Alterations of the anterior lens capsule associated with climatic keratopathy

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SUMMARY We describe changes in the anterior lens capsules of older people in Somalia, a country which is close to the equator and with large areas of sand, often highly reflective of sunlight. The capsule changes are confined to the central pupillary area. In order of apparently increasing severity they consist of a white opalescence ('frosting'), an elevation in front of the contour of the rest of the lens to form a plateau, and a 'bag' or herniation of the lens capsule through the pupil. Plateau and bagging taken together are strongly associated with climatic keratopathy (and by inference with reflected ultraviolet exposure), weakly associated with exfoliation syndrome, inversely related to the degree of cataract, and interfere severely with vision.

Deformations of the anterior lens capsule have been noted among approximately 10% of patients aged over 50 in a new eye unit in Afgooye, Somalia, and less frequently among females. These range from a plateau shaped elevation in the pupillary area through an anterior lenticonus to a frank herniation or 'bagging' of the lens capsule through the pupil. At the same time it has been frequently observed that the anterior lens capsule is thinner than normal and much more friable at lens extraction than would be considered normal in other parts of Africa or in Europe.

Severe climatic droplet keratopathy (synonyms: spheroidal degeneration of the cornea, actinic keratopathy, solar keratopathy, Labrador keratopathy) and exfoliation syndrome (pseudoexfoliation of the lens) were both frequent in this population. The questions we asked were whether the anterior lens capsule deformation could be firmly related to climatic keratopathy and by inference to ultraviolet light (UV) exposure, to exfoliation, or to the presence or absence of cataract.

Subjects and methods

The study was conducted at Centre Afgooye, a small eye unit 30 km from the capital city, Mogadishu. This is a joint project of the Government of Somalia and

the Christoffel Blindenmission, in partnership with the International Centre for Eye Health. The patients came not only from the surrounding area but from all over the country and occasionally from contiguous countries. The investigation was planned as a case control study. Possible cases, suspected of showing the anterior lens capsule distortion, were selected by torch and loupe examination by the screening doctors and nurses for more detailed study. Controls were obtained by examining in detail every second patient aged over 50 until sufficient numbers were obtained without lens capsule changes. The patients admitted for surgery on three successive weeks were also examined and classified as cases or controls

For each case and control the hospital number, age, sex, and, when possible through interpreters, the area of usual residence and occupation were recorded on a separate form. Visual acuity was recorded by medical assistants using an E chart. The anterior segment was examined with a Takagi slitlamp (similar to the Haag-Streit 900). A drawing of the cornea was made. Climatic keratopathy was graded according to previous definitions' (summarised in Table 1). The intraocular tensions were measured either by Schiøtz tonometry or the Mark II Perkins hand-held applanation tonometer (Clement Clarke Ltd), or occasionally confirmed as normal by palpation if there was no time for formal pressure measurement.

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Table 1 Definitions of categories of climatic keratopathy (spheroidal degeneration of the cornea)

- Grade 0: No keratopathy, or eyes with traces of keratopathy—that is, few 'droplets' in one or other margin but not symmetrically on both sides of the cornea
- Grade 1: The fine, translucent grey colour or colourless deposits are confined to both nasal and temporal marginal strips of the cornea
- Grade 2: The deposits extend into the optic axis but are not sufficiently dense to measurably reduce vision
- Grade 3: The grey deposits extend across the pupillary area in a band which reduces vision
- Grade 4: In addition there are raised yellow or brown nodules, elevating the epithelium, which, however, is intact unless the excrescences are in the process of flaking off

The pupils of each person included in the study were dilated with one drop of phenylephrine hydrochloride 5%. If dilatation was inadequate this was repeated, and occasionally cyclopentolate hydrochloride 1% added. The lens of each eye was then re-examined with the slit-lamp and direct ophthalmoscope with particular attention to three features: contour of anterior lens capsule, presence of exfoliation syndrome, and presence and stage of cataract.

The configuration of the centre of the anterior lens capsule was classified into one of four categories, as outlined in Table 2. The eye was disqualified if there was a mature cataract so that the anterior lens capsule was wrinkled or stretched without unequivocal forward protrusion in the pupillary area.

