

## Clinical Study of Pseudoexfoliation of Lens

L.R. Murthy<sup>1</sup>, Sangeeta Das<sup>2</sup>

### Abstract

Pseudoexfoliation [PEX] syndrome is a common age - related disorder of the extracellular matrix that is frequently associated with severe chronic secondary open angle glaucoma and cataract. PEX syndrome may affect up to 30% of people older than 60 in a worldwide distribution and is biomicroscopically diagnosed by abnormal fibrillar deposits on ocular structures that line the aqueous bathed surfaces of the anterior segment. Often associated with the need for intraocular surgery, results of cataract and glaucoma filtering surgery in eyes with PEX in the early stages of disease may be comparable to those in eyes without PEX, in the later stages morbidity is significantly increased due to periocular surgical complications and the outcome is worse. Surgical and postoperative difficulties are often multifactorial and are directly related to the pathological changes of PEX on intraocular structures. Although the visible areas of the anterior capsule are most obviously involved, this is only a small part of the picture and of least significance. Biomicroscopically invisible changes of the zonules and their attachments are of the greatest consequence as they lead to zonular dialysis and subluxation of lens. In relation to the surgical management of these patients, the importance of early recognition of the pathological features, expectations of higher complication rate and having a plan to deal with them, taking adequate time for the surgery, close attention to postoperative follow-up and a well informed patient with realistic expectations cannot be overemphasized.

**Keywords:** Pseudoexfoliation Syndrome; Zonular Dialysis; Subluxation.

### Introduction

Since the past several years, number of ophthalmologists have been intrigued by the dandruff - like greyish white fibrillar material seen at the papillary margin and on the lens surfaces of many eyes, first described by Lindberg [1] in 1917. Malling [2] later noted areas of central and peripheral deposits separated by a clear intermediate zone on the anterior lenticular surface. Vogt [3] associated these findings with open angle glaucoma and proposed the name "glaucoma capsulare" or "exfoliation of the lens capsule".

Dvorak - Theobald [4] in 1954 proposed that the lens capsule was not involved pathologically and that the term pseudoexfoliation of the lens capsule was more descriptive to distinguish the condition from capsular delamination or true exfoliation and the term pseudoexfoliation syndrome is commonly used in modern literature. However, more recent ultrastructural studies have suggested that the exfoliative material on the lens capsule is derived, at least in part, from the lens and it has been proposed that this entity be called the "exfoliation syndrome" [5,6,7]. Both terms pseudoexfoliation syndrome and exfoliation syndrome are still commonly used in current literature, Exfoliation syndrome is characterized by the deposition of greyish white fibrillar material in a distinct pattern throughout the ocular anterior segment. The hallmark of exfoliation syndrome is the characteristic pattern of whitish pseudoexfoliation material on the anterior lens capsule which has three zones: a) a translucent, central disc with occasional curled edges b) a clear intermediate zone, corresponding to contact with the moving iris c) a peripheral granular zone, which may have radial striations [6].

**Author Affiliation:** <sup>1</sup>Professor & Head <sup>2</sup>Associate Professor, Dept. of Ophthalmology, Malla Reddy Institute of Medical Sciences, Suraram, Hyderabad, Telangana 500055, India.

**Corresponding Author:** Sangeeta Das, Associate Professor, Dept. of Ophthalmology, Malla Reddy Institute of Medical Sciences, Suraram, Hyderabad, Telangana 500055, India.

**E-mail:** [drsangeetadas@yahoo.co.in](mailto:drsangeetadas@yahoo.co.in)

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Exfoliation material also accumulates on the pupillary margin of the iris, iris surface, corneal endothelium, trabecular meshwork, lens zonules and ciliary body. Exfoliation material is also seen on the anterior hyaloid in aphakic eyes and on a posterior - chamber intraocular lens in pseudophakic eyes [8].

