

# Capsular exfoliation syndrome

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The nature of senile capsular exfoliation is not clear, although at present it is often regarded, on insufficient evidence, as pseudoxfoliation, exudative in character, and possibly derived from the uvea as a result of low-grade inflammation. We have recently seen nine cases which throw light on the pathogenesis of capsular exfoliation.

## Observations

In the Eye Outpatient Department of H.P. Medical College Hospital, Simla, nine cases of capsular exfoliation affecting seventeen eyes were seen in the course of 6 months. Their main features are given in Table I (overleaf, pp. 122 and 123). All the patients were males between the ages of 60 and 76 years. The capsular exfoliation was associated with phakodonesis, iridodonesis, glaucoma, primary optic atrophy, rubeosis iridis, and choroidal sclerosis (Table II).

**Table II** *Associated findings in 17 eyes in 9 cases of exfoliation of the lens capsule*

<i>Findings</i>	<i>No. of eyes</i>
Phakodonesis	11
Mature cataract	7
Immature cataract (Nuclear sclerosis)	10
Dislocated lens	2
Glaucoma	4
Iridodonesis	5
Corneal dystrophy	1
Rubeosis iridis	1
Primary optic atrophy	1
Choroidal sclerosis	1

Seven eyes were treated surgically: five by combined lens extraction with sector iridectomy, one by lens extraction with anterior punch sclerectomy, and one by punch sclerectomy only (Table III, opposite). There were no complications; non-recurrent hyphaema appeared in three eyes soon after the operation but responded to conventional treatment.

## Discussion

Exfoliation of the lens capsule is a disease of unknown aetiology characterized by a whitish flaky or powdery substance on the anterior surface of the lens. Flakes may also be

**Table III** Operative and post-operative particulars of 7 eyes in 6 cases of exfoliation treated surgically

S. no.	Case no.	Age (yrs)	Sex	Eye	Operation	Post-operative complications	Fundus	Final visual acuity	Postoperative intraocular pressure (mm. Hg Schiödtz)
1	1	60	M	L	Punch sclerectomy Sector iridectomy Intra capsular cataract extraction	Hyphaema 4th day	Normal	6/12 with + 12 D sph.	10
2	2	60	M	R	Sector iridectomy Intracapsular cataract extraction	Nil	Normal	6/12 with + 10 D sph.	14.5
3	3	65	M	R	Sector iridectomy Intracapsular cataract extraction	Hyphaema 4th day	Normal	6/12 with + 9 D sph., 2.5 D cyl., axis 180°	17
4	5	65	M	R	Sector iridectomy Intracapsular cataract extraction	Nil	Marked choroidal sclerosis	6/24 with + 9 D sph., + 2 D cyl., axis 180°	17
5	5	65	M	L	Punch sclerectomy with sector iridectomy	Nil	Not visible	as before	20.5
6	6	60	M	L	Sector iridectomy with intracapsular cataract extraction	Nil	Normal	6/36 with + 11 D sph.	12
7	8	68	M	R	Intracapsular cataract extraction with peripheral iridectomy	Hyphaema 5th day Iridocyclitis ++	Normal	6/60 with + 9 D sph., + 1 D cyl., axis 180°	14.5

seen in the angle of the anterior chamber, pupillary margin, and iris crypts, and in association with the zonular fibres. The condition is frequently associated with glaucoma (Cambiaggi, 1959; Lowe, 1964; Sood and Ratnaraj, 1968).

Lindberg (1917) and Malling (1923) observed this condition, and Vogt (1925) made a detailed study and described it as an exfoliation of the superficial layers of the anterior lens capsule. Busacca (1927) postulated that the flakes were derived from aqueous humour and not from the lens capsule.

Dvorak-Theobald (1954) thought that the material was of independent origin and had no connection with capsular exfoliation, and he therefore termed the condition 'pseudo-exfoliation'. Weekers, Prijot, Delmarcelle, Lavergne, Watillon, Gougard, Gougard-Rion, and Gustin (1959) suggested the term 'senile uveal exudation', and the condition has since been regarded as a mild exudative process of uveal origin.

In our cases the high incidence of phakodonesis, iridodonesis, and posterior dislocation of the lens was suggestive of as a primary weakness of the zonule. Bartholomew (1970) regarded phakodonesis as a sign of incipient lens displacement. In six of his seven cases capsular exfoliation and phakodonesis co-existed. The granulo-fibrillary nature of the flaky material suggests that it is derived from the lens capsule and zonule. The presence of similar material on the deeper aspect of the lens capsule and around the iris blood vessels which was revealed by electron microscopy (Ashton, Shakib, Collyer, and Blach, 1965) favours a local origin. The presence of similar material within the internal limiting membrane, stroma, and basement membrane of the iris and trabeculae at the angle of the anterior chamber may be due to a widespread degenerative process occurring locally at all these sites. We believe that this material arises from the degradation of a substance present at these sites. This substance has not been identified, but is likely to be a specific type of mucopolysaccharide undergoing denaturation. The associated lesions (corneal dystrophy, rubeosis iridis, choroidal sclerosis, and optic atrophy) may result from a similar degenerative process. The high incidence of hyphaema after surgery may be caused by