Careful inspection of the capsule of controls often revealed a slightly whiter area of the grey capsule in the area exposed by the undilated pupil, which we called 'frosting' (Fig. 1). This was remarkably frequent in eyes without any other apparent intraocular

Table 2 Definitions of categories of lens capsule change

The configuration of the anterior lens capsule in the pupillary area was allocated to one of four categories

- 1 Normal: Normal curvature, normal shagreen and no 'frosting' of surface
- 2 Frosting: This was a central zone of capsule representing the area defined by the constricted pupil, which had a whitish opalescence (Fig. 1). It resembled the appearance of the same area in the exfoliation syndrome, but without the associated features of exfoliation. The curvature of the anterior lens capsule was smooth and continuous
- 3 Plateau: In addition to 'frosting' the pupillary area of capsule was elevated above the surrounding lens when viewed in the slit beam and protruded forward, but the central area remained flat (Fig. 2)
- 4 Bagging: The capsule and lens contour protruded forwards within the confines of the undilated pupil. The contour was often conical, and could be described as an anterior lenticonus. In other cases the 'bag' was irregular in shape and cross section and was sometimes a frank 'herniation' of lens capsule and contents through the pupil (Figs. 3-6).

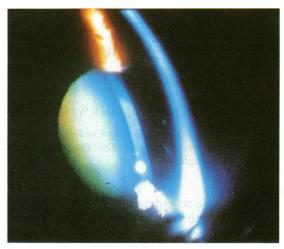


Fig. 1 The whitish appearance of the central anterior lens capsule described as 'frosting', in the presence of early grade 3 climatic keratopathy. The curve of the anterior lens capsule remains smooth and continuous.

pathology. Therefore, for a diagnosis of exfoliation to be made, it was necessary to see in addition either white/grey flakes resembling dandruff on the pupil border or the characteristic light grey membrane of exfoliation material on the periphery of the lens capsule, often with curled anterior edges.

When severe grade 3 or grade 4 climatic keratopathy was present and it was not possible to see clearly the centre of the lens capsule with the eye looking straight ahead, the patient's gaze was directed downwards. It was then possible to examine the upper half of the pupillary area of capsule with the slit-lamp, looking through clear cornea over the band of keratopathy; similarly the lower part of the pupillary area was seen with the eyes turned upwards. With the mirror of a gonioscopic lens in the 12 or 6 o'clock positions additional detail of the contour of the central capsule could be studied and signs of exfoliation confirmed. We are thus confident that the observed capsule changes are not an optical illusion caused by a distorted view through a band of keratopathy.

Cataract, if present, was classified into one of three stages, summarised in Table 3, against the red glow of the fundus. The appearance of the fundus was also recorded if it could be seen with the direct ophthalmoscope.

The diagnoses and visual acuities of all the new patients seen in this clinic are being monitored. In the first three weeks of this study 25% of all new patients were bilaterally blind (corrected acuity less than 3/60). Similarly, in the first four months of the clinic's operation 33% of all eyes examined had been blind. With this proportion of severe pathology it can be

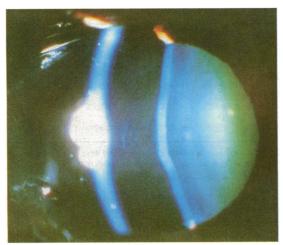


Fig. 2 Plateau formation in left eye of 79-year-old male, with the pupillary area of anterior lens capsule elevated and protruded forwards, while the centre remains flat. The pupillary zone of capsule has the whitish opalescence of frosting.

understood that many potential case and control eyes had to be disqualified from further analysis because of additional pathology, such as uveitis, severe glaucoma, or a mature cataract where the details of the lens capsule could not be clearly seen or where it was already wrinkled.

DATA ANALYSIS

Patients with plateau or bag herniation of the anterior lens capsule in either eye were regarded as 'cases' and were compared with controls who did not have such capsule changes. Similarly, eyes with plateau or bagging of the lens capsule were regarded



Fig. 3 Transition between plateau and 'bagging' of the anterior lens capsule. The contour is tending towards anterior lenticonus. Right eye of same patient as Fig. 2.