Patients who have pseudoexfoliation syndrome tend to have increased pigmentation of the anterior chamber angle. It has been associated with non-guttate endothelial loss and subsequent corneal decompensation.

Ultrastructural evaluations indicate that the exfoliation material can be found in the conjunctiva [9], extraocular muscles, orbital septa, posterior ciliary arteries, vortex veins, and central retinal vessels passing through the optic nerve sheaths. Ultrastructural studies performed on eyes during autopsy suggest that pseudoexfoliation syndrome is a systemic disorder. Exfoliation material has been found in a number of organs, which include skin, lung, gall bladder, liver, myocardium, kidney, bladder, and meninges.

The condition is more common in older age groups and is often found to be associated with raised intraocular pressure and cataract. Phacodonesis and subluxation of the lens is also seen in some patients, due to degenerative changes in the zonular fibers of these individuals [10,11]. The incidence of glaucoma in patients with pseudoexfoliation is as much as 10 to 15 fold higher than in the general population. Compared with eyes with primary open angle glaucoma, eyes with pseudoexfoliation generally have more severe optic disc and visual field damage on diagnosis.

The frequency of ocular hypertension in patients with pseudoexfoliation has been reported to be 5 to 10 times higher than in the general population. The amount of exfoliation material deposited in and around the Trabecular meshwork has been associated with increased intraocular pressure and the presence of glaucoma.

Patients with pseudoexfoliation syndrome have higher rates of intraoperative complications during and after cataract surgery compared to patients without the condition. Zonular attachments in pseudoexfoliation syndrome are weakened due to accumulation of pseudoexfoliative material resulting in higher incidence of lens subluxation, zonular dialysis and vitreous loss. Another factor which may complicate surgery is the reduced response to pharmacological dilatation in patients who have pseudoexfoliation syndrome [12].

Thus, considering the clinical importance of pseudoexfoliation, a study has been carried out in

this institution taking into account various aspects of this condition.

### Materials and Methods

In the present study 3200 patients of cataract visiting the outpatient department of ophthalmology at our Malla Reddy institute of medical sciences hospital between January 2015 to December 2015 were examined under a hospital based, observational, non - interventional, prospective, non-randomized, clinical study. 185 eyes of 115 patients diagnosed with pseudoexfoliation of lens were studied.

Patients of any age with cataract changes, patients with pseudoexfoliation lens and patients with systemic illness like diabetes, hypertension or ischaemic heart disease with due medical fitness were the inclusion criteria. Patients with evidence of ocular trauma, present or past uveitis and patients with intraocular surgery and laser treatment except for glaucoma were the exclusion criteria.

A detailed medical and ophthalmic history was obtained from each patient and a comprehensive vision and eye examination was carried out using tonometry, pupil reflex examination, Gonioscopy and slit-lamp examination, before and after dilatation of pupil for assessment of the type of cataract and for fundus examination.

Gonioscopy was done in all cases of pseudoexfoliation of lens using Goldman 3 mirror lens to determine the presence of pseudoexfoliation material and pigmentation in angle and type of the angle. 10% phenylephrine eye drops were used for mydriasis. Patients with angle closure were not dilated. Glaucoma capsulare was diagnosed on the basis of Foster's criteria for epidemiological studies. Optic disc and retinal examination was performed using 90D lens. The vertical CD ratio and the presence of focal notching was recorded. Eyes with visual acuity > 6/60, CDR  $\geq$  0.6 in the presence of asymmetry  $\geq$  0.3 or a neuroretinal rim width reduced to < 0.1 CDR (between 11 to 1 O' clock or 5 to 7 O' clock) underwent automated perimetry on Zeiss Humphrey system.

Keratometry, A scan and intraocular lens power estimation by SRK - II formula was done in all patients of pseudoexfoliation of lens undergoing cataract surgery. Intraoperative complications were determined during surgery. The surgeries were performed under peribulbar anesthesia under strict asepsis.

