior chamber depth are available for attachment to some slit lamps, glaucoma units should make this examination part of their routine procedure when there is any suggestion of shallowness of an anterior chamber. As experience with these measurements is developed their critical assessment may become as important as any other method of examination or provocative test.

The examination of the siblings of patients presenting with angle-closure glaucoma will reveal only a small percentage likely to benefit from treatment or observation.

Summary

Positive family histories of angle-closure glaucoma are very uncommon. Affected patients and their siblings have a range of anterior chamber depths approximately

only two-thirds that of the normal population. The risk of angle-closure glaucoma increases greatly in the presence of anterior chambers less than 2 mm. deep, but most siblings of patients with angle-closure glaucoma have anterior chambers deeper than this. When a person develops acute angle-closure glaucoma in one eye, the uninvolved eye should be submitted to prophylactic surgery, but for the unaffected siblings of such patients prophylactic surgery is not justified unless the anterior chambers are less than 2 mm. deep or provocative tests indicate sufficient ocular instability.

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