Fig. 4 'Bagging' of the unterior lens capsule. The limits of the protruding lens are defined by the margin of the undilated pupil. The underlying lens centre is clear.

as 'case eyes' and were compared with 'control eyes'. The latter analyses are reported in this paper. Thus the eye is taken as the experimental unit, since each eye is independently classified according to characteristic of primary interest that are local to it, such as climatic keratopathy and anterior lens capsule change. The comparisons were made within strata in which cases and controls were similar in respect of age, sex, and presence of exfoliation. Multivariate log-linear modelling was used to evaluate the inter-



Fig. 5 Frank herniation of central anterior lens capsule and contents through the pupil in 80-year-old male. The details of iris margin and lens are partially obscured by a fine granular band of climatic keratopathy.

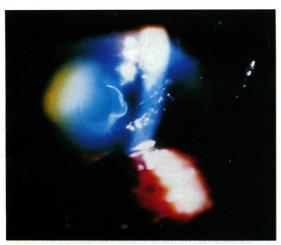


Fig. 6 Herniation or 'bag' of lens capsule protrusion through pupil, in 55-year-old male nomad. Some of the nodular changes of grade 4 climatic keratopathy are visible.

relationships between capsule changes, keratopathy, exfoliation, cataract, age, and sex.

The analyses of the association between lens capsule change and keratopathy were performed with both exclusion and inclusion of eyes with intraocular pressure over 25 mmHg, because some authors have considered that glaucoma may itself cause deposition of spheroidal bodies in the cornea or exacerbate existing actinic keratopathy.

Results

The sample comprised 52 patients with plateau or bag herniation of the anterior lens capsule in either eye (cases) and 94 controls who did not have such capsule changes, making a total of 146 patients. The age and sex distribution of the study patients are shown in Table 4.

Table 3 Definitions of categories of cataract

The presence and degree of lens opacities were judged by viewing through a direct ophthalmoscope held close to the patient's eye and against the red glow of the fundus*

- 1 Normal: No opacities detected
- 2 Slight: Very early lens opacities such as a solitary radial spoke noted. In other words, it could not be passed as completely normal
- 3 Partial cataract: An opacity was clearly present but did not obscure all the red glow
- 4 Complete cataract: No red glow visible. This did not necessarily mean that the cataract was 'mature' in the sense that the white opacities extended up to the anterior capsule when studied with the slit-lamp

Table 4 Age and sex distribution of the study patients

	Age groups					
	40-49	50-59	60-69	70–79	80+	Totals
Males % of total Females % of total Totals % of total	1 0.68% 1 0.68% 2* 1.37%	17 11·64% 12 8·22% 29 19·86%	34 23·29% 18 12·33% 52 35·62%	10 6·85% 45	15 10·27% 3 2·05% 18 12·33%	103 69·86% 44 30·14% 146 100·00%

^{*}These two patients were both controls and were excluded from analysis as there were no corresponding cases in that age group.

There were 228 eyes in which it was possible to quantify or grade keratopathy, exfoliation, and changes in the anterior lens capsule in each eye. Of these, 68 had plateau or bagging of the lens capsule. The remaining 160 were regarded as control eyes. There were two additional case eyes in which it was not possible to confirm the presence or absence of exfoliation.

Analysis revealed a strong association between capsule change (plateau or bagging) and climatic keratopathy (Table 5).

There was a marked 'dose-dependent' relationship between various grades of climatic keratopathy and

Table 5 Association of the anterior lens capsule changes with keratopathy. The data are stratified by age and sex. Case eyes are those with plateau formation or bagging of the anterior lens capsule. In this analysis eyes with frosting of the lens capsule are included among the control eyes

Age		Males		Females		Males+ - females	
		No. eyes	% with kerato- pathy	No. eyes	% with kerato- pathy	No. eyes	With kerato- pathy
50-59	Case eyes	3	100%	1	100%	4	100%
	Control eyes	25	52%	18	33%	43	42%
60-69	Case eyes	20	100%	2	100%	22	100%
	Control eyes	38	61%	32	44%	70	50%
70+	Case eyes	42	100%	2	100%	44	100%
	Control eyes	31	77%	16	63%	47	62%
All	Case eyes	65	100%	5	100%	70	100%
ages	Control eyes	94	64%	66	45%	160	51%

Results of multivariate log-linear modelling:

Significant association between: (a) Capsule change (plateau/bag) and keratopathy; (b) Capsule change and sex; (c) Capsule change and age; (d) Keratopathy and sex; (e) Keratopathy and age.