In cases of pseudoexfoliation of lens with cataract, cataract extraction was performed. Patients with raised IOP were controlled with medical therapy first, and then cataract extraction was performed. In patients with cataract and glaucoma, IOP was

controlled with medical treatment first, and then cataract extraction was performed. Cases not controlled with medical therapy alone, in these patients cataract extraction with trabeculectomy was done. PCIOL implantation was done in the cases possible.

Postoperative evaluations were done on 1<sup>st</sup> postoperative day, 1<sup>st</sup> week and at the end of 6<sup>th</sup> week for UCVA, BCVA, slit lamp biomicroscopy, intraocular pressure, direct and indirect ophthalmoscopy.

### Observations and Results

In the present study a total of 3200 patients of cataract visiting the ophthalmology outpatient department during a period of 1 year from January 2015 to December 2015 were examined. 185 eyes of 115 cases had either unilateral or bilateral pseudoexfoliation of lens capsule. The observations made are presented below

**Table 1:** Distribution of pseudoexfoliation of lens in cataract cases

Total number of patients seen	Pseudoexfoliation cases	Percentage (%)
3200	115	3.593

In the present study PEX of lens was observed in 3.593 % cases of cataract in outpatient department during a period of 1 year (Table 1).

**Table 2:** Age Distribution of PEX of Lens

Age group (years)	PEX cases total cases – 115 cases	Percentage (%)
41-50	5	4.347
51-60	25	21.739
61-70	54	46.95
71+	31	26.95

The maximum age distribution of PEX is seen between the age group of 61- 70 years while it is quite low below 50 years (Table 2).

**Table 3:** Sex Distribution of PEX of Lens

Total cases	Males	Females
115	72 (62.60%)	43 (37.39%)

PEX of lens was found predominantly in males 62.60% as compare to females 37.39% (Table 3).

**Table 4:** Distribution of laterality of PEX of lens

Unilateral		Bilateral
Rt. eye	Lt. eye	
24(20.86%)	21 (18.265)	70 (60.86%)

Total eyes = 24+21+ (70X2) = 185 eyes.

PEX was more commonly found to be bilateral than unilateral. Unilateral cases showed no specific predilection for either eye (Table 4).

**Table 5:** Distribution of PEX Material at Various Sites

Site	Number of Eyes Total Eyes – 185	Percentage (%)
Corneal endothelium	13	7.02%
Iris crypts	19	10.27%
Papillary margin	161	87.02%
Central disc on anterior lens capsule	86	46.48%
Peripheral band on anterior lens capsule	185	100%
Trabecular meshwork	58	31.355

The presence of PEX material as the peripheral band on the anterior lens capsule was the constant feature and was present in all the 185 eyes (100%), on the pupillary margin it was found in 161 eyes (87.02%) (Table 5).

**Table 6:** Distribution of PEX of lens in different types of cataract

Type of cataract	Number of eyes Total eyes – 185	Percentage (%)
Nuclear cataract(brown cataract Gr.III.N.S)	61	32.97
Cortical cataract	11	5.94
Cortical cataract + Gr.II.N.S	21	11.35
Cortical cataract + Gr.I.N.S	9	4.86
Intumescent cataract	8	4.32
Mature cataract	20	10.81
Hyper mature cataract	7	3.78
Posterior sub capsular cataract (PSC)	6	3.24
PSC + Gr.II.N.S	25	13.51
Cortical cataract + PSC	9	4.86
Cortical cataract + PSC+ Gr.II.N.S	8	4.32

PEX of lens was found in maximum number of cases of brown cataract (32.9%) (Table 6).

**Table 7:** Relation of PEX of lens with position of lens

Position of lens	Number of eyes Total eyes - 185	Percentage (%)
Normal	161	87.02
Subluxated	12	6.48
Phacodonesis	12	6.48

12 cases of subluxation and 12 cases of phacodonesis were found out of total of 185 eyes with PEX (Table 7).