**Table I** *Clinical features of 9 cases of exfoliation*

Case no.	History			Examination			
	Age (yrs)	Sex	Loss of vision Other complaints	Eye	Corrected vision	Anterior chamber	Iris
1	60	M	Progressive both eyes—6 yrs Pain right 2 yrs	R	No PL	Deep	Flakes at PM ++ Rubeosis iridis ++ Iridodonesis ++ Flakes at PM ++
				L	3/60	Normal	
2	60	M	Progressive both eyes—12 yrs Occasional headache—2 yrs	R	HM PR	Normal	White flakes at PM +
				L	CF 2 ft	Normal	White flakes at PM +
3	65	M	Gradual right eye—4 yrs Occasional headache—2 yrs	R	PL+ PR	Deep	Iridodonesis ++
				L	6/9 part	Normal	Normal
4	65	M	Gradual, painless both eyes—8 yrs	R	CF 1 ft	Deep	Flakes at PM ++ Iridodonesis + Flakes at PM ++ Iridodonesis +
				L	CF 1 ft	Deep	
5	75	M	Progressive both eyes—2 yrs	R	CF 2 ft	Normal	Normal
				L	CF 4 ft	Normal	Normal
6	60	M	Progressive left—2 yrs right—6 mths	R	6/12 pt	Normal	Normal
				L	PL+ PR	Shallow	Normal
7	76	M	Progressive both eyes—2 yrs	R	PL+	Normal	Flakes at PM ++
				L	HM+ PR	Normal	Flakes at PM ++
8	68	M	Progressive both eyes—2 yrs	R	PL+ PR	Normal	Flakes at PM +
				L	6/18 pt	Normal	Flakes at PM +
9	62	M	Progressive left—2 yrs	R L	CF 6 in.	Deep	Aphak Flakes at PM ++ Iridodonesis +++

## ABBREVIATIONS

Counting Fingers—CF, Hand movements—HM, Immature senile cataract—IS

Pupil	Lens			Fundus	Intraocular pressure (mm. Hg Schiötz)	Other pathology
	Cataract	Exfoliation	Phakodonesis			
Normal, dilated, immobile	MS Lens dislocated in vitreous	+++	+++	Not visible	43	Vitreous fluid
Circular direct reaction present Pupillary reaction absent	IS	++	++	Not visible	37	Corneal oedema
Normal	MS	++	++	Not visible	24	
Normal	IS	++	++	Primary optic atrophy	24	
Normal	MS	+	++	Not visible	24	
Normal	Incipient	+	--	Normal	20	
Normal	IS (nuclear)	+	++	Not visible	17	
Normal	IS (nuclear)	+	++	Not visible	17	
Normal	IS	++	++	Not visible	22	
Normal	IS	++	++	Not visible	45	
Normal	Incipient	+	--	Marked choroidal sclerosis	15	
Normal	MS	++	+	Not visible	20	Corneal dystrophy
Dilated and fixed	MS	++	--	Not visible	54	Trachomatous entropion Corneal opacities
Normal	MS	++	--	Not visible	17	Trachomatous entropion Corneal opacities
Normal	MS	+	--	Not visible	20	
Normal	IS	+	--	Normal	16	
Normal	IS Dislocated in vitreous	+	+++	Not visible	17	

Mature senile cataract—MS, Perception of light—PL, Projection accurate—PR, Pupillary margin —PM

pathological processes around the blood vessels and defective healing. The frequent occurrence of glaucoma with exfoliation may be due to a similar degenerative change in the trabecular meshwork. Since this process was first recognized on the anterior surface of the lens before the days of electron microscopy, it was erroneously given the name of capsular exfoliation and later of pseudoexfoliation. Further biochemical studies are needed to find out the nature of the material present at all these sites.

All our patients were men over 60 years old. The preponderance of the condition in certain races, *i.e.* Norwegians (Hørven, 1966), and in males is suggestive of a sex-linked, genetic transmission.

We recommend the removal of the lens where capsular exfoliation is associated with phakodonesis even in cases of immature cataract; this will avoid the possibility of total lens dislocation.

### Summary

Nine cases of the capsular exfoliation syndrome involving seventeen eyes in elderly males are presented. The condition was associated with cataract, phakodonesis, irridodonesis, dislocated lens, corneal dystrophy, rubeosis iridis, primary optic atrophy, and choroidal sclerosis. This syndrome seems to be the result of a degenerative process affecting a biochemical substance present at various sites. This is believed to be a specific type of mucopolysaccharide which has undergone denaturation.

In cases of phakodonesis removal of the lens is recommended, even if the cataract is immature.

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