Age-adjusted odds ratio for sex=7.6 (95% confidence limits 3.2 and 8), that is, plateau or bagging is about 7 times more common in males than in females of similar age.

The odds ratio for keratopathy in the sample is infinitely high, since every case eye had keratopathy.

NB: Two case eyes in this table are not included in Tables 6 and 7, as it was not possible to confirm presence or absence of exfoliation.

^{*}An eye was not included in the analysis if the capsule was wrinkled over a mature cataract.

Table 6 'Dose-dependent' association between anterior lens capsule change and keratopathy. Case eyes are those with plateau or bag herniation of the anterior lens capsule. Keratopathy is taken as an index of exposure to ultraviolet sunlight (UV), so that more severe grades of keratopathy indicate correspondingly higher levels of exposure to UV

£	Keratopa (index of	Totals			
	0-1*	2	3	4	_
Case eyes	22	7	27	12	68
Control eyes	132	13	13	2	160
Totals	154	20	40	14	228
Odds ratios	1.0	3.2	12.5	36.0	
Test for trend	$\chi^2 = 64 \cdot 1$	p<0.001			
Findings indica capsule change					isk of
The trend persi Summary	ists when th	ne data are	stratified	by exfoliati	on:
Odds ratios	1.0	3.8	10.3	45.6	

^{*}Grades 0 and 1 keratopathy are combined in order to avoid infinitely high odds ratios due to zero cell value.

changes in the anterior lens capsule. On the assumption that more severe grades of keratopathy indicate correspondingly higher cumulative exposure to UV, there was a strong trend of progressive increase in risk of lens capsule change with rising levels of exposure to UV light. These data are shown in Table 6. Fourteen eyes had intraocular pressure greater than 25 mmHg. When these were excluded from the analyses set out in Table 6, there was very little change in the odds ratios (1·0, 2·9, 16·0, and 40·1) for the different keratopathy grades: the 'dose-dependent' association between plateau and bag formation and keratopathy remained, and the trend persisted when the data were stratified by exfoliation.

A weaker association was found between lens

Table 7 Frosting of the anterior lens capsule in relation to exfoliation and keratopathy. Eyes with plateau or bagging are excluded from this analysis

		Keratopathy			
		0–1	2–4	Totals	
Exfoliation present	Frosting	17	5	22	
•	Capsule normal	10	0	10	
Exfoliation absent	Frosting	34	19	53	
	Capsule normal	71	4	75	
	Totals	132	28	160	
Results of log-linea	r modelling:				
(a) Strong association between		Frosting and keratopathy (odds ratio=9.5)			
(b) Weaker association between		Frosting and exfoliation (odds ratio=3·1)			
(c) No significant association between		Keratopathy and exfoliation			

Table 8 Inverse association of lens capsule plateau or bagging with cataract. Case eyes are those with plateau or bagging of the anterior lens capsule

Age		Grade 0–1	Grade 2	Grade 3	Grade 4	Total	% Eyes with grade 3-4 cataract
50-59	Case eyes	3	0	0	1	4	25%
	Control eyes	22	9	3	8	42	26%
60-69	Case eyes	8	11	1	2	22	14%
	Control eyes	14	22	4	28	68	47%
70-79	Case eyes	5	12	2	6	25	32%
	Control eyes	7	7	8	17	39	64%
80 +	Case eyes	2	8	1	8	19	47%
	Control eyes	1	1	2	4	8	75%
Summ	ary odds ratio):				Test o	f trend p Value
Age a	djusted	1	1-168	0.331	0.366		<0.01
adjı	ısted	1	1.218	0.432	0.496	5.4	<0.05
	only age isted	1	1.103	0.246	0.307	7.5	<0.01
Females only		Inadeq	uate sa	mple si	ze		

capsule change and presence of exfoliation syndrome (odds ratio 3.8). No significant association was found between exfoliation and climatic keratopathy.

Frosting of the anterior lens capsule in the absence of other capsule changes was positively associated with keratopathy and with exfoliation. However, the association with keratopathy was much stronger than that with exfoliation (Table 7).

A negative relationship was found between cataract and lens capsule change. Eyes with grade 3 or 4 cataract were about three times less likely to have plateau or bagging compared with eyes with grade 0-1 cataract (Table 8).