**Table 8:** Relation of PEX of lens with intraocular pressure

Normotensive eyes	Hypertensive eyes ( IOP > 21 mm Hg)
116 (62.70%)	69 (37.29%)

37.29% of eyes with PEX of the lens were associated with increased intraocular pressure (Table 8).

**Table 9:** Results of Gonioscopy in PEX of lens

Angle grade (Shaffer's classification)	Number of eyes Total eyes – 185	Percentage (%)
Wide open angle ( Grade 3-4)	156	84.32
Narrow angle ( Grade 1-2)	22	11.89
Closed angle ( Grade 0)	7	3.78

156 cases of wide open angle, 22 cases of narrow angle and 7 cases closed angle were observed on Gonioscopy (Table 9).

**Table 10:** Relation of PEX of lens with glaucoma

Pseudoexfoliation	Glaucoma		Number of eyes total eyes -185		Percentage (%)	
	Open		M	F	M	F
Glaucoma 29 (15.67%)	Angle Glaucoma 23 (12.43%)		15	8	8.10%	4.32%
	Angle Closure Glaucoma 6 (3.24%)		3	3	1.62%	1.62%

29 eyes (15.67%) with PEX were diagnosed with Glaucoma. out of which 23 eyes (12.43%) had open angle Glaucoma and 6 eyes (3.24%) had angle closure Glaucoma (Table 10).

**Table 11:** Effect of mydriasis in eyes with PEX of lens

Effect of mydriasis	Number of eyes (Total eyes - 179, 6 eyes were not dilated)	Percentage (%)
Full mydriasis	76	42.45
Partial mydriasis	103	57.45
Rise in intraocular pressure	120	67.03

Rise in IOP was frequently recorded after mydriasis of pupil in 67.03% of eyes. 57.45% of eyes with PEX showed heavy deposition of PEX material and thus partial mydriasis (Table 11).

**Table 12:** Mode of treatment used in PEX of lens

Treatment	Number of eyes Total eyes -185	Percentage (%)
SICS + PCIOL	86	46.48
Phacoemulsification + PCIOL	56	30.27
ECCE + PCIOL	9	4.86
ECCE + PCIOL + TRAB	15	8.108
ECCE + ANT. Vitrectomy	9	4.86
SICS + ANT. Vitrectomy	6	3.24
ICCE + ANT. Vitrectomy	1	0.54
No Treatment	1	0.54
Only Medical Line of Treatment	1	0.54
Referred to Posterior Segment Surgeon (Nucleus Drop)	1	0.54

151 cases underwent cataract extraction with PCIOL implantation, 15 cases cataract extraction with PCIOL implantation with trabeculectomy (Table 12)..

**Table 13:** Intraoperative complications eyes with PEX of lens

Complication	Number of eyes Total eyes - 185	Percentage (%)
Zonular dialysis	12	6.48
Posterior capsular rupture	24	12.97
Vitreous loss	19	10.27
Pigment dispersion	8	4.32
Nucleus drop	1	0.54
Posterior synechiae	7	3.78
Iris prolapsed	4	2.16
Difficulty in performing CCC	6	3.24

Zonular dialysis, posterior capsular rupture and vitreous loss frequently occurred during surgery in cases of PEX of lens (Table 13).

**Table 14:** Intraoperative complications according to type of surgery

Surgery and no. of surgeries	Posterior capsular rupture	Vitreous loss	Zonular dialysis
ECCE (33)	10(30.30%)	8(24.24%)	2(6.06%)
SICS (92)	11(11.95%)	8(8.69%)	8(8.69%)
Phacoemulsification (56)	3(5.35%)	3(5.35%)	2(3.57%)

Complications like posterior capsular rupture and vitreous loss occurred more in eyes with ECCE and less in eyes with SICS and phacoemulsification (Table 14).

**Table 15:** Postoperative complications

Complication	Number of eyes	Percentage (%)
Corneal edema	12	6.48
Shallow AC	7	3.78
Uveitis	5	2.70
Hyphaema	3	1.62
IOL decentration	4	2.16
Endothelial decompensation	3	1.62

12 cases had corneal edema and 3 cases went in to endothelial decompensation which were associated with corneal dystrophy (Table 15).

**Table 16:** postoperative rehabilitation

Visual acuity	Number of eyes	Percentage (%)
No improvement in visual acuity	14	7.56
Upto finger counting 6 meters	9	4.86
6/60 to 6/18	52	28.10
6/12 to 6/6	110	59.45

Six weeks after cataract extraction, most patients showed good visual outcome with 59.45% cases having visual acuity 6/12 or better and 28.10% cases showed visual acuity 6/60 or better as seen above Table 16.

Intra - ocular pressure was well controlled after cataract extraction. This shows that intraocular pressure can be well controlled by cataract extraction.

Poor visual outcome in some cases was due to intraoperative vitreous loss and postoperative complications like corneal, endothelial decompensation and associated glaucoma.

## Discussion

Comparing the present study with previous studies, we found that most of the results are in accordance with those in literature.

Although pseudoexfoliation syndrome has worldwide distribution, reported prevalence rates vary extensively. There is a well recognized tendency for the syndrome to cluster both geographically and within racial and ethnic subgroups

Country	Author	Year	Percentage (%)
India	Siva reddy <sup>13</sup>	1970	34.6
	Sood <sup>14</sup>	1968	1.9
	Lamba <sup>15</sup>	1984	7.4
	Arvind H <sup>16</sup>	2003	3.8
Finland	Lind berg <sup>1</sup>	1917	20
Greece	Trantas <sup>17</sup>	1934	13.3
Norway	Horven <sup>18</sup>	1937	17
	Bertelsen <sup>19</sup>	1964	1
	Ring vold <sup>20</sup>	1988	16.9
USA	Irvine <sup>21</sup>	1940	3
	Hiller <sup>22</sup>	1982	2.6
Germany	Aasved <sup>23</sup>	1969	4
South Africa	Bartholomew <sup>24</sup>	1973	
		Over 40	8.1
		5059	7.2
		Over 60	10.3
Present study			3.593

The distribution varies in different countries. The distribution of PEX of lens in the present study is comparable with the study done by Aravind et al. 2003.

The study of literature shows the highest prevalence to occur between 60 to 80 years, average being around 70 years. Tarkkanen (1962) [25], Siva Reddy et al. (1970) [13] and Sood et al (1968) [14] reported the maximum prevalence between 60 to 70 years.

Though Duke - Elder (1969) [26] stated that there was no appreciable sex preference for the occurrence of PEX, studies in India by Sood et al (1968) [14], Siva Reddy et al. (1970) [13] and P.A. Lamba (1984) [15] revealed PEX to predominately occur in males.

The distribution of PEX of lens was more in males in the present study was similar to the study done by others. Most workers have found bilateral cases to be more common than unilateral.

In this study flakes at the pupillary margin were found in 87.02% cases, indicating that it is a common site where exfoliative material gets deposited which is in close comparison with the studies done by other authors. The peripheral band of PEX material is a constant feature and is seen in 100% of cases, which is in accordance with the previous study by Konstas 1993 [27]. This confirms the fact that peripheral band is the most constant feature of PEX of lens and unless its presence is established PEX of lens should not be diagnosed.

PEX is commonly associated with brown cataract which is in accordance with the previous study by Sood et al. [14], Mardin C.Y [28], Rudkin et al. [29] and Rahman et al. [30].

Several studies in literature show that in patients of PEX of lens, there is a tendency for phacodonesis and spontaneous dislocation of the lens and operative dislocation of lens when grasped with forceps. This tendency of the lens for displacement occurs due to changes in zonules, which are extremely weak due to deposition of PEX material. The zonules stretch and tend to break at their attachment to ciliary body.