Discussion

From Somalia we have described a condition of the anterior lens capsule in which it either herniates through the pupil as a 'bag', or is elevated in front of the surrounding lens to form a plateau, and which is associated with severe climatic keratopathy. These two conditions frequently occur together without any sign of cataract or other pathology, and yet together may reduce vision to the level of hand movements. If the fundus is examined over the band of keratopathy with a direct ophthalmoscope with the pupil dilated, a perfectly clear view of disc and macula can be obtained. The area of protruding capsule appears as a dark ring or round spot against the red glow of the fundus.

This lens distortion is of importance for two reasons: first, as a cause of severe visual reduction due to its optical interference with image formation, and because of the difficulty of knowing the best surgical procedure to adopt; second, because of its theoretical significance.

Similar changes of the anterior lens capsule have been observed by one of the authors among the people with spheroidal degeneration in Newfoundland and Labrador,² though only three examples were noted where the deformity was described as a lenticonus or frank herniation in the pupillary area; usually it took the form of a slightly elevated plateau with a frosted surface. Somalia is located just north of the equator, and large areas are covered by sand which in places is highly reflective of sunlight. The traditional way of life is nomadic, and most of the population, even in towns, have spent a large part of their lives as nomads, herding sheep and camels.

Climatic keratopathy is generally more severe in Somalia than in Labrador. It was recorded as the third most frequent cause of blindness (less than 3/60) after cataract and glaucoma among the patients who came in the first four months of the clinic's operation. When the severe grades of keratopathy are associated, as they usually are, with the anterior lens abnormality, it is sometimes difficult to judge by observation whether the cornea or lens is making the greater contribution to visual loss. The corneal deposits may be scraped off or a lamellar keratectomy performed, yet the vision be very little improved because of the underlying capsule deformity. The experience in Labrador was that a perfectly clear corneal graft or a satisfactory lamellar keratectomy sometimes gave a disappointing visual result, interpreted as due to the abnormal shape and surface of the lens capsule.

In Somalia, in cases where the central part of the band of climatic keratopathy is not very dense, it may be that lens extraction is indicated even in the absence of significant cataract. It seems reasonable, however, first to undertake a trial of the visual results of superior optical iridectomy to allow light to bypass the abnormal corneal and lens surfaces. The lens capsules of these Somali patients are abnormally friable compared with other parts of Africa, Europe, or India. The surgeons at the Centre have learnt never to use capsule forceps, only cryoextraction or the tumbling method.

There is considerable circumstantial evidence that climatic keratopathy (primary spheroidal degeneration) is caused by exposure to excessive doses of reflected ultraviolet light. The strong association of the anterior lens capsule change with keratopathy suggests that it also could be caused by excessive light exposure. The cornea is known to filter virtually all light of wavelengths under 300 nm. It seems reasonable to think that the shortest of the wavelengths immediately above this, say 300–340 nm, which would be the most biologically active of the remain-

ing wavelengths, might be absorbed in the superficial layers of the lens, in the pupillary area, possibly by the cuboidal epithelium. This would not be surprising in view of the vulnerability of the corneal epithelium to UV light. The high ambient temperatures in the desert regions of Somalia may accelerate any resulting photochemical reaction.

The lens capsule changes are of theoretical importance in view of the extensive research being undertaken into the possible role of ultraviolet wavelengths in the aetiology of cataract. Here we have a condition confined to the anterior surface of the lens, not necessarily associated with cataract, and yet quite probably due to exposure to some of the short wavelengths of light.

Exfoliation syndrome has also been attributed to light exposure. It is extremely common in the Somali population. Although there was some association of exfoliation with lens capsule plateau formation or bagging, there was no significant association of exfoliation with climatic keratopathy.

For the analyses eyes with frosting were included among the controls rather than the cases. At the outset of the investigation it was not clear whether the frosting represented an early sign of exfoliation or was the first step in the process leading to pleateau and bag formation. Since on clinical inspection all the capsules with plateau formation had central frosting, while only a proportion of the exfoliation had frosting, it seems likely that frosting may be a stage in the development of gross anterior lens capsule deformation.

If, as suspected, the lens capsule alteration is caused primarily by ultraviolet wavelengths, both it and the keratopathy would be susceptible to prevention by the wearing of appropriate sunglasses. This has not yet become a practical possibility in the present stage of development of the nomadic population in Somalia.

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