PEX of lens showed a raised IOP which is comparable to the studies done by other workers.

In the present study, 84.32% cases of wide open angle, 11.89% cases of narrow angle and 3.78% cases of closed angle were found. The percentage of open angle was in close range with the previous studies while that of narrow angle and closed was less as compared to previous studies. Increased Trabecular pigmentation was noted in 97.72% cases, similar to the studies done by other workers.

In the present study 15.67% cases of PEX of lens was diagnosed with glaucoma. Out of 29 cases of PEX glaucoma, 23 cases of open angle glaucoma (12.43%) and 6 cases of angle closure glaucoma (3.24%) were found in the present study. the finding of open angle Glaucoma was comparable to the study done by Roth and Epstein (1980) [31] while the finding in angle closure glaucoma was much lower than this study by Gillies et al (1988) [32].

It can be said that it is difficult to control the glaucoma associated with PEX of lens with medical therapy alone and most of the patients require filtering surgeries.

In accordance with previous studies, this study shows that trabeculectomy is an effective means for control of intraocular pressure in glaucoma capsulare and it should be combined with extracapsular cataract extraction in cases where glaucoma capsulare is associated with cataract. Also, the present study shows that cataract

extraction causes reduction in IOP after cataract surgery in eyes with PEX, similar to the study done by other workers.

PEX patients have increased rates of intraoperative capsular complications, such as posterior capsular rupture, zonular dialysis, and vitreous loss, during cataract surgery.



Fig.1: PEX over ant. Lens capsule.

Fig.2: PEX over pupillary Margin.



Fig: 3. Peripheral band of PEX over anterior lens capsule.

Fig: 4. central disc of PEX Over anterior lens capsule.

### Conclusions

In the present study a total of 3200 patients of cataract visiting the ophthalmology outpatient department during the period of 1 year from January 2015 to December 2015 were examined.

1. PEX was present in 185 eyes of 115 cases. PEX of lens was found in 3.593% of patients of cataract during the period of 1 year.
2. The maximum age distribution of PEX of lens was between 61-70 years and was quite less below 50years.
3. PEX of lens was predominantly seen in males 62.60%.
4. PEX of lens was commonly found to be bilateral 60.86%
5. Presence of peripheral band of PEX material on anterior lens capsule was a constant feature and was noted in all cases.
6. PEX of lens was commonly found in brown cataract 32.97.
7. Due to zonular weakness and fragility there is a tendency of spontaneous displacement of the lens from its position. 6.48% cases had subluxation and another 6.48%cases had phacodonesis.
8. 37.29% of eyes with PEX of the lens were associated with increased intraocular pressure.
9. 156 cases of wide open angle, 22 cases of narrow angle and 7 cases closed angle were observed on Gonioscopy.
10. 29 eyes (15.67%) with PEX were diagnosed with Glaucoma, out of which 23 eyes (12.43%) had open angle Glaucoma and 6 eyes (3.24%) had angle closure Glaucoma.
11. Rise in IOP was frequently recorded after mydriasis of pupil in 67.03% of eyes. 57.45% of eyes with PEX showed heavy deposition of PEX material and thus partial mydriasis .
12. Medical line of therapy could not control IOP in patients with glaucoma capsulare
13. The IOP was well controlled in patients after combined surgery (Trabeculectomy combined with cataract extraction) and was more effective than medical therapy.
14. A reduction in IOP was observed after cataract extraction.
15. PEX of lens was found to be associated with increased intraoperative complications like posterior capsular rupture, zonular dialysis and vitreous loss.
16. Prognosis depends on the stage of glaucoma, association with subluxation, intraoperative and postoperative complications. More than half (59.45%) cases showed a very good visual prognosis while in 7.56% cases there was no improvement in visual acuity.

Thus in conclusion, PEX of lens is a condition of old age, commonly associated with brown cataract, raised intraocular pressure, open angle glaucoma and increased rates of intraoperative posterior capsular complications.

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